MONASH University

ANNUAL REPORT 2024

AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY

Data Extract Period

The data contained in this report was extracted from the ACFDR on June 12th 2025, and pertains to data related to patient events from January 1st to December 31st 2024. 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

ACFDR	Australian Cystic Fibrosis Data Registry
BAL	Broncho Alveolar Lavage
BMI	Body Mass Index
CF	Cystic Fibrosis
CFA	Cystic Fibrosis Australia
CFRD	Cystic Fibrosis Related Diabetes
CFTR	Cystic Fibrosis Transmembrane Conductance Regulator
DA	Dornase Alpha
ETI	Elexacaftor/tezacaftor/ivacaftor
FEV	Forced Expiratory Volume
FEV1 pp	Percent predicted Forced Expiratory Volume (litres) in 1 second
GLI	Global Lung Initiative
IV	Intravenous
MRSA	Methicillin–resistant *Staphylococcus aureus*
NTM	Nontuberculous Mycobacteria
PBS	Pharmaceutical Benefits Scheme
pwCF	People with Cystic Fibrosis

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CYSTIC FIBROSIS Australia

MONASH University

CONTENTS

FOREWORDS

FROM THE CYSTIC FIBROSIS AUSTRALIA CEO

The Registry continues to provide a vital window into the lives of Australians living with cystic fibrosis, guiding research, clinical care, and advocacy across the country.

The 2024 data show that 3,788 Australians are living with cystic fibrosis, and the majority, 60%, are over the age of 18. Cystic fibrosis is no longer a childhood disease. This reflects the extraordinary progress in treatment and care that has transformed cystic fibrosis from a condition that was once primarily experienced in childhood, to one in which people can live longer, healthier and hopefully more fulfilled lives.

We have seen an increase in the median age and the percentage of people with CF and an increase in the number of people with CF who are aged 40+ years. There has been a dramatic increase in the predicted survival age from 51 to 64 years. This is a giant leap in progress and care. Many of these outcomes are attributed to the innovative modulator therapies that have recently been made available in Australia. It is a testament to the commitment of clinicians, scientists, families, and, importantly, the people living with cystic fibrosis themselves, who demonstrate resilience, courage, and determination every day.

While overall these statistics mark significant progress, they also remind us of the work that remains. We cannot overlook that the data shows that still 7% of people with CF are not eligible for CFTR modulators and the prevalence of cystic fibrosis related diabetes. We still have a lot of work to ensure that all people with CF have the support and outcomes they deserve.

At Cystic Fibrosis Australia, our role as the national peak patient body is to ensure that every piece of data collected through the registry translates into meaningful outcomes for the community. Each insight guides advocacy for equitable access to care, informs research priorities, and shapes programs to support people living with CF and their families. We are committed to improving the quality, completeness, and timeliness of data, because stronger evidence leads directly to better treatment decisions, better policies, and ultimately, better lives.

This report is not just a reflection of numbers, it is a reflection of human lives, hopes, and futures. It tells a story of progress, of challenges met, and of a community whose courage drives change. My sincere thanks to everyone who has contributed to the registry. As we look to the future, Cystic Fibrosis Australia will continue to work tirelessly alongside the community, healthcare providers, and researchers to ensure that every person with CF has the opportunity to live longer, healthier, and more fulfilling lives.

Dr Jo Armstrong
Chief Executive Officer,
Cystic Fibrosis Australia

CYSTIC FIBROSIS Australia

FROM THE MONASH UNIVERSITY ACFDR LEAD

On behalf of the team at the School of Public Health and Preventive Medicine that manages the Australian Cystic Fibrosis Data Registry, I am pleased to present the 2024 Annual Report (AR). Comprising information regarding the characteristics and outcomes of people with CF from twenty-three CF centres across Australia, this report relates to 3,788 people with CF in Australia, including an additional 48 people newly diagnosed with CF.

The ACFDR Annual Reports have become increasingly comprehensive, and the 2024 AR is no exception. The registry Summary Data table (pg 4-5) highlights many of these new findings. Sixty percent of the cohort are now adult. Lung function and BMI continue to increase, as does the proportion of pwCF who are prescribed a modulator – for 2024 this was over 86% of adults and 77% of children (83% of the total cohort). Similarly, the proportion of pwCF having no hospitalisations for the year increased to 72%, highlighting the continuing reduction in acute care needs for pwCF. The AR also highlights ongoing areas of management for people with CF, including persisting respiratory infections and diabetes. Given the increasing age of the population, preventive care and management of complications of older age will become increasingly important.

I would like to thank the wonderful leadership of the ACFDR Steering Committee, and the commitment of the ACFDR Chair and Deputy Chair, Jo Armstrong, clinicians, researchers and all people with CF who have supported the registry. We are very proud of the incredible resource that is the ACFDR, and we hope you enjoy reading the 2024 ACFDR Annual Report.

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

FROM THE REGISTRY CLINICAL LEAD

The 2024 ACFDR report contains important information for people living with cystic fibrosis and the health professionals involved in their care.

The number of people in Australia with CF continues to increase with 3,788 recorded. At the same time we see an increase in the overall median age, demonstrating improving life expectancy, while there are now more than 60% of people living with cystic fibrosis who are aged 18 years and older.

A trend that appears to be emerging in recent years though is that the number of new diagnoses of CF appears to be falling. There are fewer diagnoses being recorded in children, in contrast there's been an increase in adults diagnosed for the first time with CF, having risen from 10% in 2020 to 17% in 2024.

Pleasingly lung health in all ages continues to improve. We can see a steady rise in lung function in both adults and children. While fewer people have serious chronic lung infections with bacteria such as Pseudomonas and Staphylococcus aureus and this has declined steadily since 2020.

CFTR modulators are prescribed now for 77% of all children and 86% of all adults on a regular basis.

The baby boom for people with CF also continues with no sign of slowing down with 85 pregnancies recorded this year.

Only three people required a bilateral lung transplant in 2024. Sadly, we did lose 11 people with CF in this year but that's the lowest number that has ever been recorded in the data registry.

The data shows important changes. Across the board we see improving outcomes for all people with CF, particularly in terms of lung health care while high-quality care continues to be delivered. We are very fortunate to have such high-quality data reflecting the important changes that we can see that have emerged and continue to evolve in the care of people with CF in Australia.

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.

SUMMARY OF REGISTRY DATA

	2020	2021	2022	2023	2024
PEOPLE WITH CYSTIC FIBROSIS					
Active population in the ACFDR*	3,538	3,616	3,738	3,798	3,788
Age (median)	20.2 years	20.6 years	21.1 years	21.7 years	22.4 years
Age (mean)	22.6 years	23.0 years	23.4 years	24.0 years	24.6 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%)
Adults: Males %	52.8%	52.8%	56.8%	52.5%	52.3%
Total registered patients**	5,050	5,176	5,300	5,409	5,483
CF DIAGNOSIS & GENOTYPING					
Newly diagnosed pwCF	74	92	90	68	48
% New diagnosis <1 year	82.4%	82.6%	78.9%	77.9%	83.3%
% New diagnosis ≥18 years	10.8%	12.0%	11.1%	13.2%	16.7%
Genotyped – one known allele (two alleles)	98.4% (92.2%)	98.4% (94.8%)	99.1% (95.9%)	99.2% (96.3%)	99.8% (99.7%)
% F508del Homozygous	47.0%	47.0%	46.0%	46.0%	46.0%
% F508del Heterozygous	43.0%	43.0%	44.0%	44.0%	44.0%
CLINICAL MEASURES (LUNG FUNCTION & NUTRITION)					
Median FEV1 pp children and adolescents 6–17 years	91.0	93.0	93.2	96.6	97.7
Median FEV1 pp adults 18 years and older	70.0	73.0	75.6	78.6	79.0
Median weight for length percentile <2 years	51st	51st	50th	54th	51st
Median BMI percentile children 2–17 years	57th	58th	57th	60th	61st
Median BMI adults 18 years and older	22.9	23.0	23.4	23.9	23.8
Number of lung function tests (mean)					
Paediatric	3.4	3.7	3.1	2.9	2.7
Adult	3.6	3.6	2.9	2.8	2.7
RESPIRATORY MICROBIOLOGY					
P. aeruginosa (%)	41.6%	38.9%	32.7%	22.7%	21.5%
S. aureus (%)	47.1%	47.3%	43.6%	38.1%	38.5%
Aspergillus spp (%)	18.8%	17.5%	13.2%	8.3%	7.7%
Non-tuberculous mycobacterium (%)	6.4%	8.1%	4.2%	2.8%	3.0%
Number of Microbiology samples (mean)					
Paediatric	3.63	3.73	3.70	3.57	3.61
Adult	2.09	2.38	2.24	1.42	1.50
COMPLICATIONS					
% with CF related Diabetes <12 years	1.9%	2.2%	2.2%	2.0%	2.0%
% with CF related Diabetes 12-17 years	17.3%	14.2%	13.6%	11.8%	11.7%
% with CF related Diabetes 18-29 years	21.5%	23.8%	26.0%	24.5%	25.1%
% with CF related Diabetes 30+ years	30.0%	29.9%	29.6%	26.9%	30.3%

	2020	2021	2022	2023	2024
MULTIDISCIPLINARY CARE					
% with Physiotherapy annual review	N/A	85.7%	83.9%	85.6%	88.0%
% with Dietician annual review	N/A	74.7%	76.6%	77.5%	76.8%
% with Mental Health annual review ≥12 years	N/A	80.6%	26.8%	23.6%	24.9%
% with Social review	N/A	N/A	43.2%	46.2%	48.6%
% with Gastroenterologist annual review	N/A	N/A	24.0%	23.8%	21.4%
% with Endocrinologist annual review	N/A	N/A	19.5%	19.6%	22.0%
CFTR MODULATORS					
% taking CFTR modulator – total cohort	52.6%	55.2%	68.7%	77.9%	82.8%
% taking CFTR modulator – paediatric	N/A	N/A	55.6%	68.5%	77.0%
% taking CFTR modulator – adult	N/A	N/A	78.7%	84.9%	86.3%
LUNG TRANSPLANTS AND SURVIVAL					
Total pwCF living with a transplant	128	122	116	116	106
Bilateral lung transplants	15	9	6	9	3
Deaths (Total CF deaths)	18	19	10	18	11
Deaths (post-transplant)	6	11	5	8	8
Median age of death (transplants included)	30.7 years	36.8 years	44.2 years	40.6 years	36.6 years
Median age of death (transplants excluded)	21.1 years	29.9 years	34.1 years	48.3 years	25.5 years
Survival median (cohort, 5 years)	53.0 years (2015-2019)	56.9 years (2016-2020)	58.2 years (2017-2021)	60.6 years (2018-2022)	64.0 years (2019-2023)
CF MANAGEMENT AND HEALTHCARE UTILISATION					
Clinical visits per person (mean)					
Paediatric	4.7	4.9	4.4	4.3	3.9
Adult	5.1	5.0	4.9	4.3	4.0
Hospitalisations (mean)					
Paediatric	0.7	0.7	0.7	0.6	0.5
Adult	0.7	0.7	0.6	0.4	0.5
% of pwCF with 0 hospitalisations	62.5%	61.2%	64.2%	70.4%	71.9%
PREGNANCY					
Total pregnancies	41	46	70	88	85

* Defined as number of pwCF who met the diagnostic inclusion criteria and had acceptable data quality
** Defined as total cumulative number of pwCF registered regardless of 2024 registry status
Note: pwCF with transplants may not be captured in the ACFDR post-transplant.

1.

AUSTRALIANS WITH CYSTIC FIBROSIS (CF)

1. AUSTRALIANS WITH CYSTIC FIBROSIS (CF)

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 31st December 2024.

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 2024 Report is structured in four 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

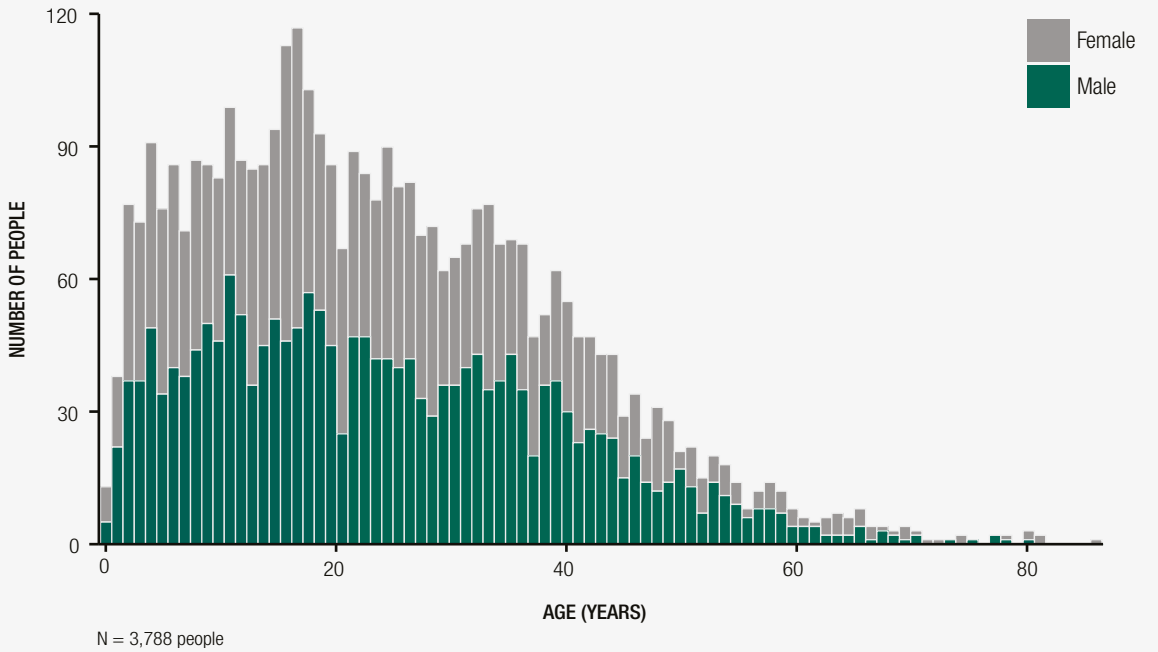
As per the 2023 Report, this report follows international registry reporting practice to **only include data from lung transplant recipients in the following sections - Demographics, Diagnostic and Genotype Information.** This is so that information regarding clinical care and outcomes reflects the majority 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 31st December 2024, the ACFDR contains data from **106 pwCF** (105 adults and 1 child/adolescent with CF) who have undergone a lung transplant. In addition, 19 pwCF were excluded from the 2024 Annual Report due to insufficient information recorded in the registry.

1.2 DEMOGRAPHICS

Age and Sex of People with CF in the Registry

As of 31st December 2024, the ACFDR held records of **3,788 active pwCF**, collected from 23 CF centres in Australia. Of the 3,788 pwCF, **2,274 were adults** (18+ years) and **1,514 were children and adolescents** (0-17 years) (Figure 1.1).

FIGURE 1.1: ACFDR 2024: PEOPLE WITH CF IN AUSTRALIA BY AGE AND SEX



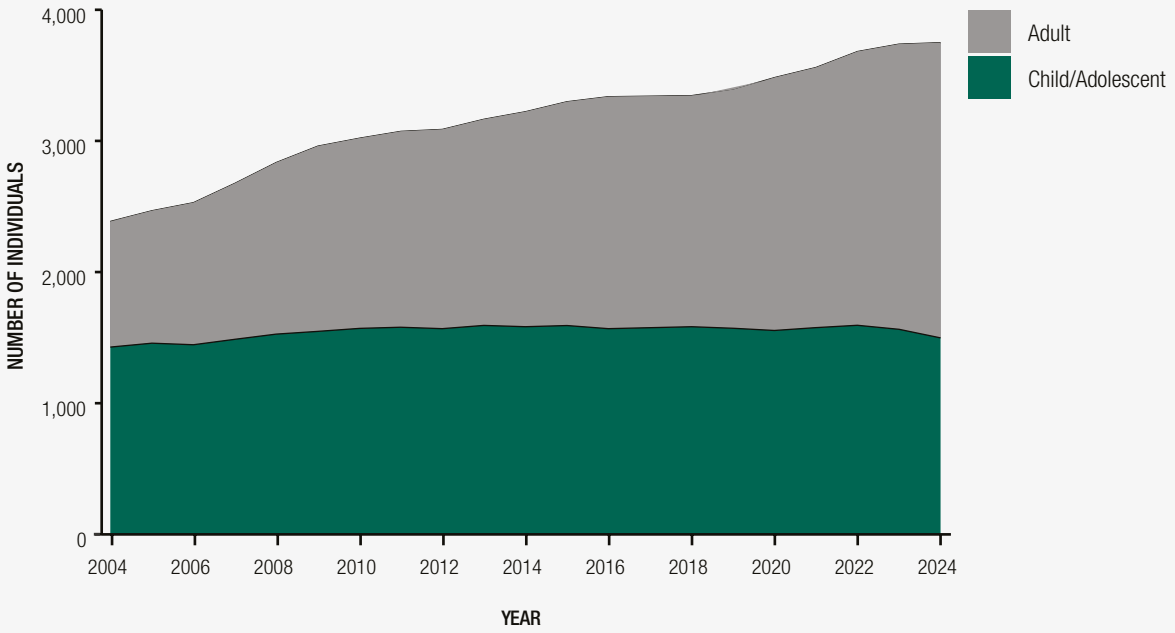
As of 31st December 2024, the proportion of males in the ACFDR was 52.3% and females were 47.7%. The largest age grouping for pwCF was those aged 18-29 years which comprises 25.7% of pwCF, followed by pwCF aged ≥40 years (17.2%) and pwCF aged 30-39 (17.1%) (Figure 1.1 and Table 1.1).

TABLE 1.1: ACFDR 2024: PEOPLE WITH CF BY AGE AND SEX

Age	Female	Male	Total
<2	55.7% (49)	44.3% (39)	88
2-5	49.7% (162)	50.3% (164)	326
6-11	45.0% (233)	55.0% (285)	518
12-17	51.0% (297)	49.0% (285)	582
18-29	49.5% (482)	50.5% (491)	973
30-39	44.8% (290)	55.2% (358)	648
≥40	44.9% (293)	55.1% (360)	653
Total	47.7% (1,806)	52.3% (1,982)	3,788

In 2024, the registry held records of 2,274 adults and 1,514 children with CF (Figure 1.2). The proportion of the registry population who were adult was 60.0%, compared with 58.2% in 2023. In 2024, the median age of the CF population was 22.4 years, with a mean age of 24.6 years. The median age for males at 22.9 years (22.2 years in 2023) remained higher than that for females at 21.8 years in 2024 (21.0 years in 2023).

FIGURE 1.2: ACFDR 2004-2024: PAEDIATRIC VS ADULTS PROPORTION

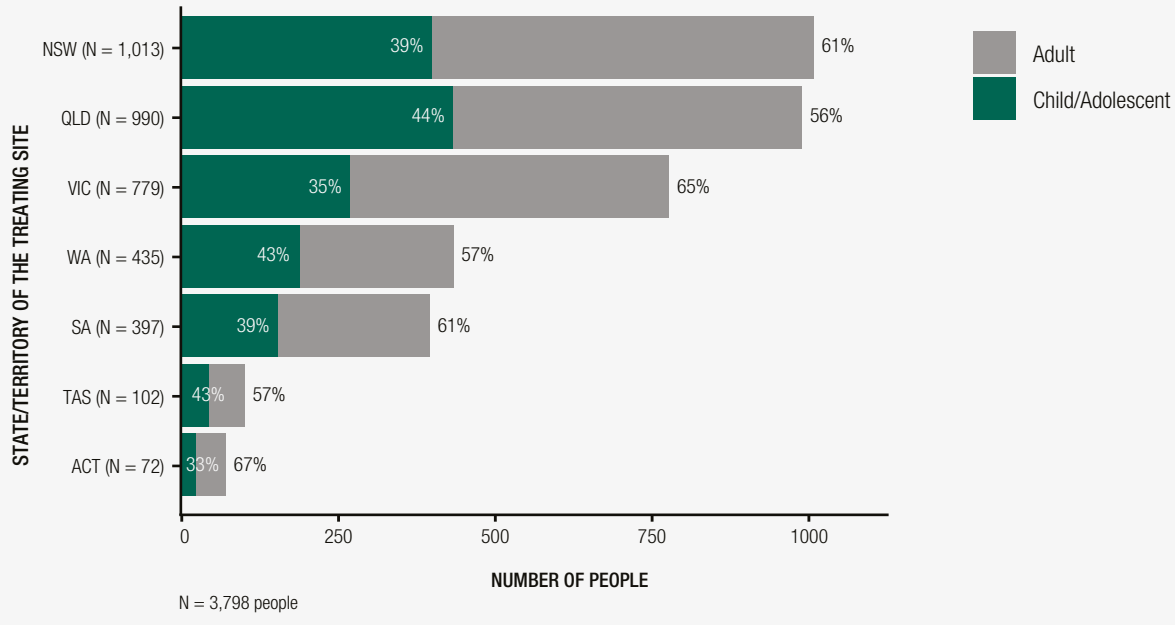


Note: Population size in 2017 was estimated based on the populations in years 2016 and 2018

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 2024, as well as the proportion of these who were children/adolescents and adults. New South Wales provided care to the most pwCF, followed by Queensland, Victoria, Western Australia, South Australia, Tasmania and the Australian Capital Territory.

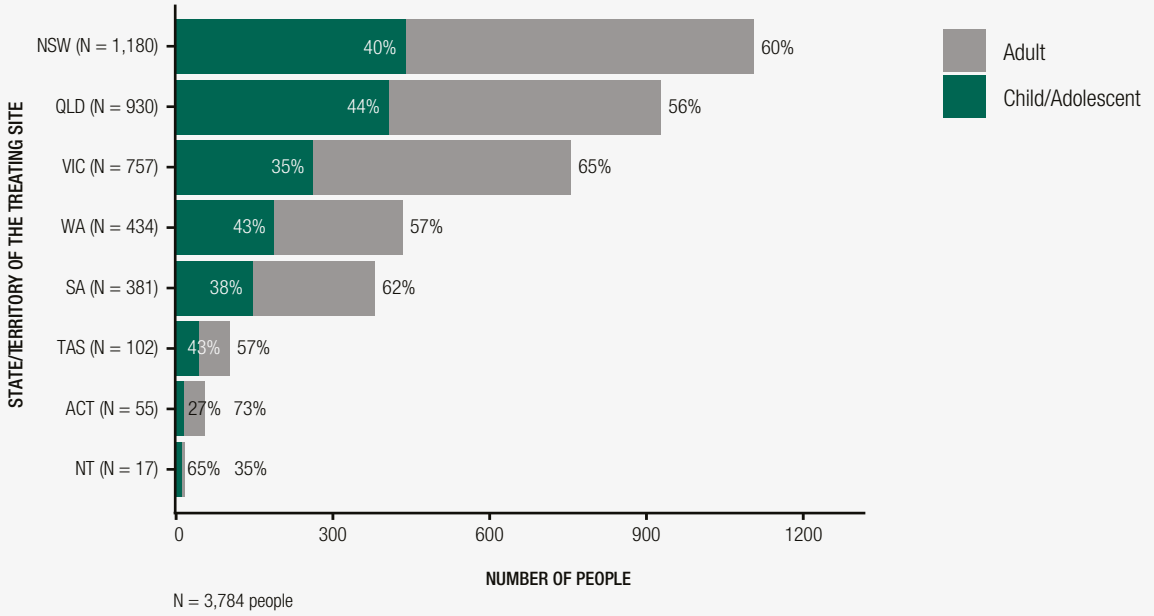
FIGURE 1.3: ACFDR 2024: DISTRIBUTION BY STATE AND TERRITORY WHERE CARE WAS PROVIDED



Note: PwCF who were shared or transferred between CF Centres in 2024 may be represented more than once.

Figure 1.4 shows the jurisdictions where pwCF lived in 2024 (or the most recent prior year), derived from postcode information collected by the registry. New South Wales had the highest number and proportion (1,108/29% of residing pwCF), followed by Queensland (930/25%), Victoria (757/20%), Western Australia (434/11%), South Australia (381/10%), Tasmania (102/3%), the Australian Capital Territory (55/1%) and the Northern Territory (17/<1%). Less than 5 pwCF did not have their postcode reported. In 2024, 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 2024: DISTRIBUTION BY PATIENT RESIDENCE



In 2024, the number of Indigenous Australians with CF was 148.

1.3 DIAGNOSTIC AND GENOTYPE INFORMATION

Diagnostic Information

New Diagnoses

The number of new diagnoses of pwCF notified to the registry in 2024 was 48, including 40 people diagnosed at less than one year of age, and 8 people diagnosed over 18 years. Seventy-one percent of all new diagnoses, and 89% of diagnoses within 8 weeks, were made by Newborn Screening (Table 1.2).

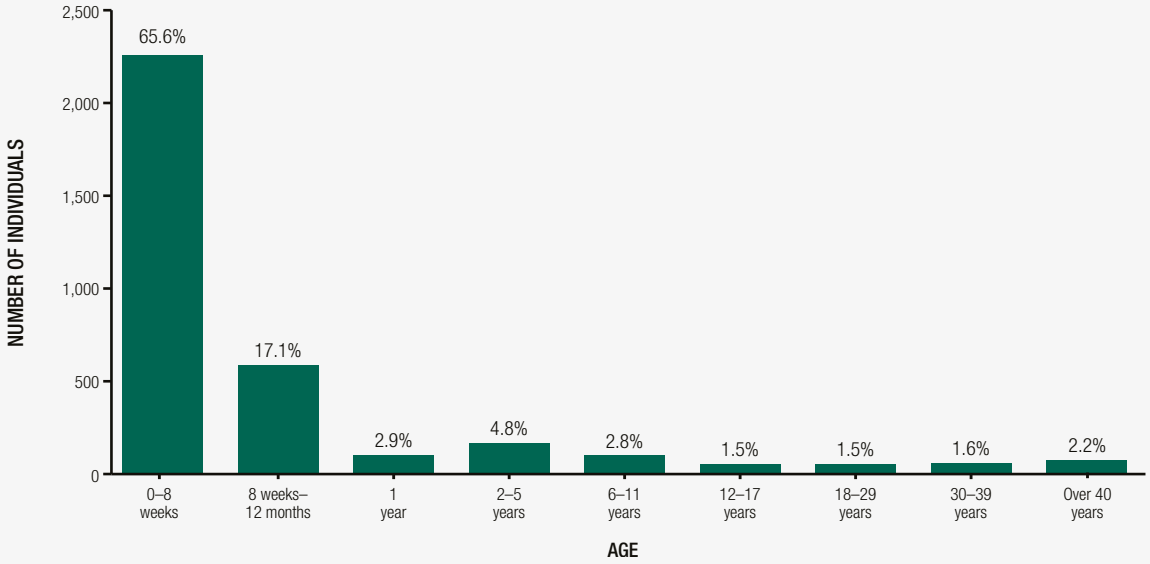
TABLE 1.2: ACFDR 2024: AGE OF NEW DIAGNOSES

Age	N (%)	Diagnosis via newborn screening test	Diagnosis by non-newborn screening test indication
Birth-8 weeks	37 (77.1%)	33 (89.2%)	<5
8 weeks-12 months	<5	<5	<5
1-17 years	0 (0.0%)	0 (NA)	0 (NA)
18+ years	8 (16.7%)	0 (0.0%)	8 (100.0%)
Total	48 (100.0%)	34 (70.8%)	14 (29.2%)

Age of Diagnosis for Registry Population

The age of CF diagnosis has been captured in the registry for 90.6% of the total registry cohort (Figure 1.5). Of these, 77.1% were diagnosed between birth and 8 weeks, 6.2% were diagnosed between 8 weeks and 12 months, and a further 16.7% of pwCF were diagnosed as adults. Compared to the 2024 diagnoses, the total cohort has fewer diagnoses within the first 8 weeks (65.6% vs 77.1%).

FIGURE 1.5: ACFDR 2024: AGE AT CF DIAGNOSIS (WHOLE COHORT)



Factors Associated with the Diagnosis of CF

For the total cohort, a diagnosis of CF was confirmed or suggested by newborn screening (59.0%), clinical signs/symptoms (35.1%), family history (8.7%), and prenatal screening (2.7%) (Figure 1.6 and Table 1.3). Of those with clinical symptoms, the most common presentations were meconium ileus/intestinal obstruction (33%), respiratory signs and symptoms (27%), and gastrointestinal symptoms (14%). With multiple responses available, often there is an overlap in clinical symptoms and newborn screening or family history in the registry. In 2024, diagnoses were more likely to be from newborn screening (70.8% vs 59.0%), family history and prenatal screening.

FIGURE 1.6: ACFDR 2024: FACTORS ASSOCIATED WITH DIAGNOSIS OF CF (WHOLE COHORT)

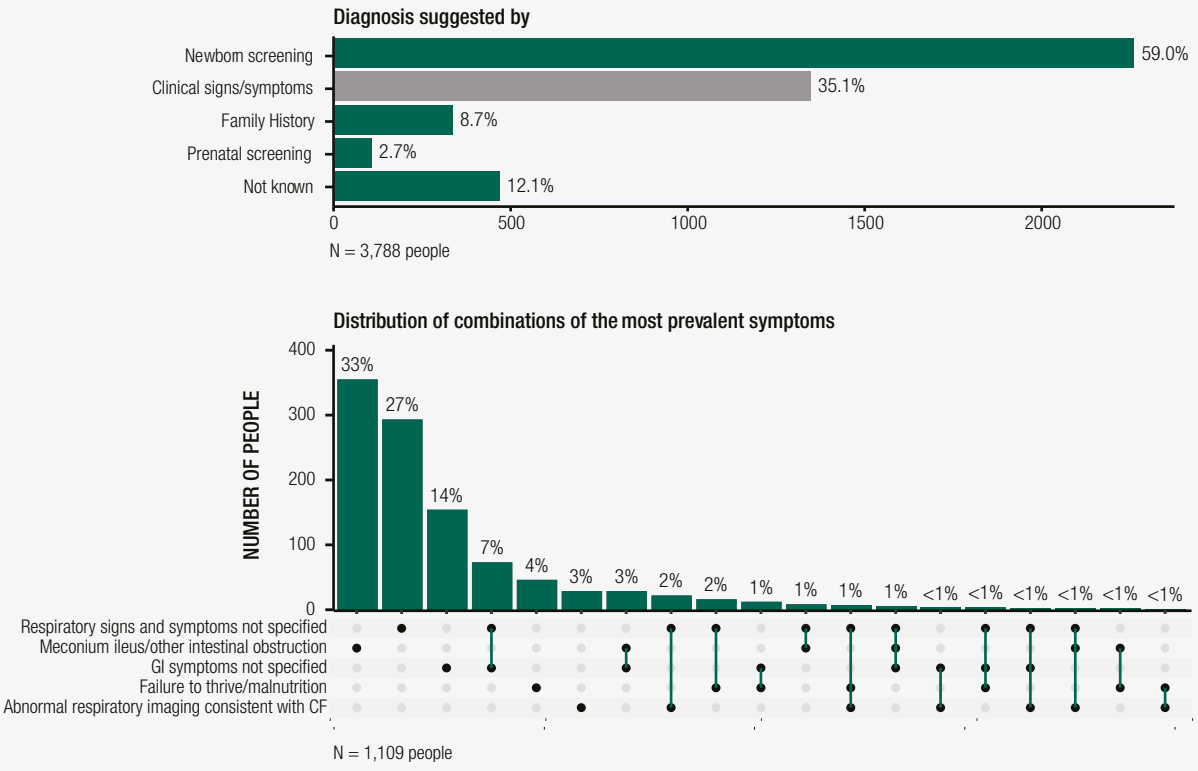


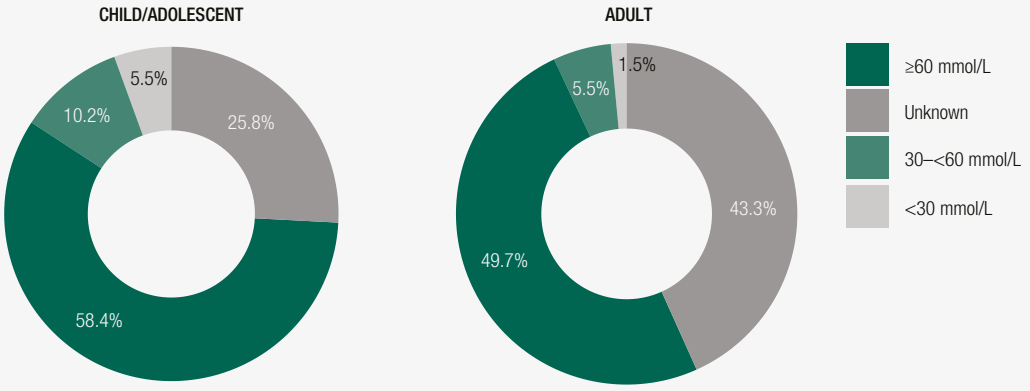
TABLE 1.3: ACFDR 1998-2024: FACTORS ASSOCIATED WITH DIAGNOSIS OF CF (2024 VS WHOLE COHORT)

Diagnosis by	Total cohort N = 3,788 (%)	2024 New diagnosis N = 48 (%)
Newborn screening	2,235 (59.0%)	34 (70.8%)
Clinical signs/symptoms	1,330 (35.1%)	13 (27.1%)
Family history	328 (8.7%)	7 (14.6%)
Prenatal screening	102 (2.7%)	<5
Not known	460 (12.1%)	<5
Sweat chloride value available	2,413 (63.7%)	37 (77.1%)

Alongside indications for diagnosis, Table 1.3 shows the proportion of pwCF with a sweat chloride value at diagnosis. For pwCF diagnosed in 2024, the documentation of sweat chloride in the registry was 77.1% compared to 63.7% 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 1.7 shows the proportion of child/adolescent and adult pwCF with sweat chloride values of <30 mmol/L, 30-<60 mmol/L, ≥60 mmol/L, and unknown. Fewer children and adolescents had unknown sweat chloride than adults (25.8% compared to 43.3%). Values for sweat chloride greater than 60 mmol/L were more common amongst children and adolescents (58.4%) than adults (49.7%), as were values of 30-<60 mmol/L (10.2% and 5.5% respectively) and <30 mmol/L (5.5% and 1.5% respectively). These differences indicate greater documentation of sweat chloride values amongst the paediatric CF population.

FIGURE 1.7: ACFDR 2024: SWEAT CHLORIDE VALUES BY AGE

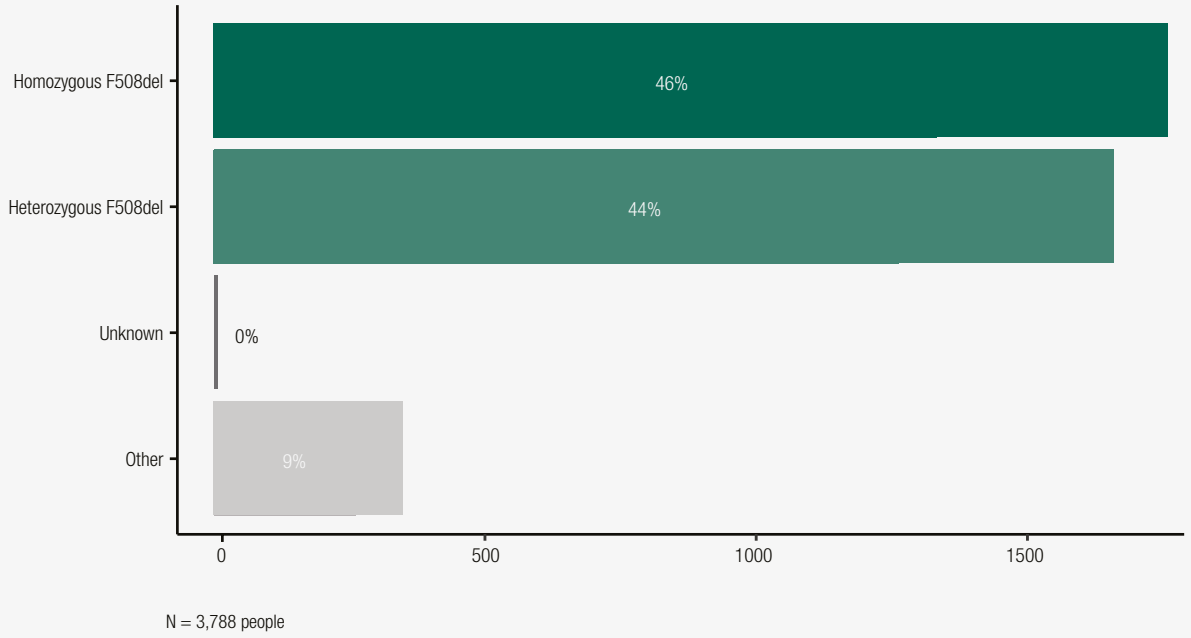


Genotype Information

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 3,788 people in the registry in 2024, 99.8% had at least one allele known and 99.7% had both alleles known. Only 8 (0.2%) of pwCF who had sufficient diagnostic information recorded had two unknown alleles, with 5 of these being adults.

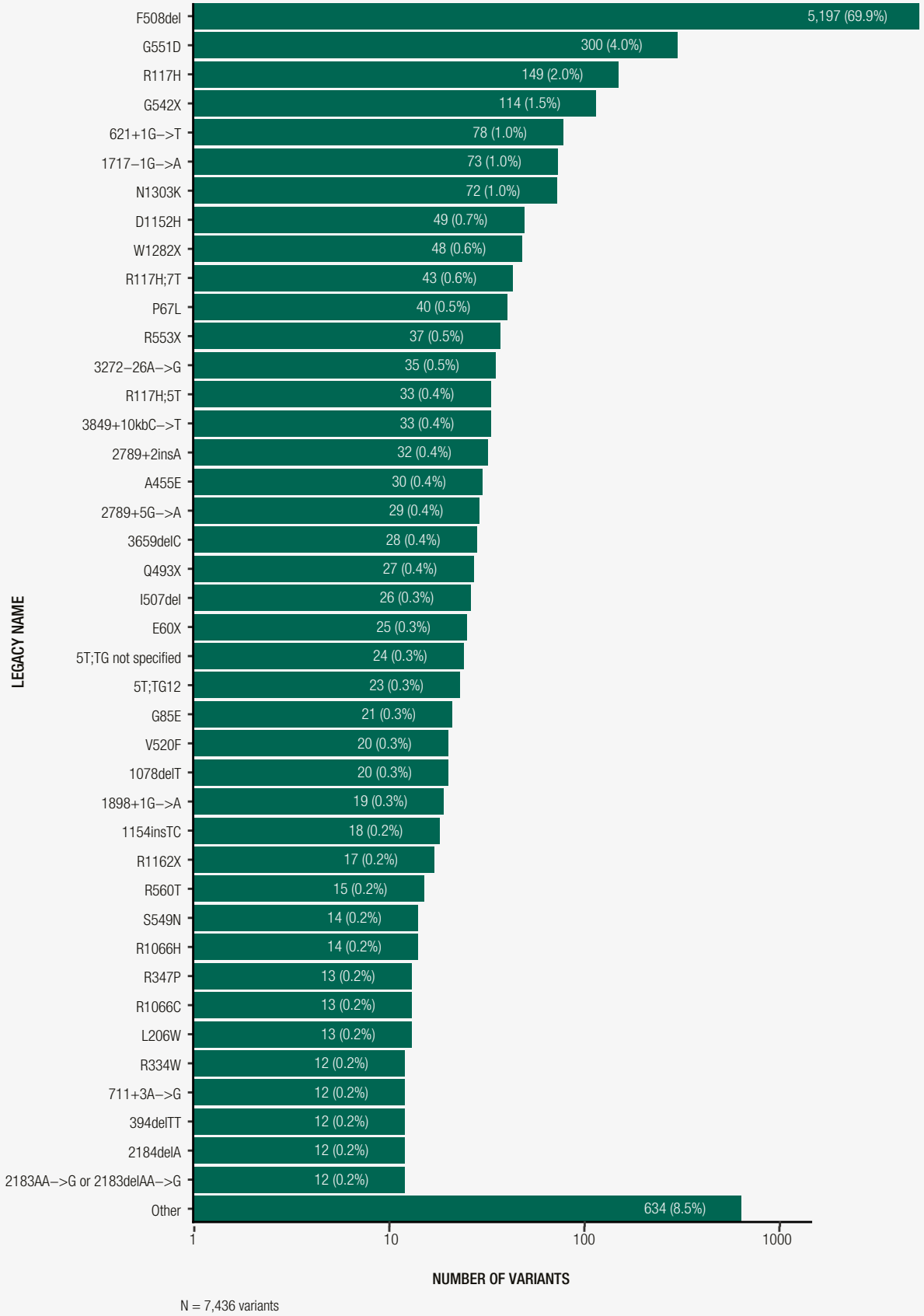
The most common CF-causing variant is F508del. PwCF can either be homozygous to F508del (have two F508del alleles) or heterozygous (have one F508del allele). In 2024, the proportion of Australian pwCF who were homozygous for the F508del variant was 46%, and the proportion who are heterozygous was 44%, thus 90% of pwCF in Australia have F508del variants. A further 9% of pwCF do not have F508del alleles (Figure 1.8).

FIGURE 1.8: ACFDR 2024: MOST COMMON CFTR VARIANT COMBINATIONS



The most common variant alleles other than F508del in 2024 were G551D (4.0%), R117H (2.0%) and G542X (1.5%) (Figure 1.9).

FIGURE 1.9: ACFDR 2024: MOST COMMON INDIVIDUAL ALLELE CFTR VARIANTS IN THE ACFDR



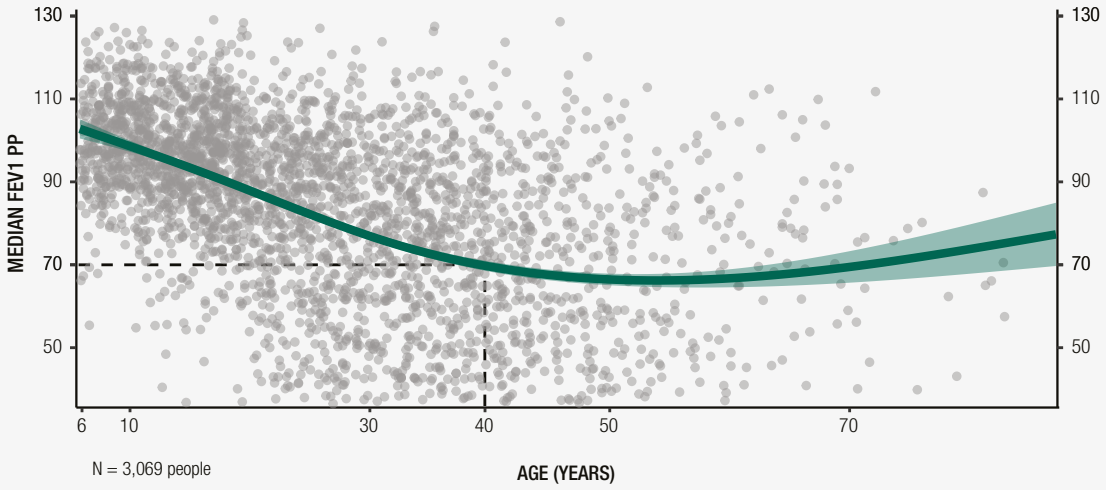
1.4 LUNG FUNCTION

For monitoring lung function in pwCF, the average of the highest Percentage Predicted Forced Expiratory Volume (litres) in 1 second (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 2024 (3,069 people). Four hundred and forty-nine (11.2%) pwCF in the registry are children younger than 6 years of age who do not routinely have lung function information recorded, and a further 7.0% of registry participants did not have lung function information recorded in 2024.

For 2024, the median lung function for pwCF, measured as FEV1 pp, is within the normal range for young children and adolescents (0-17) (Figure 1.10). At 40 years of age, the median FEV1 pp is 70.0.

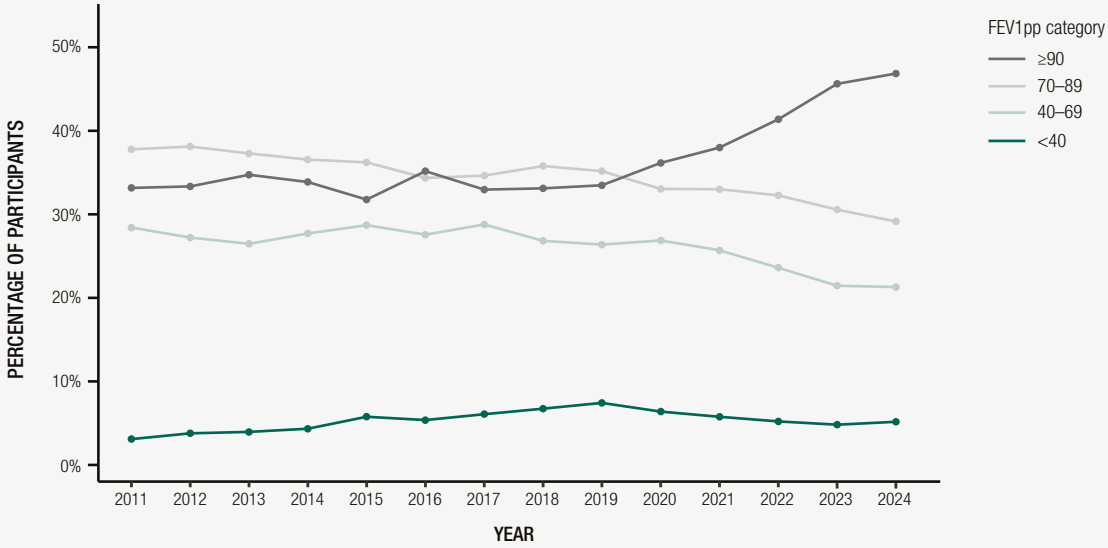
FIGURE 1.10: ACFDR 2024: MEDIAN FEV1 PP BY AGE



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

Figure 1.11 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 46.0% - 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 pp. The proportion of pwCF who have median FEV1 pp of <40% remains approximately 5%.

FIGURE 1.11: ACFDR 2011-2024: FEV1 PP DISTRIBUTION OVER TIME

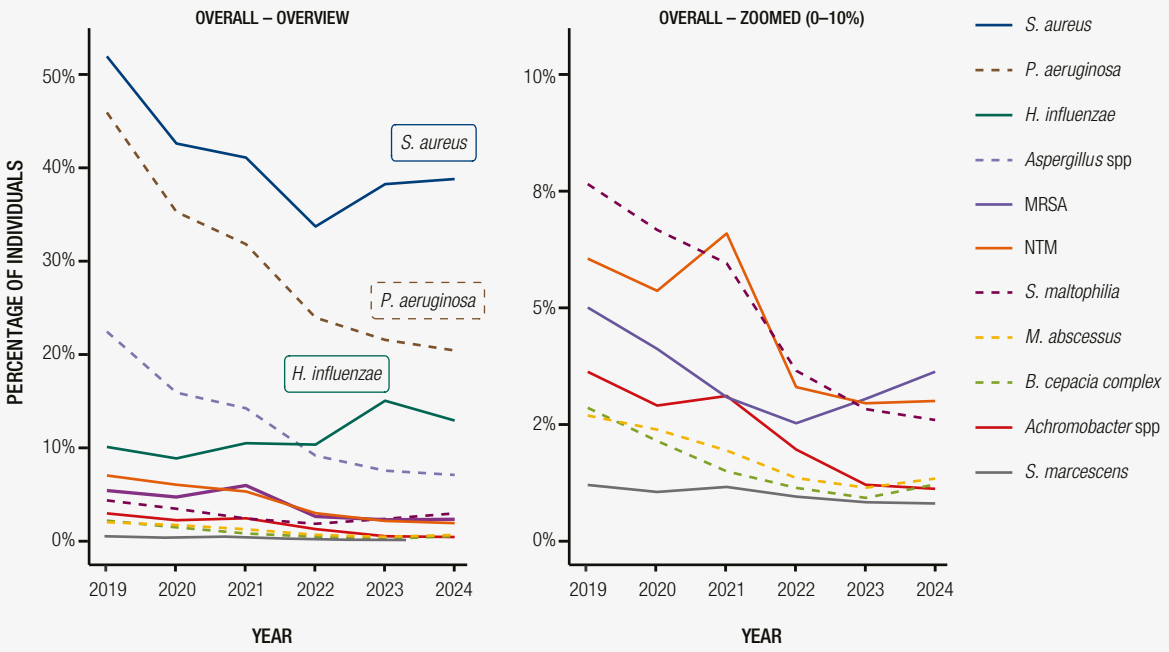


1.5 MICROBIOLOGY

Figure 1.12 shows the prevalence of respiratory organisms commonly pathogenic to pwCF. There has been a notable reduction over a number of years in many high prevalence organisms, including *S. aureus*, *P. aeruginosa* and *Aspergillus* spp, however, 2023 and 2024 have seen slight increases in the prevalence of *S. aureus* and *H. influenzae*.

As of December 2024, the overall prevalence in the registry of *S. aureus* was 39.2%; *P. aeruginosa* was 20.9%; *H. influenzae* was 13.5%; *Aspergillus* spp was 7.7%; MRSA was 3.6%; Non-Tuberculous Mycobacterium was 3.0%; *S. maltophilia* was 2.6%; *M. abscessus* was 1.4%; *B.cepacia_complex* was 1.2%; *Achromobacter* spp was 1.3%, and *S.marcescens* had a prevalence of 0.8%.

FIGURE 1.12: ACFDR 2019-2024: PREVALENCE OF RESPIRATORY MICROORGANISMS IN SPUTUM, BAL BRONCHOSCOPY AND UPPER AIRWAY SAMPLES

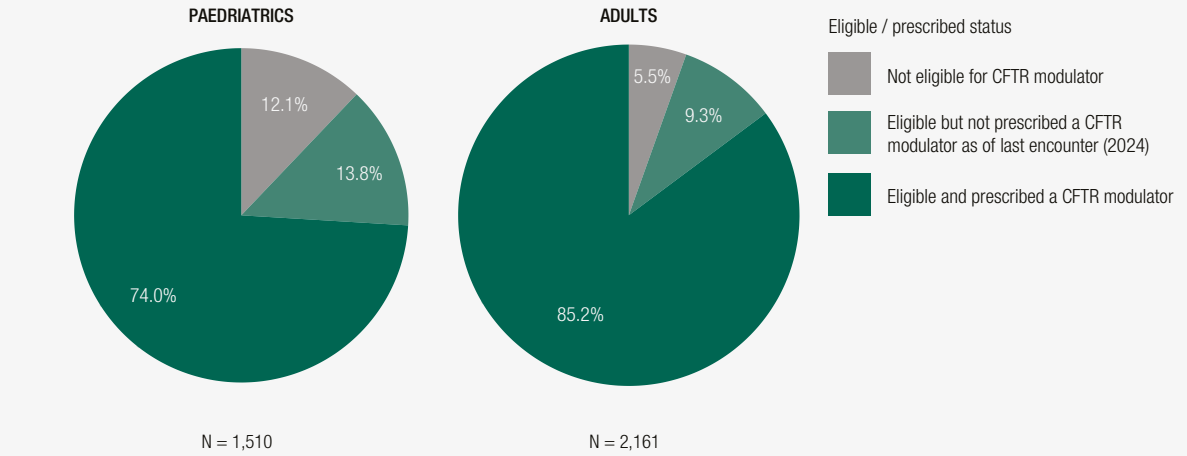


1.6 CFTR MODULATORS

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 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).

A total of 3,671 pwCF (97%) in the registry had known eligibility status for CFTR modulators based on their genotype (Figure 1.13). Of these, 3,027, or 82.5% of all pwCF in the registry have been prescribed at least one modulator during 2024.

FIGURE 1.13: ACFDR 2024: ELIGIBILITY AND PRESCRIBING OF CFTR MODULATORS BY AGE



As of 31 Dec 2024

The number of modulator prescriptions in 2024 increased by 162; up from 2,865 in 2023. Further information about CFTR modulators is provided in the paediatric and adult sections of this report.

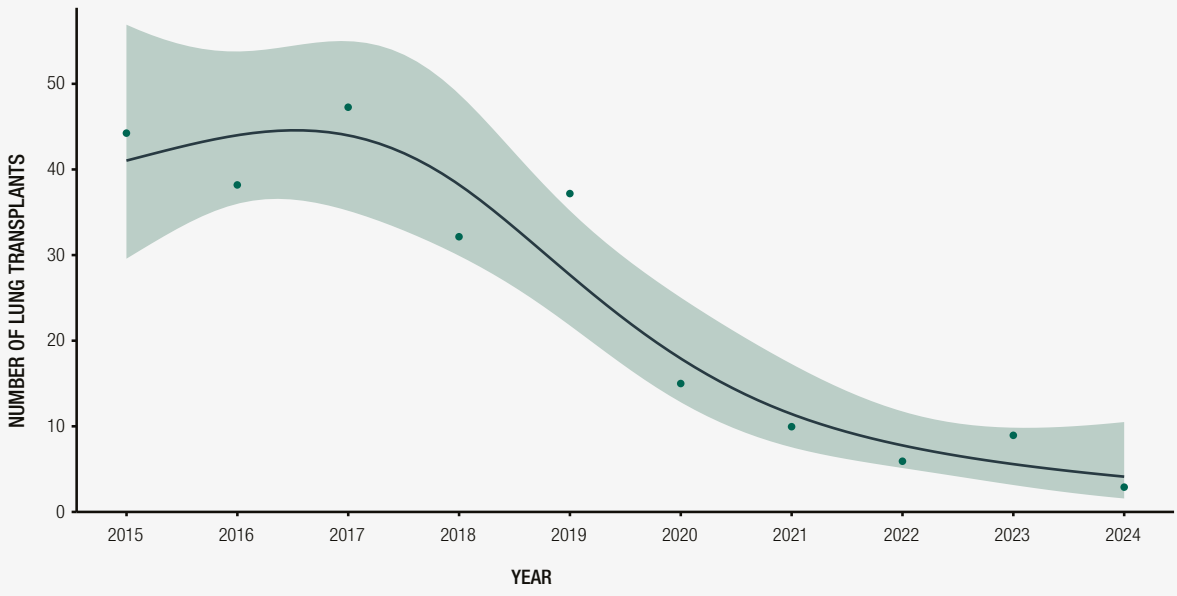
1.7 TRANSPLANTATION AND SURVIVAL

Transplantation

The most common transplantation procedure 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 2024, there were a total of 7 transplants for pwCF; 3 of which were bilateral lung transplants of which all were performed in people aged 30 years or older. The 4 non-lung transplants were comprised of liver, kidney, and pancreas transplants. There were 46 people who were evaluated for a transplant in 2024; 4 (8.7%) were waitlisted, and 5 (10.9%) were deferred from the waiting list. The number of annual bilateral lung transplants undertaken over the last decade is shown in Figure 1.14. There has been a substantial decline in bilateral lung transplants over the last few years among pwCF in Australia, consistent with international trends.

FIGURE 1.14: ACFDR 2015-2024: 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

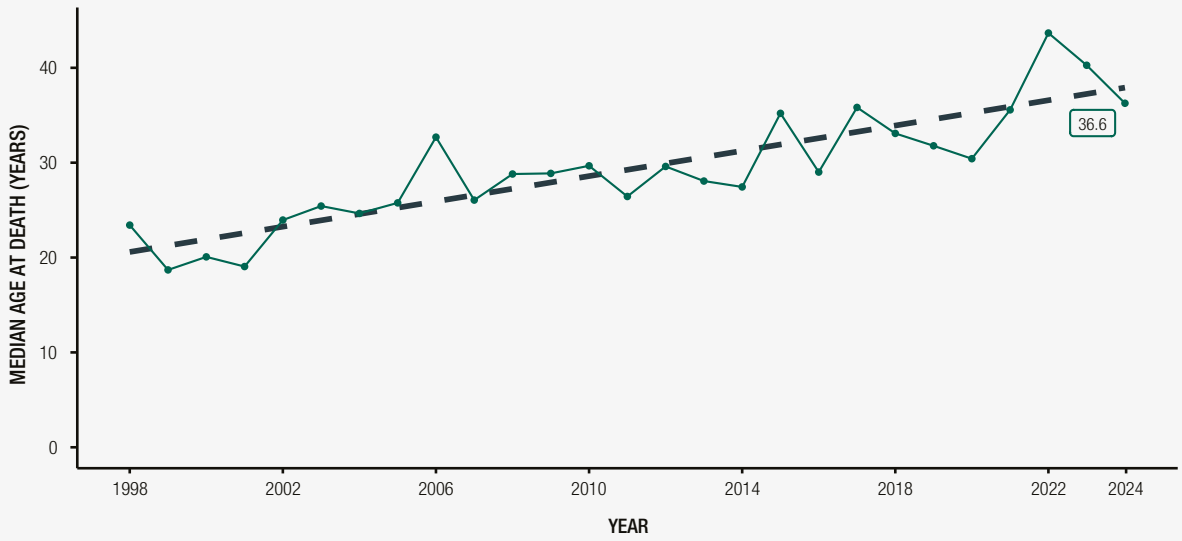
The (follow-up) status of people in the ACFDR is updated annually by CF centres. Many pwCF who have undergone organ transplantation may not have been followed up by the ACFDR, and their deaths may not be captured in the registry.

In 2024, the ACFDR recorded the deaths of 11 pwCF, eight of which (72.7%) were pwCF who had received a lung transplant. Eighty-two percent of deaths occurred in pwCF of 30 years of age or older. In 2024, the causes of the 11 deaths among individuals with CF included post-transplant complications (6), pulmonary manifestations (1), other and non-specified CF-related causes (2), and non-CF-related causes (2).

Median Age of Death

The median age of death in 2024 was 36.6 years of age for pwCF (Figure 1.15). 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 1.15: ACFDR 1998-2024: 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 1.4 is inclusive of pwCF who have undergone a lung transplant, while Table 1.5 excludes pwCF who have undergone a lung transplant. Table 1.5 (also represented in Figure 1.16) shows, that the estimated 5-year survival has increased over a 5-year period from 47.0 years for pwCF born in 2008-12, to 64.0 years for pwCF born in 2019-23. For pwCF, excluding those with lung transplants, survival is even higher, with a median survival of 82.1 years for pwCF born in 2019-23.

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 1.4: ACFDR 2010-2023: MEDIAN SURVIVAL OF PEOPLE WITH CF IN AUSTRALIA (LUNG TRANSPLANTS INCLUDED)

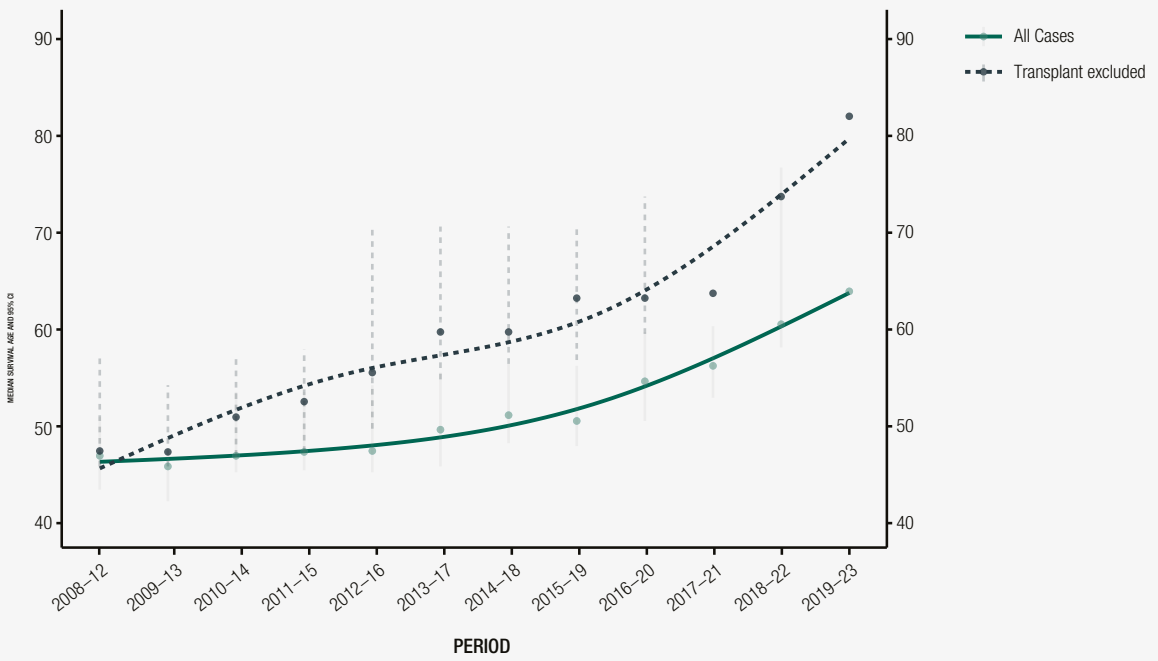
Period	Median age and 95% CI (years)	Number of deaths/Number at risk
2010-14	47.4 (45.3-53.6)	174 / 3,413
2011-15	47.4 (45.5-53.6)	174 / 3,495
2012-16	47.5 (45.4-54.3)	181 / 3,556
2013-17	49.7 (45.9-55.6)	179 / 3,587
2014-18	51.2 (48.3-56.9)	175 / 3,720
2015-19	51.2 (48-56.3)	180 / 3,789
2016-20	54.7 (50.6-59.8)	171 / 3,821
2017-21	56.9 (53-60.4)	154 / 3,861
2018-22	60.6 (58.2-76.8)	120 / 3,898
2019-23	64.0 (60.4-NA)	97 / 3,938

TABLE 1.5: ACFDR 2010-2023: MEDIAN SURVIVAL OF PEOPLE WITH CF IN AUSTRALIA (LUNG TRANSPLANTS EXCLUDED)

Period	Median age and 95% CI (years)	Number of deaths/Number at risk
2010-14	51 (47-58)	107 / 3,007
2011-15	52.6 (47.5-58)	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

Survival is also shown graphically below for all pwCF in the registry (Figure 1.16).

FIGURE 1.16: ACFDR 2008-2023: 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).

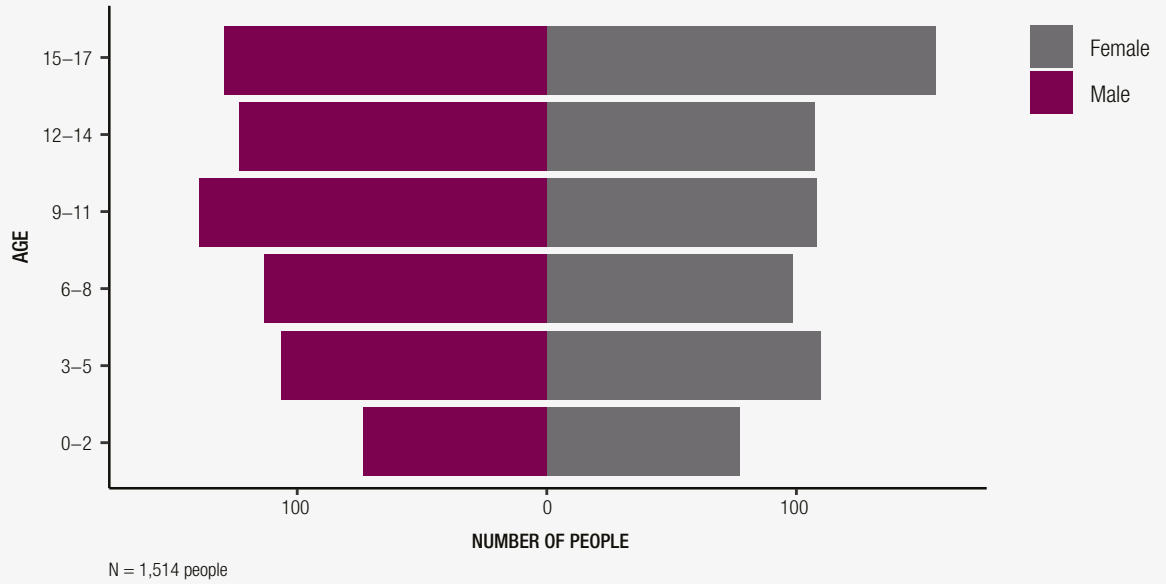
2. PAEDIATRIC DATA

2. PAEDIATRIC DATA

2.1 DEMOGRAPHIC INFORMATION

As of 31st December 2024, the ACFDR held data regarding 1,514 children and adolescents (0-17 years old) with CF, comprising 741 females and 773 males (Figure 2.1).

FIGURE 2.1: ACFDR PAEDIATRICS 2024: BY AGE AND SEX



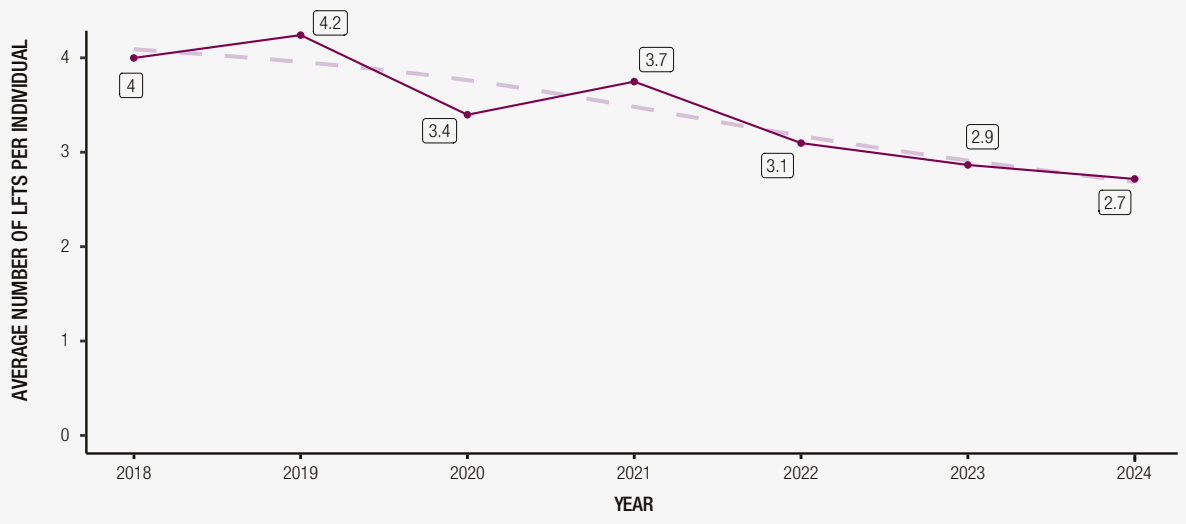
N = 1,514 people

2.2 CLINICAL MEASURES

Lung Function

Figure 2.2 illustrates the average number of spirometry tests per child recorded annually in the registry from 2018 to 2024. In 2018, children and adolescents averaged 4 tests each, however this has declined to 2.7 tests per child in 2024.

FIGURE 2.2: ACFDR PAEDIATRICS 2018-2024: AVERAGE NUMBER OF LUNG FUNCTION TESTS ENTERED PER INDIVIDUAL



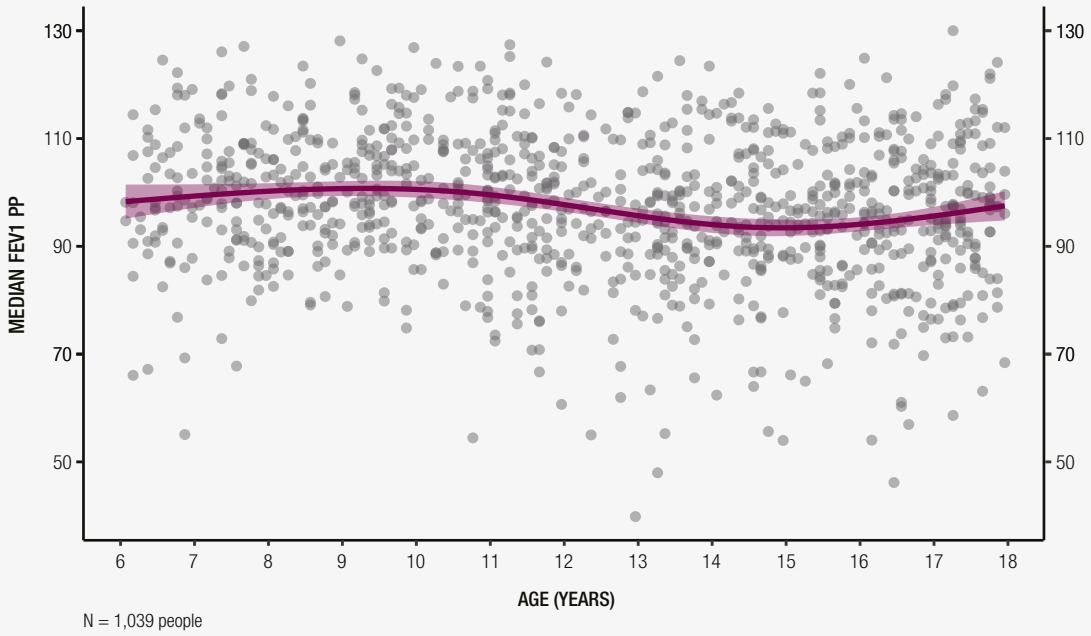
Dashed curve represents the smoothed trend (natural cubic spline, df = 3)

Median Lung Function

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 Global Lung Initiative (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 2024 median lung function is shown in the scatterplot below for 1,039 children and adolescents. 11.2% 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 4.0% of paediatric registry participants did not have lung function information recorded in 2024 (Figure 2.3).

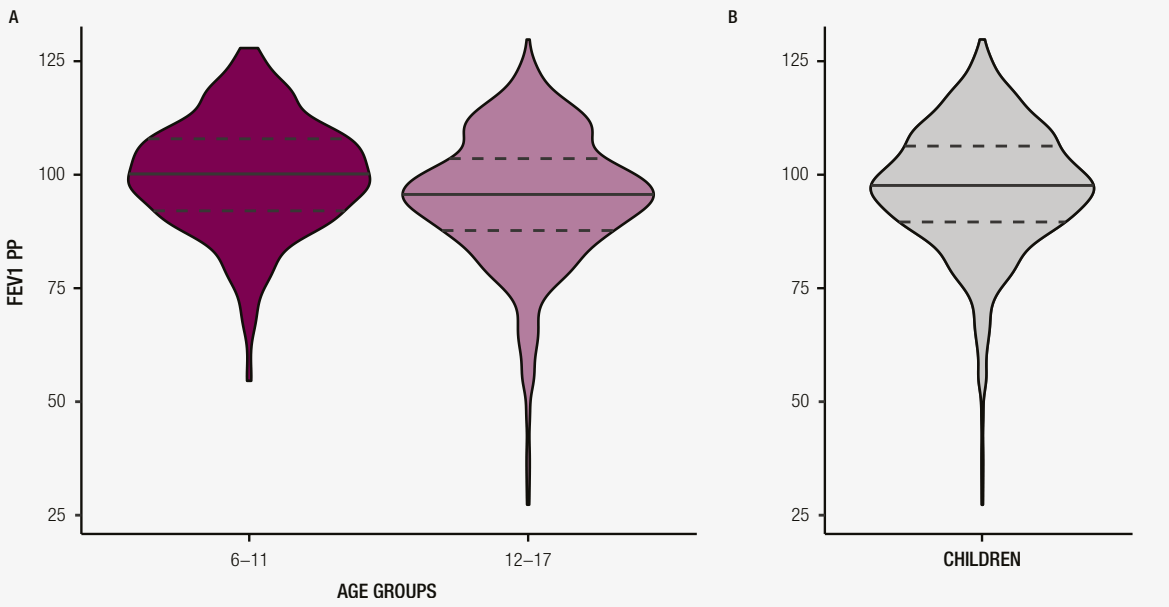
FIGURE 2.3: ACFDR PAEDIATRICS 2024: FEV1 PP BY AGE



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

In 2024, the median FEV1 pp for 6 to 11-year olds was 100.2 pp and for 12-17-year olds was 95.7 pp. The median FEV1 pp for children and adolescents 6-17 years in 2024 was 97.7 pp (Figure 2.4).

FIGURE 2.4: ACFDR PAEDIATRICS 2024: MEDIAN FEV1 PP BY AGE GROUP



The median FEV1 pp for children and adolescents has increased over time. For 6-year-olds, it has increased from 94.9 pp in 2019 to 98.1 percent predicted in 2024; for 12-year-olds it has increased from 91.8 pp to 94.4 pp; and for 17-year-olds, it has increased from 86.2 pp to 97.3 pp in 2024 (Figure 2.5).

FIGURE 2.5: ACFDR PAEDIATRICS 2019-2024: MEDIAN FEV1 PP OVER TIME

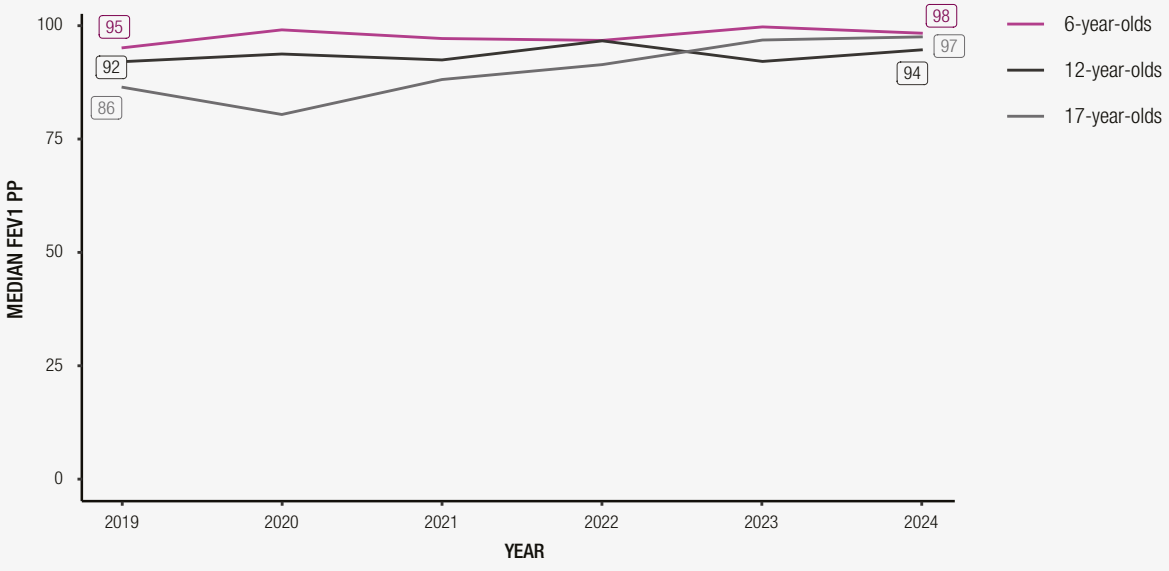
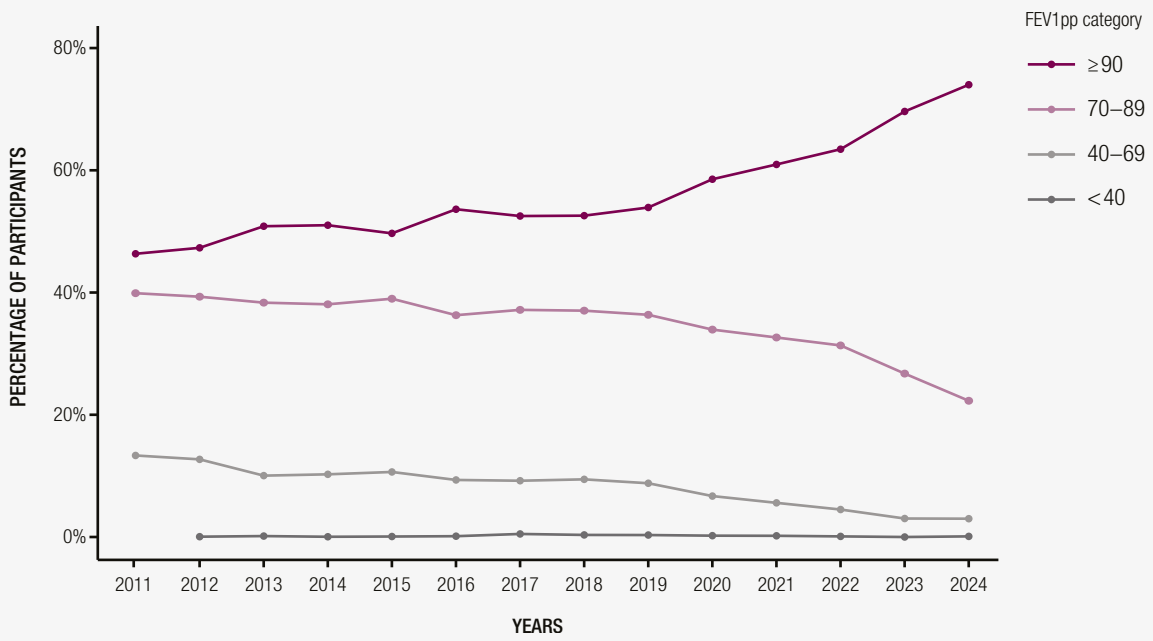


Figure 2.6 depicts the trend in FEV1 pp for children and adolescents with CF since 2011. In recent years, there has been a significant increase in the proportion of pwCF with average FEV1 pp scores above 90, from 46.5% in 2011 to 74.1% in 2024.

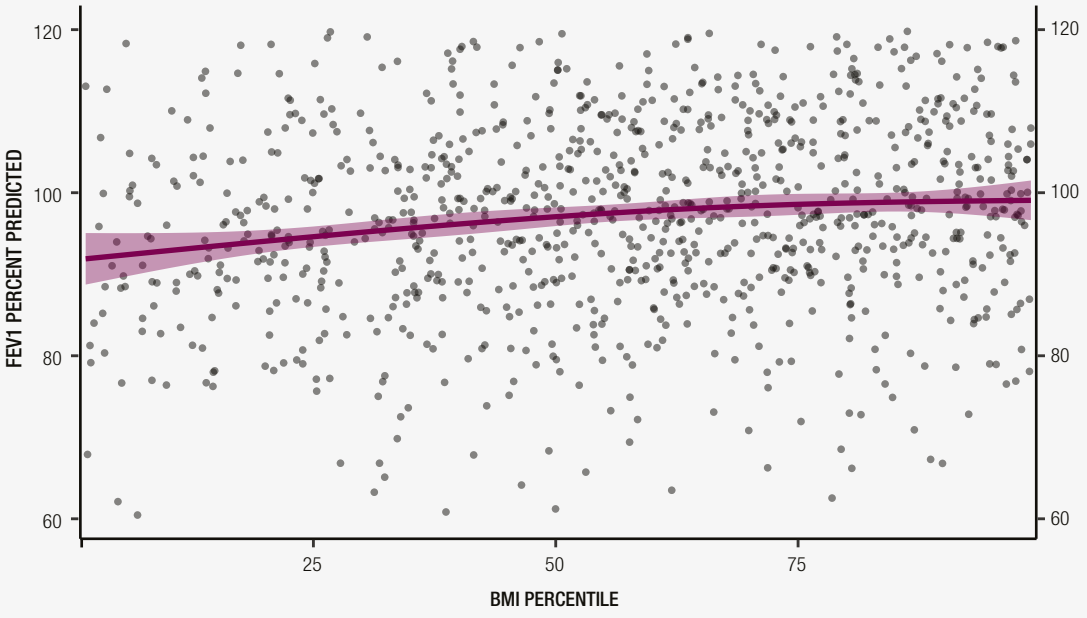
FIGURE 2.6: ACFDR PAEDIATRICS 2011-2024: FEV1 PP DISTRIBUTION OVER TIME



Lung Function and Body Mass Index

The relationship between FEV1 pp and Body Mass Index (BMI) for 2024 is shown in Figure 2.7. In general, as BMI increases, so does FEV1 pp.

FIGURE 2.7: ACFDR PAEDIATRICS 2024: FEV1 PP VS BMI PERCENTILE

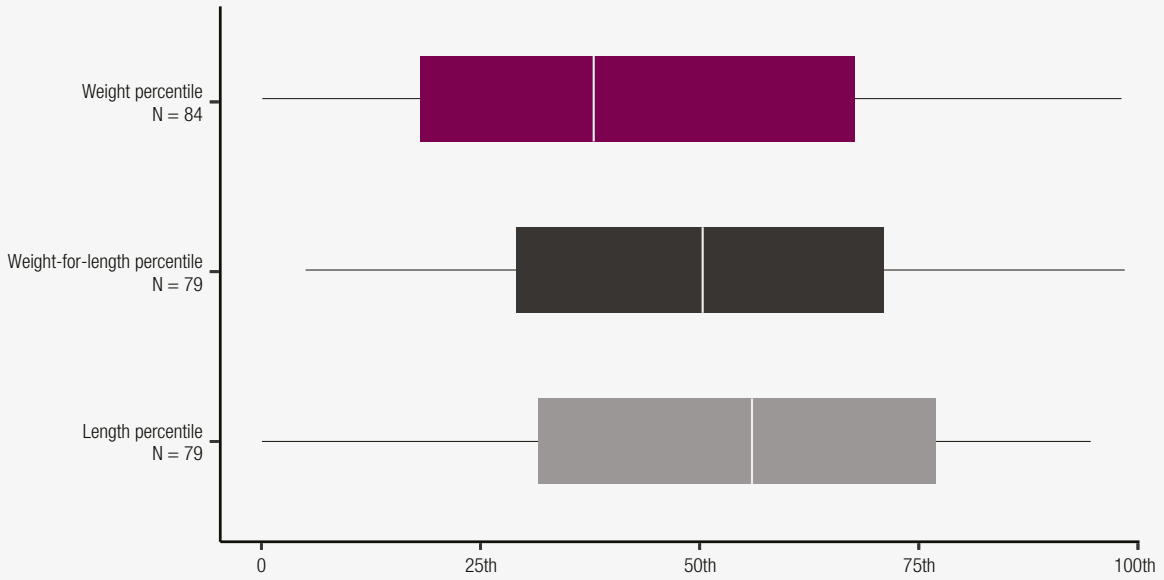


Solid line was estimated using a natural cubic spline with 3 degrees of freedom
Shaded area represents 95% confidence interval

Nutrition

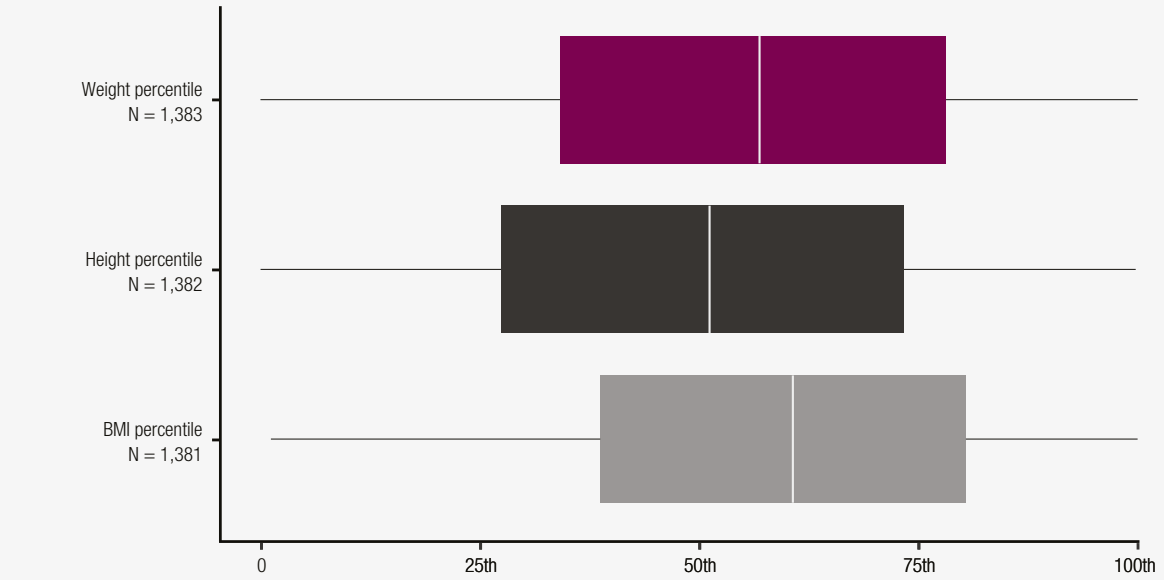
For 2024, infants (<24 months of age) had a median weight on the 38th percentile. The median length was on the 56th percentile; and the median weight-for-length was on the 51st percentile (Figure 2.8).

FIGURE 2.8: ACFDR PAEDIATRICS 2024: WEIGHT, LENGTH, AND WEIGHT-FOR-LENGTH PERCENTILES LESS THAN 24 MONTHS



In 2024, for children and adolescents aged 2-17 years, the median weight was on the 57th percentile, the median height was on the 51st percentile, and median BMI was 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 2024: 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 male and female children and adolescents with CF was in the optimal and acceptable BMI percentile ranges for 2024 (Table 2.1), with 18% being overweight/obese, and 13% having suboptimal nutritional status. The proportion of overweight/obese children/adolescents with CF has increased from 15.5% in 2023 to 18.2% in 2024.

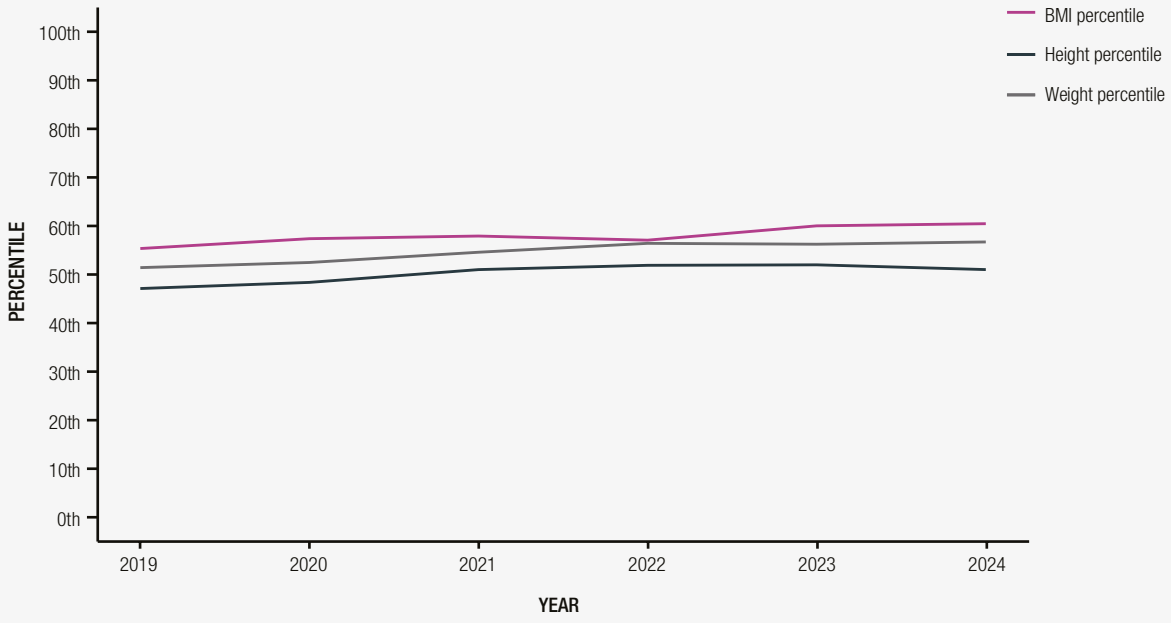
TABLE 2.1: ACFDR PAEDIATRICS 2024: <2-17 YEARS NUTRITIONAL STATUS

Nutritional Status*	<2 years	2-5 years	6-11 years	12-17 years	Total (n = 1,454)
Optimal/Acceptable	90.4% (66)	64.1% (191)	68.7% (349)	68.7% (395)	68.8% (1,001)
Overweight/Obese	0.0% (0)	27.9% (83)	17.3% (88)	16.2% (93)	18.2% (264)
Suboptimal/Undernourished	9.6% (7)	8.1% (24)	14.0% (71)	15.1% (87)	13.0% (189)

* 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 4 percentile points (from the 47th percentile to the 51st percentile), and weight has increased by 5 percentile points (from the 52nd percentile to the 57th percentile). As a result, the average BMI also increased by 5 percentile points during this time, from the 56th percentile to the 61st percentile (Figure 2.10).

FIGURE 2.10: ACFDR PAEDIATRICS 2019-2024: 2-17 YEARS MEDIAN NUTRITIONAL PERCENTILES

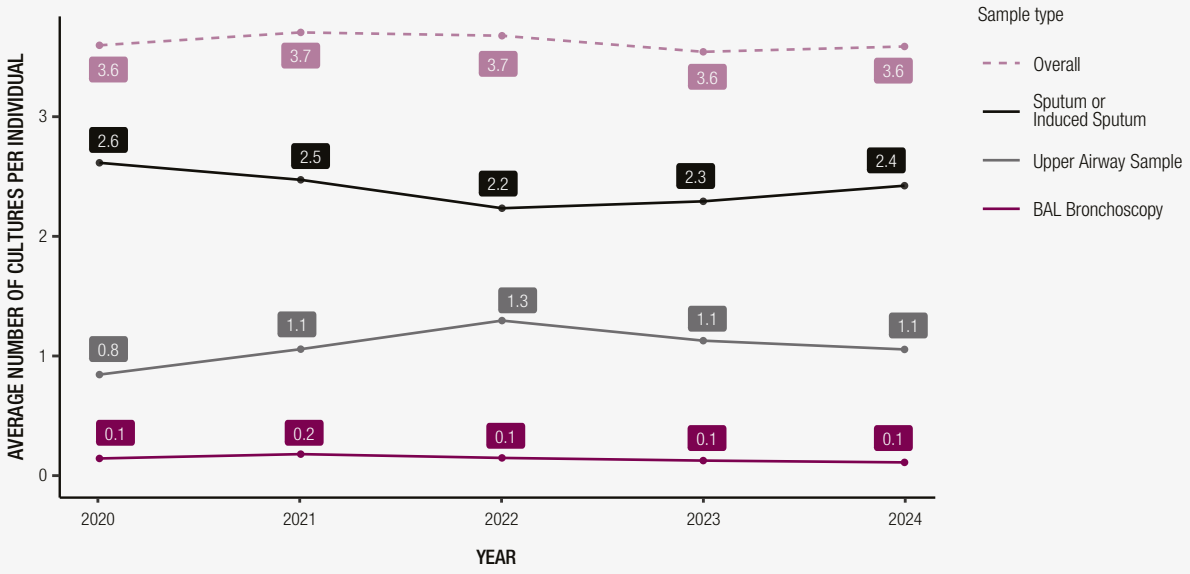


Microbiology

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 3.6 per person in 2020 to an average of 3.6 samples in 2024.

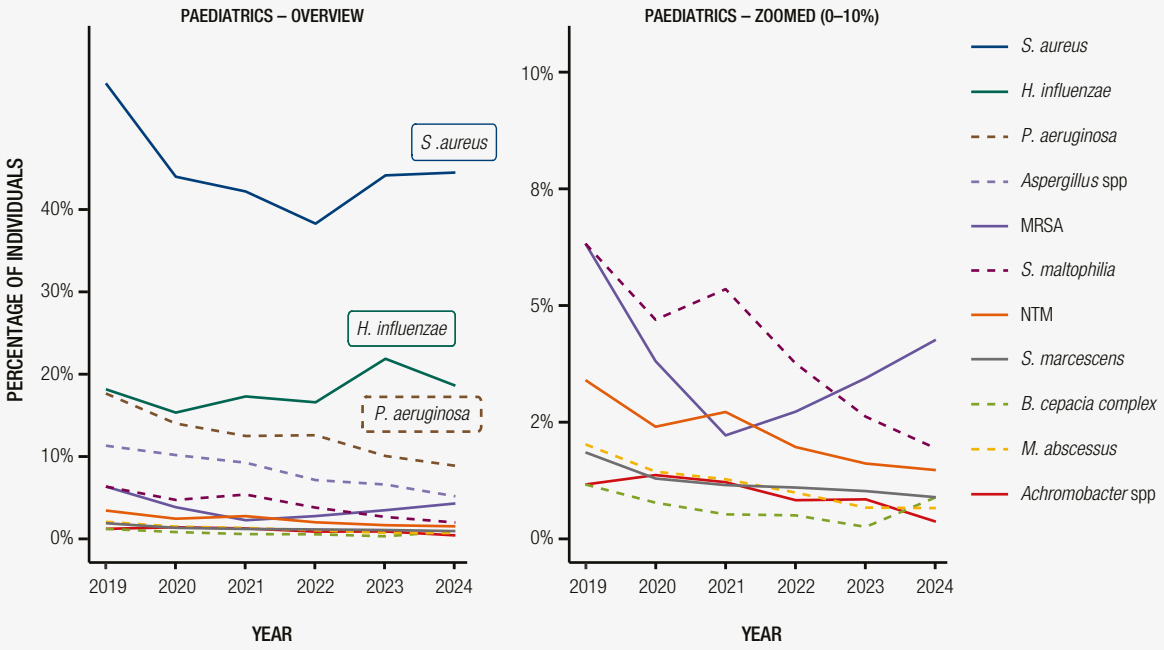
When considering the specific sample types, sputum/induced sputum samples accounted for the greatest proportion, at 2.4 samples per person in 2024. Upper airway samples averaged 1.1 per person (throat swabs, cough swabs, nasopharyngeal swabs and sinus wash), and bronchoscopy/BAL samples averaged 0.1 in 2024.

FIGURE 2.11: ACFDR PAEDIATRICS 2019-2024: MEAN NUMBER OF RESPIRATORY SAMPLES PER INDIVIDUAL



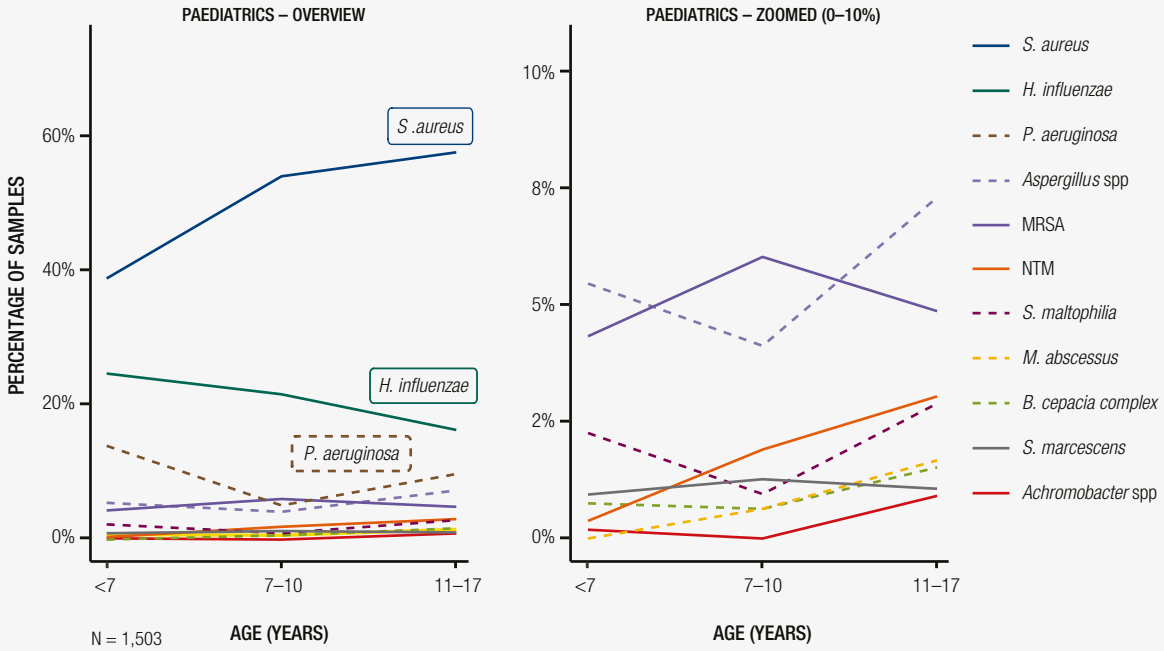
The prevalence of some of the most common paediatric microorganisms has changed over the last 6 years. The prevalence of *S. aureus* has plateaued at 44.5% in 2024; the prevalence of *H. influenzae* has decreased to 18.6%; the prevalence of *P. aeruginosa* has decreased to 8.9% in 2024; and *Aspergillus* spp has decreased to 5.2% in 2024. The prevalence of less common paediatric microorganisms has mostly decreased, except for *M. abscessus*, with a 2024 prevalence of 0.7% (Figure 2.12).

FIGURE 2.12: ACFDR PAEDIATRICS 2019-2024: PREVALENCE OF RESPIRATORY MICROBIOLOGY



Of the 1,513 microbiology culture samples collected in 2024, the most common pathogen across all age groups was *S. aureus*, with the prevalence increasing with age (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 38.8% in children <7 years, 54.0% in children and adolescents 7-11 years and 57.5% in adolescents 12-17 years. The prevalence of *H. influenzae* decreases with age, being 24.7% for children <7 years, 21.6% for children and adolescents 7-11 years, and 16.3% for adolescents aged 12-17 years. This is also shown graphically in Figure 2.13.

FIGURE 2.13: ACFDR PAEDIATRICS 2024: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE



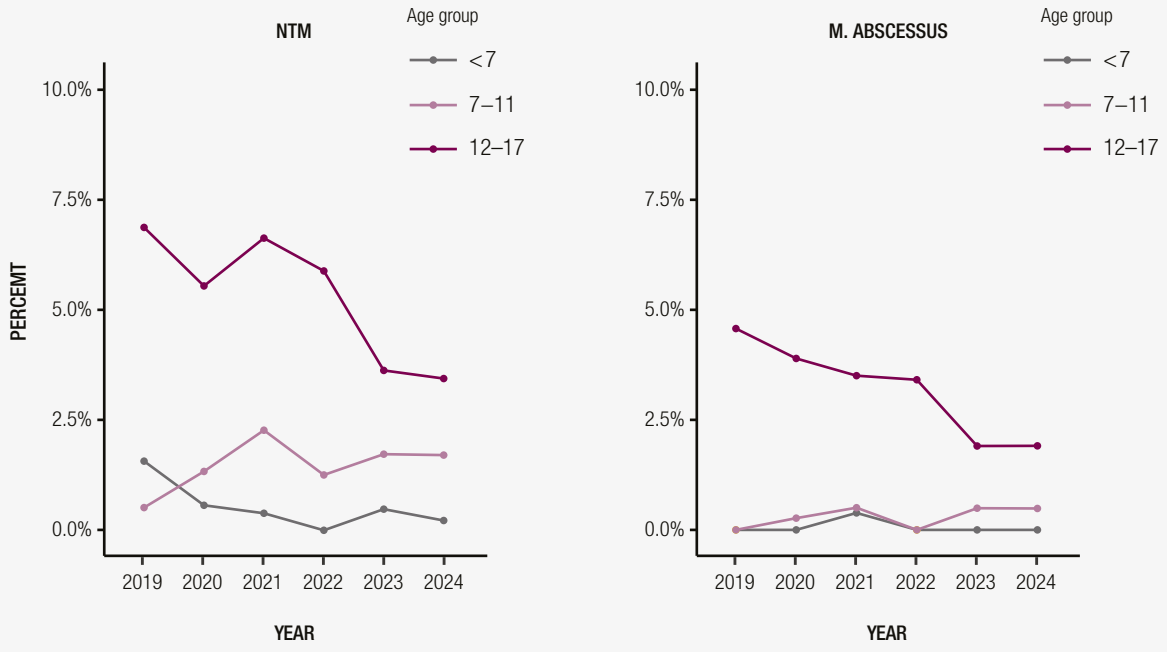
For children younger than seven years, 105 lower airway samples were collected by bronchoalveolar lavage (BAL) in 2024. The most common organisms identified in this age group in 2024 included *S. aureus* (25.7%), *H. influenzae* (22.9%), *Aspergillus* spp (13.3%), *P. aeruginosa* (11.4%), and MRSA (6.7%) (Table 2.2).

TABLE 2.2: ACFDR PAEDIATRICS 2024: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE

	BAL samples	All samples		
	<7 years	<7 years	7-11 years	12-17 years
Number of samples in the age range	493	493	331	689
Number of samples taken in 2024	105	531	315	657
Number of patients	105	453	306	631
S. aureus	27 / 105 (25.7%)	206 / 531 (38.8%)	170 / 315 (54.0%)	378 / 657 (57.5%)
H. influenzae	24 / 105 (22.9%)	131 / 531 (24.7%)	68 / 315 (21.6%)	107 / 657 (16.3%)
P. aeruginosa	12 / 105 (11.4%)	74 / 531 (13.9%)	16 / 315 (5.1%)	64 / 657 (9.7%)
Aspergillus spp	14 / 105 (13.3%)	29 / 531 (5.5%)	13 / 315 (4.1%)	48 / 657 (7.3%)
B. cepacia complex	0 / 105 (0.0%)	<5	<5	10 / 657 (1.5%)
MRSA	7 / 105 (6.7%)	23 / 531 (4.3%)	19 / 315 (6.0%)	32 / 657 (4.9%)
Achromobacter spp	0 / 105 (0.0%)	<5	0 / 315 (0.0%)	6 / 657 (0.9%)
S. maltophilia	6 / 105 (5.7%)	12 / 531 (2.3%)	<5	19 / 657 (2.9%)
S. marcescens	0 / 105 (0.0%)	5 / 531 (0.9%)	<5	7 / 657 (1.1%)
Nontuberculous mycobacteria	<5	<5	6 / 315 (1.9%)	20 / 657 (3.0%)

The prevalence of NTM among 7-11-year olds in 2024 was 1.7% and in 12-17-year olds was 3.4%. The prevalence of *M abscessus* among 7-11-year olds in 2023 was 0.5% and among 12-17-year olds was 1.9%. Since 2021, the prevalence of NTM and *M.abscessus* has decreased (Figure 2.14).

FIGURE 2.14: ACFDR PAEDIATRICS 2019-2024: PREVALENCE OF NTM INFECTIONS BY AGE



Pediatric Trends in NTM & *M. abscessus* (2019-2024)

2.3 CF MANAGEMENT

Clinic Visits

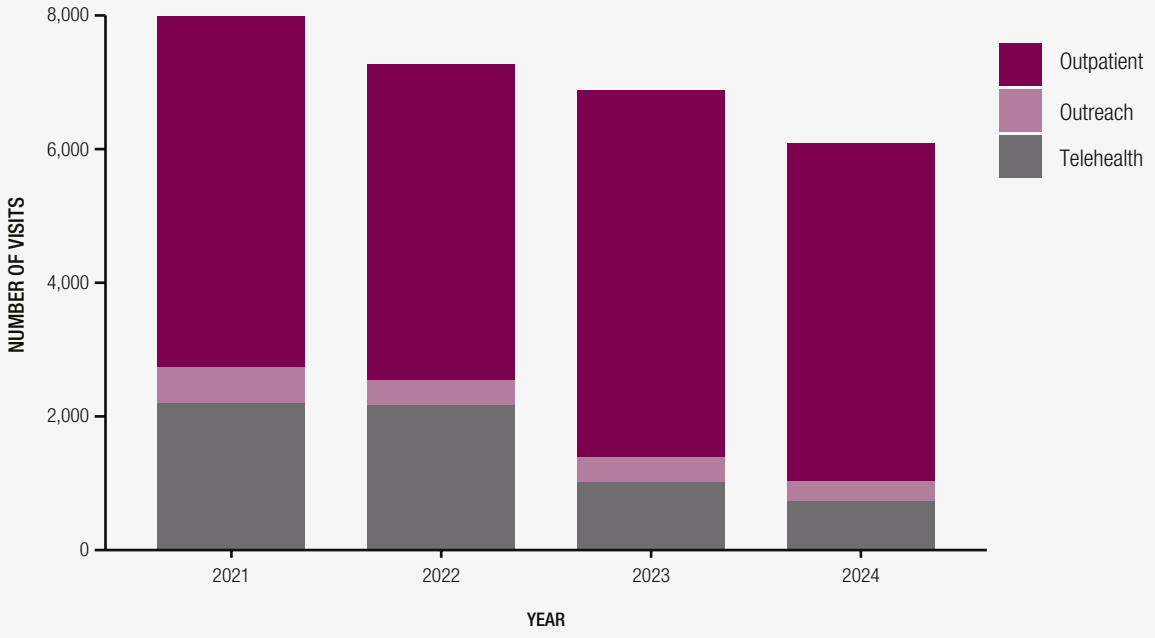
Table 2.3 and Figure 2.15 show the total number of clinical visits for the paediatric population in the registry per year over the last 4 years. The total number of clinical visits peaked in 2021 (8,089 encounters), with visits declining since then to a total of 6,135 in 2024.

The nature of clinical visits has also changed during this time. During the COVID-19 pandemic, the proportion of telehealth visits (audio and/or visual visits conducted at home or in a healthcare setting) for children and adolescents were 34% in 2020, decreasing to 12% in 2024. The proportion of outreach visits also decreased from 7% in 2020 to 5% in 2024 (Table 2.3 and Figure 2.15).

TABLE 2.3: ACFDR PAEDIATRICS 2021-2024: TOTAL NUMBER OF VISITS RECORDED PER YEAR

Visit type	2021	2022	2023	2024
Outpatient	5,337 (66.0%)	4,805 (65.0%)	5,562 (80.1%)	5,102 (83.2%)
Outreach	549 (7.0%)	370 (5.0%)	368 (5.3%)	312 (5.1%)
Telehealth	2,203 (27.0%)	2,191 (30.0%)	1,013 (14.6%)	721 (11.8%)
Total	8,089 (100%)	7,366 (100%)	6,943 (100%)	6,135 (100%)

FIGURE 2.15: ACFDR PAEDIATRICS 2021-2024: 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 2024 the number of children/adolescents with CF who had at least 4 clinic visits per year was 992 (61% of all children with CF) overall, compared to 1,213 (80%) in 2021 (Table 2.4). The proportion of children and adolescents receiving four or more clinical encounters were highest among children <2 years old at 69%, followed by 65% for 2-6-year-olds, 63% for 7-11-year-olds and 55% for adolescents.

TABLE 2.4: ACFDR PAEDIATRICS 2021-2024: AGE GROUPS WITH 4 OR MORE CLINICAL VISITS

	Number with 4+ clinic visits per year			
Age	2021	2022	2023	2024
<2	104 (76.9%)	101 (70.0%)	100 (79.0%)	61 (69.0%)
2-6	276 (79.0%)	263 (66.0%)	274 (68.0%)	263 (65.0%)
7-11	398 (69.0%)	277 (62.0%)	298 (68.0%)	278 (63.0%)
12-17	435 (70.0%)	415 (66.0%)	402 (65.0%)	320 (55.0%)
Total	1,213 (80.0%)	1,053 (65.2%)	1,074 (68.0%)	922 (61.0%)

While the majority of children/adolescents had at least 4 visits per year, 16% of children aged under 2 years, 16% of children aged 2-6 years, 19% of children aged 7-11 years, and 23% of adolescents aged 12-17 had 3 visits in 2024. Less than 10% of all children and adolescents with CF had one or no clinical visits in 2024 (Figure 2.16).

FIGURE 2.16: ACFDR PAEDIATRICS 2024: PROPORTION OF PARTICIPANTS HAVING 4 OR MORE CLINICAL VISITS

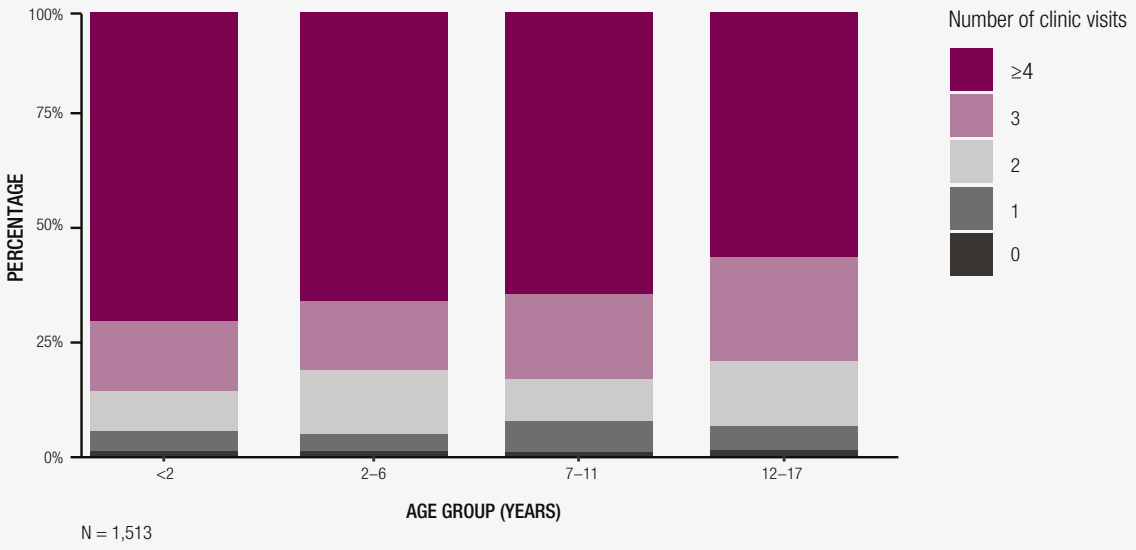


Table 2.5 shows the average number of clinical encounters per child or adolescent with CF in 2024. Average number of clinic visits decreased with age, with children aged less than two having the highest average number of clinic visits per person (5.5) and 7-17-year-olds having the fewest visits (3.8).

TABLE 2.5: ACFDR PAEDIATRICS 2024: AVERAGE NUMBER OF CLINICAL ENCOUNTERS PER PERSON

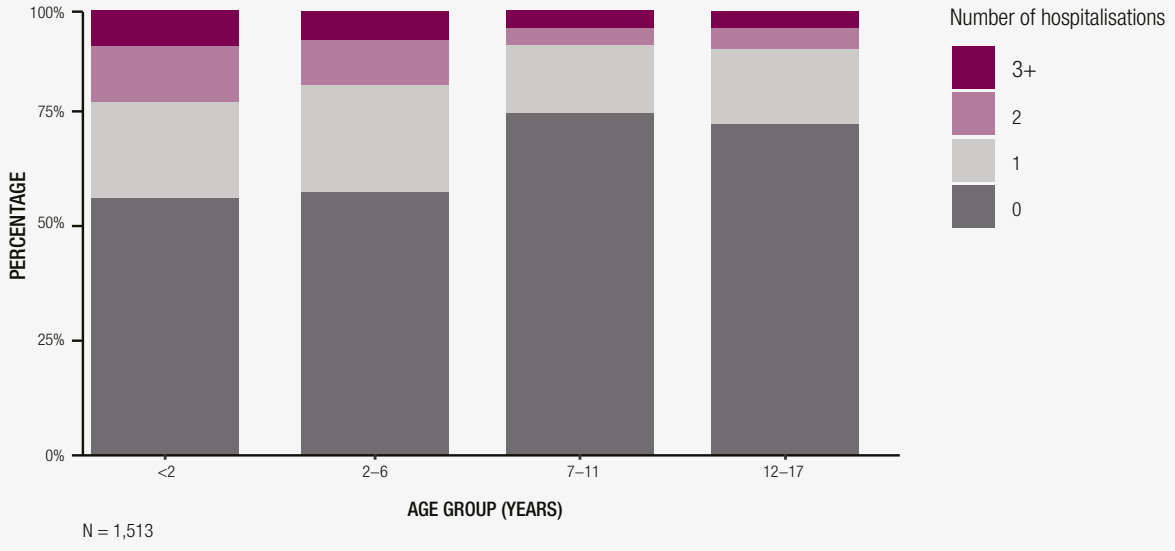
Age	Average number of clinic visits
<2	5.5
2-6	4.0
7-11	3.8
12-17	3.8
Total	3.9

Hospitalisations

There was a total of 788 hospitalisations for children and adolescents in 2024. A majority (58.0%) of children younger than 2 years of age did not have any hospitalisations in 2024, while 21.6% had 1 hospitalisation, 12.5% had 2 hospitalisations, and 8.0% had 3 or more hospitalisations (Figure 2.17). In the 2-6-year age range in 2024, 59.3% had no hospitalisations, with 24.2% having 1, 10.1% having 2, and 6.4% having 3 or more hospitalisations.

Lower proportions of hospitalisations were noted for older children and adolescents. For the 7-11 age group, 77.0% had no hospitalisations, while 15.5% had 1, 3.6% had 2, and 3.9% had at least 3 hospitalisations. Similarly, among adolescents aged 12-17, 74.7% had no hospitalisations, while 16.7% had 1, 4.8% had 2, and 3.8% had at least 3 hospitalisations (Figure 2.17).

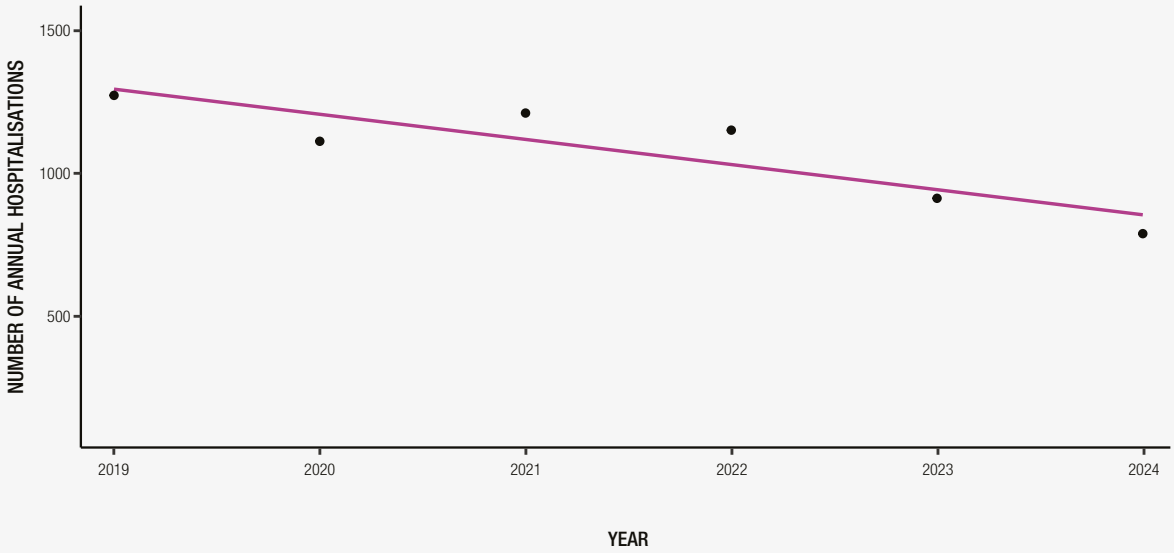
FIGURE 2.17: ACFDR PAEDIATRICS 2024: HOSPITALISATIONS BY AGE GROUP



Paediatric Hospitalisations over time

Paediatric hospitalisations have reduced since 2019, with a decrease from 1,273 admissions in 2019 to 788 admissions in 2024, a reduction of 39% over the last 5 years (Figure 2.18).

FIGURE 2.18: ACFDR PAEDIATRICS 2019-2024: HOSPITALISATIONS PER YEAR

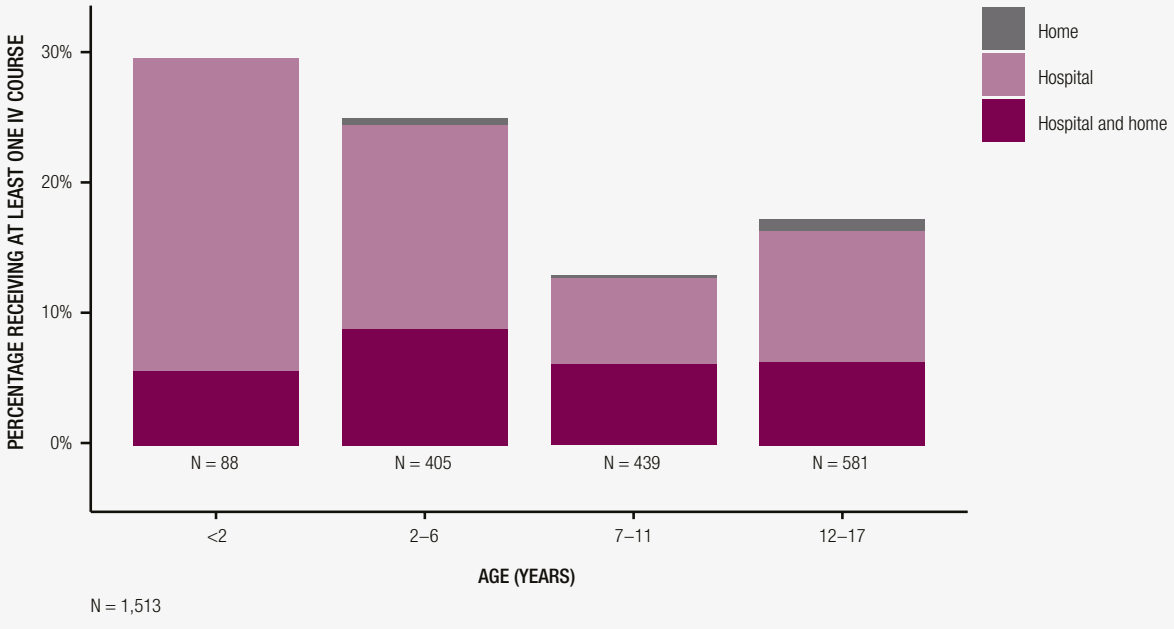


IV Antibiotic therapy

In 2024, the vast majority of IV therapy treatments for children occurred in hospital, with 23.9% of children <2 years, 15.6% of children 2-6 years, 6.6% of 7-11-year-olds, and 10.0% of 12-17-year-olds reporting receiving hospital IV therapy (Figure 2.19).

The use of combined hospital and home IV antibiotics was greatest among 2-6-year-olds, at 8.9%, followed by 6.4% of 12-17-year-olds, 6.2% of 7-11-year-olds and 5.7% of children less than 2 years of age. In 2024, very few children or adolescents received solely home-based IV therapy, with no children less than 2 years of age and only 0.2-0.9% of 2-17-year-olds reporting this (Figure 2.19).

FIGURE 2.19: ACFDR PAEDIATRICS 2024: PARTICIPANTS RECEIVING AT LEAST ONE COURSE OF IV ANTIBIOTIC THERAPY



The median duration of IV antibiotic therapy in hospital was 13 days, with 14 days for combined therapy at home and in hospital, and 9.5 days for therapy at home (Table 2.6).

TABLE 2.6: ACFDR PAEDIATRICS 2024: MEDIAN AND MEAN DAYS IV ANTIBIOTIC THERAPY

IV antibiotic therapy location	Median days receiving therapy	Mean days receiving therapy
Home	9.5	18.3
Hospital	13.0	15.4
Hospital and home	14.0	17.5

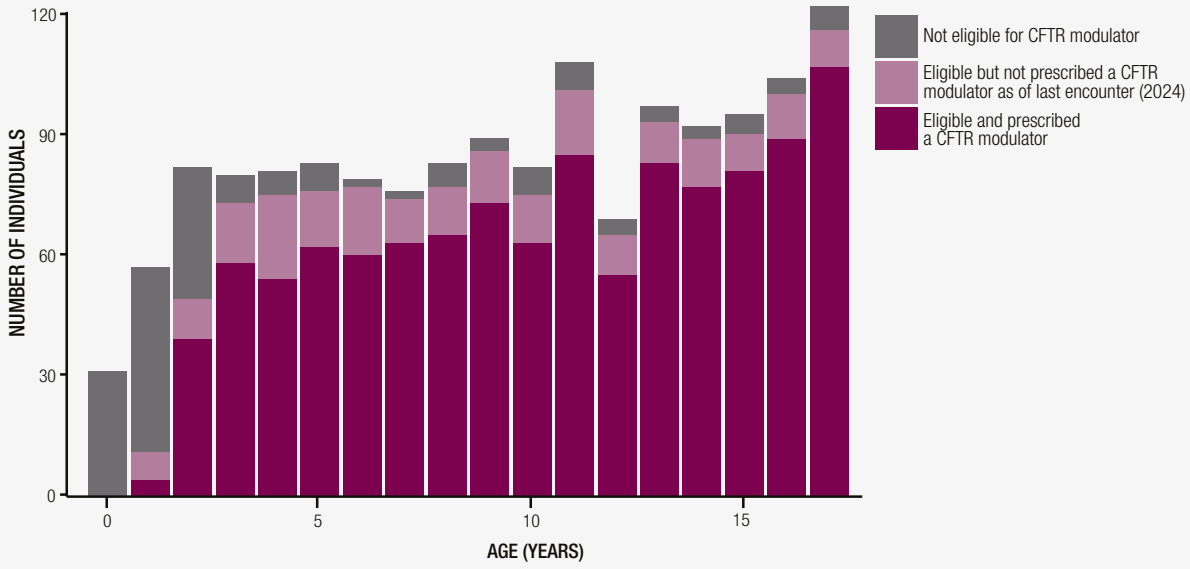
CFTR Modulators

Data were calculated from pwCF who were on a modulator as of December 31st 2024. Data presented here reflect only those pwCF (excluding transplant recipients) who had CFTR modulator data entered into the registry, which generally includes those prescribed modulators available via the PBS.

Of the 1,514 children and adolescents in the registry, eligibility status for a CFTR modulator was known/recorded for 99.7% (1,510 people) in 2024.

Of these, 183 children and adolescents (12.1%) were not eligible for a modulator; 74.0% (1,118 children and adolescents) were eligible and prescribed a modulator; and 13.8% (209 children and adolescents) were eligible and not prescribed a modulator in 2024. 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 2024: CFTR ELIGIBILITY AND PRESCRIBING BY AGE

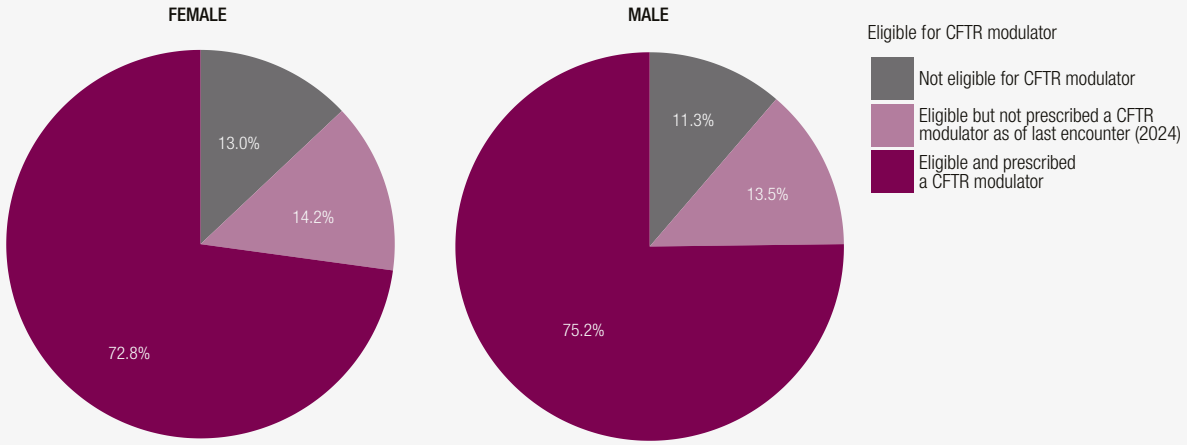


N = 1,510 individuals with known eligibility

The figure below show the eligibility and prescribing information for male and female children and adolescents with CF. The denominator is 1,510 children and adolescents with CF where eligibility for a modulator is recorded in the registry based on genotype.

Figure 2.21 shows that on 31st December 2024, 13.0% of females and 11.3% of males were not eligible for a CFTR modulator. This is a significant decrease compared to 2022, when 39% of females and 41% of males were not eligible.

FIGURE 2.21: ACFDR PAEDIATRICS 2024: ELIGIBILITY AND PRESCRIBING OF CFTR MODULATORS BY GENDER



N = 1,510 individuals with known eligibility

Of the paediatric population, 72.8% of females and 75.2% of males were eligible and prescribed a modulator, whereas 14.2% of females and 13.5% of males were eligible but not prescribed a modulator.

The following CFTR modulators were available to pwCF meeting the defined eligibility criteria.

Ivacaftor (KALYDECO®)

Ivacaftor is available on the PBS for pwCF, who are aged 4 months and older, and who have at least one mutation in the CFTR gene that is responsive to Ivacaftor potentiation based on clinical and/or in vitro data. Further information on Ivacaftor eligibility can be found via the Therapeutic Goods Administration Kalydeco Product Information sheet.

Lumacaftor/Ivacaftor (ORKAMBI®)

Lumacaftor/Ivacaftor is a combination therapy available on the PBS for pwCF, are aged one year and older, and 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, are aged 12 years and older, and who have one copy of the following changes in the CFTR gene: E56K, R117C, F508del, S977F, F1074L, 3849+10kbC→T, P67L, E193K, D579G, F1052V, D1152H, R74W, L206W, 711+3A → G, K1060T, D1270N, D110E, R352Q, E831X, A1067T, 2789+5G → A, D110H, A455E, S945L, R1070W, 3272-26A → G.

Elexacaftor/Tezacaftor/Ivacaftor (TRIKAFTA®)

Elexacaftor/tezacaftor/ivacaftor (ETI) is a triple combination therapy available on the PBS in for pwCF aged 2 years and older, with at least one copy of the F508del gene change in the CFTR gene. Trikafta was initially available on the PBS in April 2022 for pwCF aged 12 and older. In May 2023 pwCF aged 6-11 were eligible to receive Trikafta, and in August 2024 pwCF aged 2-5 were able to access the treatment.

Among children with cystic fibrosis, uptake of CFTR modulators – particularly Trikafta (ETI) – increases markedly with age. In the youngest group (<6 years), less than half (44.2%) were receiving Trikafta, and less than one quarter (22.0%) were not eligible for any CFTR modulator therapy, reflecting historical regulatory and funding restrictions for this age range. As children enter higher age bands where eligibility criteria broaden, Trikafta use becomes dominant: 73.7% 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 (3.5%-17.9%), and between 10.5% and 15.9% were eligible but not receiving treatment, suggesting potential delays in access or clinical considerations despite eligibility (Table 2.7).

Due to the majority of patients accessing Trikafta in 2024, the following modulator data has been categorised as 'Trikafta' or 'other CFTR modulator'.

TABLE 2.7: ACFDR PAEDIATRICS 2024: 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	414	183 (44.2%)	74 (17.9%)	66 (15.9%)	91 (22.0%)
6-11	517	380 (73.5%)	32 (6.2%)	81 (15.7%)	24 (4.6%)
12-17	579	474 (81.9%)	20 (3.5%)	61 (10.5%)	24 (4.1%)

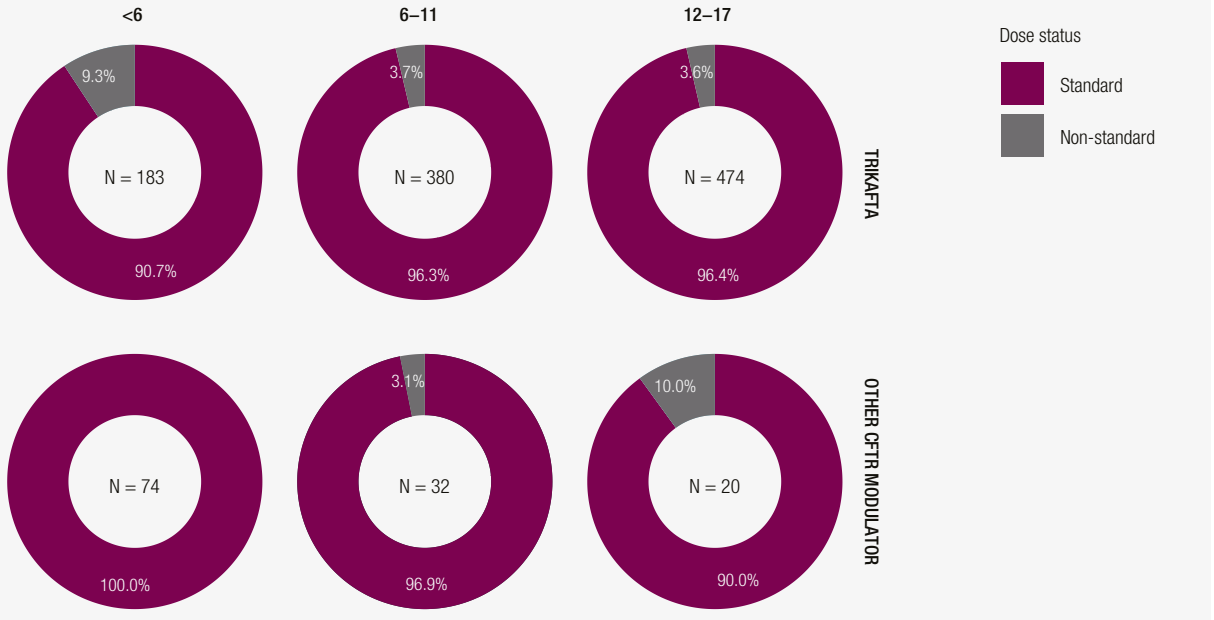
Table 2.8 describes the **reasons for permanent or temporary cessation and/or change to modulator prescriptions** throughout 2024. **Switching from a modulator other than Trikafta to another modulator was the most common reason for discontinuation** (76 pwCF). A smaller percentage of cessations were due to adverse effects, primarily liver impairment or intolerance (16 pwCF).

TABLE 2.8: ACFDR PAEDIATRICS 2024: REASON FOR DISCONTINUATION/CHANGE OF MODULATOR

Reasons for discontinuation/change	Total N	Trikafta N (%)	Other CFTR modulator N (%)
Switch to other CFTR modulator	77	<5	76 (56.3%)
Other reason (non-adverse)	64	10 (22.7%)	54 (40.0%)
Liver impairment/intolerance	16	13 (29.5%)	<5
Other intolerance/adverse event	15	13 (29.5%)	<5
Mental health	<5	<5	0 (0.0%)
Rash	<5	<5	0 (0.0%)
Pulmonary side effect/intolerance	<5	<5	0 (0.0%)
Concomitant drug interaction	0	0 (0.0%)	0 (0.0%)

In 2024, **more than 90% of children and adolescents on modulators were on a standard dose**. Figure 2.22 shows the proportion of patients aged <6, 6-11, and 12-17 years on standard and non-standard doses of Trikafta and other CFTR modulators. Less than 10% of all children and adolescents on Trikafta were on non-standard doses, and 10.0% of adolescents aged 12 and older on other CFTR modulators were on non-standard doses.

FIGURE 2.22: ACFDR PAEDIATRICS 2024: STANDARD AND NON-STANDARD DOSE BY AGE AND MODULATOR



2.4 COMPLICATIONS AND THERAPIES

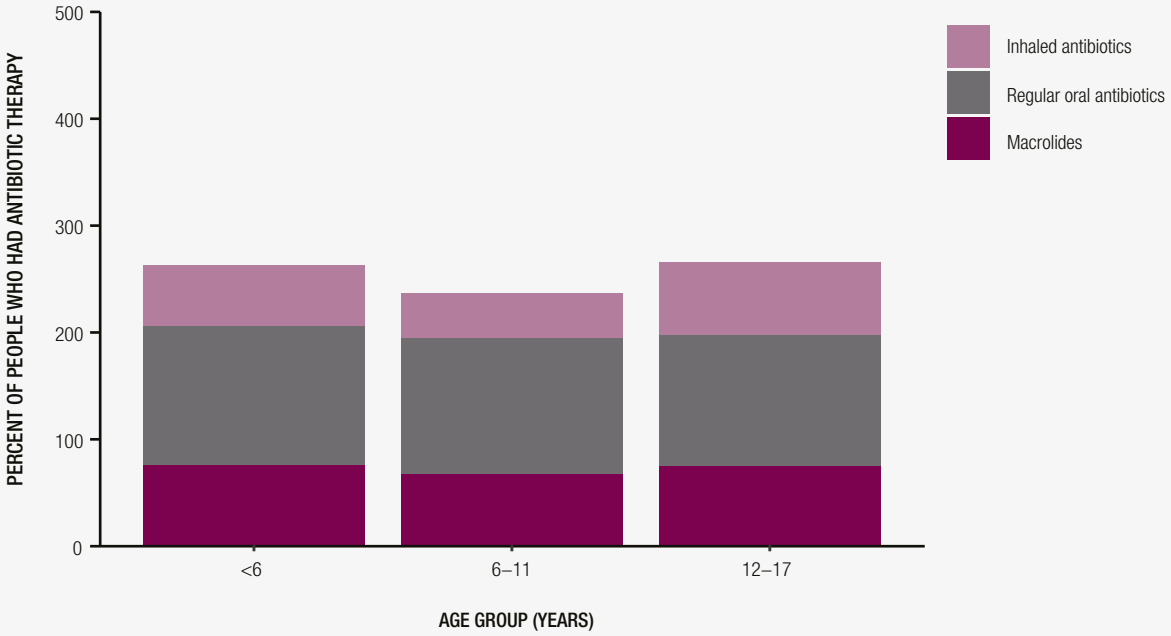
CF Pulmonary Disease

In 2024 there were 13 paediatric hospitalisations for haemoptysis, of which 2 required embolisation. One adolescent required hospitalisation for pneumothorax in 2024.

CF Pulmonary Therapies — Maintenance Antibiotics

The use of maintenance antibiotic therapy for children and adolescents is depicted in figures 2.23 and 2.24. In 2024, inhaled antibiotics were more commonly used by younger children, with 57 (13.6%) children under 6, 42 (8.1%) children 7-11 and 69 (11.8%) adolescents aged 12-17 being administered inhaled antibiotics. Use of regular oral antibiotics decreased with increasing age, with 131 (31.3%) children under 6, 129 (24.8%) children 7-11, and 123 (21.0%) adolescents aged 12-17 being prescribed regular oral antibiotics. Macrolide use was similar, with 76 (18.2%) children under 6 years, 67 (12.9%) children 7-11, and 75 (12.8%) 12-17-year-olds using macrolides in 2024.

FIGURE 2.23: ACFDR PAEDIATRICS 2024: MAINTENANCE ANTIBIOTIC THERAPY BY AGE GROUP



Given that the majority of pwCF are now receiving modulator treatments, some adjuvant antibiotic treatment use has reduced over time. This new figure **shows usage of regular oral antibiotics, macrolides and inhaled antibiotics over a five-year period**. Regular oral antibiotics use has decreased for all paediatric age groups during this time (Figures 2.24A, B, and C). Macrolide use has increased in children <6 years, remained stable in children 6-11 years, and decreased in adolescents. Similarly, use of inhaled antibiotics has remained stable for <6 year olds, but decreased in older children and adolescents.

FIGURE 2.24A: ACFDR PAEDIATRICS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN UNDER 6 YEARS

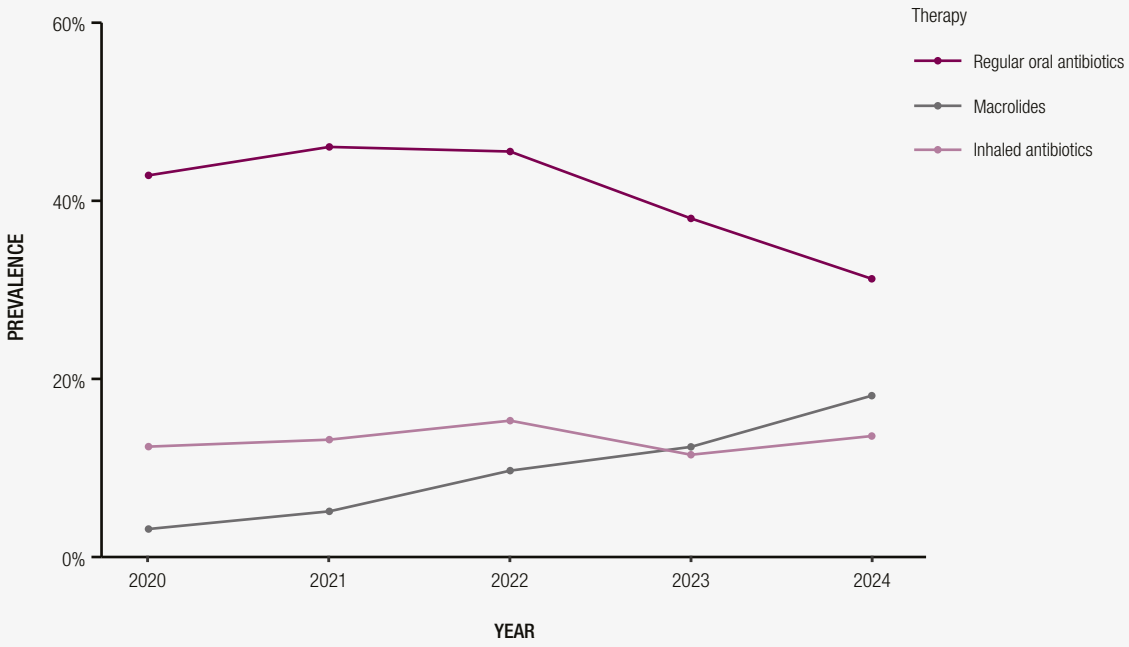


FIGURE 2.24B: ACFDR PAEDIATRICS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN AGED 6-11 YEARS

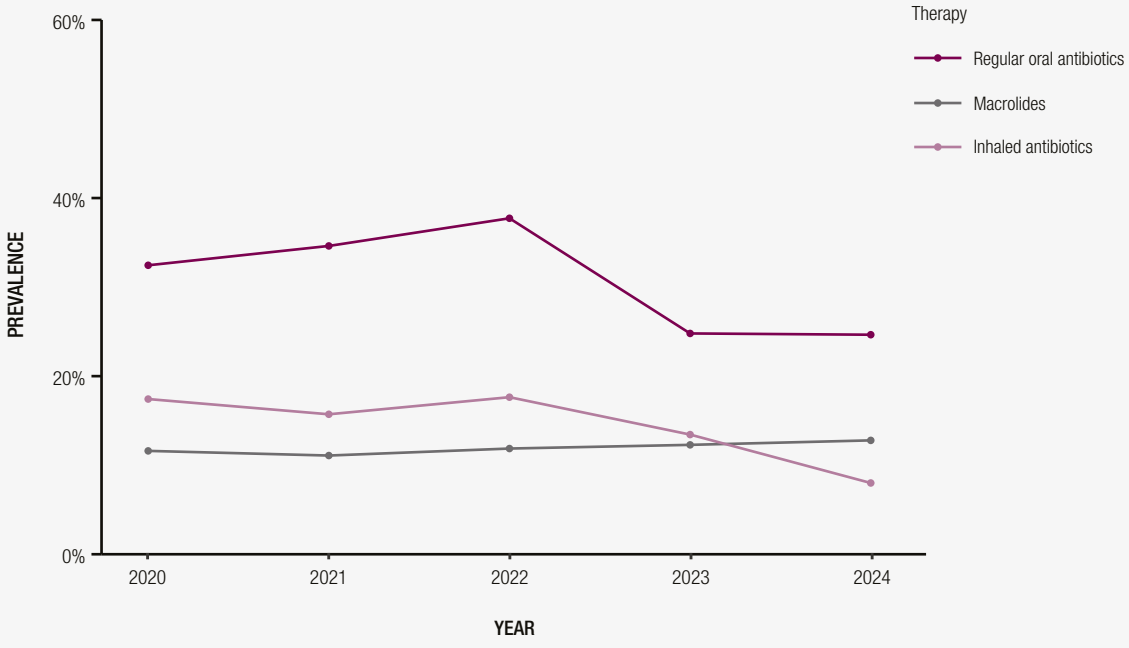
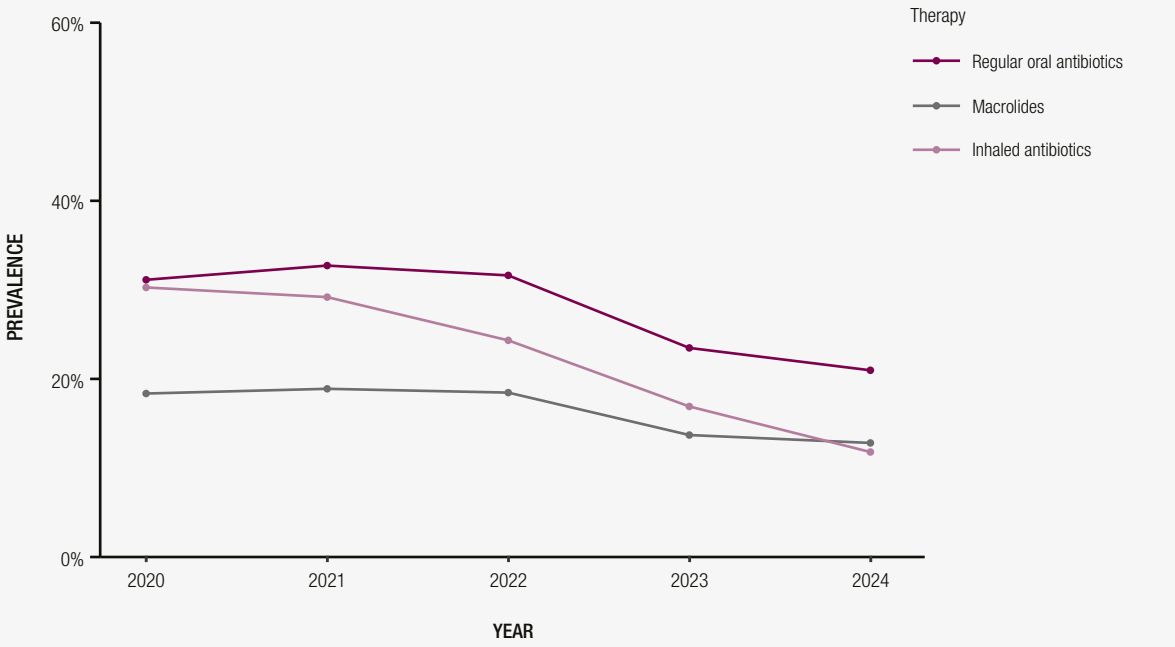


FIGURE 2.24C: ACFDR PAEDIATRICS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN AGED 12-17 YEARS

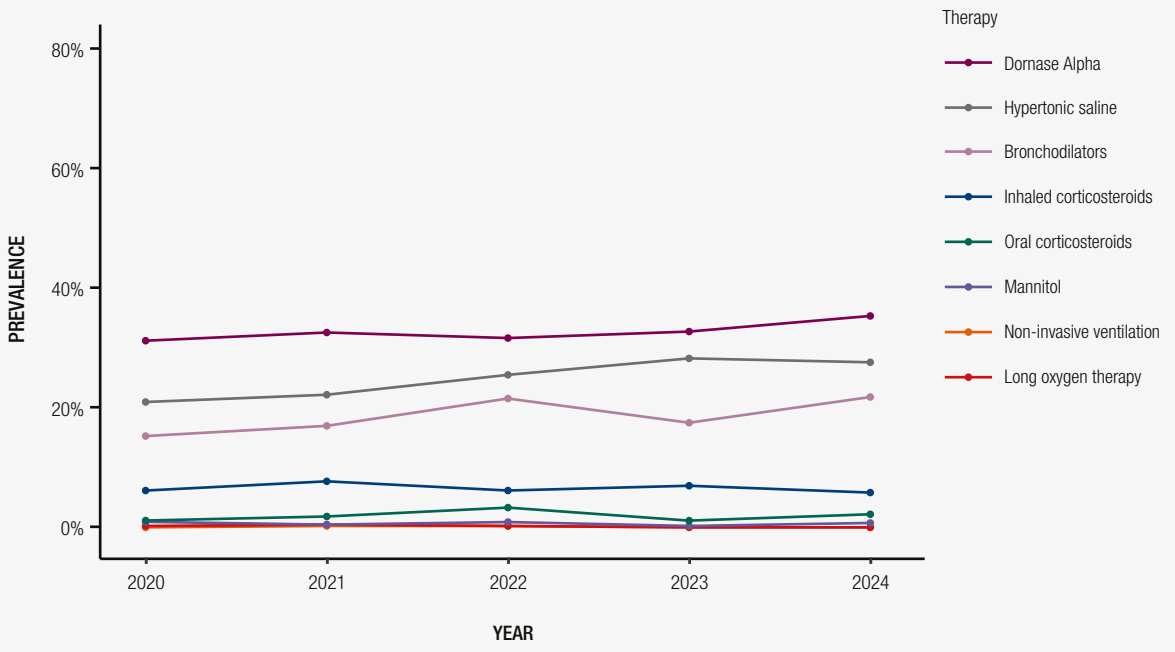


CF Lung Therapies — Non-Antibiotic Management

In 2024, the most used adjuvant lung therapies among children and adolescents, regardless of age, were dornase alpha, followed by hypertonic saline and bronchodilators, with use higher amongst the 6-11 and 12-17 age groups. Inhaled corticosteroid use increased with age, as did inhaled mannitol use. Oral corticosteroids were less commonly used, by approximately 2-4% of children and adolescents across the age groups (Figures 2.25A, B, and C). These new figures show trends over time in the use of adjuvant pulmonary therapies in the three paediatric age groups.

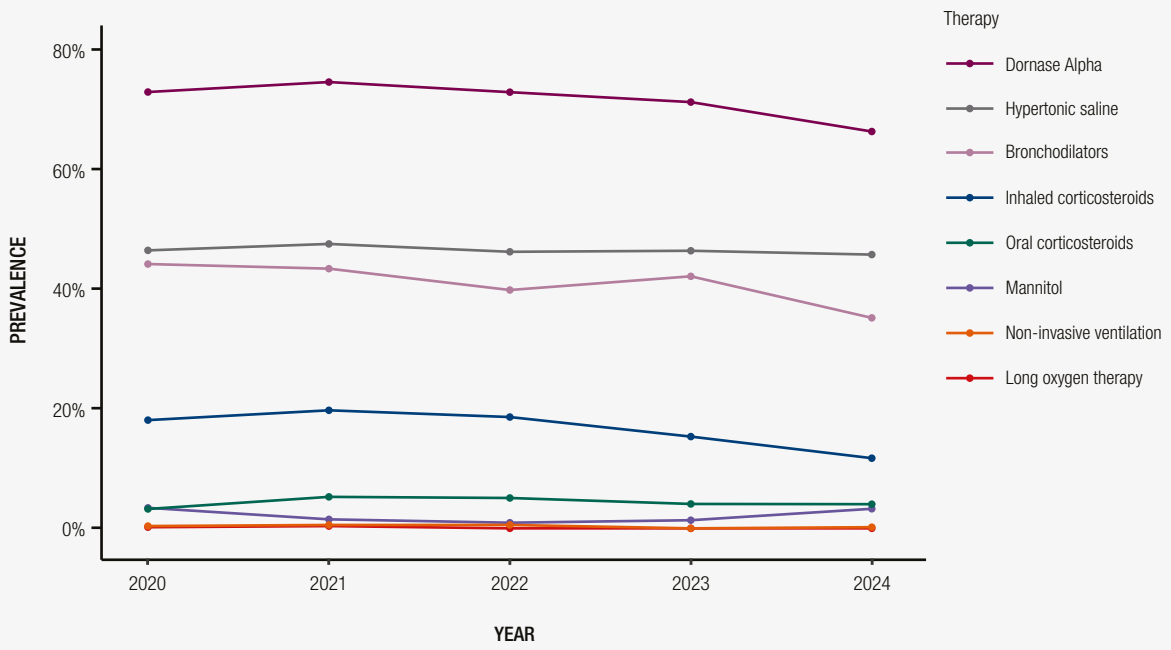
For children <6 years of age, use of dornase alpha, hypertonic saline and bronchodilators have generally increased during the last five years. In 2024, the proportions of children <6 years using these were 35.3%, 27.5% and 21.7% respectively. However, usage of inhaled and oral corticosteroids, non-invasive ventilation, long-term oxygen therapy and mannitol remained less than 10% and stable.

FIGURE 2.25A: ACFDR PAEDIATRICS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN UNDER 6 YEARS



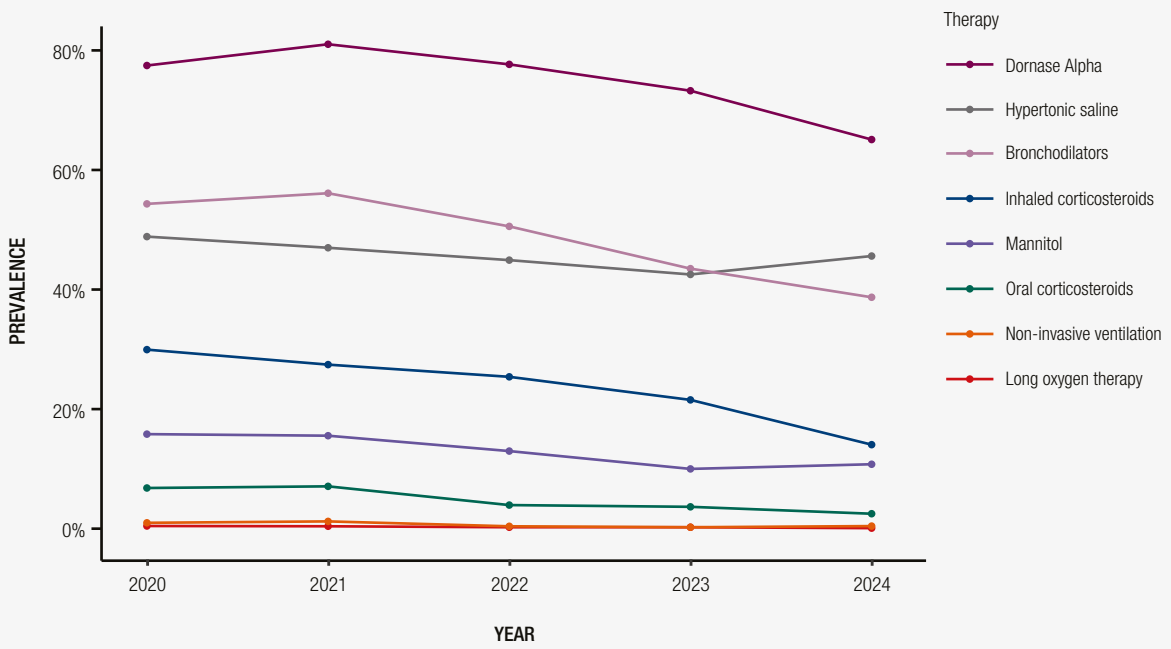
For older children (6-11 years), use of dornase alpha (DA), bronchodilators and inhaled corticosteroids decreased over the 5-year period, with 66.6% using DA and 35.3% using bronchodilators and 11.8% using inhaled corticosteroids in 2024. Other therapies remained fairly stable during this period. (Figure 2.25A).

FIGURE 2.25B: ACFDR PAEDIATRICS 2020-2024: 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 2024 for this age group were 65.1% for dornase alpha, 45.6% for hypertonic saline, 38.7% for bronchodilators, and 10.8% for mannitol. The remainder were used by less than 20% of this age group.

FIGURE 2.25C: ACFDR PAEDIATRICS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN AGED 12-17 YEARS



CF Diabetes

CF-related diabetes is a common complication in adults with CF, but may manifest in older children. Screening for diabetes and impaired glucose tolerance is recommended for pwCF aged 10 years and older. These new figures show incidence of impaired glucose tolerance and diabetes over the last five years.

The incidence of impaired glucose tolerance reported in 2024 was very low **for young children <6 years**, with 0.3% reporting impaired glucose tolerance and 0.6% 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 2024, the proportion affected were 4.3% and 3.1% respectively.

For adolescents 12-17 years, there has been a more substantial decline in the prevalence of these conditions over time. Impaired glucose intolerance has decreased from 18.8% in 2020 to 14.5% in 2024, and diabetes has reduced from 21.8% to 11.7% respectively (Figures 2.26A, B, and C).

Approximately 90% of children with diabetes were treated with insulin. Most insulin use by children and adolescents was long term (chronic) use (Table 2.9).

FIGURE 2.26A: ACFDR PAEDIATRICS 2019-2024: DIABETIC STATUS FOR CHILDREN UNDER 6 YEARS

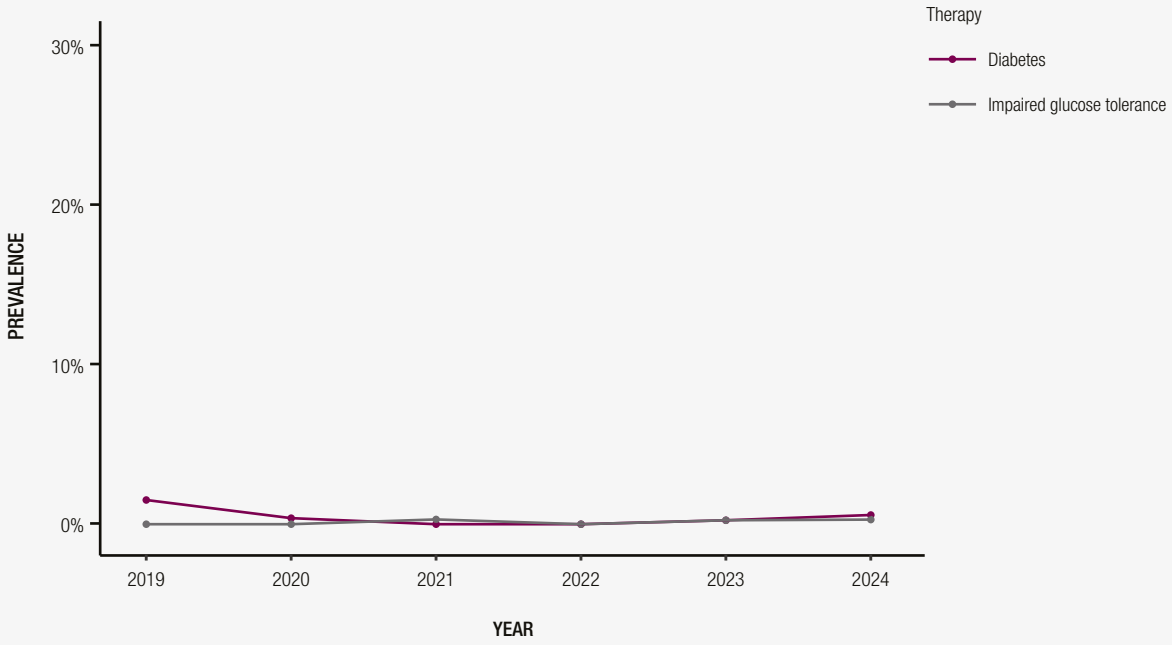


FIGURE 2.26B: ACFDR PAEDIATRICS 2019-2024: DIABETIC STATUS FOR CHILDREN AGED 6-11 YEARS

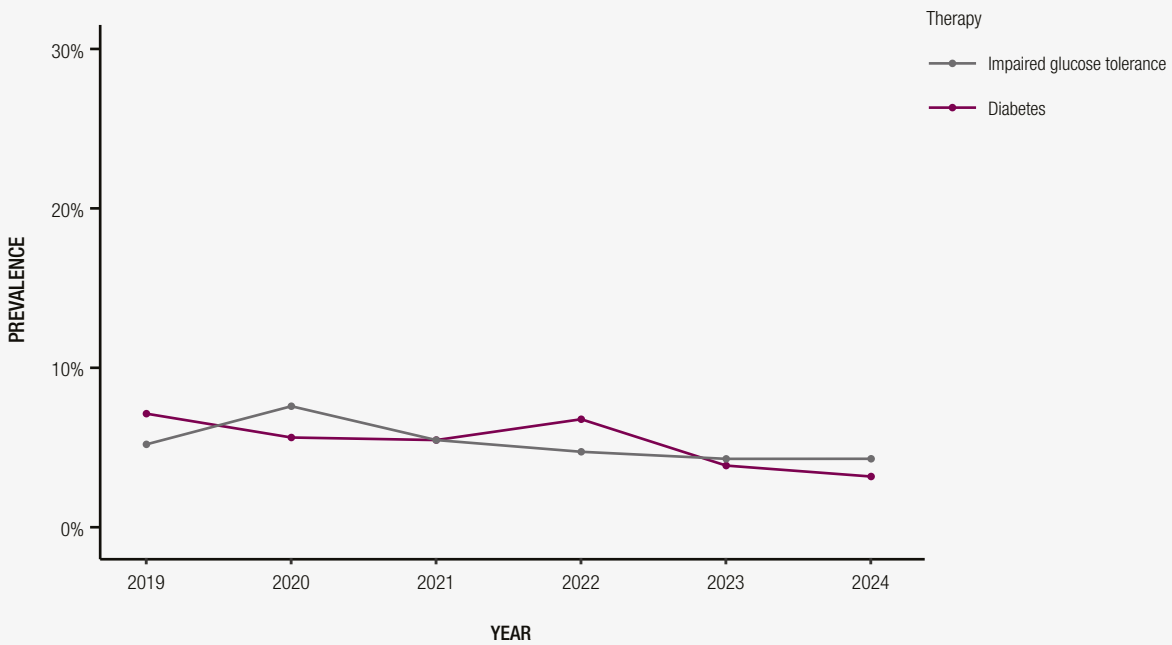


FIGURE 2.26C: ACFDR PAEDIATRICS 2019-2024: DIABETIC STATUS FOR CHILDREN AGED 12-17 YEARS

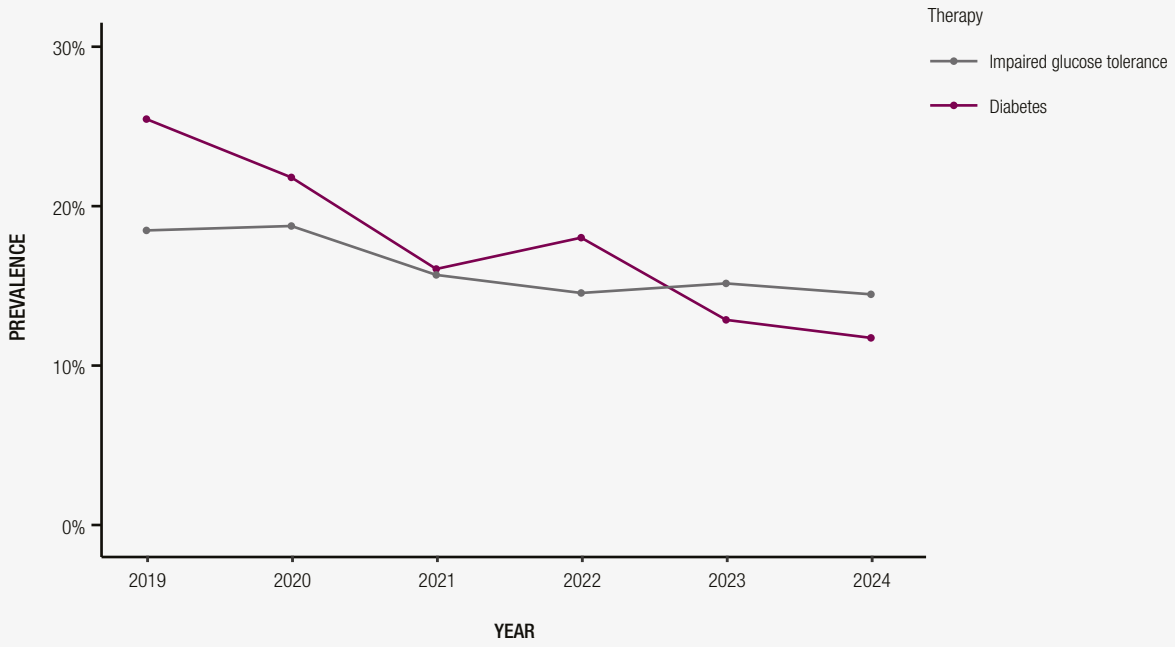


TABLE 2.9: ACFDR PAEDIATRICS 2024: DIABETIC TREATMENT BY AGE GROUP

Diabetes treatment type	<12 (N = 16)	12-17 (N = 60)
Insulin	14 (87.5%)	55 (91.7%)
Hypoglycaemics	0 (0.0%)	0 (0.0%)
Insulin and hypoglycaemics	0 (0.0%)	<5
Diet/lifestyle management only	<5	<5
No treatment for diabetes	0 (0.0%)	<5
Insulin use	**<12 (N = 14)**	**12-17 (N = 57)**
No insulin use	0 (0.0%)	<5
Intermittent insulin use	0 (0.0%)	<5
Chronic insulin use	14 (100.0%)	49 (86.0%)
Insulin use, duration unknown	0 (0.0%)	6 (10.5%)

CF Gastrointestinal Disease

Gastrointestinal complications of CF are not common in children and adolescents. In 2024, the most common gastrointestinal complication among children was gastroesophageal reflux, at 8.0% of children aged 6 or younger, 7.1% of 6-11-year-olds, and 7.1% of 12-17 year-olds. Less than 5% of children recorded hepatic complications in 2024 (Table 2.10).

TABLE 2.10: ACFDR PAEDIATRICS 2024: STOMACH AND LIVER DISEASE

	<6	6-11	12-17
Gastroesophageal reflux	33 / 414 (8.0%)	37 / 518 (7.1%)	41 / 580 (7.1%)
Liver disease, non-cirrhosis (includes viral hepatitis, fatty liver)	8 / 395 (2.0%)	23 / 494 (4.7%)	27 / 543 (5.0%)
Liver disease, cirrhosis (image confirmed)	0 / 387 (0.0%)	6 / 477 (1.3%)	7 / 523 (1.3%)
Liver disease, cirrhosis with portal hypertension	0 / 387 (0.0%)	<5 / 474	9 / 525 (1.7%)

In 2024, pancreatitis was very uncommon among children, with <1% of children/teenagers reporting acute or recurrent pancreatitis (Table 2.11). However, 73.2% of children <6 years, 75.5% of children 6-11 years, and 77.3% of adolescents 12-17 years reported pancreatic insufficiency.

TABLE 2.11: ACFDR PAEDIATRICS 2024: PANCREATITIS BY AGE

Pancreatitis	<6 (N = 406)	6-11 (N = 507)	12-17 (N = 570)
Pancreatitis (acute/recurrent/unspecified)	<5	<5	5 (0.9%)
No history of pancreatitis	405 (99.8%)	506 (99.8%)	565 (99.1%)
Pancreatic Status	<6 (N = 414)	6-11 (N = 518)	12-17 (N = 581)
Insufficient	303 (73.2%)	391 (75.5%)	449 (77.3%)

Osteopenia and Bone Density

Bone mineral density scans are not routinely performed on children younger than 10 years unless clinically indicated. For adolescents older than 10 years who had their bone density status reported to the ACFDR in 2024, 34.6% had osteopenia, 2.5% had osteoporosis, and 2.5% of 10 to 17-year-olds reported a fracture (Table 2.12). Fracture status was recorded for 757 young pwCF in 2024, with fractures reported for 19 (2.5%) 10-17-year-olds.

TABLE 2.12: ACFDR PAEDIATRICS 2024: BONE DENSITY FOR ADOLESCENTS

Bone mineral density	10-17 (N = 81)
Normal	51 (63.0%)
Osteopenia	28 (34.6%)
Osteoporosis	<5

Nutritional Supplementation

Table 2.13 reveals the usage of pancreatic enzymes and nutritional supplements across age groups from 2019-2024. In 2024, for those under 6, 72.2% used pancreatic enzymes, 69.8% used vitamin supplements, and 60.1% used salt replacement therapy. Among 6 to 11-year-olds, 75.1% used pancreatic enzymes, 69.5% took fat-soluble vitamin supplements, and 58.3% used salt replacement therapy. Among 12 to 17-year-olds, 73.8% used pancreatic enzymes, 67.8% took vitamin supplements, and 52.8% used salt replacement therapy.

TABLE 2.13: ACFDR PAEDIATRICS 2019-2024: NUTRITIONAL SUPPLEMENTS BY AGE

	<6 (N = 414)	6-11 (N = 518)	12-17 (N = 581)
Pancreatic enzymes	299 (72.2%)	389 (75.1%)	429 (73.8%)
Vitamin supplements (Fat soluble vitamins A, D, E and K)	289 (69.8%)	360 (69.5%)	394 (67.8%)
Salt replacement therapy	249 (60.1%)	302 (58.3%)	307 (52.8%)

The use of nutritional support among children and adolescents in 2024 was low (Table 2.14). Five percent of children aged less than 6 years, 6% of children aged 6-11 and 4% of children aged 12 or greater respectively required oral supplements. Otherwise, nutritional support was uncommon.

TABLE 2.14: ACFDR PAEDIATRICS 2024: NUTRITIONAL SUPPORT BY AGE

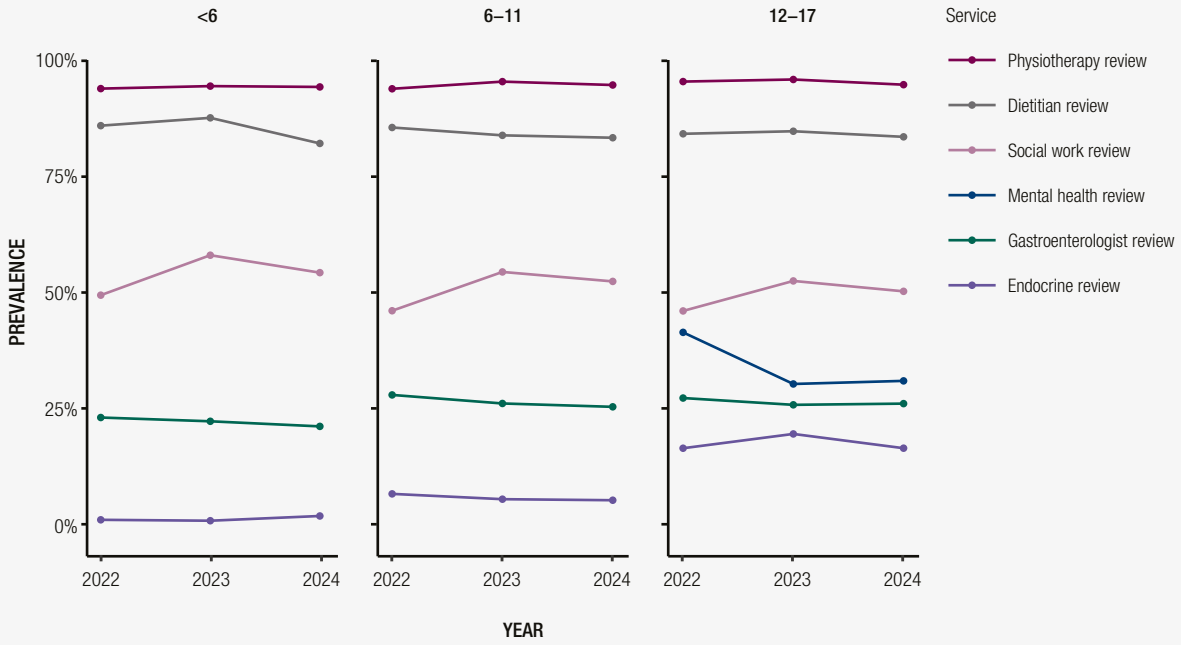
	<6 (N = 414)	6-11 (N = 518)	12-17 (N = 581)
Oral	22 (5.3%)	33 (6.4%)	24 (4.1%)
Gastrostomy tube	<5	27 (5.2%)	33 (5.7%)
Nasogastric tube	10 (2.4%)	<5	<5
Jejunostomy tube	0 (0.0%)	0 (0.0%)	0 (0.0%)
Parenteral nutrition	<5	<5	<5

Multidisciplinary Care

Multidisciplinary care is a cornerstone of effective, contemporary CF management. In 2024, a majority of children and adolescents participated in an annual physiotherapy appointments and dietician review (Figure 2.27). Approximately half of the children with CF had a social work review annually, approximately one quarter had a mental health screen or review (for children 12 years or older) or a gastroenterology review. Proportions who had reviews are generally similar for younger children and adolescents except for endocrine reviews, which were performed for over 15% of adolescents but only 4% of younger children.

From 2022 to 2024, there was minimal variation in the proportion of young pwCF who accessed physiotherapy, gastroenterology, and endocrine reviews. Social work reviews varied the most, peaking in 2023 for children under 6 (58.3%), 6-11 (54.4%), and 12-17 (52.4%) and decreasing slightly in 2024 to 54.5%, 52.4%, and 50.2% respectively. For adolescents aged 12-17, mental health reviews decreased from 41.3% in 2022 to 30.9% in 2024.

FIGURE 2.27: ACFDR PAEDIATRICS 2022-2024: ANNUAL MULTIDISCIPLINARY CARE APPOINTMENTS BY AGE GROUP

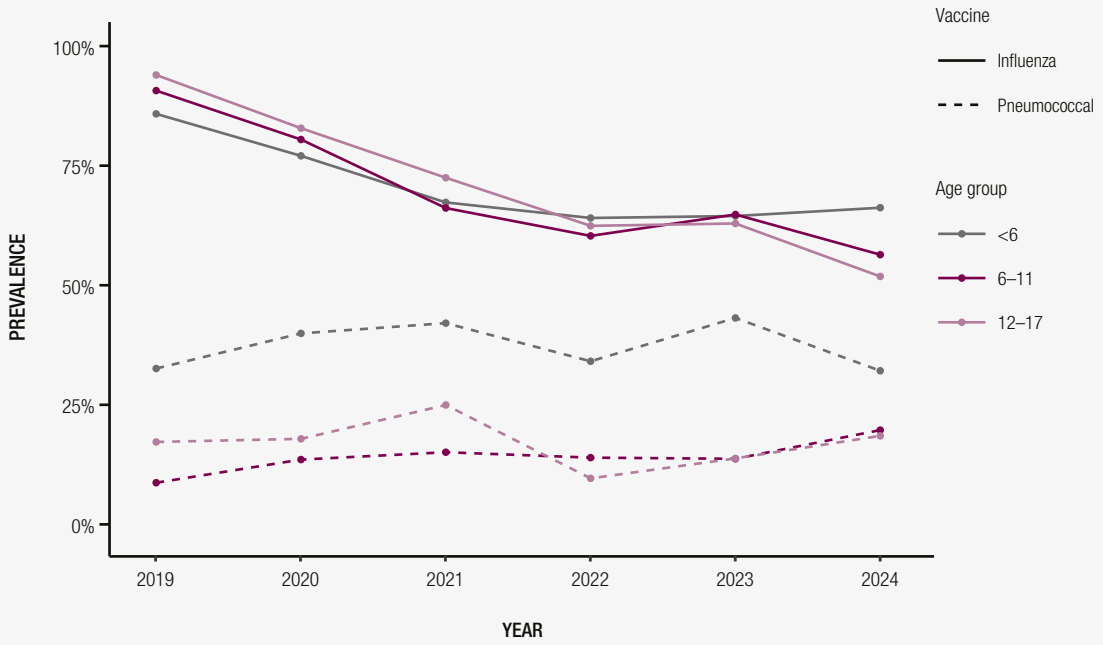


Denominator excludes Not known; cohort = patients active in each year. Mental health review not applicable to <12.

Vaccinations

Influenza immunisation is recommended for individuals with CF aged six months and older on an annual basis. In 2024, 66.2% of children aged <6, 56.4% of children aged 6-11 years, and 51.8% of children aged 12-17 years were recorded as being immunised against influenza, compared to 85.8% of children <6, 90.7% of children aged 6-11 years, and 94.0% of adolescents aged 12-17 in 2019 (Figure 2.28).

FIGURE 2.28: ACFDR PAEDIATRICS 2019-2024: VACCINATIONS BY AGE GROUP



Denominator excludes Not known/missing; cohort = patients active each year.

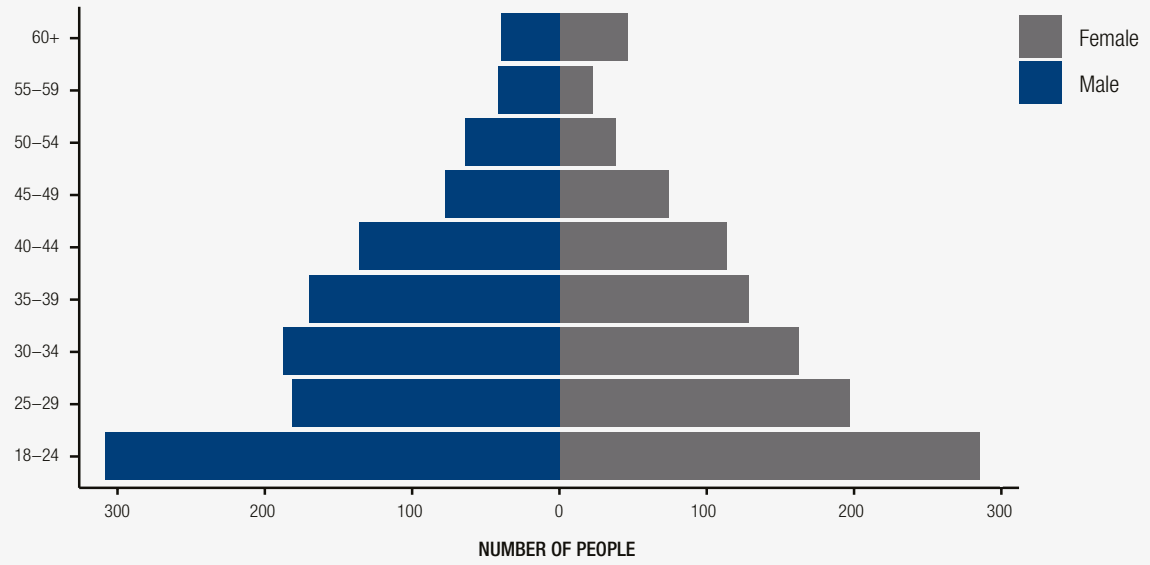
3.
ADULT DATA

3. ADULT DATA

3.1 ADULTS WITH CYSTIC FIBROSIS

As of 31st December 2024, the ACFDR held data regarding 2,274 adults with CF, of which 1,065 were female and 1,209 were male. The most common adult age group was 18-24 years for both females (16%) and males (16%), followed by 25-29 years for females (11%) and 30-34 years for males (9%). 16% of females and 18% 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 2024: AGE AND SEX



N = 2,274 people

Socioeconomic Characteristics

Educational Attainment

These **new figures** show educational and employment attainment over the last five years.

Of the 1,560 (68.6%) adults with CF for whom education information was available in the ACFDR in 2024 the most common educational attainment was senior secondary school (37%), followed by a University/Bachelor degree (29%), and a Tertiary certificate/diploma (23%). In 2024, 10% of adults nominated junior secondary school as their highest educational attainment, a decrease from 13% in 2020 (Figure 3.2).

Table 3.1 compares the level of educational attainment of pwCF to the broader Australian Population (Australian Bureau Statistics (ABS) data as of May 2024). Educational attainment for pwCF has been increasing in recent years, and in 2024, **26% of pwCF had completed a university bachelor degree**, which is higher than in the **general population** (26% compared to 21%) (Table 3.1).

FIGURE 3.2: ACFDR ADULTS 2020-2024: HIGHEST EDUCATIONAL ATTAINMENT

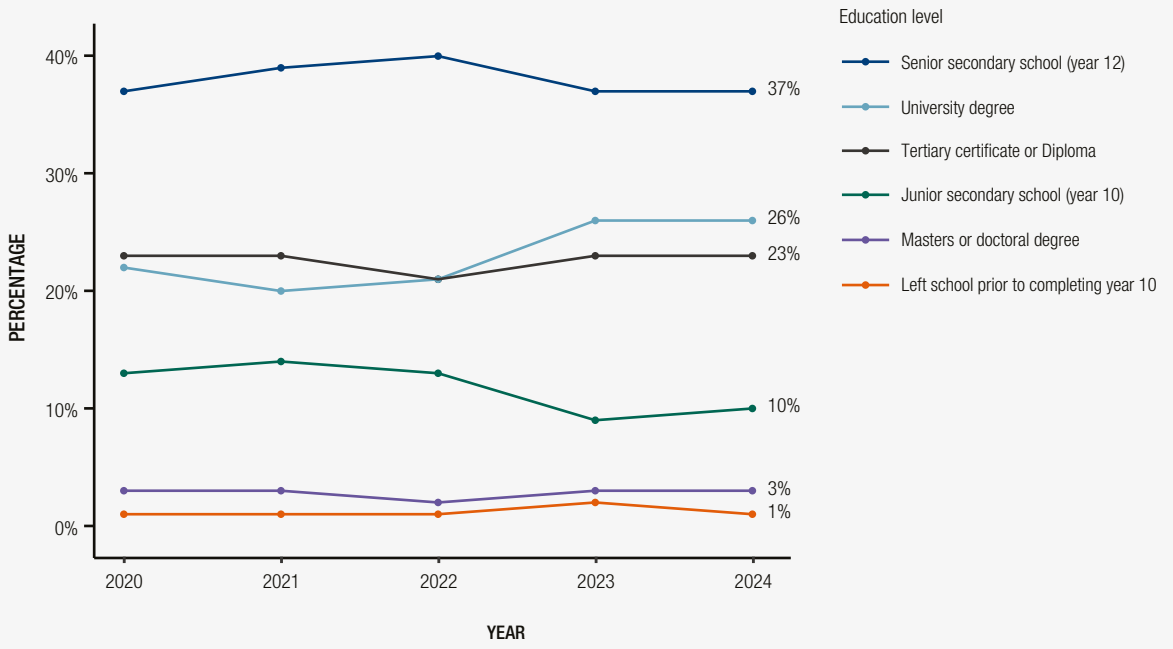


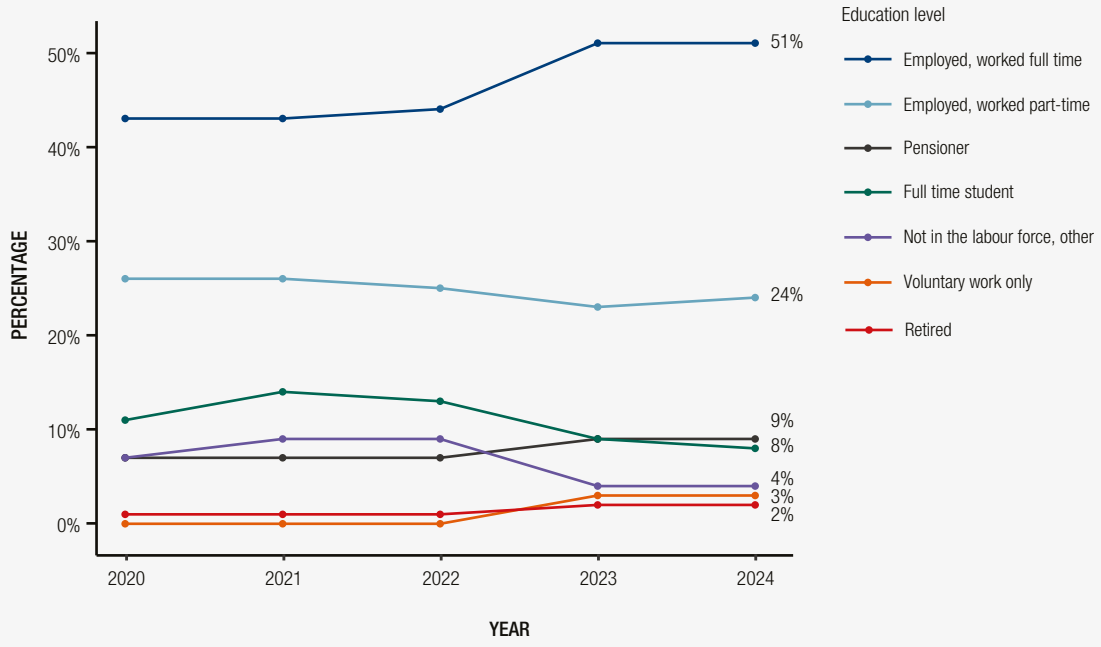
TABLE 3.1: ACFDR ADULTS 2024: HIGHEST EDUCATIONAL ATTAINMENT

Highest educational attainment	ACFDR (N = 1,593)	ABS Data (%), May 2024 for 15-74-year-olds
Masters or doctoral degree	51 (3%)	8.9%
University/Bachelor degree	407 (26%)	21.1%
Tertiary certificate or Diploma	366 (23%)	29.0%
Senior secondary school (year 12)	591 (37%)	18.5%
Junior secondary school (year 10)	155 (10%)	14.0%
Left school prior to completing year 10	23 (1%)	5.9%

Within the ACFDR dataset, information on employment status was available for 2,089 adults (90.5%) with CF. Among them, 51% were engaged in full-time employment, an increase from 43% in 2020.

Most other employment statuses remained stable or declined slightly over the last 5 years. In 2024, 24% were in part-time employment, 9% 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 4% in 2024. Adults engaged in voluntary work only, or who were retired accounted for less than 5% of all adults with CF (Figure 3.3).

FIGURE 3.3: ACFDR ADULTS 2020-2024: EMPLOYMENT STATUS



Marital Status

Information regarding marital status was available for 2,108 adults in the registry (92.7%). Of these, 54% of women and 50% of men were either married or in a de facto relationship (Figure 3.4).

FIGURE 3.4: ACFDR ADULTS 2024: MARITAL STATUS

Pregnancies for People with CF

From this point in the clinical data, adults with lung transplants (n = 105) have been excluded from the analysis.

In 2024, there were 85 pregnancies to women with CF, similar to the 88 recorded in 2023 (Figure 3.5). There were 48 (56.5%) live births and 27 (31.8%) people still pregnant as of the 31st December 2024 (Table 3.2). as of 31 December 2023 (Table 3.2).

FIGURE 3.5: ACFDR ADULTS 2018- 2024: PREGNANCY

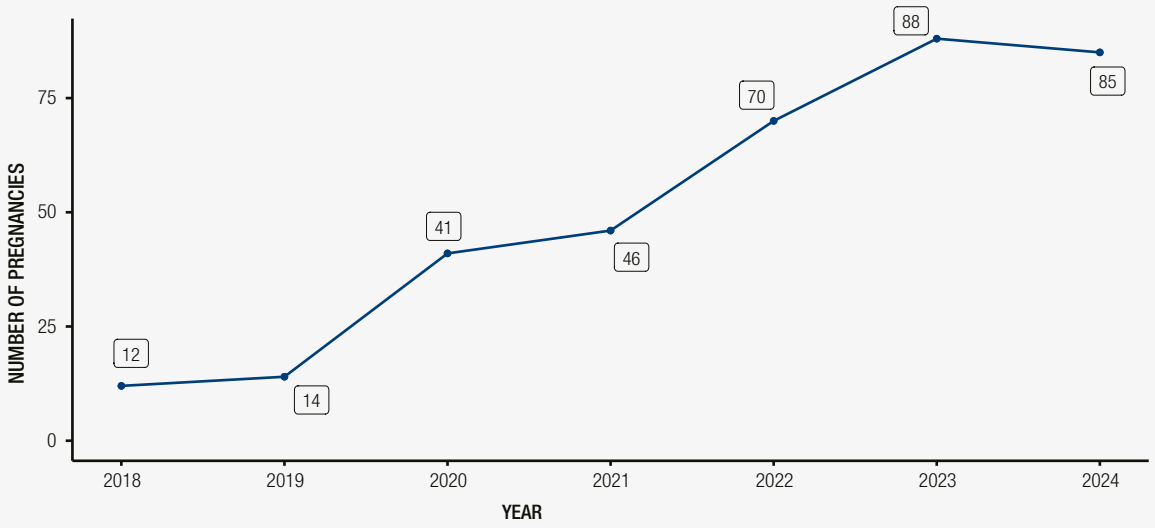


TABLE 3.2: ACFDR ADULTS 2024: PREGNANCY STATUS

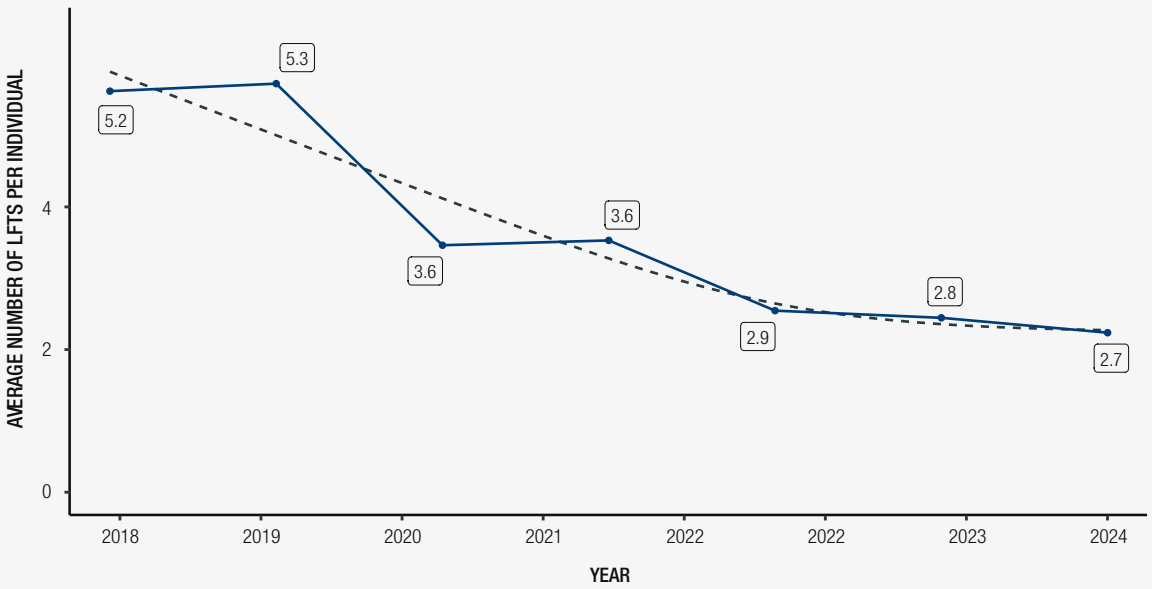
Status	≥18 (N = 85)
Currently pregnant	27 (31.8%)
Live birth	48 (56.5%)
Other (Miscarriage, stillbirth or termination)	10 (11.8%)

3.2 CLINICAL MEASURES

Lung Function

Figure 3.6 depicts the annual average number of lung function (spirometry) tests per adult recorded in the registry from 2018 to 2024. In 2018, adults averaged 5.2 tests each, however data from 2024 shows there was an average of 2.7 tests per adult.

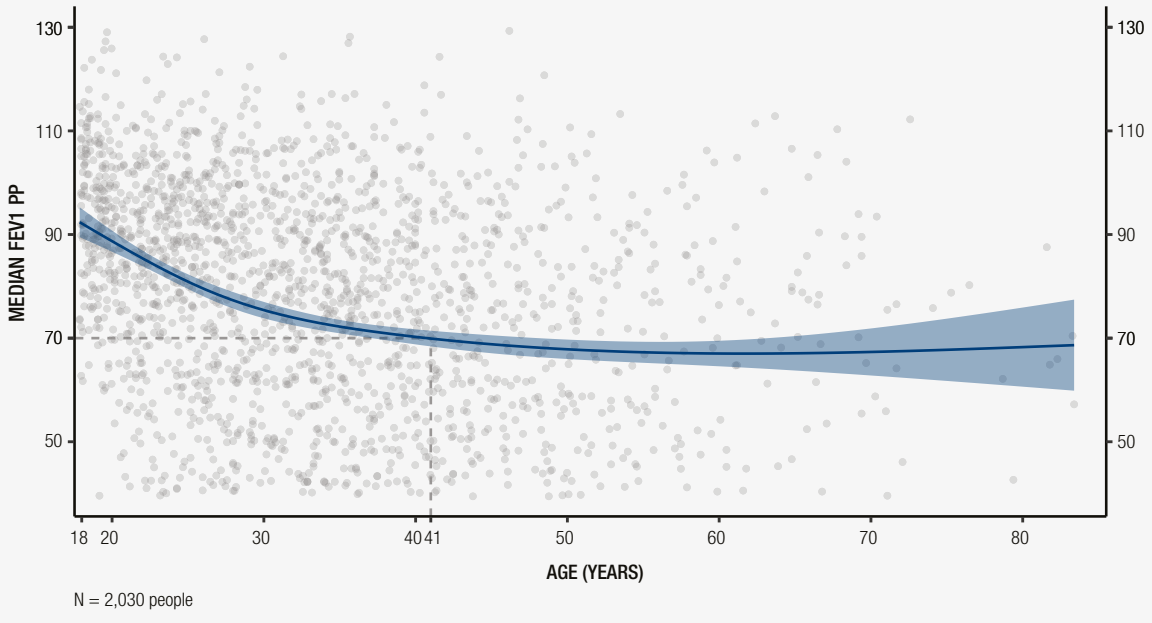
FIGURE 3.6: ACFDR ADULTS 2018-2024: AVERAGE NUMBER OF LUNG FUNCTION TESTS ENTERED PER INDIVIDUAL



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

There were 2,030 (89.3%) adults in 2024 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 3.7). The median age at which an FEV1 pp of 70.0 is recorded is 41 years in 2024, an increase from 36 years in 2022.

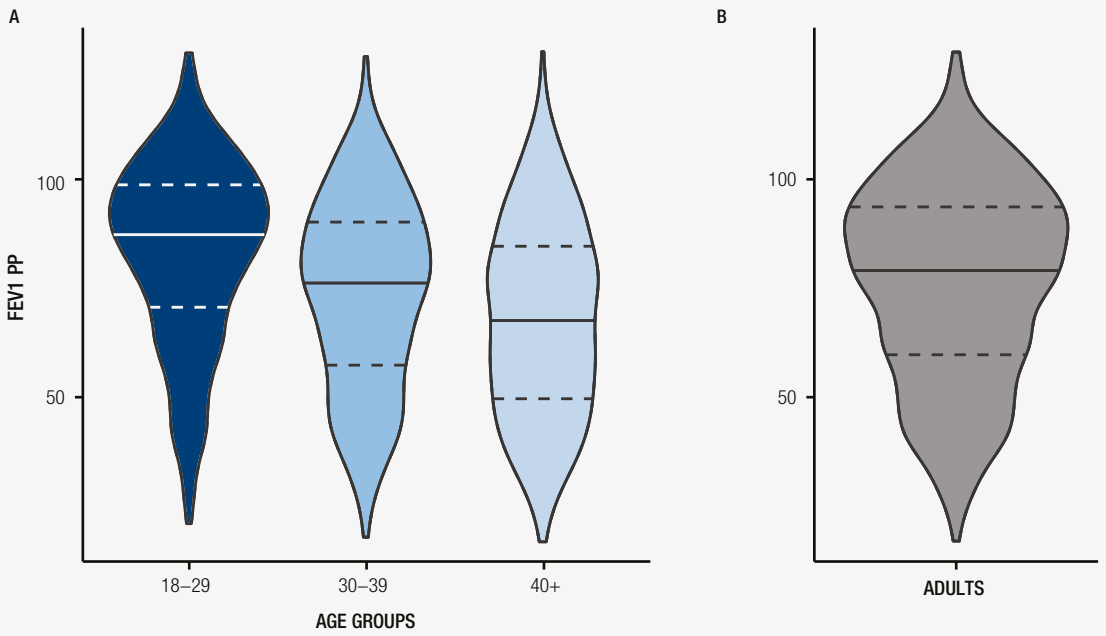
FIGURE 3.7: ACFDR ADULTS 2024: FEV1 PP BY AGE



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

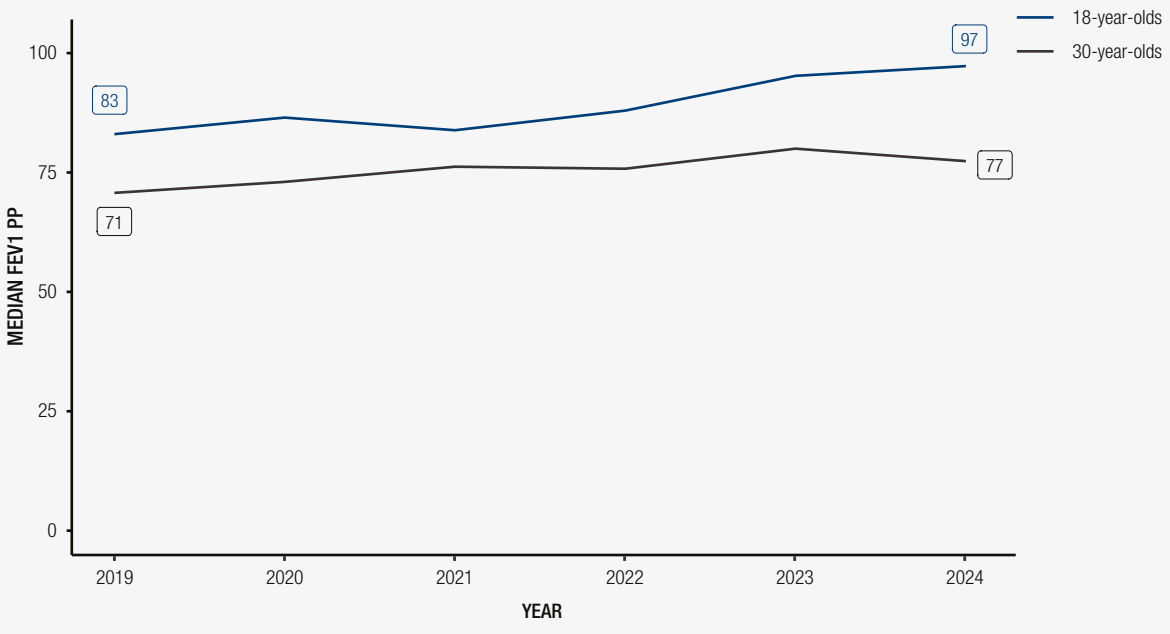
In 2024, the median FEV1 pp for adults overall was 79.0, with the median FEV1 pp for 18-29-year olds being 87.3, for 30-39-year olds being 76.2, and for 40+ year olds being 67.6 (Figure 3.8).

FIGURE 3.8: ACFDR ADULTS 2024: MEDIAN FEV1 PP



The median FEV1 pp has increased over time. For 18-year-olds, it has increased from 82.9 pp in 2019 to 97.2 pp in 2024, a 14.3% increase. For pwCF of age 30 years, the median FEV1 pp in 2024 was 77.3 pp, an increase of 6.6% from 70.7 in 2019 (Figure 3.9).

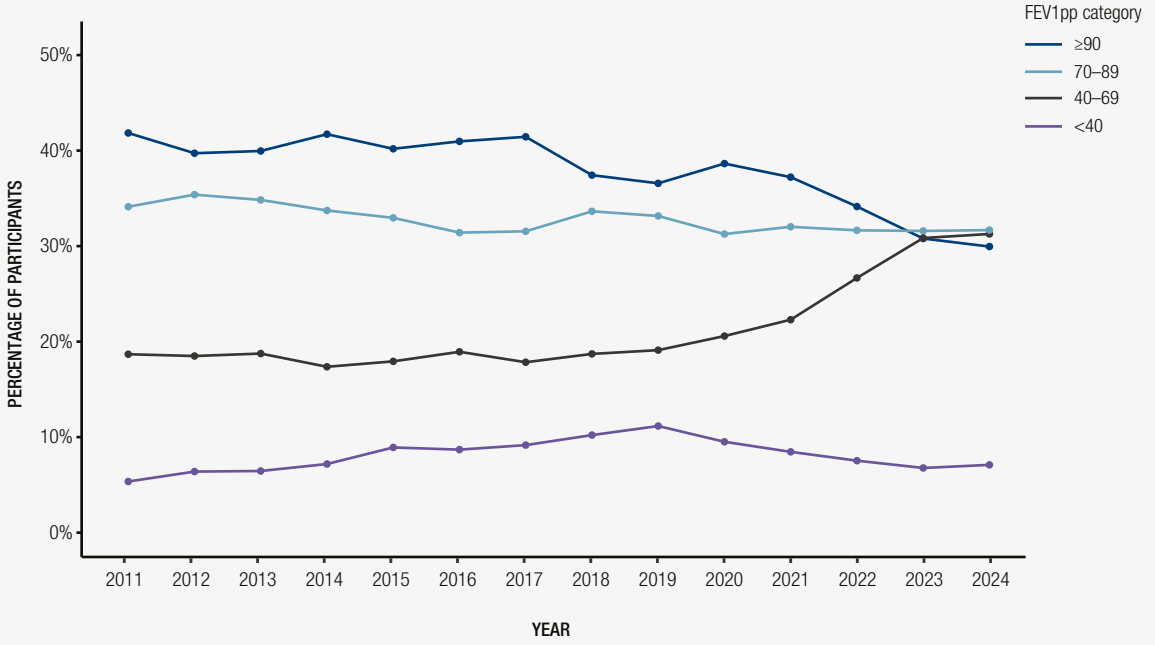
FIGURE 3.9: ACFDR ADULTS 2019-2024: MEDIAN FEV1 PP OVER TIME



Labelled percentages illustrate median predFEV1 in 2004 and 2024.

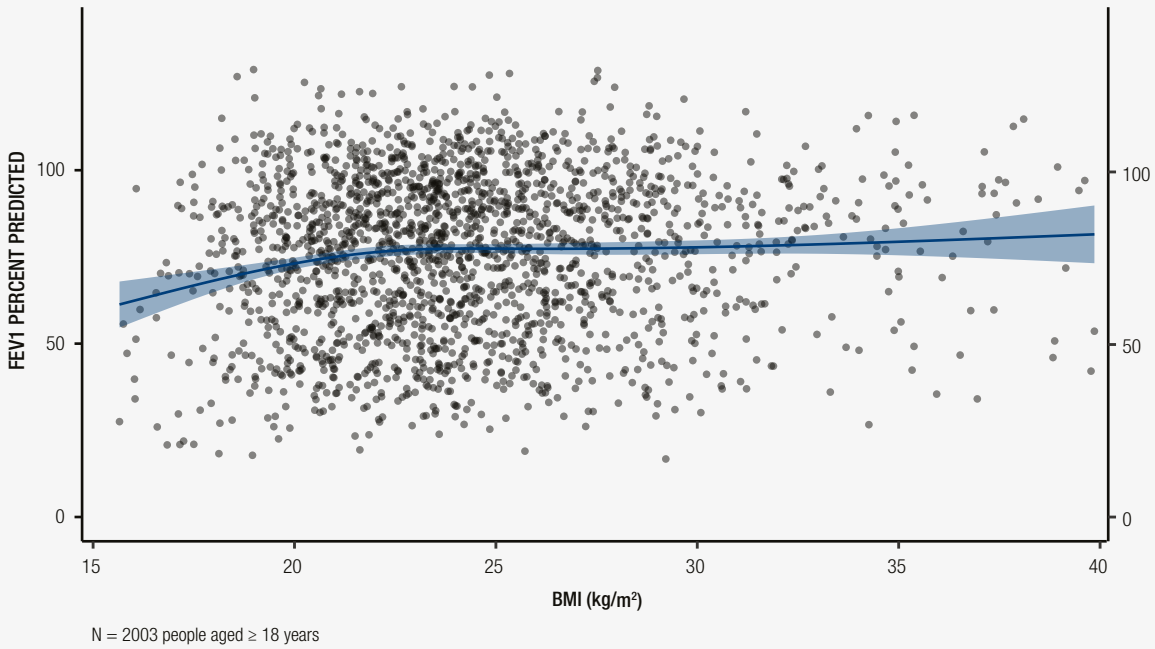
FEV1 pp has increased since 2011 (Figure 3.10). The proportion of adults with an FEV1 pp of >=90% has increased from 18.7% in 2011 to 31.3% in 2024. The proportion of adults with FEV1 pp of 70-89% has decreased from 34.1% in 2011 to 31.7% in 2024, and the proportion of adults with FEV1 pp of 40-69pp has decreased from 41.8% to 30.0%. FEV1 of <40pp has increased from 5.4% in 2011 to 7.1% in 2024.

FIGURE 3.10: ACFDR ADULTS 2011-2024: FEV1 PP DISTRIBUTION OVER TIME



For pwCF ages 18-40 years, FEV1 pp increased with increasing BMI, although, at BMIs into the high 20's, 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 3.11).

FIGURE 3.11: ACFDR ADULTS 2024 : FEV1 PP VS BMI PERCENTILE

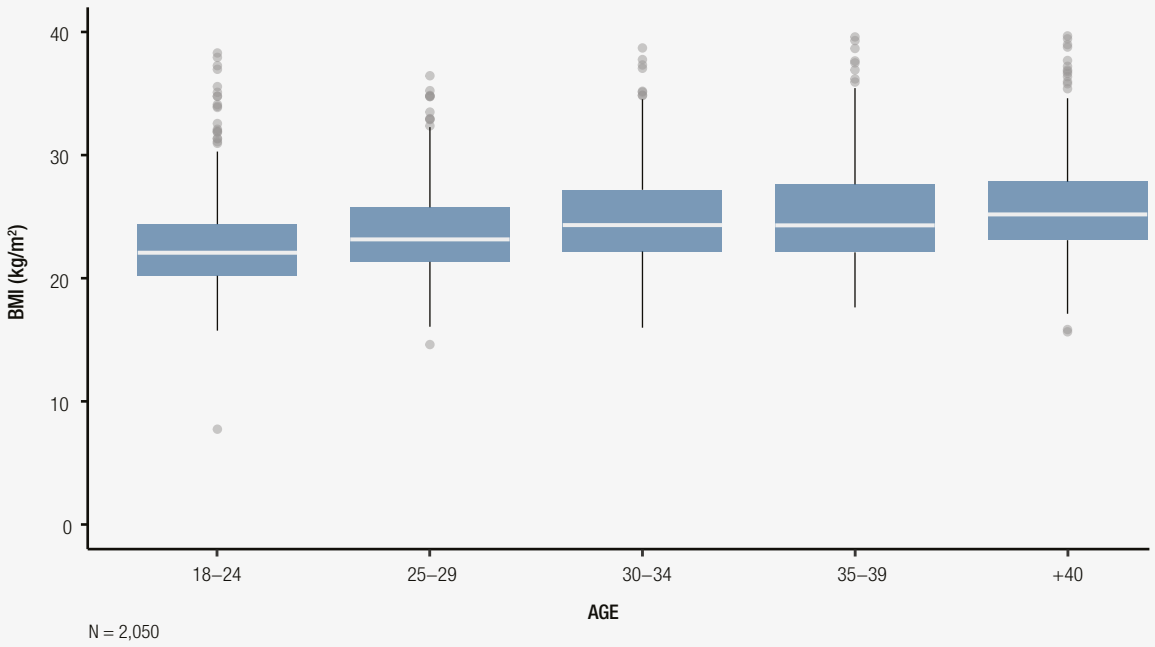


Solid line was calculated using a natural cubic spline with 3 degrees of freedom
Shaded area represents 95% confidence interval

Nutrition (Body Mass Index)

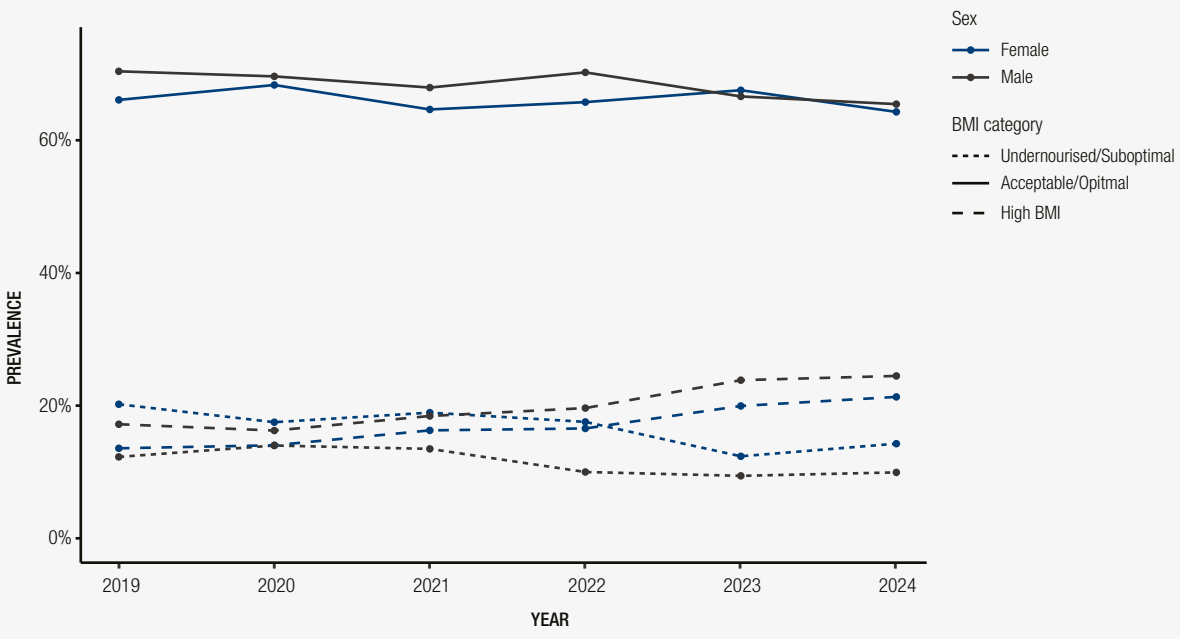
Body Mass Index (BMI) data for adults with CF increases with increasing age. For 2024, in the 18-24 age group, the median BMI was 22.1, for the 25-29 age group it was 23.2, for 30-34 age group it was 24.4 and for 35-39 age group it was 24.4. The highest median BMI was observed in the 40 and above age category, with a value of 25.3 (Figure 3.12).

FIGURE 3.12: ACFDR ADULTS 2024: BMI BY AGE



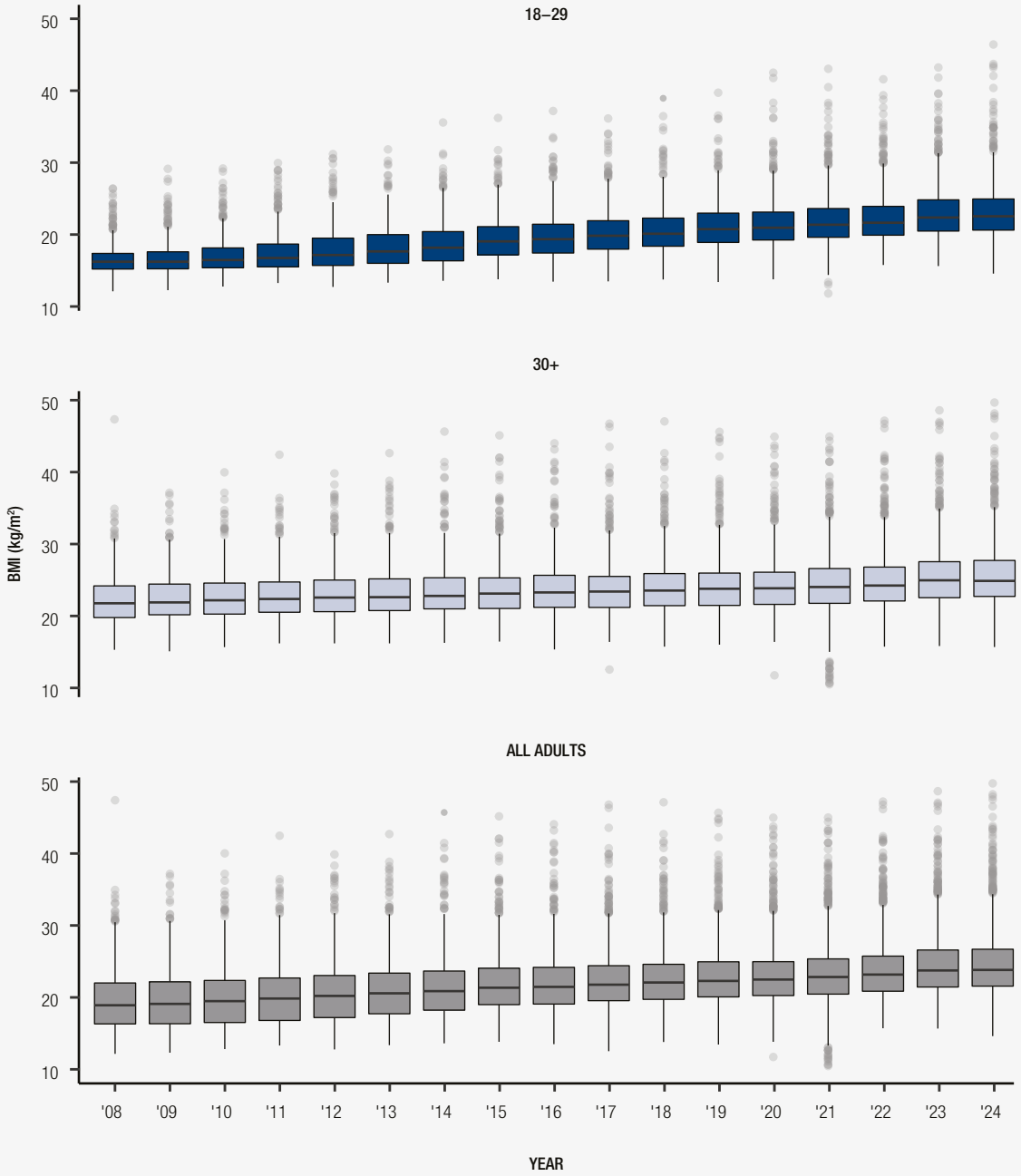
Approximately two-thirds of adults with CF had a BMI in the optimal/acceptable range recorded in 2024 (65.4% of males and 64.2% of females). 24.6% of males and 21.4% of females had a high BMI, while 14.4% of females and 10.1% of males had a suboptimal BMI (Figure 3.13).

FIGURE 3.13: ACFDR ADULTS 2024: 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 2008 to 22.6 in 2024. A similar pattern was observed in the 30-year age group, where the median BMI increased from 21.9 in 2008 to 24.9 in 2024. When considering all adults with CF, the overall median BMI has shown a continuous increase from 19.2 in 2008 to 23.8 in 2024 (Figure 3.14).

FIGURE 3.14: ACFDR ADULTS 2008-2024: MEDIAN BMI

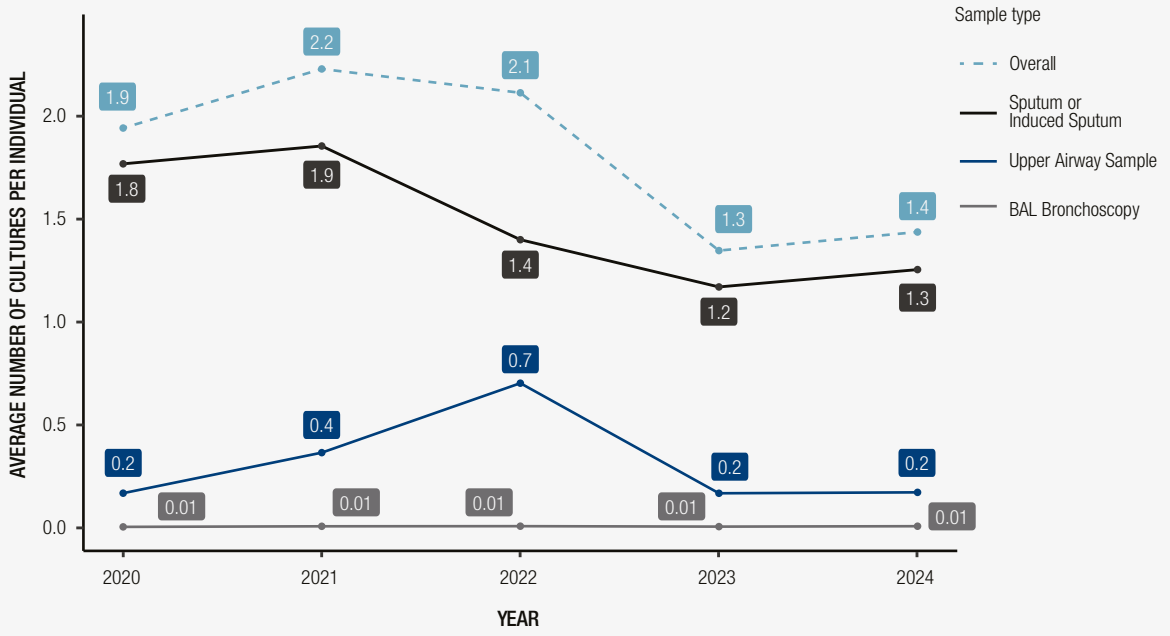


Microbiology

Microbiology Samples

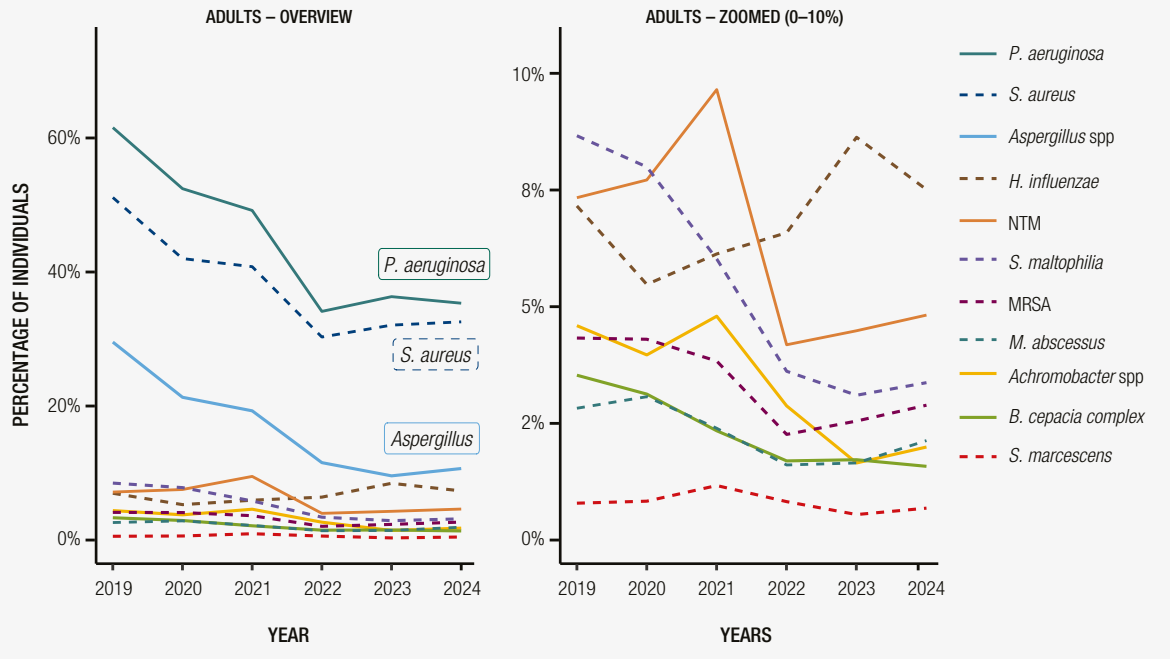
The average number of respiratory samples collected per adult from 2020 to 2024 is depicted in Figure 3.15. From 2020 to 2024 there was an increase in the overall number of respiratory samples per adult (peaking at 2.2 in 2021), however this has declined significantly in 2024 to an average of 1.4 samples. The most common respiratory sample types collected for adults are sputum/induced sputum, which has followed this trend. Upper airway samples also increased from 2020 to 2022, however declined from 2022 to 2024.

FIGURE 3.15: ACFDR ADULTS 2019-2024: AVERAGE NUMBER OF RESPIRATORY SAMPLES PER INDIVIDUAL



The prevalence of specific organisms commonly affecting pwCF has generally declined over the last six years (Figure 3.16). The prevalence of *P. aeruginosa* has decreased from 61.3% in 2019 to 35.3% by 2024. *S. aureus* has decreased from a peak of 50.9% in 2019 to 32.5% in 2024. The prevalence of *Aspergillus* spp has declined from 29.5% in 2019 to 10.8% in 2024. The prevalence of the less common microorganisms NTM, *Achromobacter* spp, *B. cepacia complex*, *S. maltophilia*, MRSA, and *M. abscessus* generally decreased, all with prevalence of less than 5% in 2024.

FIGURE 3.16: ACFDR ADULTS 2019-2024: PREVALENCE OF RESPIRATORY MICROBIOLOGY



Microbiology results for adults from 1,240 samples collected in 2024 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 20.7% among 18-24-year olds, increasing to a prevalence of 60.2% for 35 to 44-year-olds. Conversely, the prevalence of *S. aureus* decreased from 50.3% for 18 to 24-year-olds to 20.8% for pwCF at 45+ years of age (Figure 3.17 and Table 3.3). The prevalence of less common organisms is fairly consistent, at less than 5% across the age groups, with the exception of *H. influenzae*, which decreases from 15.1% for 18 to 24-year-olds to 2.9% for pwCF at 45 years of age.

FIGURE 3.17: ACFDR ADULTS 2024: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE

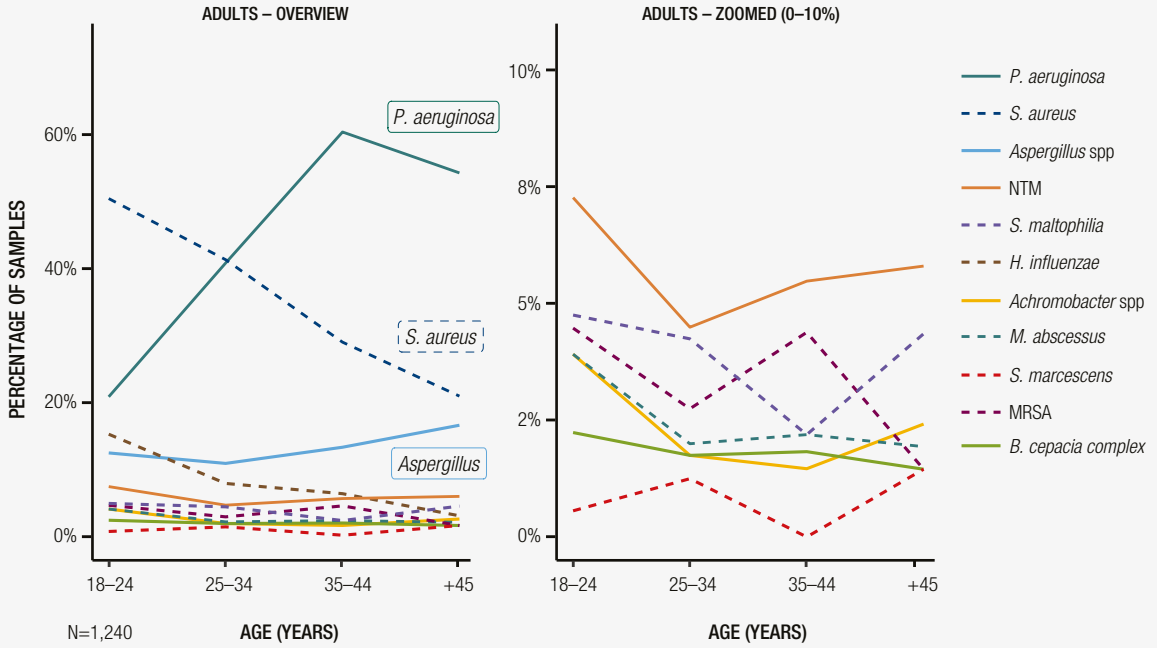


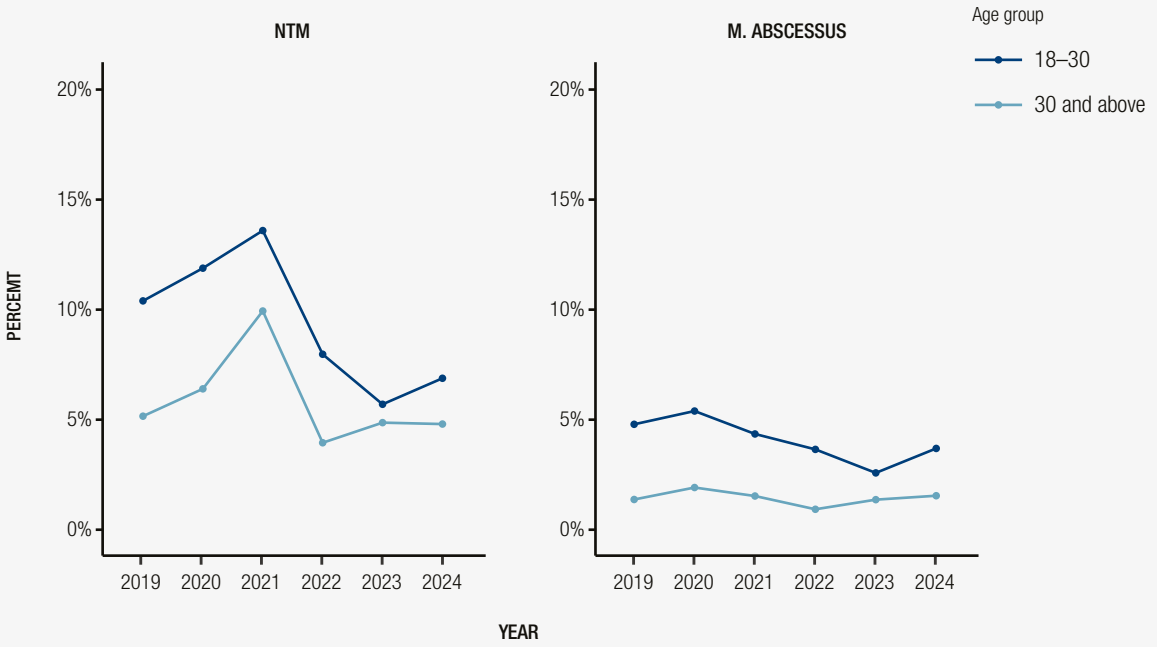
TABLE 3.3: ACFDR ADULTS 2024: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE

All samples				
	18-24	25-34	35-44	45+
Number of samples in the age range	592	708	503	366
Number of samples taken in 2023	358	401	274	207
Number of patients	348	399	271	203
P. aeruginosa	74 / 358 (20.7%)	163 / 401 (40.6%)	165 / 274 (60.2%)	112 / 207 (54.1%)
H. influenzae	54 / 358 (15.1%)	31 / 401 (7.7%)	17 / 274 (6.2%)	6 / 207 (2.9%)
B. cepacia complex	8 / 358 (2.2%)	7 / 401 (1.7%)	5 / 274 (1.8%)	<5
S. aureus	180 / 358 (50.3%)	165 / 401 (41.1%)	79 / 274 (28.8%)	43 / 207 (20.8%)
MRSA	16 / 358 (4.5%)	11 / 401 (2.7%)	12 / 274 (4.4%)	<5
Achromobacter spp	14 / 358 (3.9%)	7 / 401 (1.7%)	<5	5 / 207 (2.4%)
S. maltophilia	17 / 358 (4.7%)	17 / 401 (4.2%)	6 / 274 (2.2%)	9 / 207 (4.3%)
S. marcescens	<5	5 / 401 (1.2%)	<5	<5
Aspergillus spp	44 / 358 (12.3%)	43 / 401 (10.7%)	36 / 274 (13.1%)	34 / 207 (16.4%)
M. abscessus	14 / 358 (3.9%)	8 / 401 (2.0%)	6 / 274 (2.2%)	<5
NTM	26 / 358 (7.3%)	18 / 401 (4.5%)	15 / 274 (5.5%)	12 / 207 (5.8%)

Non Tuburculous Mycobacterium Prevalence

Non-Tuburculous Mycobacterium (NTM), particularly *M. abscessus* infection, has been associated with poorer outcomes for pwCF. NTM infection rates, including by *M. abscessus* for pwCF increased from 2017, peaking in 2021. Since then, infection rates have returned to levels similar to those in earlier years (Figure 3.18).

FIGURE 3.18: ACFDR ADULTS 2019-2024: PREVALENCE OF NTM INFECTIONS BY AGE



3.3 CF MANAGEMENT

Clinical 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 peaking in 2022 at 10,280 visits (Figure 3.19 and Table 3.4). In 2024, there were a total of 8,392 clinical visits of which 63% were in a clinic, 36% were telehealth, and 2.4% were outreach.

FIGURE 3.19: ACFDR ADULTS 2021-2024: TOTAL NUMBER OF VISITS RECORDED PER YEAR

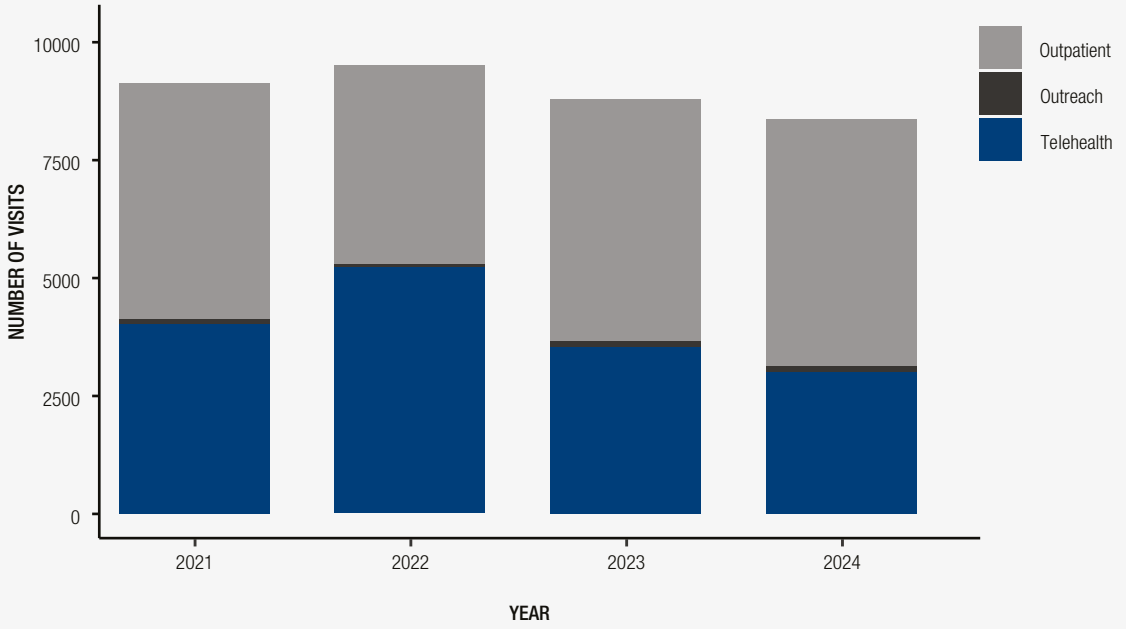


TABLE 3.4: ACFDR ADULTS 2021-2024: TOTAL NUMBER OF CLINICAL VISITS

Visit type	2021	2022	2023	2024
Outpatient	5,404 (55.0%)	4,620 (45.0%)	5,162 (58.2%)	5,263 (62.7%)
Outreach	90 (1.0%)	86 (1.0%)	131 (1.5%)	121 (1.4%)
Telehealth	4,328 (44.0%)	5,574 (54.0%)	3,571 (40.3%)	3,008 (35.8%)
Total	9,822 (100.0%)	10,280 (100.0%)	8,864 (100.0%)	8,392 (100.0%)

Standards of Care

The Australian CF Standards of Care for pwCF recommend four clinical visits per year. In 2024 the number of adults with CF who had at least 4 clinic visits was 1,092 (50.4%) overall (Figure 3.20). This was similar among pwCF of 18-24 years (48.6%) and those who were 30+ years old (51.7%). This proportion has declined over time for both age groups (Table 3.5). The average number of clinical visits per adult with CF is 3.9 for adults between the ages of 18-29 and 4 for adults thirty years or older (Table 3.6).

FIGURE 3.20: ACFDR ADULTS 2024: PROPORTION OF PATIENTS WITH 4 OR MORE CLINICAL VISITS

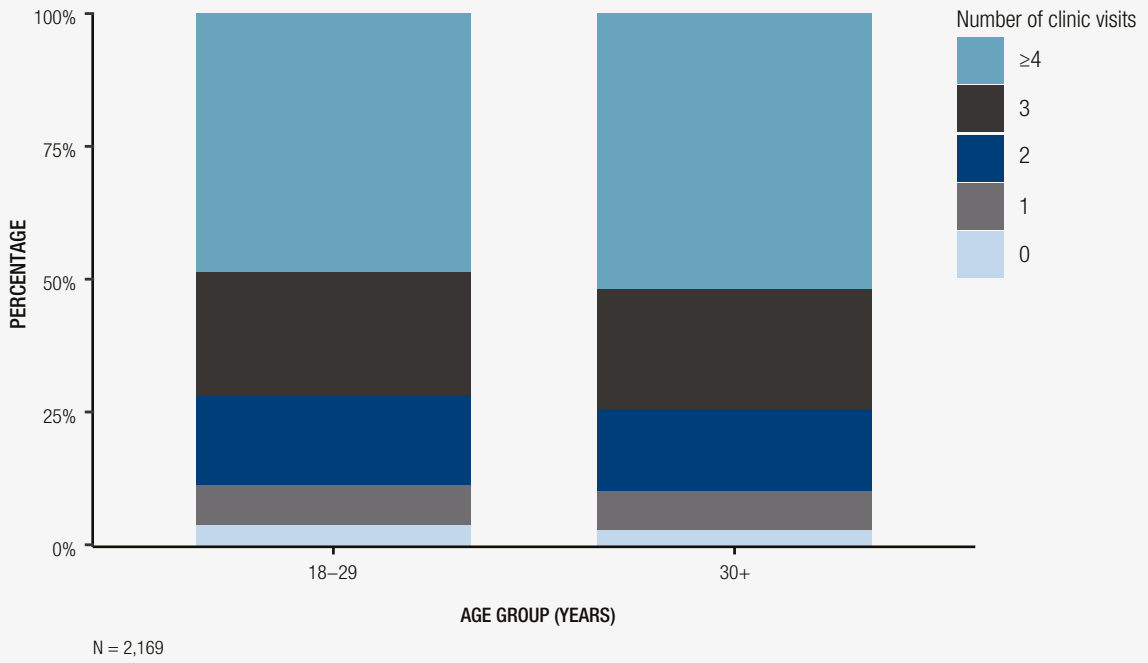


TABLE 3.5: ACFDR ADULTS 2021-2024: PROPORTION WITH 4 OR MORE CLINICAL VISITS BY AGE

	Number/% with 4+ visits			
Age	2021	2022	2023	2024
18-29	607 (67.0%)	568 (61.0%)	537 (57.0%)	468 (48.6%)
30+	669 (63.0%)	735 (62.0%)	646 (56.0%)	624 (51.7%)
Total	1,217 (60.3%)	1,303 (64.5%)	1,183 (56.0%)	1,092 (50.4%)

TABLE 3.6: ACFDR ADULTS 2024: AVERAGE NUMBER OF CLINICAL ENCOUNTERS PER PERSON

Age	Average number of clinic visits
18-29	3.9
30+	4.0
Total	4.0

Hospitalisations

In 2024, data regarding adult hospitalisations was reported for all adults, excluding those with transplants (105), totalling 2,169 adults.

Approximately three quarters (73%) of adults did not have any hospitalisations in 2024. Of 18-29-year olds, 16% had 1 hospitalisation, 5% had 2 hospitalisations, and 5% had 3 or more hospitalisations. For adults with CF aged 30 years or more, 19% had 1, 6% had 2, and 3% had 3 or more hospitalisations during 2024 (Figure 3.21 and Table 3.7).

FIGURE 3.21: ACFDR ADULTS 2024: HOSPITALISATIONS BY AGE

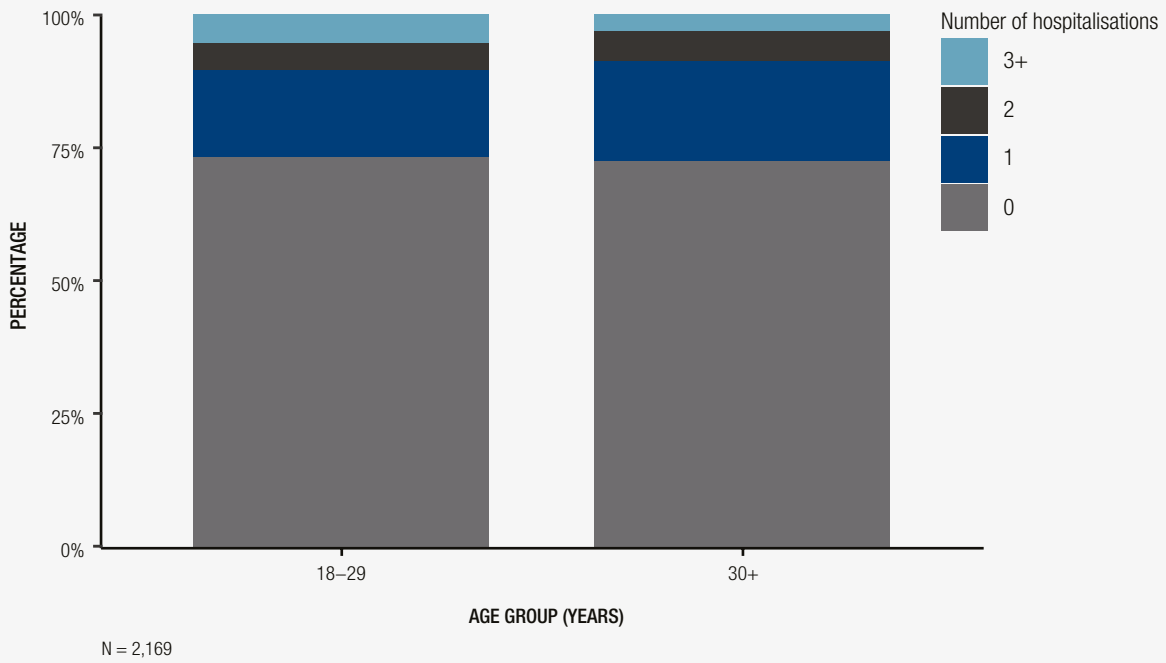


TABLE 3.7: ACFDR ADULTS 2024: HOSPITALISATIONS BY AGE

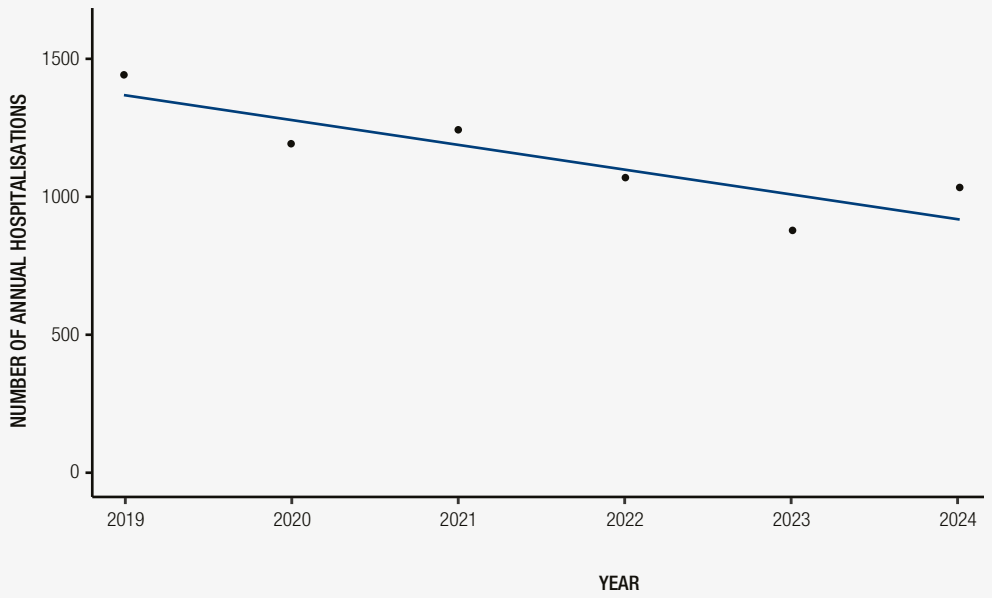
Age	Hospitalisations	N (%)
18-29	0	707 (73.4%)
	1	157 (16.3%)
	2	38 (3.2%)
	3+	52 (5.4%)

Age	Hospitalisations	N (%)
30+	0	876 (72.6%)
	1	226 (18.7%)
	2	66 (5.5%)
	3+	38 (3.2%)

Adult Hospitalisations

Adult hospitalisations per year continued to decrease from 2019 to 2024, from 1,446 admissions in 2019 to a low of 877 admissions in 2023. Hospitalisations increased slightly to 1,034 in 2024, however, the overall trend has seen a 28.5% reduction over the last six years (Figure 3.22).

FIGURE 3.22: ACFDR ADULTS 2019-2024: HOSPITALISATIONS PER YEAR



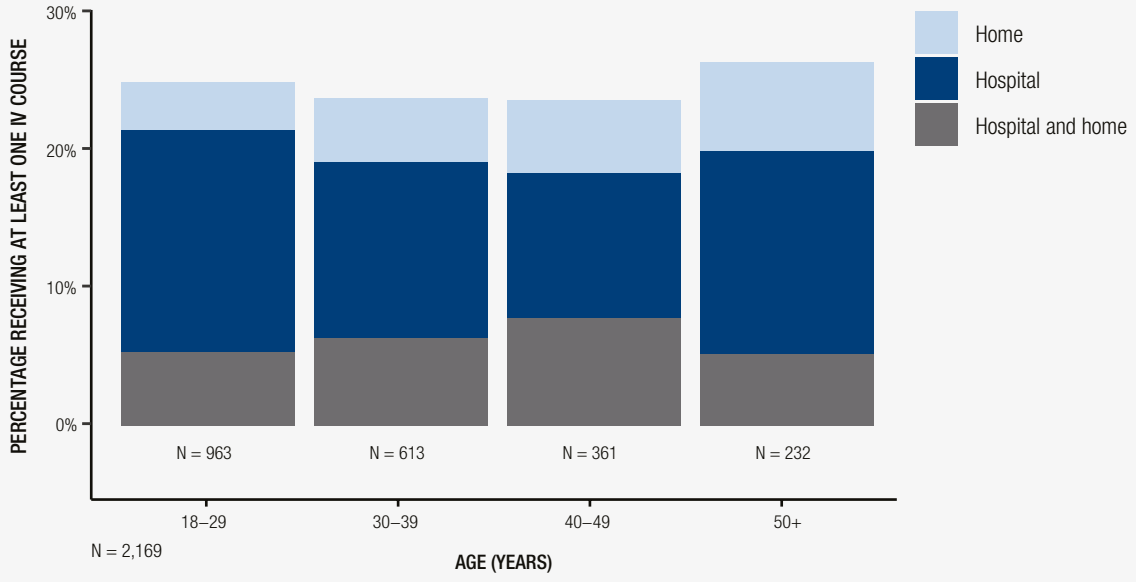
IV Antibiotic Therapy

In 2024, there were 530 adults who received at least one course of IV antibiotic therapy; this represents 24.4% of all adults with CF (excluding adults who had a lung transplant).

The most common setting for treatment with IV antibiotic therapy was hospital only, with 14.1% of adults being admitted to hospital only for IV antibiotics, followed by 6.0% of adults receiving treatment in hospital then at home, and 4.4% adults receiving IV treatment at home only (Figure 3.23).

The age groups most likely to receive IV antibiotics were those aged 50+ years (26.4% of the cohort), followed by those aged 18-29 years (24.8% of the cohort), 30-39 years (23.7% of the cohort), and 40-49 years (23.6% of the cohort).

FIGURE 3.23: ACFDR ADULTS 2024: IV ANTIBIOTIC THERAPY BY AGE



In 2024, the median duration of IV antibiotic therapy in hospital only was 12 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).

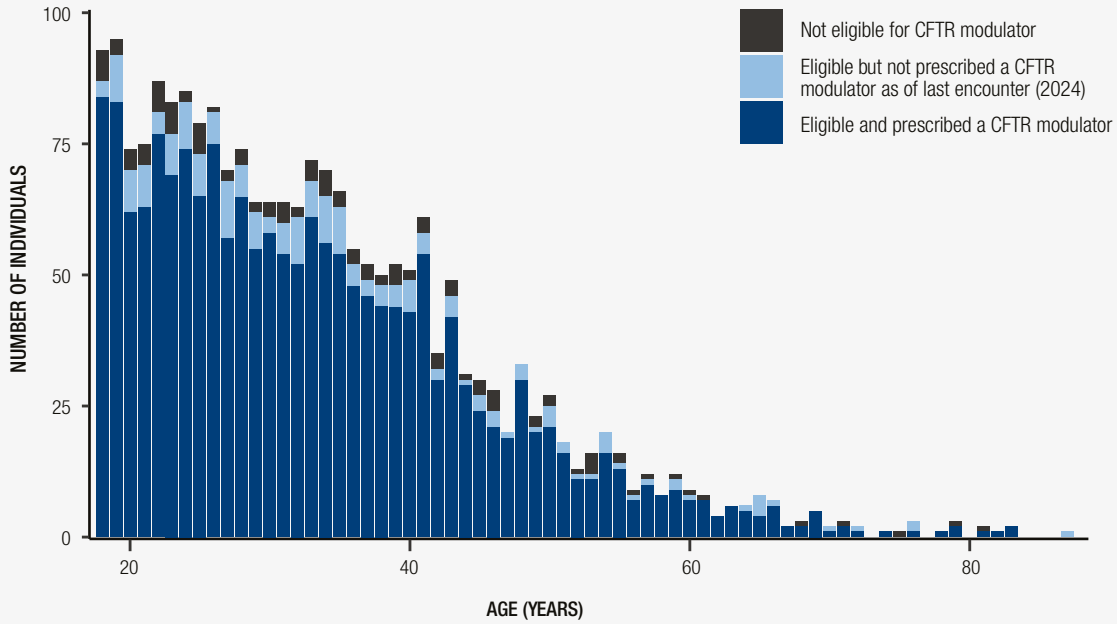
TABLE 3.8: ACFDR ADULTS 2024: MEDIAN DAYS IV ANTIBIOTICS

Location of IV antibiotic treatment	N (median days on IV antibiotics)
Home	95 (14)
Hospital	302 (12)
Hospital and home	130 (14)

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 31st December 2024. This is generally those pwCF on modulators available via the PBS. In 2024, 2,161 adults had their eligibility for a CFTR modulator (based on genotype) known (95.0% of adults). Figure 3.24 shows that the vast majority of these adults with known eligibility were prescribed a modulator at the end of 2024.

FIGURE 3.24: ACFDR ADULTS 2024: CFTR MODULATOR ELIGIBILITY AND PRESCRIBING BY AGE

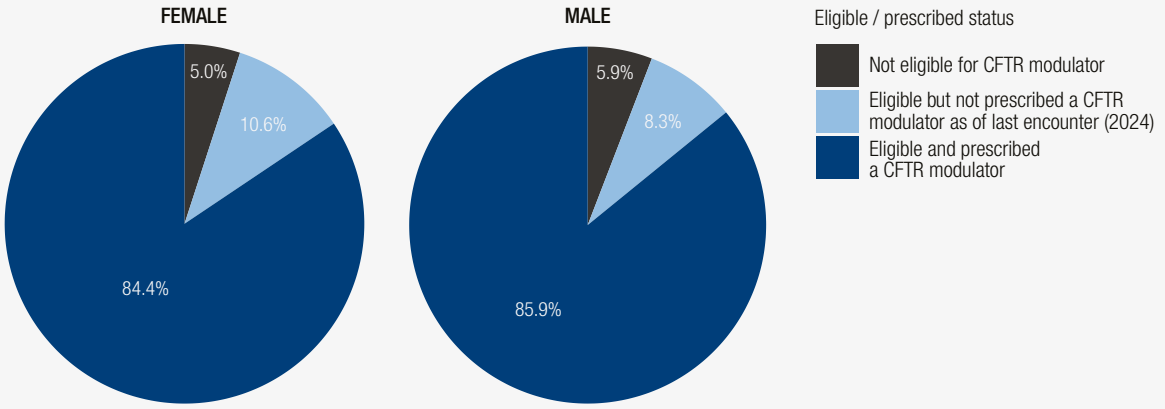


N = 2,161 individuals with known eligibility

Of the 2,161 adults in the registry in 2024 who had known eligibility for a modulator, 5.0% of females and 5.9% of males were not eligible for a CFTR modulator. 84.4% of females and 85.9% of males were eligible and prescribed a modulator, with 10.6% of females and 8.3% of males being eligible and not prescribed a modulator (Figure 3.25).

This is an increase in modulator usage since 2022, when 80% of females and 83% of males respectively were prescribed modulators, and 13% and 9% respectively were eligible but not prescribed. The proportion of adults not eligible for a modulator remained similar at 7%.

FIGURE 3.25: ACFDR ADULTS 2024: COMBINED ELIGIBILITY AND PRESCRIPTION OF CFTR MODULATORS



N = 2,161 individuals with known eligibility

The following CFTR modulators are available to pwCF meeting the defined eligibility criteria:

Ivacaftor (KALYDECO®)

Ivacaftor is available on the PBS for pwCF, who are aged 4 months and older, and who have at least mutation in the CFTR gene that is responsive to Ivacaftor potentiation based on clinical and/or in vitro data. Further information on Ivacaftor eligibility can be found via the Therapeutic Goods Administration Kalydeco Product Information sheet.

Lumacaftor/Ivacaftor (ORKAMBI®)

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

Tezacaftor/Ivacaftor (SYMDEKO®)

Tezacaftor/ivacaftor is also a combination therapy available on the PBS for pwCF, are aged 12 years and older, and who have one copy of the following changes in the CFTR gene: E56K, R117C, F508del, S977F, F1074L, 3849+10kbC→T, P67L, E193K, D579G, F1052V, D1152H, R74W, L206W, 711+3A→G, K1060T, D1270N, D110E, R352Q, E831X, A1067T, 2789+5G→A, D110H, A455E, S945L, R1070W, 3272-26A→G.

Elexacaftor/Tezacaftor/Ivacaftor (TRIKAFTA®)

Elexacaftor/tezacaftor/ivacaftor (ETI) is a triple combination therapy available on the PBS in for pwCF aged 2 years and older, with at least one copy of the F508del gene change in the CFTR gene. Trikafta was initially available on the PBS in April 2022 for pwCF aged 12 and older. In May 2023 pwCF aged 6-11 were eligible to receive Trikafta, and in August 2024 pwCF aged 2-5 were able to access the treatment.

In adults, Trikafta uptake was consistently high. Among those aged 18-29 years, 82.1% were receiving Trikafta, and this remained high (76.9%) in adults aged ≥30 years. Use of older CFTR modulators was low across adult groups (5.3%-8.6%), indicating a transition away from previous-generation therapies. Only a small minority were eligible but not yet treated (9.1%-9.7%), and very few were classified as not eligible for modulators (3.7%-4.8%), 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 patients accessing Trikafta in 2024, the following modulator data will be categorised as 'Trikafta' and 'other CFTR modulator'.

TABLE 3.9: ACFDR ADULTS 2024: 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	961	787 (81.9%)	51 (5.3%)	87 (9.1%)	36 (3.7%)
30+	1200	920 (76.7%)	106 (8.8%)	116 (9.7%)	58 (4.8%)

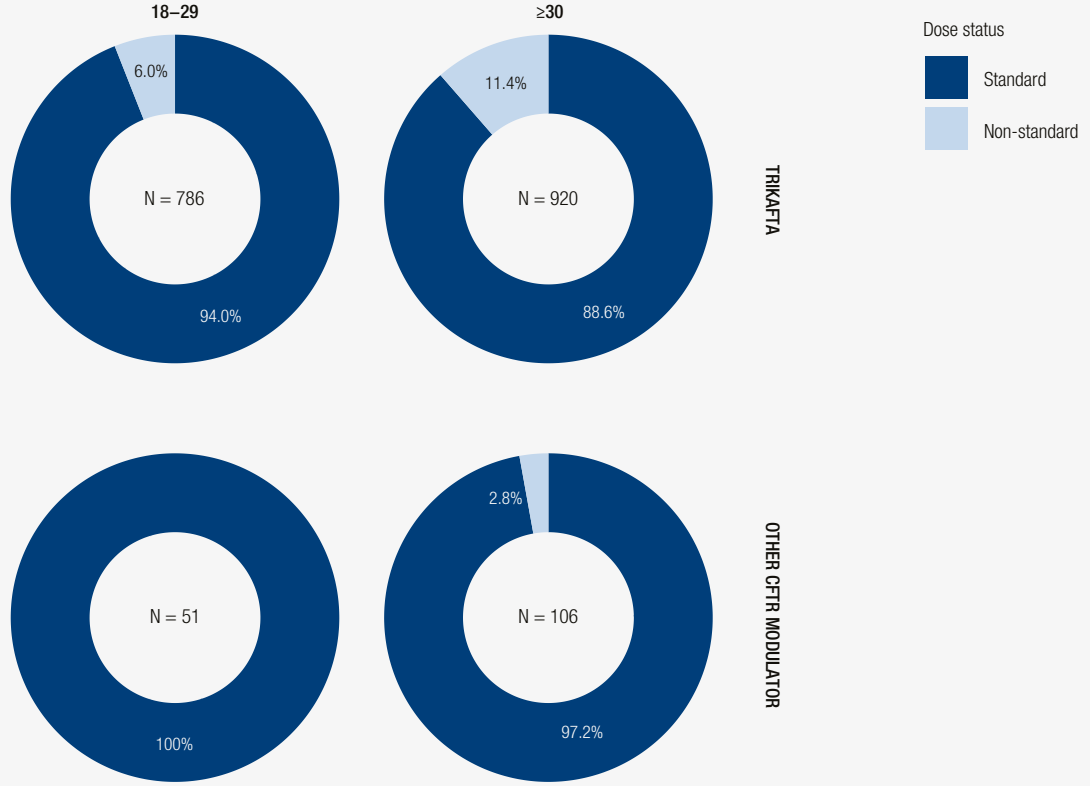
Table 3.10 shows the reasons for permanent or temporary cessation and change to modulator prescriptions. Throughout 2024, intolerance/adverse events alongside unspecified non-adverse events were the most common reason for discontinuation or switch to another modulator, followed by liver impairment or intolerance. A small proportion of pwCF discontinued a CFTR modulator due to a switch to another modulator, rash, or weight gain.

TABLE 3.10: ACFDR ADULTS 2024: REASONS FOR DISCONTINUATION/SWITCH FROM MODULATORS

Reasons for discontinuation/change	Total N	Trikafta N (%)	Other CFTR modulator N (%)
Other intolerance/adverse event	18	15 (45.5%)	<5
Other reason (non-adverse)	15	9 (27.3%)	6 (40.0%)
Liver impairment/intolerance	8	7 (21.2%)	<5
Rash	<5	<5	<5
Switch to other CFTR modulator	<5	0 (0.0%)	<5
Weight gain	<5	<5	0 (0.0%)
Pulmonary side effect/intolerance	0	0 (0.0%)	0 (0.0%)
Pregnancy	0	0 (0.0%)	0 (0.0%)
Concomitant drug interaction	0	0 (0.0%)	0 (0.0%)
Mental health	0	0 (0.0%)	0 (0.0%)

In 2024, more than 85% of adults on modulators were on standard doses. Figure 3.26 shows the proportion of patients aged 18-29 and 30 years and older on standard and non-standard doses of Trikafta and other CFTR modulators. 6.0% of adults aged 18-29 and 11.4% of adults aged 30 or older were on non-standard doses of Trikafta. Less than 3% of all adults on other CFTR modulators were on non-standard doses.

FIGURE 3.26: ACFDR ADULTS 2024: STANDARD AND NON-STANDARD DOSE BY AGE AND MODULATOR



3.4 COMPLICATIONS AND THERAPIES

CF Pulmonary Disease

In 2024, the rate of haemoptysis requiring hospital admission was 5.4% for adults 18-29 years, and 7.4% for those aged 30+. Embolisation was required for 0.6% of haemoptysis presentations for 18 to 29-year-olds and 0.4% for 30+ year-olds. Pneumothoraces occurred in 0.0% of 18-29 year olds and 0.2% 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.

These **new figures** show changes in use of adjuvant pulmonary therapies over the last five years. Overall, there has been a significant reduction in use of inhaled antibiotics and macrolides in this age group, with stable use of regular oral antibiotics.

In 2024, among individuals aged 18-29, 21.0% were prescribed inhaled antibiotics, 17.0% were on regular oral antibiotics, and 20.3% were using macrolides (Figures 3.27A and B). In the 30+ age group, 33.5% were on inhaled antibiotics, 21.7% were on regular oral antibiotics, and 42.6% were using macrolides. For 18-29-year-olds, the use of both inhaled antibiotics and macrolides has gradually decreased from approximately two-fifths in 2020 to one-fifth of in 2024. For pwCF aged 30 and older, inhaled antibiotics use decreased from approximately half of the cohort in 2020 to one-third of 30+ year-olds in 2024. Macrolides usage has also decreased with time for adults aged 30 and older, from three-fifths of the cohort in 2020 to approximately two-fifths in 2024.

FIGURE 3.27A: ACFDR ADULTS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR ADULTS AGED 18-29 YEARS

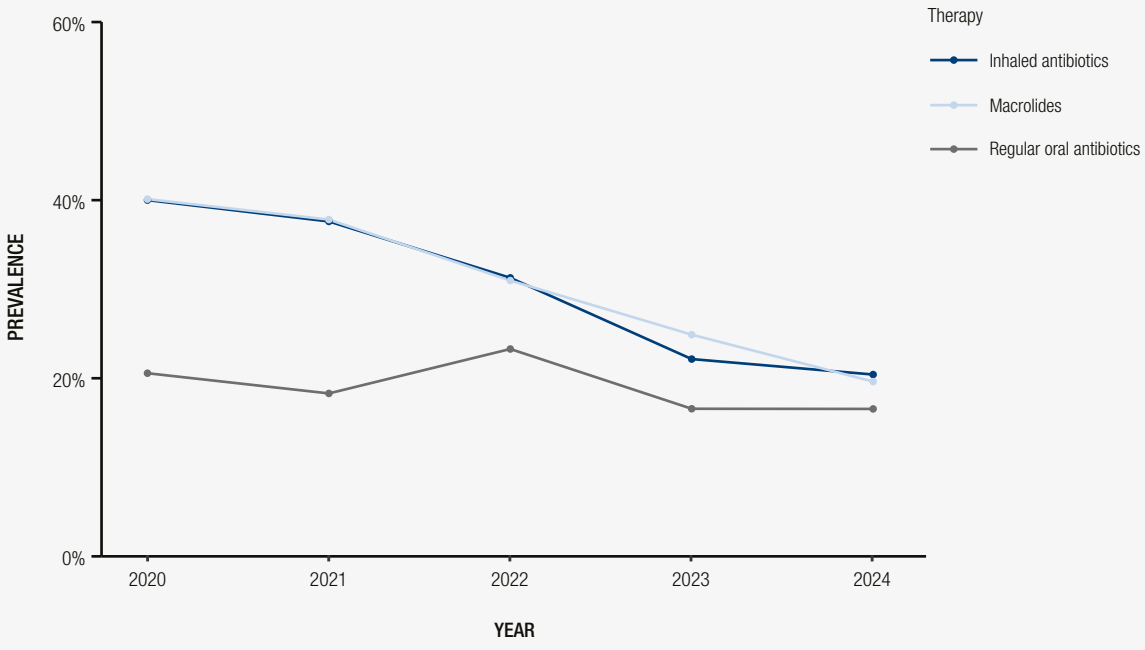
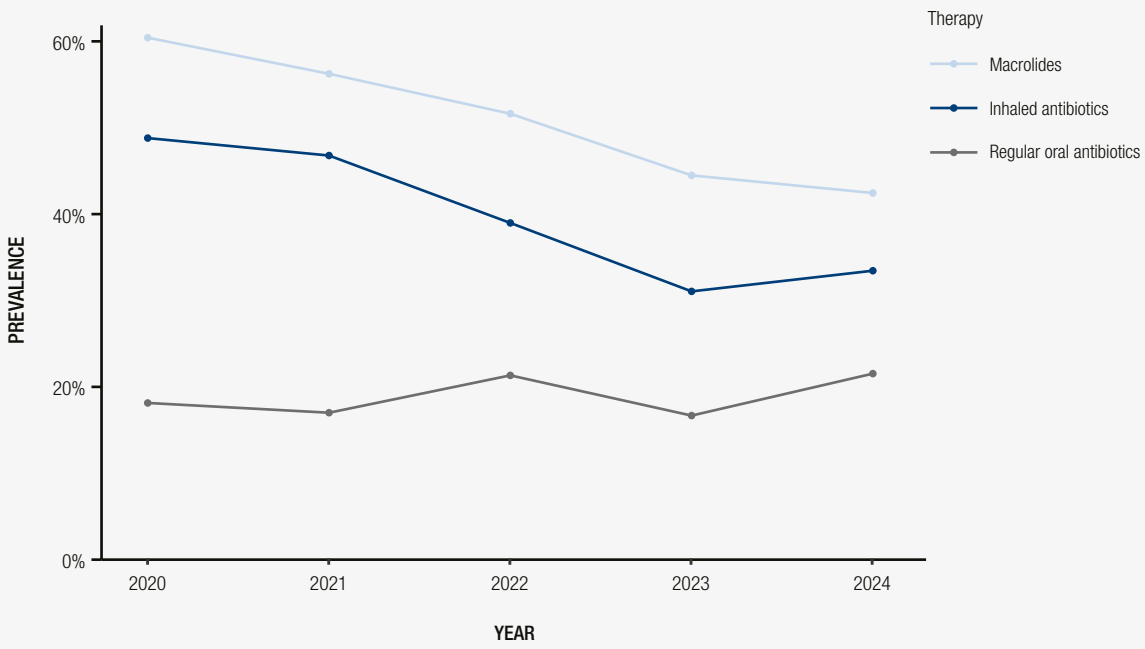


FIGURE 3.27B: ACFDR ADULTS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR ADULTS AGED 30 YEARS AND OLDER



CF Lung Therapies — Non-Antibiotic Management

These **new figures** also show a reduction in other (non-antibiotic) adjuvant pulmonary therapies. Among both age groups there has been a reduction in use of bronchodilators, dornase alpha, hypertensive saline, and inhaled corticosteroids, although the decline seems to have plateaued for older adults in 2024. There has been little change in use of the remaining therapies.

In 2024, among adults 18-29 years, approximately half used bronchodilators (51.1%) and dornase alpha (45.9%), approximately one third used inhaled corticosteroids (33.5%), and 39.5% used hypertonic saline (Figures 3.28A and 3.28B). Less commonly used drugs were inhaled mannitol (5.4%), oral corticosteroids (2.9%), non-invasive ventilation (1.2%) and long-term oxygen therapy (0.4%).

In 2024, among adults aged 30 years or older there was a slightly higher use of bronchodilators (60.3%), inhaled corticosteroids (48.4%), oral corticosteroids (4.1%), long-term oxygen therapy (2.1%) and non-invasive ventilation (3.0%). A large minority also used dornase alpha (39.1%) and hypertonic saline (38.6%), whereas the use of inhaled mannitol was uncommon compared to younger adults (2.4%).

FIGURE 3.28A: ACFDR ADULTS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR ADULTS AGED 18-29 YEARS

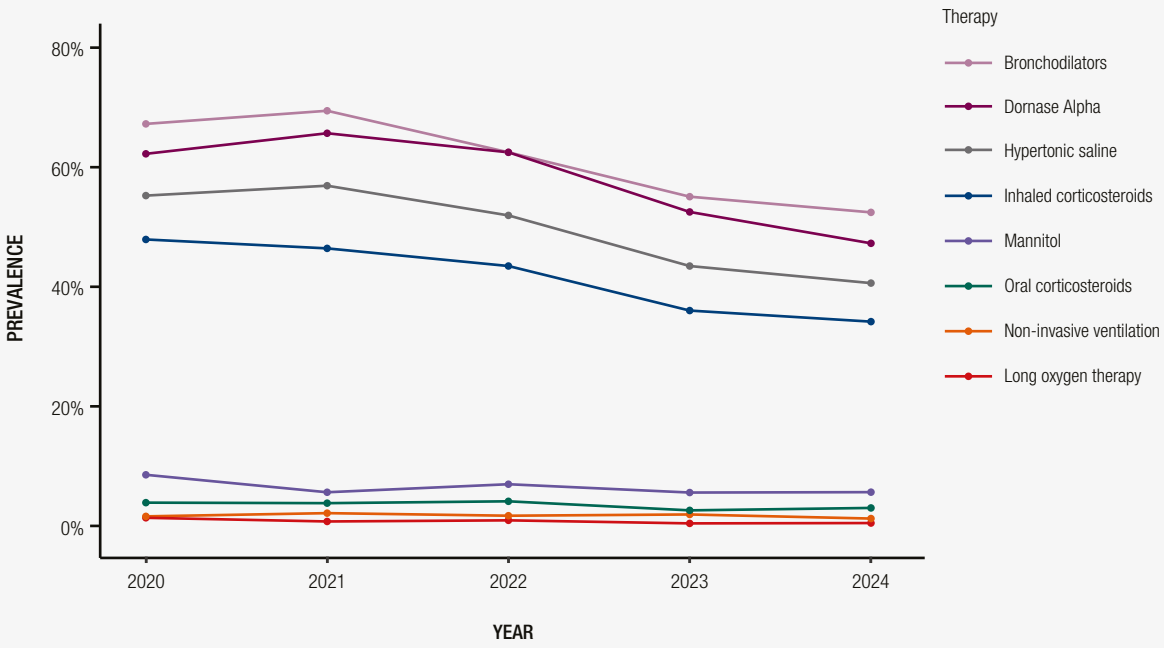
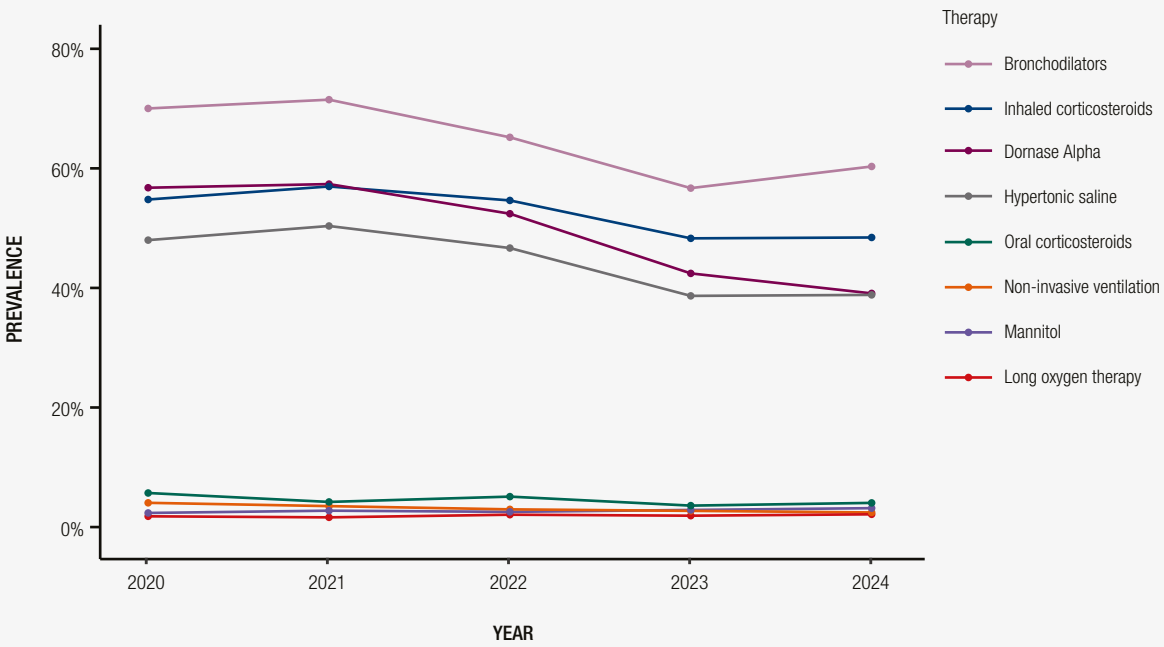


FIGURE 3.28B: ACFDR ADULTS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR ADULTS AGED 30 YEARS AND OLDER



CF Related Diabetes

For the first time, these new figures show change in diabetes status over time (2019–2024). In 2024, 14.8% of 18-29-year-olds and 12.7% of 30+ year-olds had impaired glucose tolerance. The proportion of those with diabetes was 25.1% for 18-29-year-olds and 30.3% for those aged 30 years or older. The prevalence of impaired glucose tolerance has reduced slightly since 2019 (from 20.0% of 18-29-year-olds and 20.4% of 30+ year-olds) and has stayed relatively stable from 2020 to 2024. The prevalence of diabetes has increased, from 24.1% of 18-29-year-olds and 25.7% of 30+ year-olds in 2019 (Figures 3.29A and B).

FIGURE 3.29A: ACFDR ADULTS 2019-2024: DIABETIC STATUS FOR ADULTS AGED 18-29 YEARS

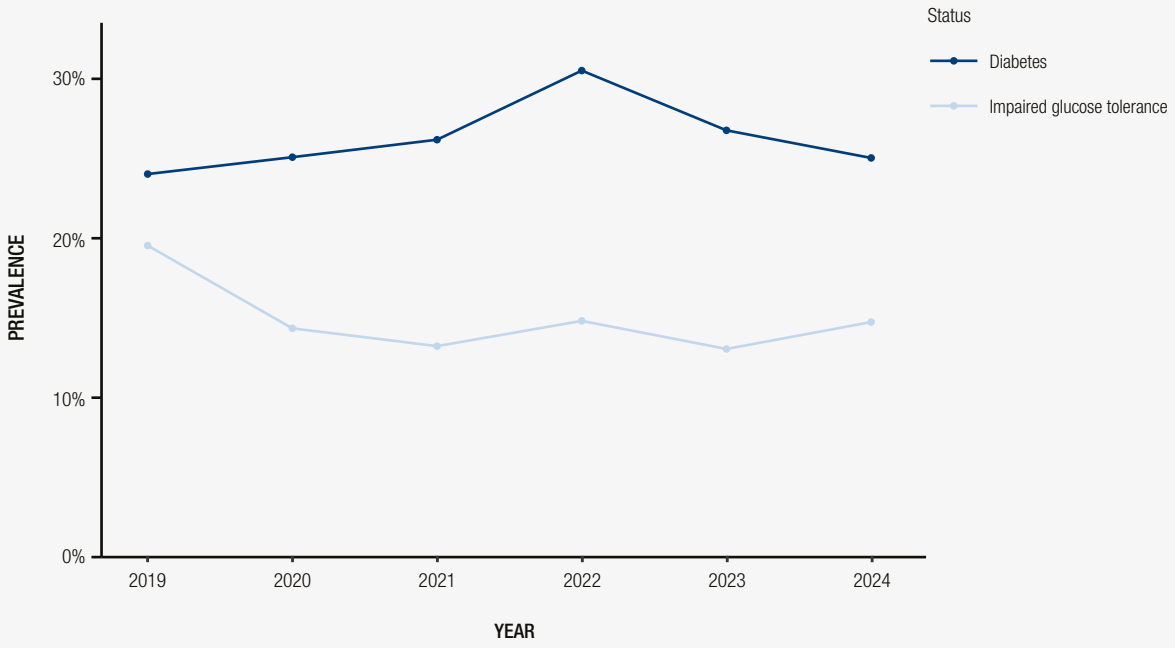
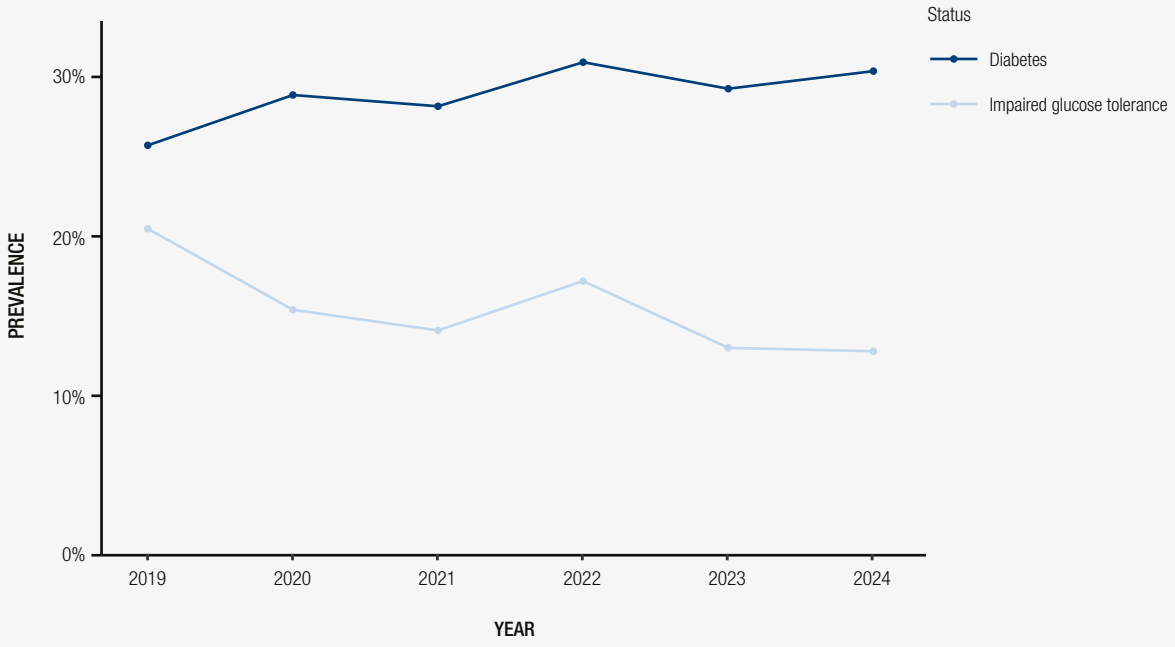


FIGURE 3.29B: ACFDR ADULTS 2019-2024: DIABETIC STATUS FOR ADULTS AGED 30 YEARS AND OLDER



Of those with diabetes, the vast majority are treated with insulin: 83.3% of 18-29-year olds and 70.8% 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. Oral 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 2024: DIABETIC TREATMENT BY AGE

Diabetes treatment type	18-29 (N = 221)	30+ (N = 339)
Insulin	184 (83.3%)	240 (70.8%)
Oral Hypoglycemics	<5	19 (5.6%)
Insulin and oral hypoglycemics	<5	6 (1.8%)
Diet/lifestyle management only	22 (10.0%)	66 (19.5%)
No treatment for diabetes	8 (3.6%)	8 (2.4%)

Of those that use insulin, the vast majority (95.9% of 18-29 year olds and 94.8% of 30+ year olds) require chronic insulin administration. Only 2.9% of 18-29 year olds and 4.4% of 30+ year olds use insulin intermittently (Table 3.12).

TABLE 3.12: ACFDR ADULTS 2024: INSULIN USE BY AGE

Insulin use	18-29 (N = 170)	30+ (N = 250)
Intermittent insulin use	5 (2.9%)	11 (4.4%)
Chronic insulin use	163 (95.9%)	237 (94.8%)
Insulin use, duration unknown	<5	<5

CF Gastrointestinal Disease

Stomach and Liver

Gastrointestinal complications for people with CF include those related to the stomach, pancreas and liver. In 2024, among adults aged 18-29, 22.8% experienced gastroesophageal reflux (compared to 24.4% in 2023), increasing to 42.3% for those aged 30 years and older (compared to 40.1% in 2023).

The most common liver disease for pwCF is acute (non-cirrhotic) liver disease which affected 9.7% of 18 to 29-year-olds (an increase from 6.1% in 2023) and 11.2% of 30+ year-olds in 2024 (compared to 6.7% in 2023). Chronic liver disease affected 6.8% of 18 to 29-year-olds (compared to 5.0% in 2023) and 6.5% of 30+ year-olds in 2024 (an increase from 5.3% in 2023) (Table 3.13).

TABLE 3.13: ACFDR ADULTS 2024: GASTROINTESTINAL COMPLICATIONS BY AGE

	18-29	30+
Gastroesophageal reflux	215 / 945 (22.8%)	500 / 1,182 (42.3%)
Liver disease, non-cirrhosis (includes viral hepatitis, fatty liver)	85 / 876 (9.7%)	124 / 1,106 (11.2%)
Liver disease, cirrhosis (image confirmed)	19 / 810 (2.3%)	36 / 1,018 (3.5%)
Liver disease, cirrhosis with portal hypertension	37 / 828 (4.5%)	30 / 1,012 (3.0%)

Pancreatic Disease

The majority of adult participants in the registry were pancreatic insufficient. In 2024, 83.8% of pwCF aged 18-29, and 73.9% 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 3.14).

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), whereas acute pancreatitis has increased (from <5 of 18 to 29-year-olds and <5 of 30+ year-olds).

TABLE 3.14: ACFDR ADULTS 2024: PANCREATIC DISEASE BY AGE

Pancreatic status	18-29 (N = 947)	30+ (N = 1,185)
Insufficient	794 (83.8%)	876 (73.9%)

Pancreatitis	18-29 (N = 909)	30+ (N = 1,138)
Acute (first pancreatitis event this current year); pancreatitis not otherwise specified	10 (1.1%)	7 (0.6%)
Recurrent pancreatitis (history of more than one event of pancreatitis)	12 (1.3%)	29 (2.5%)
No history of pancreatitis	887 (97.6%)	1,102 (96.8%)

Bone Density Status and Osteopenia

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

In 2024, of those that recorded results from bone mineral density scans, 56.4% of 18-29 year-olds and 34.9% of 30 + year-olds reported bone mineral density within the normal range. Osteopenia was reported by 36.1% of 18-29 year-olds and 47.8% of 30+ year-olds, and osteoporosis was reported by 7.4% and 17.3% respectively. Thirty-three fractures were reported, with 13 (1.6%) among 18-29 year-olds and 20 (2.1%) among 30+ year-olds (Table 3.15).

TABLE 3.15: ACFDR ADULTS 2024: BONE DENSITY STATUS

Bone mineral density	18-29 (N = 202)	30+ (N = 289)
Normal	114 (56.4%)	101 (34.9%)
Osteopenia	73 (36.1%)	138 (47.8%)
Osteoporosis	15 (7.4%)	50 (17.3%)
Bone mineral density	**18-29 (N = 800)**	**30+ (N = 972)**
Fracture	13 (1.6%)	20 (2.1%)

Cancer

Of the ACFDR's adult cohort, 18-29 year-olds had two recorded cases of cancer, whereas those aged 30 and above had nine newly diagnosed cases. One of the diagnoses were colorectal cancers.

Nutritional Supplementation

Pancreatic enzymes have been a mainstay of treatment for pancreatic insufficiency for pwCF. In 2024, 79.7% of those aged 18-29 and 75.3% of those aged 30 years or older used pancreatic enzymes. A majority (64.8% and 64.1% respectively) also used vitamin supplements, with 29.4% and 19.2% respectively using salt replacement therapy.

A small proportion of adults required nutritional support in 2024, 5% or less of the population (Table 3.16).

TABLE 3.16: ACFDR ADULTS 2024: NUTRITIONAL SUPPLEMENTS BY AGE

Nutritional supplementation	18-29 (N = 947)	30+ (N = 1,185)
Pancreatic enzymes	755 (79.7%)	892 (75.3%)
Vitamin supplements (Fat soluble vitamins A, D, E and K)	614 (64.8%)	759 (64.1%)
Salt replacement therapy	278 (29.4%)	227 (19.2%)

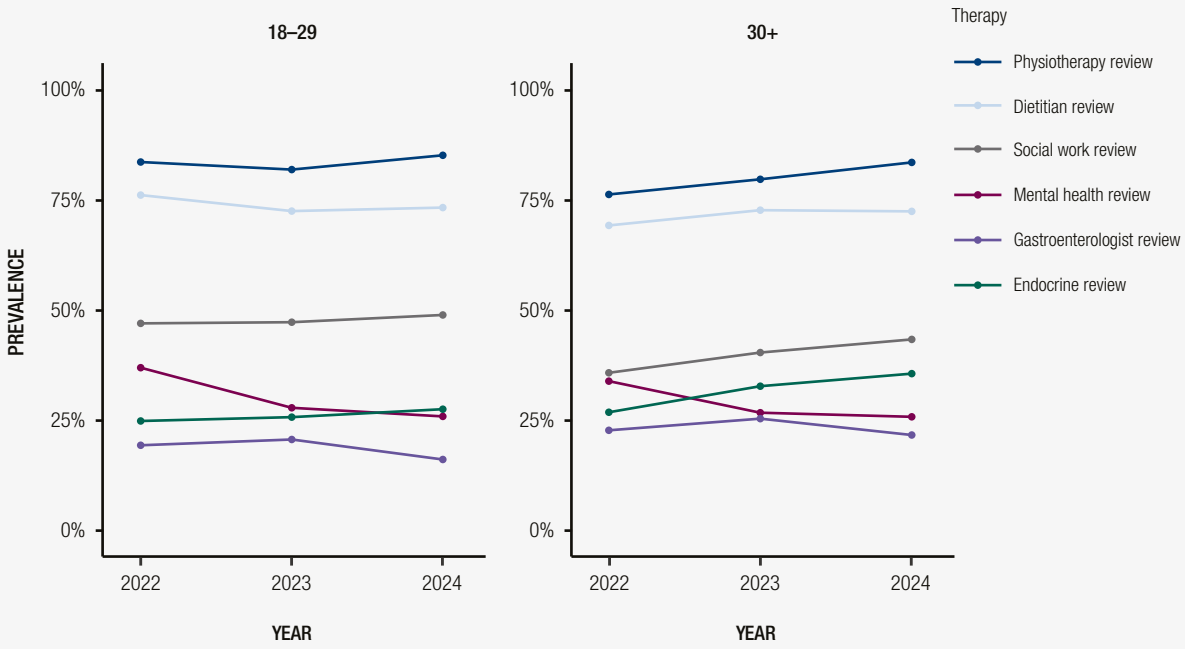
Nutritional support	18-29 (N = 947)	30+ (N = 1,185)
Oral	44 (4.6%)	19 (1.6%)
Gastrostomy tube	21 (2.2%)	<5
Nasogastric tube	<5	0 (0.0%)
Jejunostomy tube	0 (0.0%)	0 (0.0%)
Parenteral nutrition	0 (0.0%)	0 (0.0%)

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 3.30 shows that the majority of people with CF participate in annual physiotherapy and dietitian reviews. Of pwCF who are 18-29 in 2024, just under half participated in annual social work reviews; approximately one quarter participated in mental health and endocrine reviews and approximately one fifth participated in gastroenterologist reviews. Of adults with CF who are 30 years or older, just under half participated in social work reviews, approximately one third participated in endocrine reviews, and approximately one quarter participated in mental health and gastroenterologist reviews. The main trend is that mental health reviews have declined during the last 3 years for both age groups.

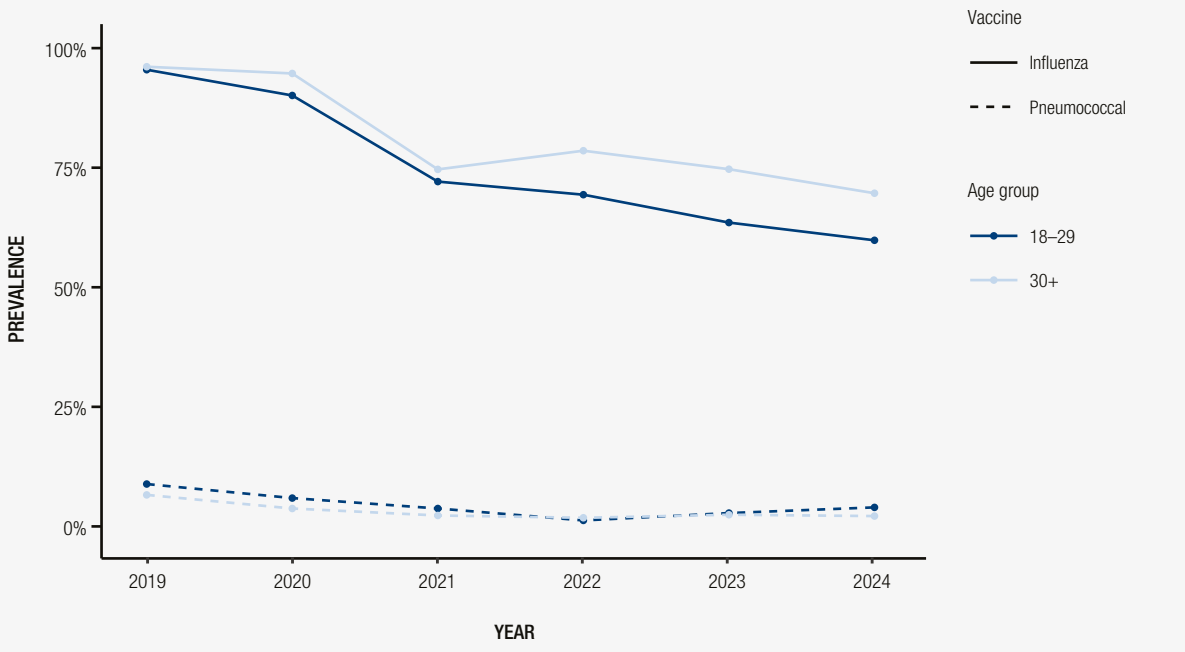
FIGURE 3.30: ACFDR ADULTS 2022-2024: MULTIDISCIPLINARY CARE REVIEWS BY AGE



Vaccination

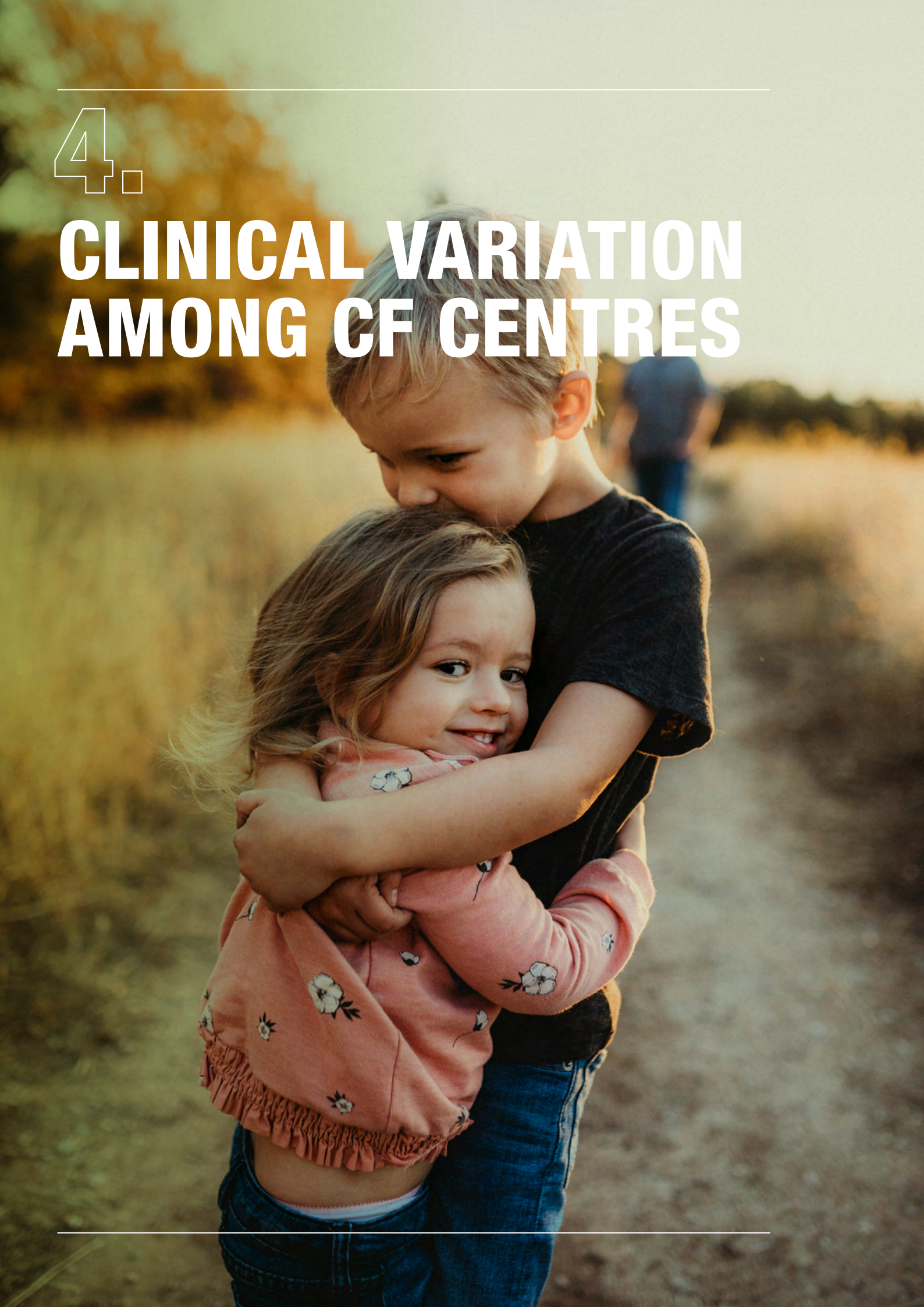
Fifty-eight percent of adults recorded influenza vaccination status in 2024. Of these, 69.6% of adults aged 30 years or older, and 59.8% of adults aged 18-29 years reported receiving an influenza vaccination. This is a decrease from 2019 when 96.0% of 30+ year-olds and 95.4% of 18-29 year-olds reported receiving an influenza vaccination. (Figure 3.31).

FIGURE 3.31: ACFDR ADULTS 2019-2024: 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 AFCDR 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 31st December 2024. 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.

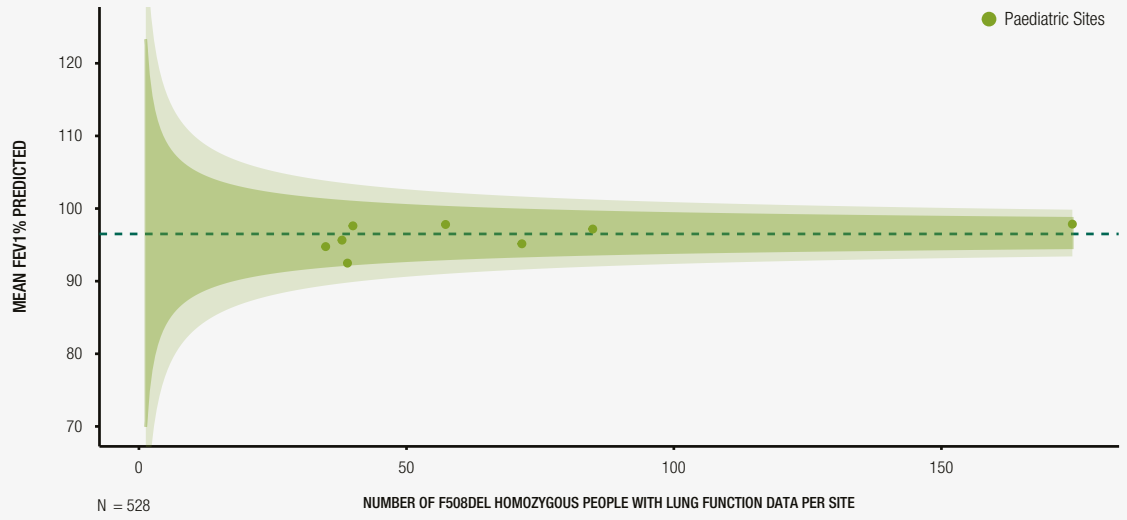
For pwCF who had measurements taken at more than one centre, annual values contributing to the charts and tables for each of these centres were compiled from all clinical measurements reported by any centre.

Height and BMI percentiles were calculated using the WHO growth chart, and Weight percentiles were calculated using the 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

Figure 4.1 funnel plot shows the mean FEV1 pp for different paediatric sites (n = 8). 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 95% 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. The remaining sites display a range of mean FEV1 pp values from 92.5% to 97.9%, with patient numbers varying between 34 and 171 per site.

FIGURE 4.1: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN FEV1 PP AT PAEDIATRIC SITES

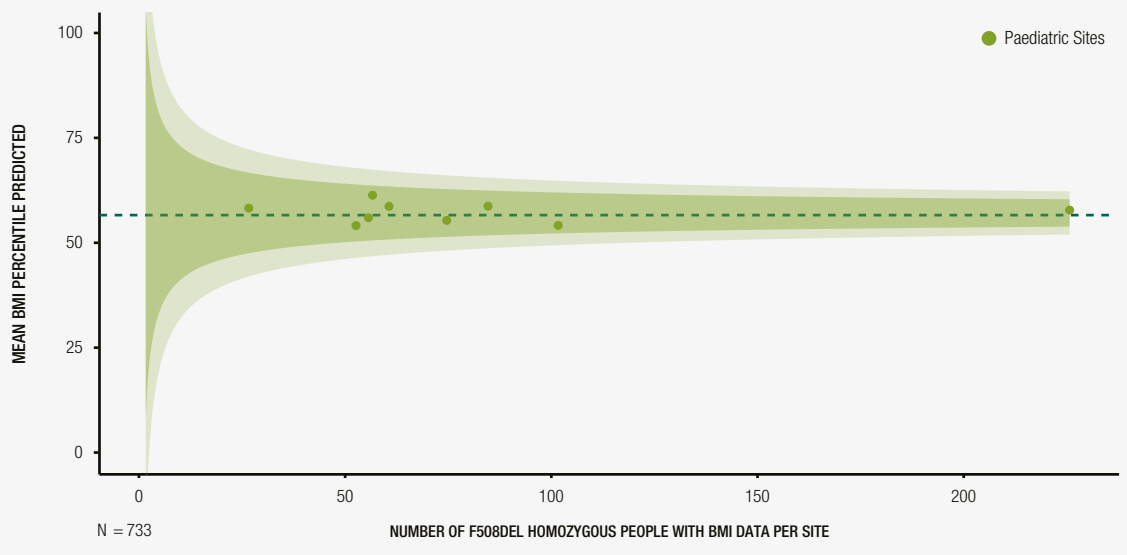


Shaded areas represent 95 and 99.8% control limits
Mean FEV1% predicted in all paediatric sites = 97%
Mean was caluclated across all the measurements
* Age, sex and height were standardised for when caluclating FEV1 % predicted
Sites with fewer than 20 cases are not shown.

Paediatrics: Variation in BMI

Figure 4.2 illustrates the average BMI percentiles across different paediatric sites (n = 9). The horizontal line at 57% 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 approximately 54.3% to 61.5%. The number of people for these sites varied from 26 to 225.

FIGURE 4.2: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN BMI% AT PAEDIATRIC SITES

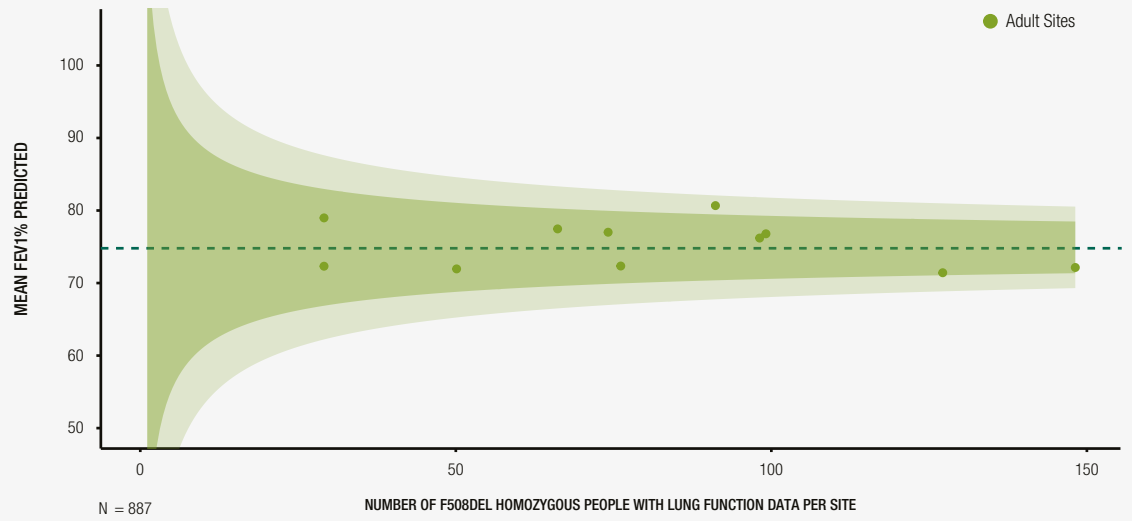


Shaded areas represent 95 and 99.8% control limits
Mean BMI percentile in all paediatric sites = 57%
Mean was caluclated across all the measurements
* Age, sex and height were standardised for when caluclating BMI predicted
Sites with fewer than 20 cases are not shown.

Adults: Variation in Lung Function

Figure 4.3 illustrates ACFDR clinical variation at adult sites, focusing on mean FEV1 pp (n = 11). 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 75%. Most sites fall within the expected variation. One site was removed from analysis due to low numbers.

FIGURE 4.3: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN FEV1 PP AT ADULT SITES

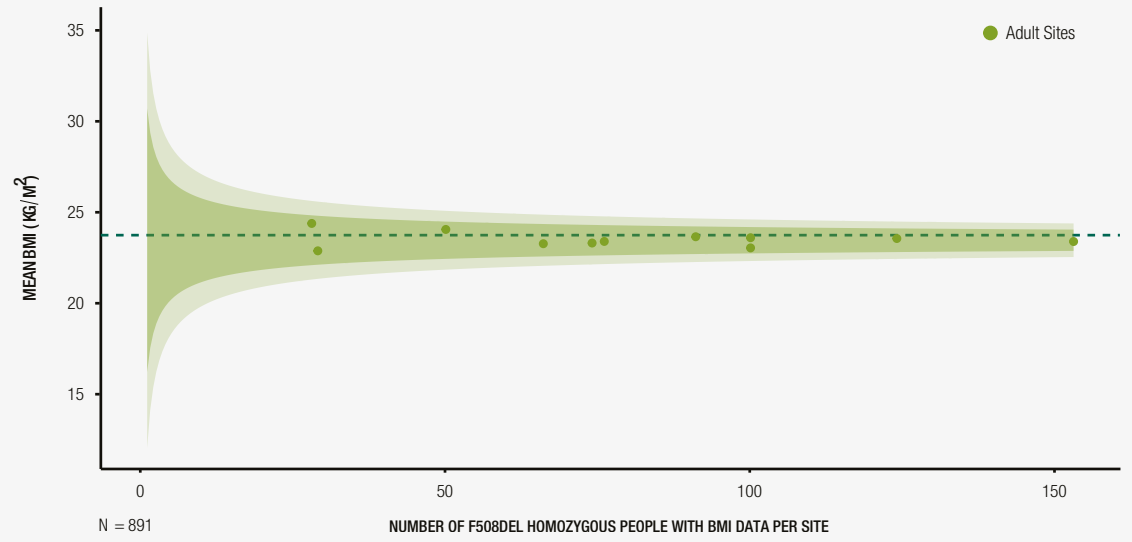


Shaded areas represent 95 and 99.8% control limits
Mean FEV1% predicted in all adult sites = 75%
Mean was caluclated across all the measurements
* Age, sex and height were standardised for when caluclating FEV1 % predicted
Sites with fewer than 20 cases are not shown.

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 = 11). The overall mean BMI across all adult sites is 24 kg/m^2. Most sites fall within the expected variation limits, with one site omitted from the analysis due to low numbers (Figure 4.4).

FIGURE 4.4: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN BMI% AT ADULT SITES

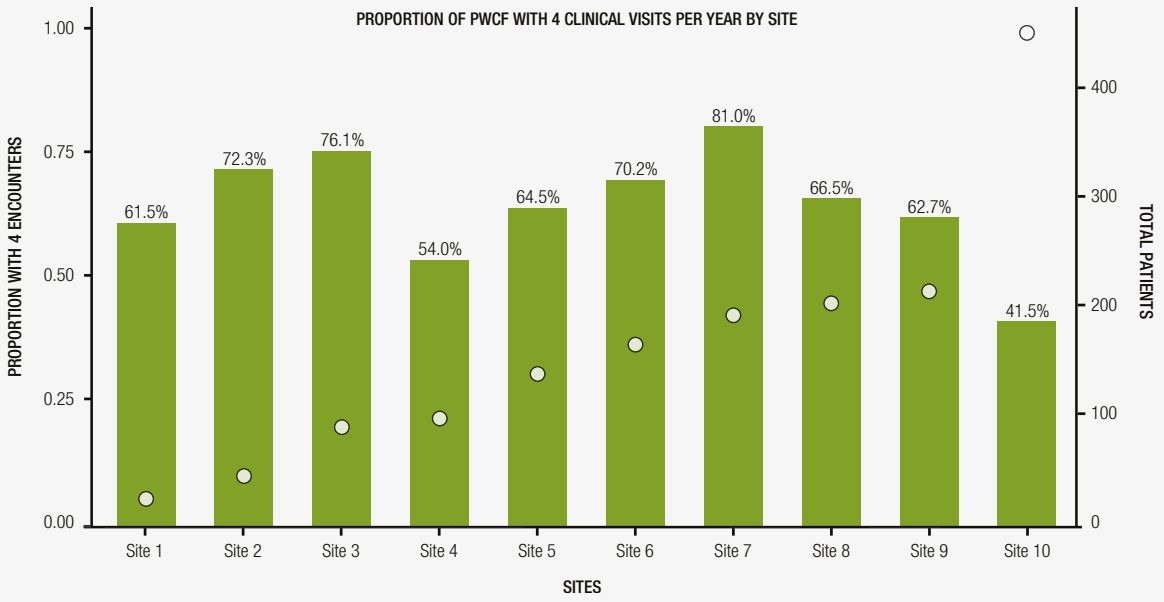


Shaded areas represent 95 and 99.8% control limits
Mean BMI in all adult sites = 24 kg/m^2
* Age, sex and height were standardised for when caluclating BMI predicted
Sites with fewer than 20 cases are not shown.

Paediatrics: Proportion of Individuals with four visits per year

The proportion of pwCF with 4 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 41.6% at Site 10 to a high of 81.0% at Site 7. Most sites have between 60-80% pwCF receiving 4 annual clinical visits. The white dots indicate the total number of pwCF at each site, with the scale shown at the right.

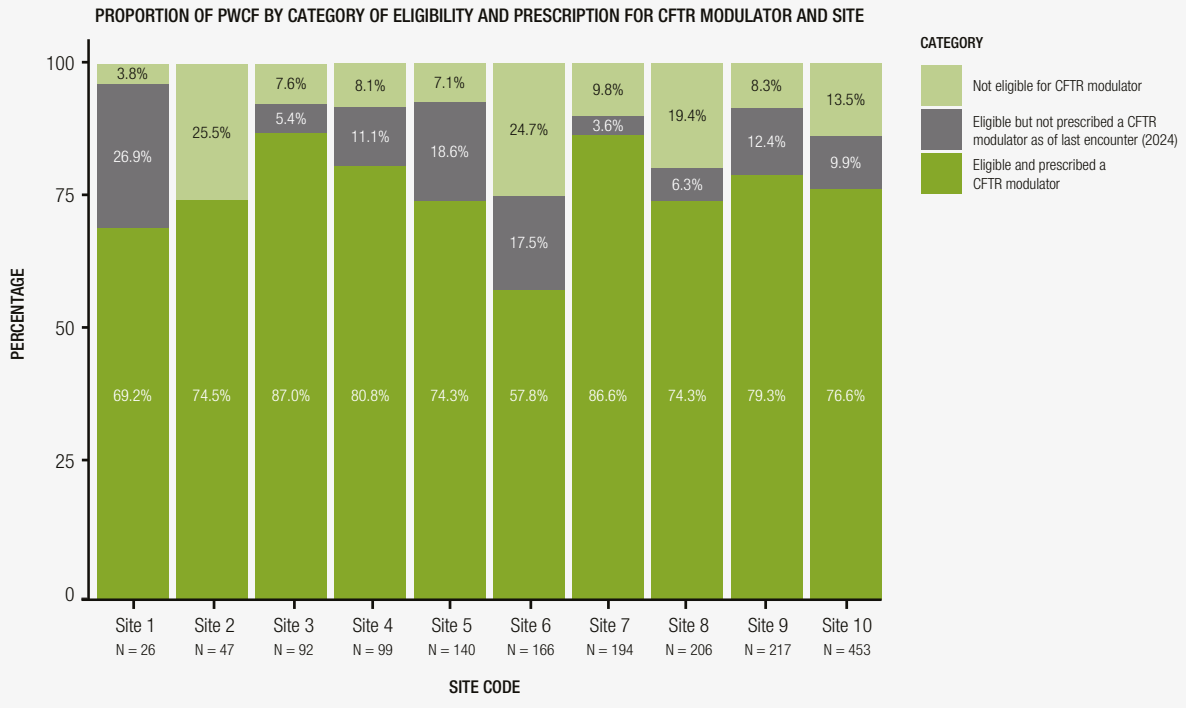
FIGURE 4.5: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: PAEDIATRIC SITES WITH 4 ANNUAL CLINICAL VISITS



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 57.8% (Site 6) to 86.6% (Site 7), with most sites falling between 70-85%. The proportion of pwCF who are not eligible for CFTR modulators ranges from 3.8% (Site 1) to 25.5% (Site 2). The proportion of pwCF at each site who are eligible but not prescribed a modulator vary from 0.0% (Site 2) to 26.9% (Site 1).

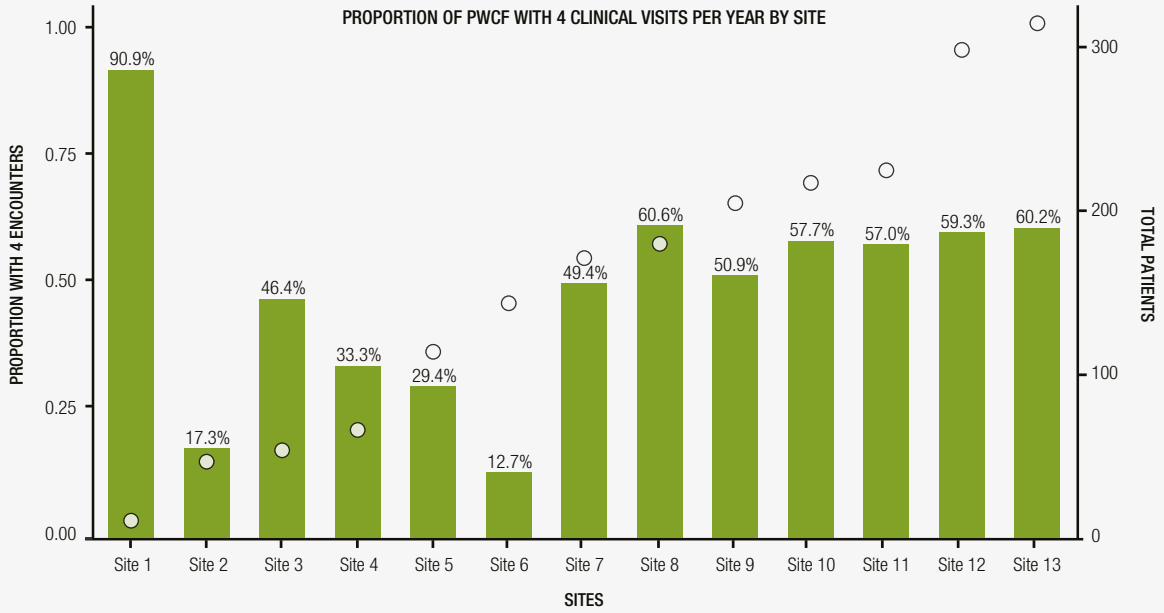
FIGURE 4.6: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: PROPORTION AT PEADIATRIC SITES THAT ARE ELIGIBLE AND PRESCRIBED A CFTR MODULATOR



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 12.7% at Site 5 to a high of 90.9% at Site 1. The majority of sites fall between 45% and 60%. 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.

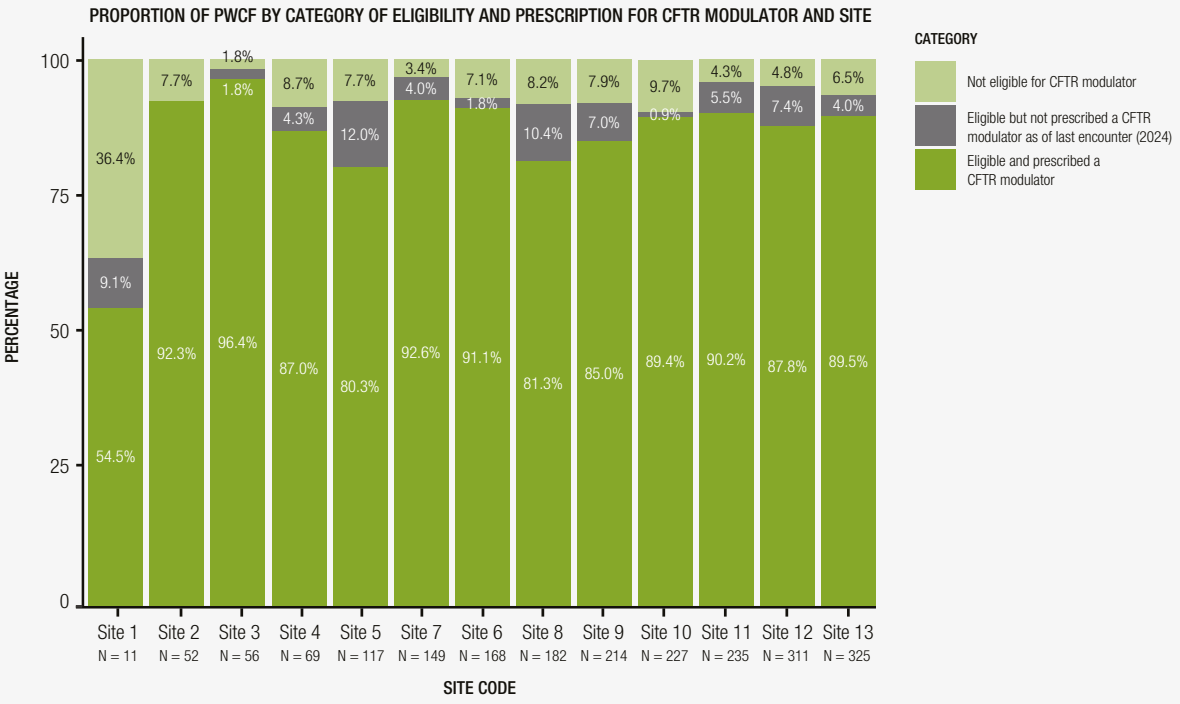
FIGURE 4.7: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: ADULT SITES WITH 4 ANNUAL CLINICAL VISITS



Adults: Proportion of eligible Individuals Prescribed a Modulator

The proportion of pwCF by category of eligibility and prescription for CFTR modulators across 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 54.5% (Site 1) to 96.4% (Site 3). The proportion of pwCF who are not eligible to be prescribed a modulator ranges from 1.8% (Site 3) to 36.4% (Site 1). A smaller proportion of pwCF are eligible but not prescribed a CFTR modulator, varying from 1.8% (Site 3) to 10.1% (Site 12).

FIGURE 4.8: ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: PROPORTION AT ADULT SITES THAT ARE ELIGIBLE AND PRESCRIBED A CFTR MODULATOR



5. 2024 ACADEMIC OUTPUTS

Publications

Semenchuk J, Naito Y, Charman SC, Carr SB, Cheng SY, Marshall BC et al. *Impact of COVID-19 infection on lung function and nutritional status amongst individuals with cystic fibrosis: A global cohort study.* Journal of Cystic Fibrosis. 2024 Sept;23(5):815-822. doi: 10.1016/j.jcf.2024.07.019

Conference Presentations

NACFC 2024, Boston, MA, 2024. Ruseckaite R, Wark P, Pourghaderi A, Caruso M, Ahern S. *Use of elexacaftor/tezacaftor/ivacaftor in Australians with cystic fibrosis: a registry-based study.* Poster presentation.

ACFC 2024, Brisbane, AUS, 2024. Ruseckaite R, Wark P, Pourghaderi A, Caruso M, Ahern S. *Tolerance of elexacaftor/tezacaftor/ivacaftor in Australians with cystic fibrosis.* Oral presentation.

ACFC 2024, Brisbane, AUS, 2024. Orre M, Bresnick K, Higgins M, Zahigian R, Chen Y-C, Pourghaderi A, Ruseckaite R. *Real-world outcomes associated with LUM/IVA treatment initiation in children with cystic fibrosis from Australia.*

ACFC 2024, Brisbane, AUS, 2024. Pourghaderi AR, Ahern S, Corda J, Earnest A, Garduce P, Mulrennan S, Ranganathan S, Ruseckaite R, Shanthikumar S, Dougas T. *Impact of telehealth on health outcomes in cystic fibrosis care across Australia.*

ABF 2024, Melbourne, AUS, 2024. Ruseckaite, R. *Clinical quality registries: using data to improve quality of care.* Oral presentation.

6. 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 2024.

Date	Name	Organisation	Request type	Request
22-Jan	Claire Wainwright	Queensland Children's Hospital	Non-research	A spatial distribution of individuals for presentation to the state government around providing state-wide services for CF.
24-Jan	Sean Beggs/ Rasa Ruseckaite	Royal Hobart Hospital/ Monash University	Research	Incorporating patient-reported outcome measures for children and adolescents into the Australian Cystic Fibrosis Data Registry
25-Jan	Sheila Sivam	Royal Prince Alfred Hospital	Research	Reducing the frequency of outpatient review in people with cystic fibrosis on high-efficacy modulator therapy (REFORM)
11-Mar	Charlotte Hawthorn	Children's Health QLD	Research	Anthropometric measures an CFTR modulator use at QCH
4-Apr	Maxine Orre	Vertex Pharmaceuticals	Non-research	Rare mutations
10-Apr	Arul Earnest/ Zemenu Tadesse Tessema	Monash University	Research	Bayesian spatio-temporal modelling of specific types of infection, poor lung function (FEV1%), and optimal BMI% among cystic fibrosis patients in Victoria and areal level determinants
3-May	Lisa Bayakly	Cystic Fibrosis WA	Non-Research	Paediatric and adult breakdowns of WA microbiology data
13-May	Petrina Fraccaro	Cystic Fibrosis QLD	Non-Research	Data request regarding CF people residing in QLD/Gladstone region
15-Jul	Maxine Orre	Vertex Pharmaceuticals	Non-research	CF patient population by age and mutation
17-Jul	David Armstrong	Monash Health	Research	Monash MC patients list to identify those who potentially have CFRD
19-Aug	Scott Bell/ Christine Duplancic	QIMR Berghofer Medical Research Institute	Research	The emerging problem of nontuberculous mycobacteria infection in people with cystic fibrosis
3-Oct	Darsy Darssan/ Brittnee Bryer	University of Queensland	Research	Spatial distribution of microbiological infections in people with Cystic Fibrosis and the relationship with risk factors, comorbidities, and treatments
13-Nov	Claire Wainwright	Queensland Children's Hospital	Non-research	Outcomes for QCH Cf service for CF service planning purposes 2024
14-Nov	Astrid Gardiner	Royal Prince Alfred Hospital/ University of Sydney	Research	NIV and Respiratory Failure in Cystic Fibrosis
18-Nov	Maxine Orre	Vertex Pharmaceuticals	Non-research	CF patient population: number of patients by age and weight range

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.

APPENDICES

LIST OF FIGURES

2.22:	ACFDR PAEDIATRICS 2024: STANDARD AND NON-STANDARD DOSE BY AGE AND MODULATOR
2.23:	ACFDR PAEDIATRICS 2024: MAINTENANCE ANTIBIOTIC THERAPY BY AGE GROUP
2.24A:	ACFDR PAEDIATRICS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN UNDER 6 YEARS
2.24B:	ACFDR PAEDIATRICS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN AGED 6-11 YEARS
2.24C:	ACFDR PAEDIATRICS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN AGED 12-17 YEARS
2.25A:	ACFDR PAEDIATRICS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN UNDER 6 YEARS
2.25B:	ACFDR PAEDIATRICS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN AGED 6-11 YEARS
2.25C:	ACFDR PAEDIATRICS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN AGED 12-17 YEARS
2.26A:	ACFDR PAEDIATRICS 2019-2024: DIABETIC STATUS FOR CHILDREN UNDER 6 YEARS
2.26B:	ACFDR PAEDIATRICS 2019-2024: DIABETIC STATUS FOR CHILDREN AGED 6-11 YEARS
2.26C:	ACFDR PAEDIATRICS 2019-2024: DIABETIC STATUS FOR CHILDREN AGED 12-17 YEARS
2.27:	ACFDR PAEDIATRICS 2022-2024: ANNUAL MULTIDISCIPLINARY CARE APPOINTMENTS BY AGE GROUP
2.28:	ACFDR PAEDIATRICS 2019-2024: VACCINATIONS BY AGE GROUP
3.1:	ACFDR ADULTS 2024: AGE AND SEX
3.2:	ACFDR ADULTS 2020-2024: HIGHEST EDUCATIONAL ATTAINMENT
3.3:	ACFDR ADULTS 2020-2024: EMPLOYMENT STATUS
3.4:	ACFDR ADULTS 2024: MARITAL STATUS
3.5:	ACFDR ADULTS 2018-2024: PREGNANCY
3.6:	ACFDR ADULTS 2018-2024: AVERAGE NUMBER OF LUNG FUNCTION TESTS ENTERED PER INDIVIDUAL
3.7:	ACFDR ADULTS 2024: FEV1 PP BY AGE
3.8:	ACFDR ADULTS 2024: MEDIAN FEV1 PP
3.9:	ACFDR ADULTS 2019-2024: MEDIAN FEV1 PP OVER TIME
3.10:	ACFDR ADULTS 2011-2024: FEV1 PP DISTRIBUTION OVER TIME
3.11:	ACFDR ADULTS 2024 : FEV1 PP VS BMI PERCENTILE
3.12:	ACFDR ADULTS 2024: BMI BY AGE
3.13:	ACFDR ADULTS 2024: BMI BY SEX
3.14:	ACFDR ADULTS 2008-2024: MEDIAN BMI
3.15:	ACFDR ADULTS 2019-2024: AVERAGE NUMBER OF RESPIRATORY SAMPLES PER INDIVIDUAL
3.16:	ACFDR ADULTS 2019-2024: PREVALENCE OF RESPIRATORY MICROBIOLOGY
3.17:	ACFDR ADULTS 2024: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE
3.18:	ACFDR ADULTS 2019-2024: PREVALENCE OF NTM INFECTIONS BY AGE
3.19:	ACFDR ADULTS 2021-2024: TOTAL NUMBER OF VISITS RECORDED PER YEAR
3.20:	ACFDR ADULTS 2024: PROPORTION OF PATIENTS WITH 4 OR MORE CLINICAL VISITS
3.21:	ACFDR ADULTS 2024: HOSPITALISATIONS BY AGE
3.22:	ACFDR ADULTS 2019-2024: HOSPITALISATIONS PER YEAR
3.23:	ACFDR ADULTS 2024: IV ANTIBIOTIC THERAPY BY AGE
3.24:	ACFDR ADULTS 2024: CFTR MODULATOR ELIGIBILITY AND PRESCRIBING BY AGE
3.25:	ACFDR ADULTS 2024: COMBINED ELIGIBILITY AND PRESCRIPTION OF CFTR MODULATORS
3.26:	ACFDR ADULTS 2024: STANDARD AND NON-STANDARD DOSE BY AGE AND MODULATOR
3.27A:	ACFDR ADULTS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR ADULTS AGED 18-29 YEARS
3.27B:	ACFDR ADULTS 2019-2024: MAINTENANCE ANTIBIOTIC THERAPY FOR ADULTS AGED 30 YEARS AND OLDER
3.28A:	ACFDR ADULTS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR ADULTS AGED 18-29 YEARS

3.28B:	ACFDR ADULTS 2020-2024: NON-ANTIBIOTIC LUNG THERAPIES FOR ADULTS AGED 30 YEARS AND OLDER
3.29A:	ACFDR ADULTS 2019-2024: DIABETIC STATUS FOR ADULTS AGED 18-29 YEARS
3.29B:	ACFDR ADULTS 2019-2024: DIABETIC STATUS FOR ADULTS AGED 30 YEARS AND OLDER
3.30:	ACFDR ADULTS 2022-2024: MULTIDISCIPLINARY CARE REVIEWS BY AGE
3.31:	ACFDR ADULTS 2019-2024: VACCINATIONS BY AGE
4.1:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN FEV1 PP AT PAEDIATRIC SITES
4.2:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN BMI% AT PAEDIATRIC SITES
4.3:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN FEV1 PP AT ADULT SITES
4.4:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: MEAN BMI% AT ADULT SITES
4.5:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: PAEDIATRIC SITES WITH 4 ANNUAL CLINICAL VISITS
4.6:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: PROPORTION AT PEADIATRIC SITES THAT ARE ELIGIBLE AND PRESCRIBED A CFTR MODULATOR
4.7:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: ADULT SITES WITH 4 ANNUAL CLINICAL VISITS
4.8:	ACFDR CLINICAL VARIATION AMONG CF CENTRES 2024: PROPORTION AT ADULT SITES THAT ARE ELIGIBLE AND PRESCRIBED A CFTR MODULATOR

LIST OF TABLES

ACFDR STEERING COMMITTEE MEMBERSHIP 2024

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 André Schultz	Deputy Lead/CF Paediatric Physician	Perth Children's Hospital, WA
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 Katherine Frayman	CF Paediatric Physician	Royal Children's Hospital, VIC
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
Honor Rose	Consumer Representative	VIC
Caz Boyd	Consumer Representative	WA
Sophie Worthington	Consumer Representative	WA
Penny Jones	Consumer Representative	NSW
Rebecca Edwards	Consumer Representative	NSW

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 & 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: 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 was 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]

SPONSORS

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395859 February 2026