



Port CF 2022

New Zealand National Data Registry

cf CYSTIC
FIBROSIS NZ

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The Port CF National Data Registry is a research project of Cystic Fibrosis NZ. For further information about Cystic Fibrosis NZ visit cfnz.org.nz

Source of Data:

Children, young persons and adults with cystic fibrosis in New Zealand who have consented to have their data recorded as part of the National Data Registry.

Suggested Citation:

Port CF National Data Registry, 2022 Registry Report, Cystic Fibrosis NZ. <http://cfnz.org.nz/>

Introduction

From the Chair of the Port CFNZ Steering Committee

Cystic Fibrosis NZ and the Port CF Steering Committee are pleased to present the National Data Registry 2022 Report containing data collected from children, young persons and adults with cystic fibrosis (CF) in New Zealand.

We would like to thank:

- The children and adults with CF and their families for participating in this process.
- Cystic Fibrosis NZ for providing funding for database analysis and data entry.
- The nurses, specialists and administrators who have worked to enter data, enabling a detailed analysis for New Zealand presented in this report.
- Health NZ (Canterbury) for their on-going information technology service to maintain the National Data Registry (Registry).

This twelfth Registry Report provides an increasingly accurate picture of people with CF and their outcomes for New Zealand, with 97-98% opting to provide this anonymised data, bringing the total number of people with CF (PWCF) included to 576.

The 2022 Registry Report continues to inform future care and the future resources needed.

We hope you find the information in this 2022 Report informative and useful.



Associate Professor Cass Byrnes

Chair Port CF
Port CFNZ Principal Investigator (2017 - 2021)



Lisa Burns

Chief Executive, Cystic Fibrosis NZ
November 2021 to October 2025

Report completed by:

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A special thanks to:

Andrew Watson, Health NZ (Canterbury), Alexia Searchfield



CF Clinics in New Zealand

Northland (Paediatrics)

Whangarei Hospital, Whangarei

Auckland (Paediatrics and Adults)

Starship Child Health
Greenlane Clinical Centre

Waikato (Paediatrics and Adults)

Waikato Hospital, Hamilton

Taranaki (Paediatrics and Adults)

Taranaki Base Hospital, New Plymouth

Bay of Plenty (Paediatrics and Adults)

Tauranga Hospital, Tauranga
Whakatane Hospital, Whakatane
Lakes Hospital, Rotorua

Central Districts (Paediatrics and Adults)

Palmerston North Hospital, Palmerston North

Hawkes Bay (Paediatrics and Adults)

Hawkes Bay District Hospital, Hastings
Tairāwhiti Hospital, Gisborne

Wellington (Paediatrics and Adults)

Capital and Coast Hospital, Wellington
Hutt Valley Hospital, Lower Hutt

Nelson/Marlborough (Paediatrics)

Nelson Hospital, Nelson
Wairau Hospital, Blenheim

Canterbury (Paediatrics and Adults)

Christchurch Hospital, Christchurch

Otago (Paediatrics and Adults)

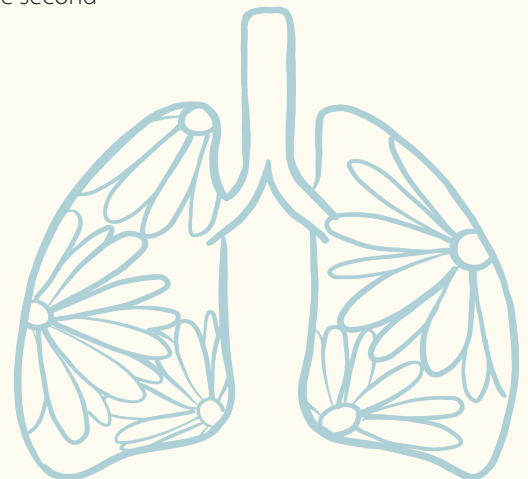
Dunedin Hospital, Dunedin

Southland (Paediatrics)

Kew Hospital, Invercargill

Glossary of Terms

CFNZ	Cystic Fibrosis NZ
FEV₁	Lung function measurement as forced expiratory volume in one second
FVC	Lung function measurement as the total forced vital capacity
BMI	Body Mass Index: measurement of weight relative to height
Median	Middle number in a numerically arranged range of numbers
Range	Upper and lower values in a dataset
Paediatric	Under 16 years of age
Adult	16 years and over
PWCF	Person with CF



Notes to the Registry

New Zealand has a total CF population comparative to a single clinic in the United States (USA) or the United Kingdom (UK). The data collected from our CF population provides our national statistics. Our smaller population provides significant challenges to statistical interpretation as outliers, in terms of late diagnoses and key markers, will have an impact on outcomes reported.

The brief commentary provided throughout this report reflects opinions based on our data and, when cited as compared to other registries, these are from Australia, the UK and the USA. Although we have a total of 576 registered in Port CF, not all individuals had an input for all categories. While the total is 576 (231 children under 16 years of age and 345 adults 16 years and over) the number of PWCF shown at the top of each table or figure is the total number with a response for that category.

New Zealand Registry data is increasingly robust and accurate and **we welcome its use in audit and research projects by researchers from reputable institutions.** Enquiries regarding the use of data can be made to either the CFNZ Chief Executive or the Project Co-ordinator.

Project Co-ordinator:

data@cfnz.org.nz

OR

Chief Executive:

ceo@cfnz.org.nz



Key Indicators

Key Indicators								576 PWCF
	2022	2021	2020	2019	2018	2017	2016	2015
CF Patients Registered	576	577	572	531	514	498	501	449
Diagnosis								
Diagnosis age <1 year	7	14	15	11	15	15	6	5
Diagnosis age <16 years	1	4	4	1	0	2	3	
Diagnosis age ≥16 years	1	3	0	2	0	1	2	0
Age								
Median Age (in years)	20	19.0	19	18.87	18.54	18.27	17.38	18.25
Mean Age (in years)	22.4	21.7	21.6	21.49	21.2	20.79	20.04	
PWCF <16 years								
Number	231	239	236	223	224	279	233	192
Percent	40.1%	41.4%	41.3%	42%	43.6%	56.0%	46.5%	42.8%
PWCF ≥16 years								
Number	345	338	336	308	290	219	268	257
Percent	59.9%	58.6%	58.7%	58%	56.4%	44.0%	53.5%	57.2%
Gender								
Males								
Number	321	321	319	297	285	273	275	247
Percent	55.7%	55.6%	55.8%	56%	55.4%	54.9%	54.9%	55.0%
Females								
Number	255	256	253	233	229	224	226	202
Percent	44.3%	44.4%	44.2%	44%	44.6%	45.1%	45.1%	45.0%
Genotyped								
Number	576	577	572	495	466	484	450	400
Percent	100.0%	100%	100.0%	93.2%	90.7%	97.4%	90.0%	89.1%
FEV1 (% predicted)								
Mean	81.4%	79.8	78.4%	76.6%	81.8	85.1%	85.0%	
Median	85.2%	84.8%	82.8%	79%	86.2	86.5%	88.4%	85.6%
FEV1 < 16 Years								
Mean	93.1%	94.0%	91.4%	95.8%	96.70%	96.8%	97.3%	
Median	96.2%	95.2%	93.7%	97.9%	98.80%	99.3	99.3%	98.9%
FEV1 ≥16 Years								
Mean	75.6%	72.5%	71.7%	74.7%	75.30%	72.60%	72.6%	
Median	76.9	75.8%	74.2%	76.8%	79.20%	77.4	77.4%	77.0%
FEV1 < 18 Years								
Mean	92.2	91.8%	89.7%	94.5%	95.40%	95.1%	95.0%	
Median	95.6	93.8%	93.1%	97.2%	98.30%	98.3%	98.0%	
FEV1 ≥18 Years								
Mean	74.3	77.0%	70.9%	73.1%	73.7%	72.2%	71.2%	
Median	74.82	75.6%	72.7%	74.7%	77.6%	75.6%	75.1%	

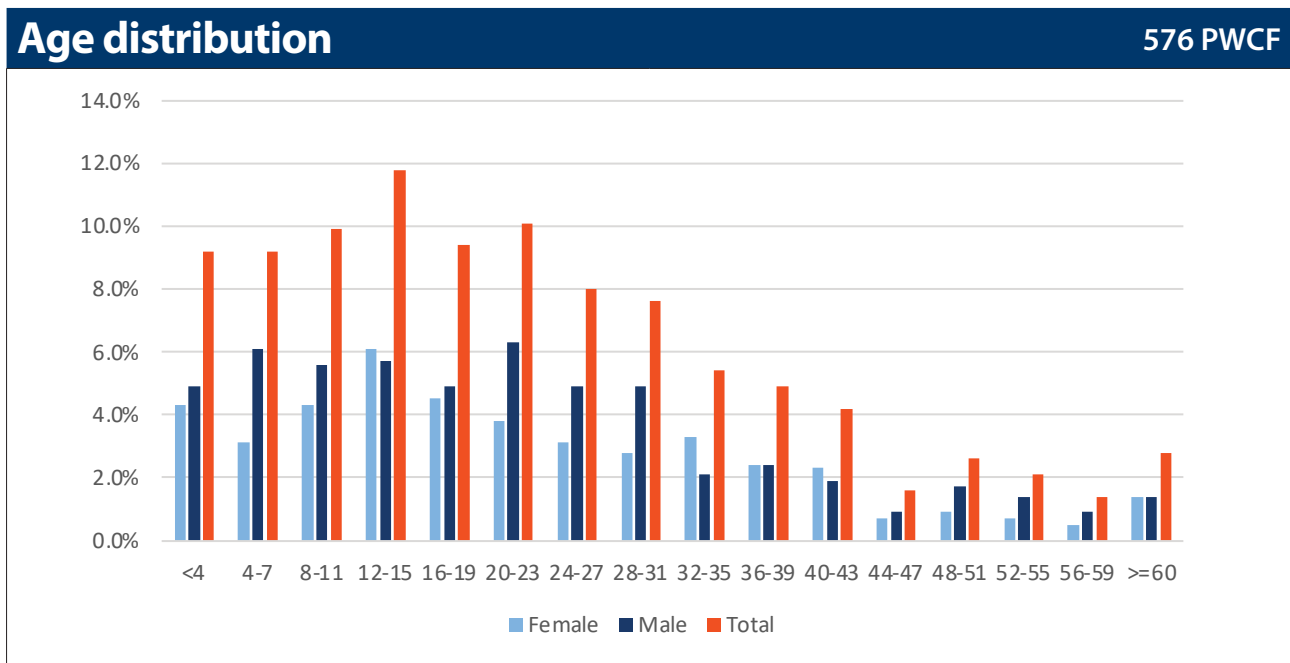
Key Indicators cont.

Key Indicators continued								
	2022	2021	2020	2019	2018	2017	2016	2015
FVC (% predicted)								
Mean	97.0%	94.7%	93.4%					
Median	100.4%	98.8%	96.0%					
FVC < 16 Years								
Mean	99.3%	100.5%	97.0%					
Median		101.4%	98.3%					
FVC ≥ 16 Years								
Mean	95.9%	91.7%	91.5%					
Median	101.2	95.9%	95.1%					
FVC < 18 Years								
Mean	99.7	99.5%	96.9%					
Median	95.2	101.3%	98.3%					
FVC ≥ 18 Years								
Mean	95.2	91.5%	91.1%					
Median	100.8	95.5%	93.7%					

FEV₁ is still increasing a little on previous years in the adult group but is static in the paediatric group. We now have three years presenting FVC which has gone up as a mean overall, but largely reflects a slight increase in the adult population.

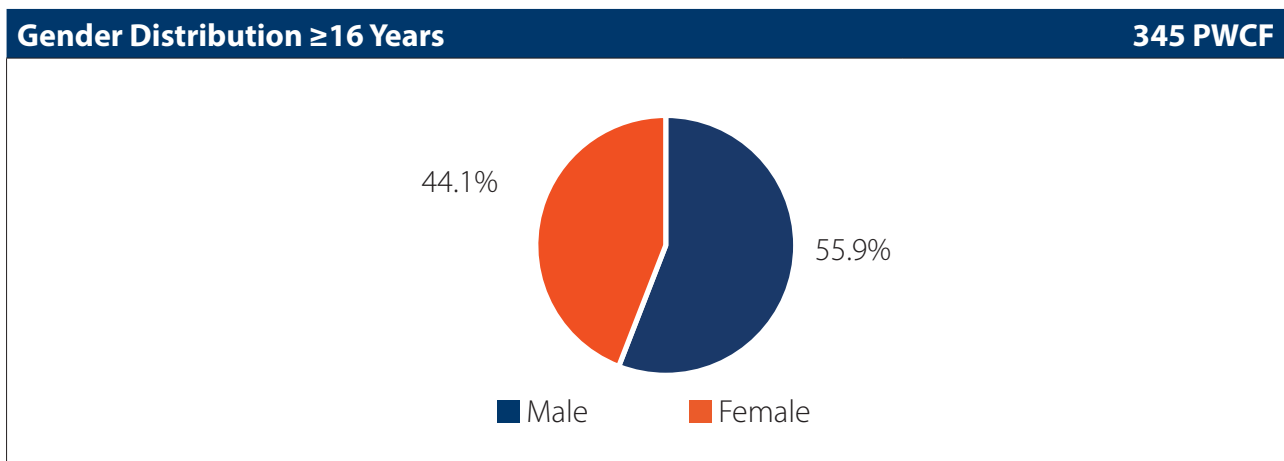
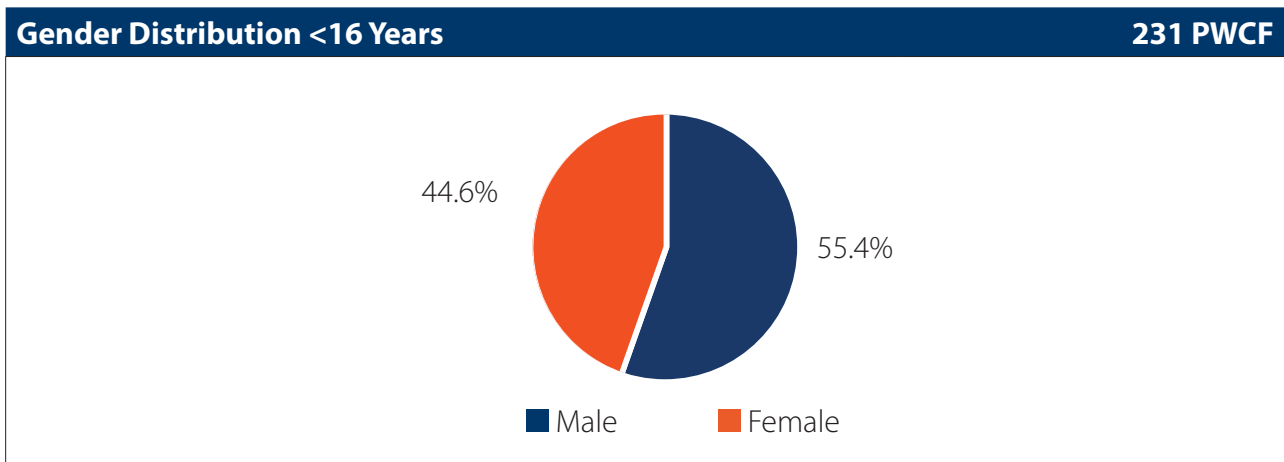
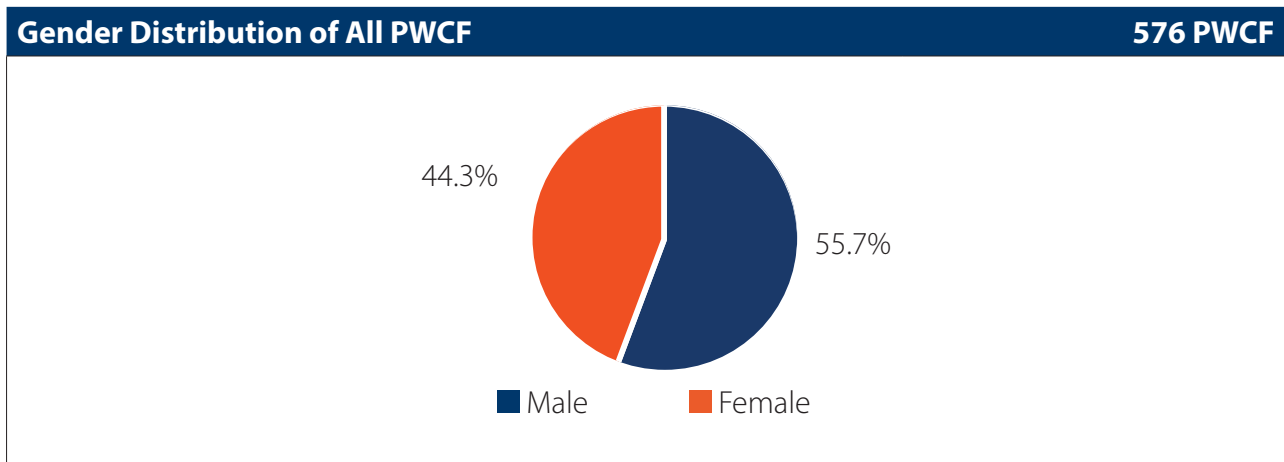
1. Demographics

1.1 Age distribution							576 PWCF
Age Group	All		Male		Female		
0-3	53	9.2%	28	4.9%	25	4.3%	
4-7	53	9.2%	35	6.1%	18	3.1%	
8-11	57	9.9%	32	5.6%	25	4.3%	
12-15	68	11.8%	33	5.7%	35	6.1%	
16-19	54	9.4%	28	4.9%	26	4.5%	
20-23	58	10.1%	36	6.3%	22	3.8%	
24-27	46	8.0%	28	4.9%	18	3.1%	
28-31	44	7.6%	28	4.9%	16	2.8%	
32-35	31	5.4%	12	2.1%	19	3.3%	
36-39	28	4.9%	14	2.4%	14	2.4%	
40-43	24	4.2%	11	1.9%	13	2.3%	
44-47	9	1.6%	5	0.9%	4	0.7%	
48-51	15	2.6%	10	1.7%	5	0.9%	
52-55	12	2.1%	8	1.4%	4	0.7%	
56-59	8	1.4%	5	0.9%	3	0.5%	
≥60	16	2.8%	8	1.4%	8	1.4%	
Total	576	100.0%	321	55.7%	255	44.3%	
Median	20						
Range	0 - 80						



The median age of persons with CF in New Zealand has increased from 16 to 20 years over the twelve years that we have had Registry data. Children in international registries are defined as either up to 16 years or up to 18 years of age. In New Zealand, if we include children as being up to 16 years, we have 231 children (40.1% total) and 345 adults (59.9% total). If we include children as being up to 18 years, which is our more usual clinical practice, we have 258 children (44.8% total) and 318 adults (55.2% total).

1.2 Gender Distribution						576 PWCF
	All		<16		≥16	
	Number in age	Percent of all PWCF	Number in age group	Percent of all PWCF	Number in age group	Percent of PWCF
Male	321	55.7%	128	22.2%	193	33.5%
Female	255	44.3%	103	17.9%	152	26.4%
Totals	576	100.0%	231	40.1%	345	59.9%



“The median age of persons with CF in New Zealand has increased from 16 to 20 years over the twelve years that we have had National Registry data.”



2. Genotypes

2.1 Number and Percent Genotyped			576 PWCF
Mutations	Number of PWCF Genotyped	Percentage of PWCF Genotyped	
Homozygous F508del	263	45.7%	
Heterozygous F508del	241	41.9%	
No F508del	72	12.5%	
Total	577		

2.2 Second Allele of Heterozygous F508del			241 PWCF
Second Allele	c.DNA Name	Number of PWCF	Percent of PWCF
R117H	c.350G>A	29	5.0%
G551D	c.1652G>A	25	4.3%
G542X	c.1624G>T	20	3.5%
3849+10kbC->T	c.3717+12191C>T	9	1.6%
N1303K	c.3909c>G	8	1.4%
1898+1G->A	c.1766+1G>A	4	0.7%
G85E	c.254G>A	4	0.7%
3659delC	c.3528delC	4	0.7%
^1507	c.1519_1521delATC	4	0.7%
D1152H	c.3454G>C	4	0.7%
621+1G->T	c.489+1G>T	3	0.5%
R560T	c.1679G>C	3	0.5%
Q493X	c.1477C>T	3	0.5%
2789+5G->A	c.2657+5G>A	3	0.5%
1717-1G->A	c.1585-1G>A	2	0.3%
R352Q	c.1055G>A	2	0.3%
Y563D	c.1687T>G	2	0.3%
A455E	c.1364C>A	2	0.3%
1078delT	c.948delT	2	0.3%
E60X	c.178G>T	2	0.3%
L206W	c.617T>G	1	0.2%
R334W	c.1000C>T	1	0.2%
P574H	c.1721C>A	1	0.2%
3120+1G->A	c.2988+1G>A	1	0.2%
R347H	c.1040G>A	1	0.2%
R347P	c.1040G>C	1	0.2%
W1282X	c.3846G>A	1	0.2%
1898+5G->T	c.1766+5G>T	1	0.2%
D1270N	c.3808G>A	1	0.2%
R117C	c.349C>T	1	0.2%
R1162X	c.3484C>T	1	0.2%
Other genetic mutation		88	15.3%
Not identified		7	1.2%

Note: Because people have two genes, they are counted twice, once for each gene.

Our high percentage of F508del is in keeping with the international registries from European derived populations. In total, only 72 persons in New Zealand do not have at least one F508 mutation. Looking at the gene mutations recorded in 2022 Registry data, 47 of the 576 people who have been genotyped (8.2%) would not be detected by the current new born screening programme.

2.3 No F508del Mutations

72 PWCF

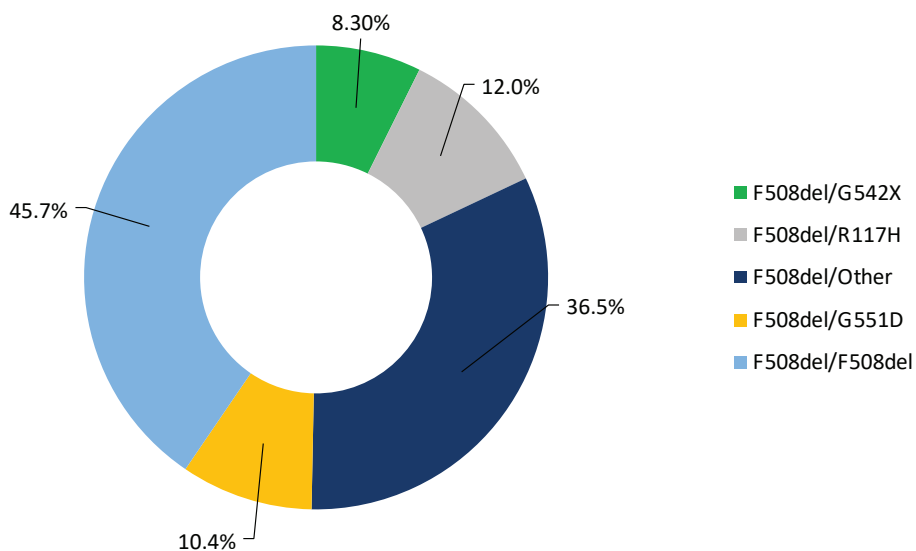
	1717-1G->A	2143delT	3849+10kbC->T	621+1G->T	G542X	G551D	Not Identified	Other	Q493X	R1162X	R117H	R553X	W1282X	ΔI507
1078delT	0	0	0	0	0	0	0	1	0	0	0	0	0	0
1161delC	0	0	0	0	0	0	0	2	0	0	0	0	0	0
1898+1G->A	0	0	0	0	0	0	0	1	0	0	0	0	0	0
2789+5G->A	0	0	0	0	0	0	0	1	0	0	0	0	0	0
3120+1G->A	0	0	0	0	0	0	0	2	0	0	0	0	0	0
3849+10kbC-	1	0	0	0	0	0	0	0	0	0	0	0	0	0
621+1G->T	0	0	0	0	0	0	0	1	0	0	0	0	0	0
A455E	0	0	0	0	0	0	0	1	0	0	0	0	0	0
G178R	0	0	0	0	0	0	0	1	0	0	0	0	0	0
G542X	0	1	0	1	1	0	1	0	0	0	2	0	0	0
G551D	0	0	1	0	1	1	1	4	4	0	5	1	0	0
G85E	0	0	0	0	0	0	0	1	0	0	0	0	0	0
N1303K	0	0	0	0	0	0	0	1	0	0	0	0	0	0
Not Identified	0	0	0	0	0	0	7	0	0	0	0	0	0	0
Other	0	0	0	0	0	0	3	11	0	0	0	0	0	0
R1162X	0	0	0	0	0	0	0	1	0	1	0	0	0	0
R117H	0	0	0	0	0	0	3	1	1	0	0	0	1	0
R553X	0	0	0	0	0	0	0	1	0	0	0	0	0	0
S549N	0	0	0	0	0	0	1	0	0	0	0	0	0	0
W1282X	0	0	0	0	0	0	1	2	0	0	0	0	0	1

2.4 Genotype Major Categories

Mutations	Number	Percentage of all PWCF
F508del	504	87.5%
G551D	43	7.5%
R117H	41	7.1%
G542X	27	4.7%

Note: Because people have two genes they are counted twice, once for each gene. The total number of PWCF is 576

2.5 Common Genotype Combinations

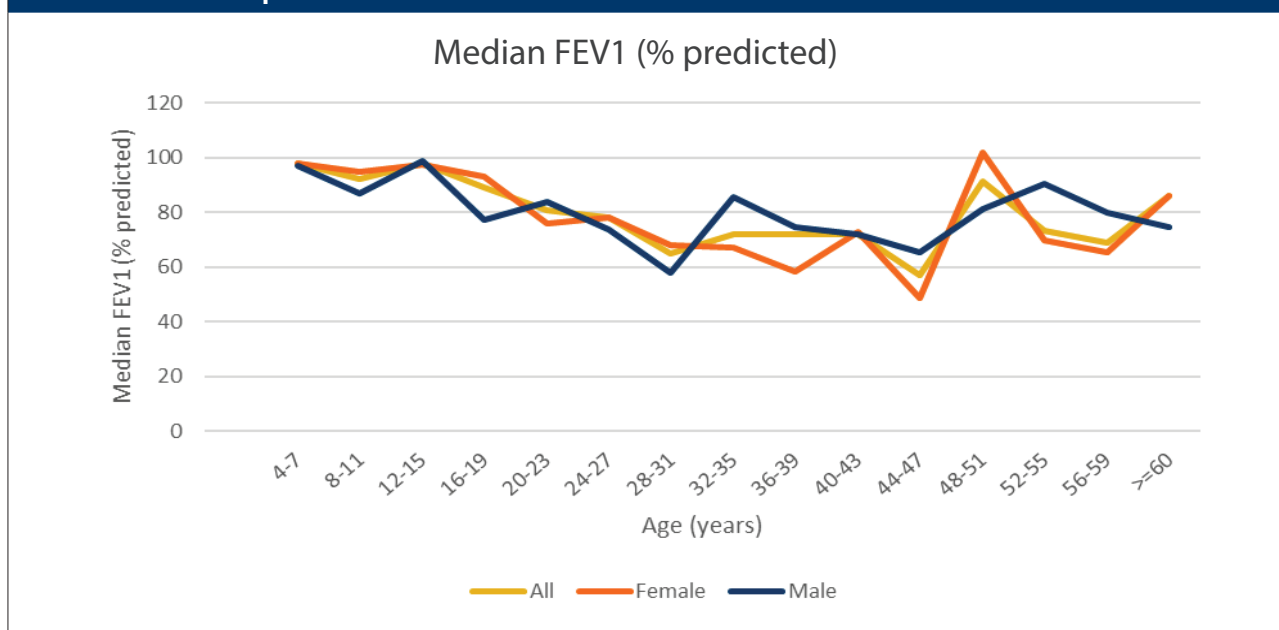


3. Respiratory

3.1 Median FEV₁ 413 PWCF

Age Group	All		Female		Male	
	Number in age group	Median FEV ₁	Number in age group	Median FEV ₁	Number in age group	Median FEV ₁
4-7	31	97.8	21	97.8	10	97.2
8-11	54	92.1	30	94.8	24	86.9
12-15	62	97.8	29	97.3	33	98.7
16-19	48	89.1	25	92.9	23	77.3
20-23	46	80.8	30	75.7	16	83.8
24-27	38	78.0	26	78.1	12	73.9
28-31	29	64.9	20	68.0	9	57.9
32-35	22	71.9	9	67.2	13	85.7
36-39	18	72.1	8	58.3	10	74.7
40-43	18	71.8	8	72.9	10	71.8
44-47	7	57.0	3	48.6	4	65.5
48-51	12	91.3	7	101.7	5	81.0
52-55	8	73.1	6	69.8	2	90.5
56-59	7	68.8	4	65.5	3	80.0
≥60	13	86.1	6	86.1	7	74.7
Totals	413		232		181	

Median FEV₁ 413 PWCF



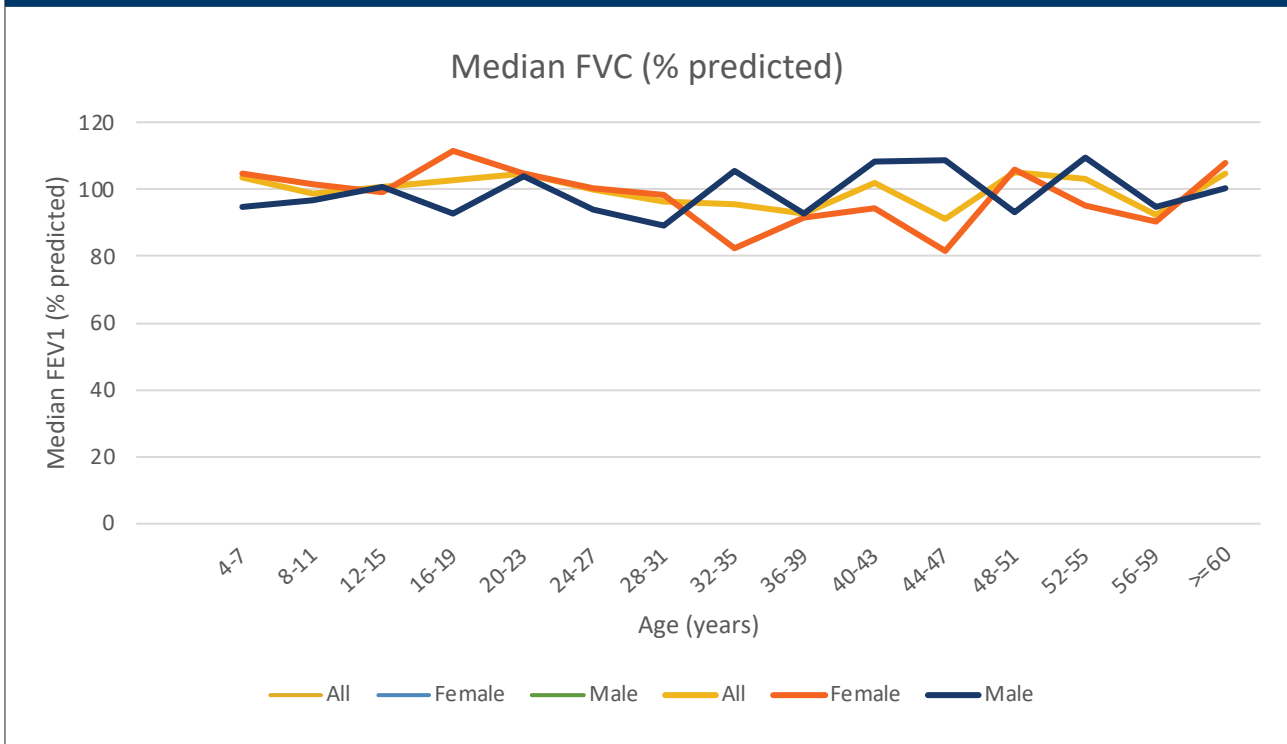
3.2 Median FVC

413 PWCF

Age Group	All		Female		Male	
	Number in age group	Median FVC	Number in age group	Median FVC	Number in age group	Median FVC
4-7	31	103.5	21	104.5	10	94.9
8-11	54	98.7	30	101.4	24	96.9
12-15	62	100.6	29	99.0	33	100.9
16-19	48	102.8	25	111.4	23	92.6
20-23	46	104.6	30	104.6	16	103.9
24-27	38	99.8	26	100.5	12	93.9
28-31	29	96.4	20	98.4	9	89.0
32-35	22	95.6	9	82.2	13	105.4
36-39	18	92.9	8	91.5	10	92.9
40-43	18	101.8	8	94.3	10	108.1
44-47	7	91.2	3	81.4	4	108.8
48-51	12	105.0	7	106.0	5	93.1
52-55	8	103.3	6	95.2	2	109.7
56-59	7	92.5	4	90.3	3	94.8
≥60	13	104.7	6	108.0	7	100.4
Totals	413		232		181	

Median FVC

413 PWCF



The median FEV1 of the population able to do lung function has always been 80% predicted since we started the Registry and the median this year is 85.2% (96.2% in children, 76.9% in adults). This necessarily excludes very young children who are unable to do lung function or those that find it very difficult because of technique or severity of disease. FVC is less impacted by early CF lung disease.



4. Nutrition

4.1 Paediatric BMI									195 PWCF
All <16 Years			Female <16 Years			Male <16 Years			
BMI Percentile			BMI Percentile			BMI Percentile			
Age Group	Number in group	Median BMI	Age Group	Number in group	Median BMI	Age Group	Number in group	Median BMI	
<4	26	44.4	<4	11	55	<4	15	36.4	
4-7	50	68.6	4-7	18	61.2	4-7	32	69.4	
8-11	55	45.5	8-11	25	44.5	8-11	30	47.4	
12-15	64	54.8	12-15	33	59.0	12-15	31	47.4	
Totals	195			87			108		

4.2 Adult BMI									261 PWCF
All ≥16 Years			Female ≥16 Years			Male ≥16 Years			
BMI			BMI			BMI			
Age Group	Number in group	Median BMI	Age Group	Number in group	Median BMI	Age Group	Number in group	Median BMI	
16-19	45	20.2	16-19	20	19.9	16-19	25	20.5	
20-23	46	22.9	20-23	16	21.9	20-23	30	23.4	
24-27	38	23	24-27	12	24.4	24-27	26	22.8	
28-31	29	21.2	28-31	9	21.0	28-31	20	21.2	
32-35	21	23.5	32-35	12	23.2	32-35	9	24.9	
36-39	18	22.3	36-39	10	22.2	36-39	8	24.1	
40-43	18	23.6	40-43	10	22	40-43	8	24.7	
44-47	7	23	44-47	4	22.7	44-47	3	24.2	
48-51	12	25.1	48-51	5	23.2	48-51	7	25.8	
52-55	8	2	52-55	2	35.7	52-55	6	25.7	
56-59	6	3	56-59	3	25.8	56-59	3	22.6	
≥60	13	7	≥60	7	22.5	≥60	6	25.1	
Totals	261			110			151		

The relationship between nutrition, lung function and survival in CF is well established with normal body weight associated with better preservation of lung function. The Cystic Fibrosis Foundation (USA) suggest the following targets for optimal weight status:

- 1) Infants (0 to 24 months): weight-for-length ≥50th percentile using WHO growth charts
- 2) Children and adolescents (2-18 years): BMI 50-85th percentile (CDC growth charts) or 50-91st percentile (WHO growth charts)
- 3) Adults: males BMI 23 - 27 kg/m², females BMI 22 - 27 kg/m²

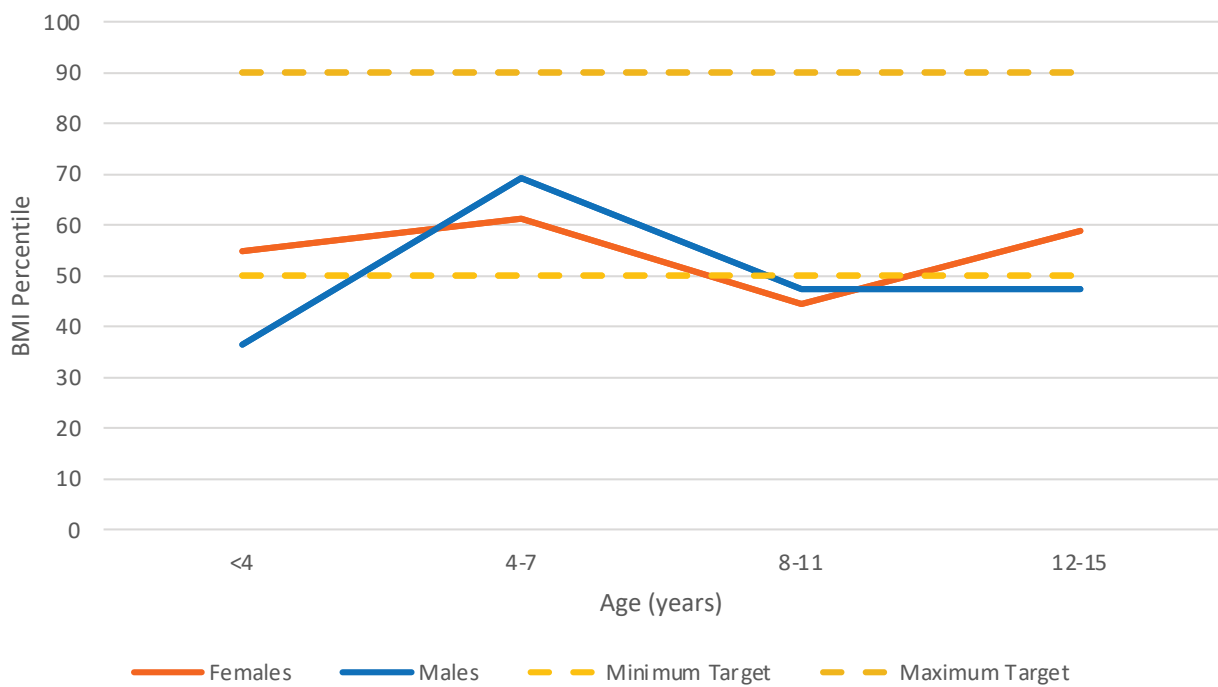
This is also reflected in the **Nutrition Guidelines for Cystic Fibrosis in Australia and New Zealand** (<https://www.thoracic.org.au/documents/item/1045>)

For infants under 4 years of age, the median BMI is 44.4 percentile. For children and adolescents, the median BMI is 55.2 percentile. For adults' 46.4% of males and 50% of females are above the minimum target range.

Similar to 2020 and 2021, infants and children, particularly male, appear to have had a decrease in their BMI which remains unexplained.

4.3 Median BMI Percentile <16 Years

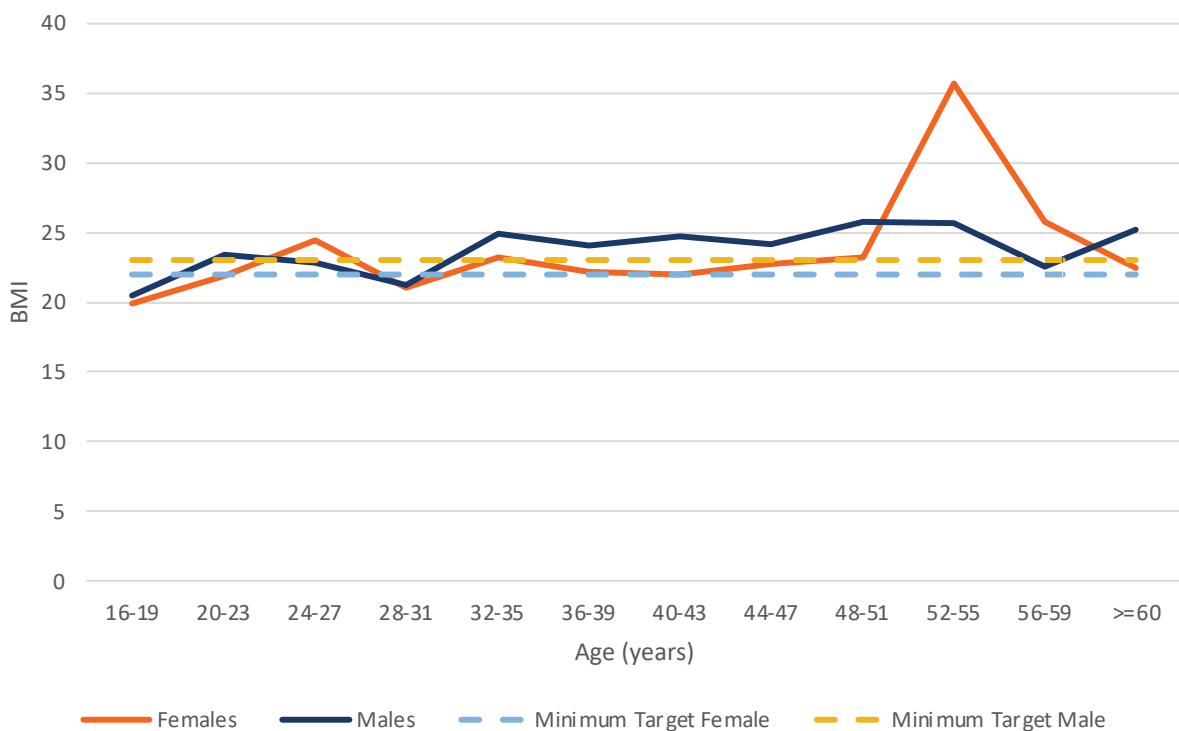
195 PWCF



The optimal BMI for children 2 - 16 is 50 - 91 percentile using the WHO-NZ growth chart. The dotted yellow lines shows the target range.

4.4 Median BMI ≥16 Years

261 PWCF



The optimal BMI for women is 22 - 27 and the dotted yellow line shows the minimum BMI for women in the target range. The optimal BMI for men is 23 - 27. The dotted blue line shows the minimum BMI for men in the target range.

4.5 Supplemental Feeding

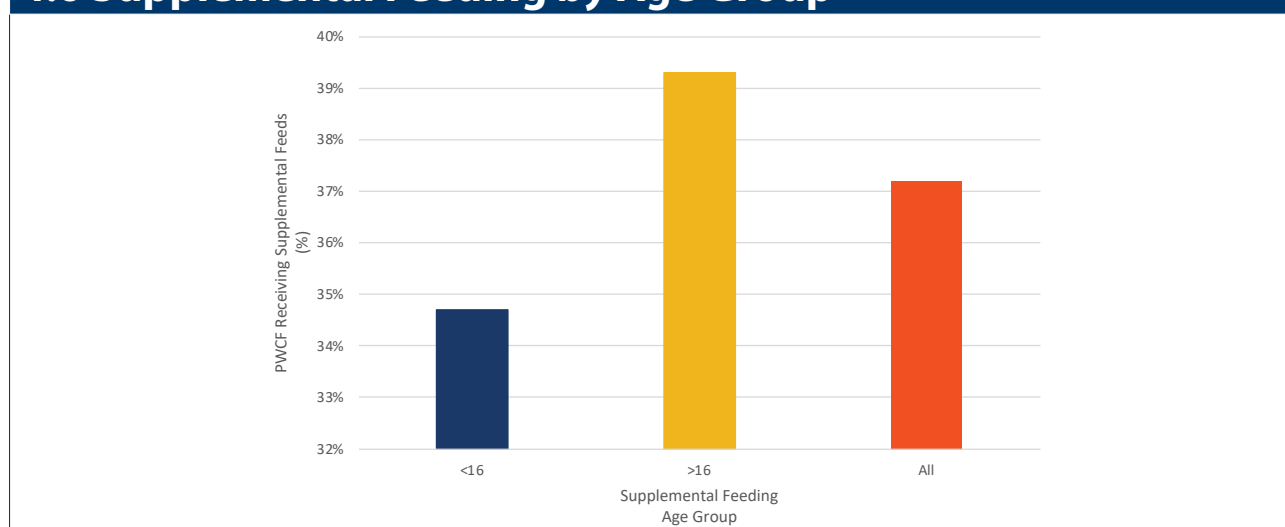
494 PWCF

	<16 years, n = 224	
	Yes	Percent supplemented
Supplemental Feeding Total	76	34.7%
Nasogastric	11	5.0%
Gastrostomy	9	4.1%
Oral	69	31.5%
	≥16 years, n = 270	
	Yes	% ≥16 years supplemented
Supplemental Feeding Total	106	39.3%
Nasogastric	4	1.5%
Gastrostomy	15	5.6%
Oral	95	35.2%

Some individuals may be on more than one type of supplemental feeding.

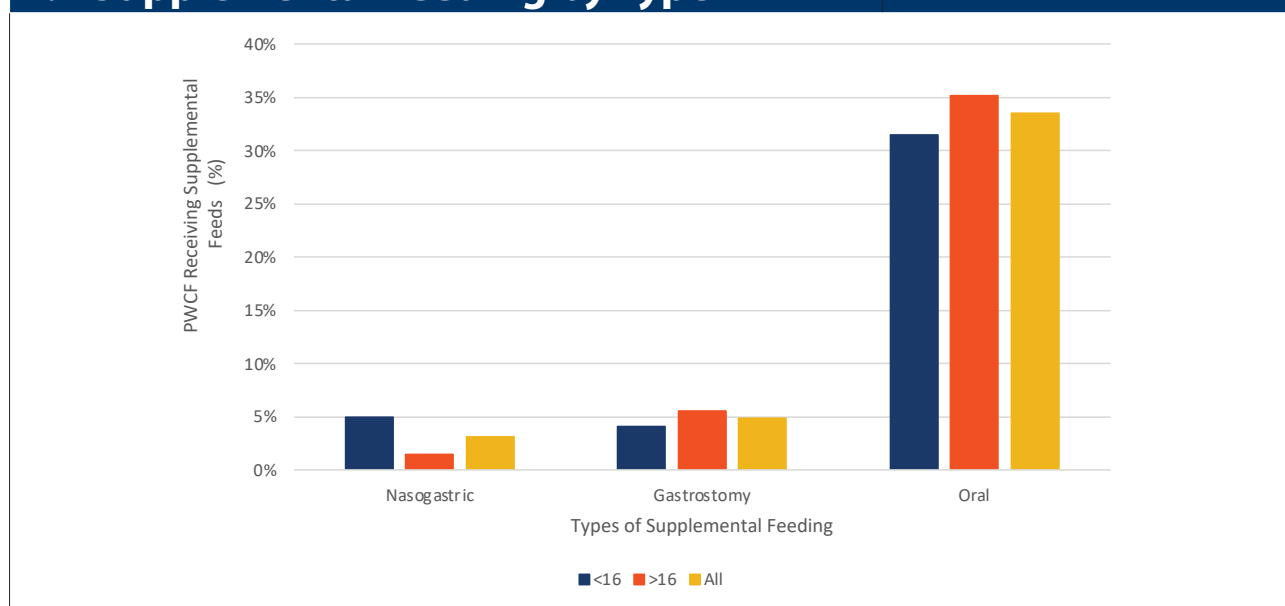
4.6 Supplemental Feeding by Age Group

494 PWCF



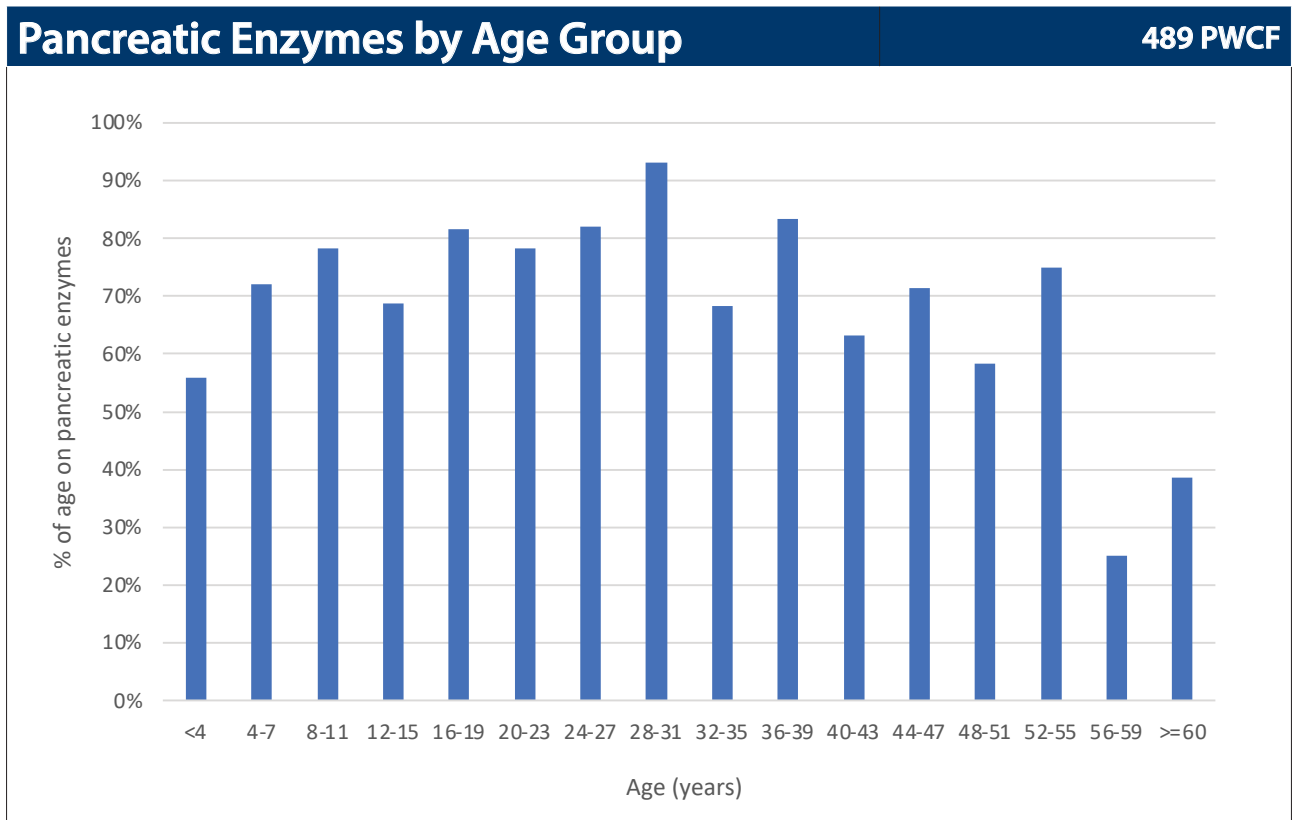
4.7 Supplemental Feeding by Type

494 PWCF



5. Pancreatic Enzymes

5.1 Pancreatic Enzymes by Age Group				489 PWCF
Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of PWCF
<4	50	28	56.0%	5.7%
4-7	50	36	72.0%	7.4%
8-11	55	43	78.2%	8.8%
12-15	64	44	68.8%	9.0%
16-19	49	40	81.6%	8.2%
20-23	46	36	78.3%	7.4%
24-27	39	32	82.1%	6.5%
28-31	29	27	93.1%	5.5%
32-35	22	15	68.2%	3.1%
36-39	18	15	83.3%	3.1%
40-43	19	12	63.2%	2.5%
44-47	7	5	71.4%	1.0%
48-51	12	7	58.3%	1.4%
52-55	8	6	75.0%	1.2%
56-59	8	2	25.0%	0.4%
≥60	13	5	38.5%	1.0%
Totals	489	353		72.2%



Of the 489 PWCF for whom there is data, 353 are on pancreatic enzymes and 136 are not



6. Airway Clearance Techniques

6.1 Primary Airway Clearance

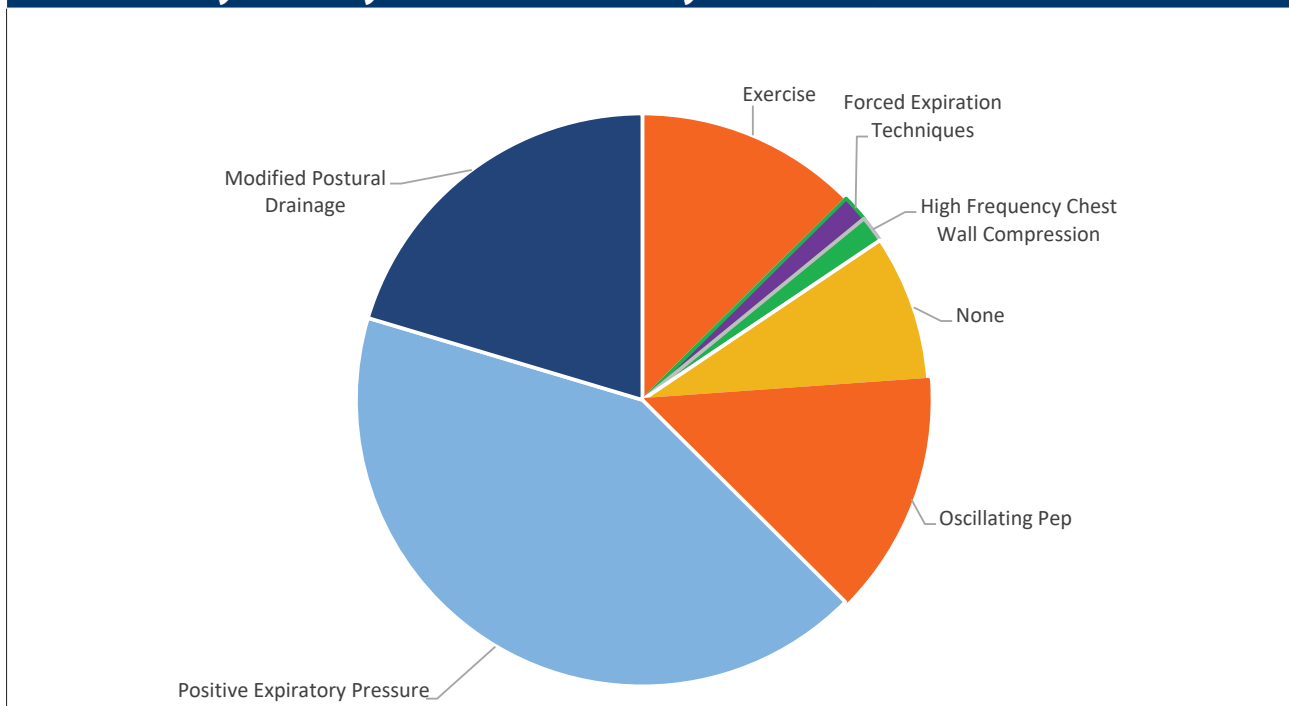
489 PWCF

	<16 years, n = 219	
	Number of PWCF	Percent of PWCF
Positive Expiratory Pressure	87	42.2%
Modified Postural Drainage	42	20.4%
Exercise	26	12.6%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	28	13.6%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	3	1.5%
High Frequency Chest Wall Compression (e.g. vest)	3	1.5%
None	17	8.3%

	≥16 years, n = 270	
	Number of PWCF	Percent of PWCF
Positive Expiratory Pressure	113	43.1%
Modified Postural Drainage	1	0.4%
Exercise	79	30.2%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	38	14.5%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	22	8.4%
High Frequency Chest Wall Compression (e.g. vest)	0	0.0%
None	9	3.4%

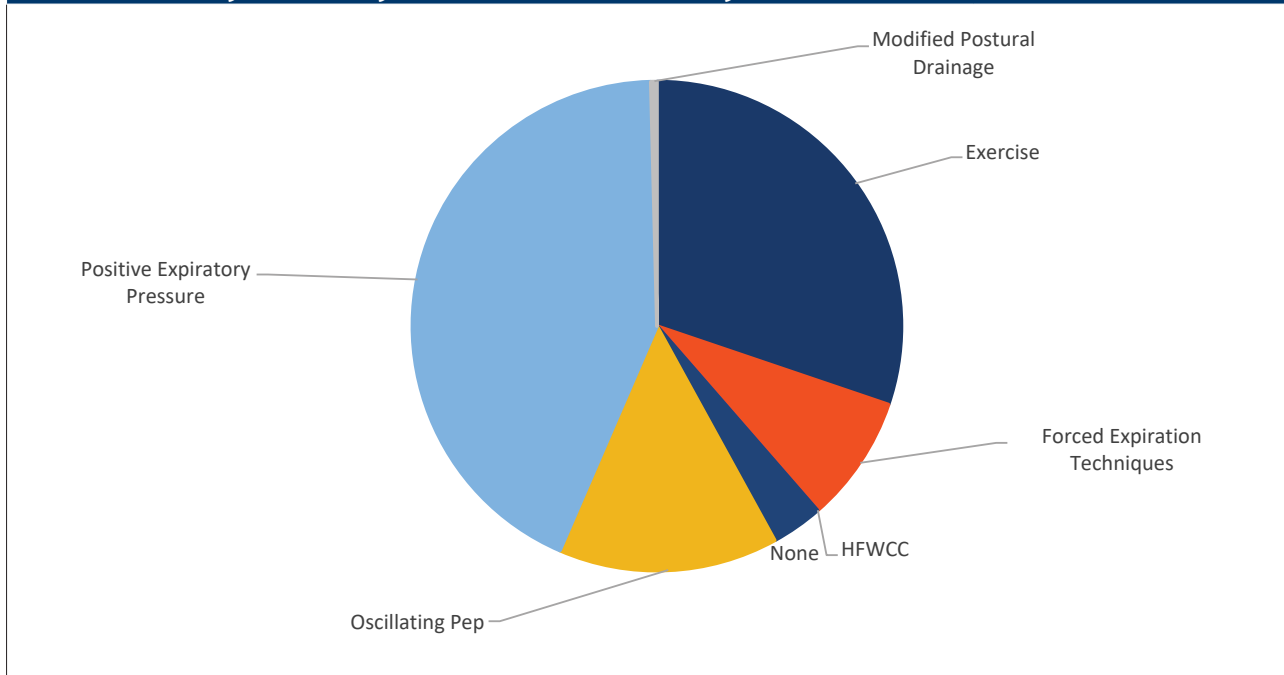
6.2 Primary Airway Clearance <16 years

219 PWCF



6.3 Primary Airway Clearance ≥16 years

270 PWCF



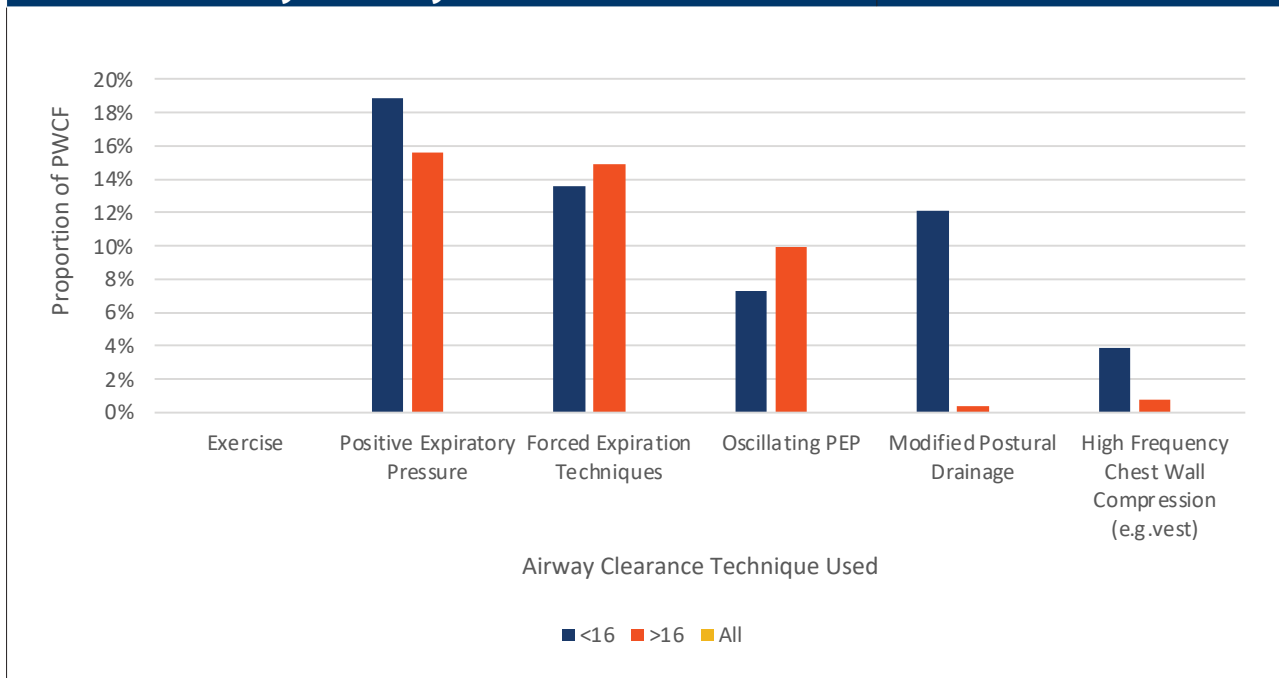
NOTE: Some patients may have used more than one technique as their primary airway clearance technique over the course of a year.

The nominated primary airway clearance technique adds up to greater than 100%. This is because, over the clinic visits for a year, an individual may use differing techniques as his/her main option at different times.

Over the years of the Registry reports, there has been an increase in the use of Oscillating Pep by children as these devices become more readily available. In paediatrics, there has been a significant drop in the use of modified postural drainage, by about a third, from 2021.

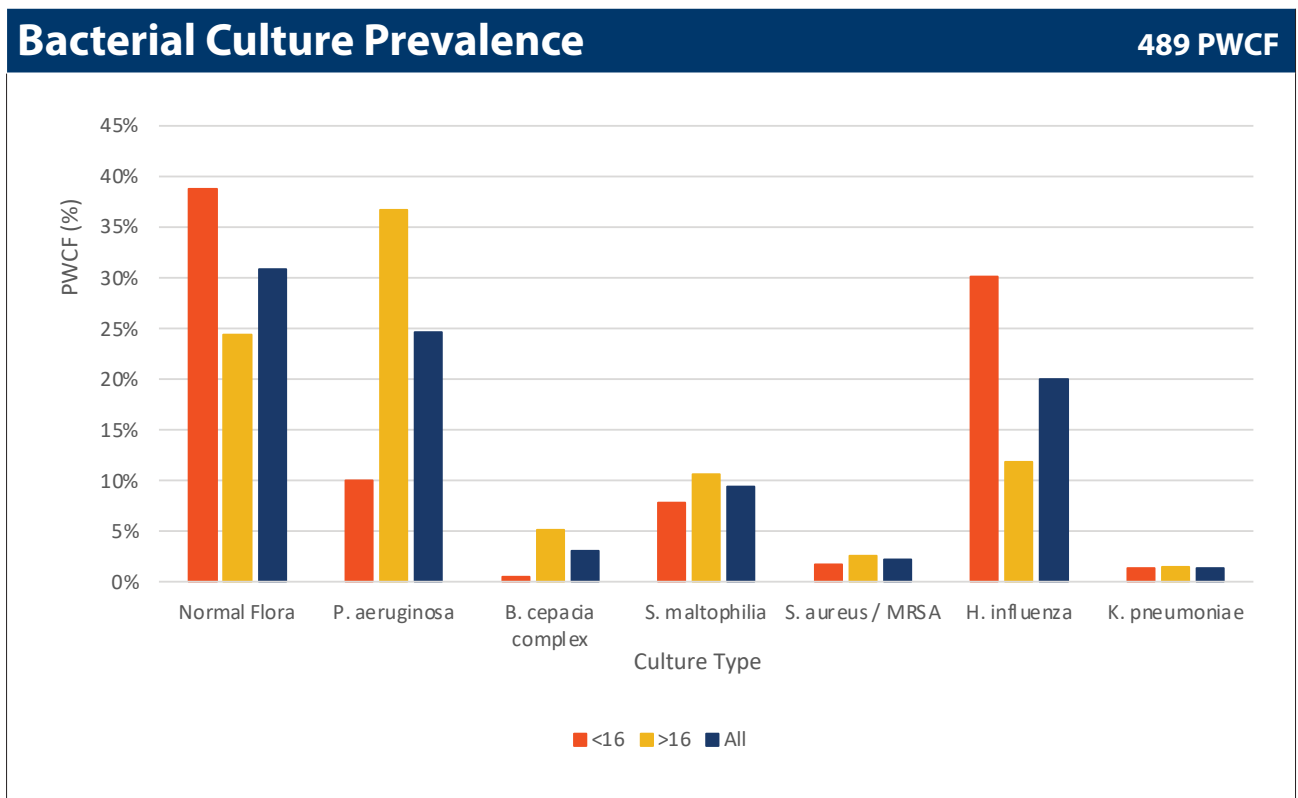
6.4 Secondary Airway Clearance

232 PWCF



7. Microbiology

7.1 Bacterial Culture Prevalence						489 PWCF
	<16 years 219 PWCF		≥16 years 270 PWCF		Total PWCF	
	Number	Percent	Number	Percent	Number	Percent
Normal Flora	85	38.8%	66	24.4%	151	30.9%
Haemophilus influenzae	66	30.1%	32	11.9%	98	20.0%
Escherichia coli	7	3.2%	0	0.0%	7	1.4%
Klebsiella pneumoniae	3	1.4%	4	1.5%	7	1.4%
Stenotrophomonas Maltophilia	17	7.8%	29	10.7%	46	9.4%
Pseudomonas Aeruginosa	22	10.0%	99	36.7%	121	24.7%
Mucoid	8	3.7%	61	22.6%	69	14.1%
Non Mucoid	14	6.4%	71	26.3%	85	17.4%
Staphylococcus Aureus	126	57.5%	137	50.7%	263	53.8%
MSSA	122	55.7%	130	48.1%	252	51.5%
MRSA	4	1.8%	7	2.6%	11	2.2%
Burkholderia Cepacia Complex	1	0.5%	14	5.2%	15	3.1%
Cenocepacia	0	0.0%	2	0.7%	2	0.4%
Multivorans	1	0.5%	6	2.2%	7	1.4%
Other	29	13.2%	18	6.7%	47	9.6%

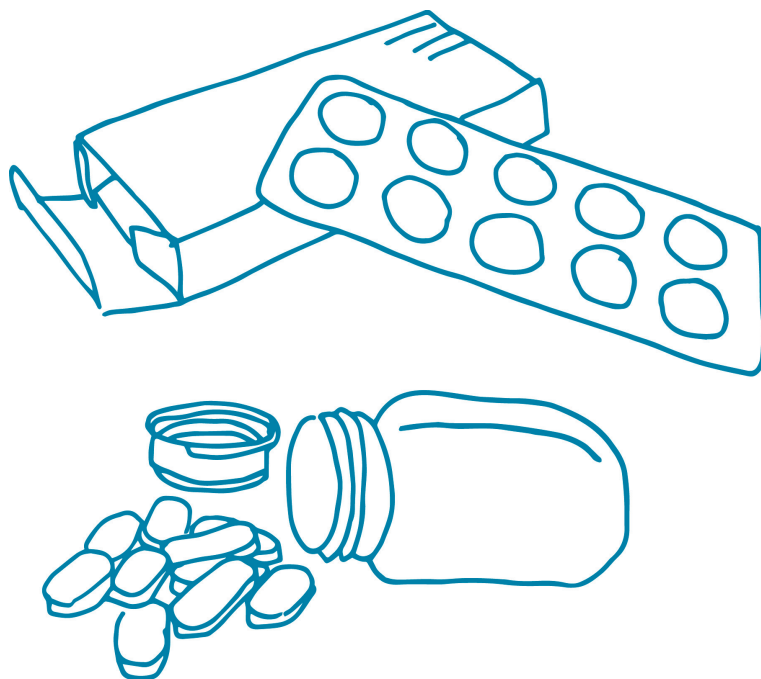
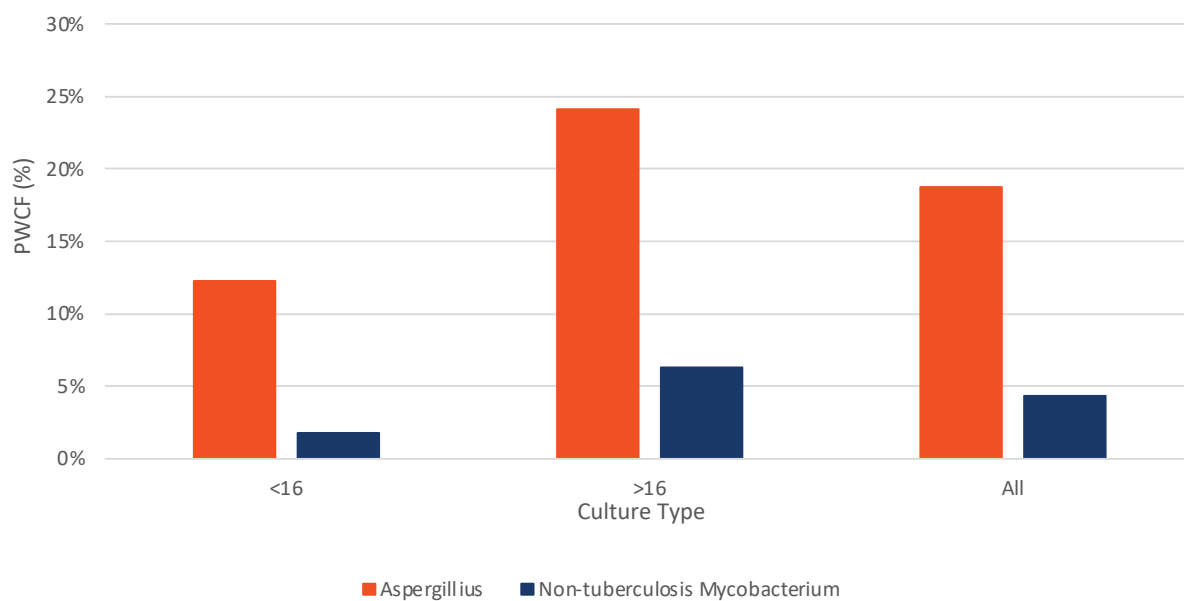


Pseudomonas aeruginosa is found in 10% of children and increases to 36.7% in adults. Our MRSA rates are relatively low overall, dropping from 4.3% in 2021 to 2.2% this year.

Compared to 2020, Haemophilus influenzae in 2022 remains lower and is similar to 2021 at 20%.

7.2 Nonbacterial/Fungal Prevalence

486 PWCF



8. Medications

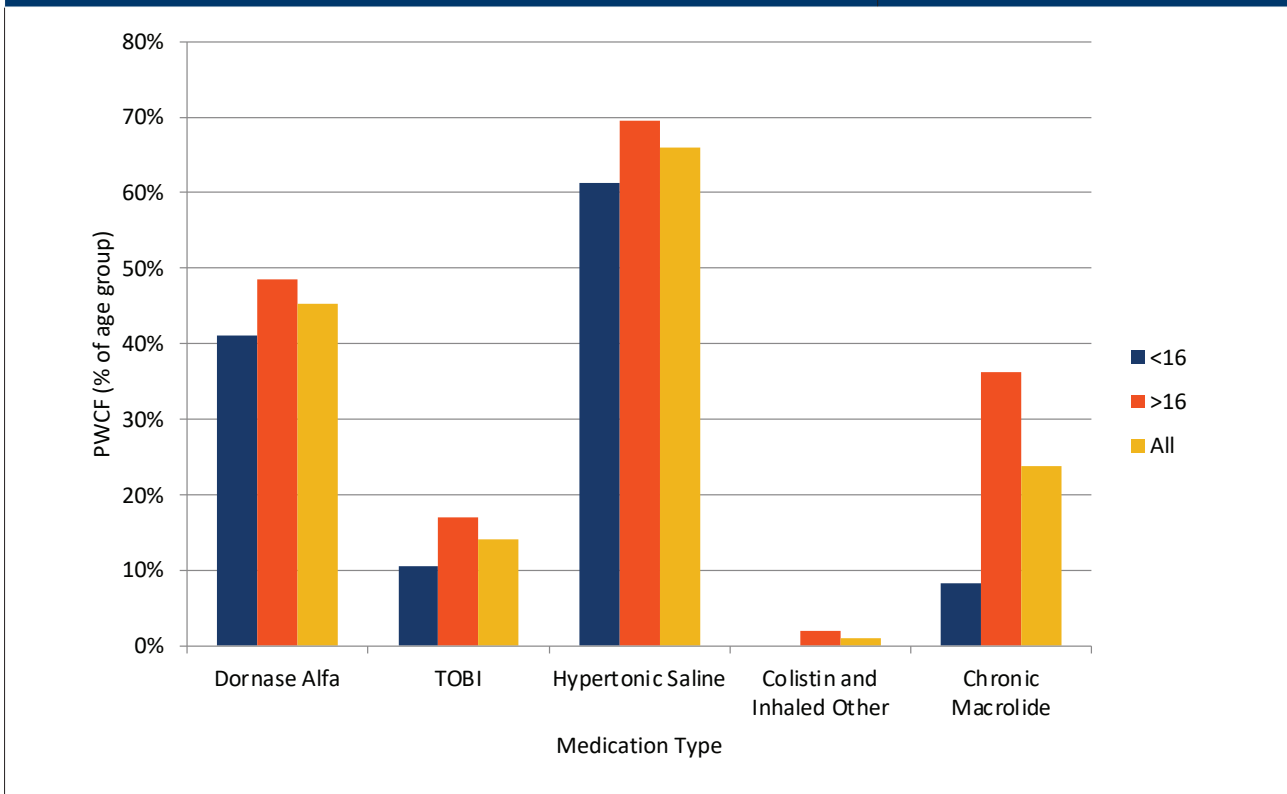
8.1 Medications Prescribed

489 PWCF

Medication	<16 Years	≥16 Years	All
Hypertonic Saline	61.2%	69.6%	65.9%
Dornase alfa	41.1%	48.5%	45.2%
TOBI	10.5%	17.0%	14.1%
Chronic Macrolide	8.2%	36.3%	23.7%
Corticosteroids Inhaled	10.1%	11.9%	11.0%
Corticosteroids Oral	2.3%	4.4%	3.5%
Antifungals	90.0%	2.6%	1.8%
Influenza Vaccine	73.2%	61.2%	66.3%

Medications Prescribed

489 PWCF



Inhaled other - This includes nebulised Colistin, Tobramycin (intravenous solution), Gentamicin and Aztreonam.

Our use of inhaled antibiotics, nebulised dornase alfa, and oral chronic macrolide therapy is lower than other international registries, but we are high users of nebulised hypertonic saline.

9. Intravenous Antibiotic Treatment

9.1 Home IV Days 477 PWCF

Age	Number In Age Group	Number Who Had IV Days	Percent PWCF Who Had IV Days	Mean Days For PWCF Who Had IV Days	Mean Days For All PWCF
<4	38	13	34.2%	13.5	4.6
4-7	49	10	20.4%	9.2	1.9
8-15	54	13	24.1%	24.2	5.8
16-18	65	21	32.3%	24	7.9
16-19	49	14	28.6%	21.6	6.2
20-23	45	16	35.6%	28.2	10.0
24-27	39	10	25.6%	14.8	3.8
28-31	29	11	37.9%	17.4	6.6
32-35	22	7	31.8%	7.1	2.3
36-39	19	8	42.1%	20.9	8.8
40-43	19	6	31.6%	6.2	1.9
44-47	8	1	12.5%	6	0.8
48-51	12	1	8.3%	11	0.9
52-55	8	1	12.5%	9	1.1
56-59	8	2	8.3%	8.5	2.1
≥60	13	2	12.5%	11.5	1.8
Totals	477	136	28.5%	18.4	2.3

9.2 Home IV Days 477 PWCF

Age	Number In Age Group	Number Who Had IV Days	Percent PWCF Who Had IV Days	Mean Days For PWCF Who Have Had IV Days	Mean Days For All PWCF
<4	38	1	2.6%	8	0.2
4-7	49	7	14.3%	10.7	1.5
8-11	54	3	5.6%	28.0	1.6
12-15	65	5	7.7%	25.0	2
16-19	49	8	16.3%	15.6	2.6
20-23	45	9	20.0%	14.7	2.9
24-27	39	8	20.5%	16.8	3.4
28-31	29	6	20.7%	13.7	2.8
32-35	22	5	22.7%	11.0	2.5
36-39	19	4	21.1%	31.0	6.5
40-43	19	6	31.6%	11.7	3.7
44-47	8	0	0.0%	-	-
48-51	12	1	8.3%	35.0	2.9
52-55	8	1	12.5%	35.0	4.4
56-59	8	1	12.5%	8.0	1.0
≥60	13	1	7.7%	14	1.1
Totals	477	66	13.8%	16.8	2.3



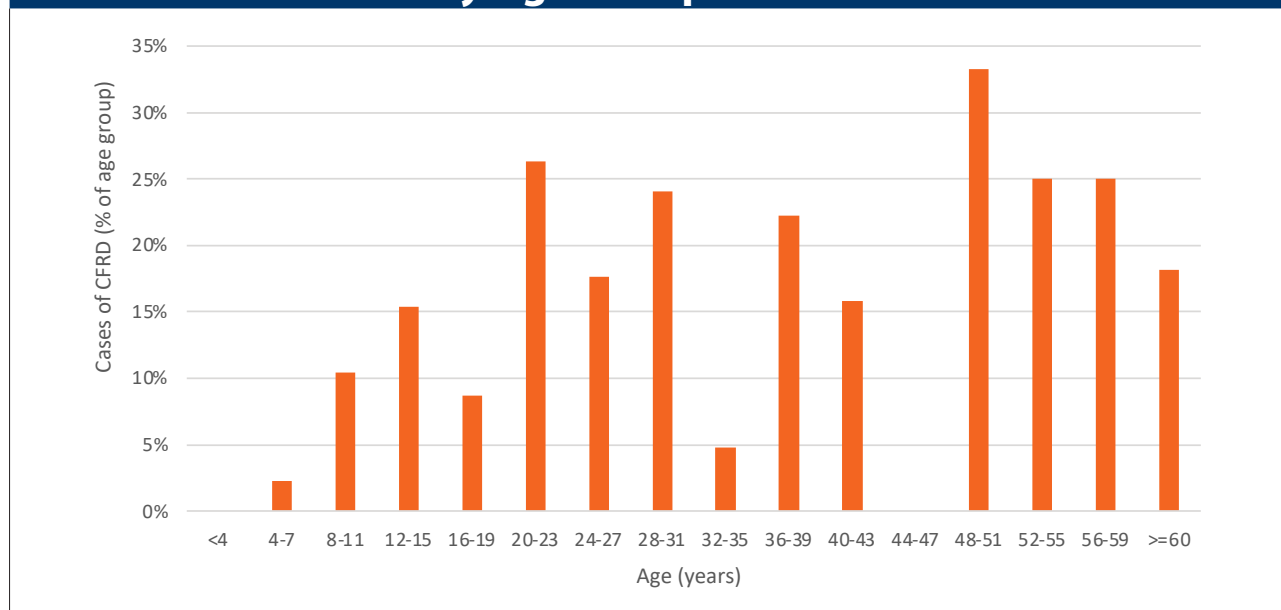
10. Complications

10.1 CF Related Diabetes By Age Group 430 PWCF

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<4	36	0	0.0%	0.0%
4-7	43	1	2.3%	0.2%
8-11	48	5	10.4%	1.2%
12-15	52	8	15.4%	1.9%
16-19	46	4	8.7%	0.9%
20-23	38	10	26.3%	2.3%
24-27	34	6	17.6%	1.4%
28-31	29	7	24.1%	1.6%
32-35	21	1	4.8%	0.2%
36-39	18	4	22.2%	0.9%
40-43	19	3	15.8%	0.7%
44-47	7	0	0.0%	0.0%
48-51	12	4	33.3%	0.9%
52-55	8	2	25.0%	0.5%
56-59	8	2	25.0%	0.5%
≥60	11	2	18.2%	0.5%
Total	430	59		13.7%

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<16	179	14	7.8%	3.3%
≥16	251	45	17.9%	10.5%
Total	430	59		13.7%

CF Related Diabetes By Age Group 430 PWCF



Diabetes diagnosis is decreasing as a percentage of the paediatric population from 2020, 2021 and for 2022.

10.2 Liver Function by Ultra Sound					61 PWCF
		Normal		Abnormal	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	24	22	91.7%	2	8.3%
Adults	37	24	64.9%	13	35.1%
Total	61	46	75.4%	15	24.6%

10.3 Bone Density by DEXA Scans					150 PWCF
		Normal		Abnormal	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	90	72	80.0%	18	20.0%
Adults	60	40	66.7%	20	33.3%
Total	150	112	74.7%	38	25.3%

