Port CF 2021

New Zealand National Data Registry

CYSTIC FIBROSIS NZ

Contents

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, 2021 Registry Report, Cystic Fibrosis NZ. http://cfnz.org.nz/

Introduction

From the Chair of the Port CFNZ Steering Commmittee

Cystic Fibrosis NZ and the Port CF Steering Committee are pleased to present the National Data Registry 2021 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 eleventh Registry Report provides an increasingly accurate picture of people with CF in New Zealand and their outcomes with 97-98% opting to provide this anonymised data, bringing the total number to 577.

The 2021 Registry Report continues to inform future care and the future resources needed. It is particularly important to understand the number of people with CF and those who may benefit from modulator therapy.

We hope you find the information in this 2021 Report informative and useful.

Associate Professor Cass Byrnes
Chair Port CF Steering Committee
Port CFNZ Principal Investigator (2017 - 2021)

Jane Bollard
Chief Executive, Cystic Fibrosis NZ
(until September 2021)

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
Tairawhiti 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
FEV_1	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 some challenges to statistical interpretation as outliers, in terms of late diagnoses and key markers, may 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 there are a total of 577 registered in Port CF, not all individuals had an input for all categories. While the total is 577 (239 children under 16 year yof age and 338 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								577 PWCF
	2021	2020	2019	2018	2017	2016	2015	2014
CF Patients Registered	577	572	531	514	498	501	449	443
Diagnosis								
Diagnosis age <1 year	14	15	11	15	15	6	5	7
Diagnosis age <16 years	4	4	1	0	2	3		
Diagnosis age ≥16 years	3	0	2	0	1	2	0	2
Age								
Median Age (in years)	19.0	19	18.87	18.54	18.27	17.38	18.25	18.11
Mean Age (in years)	21.7	21.6	21.49	21.2	20.79	20.04		
PWCF <16 years								
Number	239	236	223	224	279	233	192	196
Percent	41.4%	41.3%	42%	43.6%	56.0%	46.5%	42.8%	44.2%
PWCF ≥16 years								
Number	338	336	308	290	219	268	257	247
Percent	58.6%	58.7%	58%	56.4%	44.0%	53.5%	57.2%	55.8%
Gender								
Males								
Number	321	319	297	285	273	275	247	240
Percent	55.6%	55.8%	56%	55.4%	54.9%	54.9%	55.0%	54.2%
Females								
Number	256	253	233	229	224	226	202	203
Percent	44.4%	44.2%	44%	44.6%	45.1%	45.1%	45.0%	45.8%
Genotyped								
Number	577	572	495	466	484	450	400	429
Percent	100%	100.0%	93.2%	90.7%	97.4%	90.0%	89.1%	96.8%
FEV1 (% predicted)								
Mean	79.8	78.4%	76.6%	81.8	85.1%	85.0%		
Median	84.8%	82.8%	79%	86.2	86.5%	88.4%	85.6%	85.1%
FEV1 < 16 Years								
Mean	94.0%	91.4%	95.8%	96.70%	96.8%	97.3%		
Median	95.2%	93.7%	97.9%	98.80%	99.3	99.3%	98.9%	97.7%
FEV1 ≥16 Years								
Mean	72.5%	71.7%	74.7%	75.30%	72.60%	72.6%		
Median	75.8%	74.2%	76.8%	79.20%	77.4	77.4%	77.0%	78.0%
FEV1 < 18 Years								
Mean	91.8%	89.7%	94.5%	95.40%	95.1%	95.0%		
Median	93.8%	93.1%	97.2%	98.30%	98.3%	98.0%		
FEV1 ≥18 Years								
Mean	77.0%	70.9%	73.1%	73.7%	72.2%	71.2%		
Median	75.6%	72.7%	74.7%	77.6%	75.6%	75.1%		

Key Indicators cont.

Key Indicators								
	2021	2020	2019	2018	2017	2016	2015	2014
FVC (% predicted)								
Mean	94.7%	93.4%						
Median	98.8%	96.0%						
FVC < 16 Years								
Mean	100.5%	97.0%						
Median	101.4%	98.3%						
FVC ≥16 Years								
Mean	91.7%	91.5%						
Median	95.9%	95.1%						
FVC < 18 Years								
Mean	99.5%	96.9%						
Median	101.3%	98.3%						
FVC ≥ 18 Years								
Mean	91.5%	91.1%						
Median	95.5%	93.7%						

Compared to previous years, there has been a slight increase in FEV1 and FVC in both our paediatric and adult populations.

1. Demographics

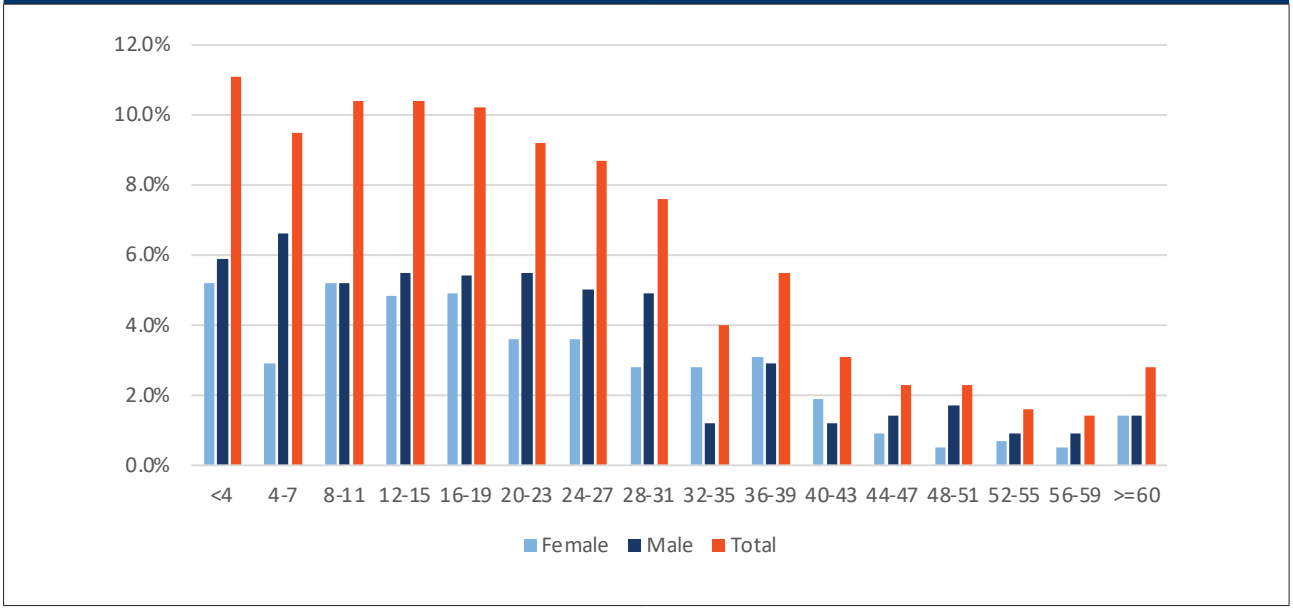
1.1 Age distribution

577 PWCF

Age Group	All		Male		Female	
0-3	63	11.1%	34	5.9%	30	5.2%
4-7	55	9.5%	38	6.6%	17	2.9%
8-11	60	10.4%	30	5.2%	30	5.2%
12-15	60	10.4%	32	5.5%	28	4.9%
16-19	59	10.2%	31	5.4%	28	4.9%
20-23	53	9.2%	32	5.5%	21	3.6%
24-27	50	8.7%	29	5.0%	21	3.6%
28-31	44	7.6%	28	4.9%	16	2.8%
32-35	23	4.0%	7	1.2%	16	2.8%
36-39	32	5.5%	17	2.9%	15	3.1%
40-43	18	3.1%	7	1.2%	11	1.9%
44-47	13	2.3%	8	1.4%	5	0.9%
48-51	13	2.3%	10	1.7%	3	0.5%
52-55	9	1.6%	5	0.9%	4	0.7%
56-59	8	1.4%	5	0.9%	3	0.5%
>=60	16	2.8%	8	1.4%	8	14%
Total	**577**		**321**	**55.6%**	**256**	**44.4%**
Median	**19**					
Range	**0 - 79**					

Age distribution

577 PWCF

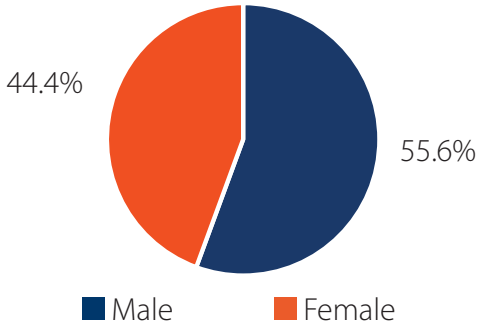


The median age of persons with CF in New Zealand has increased from 16 to 19 years over the eleven years that we have had Registry data. Children in international registries are defined as either up to 16 years or up to 18 years of age. In New Zealand, if we include children as being up to 16 years, we have 239 children (41.4% total) and 338 adults (58.6% total), if we include children as being up to 18 years, which is our more usual clinical practice, we have 263 children (45.6% total) and 314 adults (54.4% total).

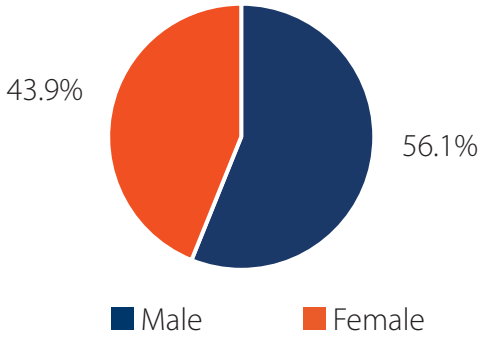
1.2 Gender Distribution — 577 PWCF

	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	55.8%	134	56.1%	187	55.3%
Female	256	44.4%	105	43.9%	151	44.7%
Totals	**577**	**100%**	**239**	**41.4%**	**338**	**58.6%**

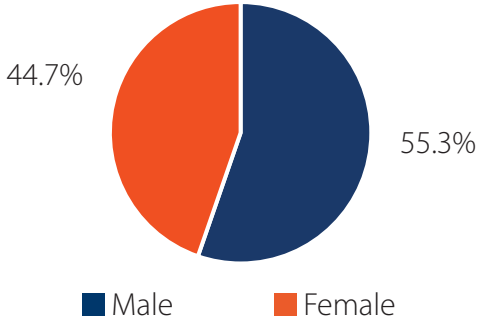
Gender Distribution of All PWCF — 577PWCF



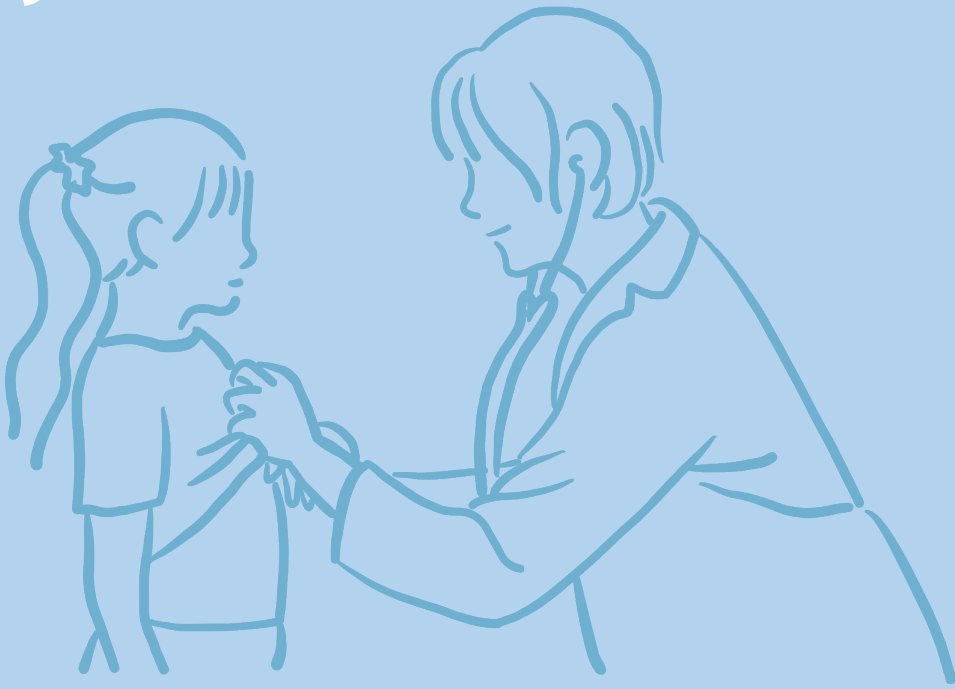
Gender Distribution <16 Years — 239 PWCF



Gender Distribution ≥16 Years — 338 PWCF



“The median age of persons with CF in New Zealand has increased from 16 years to 19 years over the eleven years that we have had National Registry data.”



2. Genotypes

2.1 Number and Percent Genotyped — 577 PWCF

Mutations	Number of PWCF Genotyped	Percentage of PWCF Genotyped
Homozygous F508del	267	46.3%
Heterozygous F508del	237	41.1%
No F508del	73	12.7%
Total	**577**	

2.2 Second Allele of Heterozygous F508del — 237 PWCF

Second Allele	c.DNA Name	Number of PWCF	Percent of PWCF
R117H	c.350G>A	29	5.0%
G551D	c.1652G>A	24	4.2%
G542X	c.1624G>T	21	3.6%
3849+10kbC->T	c.3717+12191C>T	9	1.6%
N1303K	c.3909c>G	8	1.4%
^1507	c.1519_1521delATC	4	0.7%
1898+1G->A	c.1766+1G>A	4	0.7%
3659delC	c.3528delC	4	0.7%
G85E	c.254G>A	4	0.7%
D1152H	c.3454G>C	4	0.7%
1717-1G->A	c.1585-1G>A	3	0.5%
621+1G->T	c.489+1G>T	3	0.5%
Q493X	c.1477C>T	3	0.5%
1078delT	c.948delT	2	0.3%
A455E	c.1364C>A	2	0.3%
E60X	c.178G>T	2	0.3%
L206W	c.617T>G	2	0.3%
R352Q	c.1055G>A	2	0.3%
R560T	c.1679G>C	2	0.3%
Y563D	c.1687T>G	2	0.3%
2789+5G->A	c.2657+5G>A	2	0.3%
R334W	c.1000C>T	1	0.2%
1898+5G->T	c.1766+5G>T	1	0.2%
3120+1G->A	c.2988+1G>A	1	0.2%
P574H	c.1721C>A	1	0.2%
R1162X	c.3484C>T	1	0.2%
R347H	c.1040G>A	1	0.2%
R347P	c.1040G>C	1	0.2%
W1282X	c.3846G>A	1	0.2%
D1270N	c.3808G>A	1	0.2%
R117C	c.349C>T	1	0.2%
Other genetic mutation		84	14.6%
Not identified		7	1.2%
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 73 persons in New Zealand do not have at least one F508del mutation. Looking at the gene mutations recorded in 2021 Registry, 47 of the 577 people who have been genotyped (8.1%) would not be detected by the current new born screening programme.

2.3 No F508del Mutations

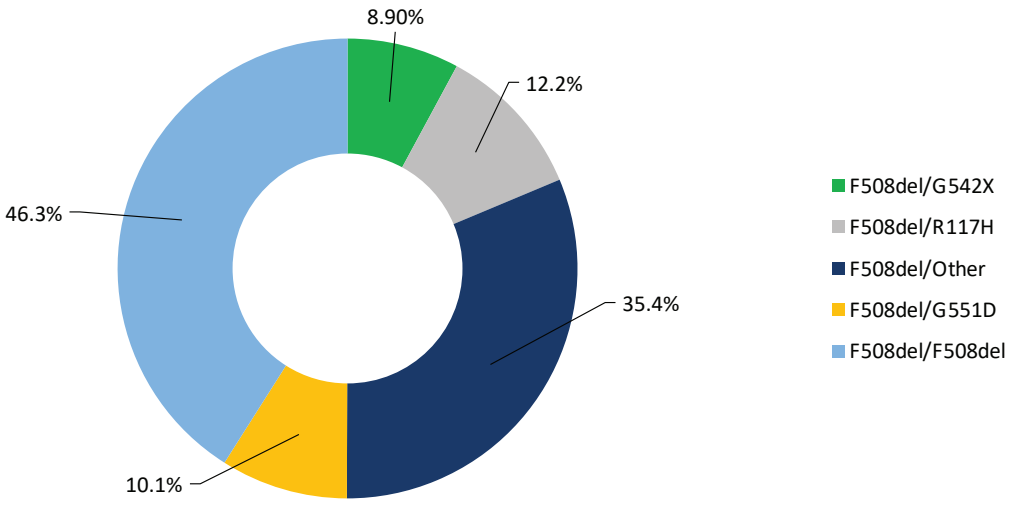
73 PWCF

	1717-1G->A	2143delT	3849+10kbC->T	621+1G->T	G542X	G551D	Not Identified	Other	Q493X	R1162X	R117H	R553X	W1282X	^I507
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	1	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	8	0	0	0	0	0	0	0
Other	0	0	0	0	0	0	3	10	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 PWCF Identified	Percentage PWCF Identified
F508del	504	87.3%
G551D	43	7.5%
R117H	41	7.1%
G542X	28	4.9%

2.5 Common Genotype Combinations

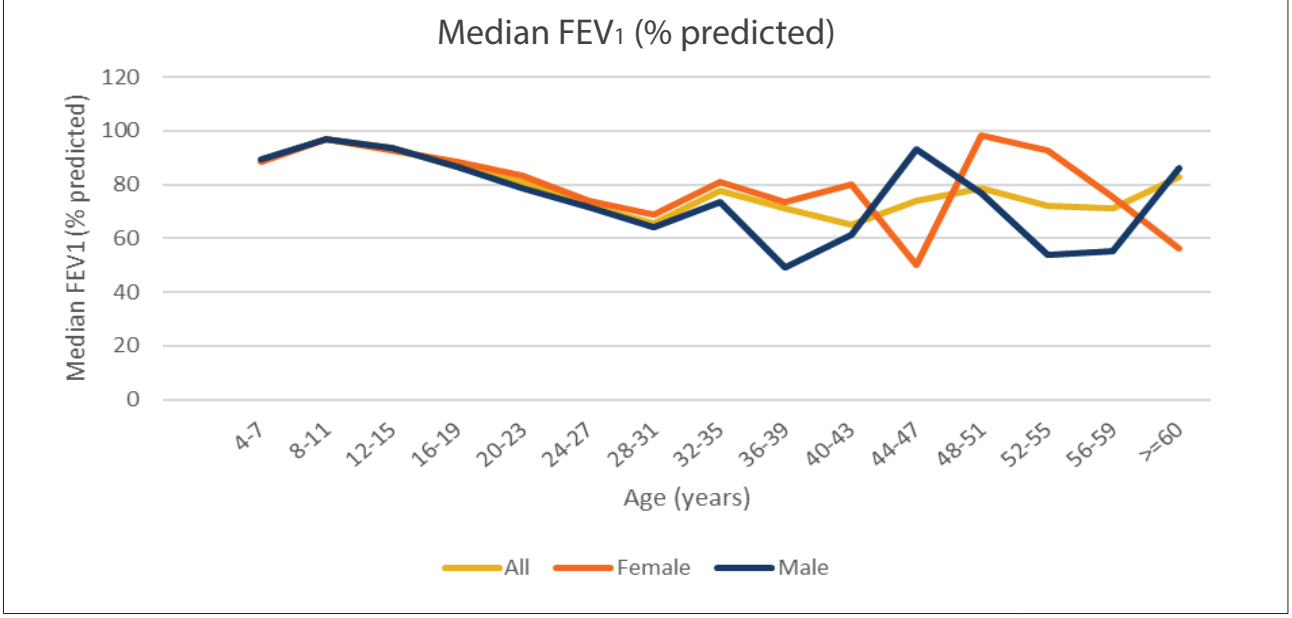


3. Respiratory

3.1 Median FEV_1 — 407 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	38	89.0	13	88.4	25	89.2
8-11	56	96.9	29	96.9	27	96.9
12-15	55	93.4	25	92.8	30	93.6
16-19	49	86.5	23	88.6	26	86.4
20-23	41	80.8	15	83.3	26	78.7
24-27	38	72.3	15	74.0	23	71.8
28-31	28	65.6	6	68.8	22	64.0
32-35	18	77.7	12	80.9	6	73.5
36-39	25	71.2	13	73.4	12	49.4
40-43	13	65.1	8	79.9	5	61.4
44-47	9	73.8	3	50.3	6	93.2
48-51	12	78.7	3	98.5	9	76.8
52-55	6	72.3	2	92.9	4	53.8
56-59	7	71.1	3	75.1	4	55.5
≥60	12	82.8	6	56.1	6	86.0
Totals	**407**		**176**		**231**	

Median FEV_1 — 407 PWCF



The median FEV_1 of the population able to do lung function has always been greater than 80% predicted since we started our Registry and the median this year is 84.8% (95.2% in children, 75.8% 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. Nonetheless FEV1 is an important prognostic indicator.

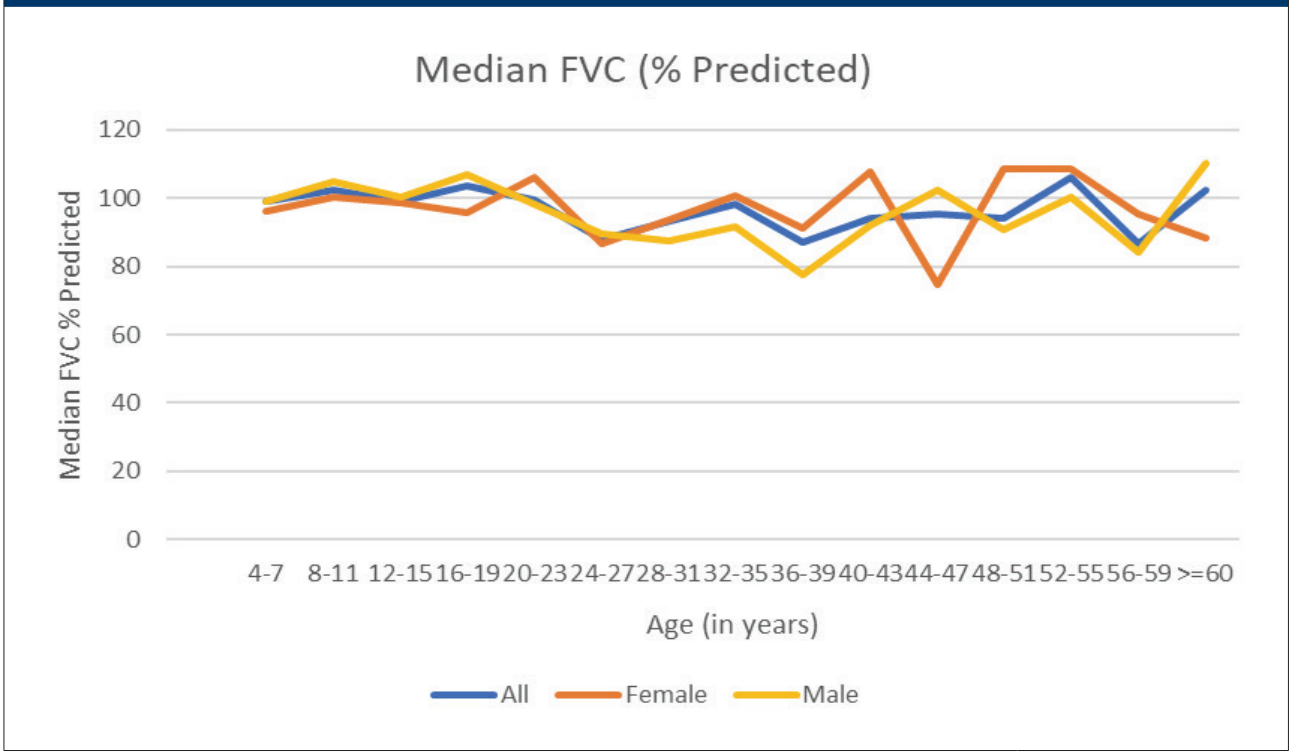
3.2 Median FVC

407 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	38	98.9	13	96.3	25	98.9
8-11	56	102.3	29	100.2	27	104.9
12-15	55	99.3	25	98.8	30	100.4
16-19	49	103.5	23	95.7	26	106.7
20-23	41	99.6	15	106.0	26	98.2
24-27	38	88.0	15	86.6	23	89.7
28-31	28	93.3	6	93.8	22	87.4
32-35	18	98.2	12	100.7	6	91.6
36-39	25	86.9	13	91.3	12	77.5
40-43	13	94.1	8	107.7	5	91.8
44-47	9	95.1	3	74.6	6	102.4
48-51	12	94.1	3	108.4	9	90.8
52-55	6	106.2	2	108.6	4	100.4
56-59	7	86.6	3	95.2	4	84.2
≥60	12	102.2	6	88.2	6	110.3
Totals	**407**		**176**		**231**	

Median FVC

407 PWCF



4. Nutrition

4.1 Paediatric BMI

197 PWCF

All <16 Years			Female <16 Years			Male <16 Years		
BMI Percentile			BMI percentile			BMI percentile		
Age group	Number in group	Median percentile	Age group	Number in group	Median percentile	Age group	Number in group	Median percentile
<4	34	63.1	<4	13	68.3	<4	21	51.3
4-7	51	63.8	4-7	17	57.6	4-7	34	67.6
8-11	56	54.8	8-11	29	46	8-11	27	57.3
12-15	56	48.6	12-15	25	67.9	12-15	31	40.8
Totals	**197**			**84**			**113**	

4.2 Adult BMI

254 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	48	20.3	16-19	22	20.2	16-19	26	20.7
20-23	41	22.1	20-23	15	22.7	20-23	26	21.4
24-27	38	22.1	24-27	15	22.1	24-27	23	21.9
28-31	28	22.1	28-31	6	22.6	28-31	22	22
32-35	17	22.7	32-35	11	22.7	32-35	6	26.4
36-39	23	23.1	36-39	12	22.6	36-39	11	24
40-43	13	23.3	40-43	8	22.8	40-43	5	24.4
44-47	9	24.4	44-47	3	22.2	44-47	6	27.6
48-51	12	25.1	48-51	3	23.2	48-51	9	25.3
52-55	6	26.1	52-55	2	35.3	52-55	4	25.4
56-59	7	26.5	56-59	3	24.9	56-59	4	27
≥60	12	25.2	60+	6	24.4	60+	6	25.9
254			**106**			**148**		

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 suggests 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)

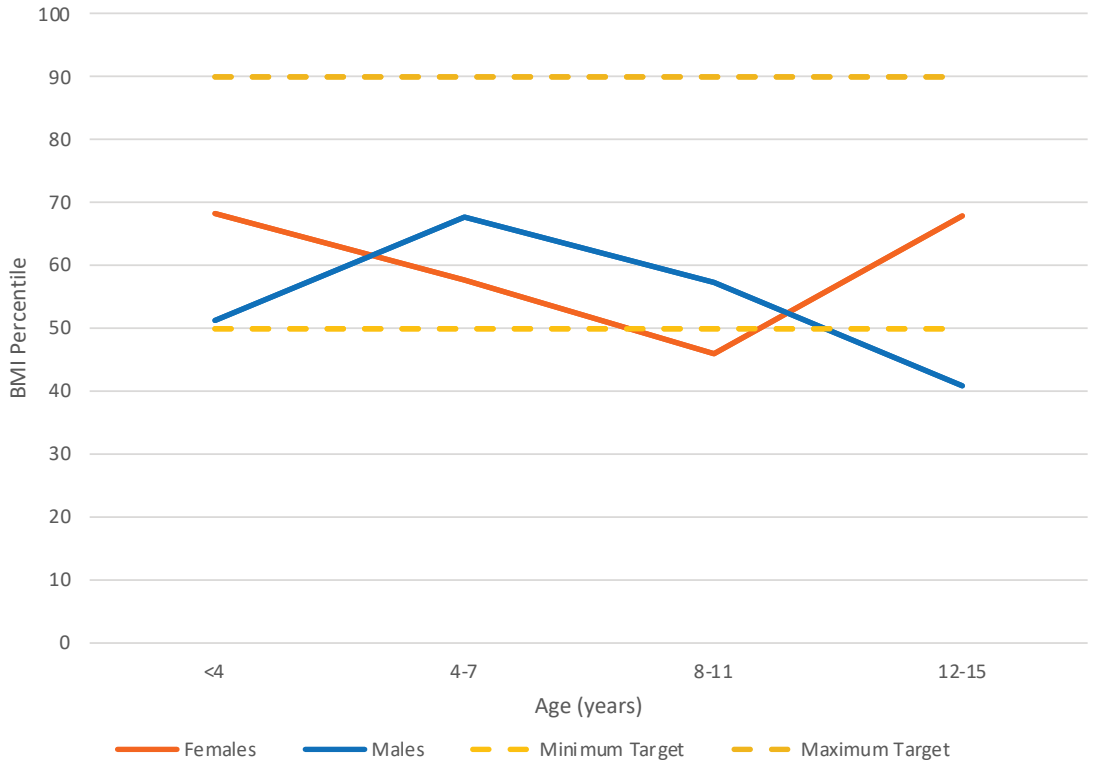
3) Adults: males BMI 23 - 27 kg/m2, females BMI 22 - 27 kg/m2

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 63.1 percentile. For children and adolescents, the median BMI is 57.3 percentile. For adults 49% of males and 55% of females are above the minimum target range.

4.3 Median BMI Percentile <16 Years

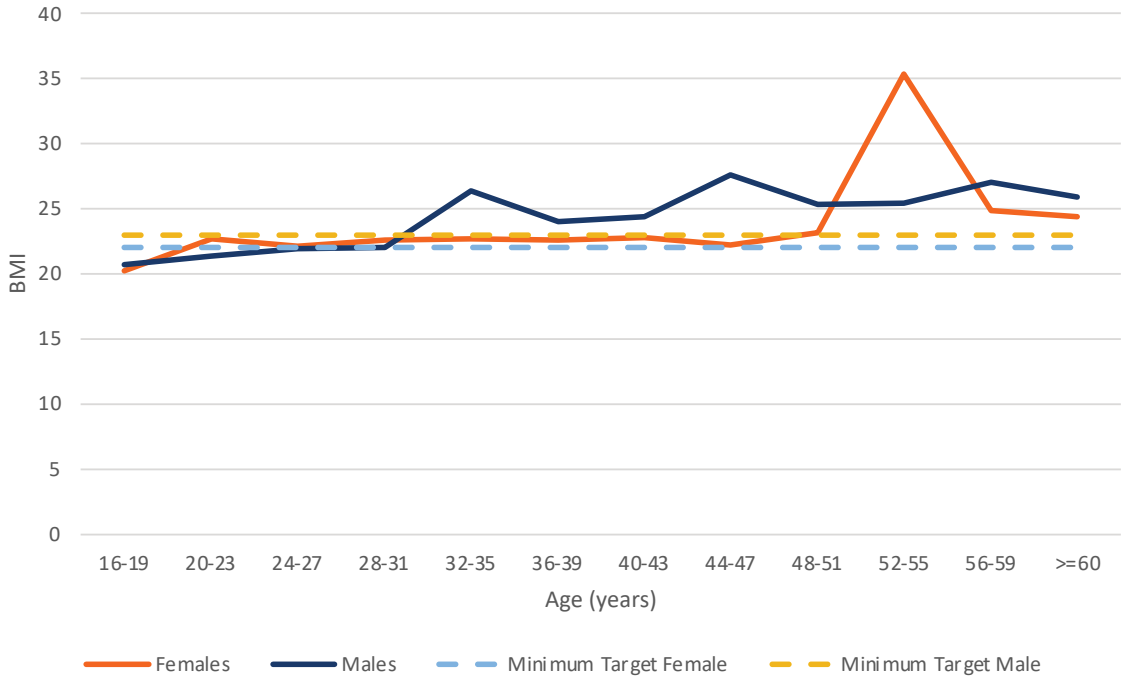
197 PWCF



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

254 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

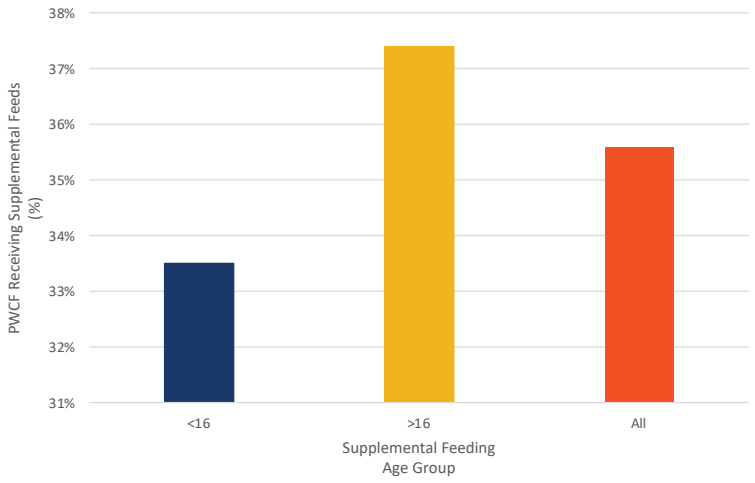
486 PWCF

	<16 years, n =224	
	Yes	Percent supplemented
Supplemental Feeding Total	**75**	**33.5%**
Nasogastric	5	2.2%
Gastrostomy	9	4.0%
Oral	69	30.8%
	≥16 years, n = 262	
	Yes	Percent supplemented
Supplemental Feeding Total	**98**	**37.4%**
Nasogastric	3	1.2%
Gastrostomy	17	6.5%
Oral	86	32.8%

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

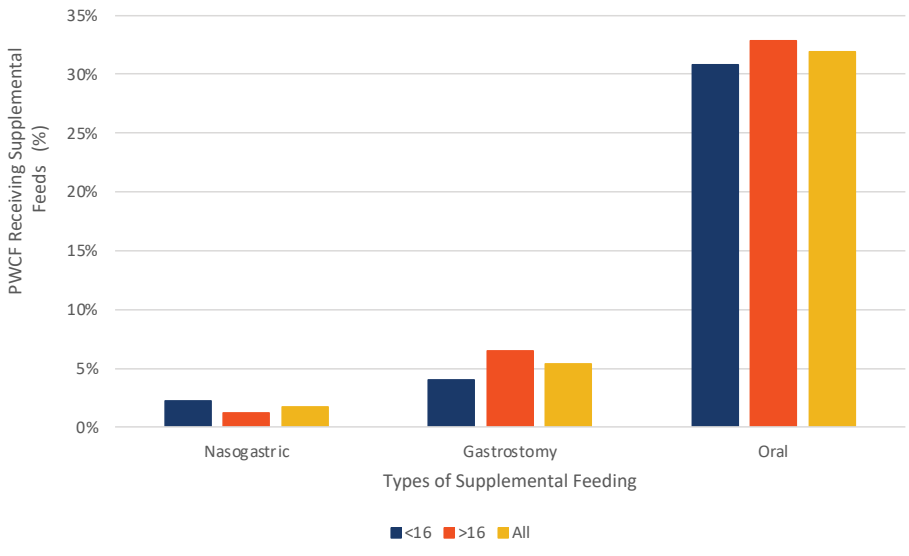
4.6 Supplemental Feeding by Age Group

486 PWCF



4.7 Supplemental Feeding by Type

486 PWCF



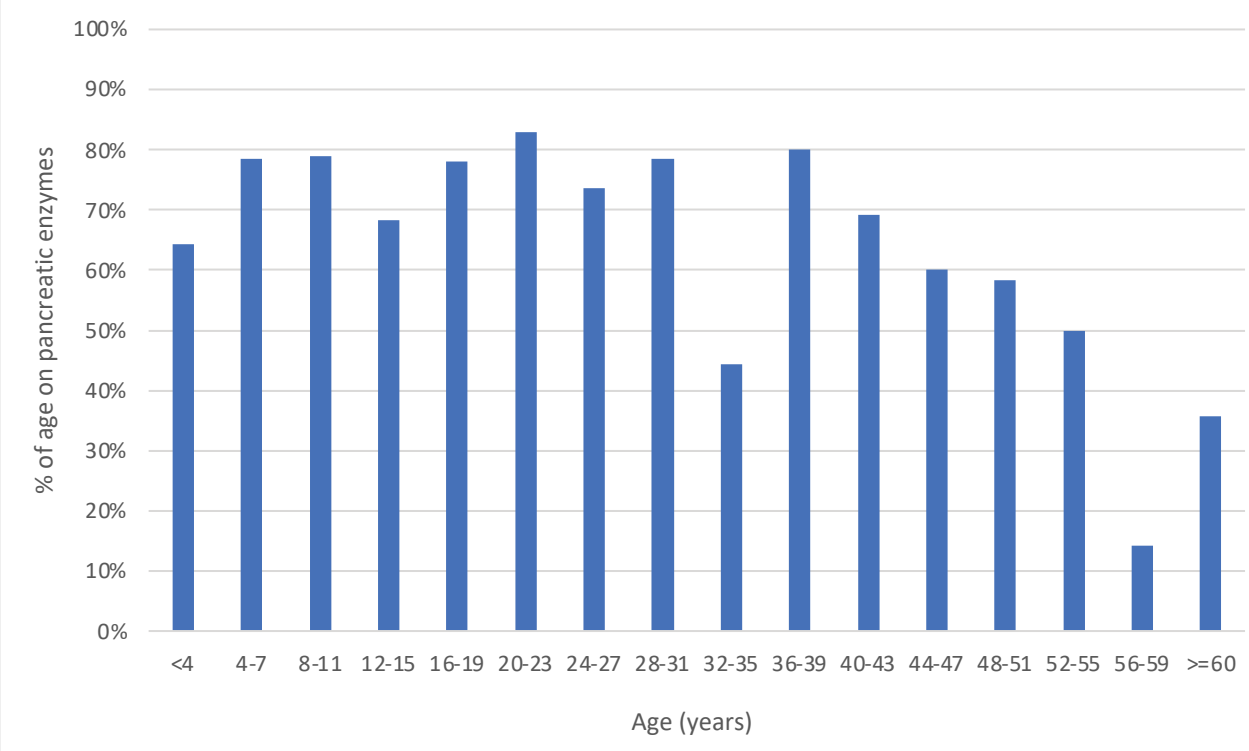
Supplemental feeding use remains high.

5. Pancreatic Enzymes

5.1 Pancreatic Enzymes by Age Group — 486 PWCF

Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of PWCF
<4	59	38	64.4%	7.8%
4-7	51	40	78.4%	8.2%
8-11	57	45	78.9%	9.3%
12-15	57	39	68.4%	8.0%
16-19	50	39	78.0%	9.0%
20-23	41	34	82.9%	7.0%
24-27	38	28	73.7%	5.8%
28-31	28	22	78.6%	4.5%
32-35	18	8	44.4%	1.6%
36-39	25	20	80.0%	4.1%
40-43	13	9	69.2%	1.9%
44-47	10	6	60.0%	1.2%
48-51	12	7	58.3%	1.4%
52-55	6	3	50.0%	0.6%
56-59	7	1	14.3%	0.2%
>=60	14	5	35.7%	1.0%
Totals	**486**	**344**		**71.6%**

Pancreatic Enzymes by Age Group — 486 PWCF



Of the 486 PWCF for whom there is data, 344 are on pancreatic enzymes and 142 are not.

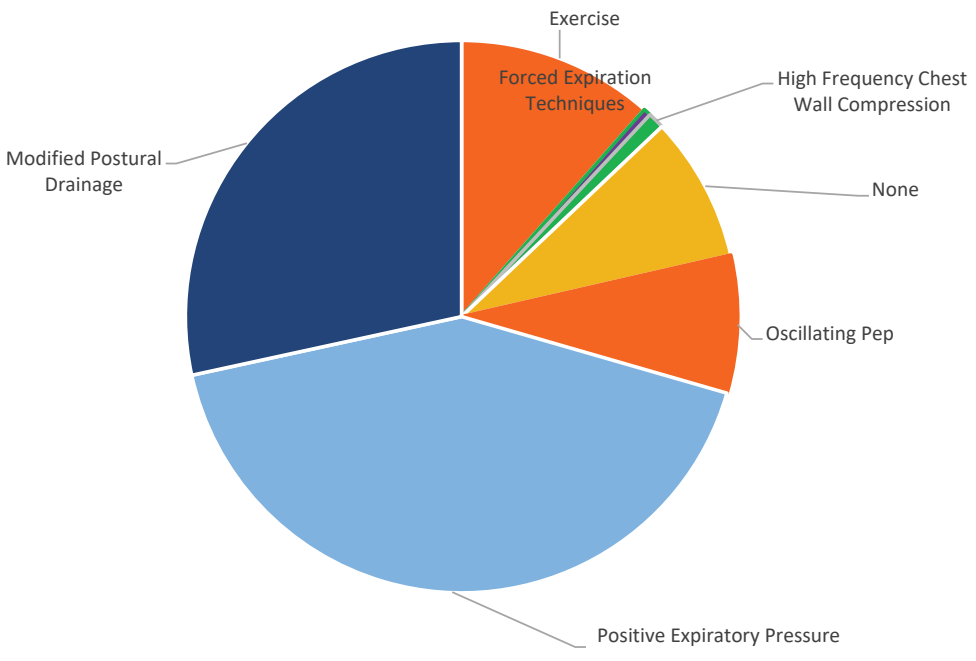
6. Airway Clearance Techniques

6.1 Primary Airway Clearance — 486 PWCF

	<16 years, n = 224	
	Number of PWCF	Percent of PWCF
Positive Expiratory Pressure	89	47.6%
Modified Postural Drainage	60	32.1%
Exercise	28	13.0%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	17	9.1%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	1	0.5%
High Frequency Chest Wall Compression (e.g. vest)	2	1.1%
None	18	9.6%

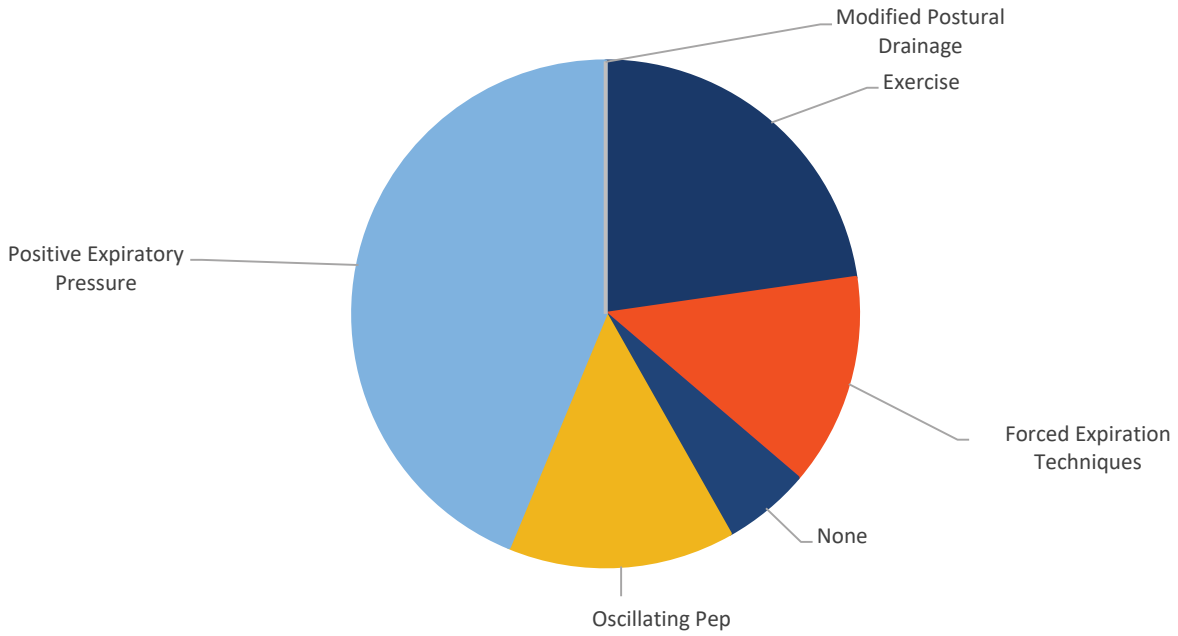
	≥16 years, n = 262	
	Number of PWCF	Percent of PWCF
Positive Expiratory Pressure	94	56.6%
Modified Postural Drainage	0	0.0%
Exercise	69	29.4%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	31	18.7%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	29	17.5%
High Frequency Chest Wall Compression (e.g. vest)	0	0.0%
None	12	7.2%

6.2 Primary Airway Clearance <16 years — 224 PWCF



6.3 Primary Airway Clearance ≥16 years

262 PWCF



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

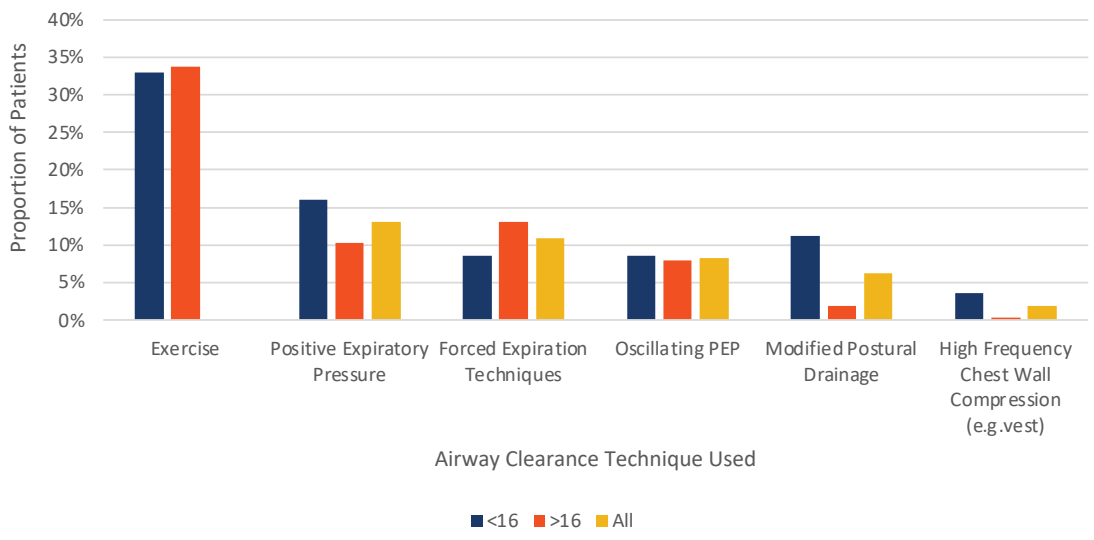
The nominated primary airway clearance technique adds up to greater than 100% because, over the clinic visits for a year, an individual may use differing techniques as his/her main option at different times.

Over the years of the Registry reports, there is a trend for an increased percentage of children to have nominated 'none' as their primary airway clearance, from 2.3% in 2013 to 9.6% in 2021. Overall, the trend is the opposite for the adults, from 18.8% in 2013 down to 6.5% in 2020. This year it has increased slightly to 7.2%

In 2020, significantly fewer children than in past reports chose positive expiratory pressure as their main source of airway clearance, from 59.9% in 2019 to 45.8% in 2020. This year has seen that number grow slightly with 47.6% of children using positive expiratory pressure as their primary airway clearance.

In 2020, we saw a slight drop in the numbers of adults using positive expiratory pressure as their main source of airway clearance, from 42.2% in 2019 to 40.4% in 2020. This year we see a large shift in the opposite direction, with 56.6% of adults choosing positive expiratory pressure as their primary form of airway clearance.

6.4 Secondary Airway Clearance



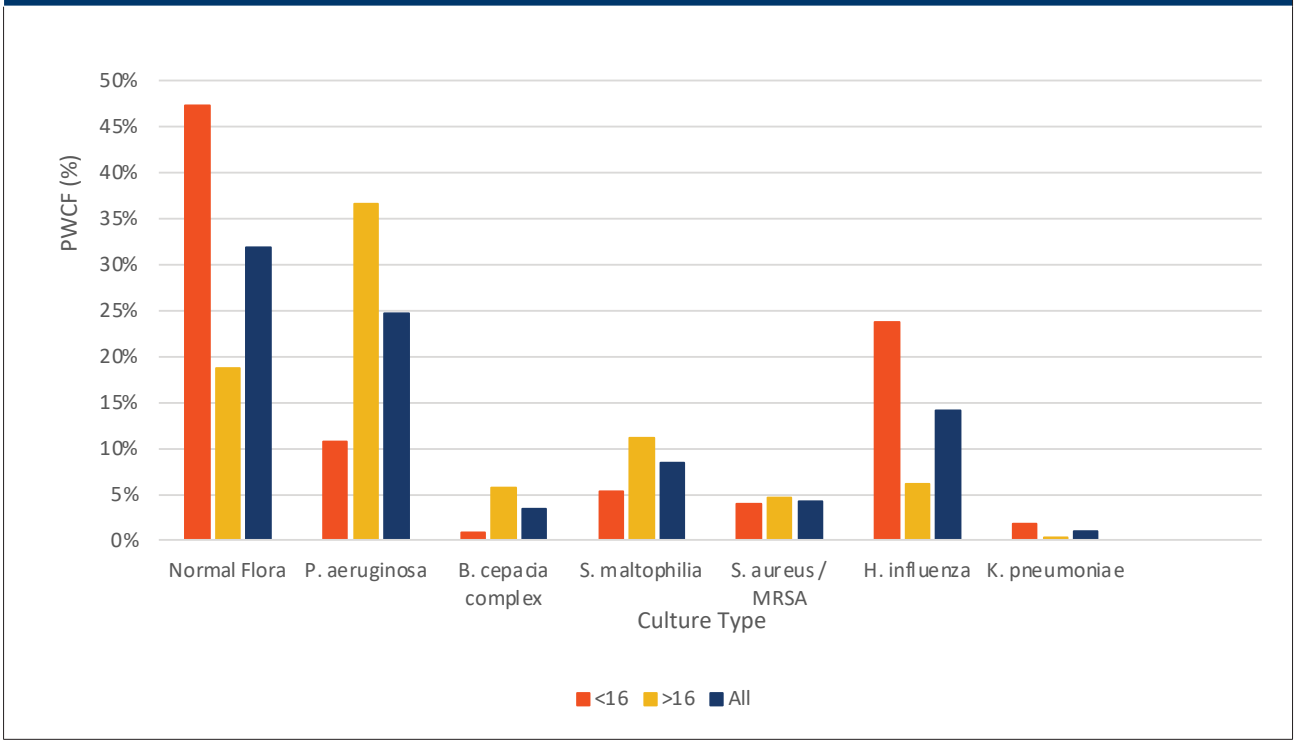
7. Microbiology

7.1 Bacterial Culture Prevalence

486 PWCF

	<16 years 224PWCF		≥16 years 262 PWCF		Total 486 PWCF	
	Number	Percent	Number	Percent	Number	Percent
Normal Flora	106	47.3%	49	18.7%	155	31.9%
Haemophilus influenzae	53	23.7%	16	6.1%	69	14.2%
Escherichia coli	7	3.1%	2	0.8%	9	1.9%
Klebsiella pneumoniae	4	1.8%	1	0.4%	5	1.0%
Stenotrophomonas Maltophilia	12	5.4%	29	11.1%	41	8.4%
Pseudomonas Aeruginosa	**24**	**10.7%**	**96**	**36.6%**	**120**	**24.7%**
Mucoid	6	2.7%	60	22.9%	66	13.6%
Non Mucoid	17	7.6%	69	26.3%	86	17.7%
Staphylococcus Aureus	**126**	**56.3%**	**120**	**45.8%**	**246**	**50.6%**
MSSA	117	51.8%	108	41.2%	225	46.3%
MRSA	9	4.0%	12	4.6%	21	4.3%
Burkholderia Cepacia Complex	**2**	**0.9%**	**15**	**5.7%**	**17**	**3.5%**
Cenocepacia	0	0.0%	1	0.4%	1	0.2%
Multivorans	2	0.9%	7	2.7%	9	1.9%
Other	22	9.8%	24	9.2%	46	9.5%

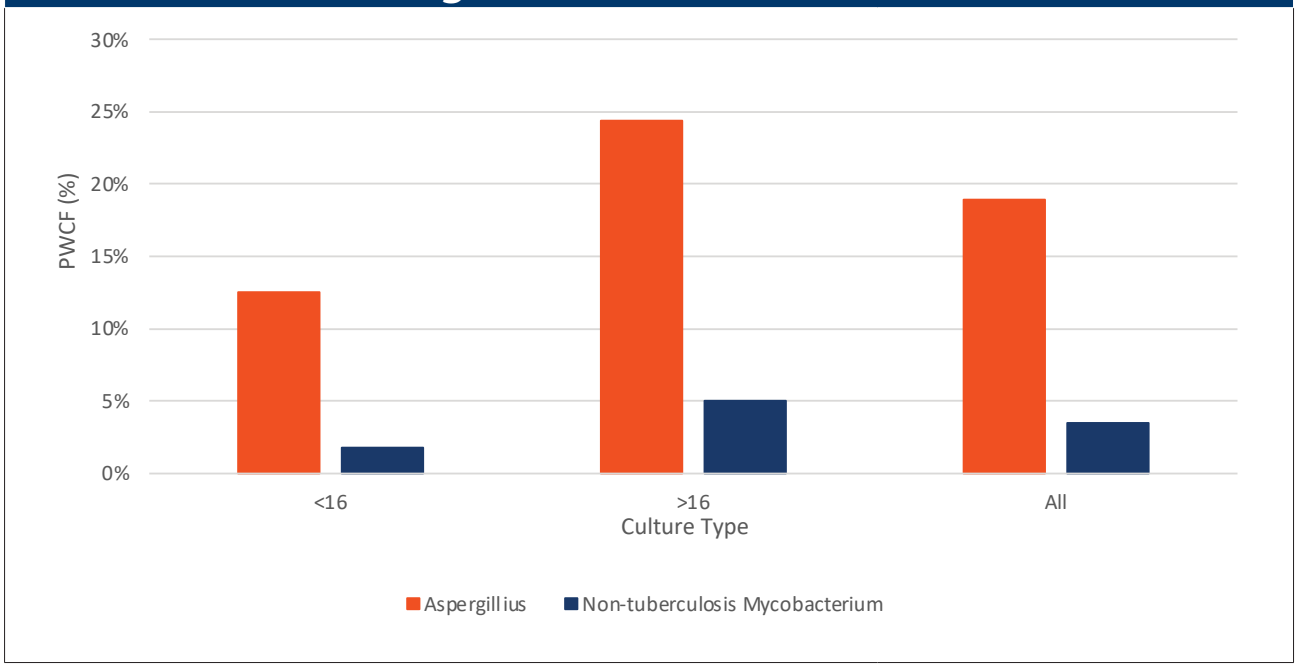
Bacterial Culture Prevalence



In 2019, Pseudomonas aeruginosa was found in 16% of children and 43% of adults. In 2020, this dropped to 11.1% for children and 27.2% for adults and it was speculated that this was due to COVID lockdowns reducing the number of samples collected. This year the child rate remains low at 10.7% but the adults' rate has increased to 36.6%.

7.2 Nonbacterial/Fungal Prevalence

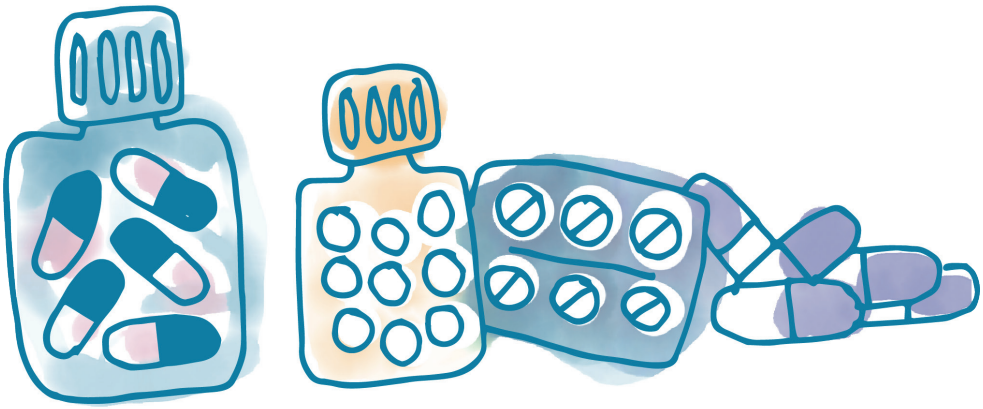
486 PWCF



Normal flora continues to increase in the paediatric cohort.

In 2020, we saw a significant increase in Haemophilus influenzae while this year it has dropped to the lowest rate seen since the first Registry report in 2011. This may now reflect the difficulty in obtaining sputum results post COVID lockdowns.

Burkholderia Cepacia Complex (Other) has dropped since last year but is still higher than in 2019.



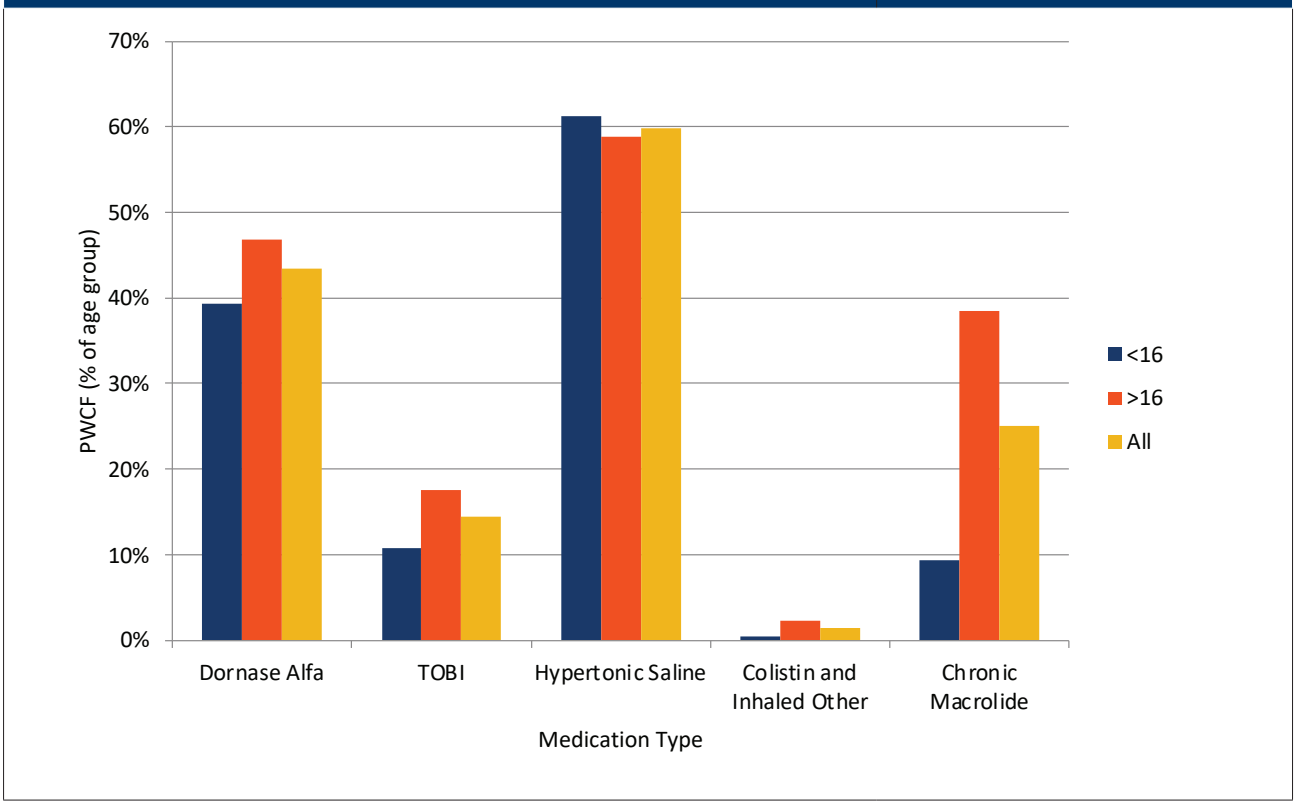
8. Medications

8.1 Medications Prescribed

476 PWCF

Medication	<16 Years	≥16 Years	All
Hypertonic Saline	61.2%	58.8%	59.9%
Dornase alfa	39.3%	47%%	43.4%
TOBI	10.7%	17.6%	14.4%
Chronic Macrolide	9.4%	38.5%	25.1%
Corticosteroids Inhaled	7.6%	13.0%	10.5%
Corticosteroids Oral	2.7%	5.7%	4.3%
Antifungals	2.2%	1.9%	2.1%
Influenza Vaccine	74.9%	54.4%	63.7%

Medications Prescribed



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.

The trend with the Influenza vaccine is opposite to last year, with more children and less adults getting the vaccine. It is disappointing that a third of adults and nearly half of children and youth did not receive the nfluenza vaccine (or if they did, it was not recorded in the Registry).

9. Intravenous Antibiotic Treatment

9.1 Home IV Days

476 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	52	9	17.3%	17.9	3.1
4-7	51	10	19.6%	12.8	2.5
8-15	56	12	21.4%	22.9	4.9
16-18	56	16	28.6%	18.5	5.3
16-19	50	22	44.0%	33.8	14.9
20-23	41	14	34.1%	33.8	11.5
24-27	38	12	31.6%	23.3	7.4
28-31	28	11	39.3%	18.7	7.4
32-35	17	5	29.4%	8.2	2.4
36-39	24	9	37.5%	15.8	5.9
40-43	13	5	38.5%	11.8	4.5
44-47	11	4	36.4%	21.5	7.8
48-51	12	4	33.3%	14.3	4.8
52-55	6	0	0.0%	9.0	0
56-59	7	1	14.3%	0.0	1.3
≥60	14	2	14.3%	12	1.7
Totals	**476**	**136**	**28.6%**	**21.9**	**6.3**

9.2 Home IV Days

476 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	52	0	0.0%	0	0.0
4-7	51	5	9.8%	1.6	1.6
8-11	56	6	10.7%	1.4	1.4
12-15	56	11	19.6%	2.8	2.8
16-19	50	16	32.0%	4.6	4.6
20-23	41	5	12.2%	1.7	1.7
24-27	38	9	23.7%	5.6	5.6
28-31	28	6	21.4%	2.5	2.5
32-35	17	4	23.5%	1.8	1.8
36-39	24	7	29.2%	6.3	6.3
40-43	13	4	30.8%	6.2	6.2
44-47	11	3	27.3%	2.3	2.3
48-51	12	4	33.3%	4.3	4.3
52-55	6	0	0.0%	0	0.0
56-59	7	0	0.0%	0	0
≥60	14	1	7.1%	0.4	0.4
Totals	**476**	**81**	**17%**	**15.3**	**2.6**


KEEP OUT OF REACH OF CHILDREN
GO
GO MAGNE
Sandoz
1mg
SANDOZ
Batch No. JL1945
Exp Date: 03 2020
Take ONE
May cause
not stop
Take TWO capsules once a month on
the same day each month.

10. Complications

10.1 CF Related Diabetes By Age Group

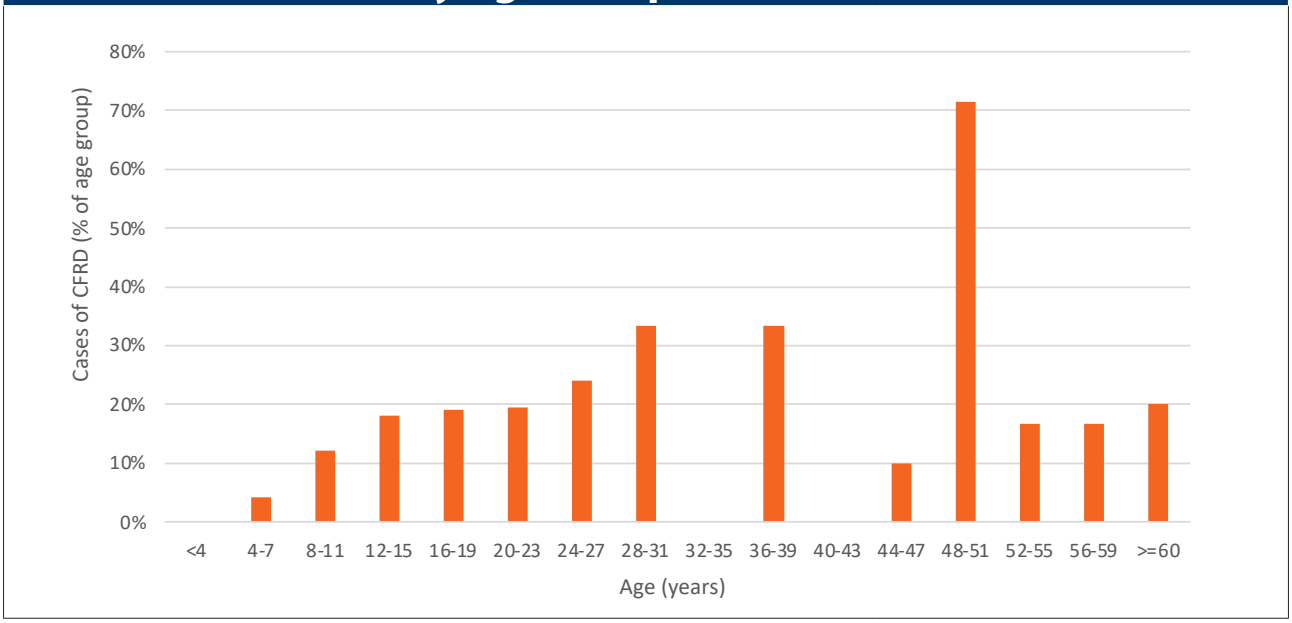
408 PWCF

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<4	46	0	0.0%	0.0%
4-7	47	2	4.3%	0.5%
8-11	49	6	12.2%	1.5%
12-15	50	9	18.0%	2.2%
16-19	47	9	19.1%	2.2%
20-23	36	7	19.4%	1.7%
24-27	29	7	24.1%	1.7%
28-31	21	4	33.3%	1.0%
32-35	11	0	0.0%	0.0%
36-39	21	7	33.3%	1.7%
40-43	12	0	0.0%	0.0%
44-47	10	1	10.0%	0.2%
48-51	7	5	71.4%	1.2%
52-55	6	1	16.7%	0.2%
56-59	6	1	16.7%	0.2%
≥60	10	2	20.0%	0.5%
Total	**408**	**61**		**15.0%**

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<16	192	17	8.9%	4.2%
≥16	216	44	20.4%	10.8%
Total	**408**	**61**		

CF Related Diabetes by Age Group

408 PWCF



There are fewer cases of CF Related diabetes in the paediatric group this year.

10.2 Liver Function by Ultra Sound

111 PWCF

		Normal		Abnormal	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	59	59	100.0%	0	0.0%
Adults	52	29	55.8%	23	44.2%
Total	**111**	**88**	**79.3%**	**23**	**20.7%**

10.3 Bone Density by DEXA Scans

73 PWCF

		Normal		Abnormal	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	18	18	100.0%	0	0.0%
Adults	55	32	58.2%	23	41.8%
Total	**73**	**50**	**68.5%**	**23**	**31.5%**

CYSTIC FIBROSIS NZ

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