Port CF 2020

New Zealand National Data Registry

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, 2020 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 2020 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 tenth Registry Report provides an increasingly accurate picture of people with CF in New Zealand and their outcomes with 97% of CF patients opting to provide this anonymised data, bringing the total number for 2020 to 572.

The 2020 Registry Report represents the most data captured on our population with CF, to inform future care and what future resources are needed.

We hope you find the information in this 2020 Report informative and useful.

Associate Professor Cass Byrnes
Chair Port CF Steering Committee
Port CFNZ Principal Investigator (2017 - 2020)

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

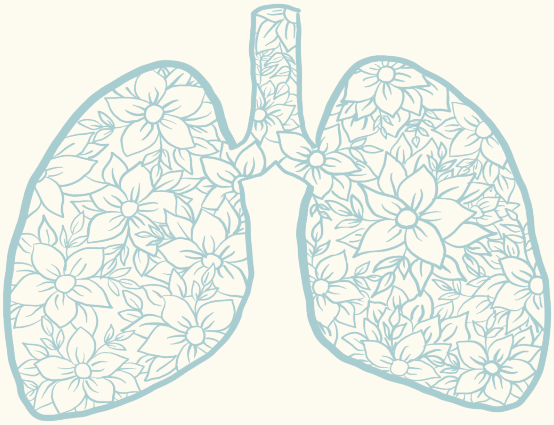
Canterbury (Paediatrics and Adults)
Christchurch Hospital, Christchurch

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 comparable to a single clinic in the United States (USA) or the United Kingdom (UK). The data collected from the New Zealand CF population provides our national statistics. Our small 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 New Zealand data and when cited as compared to other registries these are from Australia, the UK and the USA. Although there are a total of 572 people registered in Port CF, not all individuals had an input for every category. While the total is 572 (236 children under 16 years and 336 adults 16 years and over), the number of PWCF 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								572 PWCF
	2020	2019	2018	2017	2016	2015	2014	2013
CF Patients Registered	572	531	514	498	501	449	443	444
Diagnosis								
Diagnosis age <1 year	15	11	15	15	6	5	7	5
Diagnosis age <16 years	4	1	0	2	3			
Diagnosis age ≥16 years	0	2	0	1	2	0	2	3
Age								
Median Age (in years)	19	18.87	18.54	18.27	17.38	18.25	18.11	17.55
Mean Age (in years)	21.6	21.49	21.2	20.79	20.04			
PWCF <16 years								
Number	236	223	224	279	233	192	196	205
Percent	41.3%	42%	43.6%	56.0%	46.5%	42.8%	44.2%	46.2%
PWCF ≥16 years								
Number	336	308	290	219	268	257	247	239
Percent	58.7%	58%	56.4%	44.0%	53.5%	57.2%	55.8%	53.8%
Gender								
Males								
Number	319	297	285	273	275	247	240	240
Percent	55.8%	56%	55.4%	54.9%	54.9%	55.0%	54.2%	54.1%
Females								
Number	253	233	229	224	226	202	203	204
Percent	44.2%	44%	44.6%	45.1%	45.1%	45.0%	45.8%	45.9%
Genotyped								
Number	572	495	466	484	450	400	429	426
Percent	100.0%	93.2%	90.7%	97.4%	90.0%	89.1%	96.8%	95.9%
FEV1 (% predicted)								
Mean	78.4%	76.6%	81.8	85.1%	85.0%			
Median	82.8%	79%	86.2	86.5%	88.4%	85.6%	85.1%	84.3%
FEV1 < 16 Years								
Mean	91.4%	95.8%	96.70%	96.8%	97.3%			
Median	93.7%	97.9%	98.80%	99.3	99.3%	98.9%	97.7%	96.6%
FEV1 ≥16 Years								
Mean	71.7%	74.7%	75.30%	72.60%	72.6%			
Median	74.2%	76.8%	79.20%	77.4	77.4%	77.0%	78.0%	70.7%
FEV1 < 18 Years								
Mean	89.7%	94.5%	95.40%	95.1%	95.0%			
Median	93.1%	97.2%	98.30%	98.3%	98.0%			
FEV1 ≥18 Years								
Mean	70.9%	73.1%	73.7%	72.2%	71.2%			
Median	72.7%	74.7%	77.6%	75.6%	75.1%			

Key Indicators cont.

Key Indicators								
	2020	2019	2018	2017	2016	2015	2014	2013
FVC (% predicted)								
Mean	93.4%							
Median	96.0%							
FVC < 16 Years								
Mean	97.0%							
Median	98.3%							
FVC ≥16 Years								
Mean	91.5%							
Median	95.1%							
FVC < 18 Years								
Mean	96.9%							
Median	98.3%							
FVC ≥ 18 Years								
Mean	91.1%							
Median	93.7%							

For the first time, we are presenting FVC alongside FEV_1. We are also presenting the spirometry values for those under 16 years of age and under 18 years of age to compare with international registries that variably include their paediatric population as being up to 16 years or up to 18 years.

FEV1 has remained relatively stable over the last years.

There is an increase in the number of individuals who have been genotyped. In 2020, for the first time, it is 100 percent. This

1. Demographics

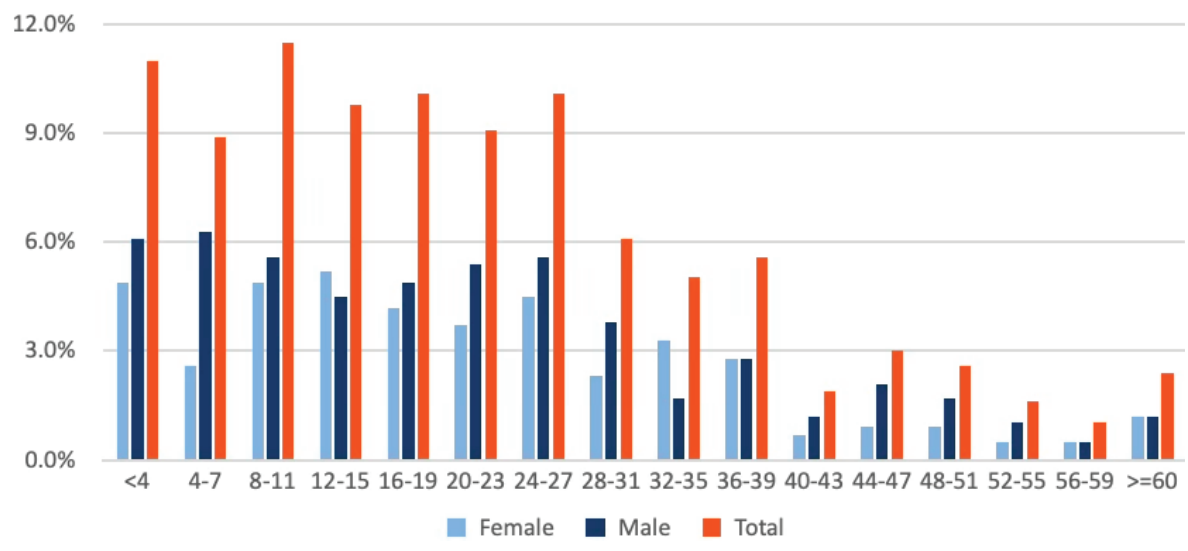
1.1 Age distribution

572 PWCF

Age Group	All		Male		Female	
0-3	63	11.0%	35	6.1%	28	4.9%
4-7	51	8.9%	36	6.3%	15	2.6%
8-11	66	11.5%	32	5.6%	34	4.9%
12-15	56	9.8%	26	4.5%	30	5.2%
16-19	58	10.1%	34	4.9%	24	4.2%
20-23	52	9.1%	31	5.4%	21	3.7%
24-27	58	10.1%	32	5.6%	26	4.5%
28-31	35	6.1%	22	3.8%	13	2.3%
32-35	29	5.1%	10	1.7%	19	3.3%
36-39	32	5.6%	16	2.8%	16	2.8%
40-43	11	1.9%	7	1.2%	4	0.7%
44-47	17	3.0%	12	2.1%	5	0.9%
48-51	15	2.6%	10	1.7%	5	0.9%
52-55	9	1.6%	6	1.0%	3	0.5%
56-59	6	1.0%	3	0.5%	3	0.5%
>=60	14	2.4%	7	1.2%	7	1.2%
Total	**572**		**319**	**55.8%**	**253**	**44.2%**
Median	**19**					
Range	**0 - 78**					

Age distribution

572 PWCF

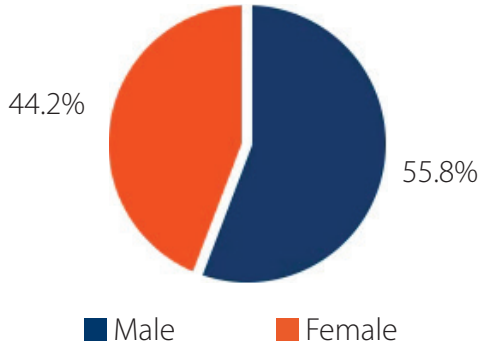


The median age of persons with CF in New Zealand has increased from 16 to 19 years over the ten years that we have had Registry data. Children in international registries are defined as either up to 16 years or up to 18 years of age. In New Zealand, if we include children as being up to 16 years we have 236 children (41.1% total) and 336 adults (58.9% total) and if we include children as being up to 18 years, which is our more usual clinical practice, we have 262 children (45.8% total) and 310 adults (54.2% total).

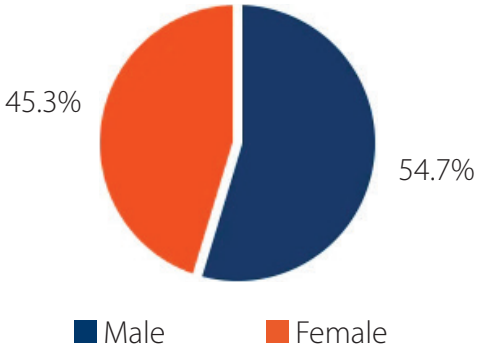
1.2 Gender Distribution — 572 PWCF

	All		<16		≥16	
	Number in age group	**Percent** of PWCF	**Number** in age group	**Percent** of PWCF	**Number** in age group	**Percent** of PWCF
Male	319	55.8%	129	54.7%	190	56.6%
Female	253	44.2%	107	45.3%	146	43.5%
Totals	**572**	**100.0%**	**236**	**41.3%**	**336**	**58.7%**

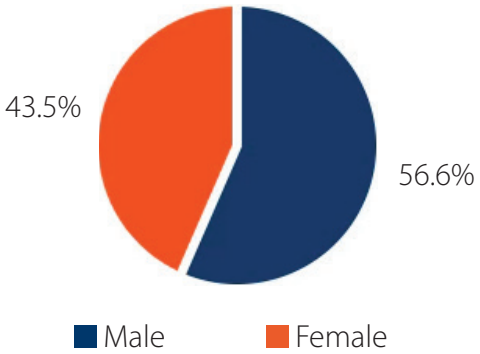
Gender Distribution of All PWCF — 572 PWCF



Gender Distribution <16 Years — 236 PWCF



Gender Distribution ≥16 Years — 336 PWCF



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

2. Genotypes

2.1 Numbered and Percent Genotyped — 570 PWCF

Mutations	Number of PWCF Genotyped	Percentage of PWCF Genotyped
Homozygous F508del	268	46.9%
Heterozygous F508del	231	40.4%
No F508del	71	12.8%
Total	**570**	

2.2 Second Allele of Heterozygous F508del — 231 PWCF

Second Allele	c.DNA Name	Number of PWCF	Percent of PWCF
R117H	c.350G>A	27	4.7%
G551D	c.1652G>A	24	4.2%
G542X	c.1624G>T	21	3.7%
3849+10kbC->T	c.3717+12191C>T	9	1.6%
N1303K	c.3909c>G	7	1.2%
1717-1G->A	c.1585-1G>A	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%
^I507	c.1519_1521delATC	4	0.7%
621+1G->T	c.489+1G>T	3	0.5%
D1152H	c.3454G>C	3	0.5%
Q493X	c.1477C>T	3	0.5%
1078delT	c.948delT	2	0.4%
A455E	c.1364C>A	2	0.4%
E60X	c.178G>T	2	0.4%
L206W	c.617T>G	2	0.4%
R352Q	c.1055G>A	2	0.4%
R560T	c.1679G>C	2	0.4%
Y563D	c.1687T>G	2	0.4%
1898+5G->T	c.1766+5G>T	1	0.2%
2789+5G->A	c.2657+5G>A	1	0.2%
3120+1G->A	c.2988+1G>A	1	0.2%
D1270N	c.3808G>A	1	0.2%
P574H	c.1721C>A	1	0.2%
R1158X	c.3472C>T	1	0.2%
R1162X	c.3484C>T	1	0.2%
R117C	c.349C>T	1	0.2%
R334W	c.1000C>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
Other		81	14.2
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 71 people in New Zealand do not have at least one F508del mutation. Looking at the gene mutations recorded in 2020 Registry, 44 of the 572 people who have been genotyped (7.7%) would not be detected by the current new born screening programme.

2.3 No F508del Mutations

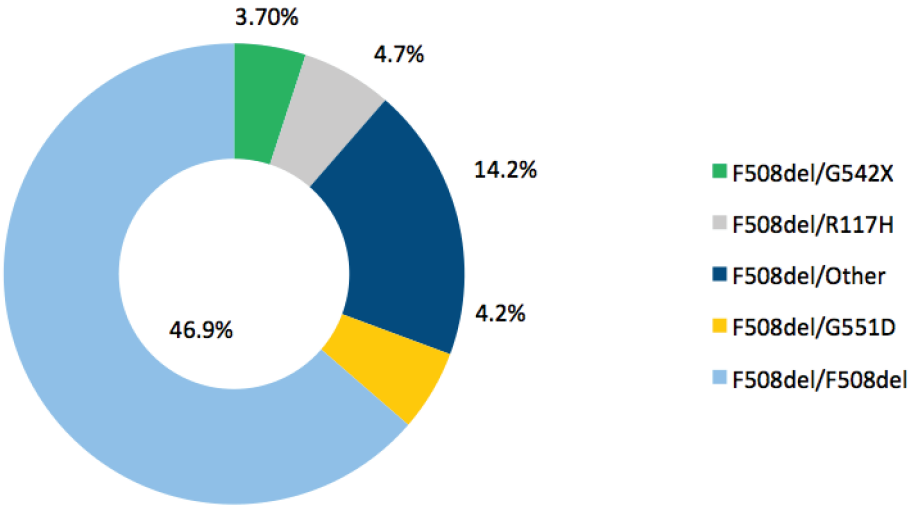
71 PWCF

	1717-1G->A	2143delT	3849+10kbC->T	621+1G->T	G542X	G551D	N1303K	Not Identified	Other	Q493X	R117H	R553X	W1282X	^I507
1078delT	0	0	0	0	0	0	0	0	1	0	0	0	0	0
1161delC	0	0	0	0	0	0	0	0	2	0	0	0	0	0
2789+5G->A	0	0	0	0	0	0	0	0	1	0	0	0	0	0
3120+1G->A	0	0	0	0	0	0	0	0	2	0	0	0	0	0
3849+10kbC->T	1	0	0	0	0	0	0	0	0	0	0	0	0	0
621+1G->T	0	0	0	0	0	0	0	0	1	0	0	0	0	0
A455E	0	0	0	0	0	0	0	0	1	0	0	0	0	0
G178R	0	0	0	0	0	0	0	0	1	0	0	0	0	0
G542X	0	1	0	1	1	0	0	1	0	0	2	0	0	0
G551D	1	0	1	0	1	1	1	2	4	4	5	1	0	0
G85E	0	0	0	0	0	0	0	0	1	0	0	0	0	0
N1303K	0	0	0	0	0	0	0	0	1	0	0	0	0	0
Not Identified	0	0	0	0	0	0	0	8	0	0	0	0	0	0
Other	0	0	0	0	0	0	0	3	8	0	0	0	0	0
R1162X	0	0	0	0	0	0	0	0	1	0	0	0	0	0
R117H	0	0	0	0	0	0	0	3	1	1	0	0	1	0
R553X	0	0	0	0	0	0	0	0	1	0	0	0	0	0
S549N	0	0	0	0	0	0	0	1	0	0	0	0	0	0
W1282X	0	0	0	0	0	0	0	1	2	0	0	0	0	1

2.4 Genotype Major Categories

Mutations	Number	Percentage
F508del	499	87.2%
G551D	44	7.7%
R117H	40	7.0%
G542X	26	4.5%

2.5 Common Genotype Combinations

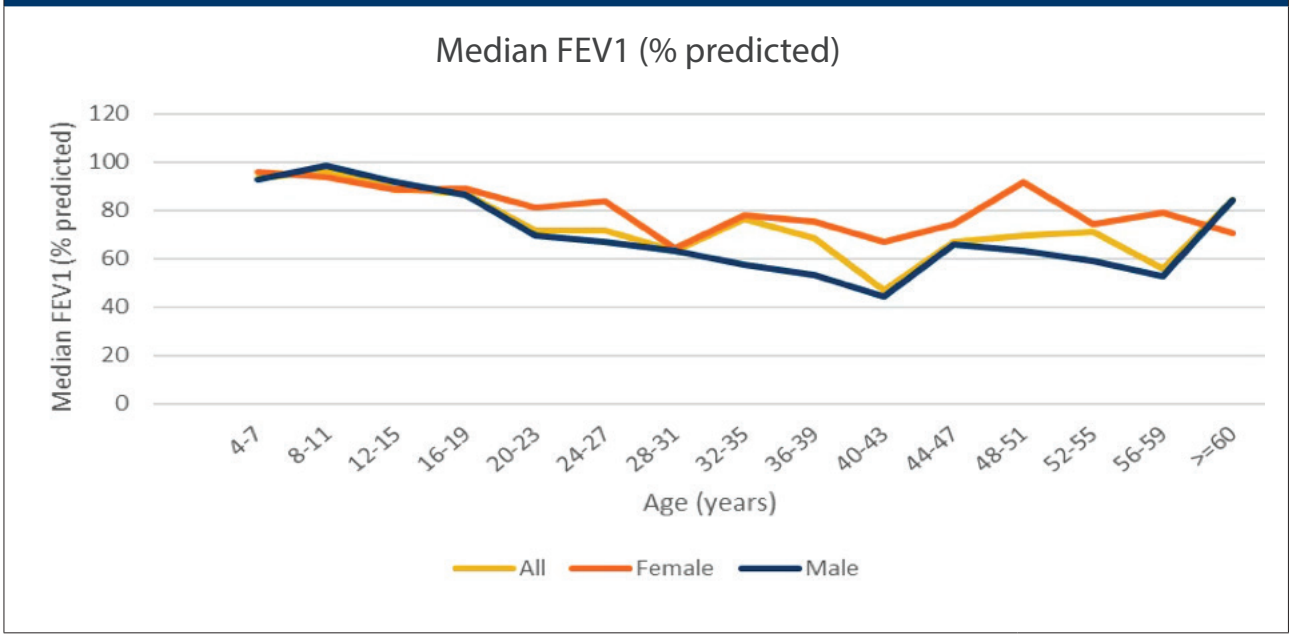


3. Respiratory

3.1 Median FEV_1 — 405 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
5-7	38	93.40	11	95.97	27	92.61
8-11	62	96.56	32	94.06	30	98.36
12-15	52	88.94	28	88.67	24	91.87
16-19	50	86.99	19	89.11	31	86.67
20-23	40	72.00	15	81.46	25	69.78
24-27	41	71.68	16	83.86	25	67.10
28-31	21	63.30	6	64.21	15	63.22
32-35	23	76.32	17	78.12	6	57.65
36-39	24	68.43	10	75.34	14	53.27
40-43	7	46.85	3	66.95	4	44.38
44-47	12	66.78	5	74.19	7	65.91
48-51	12	69.89	4	91.54	8	63.59
52-55	7	71.17	2	74.48	5	59.16
56-59	5	55.78	3	79.06	2	52.97
>=60	11	84.22	5	70.94	6	84.22
Totals	**405**		**176**		**229**	

Median FEV_1 — 405 PWCF



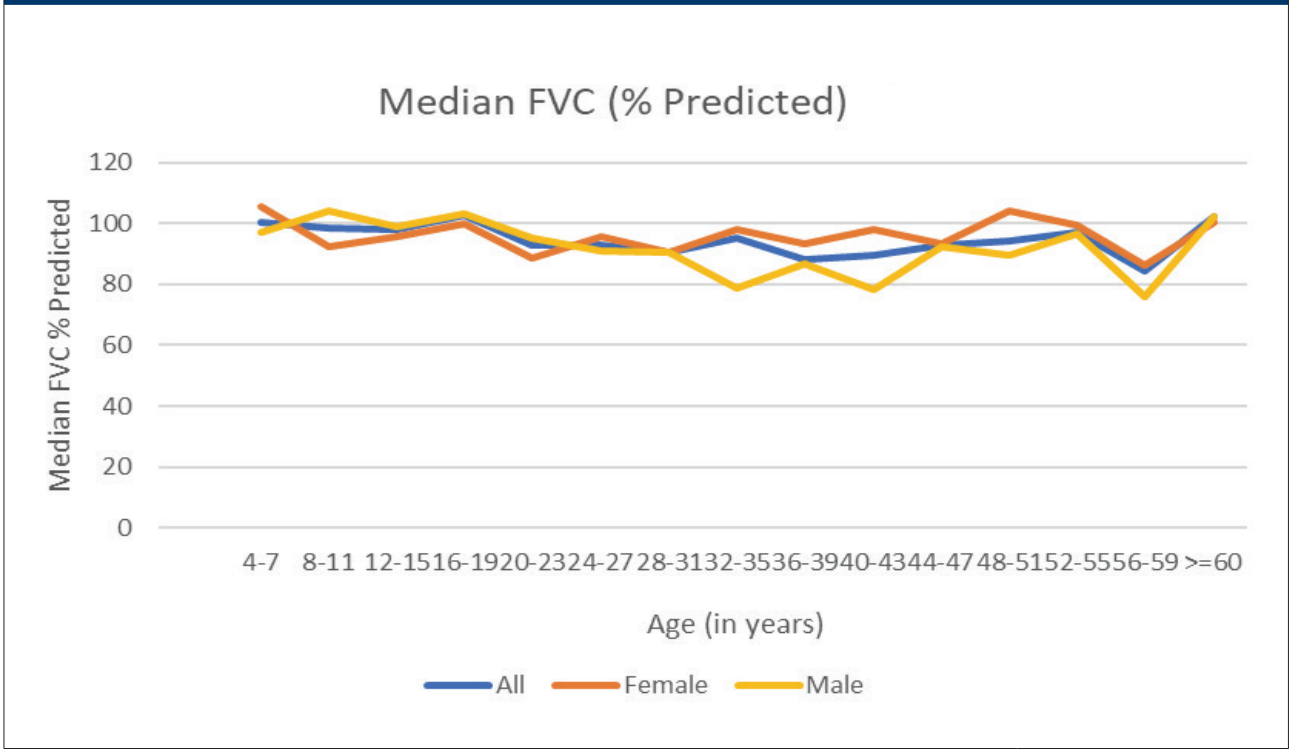
3.2 Median FVC

405 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	100.6	11	105.5	27	97.3
8-11	62	98.3	32	92.6	30	104.2
12-15	52	98.1	28	95.5	24	98.9
16-19	50	102.9	19	99.9	31	103.0
20-23	40	92.7	15	88.4	25	95.2
24-27	41	93.0	16	95.5	25	90.9
28-31	21	90.6	6	90.5	15	90.6
32-35	23	95.0	17	98.2	6	78.8
36-39	24	88.3	10	93.3	14	86.9
40-43	7	89.5	3	97.9	4	78.3
44-47	12	93.0	5	93.5	7	92.5
48-51	12	94.3	4	104.0	8	89.4
52-55	7	96.9	2	99.3	5	96.5
56-59	5	84.2	3	86.5	2	75.9
≥60	11	102.1	5	100.4	6	102.1
Totals	**405**		**176**		**229**	

Median FVC

405 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 82.8% (93.7% in children, 74.2% 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, FEV_1 is an important prognostic indicator.

4. Nutrition

4.1 Paediatric BMI

194 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	27	64.8	<4	11	72.4	<4	16	59.2
4-7	48	63.0	4-7	15	68.8	4-7	33	61.0
8-11	64	56.0	8-11	34	58.3	8-11	30	53.0
12-15	55	54.3	12-15	30	56.1	12-15	25	49.7
Totals	**194**			**90**			**104**	

4.2 Adult BMI

247 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	46	20.6	16-19	18	20.8	16-19	28	20.3
20-23	40	21.7	20-23	15	22.8	20-23	25	21.1
24-27	41	22.6	24-27	16	22.8	24-27	25	22.3
28-31	21	21.6	28-31	6	22.0	28-31	15	21.3
32-35	22	23.2	32-35	16	22.0	32-35	6	26.8
36-39	24	22.9	36-39	10	22.1	36-39	14	23.0
40-43	7	25.0	40-43	3	22.2	40-43	4	25.4
44-47	12	23.6	44-47	5	22.1	44-47	7	23.9
48-51	12	25.6	48-51	4	29.9	48-51	8	24.6
52-55	7	24.9	52-55	2	26.6	52-55	5	24.6
56-59	5	27.6	56-59	3	23.2	56-59	2	29.3
≥60	10	23.2	60+	5	22.3	60+	5	24.0
Totals	**247**			**103**			**144**	

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:

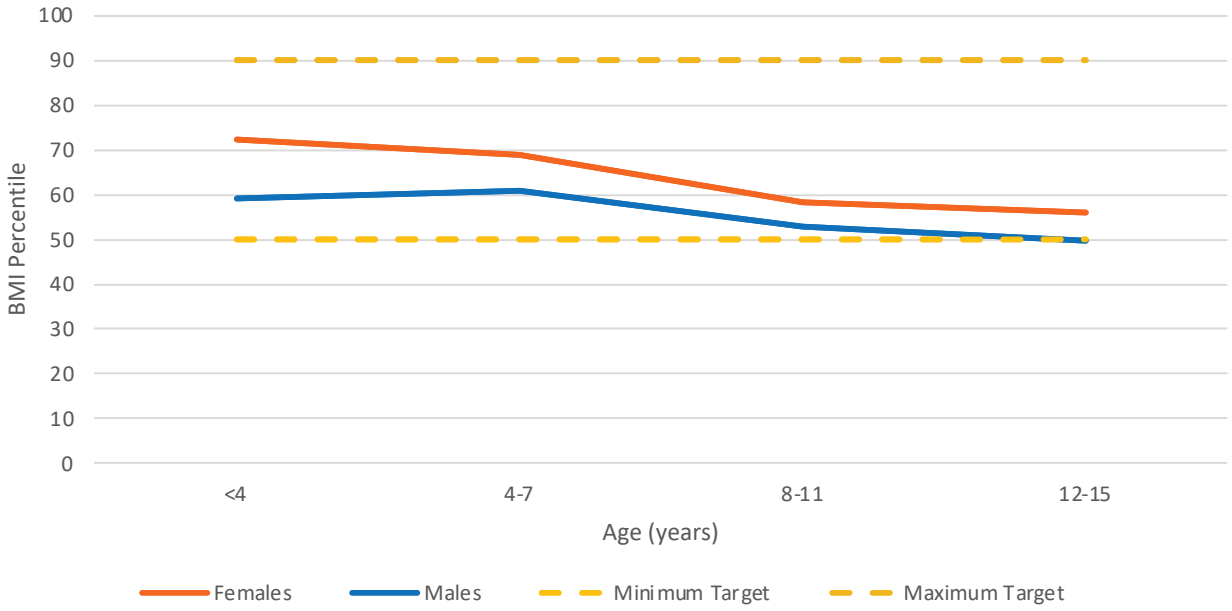
1) Infants (0 to 24 months): weight-for-length ≥50th percentile using WHO growth charts.
2) Children and adolescents (2-18 years): BMI 50-85th percentile (CDC growth charts) or 50- 91st percentile (WHO growth charts.)
3) Adults: males BMI 23 - 27 kg/m2, females BMI 22 - 27 kg/m2.

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 64.8 percentile. For children and adolescents, the median BMI is 57.1 percentile. For adults, 42.7% of males and 52.8% of females are above the minimum target range.

4.3 Median BMI Percentile <16 Years

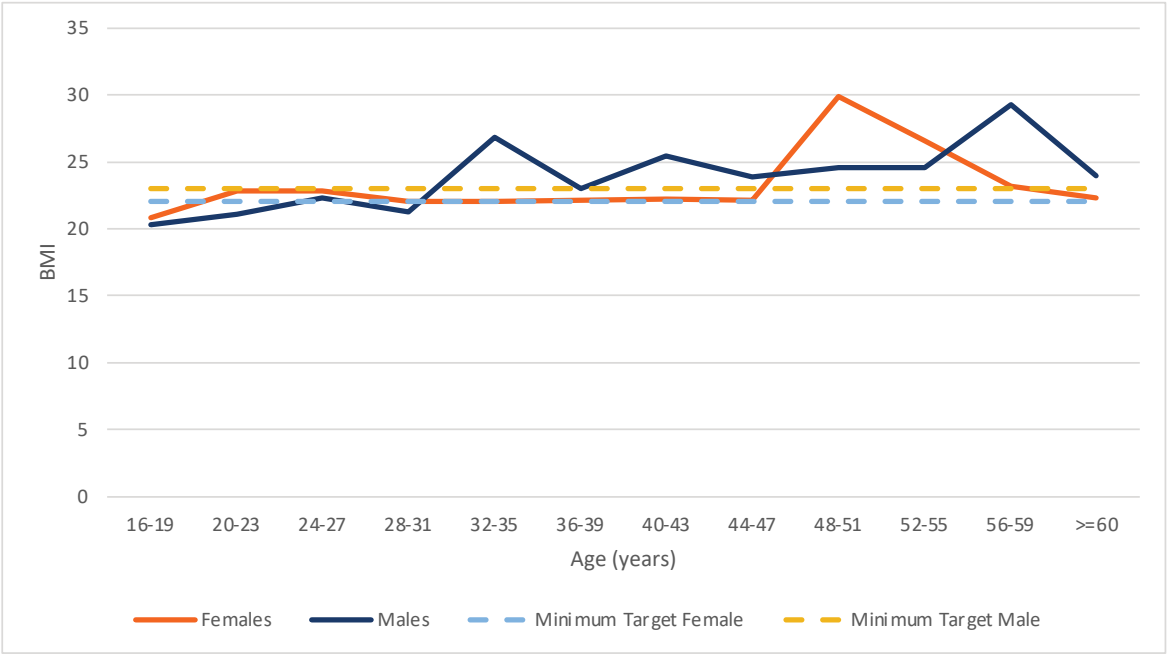
194 PWCF



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

247 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.
It is noted that the BMI for <4 years of age has dropped compared to previous years which has been checked and is an accurate finding. There is currently no explanation for this.

4.5 Supplemental Feeding

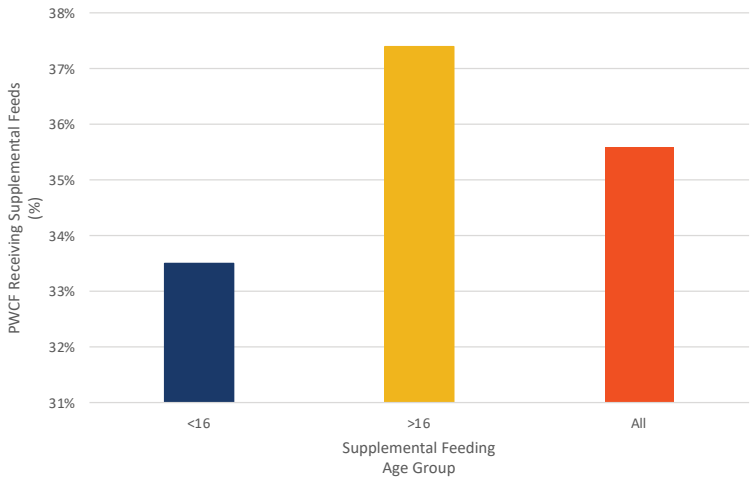
486 PWCF

	<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	**% >=16 years 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.

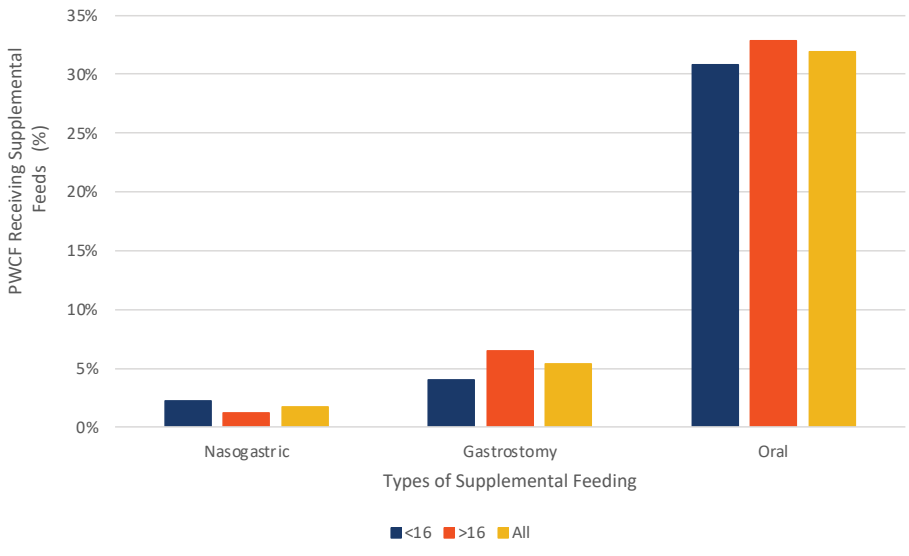
4.6 Supplemental Feeding by Age Group

486 PWCF



4.7 Supplemental Feeding by Type

486 PWCF



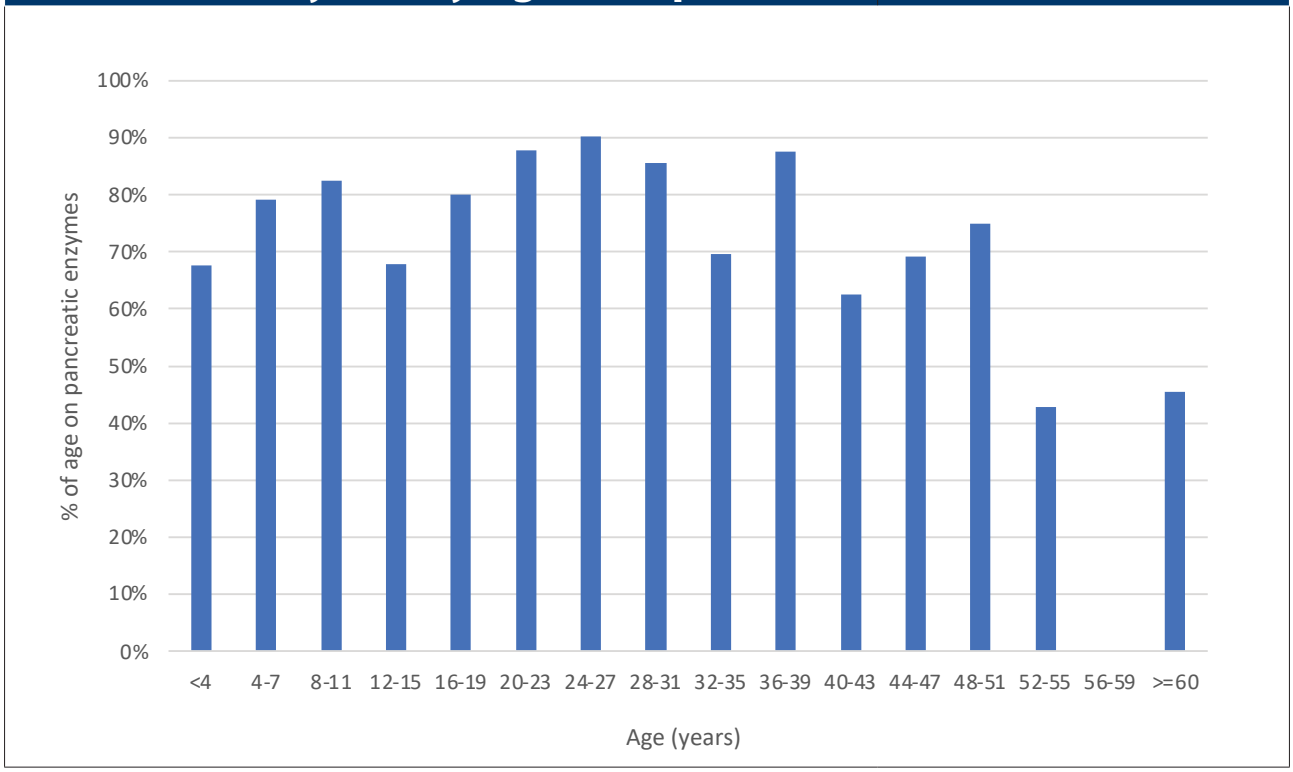
There has been no significant change in the numbers of children and adults on supplemental feeding this year.

5. Pancreatic Enzymes

5.1 Pancreatic Enzymes by Age Group — 482 PWCF

Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of PWCF
<4	62	42	67.7%	8.7%
4-7	48	38	79.2%	7.9%
8-11	63	52	82.5%	10.8%
12-15	53	36	67.9%	7.5%
16-19	50	40	80.0%	8.3%
20-23	41	36	87.8%	7.5%
24-27	41	37	90.2%	7.7%
28-31	21	18	85.7%	3.7%
32-35	23	16	69.6%	3.3%
36-39	24	21	87.5%	4.4%
40-43	8	5	62.5%	1.0%
44-47	13	9	69.2%	1.9%
48-51	12	9	75.0%	1.9%
52-55	7	3	42.9%	0.6%
56-59	5	0	0.0%	0.0%
≥60	11	5	45.5%	1.0%
Totals	**482**	**367**		**78.2%**

Pancreatic Enzymes by Age Group — 367 PWCF



Of the 482 PWCF for whom there is data, 367 are on pancreatic enzymes and 115 are not.

19

6. Airway Clearance Techniques

6.1 Primary Airway Clearance

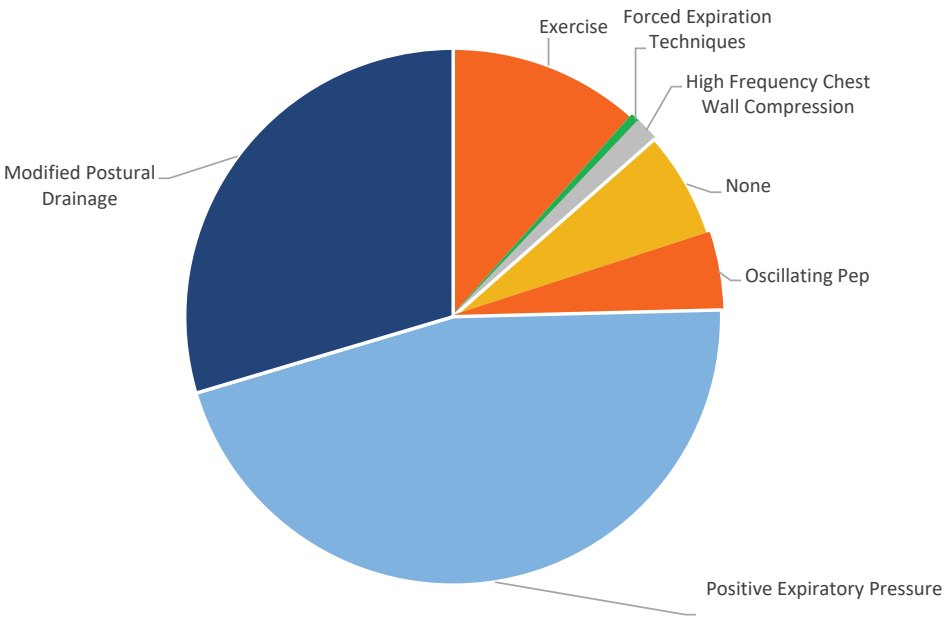
461 PWCF

	<16 years, n= 216	
	Number of Patients	Percent of Patients
Positive Expiratory Pressure	99	45.8%
Modified Postural Drainage	64	29.6%
Exercise	25	11.6%
Oscillating Pep (e.g. Flutter, Acapella, IPV)	10	4.6%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	1	0.5%
High Frequency Chest Wall Compression (e.g. vest)	3	1.4%
None	14	6.5%

	≥16 years, n = 245	
	Number of Patients	Percent of Patients
Positive Expiratory Pressure	99	40.4%
Modified Postural Drainage	1	0.4%
Exercise	67	27.4%
Oscillating Pep (e.g. Flutter, Acapella, IPV)	36	14.7%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	25	10.2%
High Frequency Chest Wall Compression (e.g. vest)	1	04%
None	16	6.5%

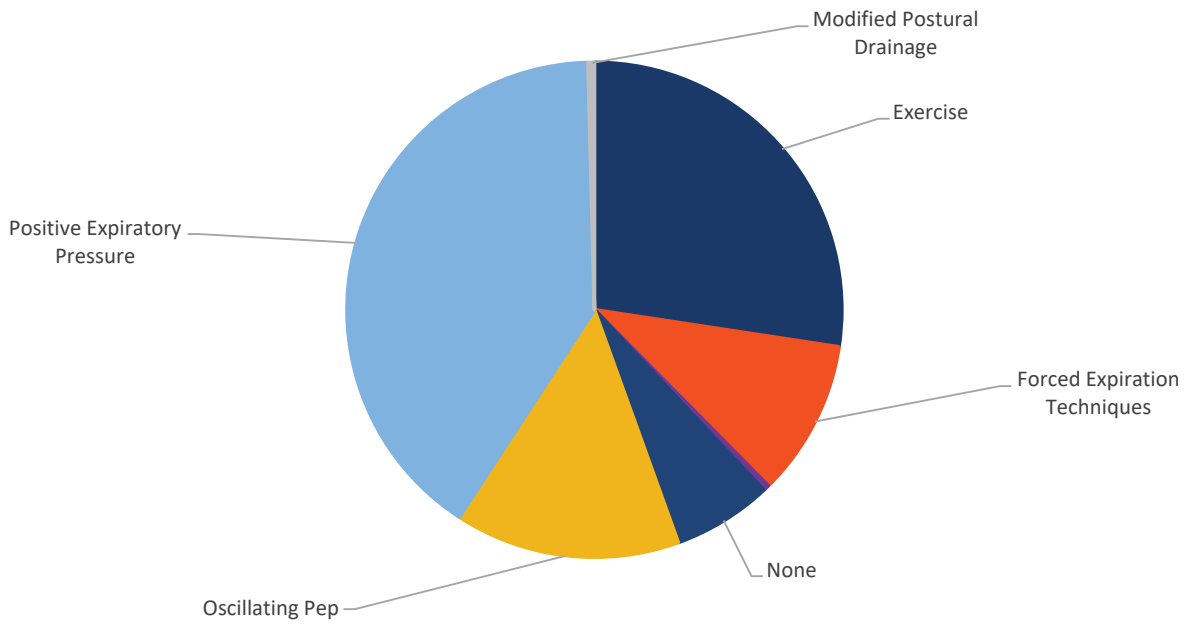
6.2 Primary Airway Clearance <16 years

216 PWCF



6.3 Primary Airway Clearance ≥16 years

245 PWCF



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

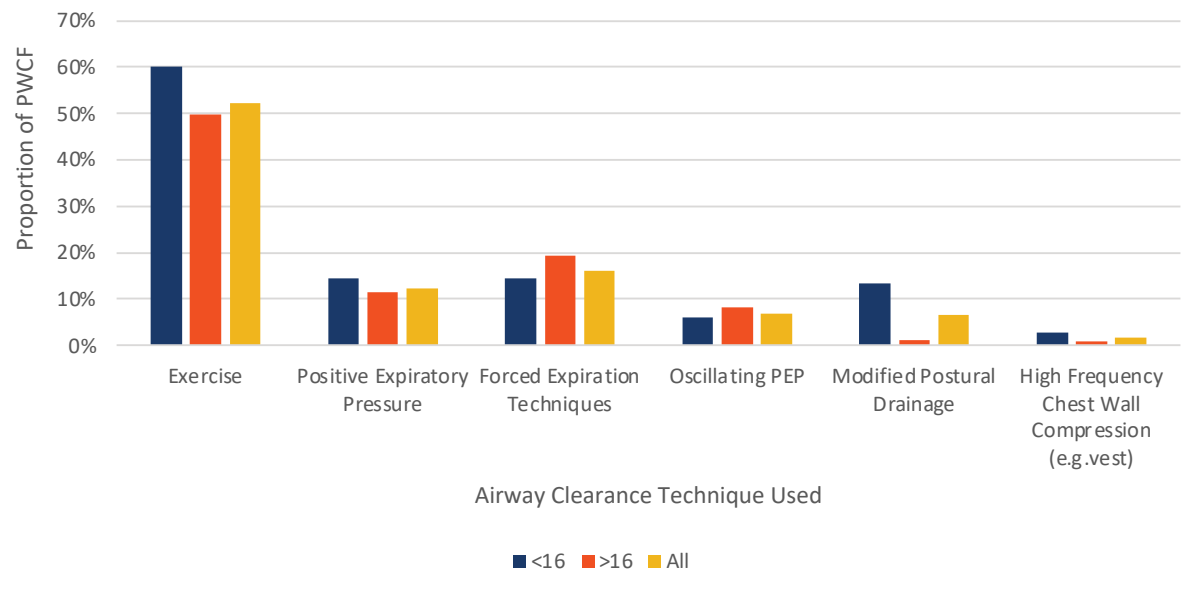
The nominated primary airway clearance technique adds up to greater than 100% because, over the clinic visits for a year, an individual may use differing techniques as his/her main option at different times.

Over the years of the Registry reports, there has been a trend for an increased percentage of children to nominate 'none' as their primary airway clearance (from 2.3% in 2013 to 6.5% in 2020). The trend in adults is the opposite, dropping from 18.8% in 2013 to 6.5% in 2020. There has been a corresponding drop in the numbers for both children and adults who have nominated Positive Expiratory Pressure as their preferred option.

There is a drop in children selecting exercise as their primary airway clearance technique.

6.4 Secondary Airway Clearance

461 PWCF



7. Microbiology

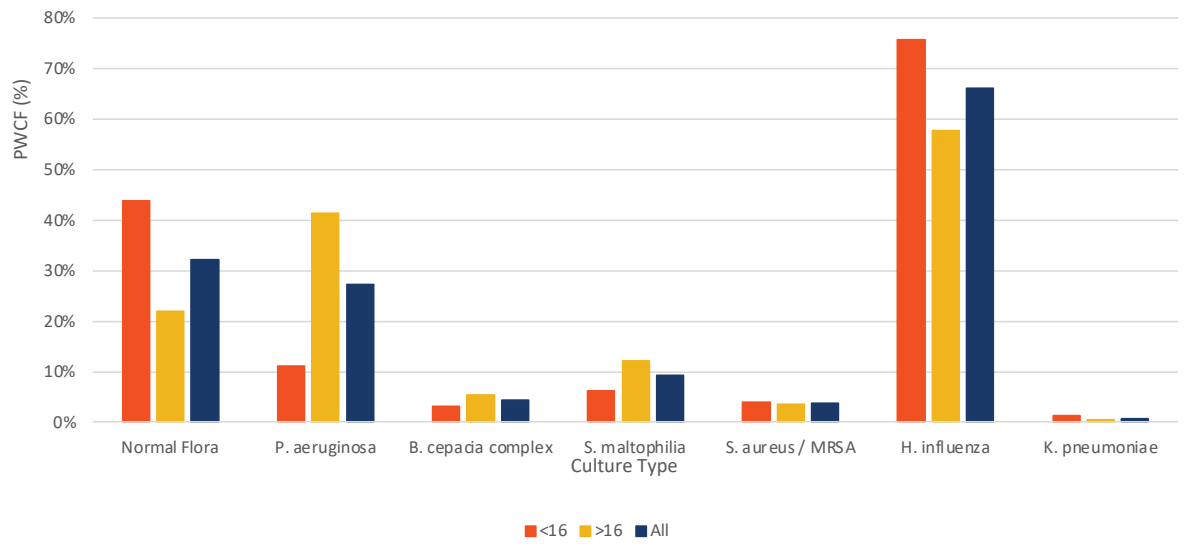
7.1 Bacterial Culture Prevalence

482 PWCF

	<16 years 226 PWCF		≥16 years 256 PWCF		Total PWCF	
	Number	Percent	Number	Percent	Number	Percent
Normal Flora	99	43.8%	56	21.9%	155	32.2%
Haemophilus Influenza	171	75.7%	148	57.8%	319	66.2%
Escherichia coli	6	2.7%	3	1.2%	9	1.9%
Klebsiella Pneumoniae	3	1.3%	1	0.4%	4	0.8%
Stenotrophomonas maltophilia	14	6.2%	31	12.1%	45	9.3%
Pseudomonas Aeruginosa	**25**	**11.1%**	**106**	**41.4%**	**131**	**27.2%**
Mucoid	6	2.7%	73	28.5%	79	16.4%
Non Mucoid	18	8.0%	71	27.7%	89	18.5%
Staphylococcus Aureus	**120**	**53.1%**	**129**	**50.4%**	**249**	**51.7%**
MSSA	111	49.1%	120	46.9%	231	47.9%
MRSA	9	4.0%	9	3.5%	18	3.7%
Burkholderia Cepacia Complex	**7**	**3.1%**	**14**	**5.5%**	**21**	**4.4%**
Cenocepacia	1	0.4%	2	0.8%	3	0.6%
Multivorans	4	1.8%	5	2.0%	9	1.9%
Other	43	19.0%	24	9.4%	67	13.9%

Bacterial Culture Prevalence

482 PWCF



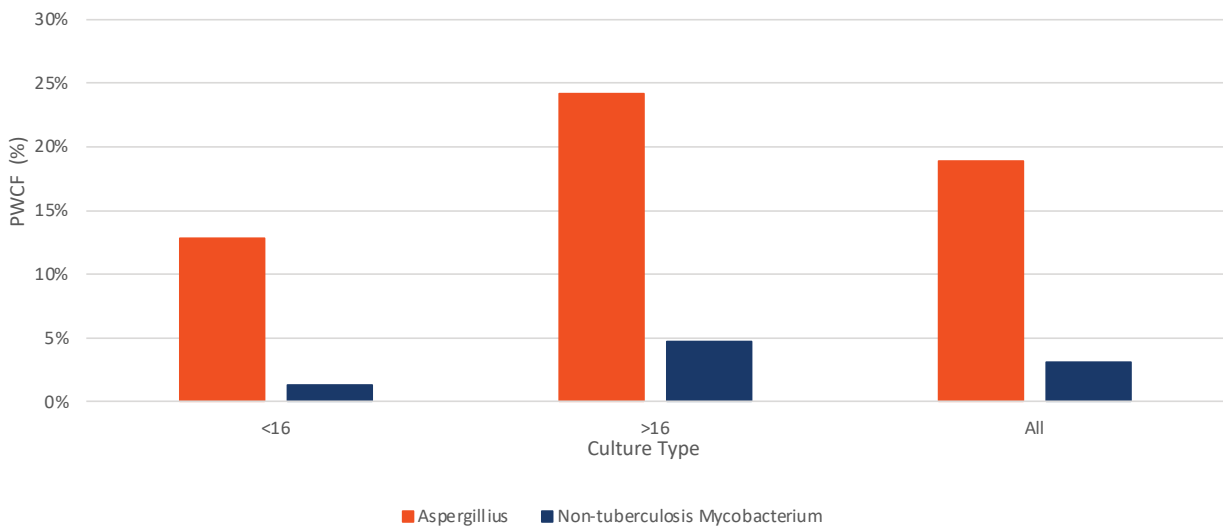
The percentages of the CF population having had specific respiratory pathogens identified such as Staphylococcus aureus, Pseudomonas aeruginosa etc. are very similar to other international registries, with the exception of much higher percentages of Haemophilus influenzae in New Zealand.

Pseudomonas aeruginosa was found in 11.1% of children. This is a reduction of more than 30% of positives in the previous year and we suspect this reflects COVID19 lockdown periods when less respiratory samples were taken. Our MRSA rates are relatively low at 3.7% overall.

There are significant changes in some of the respiratory cultures with Normal flora dropping from 44% (total) in 2019 to 32.2% (total) in 2020. There has been an increase in positive Haemophilus influenzae from 22.8% (total) in 2019 to 66.2% (total) in 2020. This may reflect the increased number of sputum cultures being taken as we worked to assess the impact of COVID19 on CF respiratory health.

7.2 Nonbacterial/Fungal Prevalence

482 PWCF



8. Medications

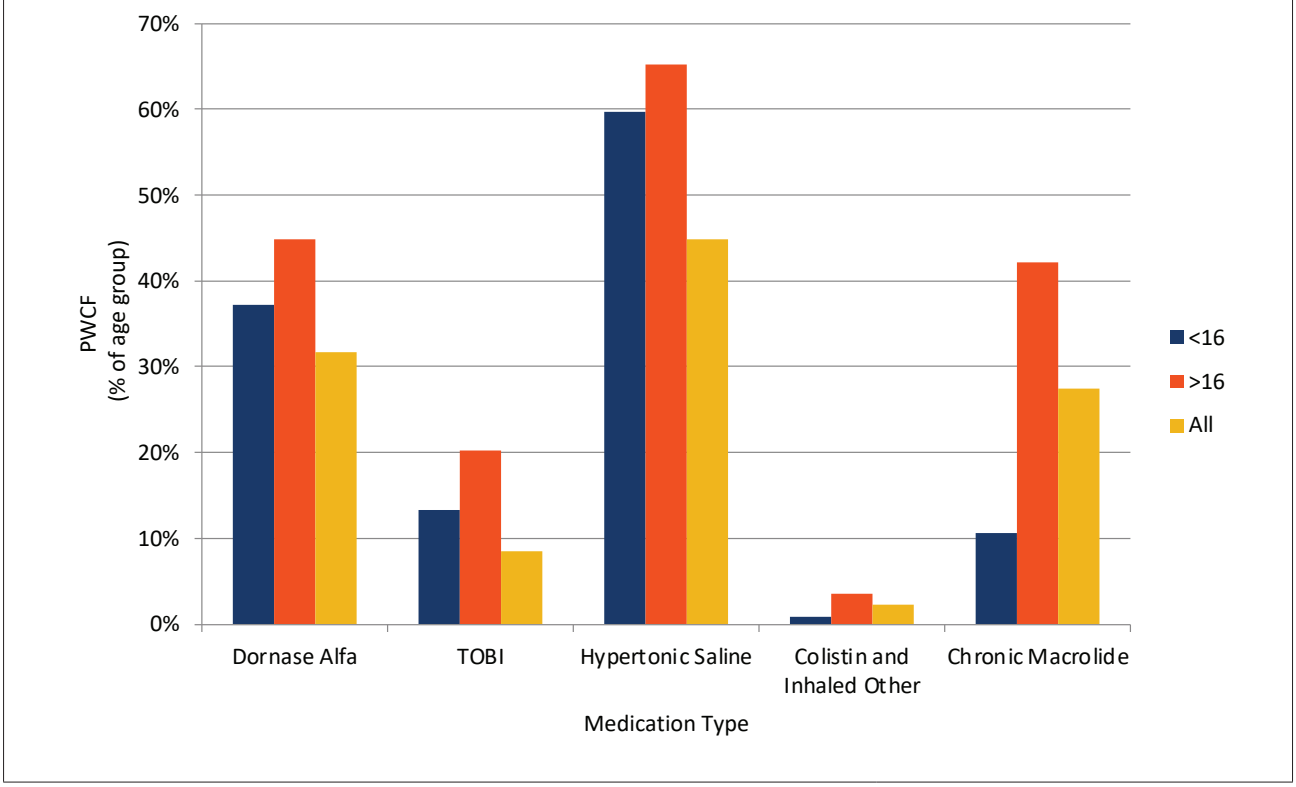
8.1 Medications Prescribed

467 PWCF

Medication	<16 Years	≥16 Years	All
Hypertonic Saline	59.7%	65.2%	62.7%
Dornase alfa	37.2%	44.9%	41.3%
TOBI	13.3%	20.3%	17.0%
Chronic Macrolide	10.6%	45.2%	27.4%
Corticosteroids Inhaled	9.7%	10.6%	10.2%
Corticosteroids Oral	2.7%	5.1%	3.9%
Antifungals	2.2%	3.1%	2.7%
Influenza Vaccine	68.8%	67.9%	68.3%

Medications Prescribed

467 PWCF



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. We have had no access to some newer medications except on research programmes - notably some of the modulator drugs.

In 2020, Pharmac funded the first modulator drug Ivacaftor for 9 eligible CFTR genetic mutations from March 2020. We saw 13.3% less children, but 11.2% more adults, receiving the Influenza vaccine this year.

9. Intravenous Antibiotic Treatment

9.1 Home IV Days

470 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	5	9.6%	14.8	1.4
4-7	48	9	18.8%	10.6	2
8-15	62	20	32.3%	15.3	4.9
16-18	53	18	34.0%	27.8	9.4
16-19	49	25	51.0%	29.4	15
20-23	41	18	43.9%	27.3	12
24-27	40	11	27.5%	18.9	5.2
28-31	21	7	33.3%	13.4	4.5
32-35	21	8	38.1%	17.9	6.8
36-39	26	11	42.3%	16.1	7.1
40-43	8	1	12.5%	77	9.6
44-47	14	3	21.4%	13.3	3.1
48-51	12	5	41.7%	23.2	9.7
52-55	6	0	0.0%	-	-
56-59	5	1	20.0%	8	1.6
≥60	12	0	0.0%	-	
Totals	**470**	**142**	**30.2**	**21.6**	**6.6**

9.2 Home IV Days

470 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	1	1.9%	11.0	0.2
4-7	48	8	16.7%	15.4	2.6
8-11	62	12	19.4%	15.7	3
12-15	53	10	18.9%	13.8	2.6
16-19	49	14	28.6%	31.1	8.9
20-23	41	12	29.3%	15.3	4.5
24-27	40	7	17.5%	17.9	3.1
28-31	21	5	23.8%	15.8	3.8
32-35	21	7	33.3%	22.9	7.6
36-39	26	10	38.5%	15.6	6.2
40-43	8	2	25.0%	15.5	3.9
44-47	14	2	14.3%	16.5	2.5
48-51	12	4	33.3%	22.5	7.5
52-55	6	0	0.0%	-	-
56-59	5	0	0.0%	-	-
≥60	12	0	0.0%	-	-
Totals	**470**	**94**	**20%**	**18.7**	**3.75**

10. Complications

10.1 CF Related Diabetes by Age Group

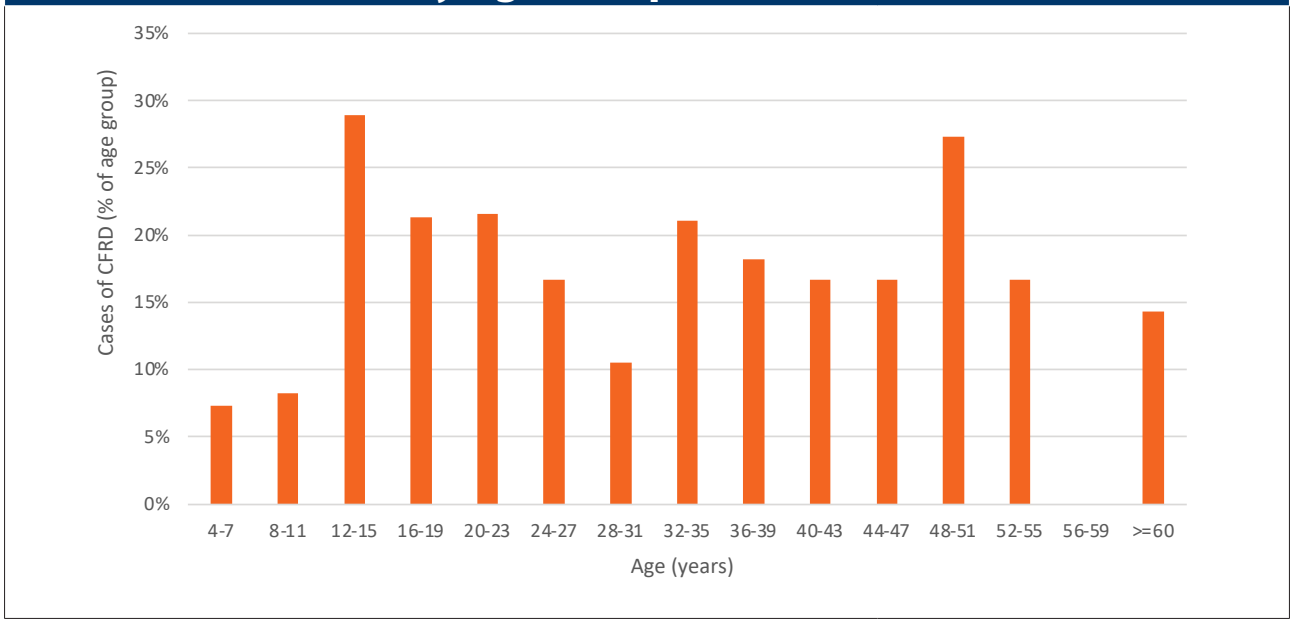
416 PWCF

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<4	47	0	0.0%	0.0%
4-7	41	3	7.3%	0.7%
8-11	49	4	8.2%	1.0%
12-15	52	15	28.9%	3.6%
16-19	47	10	21.3%	2.4%
20-23	37	8	21.6%	1.9%
24-27	36	6	16.7%	1.4%
28-31	19	2	10.5%	0.5%
32-35	19	4	21.1%	1.0%
36-39	22	4	18.2%	1.0%
40-43	6	1	16.7%	0.2%
44-47	12	2	16.7%	0.5%
48-51	11	3	27.3%	0.7%
52-55	6	1	16.7%	0.2%
56-59	5	0	0.0%	0.0%
≥60	7	1	14.3%	0.2%
Total	**416**	**64**		**15.4%**

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<16	189	22	11.6%	5.3%
≥16	227	42	18.5%	10.1%
Total	**416**	**64**		**15.4%**

CF Related Diabetes by Age Group

416 PWCF



There has been little change in the number of those with CF Related diabetes this year.

10.2 Liver Function by Ultra Sound — 153 PWCF

		Normal		Abnormal	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	99	81	81.8%	18	18.2%
Adults	54	36	66.7%	18	33.3%
Total	**153**	**117**	**76.5**	**36**	**23.5%**

10.3 Bone Density by DEXA Scans — 93 PWCF

		Normal		Abnormal	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	34	32	94.1%	2	5.9%
Adults	59	35	59.3%	24	40.7%
Total	**93**	**67**	**72.0%**	**26**	**28%**

CYSTIC FIBROSIS NZ

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