Port CFNZ

2015

National Data Registry

CYSTIC FIBROSIS NZ

The Port CFNZ National Data Registry is a research project of

Cystic Fibrosis New Zealand

For further information about CFNZ

visit www.cfnz.org.nz

Published 29 March 2019

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 this national registry

Suggested Citation: Port CFNZ National Data Registry, 2015 Registry Report, Cystic Fibrosis New Zealand. http://cfnz.org.nz/

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Introduction & Acknowledgements

Cystic Fibrosis New Zealand and the Port CFNZ Steering Committee are pleased to present The National Cystic Fibrosis Data Registry 2015 Report; data collected from children, young persons and adults with Cystic Fibrosis in New Zealand.

We would like to thank:

- The children and adults with CF and their families for participating in this process.
- CFNZ for providing pivotal funding for database and data entry.
- The Nurses, Specialists and Administrators who have worked to enter data, enabling a detailed analysis for NZ – presented in this report.
- Canterbury District Health Board for their on-going information technology service to maintain the registry.

This fifth registry report continues to give an accurate picture of people with CF and outcomes for New Zealand with greater than 95% opting to provide anonymous data.

We plan on developing the database further over the next years. Firstly, with possible resourcing for data entry and analysis, secondly, in considering changes to questions to more closely align to other international registries. There is a group working on 'harmonisation' of data registries for cystic fibrosis involving representation from all countries that have a CF registry.

We hope you continue to find the information in the report informative and useful.

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

Dr Richard Laing — ***Port CFNZ Principal Investigator (2011-2016)***

Port CFNZ Steering Committee

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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)
Taranaki Base Hospital, New Plymouth

Bay of Plenty (Paediatrics)
Tauranga Hospital, Tauranga
Whakatane Hospital, Whakatane
Lakes Hospital, Rotorua

Central Districts (Paediatrics and Adults)
Whanganui Hospital, Whanganui
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 and Adults)
Nelson Hospital, Nelson
Wairau Hospital, Blenheim

Canterbury/ Westland (Paediatrics and Adults)
Christchurch Hospital, Christchurch

Otago (Paediatrics and Adults)
Dunedin Hospital, Dunedin

Southland (Paediatrics and Adults)
Kew Hospital, Invercargill

Notes to the Registry

New Zealand has a total CF population comparative to a single clinic in USA/UK and this data gives 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 opinion based on our data, and when cited as compared to other registries these are from Australia, UK and USA. Although we have a total of 449 registered in Port CFNZ, not all individuals had an input for all questions. While the total is 449 (192 children <16 year years, 257 adults > 16 years) at the top of each table or figure is the total number that had a response to the question. For example, on supplemental feeding a total response was obtained from 150 children and 203 adults on page 16 (the rest of the data for those individuals are missing).

The NZ registry data is becoming more robust and accurate; **we welcome its use in audit and research projects for researchers from reputable institutions.** Enquiries regarding the use of data can be made either to Jane Bollard, CE, CFNZ or to the Project Co-ordinator Jan Tate.

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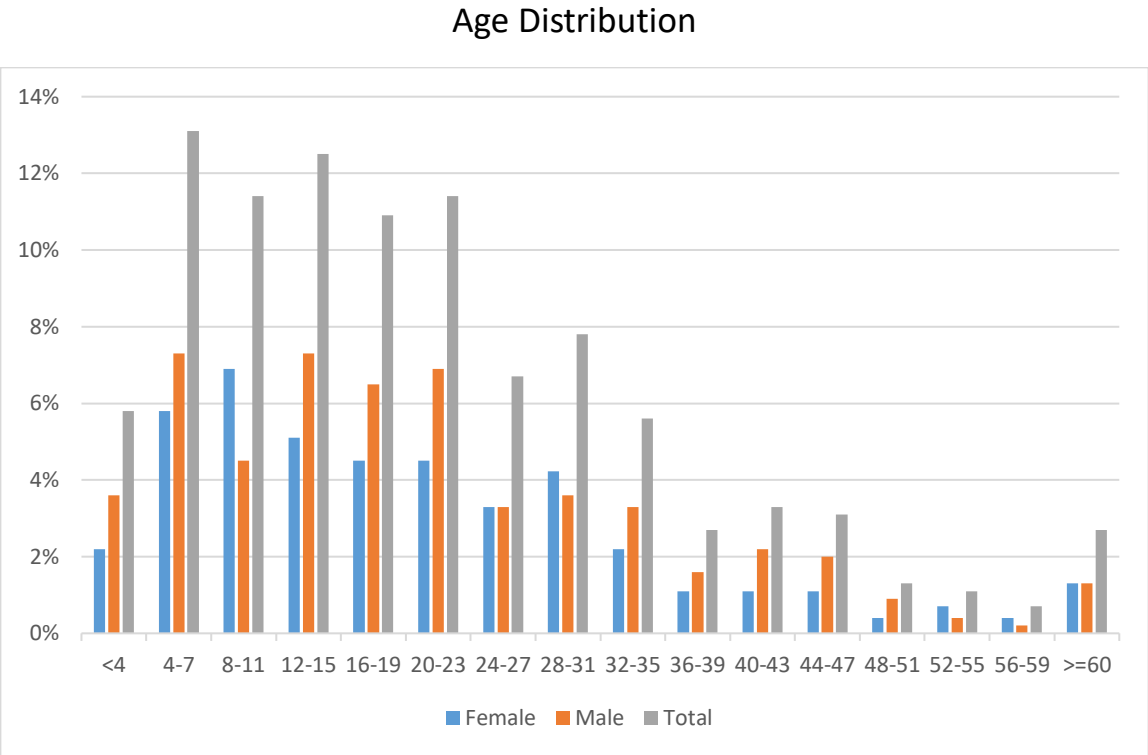
Key Indicators

	2015	2014	2013	2012	2011
CF patients registered	449	443	444	423	415
Diagnosis age <1 year	5	7	5	11	11
Diagnosis age >16 years	0	2	3	2	
Age in years; median	18.25	18.11	17.55	16.15	15.71
PWCF aged >16 years	257 57.20%	247 55.80%	239 53.80%	214 50.60%	206 49.60%
Males	247 55%	240 54.20%	240 54.10%	228 53.90%	226 54.60%
Genotyped	400 89.10%	429 96.80%	426 95.90%	407 96.20%	364 87.70%
Median FEV1 (% predicted)	85.60%	85.10%	84.30%	84.50%	80.50%
<16 years	98.90%	97.70%	96.60%	97.20%	91.60%
>16 years	77%	78.00%	70.70%	70.60%	70.70%

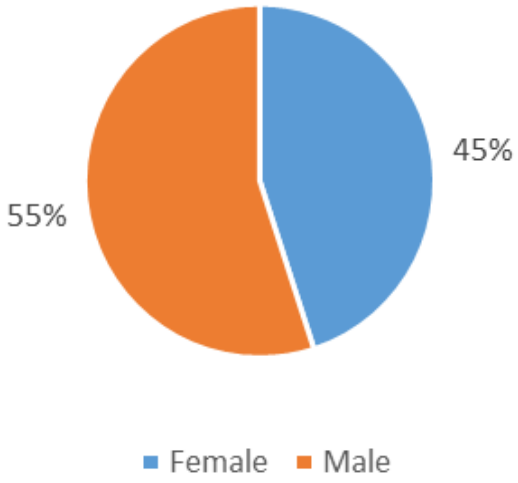
Demographics



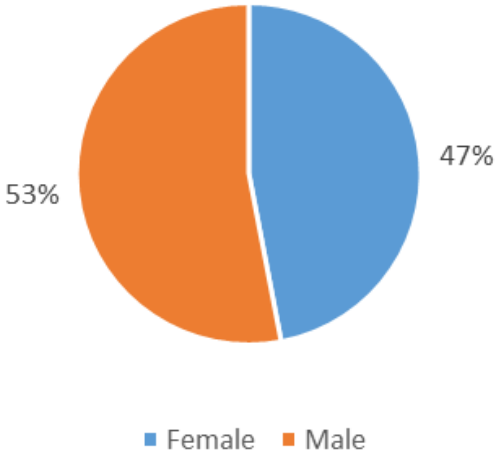
Age (years)	All		Male		Female	
	n	%	n	%	n	%
0-3	26	5.7	16	6.48	10	4.9
4-7	59	13.1	33	13.4	26	12.9
8-11	51	11.4	20	8.1	31	15.3
12-15	56	12.5	33	13.4	23	11.4
16-19	49	10.9	29	11.7	20	9.9
20-23	51	11.4	31	12.5	20	9.9
24-27	30	6.7	15	6.1	15	7.4
28-31	35	7.8	16	6.5	19	9.4
32-35	25	5.6	15	6.1	10	4.9
36-39	12	2.7	7	2.8	5	2.5
40-43	15	3.3	10	4	5	2.5
44-47	14	3.1	9	3.6	5	2.5
48-51	6	1.3	4	1.6	2	1
52-55	5	1.1	2	0.8	3	1.5
56-59	3	0.7	1	0.4	2	1
>=60	12	2.7	6	2.4	6	3
Total	449		247		202	
Median	18.25					
Range	0.28 – 73.0 years					

The median age of all persons with cystic fibrosis in New Zealand continues to increase. International registries differ in presenting data for children and adults; defining 'children' as being either up to 16 years or up to 18 years of age. If <16 years, we have 192 (42.8%) children and 257 (57.2%) adults. If <18 years, we have 224 (49.8%) children and 225 (50.1%) adults.

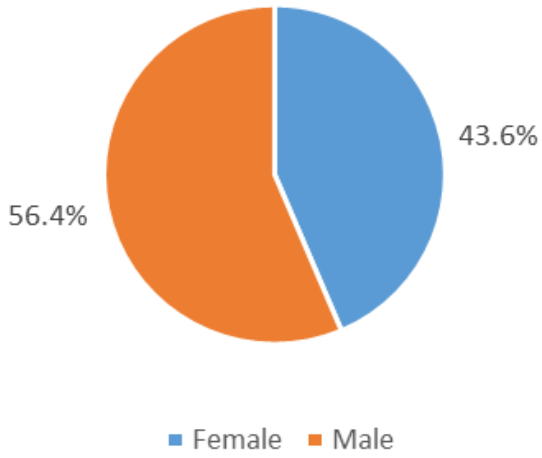
Gender Distribution



Gender Distribution <16 years



Gender Distribution >16 years



	All	
	n	%
Male	240	54.2
Female	203	45.8
Total	443	

	<16 years	
	n	%
Male	102	53
Female	90	47
Total	192	

	>16 years	
	n	%
Male	145	56.4
Female	112	43.6
Total	257	

Genotype

F508del Mutations	n	%
Homozygous F508del	221	55.3
Heterozygous F508del	146	36.5
No F580del or both unidentified	33	8.3
Total	400	

Genetic Mutations	c.DNA Name	n	%
F508del	c.1521_1523delCTT	588	77.9
Other		59	7.8
Unidentified		21	2.8
G542X	c.1624G>T	19	2.5
G551D	c.1652G>A	16	2.1
R117H	c.350G>A	15	2
G85E	c.254G>A	5	0.7
^1507	c.1519_1521delATC	4	0.5
3849+10kbC->T	c.3717+12191C>T	3	0.4
N1303K	c.3909c>G	3	0.4
1717-1G->A	c.1585-1G>A	2	0.3
1898+1G->A	c.1766+1G>A	2	0.3
A455E	c.1364C>A	2	0.3
1078delT	c.948delT	2	0.3
D1152H	c.3454G>C	2	0.3
R334W	c.1000C>T	2	0.3
Q493X	c.1477C>T	1	0.1
2789+5G->A	c.2657+5G>A	1	0.1
3120+1G->A	c.2988+1G>A	1	0.1
3659delC	c.3528delC	1	0.1
621+1G->T	c.489+1G>T	1	0.1
712-1G->T	c.580-1G>T	1	0.1
R347H	c.1040G>A	1	0.1
R347P	c.1040G>C	1	0.1
R560T	c.1679G>C	1	0.1
W1282X	c.3846G>A	1	0.1
R553X	c.1657C>T	0	0

The knowledge of each individual's genetic determination is increasingly important with the development of genetic mutation specific drugs; the so-called "correctors" and "potentiators". We recommend any individual who does not know their genetic mutation, especially if only tested in early childhood, to have a further genetic evaluation.

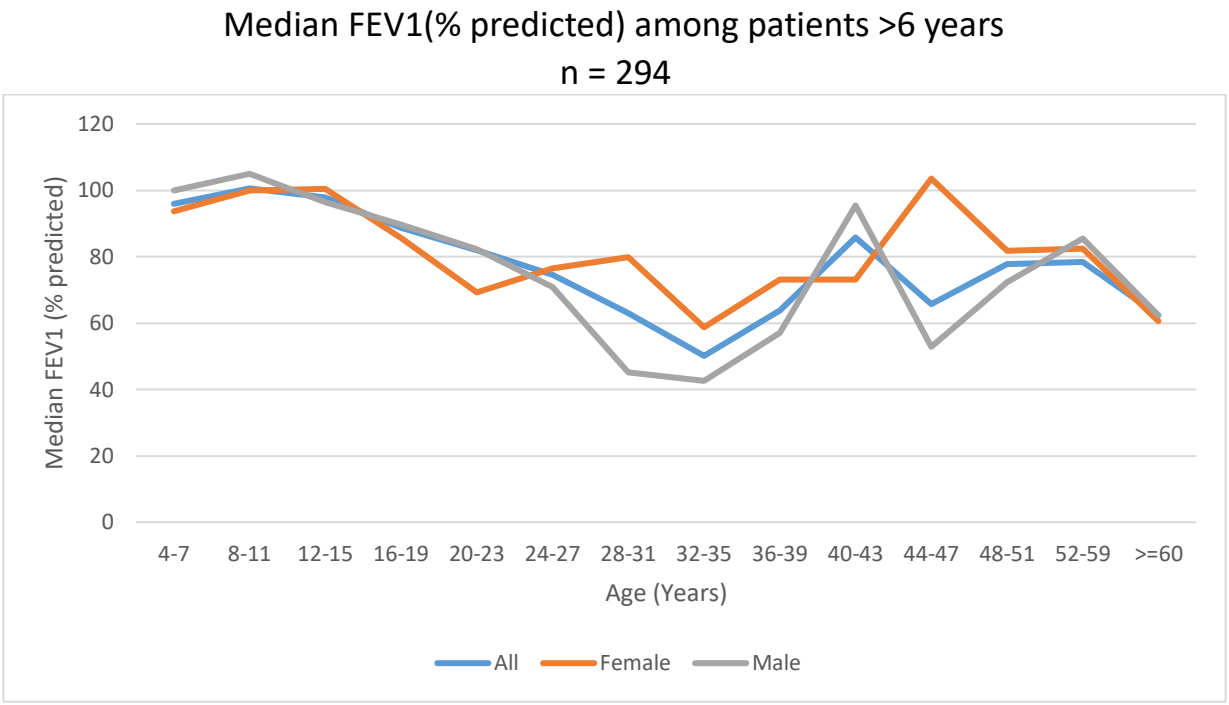
Individuals with no F508del genetic mutations (n=44)

	G542X	G551D	G85E	R1162X (c.3484C>T)	R117H	1717-1G->A	Other	Not Identified
G542X	1						2	1
G551D	1	1			2		2	2
N1303K		1					1	
Q493X		2		1				
R553X		1						
3849+10kbC->T						1		
W1282X								1
Other			1	1			15	1
Not Identified					2			9

This table describes those individuals who have no F508del genetic mutations. It is important to know all combinations to inform our new-born screening programme and later diagnostic screening programmes. It allows us to determine whether any people in NZ will be missed on current diagnostic panels.

	n	%
G542X	4	8.2
G551D	8	16.3
N1303K	2	4.1
Q493X	3	6.1
R553X	1	2
3849+10kbC->T	1	2
W1282X	1	2
Other	18	36.7
Not Identified	11	22.4

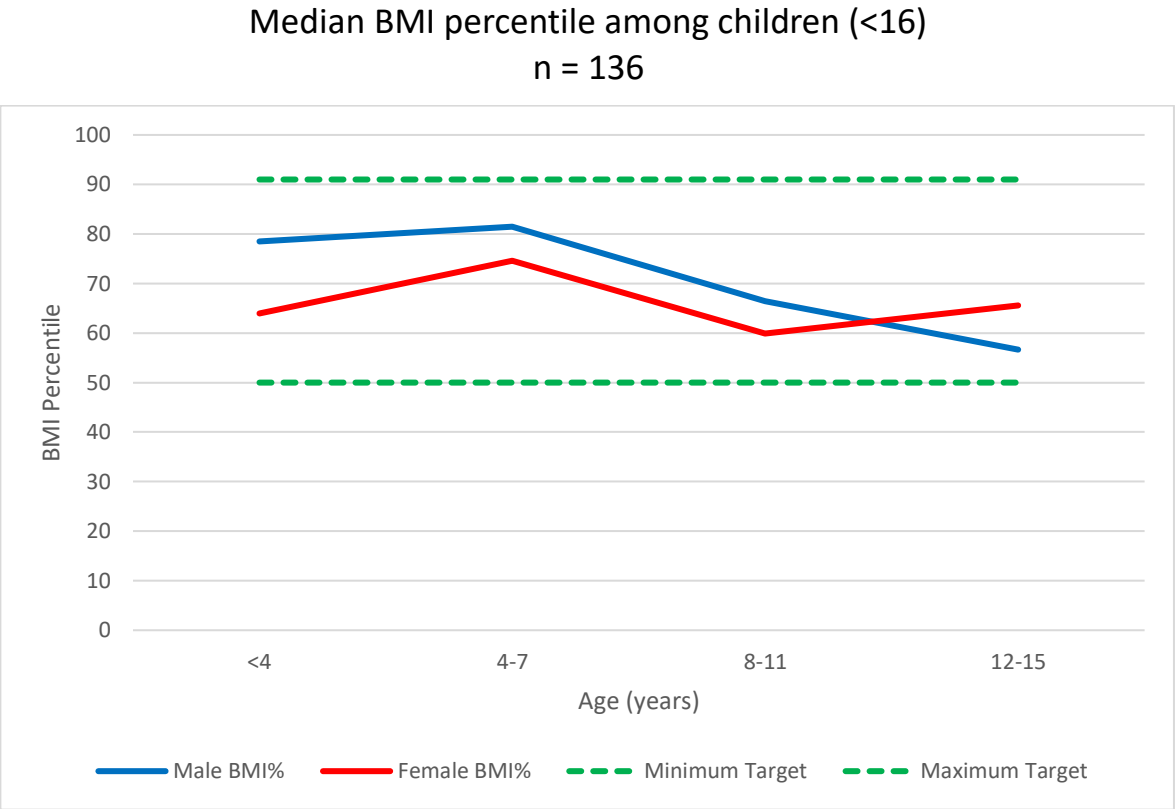
Respiratory



Age (Years)	All		Female		Male	
	n	median	n	median	n	median
4-7	17	95.9	6	93.7	11	100
8-11	38	100.6	23	99.9	15	105
12-15	37	97.9	14	100.5	23	96.5
16-19	40	88.8	15	85.7	25	89.7
20-23	41	82	15	69.3	26	82.3
24-27	23	74.6	11	76.5	12	70.8
28-31	25	63	13	79.9	12	45.2
32-35	19	50.1	8	58.7	11	42.6
36-39	10	63.7	4	73.1	6	57.1
40-43	11	85.8	3	73.1	8	95.5
44-47	11	65.7	3	103.5	8	52.9
48-51	5	77.7	2	81.8	3	72.3
52-55	3	71.3	3	71.3	0	.
56-59	3	85.4	2	93.6	1	85.4
>=60	11	62.4	6	60.6	5	62.4
Total	294		128		166	

This data necessarily excludes very young children in whom we cannot measure lung function or those who find doing accurate lung function testing difficult. The trend of mean and median lung function over the time we have been collecting registry data (2011-2015) has seen a gradual increase. The trend regarding lung function with age is of gradual deterioration from early childhood to early adulthood. The late maintenance of lung function reflects those living longer with more mild disease and late diagnoses of people with milder CF phenotype.

Nutrition



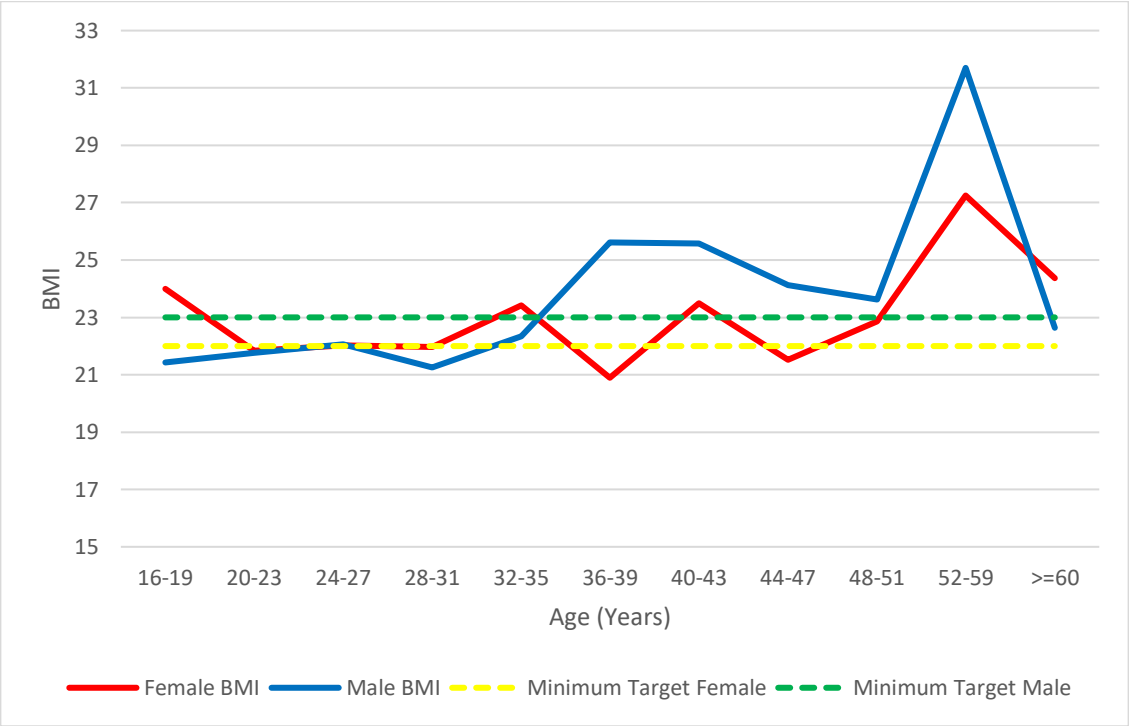
The optimal BMI for children 2-16 years is 50-91 percentile using WHO-NZ growth chart.
The dotted lines show the target range.

Age (years)	BMI Percentile	
	n	median
<4	10	70.7
04-Jul	48	71.4
08-Nov	40	60.2
Dec-15	38	53.7
Total	136	

The pattern suggests that nutrition becomes more at risk during adolescent years with greater need given to the pubertal growth spurts and it is often a time of increased sport activities – both with an increasing calorie requirement. CF Related Diabetes also begins to be diagnosed more frequently in teenage years. Any issues regarding nutrition will be covered by the excellent and recently published Australasian Cystic Fibrosis Nutritional Guidelines.

http://www.knowledge.scot.nhs.uk/media/CLT/ResourceUploads/1008938/Austrailian%20CF_Nutrition_Guidelines.pdf

Median BMI among Adults (>16)
n = 201



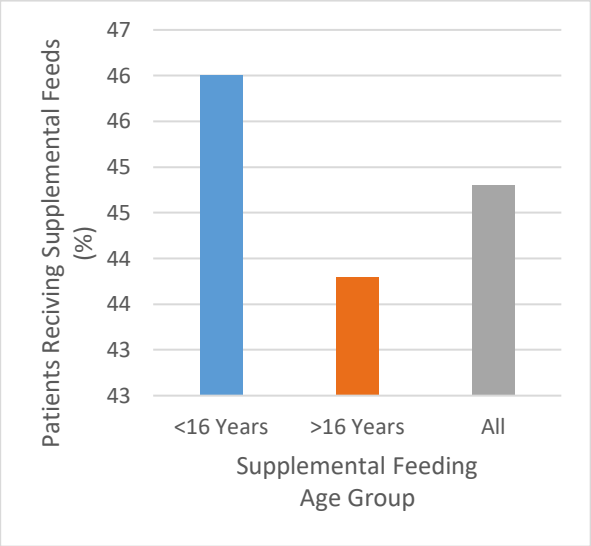
The optimal BMI for woman is 22-27 and the dotted yellow line shows the minimum BMI for women in the target range. The optimal BMI for men is 23-27. The dotted blue line shows the minimum BMI for men in the target range.

Females 16>		
Age (Years)	BMI Percentile	
	n	median
16-19	15	24
20-23	15	21.8
24-27	10	22
28-31	13	22
32-35	8	23.4
36-39	4	20.9
40-43	3	23.5
44-47	3	21.5
48-51	2	22.9
52-55	3	28.3
56-59	2	26.2
>=60	6	24.4

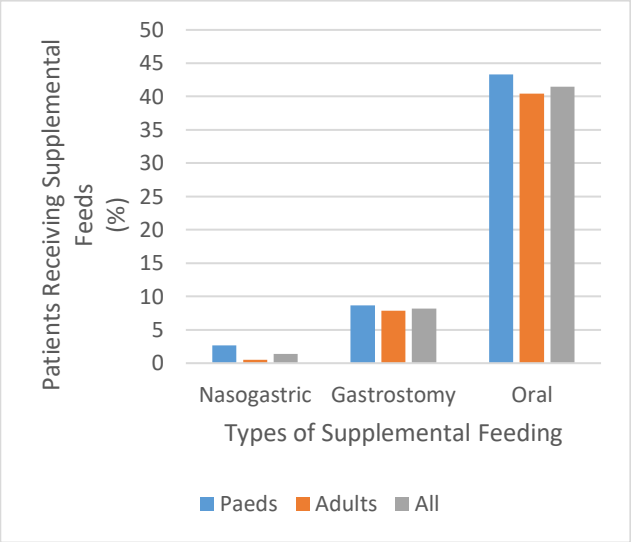
Males 16>		
Age (Years)	BMI	
	n	median
16-19	25	21.4
20-23	26	21.8
24-27	12	22.1
28-31	12	21.3
32-35	11	22.3
36-39	6	25.6
40-43	8	25.6
44-47	8	24.1
48-51	3	23.6
52-55	0	.
56-59	1	31.7
>=60	5	22.6

Patients receiving supplemental feeding

<16 yrs n=150 >16 yrs n=203 ALL n=353



Types of Supplemental Feeding (Shown as % of those receiving supplemental feeds)

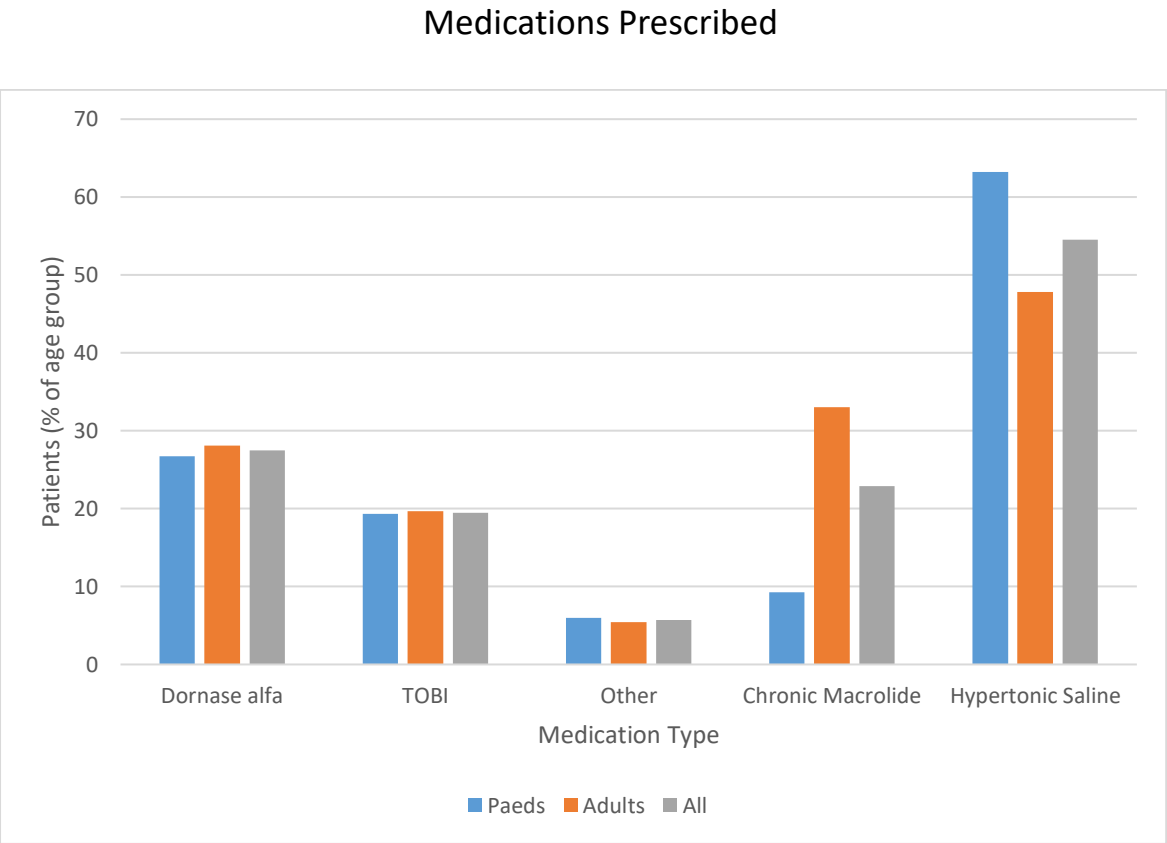


In NZ, we are high users of supplemental feeds. It may be associated with improved nutritional profile in our persons with CF when compared to other international registry data.

	<16 years, n = 150	
	Yes	% <16 years supplemented
Supplemental Feeding	69/150	46
Nasogastric	4	2.7
Gastrostomy	13	8.7
Oral	65	43.3

	>16 years, n = 203	
	Yes	% >16 years supplemented
Supplemental Feeding	89/203	43.8
Nasogastric	1	0.5
Gastrostomy	16	7.9
Oral	82	40.4

Medications

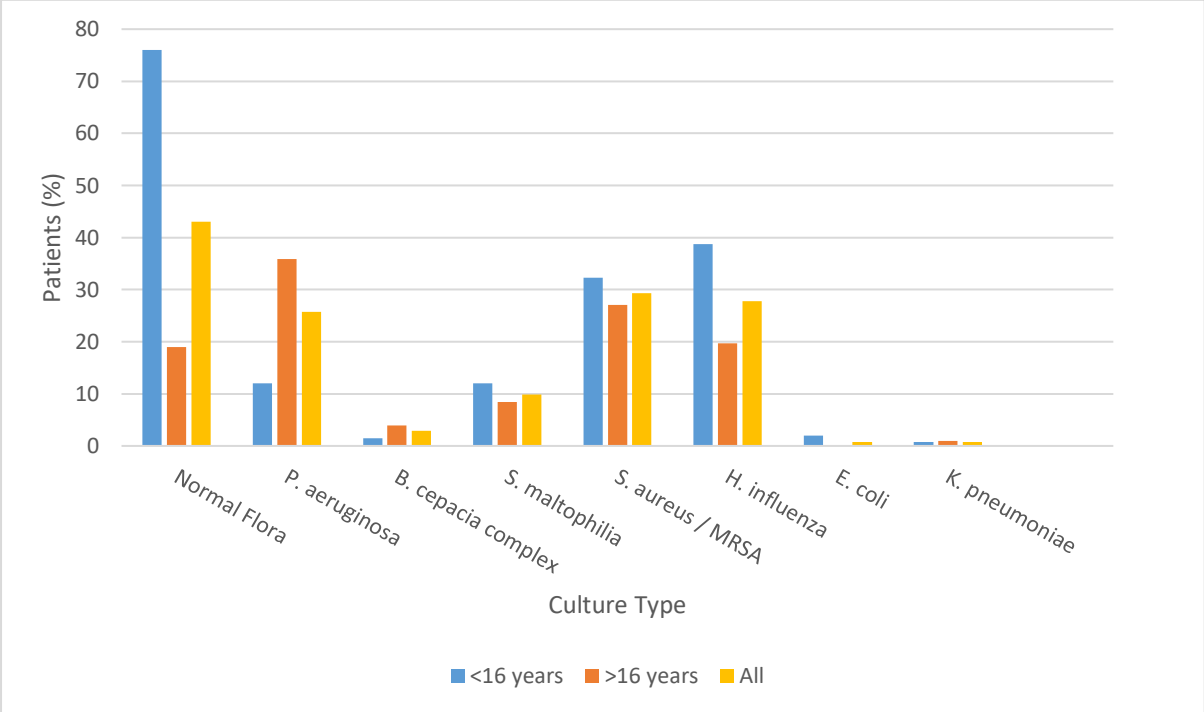


Other - This reflects nebulised Colistin, Tobramycin (Intravenous Solution), Gentamicin and Aztreonam.

Medication	<16 years	>16 years	All
Dornase alfa	26.70%	28.10%	27.50%
TOBI	19.30%	19.70%	19.50%
Other	6.00%	5.40%	5.60%
Chronic Macrolide	9.30%	33.00%	22.90%
Hypertonic Saline	63.20%	47.80%	54.50%

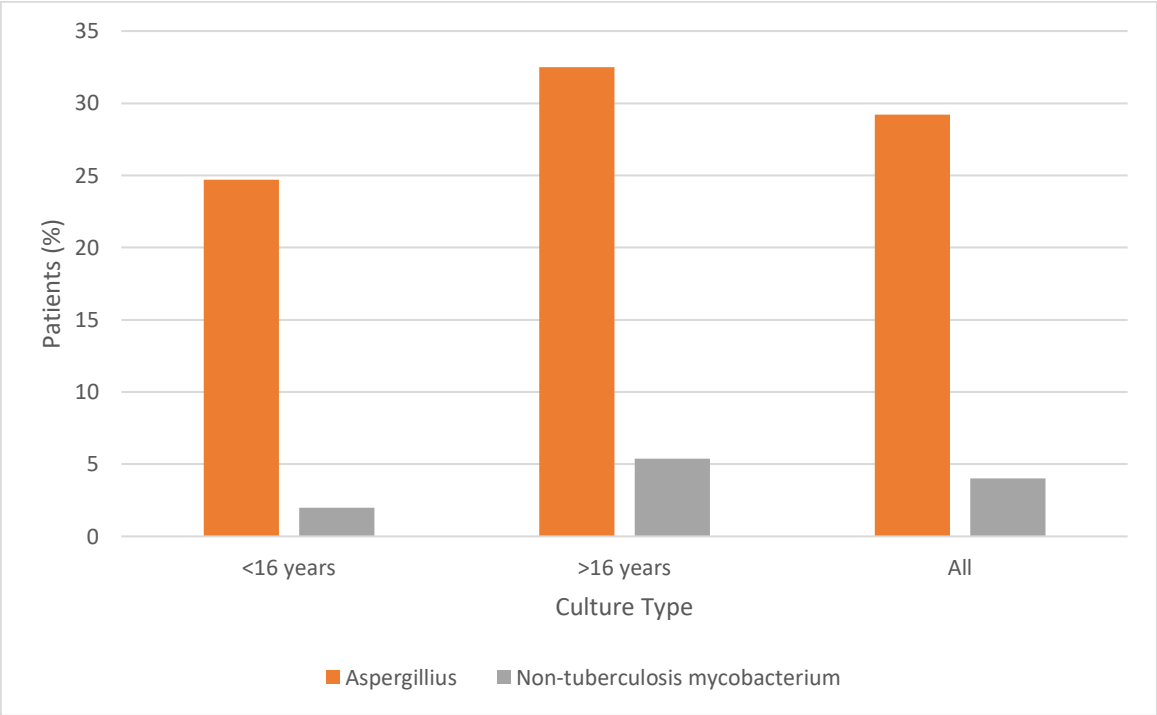
Microbiology

Culture Prevalence
<16 years n = 150, >16 years n = 203, All n = 353



	<16 years, n = 150		>16 years, n = 203		All, n = 353	
	n	%	n	%	n	%
Normal Flora	12	8	2	1	14	4
Pseudomonas aeruginosa	27	18	92	45.3	119	33.7
Mucoid	7	4.7	65	32	72	20.4
Non mucoid	20	13.3	62	30.5	82	23.2
Staphylococcus aureus	90	60	101	49.8	191	54.1
MSSA	83	55.3	92	45.4	175	49.6
MRSA	7	4.7	9	4.4	16	4.5
Stenotrophomonas Maltophilia	18	12	17	8.4	35	9.9
Burkholderia Cepacia Complex	4	2.7	18	8.9	22	6.2
Cenocepacia	0	0	2	1	2	0.6
Multivorans	4	2.7	9	4.4	13	3.7
Other	1	0.7	3	1.5	4	1.1

Nonbacterial/Fungal Culture Prevalence
<16 years n = 150, >16 years n = 203, All n = 353



Intravenous Antibiotic Treatment Episodes

Home IV Days

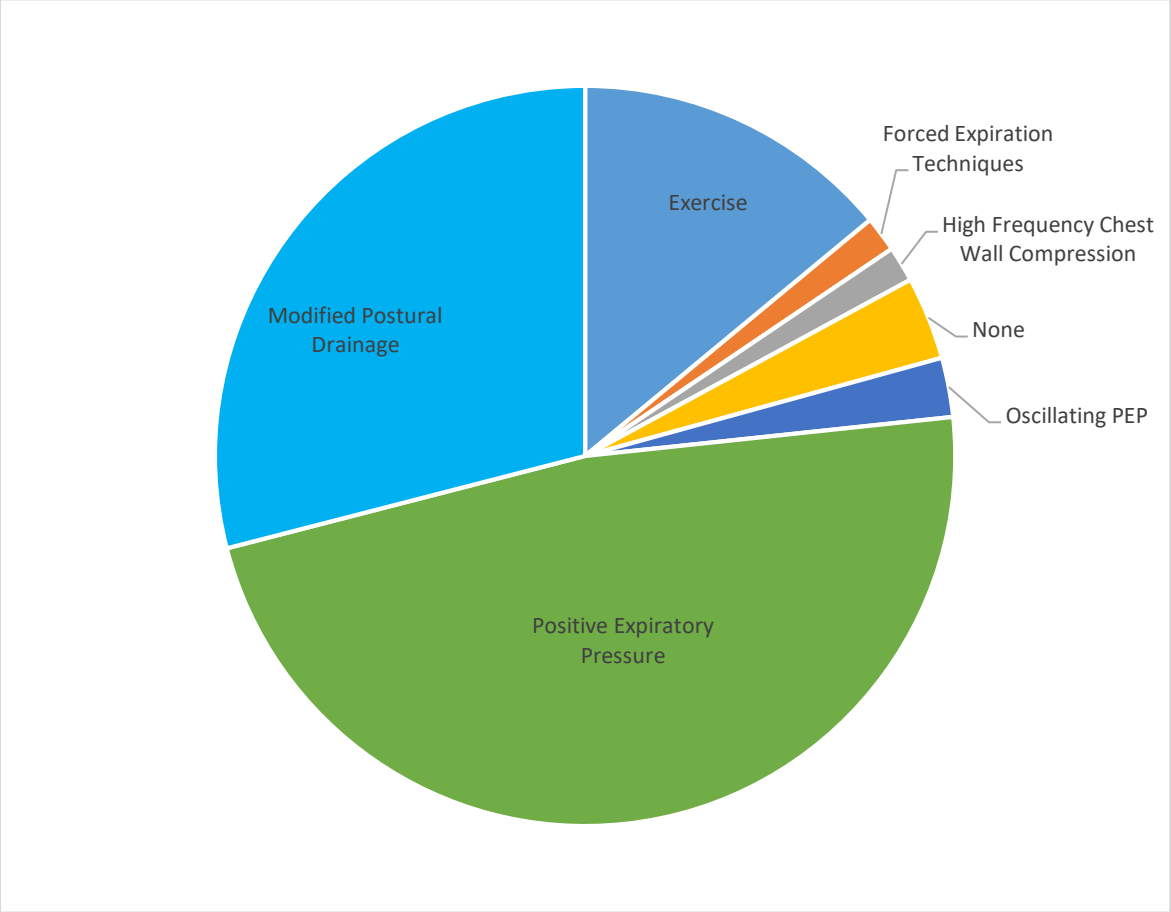
Age	n	n = people who had IV days	%	Mean days (for people who had IV days)	Mean (Overall)
<4	20	3	15%	12	1.8
4-7	46	13	28%	10.2	2.9
8-11	40	7	18%	71.6	3.1
12-15	37	10	27%	19.8	5.4
16-19	37	11	30%	24.3	7.2
20-23	41	11	27%	14.8	4
24-27	17	2	12%	17.5	2.1
28-31	24	10	42%	32.9	13.7
32-35	19	6	32%	15.2	4.8
36-39	10	4	40%	21.5	8.6
40-43	11	2	18%	14.5	2.6
44-47	11	1	9%	9	0.8
48-51	5	0	0%	0	0
52-55	3	0	0%	0	0
56-59	2	0	0%	0	0
>=60	10	1	10%	7	0.7

Hospital IV Days

Age	n	n = people who had IV days	%	Mean (for people who had IVA days)	Mean (Overall)
<4	20	8	40%	10.9	4.4
04-Jul	46	20	43%	14.9	6.5
08-Nov	40	16	40%	14.6	5.9
Dec-15	37	19	51%	18.7	9.6
16-19	37	20	54%	24.6	13.3
20-23	41	21	51%	25.6	13.1
24-27	17	6	35%	25.2	8.9
28-31	24	10	42%	27.1	11.3
32-35	19	7	37%	30.1	11.1
36-39	10	5	50%	6.8	3.4
40-43	11	2	18%	6.5	1.2
44-47	11	3	27%	38	10.4
48-51	5	1	20%	10	2
52-55	3	1	33%	6	2
56-59	2	0	0%	0	0

Airway Clearance Techniques

Primary Airway Clearance Technique <16 years
n = 150
(Some patients may have used more than one technique as their primary form of physiotherapy in different clinical encounters)

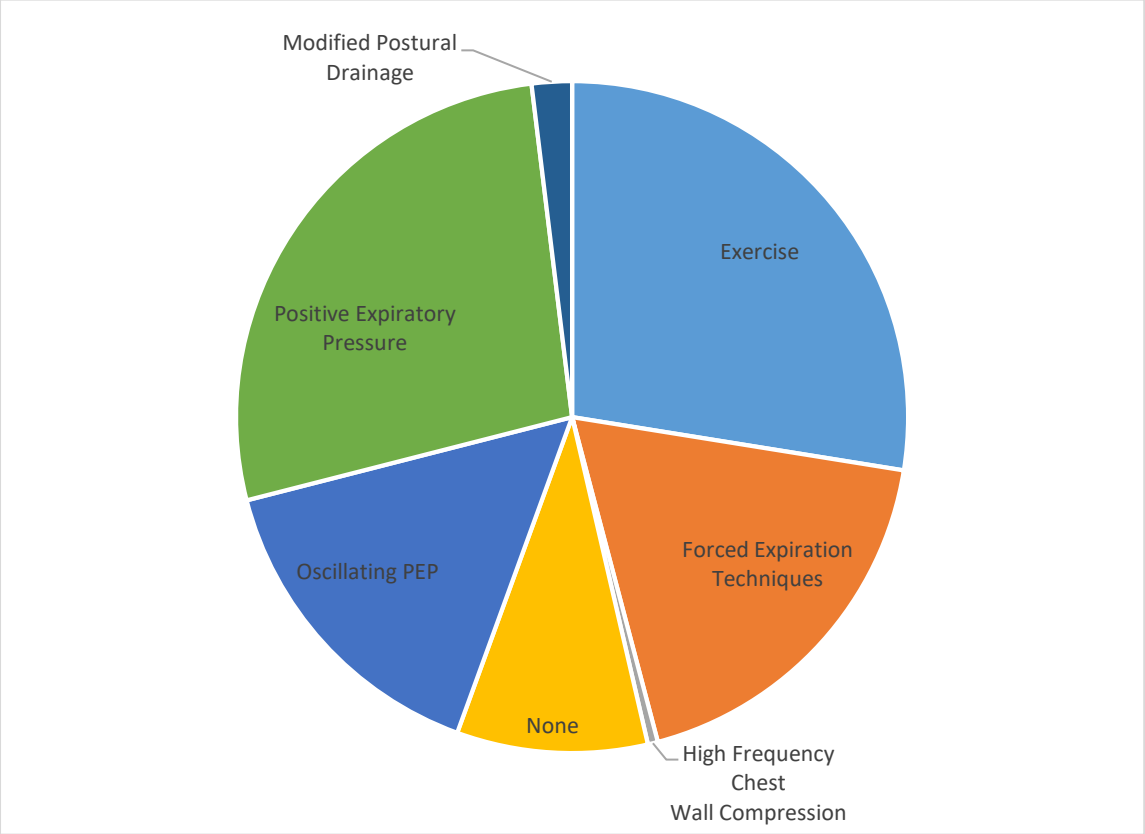


* Number of individuals employing each technique at least once a year. Data collected from 150 patients.

Technique	< 16 years	
	n	%
Positive Expiratory Pressure	92	61
Modified Postural Drainage	56	37.3
Exercise	27	18
Oscillating Pep (e.g.Flutter, Acapella, IPV)	5	3.3
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	3	2
High Frequency Chest Wall Compression (e.g. VEST)	3	2
None	7	4.7

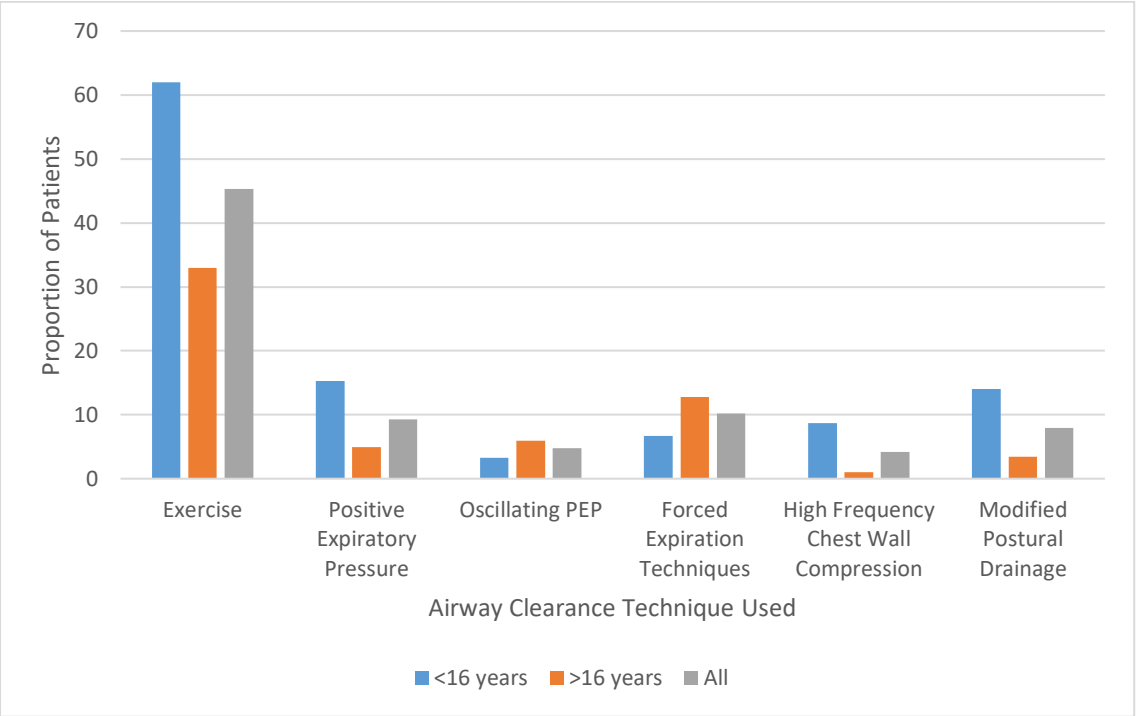
Primary Airway Clearance Technique >16 Years
n = 203

(Some patients may have used more than one technique as their primary form of physiotherapy in different clinical encounters)



* Number of individuals employing each technique at least once a year. Data collected from 203 patients.

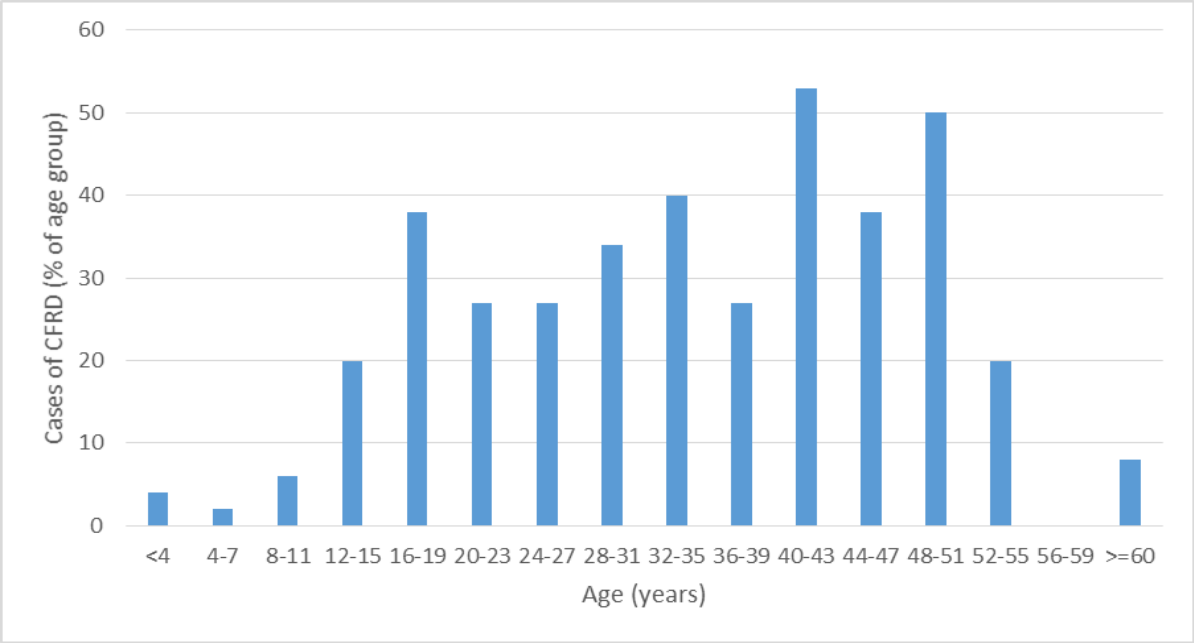
Technique	> 16 years	
	n	%
Positive Expiratory Pressure	56	27.6
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	38	18.7
Oscillating PEP (e.g. Flutter, Acapella, IPV)	32	15.8
Modified Postural Drainage	4	2
High Frequency Chest Wall Compression (e.g. VEST)	1	0.5
None	19	9.4



Data Collected in 150 <16 years, 203 >16 years. Some patients may have used more than one technique

Technique	< 16 years		> 16 years		All	
	n	%	n	%	n	%
Exercise	93	62	67	33	160	45.3
Positive Expiratory Pressure	23	15.3	10	4.9	33	9.3
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	10	6.7	26	12.8	36	10.2
Oscillating PEP (e.g. Flutter, Acapella, IPV)	5	3.3	12	5.9	17	4.8
Modified Postural Drainage	21	14	7	3.4	28	7.9
High Frequency Chest Wall Compression (e.g. VEST)	13	8.7	2	1	15	4.2

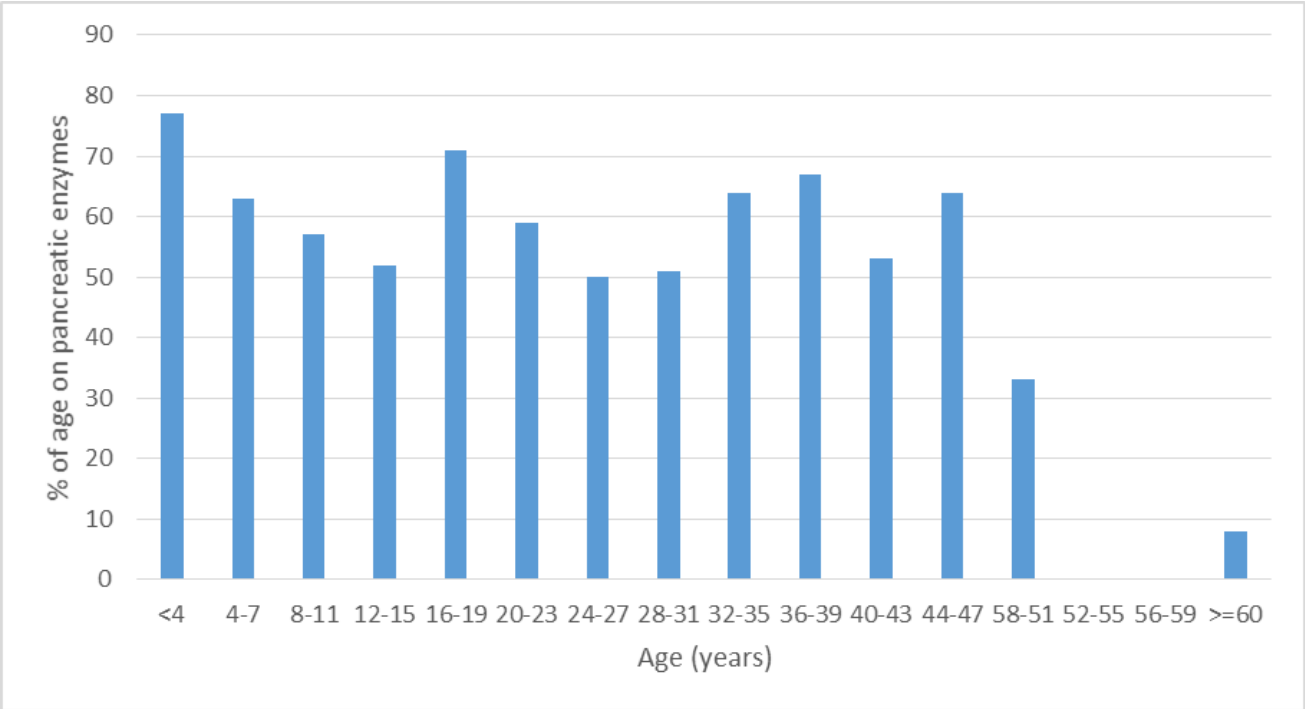
CF Related Diabetes



Age (years)	Age n	CFRD n	% of Age Group	% of CF Population
<16	188	16	9	3.65
>16	250	82	33	18.72
Total	438	98	22	22.37

Age (years)	Age n	CFRD n	% of Age Group	% of CF Population
<4	23	1	4	0.2
4-7	58	1	2	0.2
8-11	51	3	6	0.7
12-15	56	11	20	2.5
16-19	48	18	38	4.1
20-23	51	14	27	3.2
24-27	30	8	27	1.8
28-31	32	11	34	2.5
32-35	25	10	40	2.3
36-39	11	3	27	0.7
40-43	15	8	53	1.8
44-47	13	5	38	1.1
48-51	6	3	50	0.7
52-55	5	1	20	0.2
56-59	2	0	0	0
>=60	12	1	8	0.2

Pancreatic Enzymes



Age (years)	n	On PE	% of Age Group	% of CF Population
<16	192	115	60	25.6
>16	257	142	55	31.6
Total	449	257	57.3	57.3

Age (years)	n	On PE	% of Age Group	% of CF Population
<4	26	20	77	4.5
4-7	59	37	63	8.2
8-11	51	29	57	6.5
12-15	56	29	52	6.5
16-19	49	35	71	7.8
20-23	51	30	59	6.7
24-27	30	15	50	3.3
28-31	35	18	51	4
32-35	25	16	64	3.6
36-39	12	8	67	1.8
40-43	15	8	53	1.8
44-47	14	9	64	2
58-51	6	2	33	0.4
52-55	5	0	0	0
56-59	3	0	0	0
>=60	12	1	8	0.2
Total	449	257	57.3	57.3

Glossary of Terms

FEV1	Measurement of lung capacity as Forced Expired Volume in one second
BMI	Body Mass Index: measurement of weight relative to height
N (n)	Total number of people in a dataset
Median	Middle number in a numerically arranged range of numbers
Range	Upper and lower values in a dataset
Paediatric	0 – 16 years of age
Adult	>16 years of age

All enquiries regarding this Data Registry should be forwarded in the first instance, to:

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