PORT CFNZ

2014 National Data Registry

CYSTIC FIBROSIS NZ
Creating Better Tomorrows

PORT CFNZ NATIONAL DATA REGISTRY

The Port CFNZ National Data Registry is a research project of Cystic Fibrosis New Zealand.
For further information about the CFNZ visit www.cfnz.org.nz

The production of this Data Registry is funded through a conditional grant from

Shares in Life Foundation
Putting hope into action

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, 2014 Registry Report, Cystic Fibrosis New Zealand.

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Introduction & Acknowledgements

The Cystic Fibrosis New Zealand and the PORT CFNZ Steering Committee, are pleased to present The National Cystic Fibrosis Data Registry 2014 Report; data collected on children, young persons and adults with Cystic Fibrosis in New Zealand.

We would like to thank:

- The Nurses, Specialists and Administrators who have worked to enter data enabling a detailed analysis for NZ – presented in this report.
- Shares in Life Foundation for providing pivotal funding for database and data entry.
- Canterbury District Health Board for their ongoing commitment to maintain the registry.
- Above all, the children and adults with CF and their families for participating in this process.

This fourth 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.

Development of the database will lead to slightly different questions used next year, written exclusively with the New Zealand clinical environment in mind and provide more accurate information for our environment.

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

Associate Professor Cass Byrnes
Chair Port CFNZ Steering Committee

Dr Richard Laing
Port CFNZ Principal Investigator

Port CFNZ Steering Committee

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CF Clinics in New Zealand

Northland (Paediatrics)
Whangarei Hospital, Whangarei

Auckland (Paediatrics and Adults)
Starship Children's 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

The Data registry gives national statistics. New Zealand has a total CF population comparative to a single clinic in USA/UK. Statistically accurate and relevant data for smaller clinics is difficult given the smaller numbers.

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 given the smaller numbers. Some decisions were made by the steering committee to exclude those outlier ages and statistics in order to give a more accurate picture of the overall patient outcomes for the country.

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 in the main.

The NZ registry data is becoming more robust and accurate, **we welcome its use in audit and research projects.** A proposal for a project involving this national data base can be made in writing using the form found on the CFNZ website the PORTCFNZ steering committee.

Link: http://www.cfnz.org.nz/our-services/library/downloads/#other

Port CF Steering Committee
C/- CFNZ
P O Box 8241
Riccarton
Christchurch

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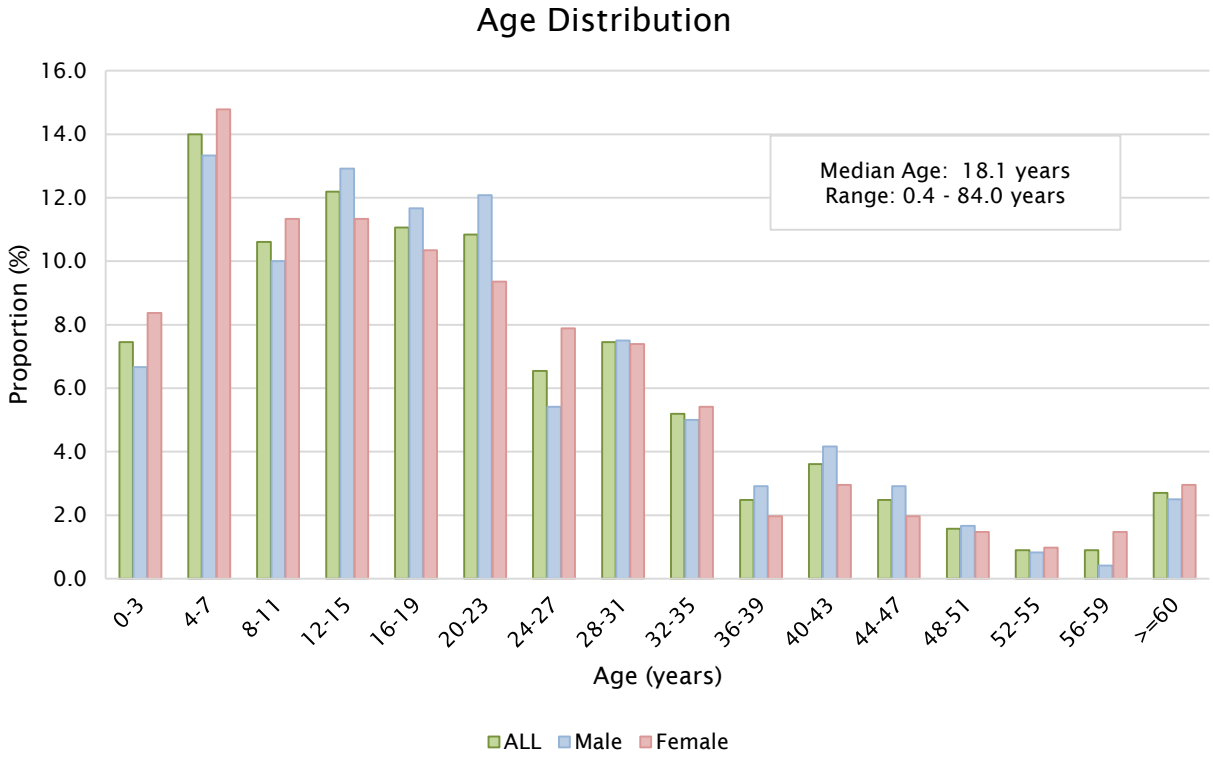
Key Indicators

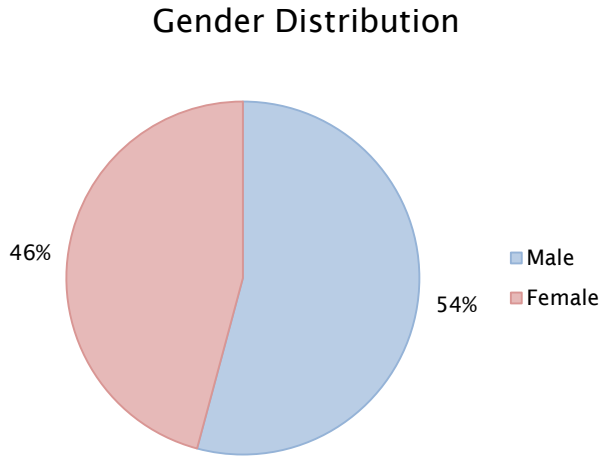
	2014	2013	2012	2011
CF patients registered	443	444	423	415
Diagnosis age <1 year	7	5	11	11
Diagnosis age >16 years	2	3	2	
Age in years; median	18.11	17.55	16.15	15.71
PWCF aged >16 years	247 *55.8%*	239 *53.8%*	214 *50.6%*	206 *49.6%*
Males	240 *54.2%*	240 *54.1%*	228 *53.9%*	226 *54.6%*
Genotyped	429 *96.8%*	426 *95.9%*	407 *96.2%*	364 *87.7%*
Median FEV1 (% predicted)	85.1%	*84.3%*	*84.5%*	*80.5%*
<16 years	97.7%	*96.6%*	*97.2%*	*91.6%*
>16 years	78.0%	*70.7%*	*70.6%*	*70.7%*

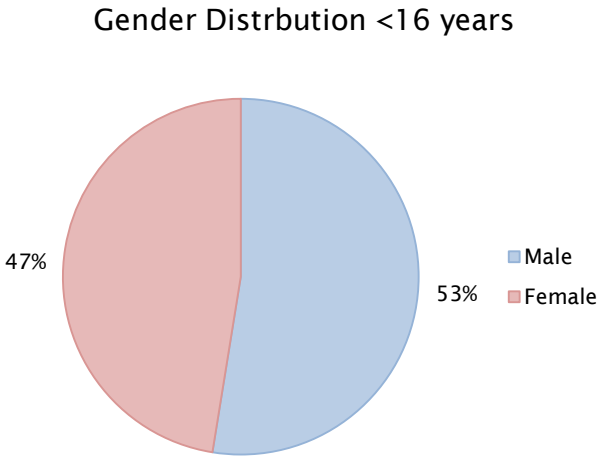
Demographics

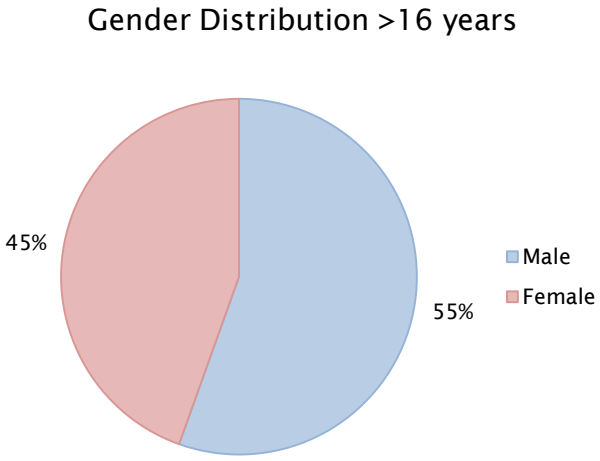


Age (yrs)		All			Male			Female	
		n	%		n	%		n	%
0-3		33	*7.4*		16	*6.7*		17	*8.4*
4-7		62	*14.0*		32	*13.3*		30	*14.8*
8-11		47	*10.6*		24	*10.0*		23	*11.3*
12-15		54	*12.2*		31	*12.9*		23	*11.3*
16-19		49	*11.1*		28	*11.7*		21	*10.3*
20-23		48	*10.8*		29	*12.1*		19	*9.4*
24-27		29	*6.5*		13	*5.4*		16	*7.9*
28-31		33	*7.4*		18	*7.5*		15	*7.4*
32-35		23	*5.2*		12	*5.0*		11	*5.4*
36-39		11	*2.5*		7	*2.9*		4	*2.0*
40-43		16	*3.6*		10	*4.2*		6	*3.0*
44-47		11	*2.5*		7	*2.9*		4	*2.0*
48-51		7	*1.6*		4	*1.7*		3	*1.5*
52-55		4	*0.9*		2	*0.8*		2	*1.0*
56-59		4	*0.9*		1	*0.4*		3	*1.5*
>=60		12	*2.7*		6	*2.5*		6	*3.0*
Total		443			240			203	
Median		18.1 years							
Range		0.4 - 84.0 years							

Over our first years of collecting registry data the median age of PWCF has increased with a corresponding increase in the proportion of adults.







	All		<16 years		>16 years	
	n	%	n	%	n	%
Male	240	*54.2*	103	*52.6*	137	*55.5*
Female	203	*45.8*	93	*47.4*	110	*44.5*
Total	443		196		247	

The gender distribution can become less even in adult years – in part because young women can have accelerated disease.

Genotype

429 (96.8%) of 443 patients have been genotyped with a recorded value.

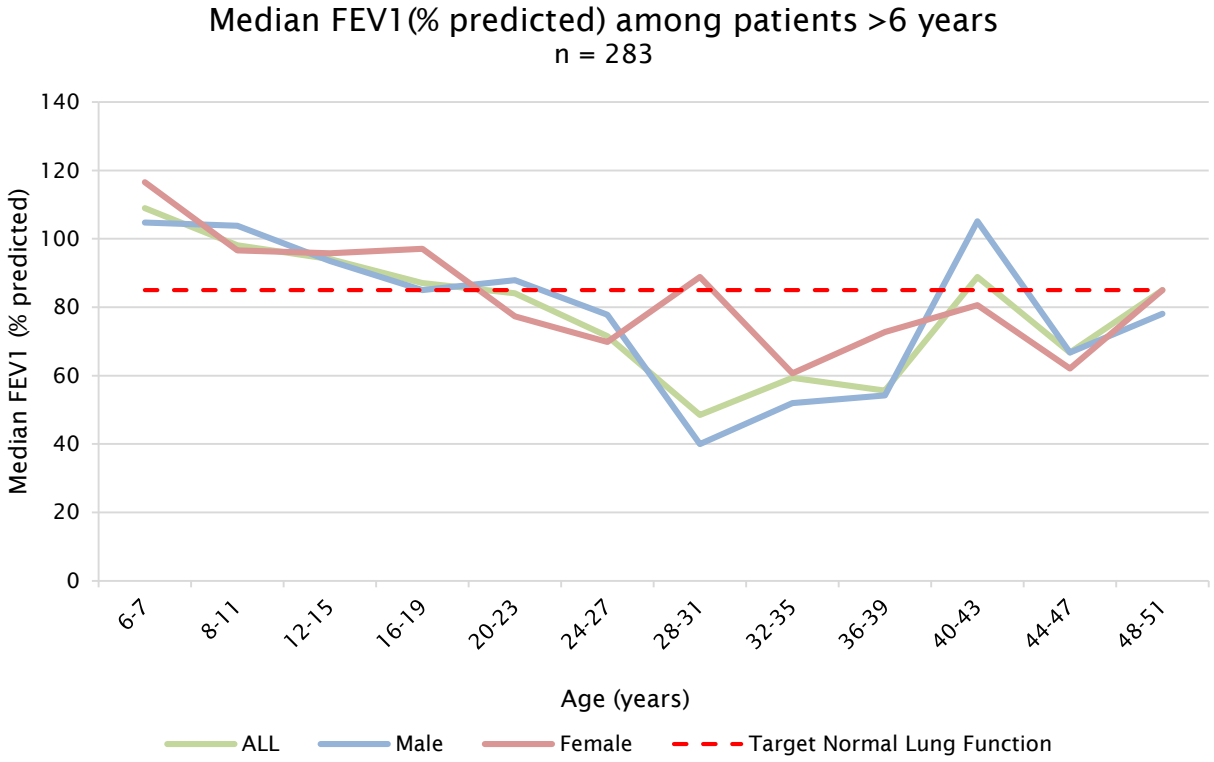
F508del Mutations	n	%
Homozygous F508del	221	51.5
Heterozygous F508del	162	37.8
No F508del or both unidentified	46	10.7
Total	429	

Mutations Identified	c.DNA Name	n	%
F508del	c.1521_1523delCTT	604	70.4
G551D	c.1652G>A	30	3.5
G542X	c.1624G>T	27	3.1
R117H	c.350G>A	21	2.4
G85E	c.254G>A	6	0.7
N1303K	c.3909c>G	5	0.6
3272-26A>G	c.3140-26A>G	5	0.6
^I507	c.1519_1521delATC	4	0.5
1717-1G->A	c.1585-1G>A	4	0.5
3849+10kbC->T	c.3717+12191C>T	4	0.5
Q493X	c.1477C>T	4	0.5
1898+1G->A	c.1766+1G>A	3	0.3
2789+2insA	c.2657+2_2657+3insA	2	0.2
A455E	c.1364C>A	3	0.3
c.3718-2477C>T	c.3718-2477C>T	3	0.3
Other		88	10.3
Unidentified		45	5.2
		858	

Increasingly the genetic mutations will be presented named for their DNA abnormalities (column 2) standardised world-wide. We still know them best by what has come to be called their legacy names (column 1).

It is encouraging to see that the majority have had their genotype determined. This will become increasingly important as new genotype specific drugs potentially become available. As previously mentioned, here in NZ while F508 remains the dominant gene, this is seen less here than in registries from other English speaking countries where it is 85% to 92%, suggesting that our ethnic diversity may contribute less common genes.

Respiratory

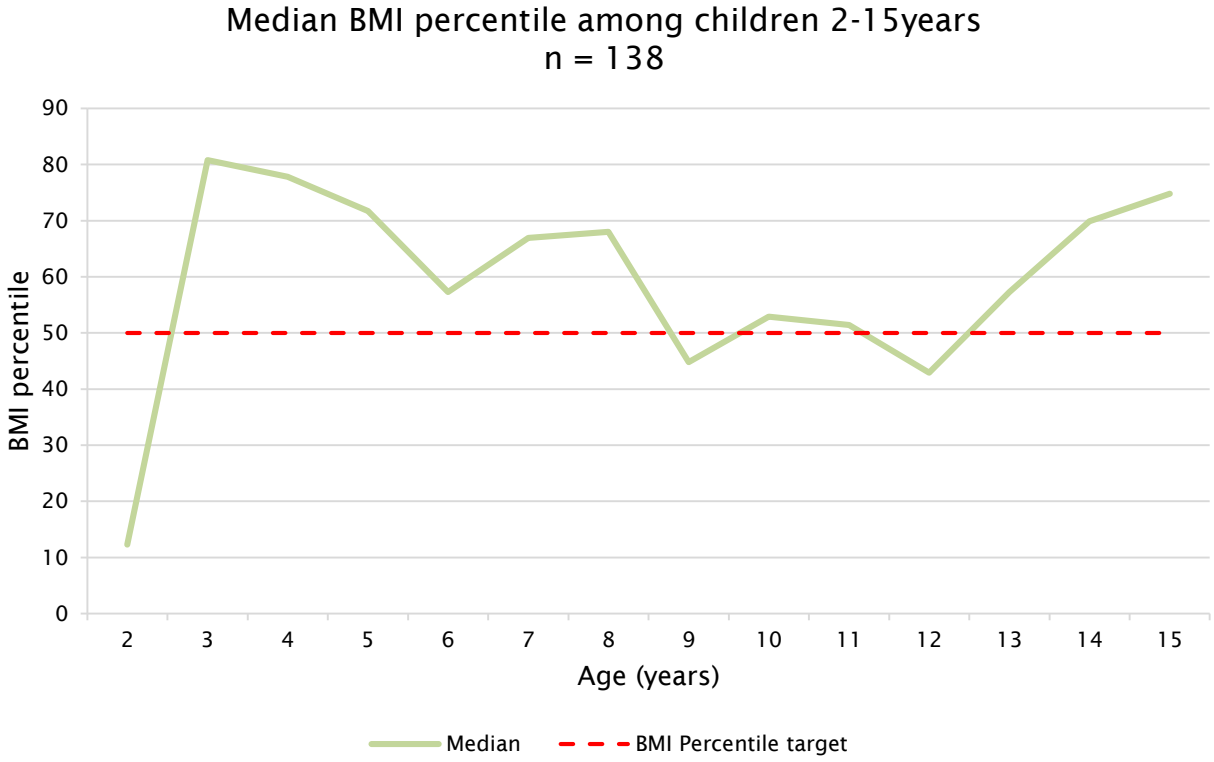


Age (yrs)	All		Male		Female	
	n	median	n	median	n	median
6-7	18	109.0	9	104.8	9	116.6
8-11	37	98.1	18	103.9	19	96.6
12-15	37	94.1	22	93.5	15	95.8
16-19	38	87.0	23	85.0	15	97.1
20-23	33	84.1	21	87.9	12	77.3
24-27	25	71.5	12	77.8	13	69.8
28-31	28	48.5	17	40.0	11	88.8
32-35	19	59.4	11	52.0	8	60.7
36-39	9	55.6	6	54.2	3	72.8
40-43	14	88.9	9	105.1	5	80.6
44-47	7	66.8	5	66.8	2	62.1
48-51	5	85.0	2	78.1	3	85.0
52-55	2	69.2	1	85.6	1	52.8
56-59	4	91.4	1	87.6	3	95.2
>=60	7	61.7	4	64.2	3	61.5
Total	283		161		122	

The slope of lung function (FEV1) over time is very similar to the other registries with our target lung function being greater than 85% which is in keeping with the UK registry. Our median FEV1 for those <16 years and >16 years is comparative to other registries.

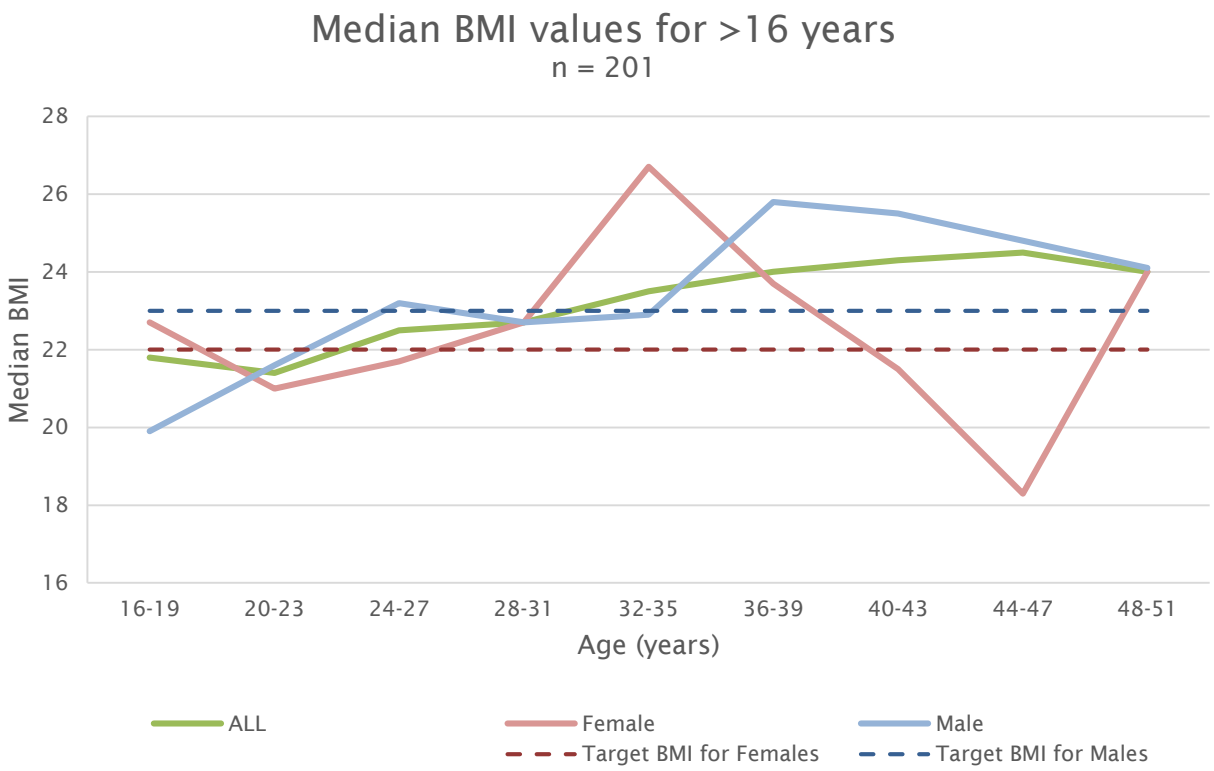
The spike in older ages reflects late diagnoses with CF reflecting a more mild or atypical disease, plus loss of those with severe disease. It is more obvious within the smaller numbers that we have in each age bracket in NZ. Other registries have presented the data as the percentage of children and adults with normal, mild, moderate or severely affected lung function.

Nutrition

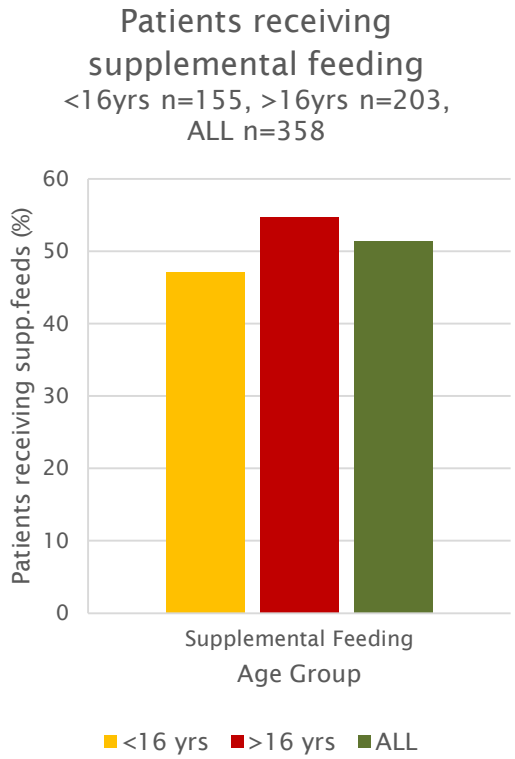


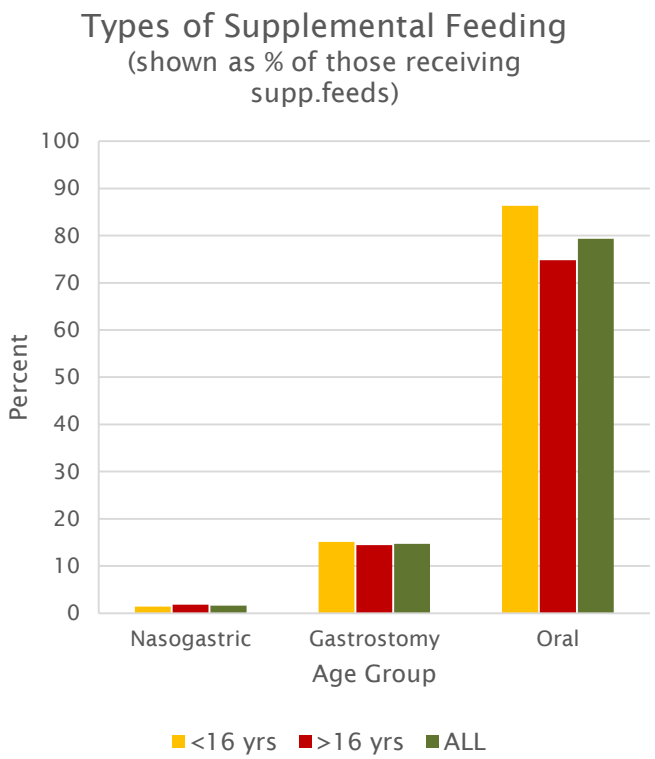
Age (yrs)	BMI Percentile	
	n	median
2	1	12.3
3	10	80.8
4	12	77.8
5	14	71.7
6	11	57.3
7	10	66.9
8	13	68.0
9	11	44.8
10	10	52.9
11	8	51.4
12	11	42.9
13	8	57.3
14	11	69.9
15	8	74.8
Total	138	

The dotted line is the marker to target weight for height in children. It appears that we perform well in this parameter in NZ.



Age (yrs)	All		Female		Male	
	n	median	n	median	n	median
16-19	38	21.8	15	22.7	23	19.9
20-23	40	21.4	15	21.0	25	21.6
24-27	25	22.5	13	21.7	12	23.2
28-31	28	22.7	11	22.7	17	22.7
32-35	19	23.5	8	26.7	11	22.9
36-39	9	24.0	3	23.7	6	25.8
40-43	15	24.3	6	21.5	9	25.5
44-47	8	24.5	3	18.3	5	24.8
48-51	5	24.0	3	24.0	2	24.1
52-55	2	26.1	1	28.4	1	23.8
56-59	4	27.2	3	24.1	1	32.0
>=60	8	25.5	4	28.6	4	22.7
Total	201		85		116	

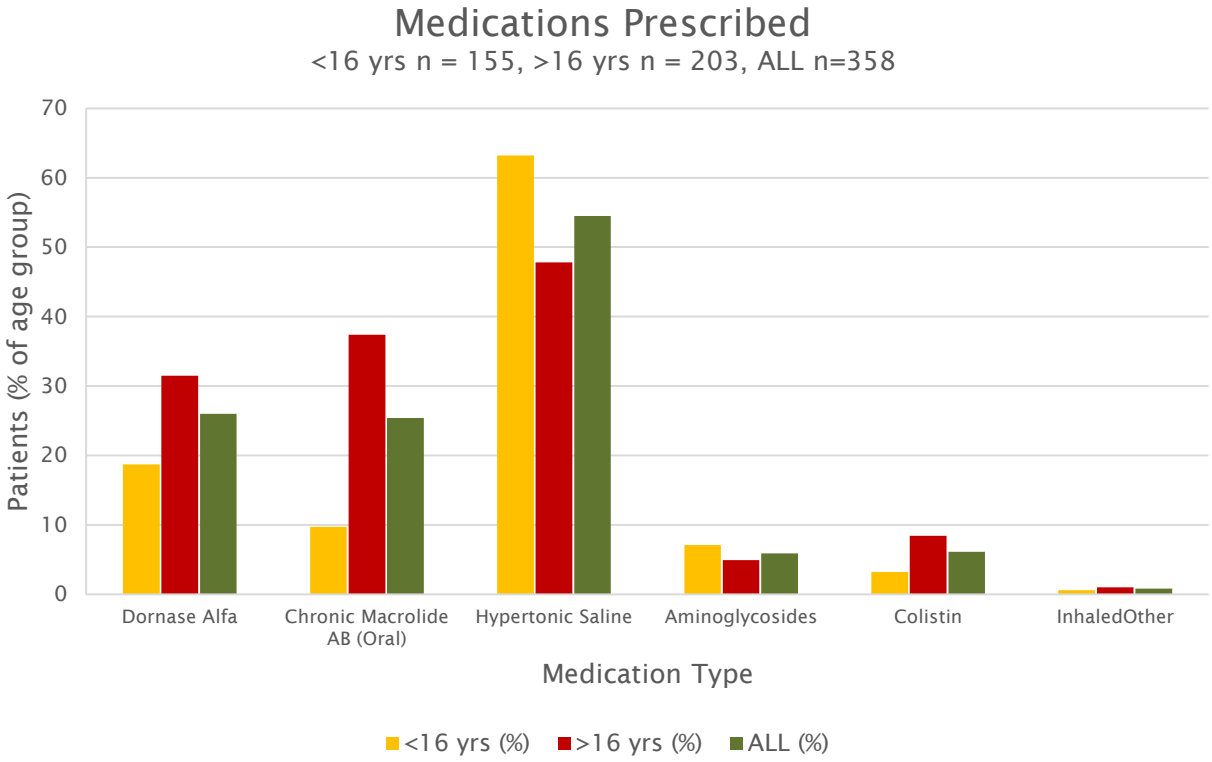




	<16 yrs, n = 155			>16 yrs, n = 203			All, n = 358		
	Yes	%	% <16yrs supp.	Yes	%	% >16 yrs supp.	Yes	%	% All supp.
Supplemental Feeding	73		47.1	111		54.7	184		51.4
Nasogastric	1	1.4	0.6	2	1.8	1.0	3	1.6	0.8
Gastrostomy	11	15.1	7.1	16	14.4	7.9	27	14.7	7.5
Oral	63	86.3	40.6	83	74.8	40.9	146	79.3	40.8

The early and high use of supplemental feeding may reflect on the overall good nutritional percentiles presented in previous graphs.

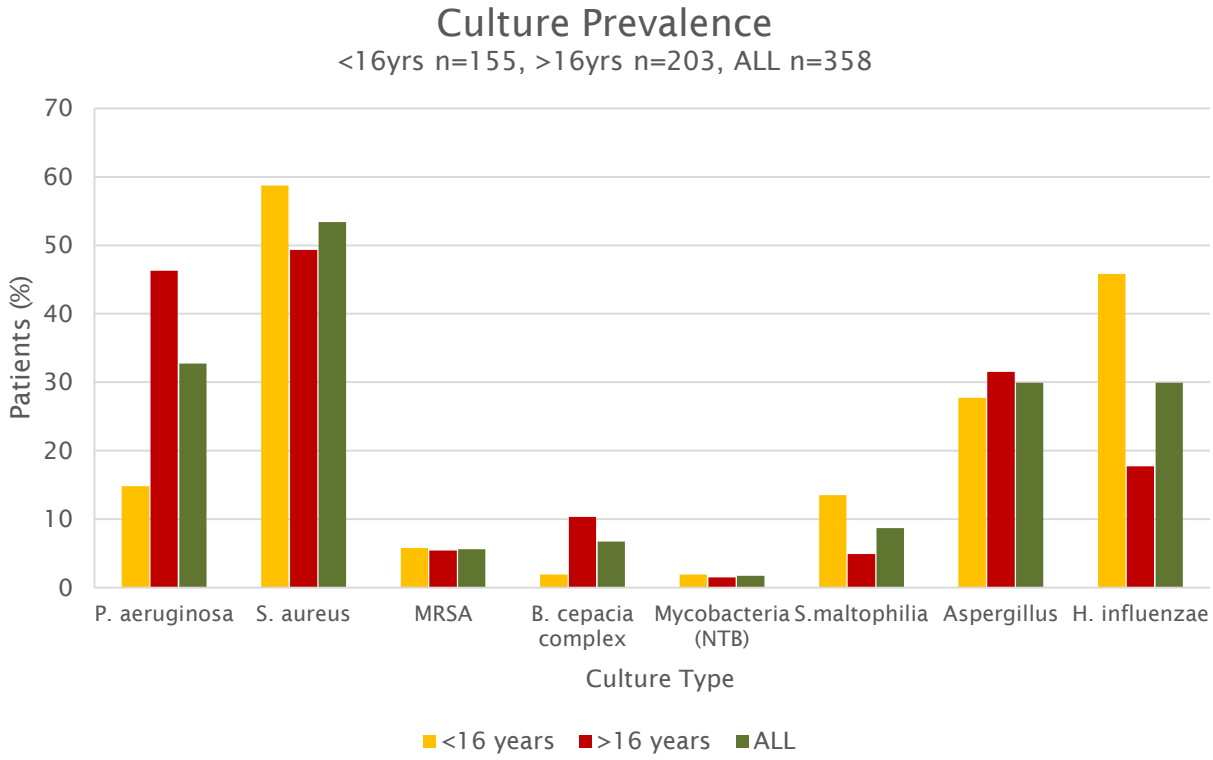
Medications



Medication	<16 yrs, n = 155		>16 yrs, n = 203		All, n = 358	
	Yes	%	Yes	%	Yes	%
Dornase Alfa	29	18.7	64	31.5	93	26.0
Chronic Macrolide AB (Oral)	15	9.7	76	37.4	91	25.4
Hypertonic Saline	98	63.2	97	47.8	195	54.5
Aminoglycosides	11	7.1	10	4.9	21	5.9
Colistin	5	3.2	17	8.4	22	6.1
InhaledOther	1	0.6	2	1.0	3	0.8

In New Zealand there is greater use of nebulised hypertonic saline, but less use of the other medications compared to that documented in other registries.

Microbiology



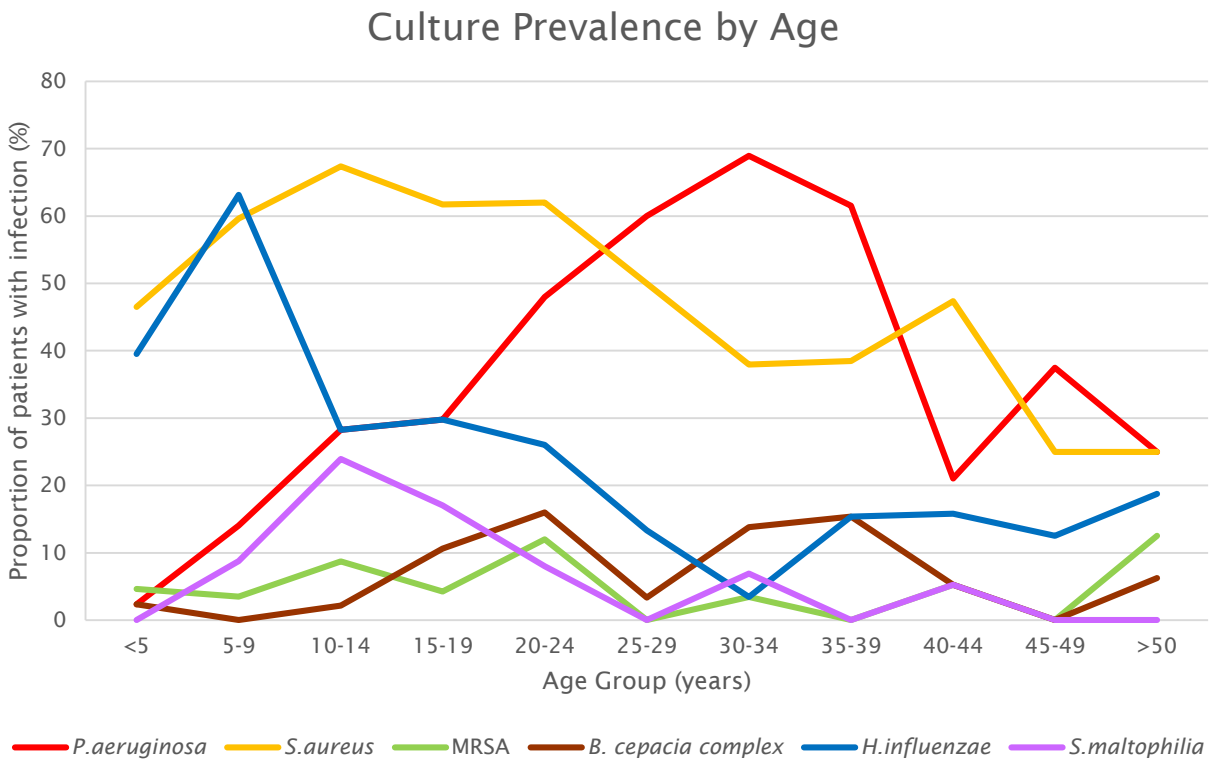
	<16 yrs, n = 155		>16 yrs, n = 203		All, n = 358	
	Yes	%	Yes	%	Yes	%
P. aeruginosa	23	14.8	94	46.3	117	32.7
S. aureus	91	58.7	100	49.3	191	53.4
MRSA	9	5.8	11	5.4	20	5.6
B. cepacia complex	3	1.9	21	10.3	24	6.7
Mycobacteria (NTB)	3	1.9	3	1.5	6	1.7
S.maltophilia	21	13.5	10	4.9	31	8.7
Aspergillus	43	27.7	64	31.5	107	29.9
H. influenzae	71	45.8	36	17.7	107	29.9

Our levels of *Staphylococcus aureus* are higher that the registries that capture data as 'chronic infection', but similar to the USA which captures the data as 'ever' in the last year.

Our levels of *Pseudomonas aeruginosa* infections seem lower if 'ever' or 'intermittent' infections as well as 'chronic' infection are included. One goal in the UK is to have only 30% of children having P. aeruginosa at the time of transfer to adult clinic.

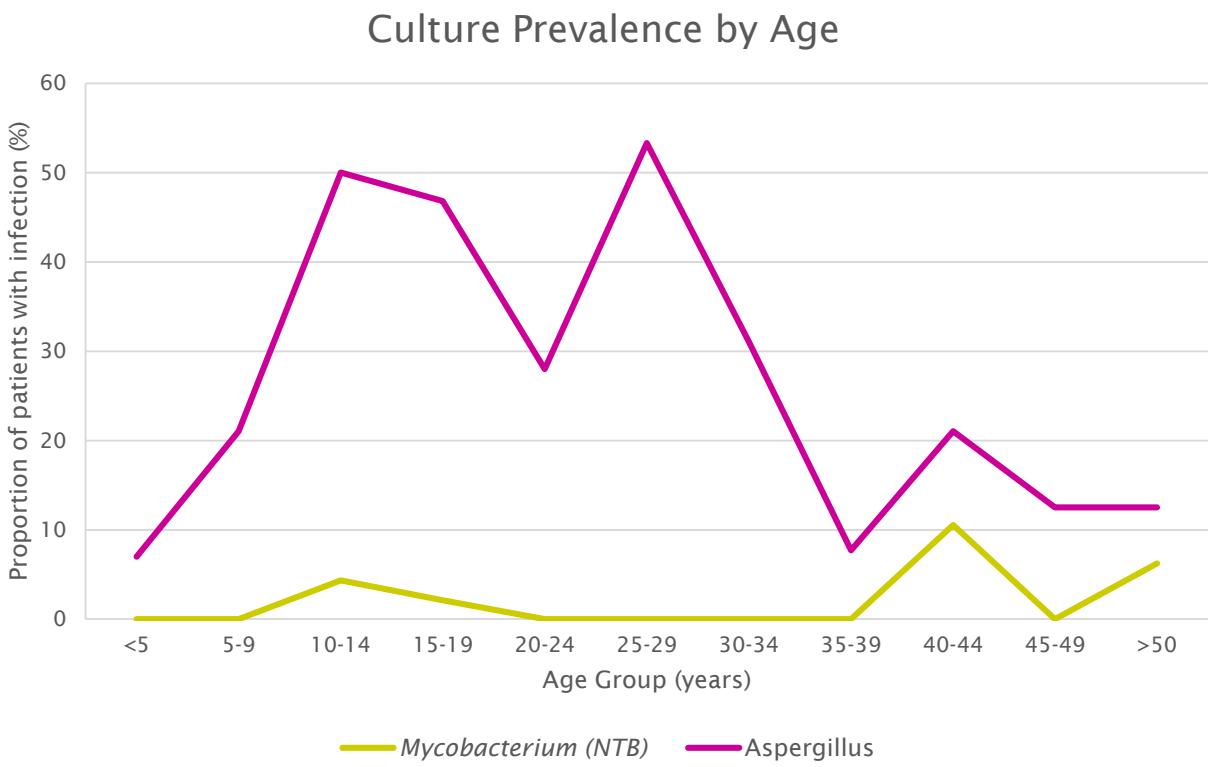
We have more *Burkholderia cepacia*, and less *Stenotrophomonas maltophilia* than elsewhere. We still have low levels of MRSA compared to other registry data but this appears to be increasing over recent years here. Not mentioned here is the division within the *Mycobacteria* categories – in the main we see *Mycobacteria avium intracellulare*, and while that is the same across countries, in some such as Australia they are reporting increased numbers of those with *Mycobacterium abscessus* – a more pathogenic organism.

It remains critical to check for *Mycobacterium* growth in sputum before commencing macrolide therapy and to review this at least annually. If *Mycobacteria* are present – macrolide therapy must be stopped.



Age (yrs)		*P.aeruginosa*		*S. aureus*		MRSA		*B. cepacia* complex		*H. infuenzae*		*S. maltophilia*	
	n	n	%	n	%	n	%	n	%	n	%	n	%
<5	43	1	2.3	20	46.5	2	4.7	1	2.3	17	39.5	0	0.0
5-9	57	8	14.0	34	59.6	2	3.5	0	0.0	36	63.2	5	8.8
10-14	46	13	28.3	31	67.4	4	8.7	1	2.2	13	28.3	11	23.9
15-19	47	14	29.8	29	61.7	2	4.3	5	10.6	14	29.8	8	17.0
20-24	50	24	48.0	31	62.0	6	12.0	8	16.0	13	26.0	4	8.0
25-29	30	18	60.0	15	50.0	0	0.0	1	3.3	4	13.3	0	0.0
30-34	29	20	69.0	11	37.9	1	3.4	4	13.8	1	3.4	2	6.9
35-39	13	8	61.5	5	38.5	0	0.0	2	15.4	2	15.4	0	0.0
40-44	19	4	21.1	9	47.4	1	5.3	1	5.3	3	15.8	1	5.3
45-49	8	3	37.5	2	25.0	0	0.0	0	0.0	1	12.5	0	0.0
>50	16	4	25.0	4	25.0	2	12.5	1	6.3	3	18.8	0	0.0
Total	358	117	32.7	191	53.4	20	5.6	24	6.7	107	29.9	31	8.7

The pattern of acquisition of these organisms with age are similar worldwide. The drop off in *P. aeruginosa* infection towards the older years reflects the more mild or atypical CF diagnosed in these older age brackets. It is more marked in this graph as it is based on the smaller numbers than elsewhere.



Age (yrs)		*Mycobacterium* (NTB)		*Aspergillus*	
	n	n	%	n	%
<5	43	0	0.0	3	7.0
5-9	57	0	0.0	12	21.1
10-14	46	2	4.3	23	50.0
15-19	47	1	2.1	22	46.8
20-24	50	0	0.0	14	28.0
25-29	30	0	0.0	16	53.3
30-34	29	0	0.0	9	31.0
35-39	13	0	0.0	1	7.7
40-44	19	2	10.5	4	21.1
45-49	8	0	0.0	1	12.5
>50	16	1	6.3	2	12.5
Total	358	6	1.7	107	29.9

Rates of *Aspergillus* presence in respiratory sections here are similar to that reported in Australia.

The presence of NTB seems low - not mentioned here is the division within the *Mycobacteria* categories – in the main we see *M. avium intracellulare*, and while that is the same across countries, in some such as Australia they are reporting increased numbers of those with *M. abscessus* – a more pathogenic organism.

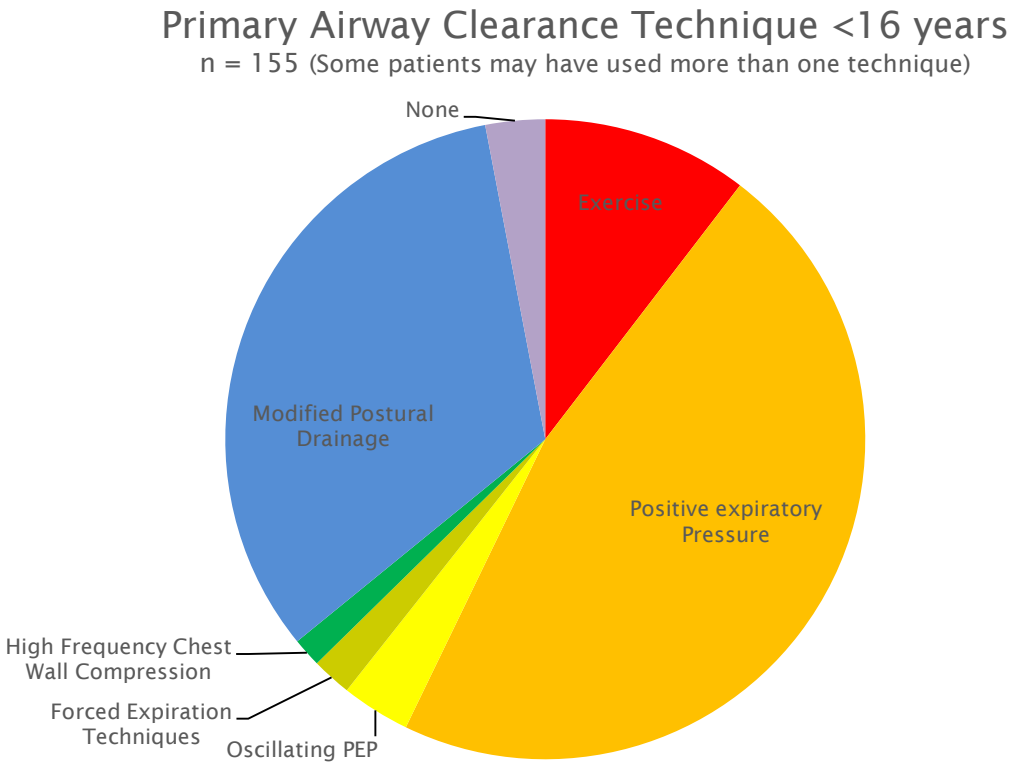
Hospital & Home IVA Days

		Home IV Days				Hospital IV Days				Total IVA Days
Age	n	n	%	Total Days	Mean IVA	n	%	Total Days	Mean IVA	
0-3	21	4	19.0	37	9.3	7	23.7	166	24	203
4-7	42	9	21.4	79	8.8	17	10.1	172	10	251
8-11	36	9	25.0	108	12.0	15	17.4	261	17	369
12-15	36	12	33.3	213	17.8	20	21.8	435	22	648
16-19	38	11	28.9	174	15.8	20	36.6	731	37	905
20-23	40	9	22.5	93	10.3	20	25.0	500	25	593
24-27	25	4	16.0	75	18.8	10	39.0	390	39	465
28-31	23	8	34.8	207	25.9	10	34.6	246	25	453
32-35	20	7	35.0	114	16.3	13	16.5	214	17	328
36-39	9	2	22.2	27	13.5	2	6.0	12	6	39
40-43	16	7	43.8	136	19.4	7	18.3	128	18	264
44-47	8	1	12.5	28	28.0	2	45.5	91	46	119
48-51	7	2	28.6	53	26.5	3	11.0	33	11	86
>52	17	3	17.6	25	8.3	4	5.8	23	6	48
	338	88		1369		150		3402		4771

29% of intravenous antibiotic therapy was given in the home – now even in the very young. In part it depends on the ability to have excellent intravenous access, the severity of the current infection in the individual, and the home environment at any particular time.

One factor that is missing when using home IV therapy is the ability to deliver professional physiotherapy which is equally important in overcoming an exacerbation.

Airway Clearance Techniques

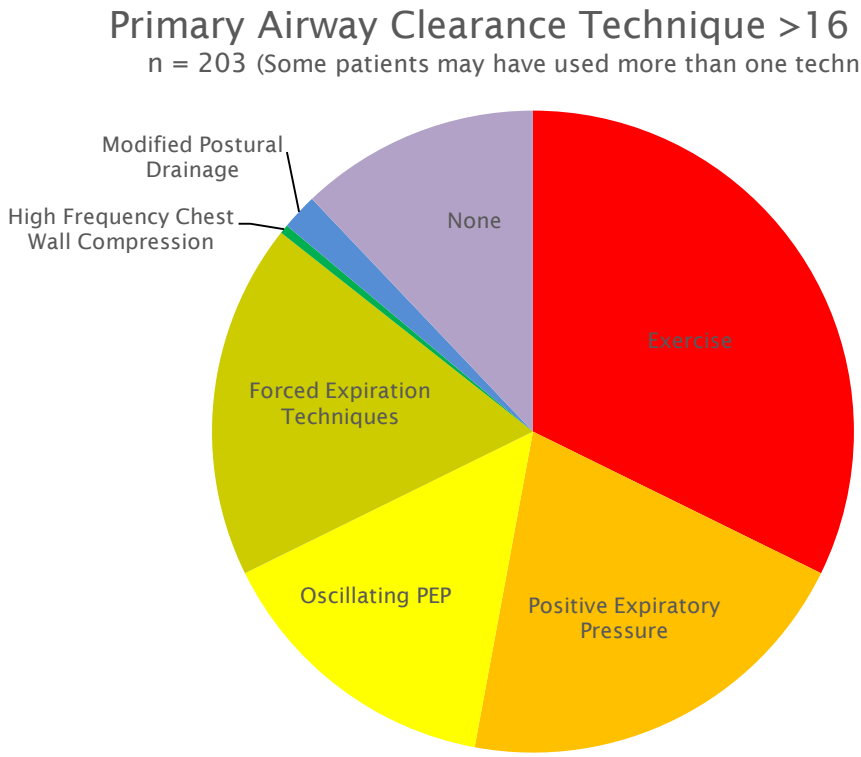


* number of individuals employing each technique at least once in the year.
Data collected from 155 patients

Technique	<16 years	
Exercise	21	13.5
Positive Expiratory Pressure	94	60.6
Oscillating PEP (e.g.: Flutter, Acapella, IPV)	7	4.5
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	4	2.6
High Frequency Chest Wall Compression (e.g.: vest)	3	1.9
Modified Postural Drainage	66	42.6
None	6	3.9
Total	201	

There are a variety of techniques used as a first option for airway clearance, with nearly half using some airway resistance device.

The percentage of children and young people using no airway clearance technique has generally decreased over the years that we have been collecting the data but with a slight increase from 2.3% last year to 3.9% this year.

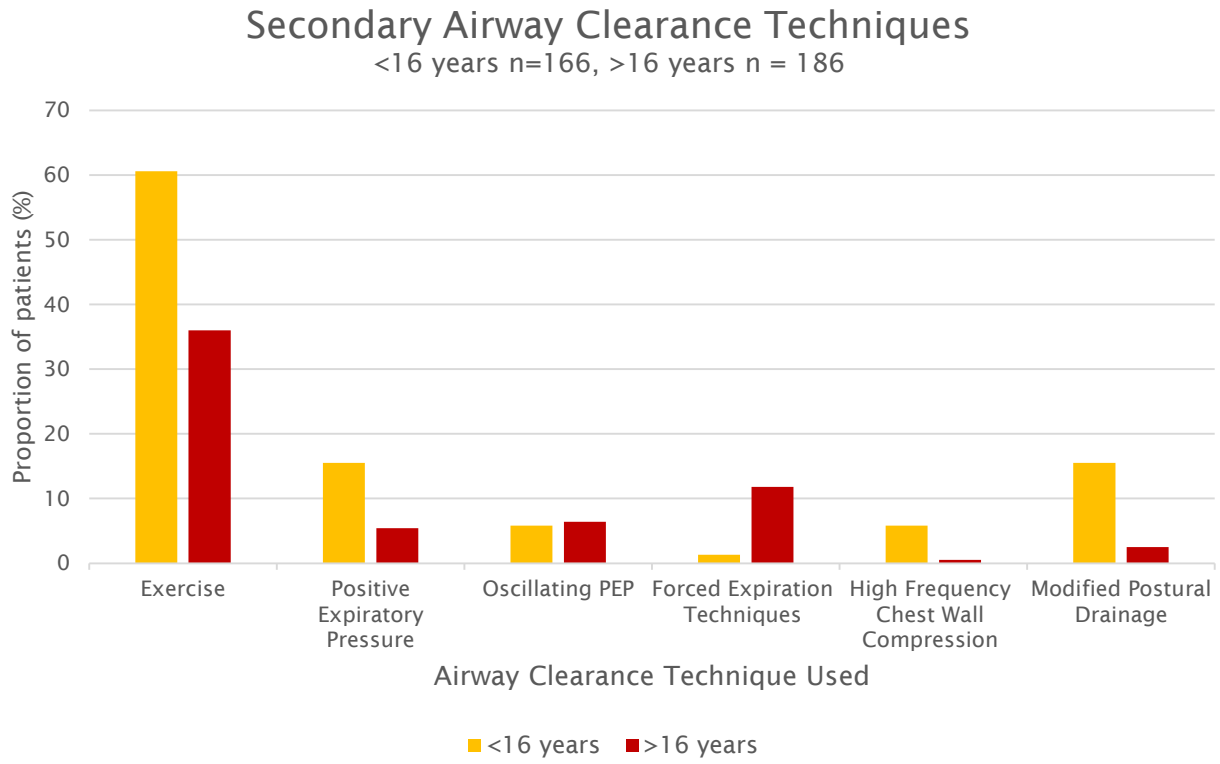


* number of individuals employing each technique at least once in the year.
Data collected from 203 patients

Technique	>16 years	
Exercise	72	35.5
Positive Expiratory Pressure	46	22.7
Oscillating PEP (e.g.: Flutter, Acapella, IPV)	33	16.3
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	40	19.7
High Frequency Chest Wall Compression (e.g.: vest)	1	0.5
Modified Postural Drainage	4	2.0
None	27	13.3
Total	223	

More adults than children and younger people use exercise as their primary airway clearance technique, with a similar number across the components using resistance devices.

Those using no airway clearance are higher in the adults than in children.

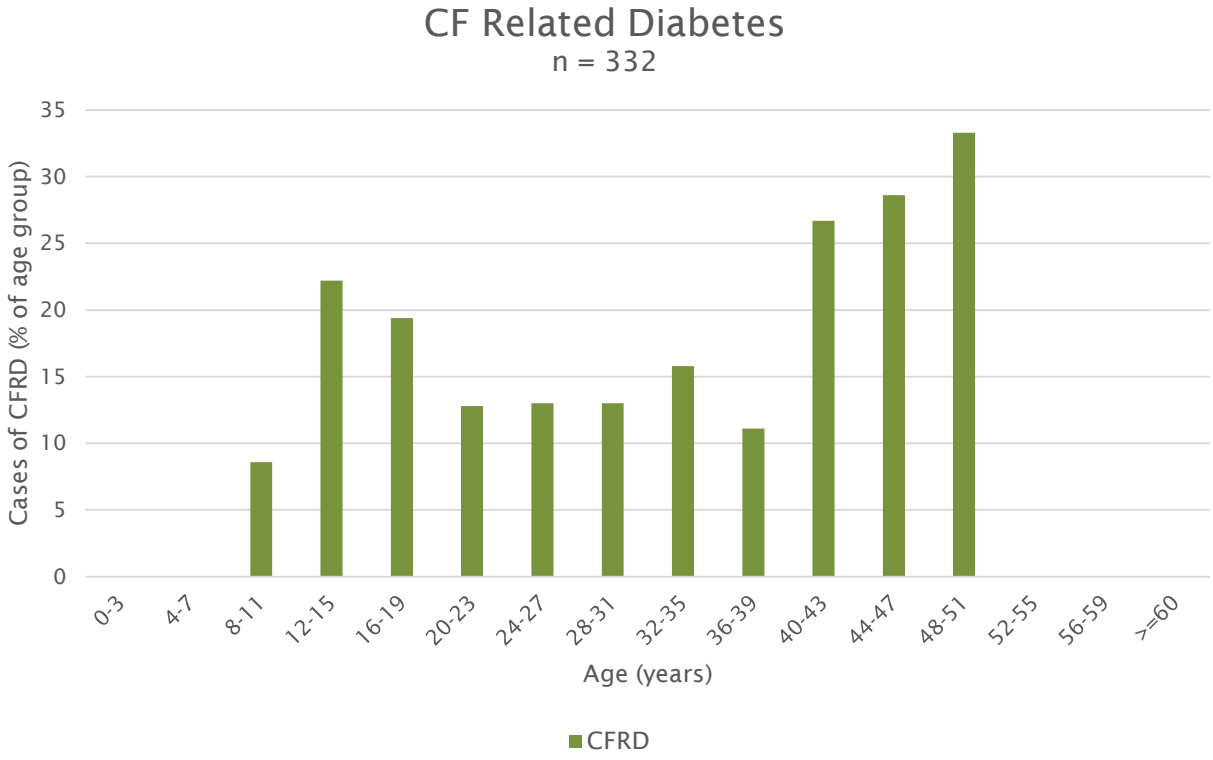


Data collected in 166 <16 years, 186 >16 years; Some patients may use more than one technique

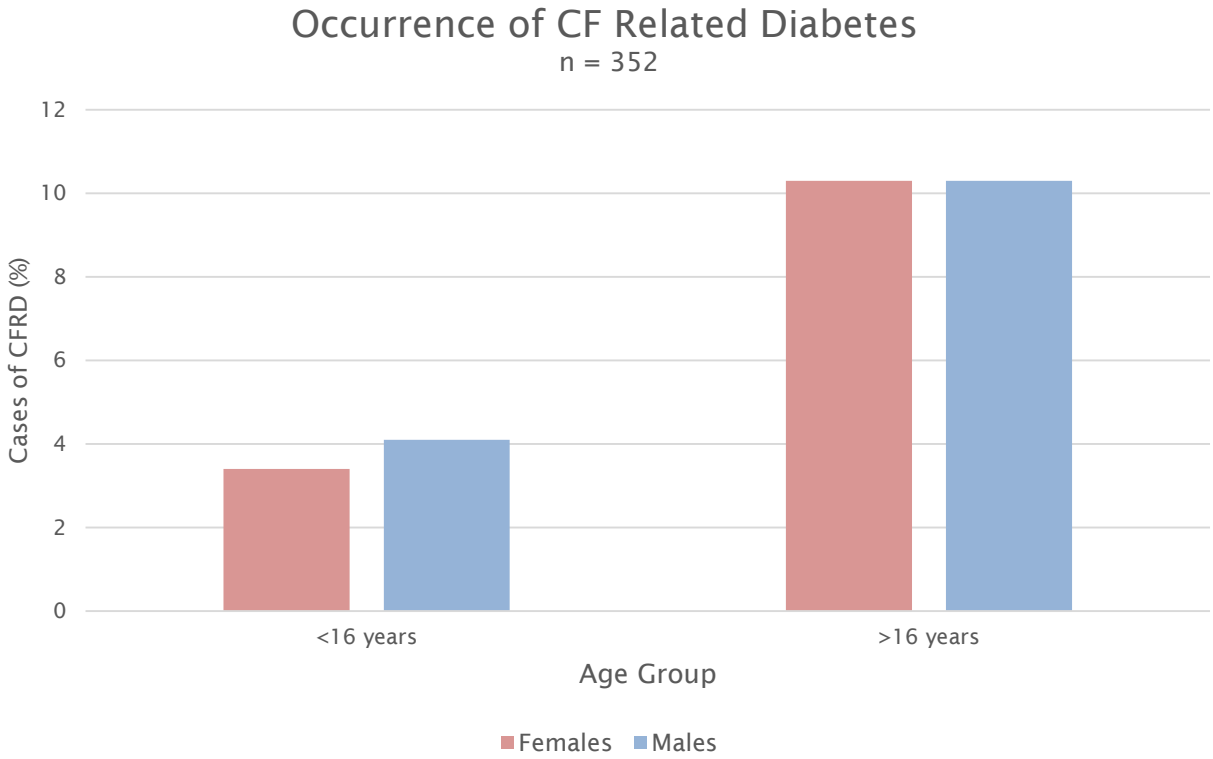
	<16 years		>16 years	
Technique	n	%	n	%
Exercise	94	60.6	73	36.0
Positive Expiratory Pressure	24	15.5	11	5.4
Oscillating PEP (eg: Flutter, Acapella, IPV)	9	5.8	13	6.4
Forced Expiration Techniques (eg:huff cough, active cyce breathing, autogenic drainage)	2	1.3	24	11.8
High Frequency Chest Wall Compression (eg: vest)	9	5.8	1	0.5
Modified Postural Drainage	24	15.5	5	2.5
Total	162		127	

Exercise remains a strong component of airway clearance

CF-Related Diabetes



Age (years)	Age n	CFRD n	% Age Group	%CF Population
0-3	25	0	0.0	0.0
4-7	44	0	0.0	0.0
8-11	35	3	8.6	0.9
12-15	36	8	22.2	2.4
16-19	36	7	19.4	2.1
20-23	39	5	12.8	1.5
24-27	23	3	13.0	0.9
28-31	23	3	13.0	0.9
32-35	19	3	15.8	0.9
36-39	9	1	11.1	0.3
40-43	15	4	26.7	1.2
44-47	7	2	28.6	0.6
48-51	6	2	33.3	0.6
52-55	2	0	0.0	0.0
56-59	4	0	0.0	0.0
>=60	9	0	0.0	0.0
	332	41		12.3



	n	CFRD n	%	<16 years	%	>16 years	%
Females	145	20	13.8	5	3.4	15	10.3
Males	187	21	14.5	6	4.1	15	10.3
Total	332	41	28.3	11	7.6	30	20.7

The overall percentage of persons with CF affected by CFRD is similar to other reports, but the younger age group seems less with similar results since collecting the data. A review of the timeliness and accuracy of our screening may be appropriate.

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

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