PORT CFNZ

2013 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 the Cystic Fibrosis Association of New Zealand.

For further information about the Association

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: Cystic fibrosis patients under are in New Zealand CF clinics, who have consented to have their data recorded in the Registry.

Suggested Citation: PORT CFNZ National Data Registry, 2013 Registry Report, Cystic Fibrosis Association of New Zealand.

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Introduction & Acknowledgements

On behalf of the Cystic Fibrosis Association of New Zealand and the Port CFNZ Steering Committee, we are delighted to present the New Zealand Cystic Fibrosis Patient Data Registry 2013 Report.

We would like to thank all the Nurses, Specialists and Administrators who have worked hard to get this data entered to enable a detailed analysis for NZ and the presentation of this report.

We also thank Shares in Life Foundation that has provided pivotal funding to maintain the database and assist centres with data entry.

This third registry report from the Port CFNZ database provides us with an increasingly accurate picture of CF outcomes for New Zealand with a high proportion of patients opted into providing data.

Further development of the database at the Canterbury District Health Board has been undertaken with a new database being written exclusively with the New Zealand clinical environment in mind and should provide improvement and gains in efficiency in data entry processes. Our sincere thanks to the Canterbury District Health Board for their ongoing commitment to this project

Above all, thank you to the persons with CF (children and adults alike) and their families for participating in this process. We hope you 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

At this stage, the Data registry gives national statistics only. As a nation, New Zealand has a total CF population that is close to those of a single clinic in larger countries. Because of this, statistically accurate and relevant data by clinic is not feasible.

However, our aim from 2014 onwards is to provide individual clinics a service of reporting on their own patient statistics to see where they sit against the national median, in order to provide a good platform for quality improvement and goal setting into the future. We will also encourage clinics to share this data with their patients.

Our smaller population size provides significant challenges to our Statistician as the 'outliers' in terms of age and key markers will have a much larger impact on statistics than they would on a larger data set. Because of this, 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.

As our NZ registry data becomes more robust and more 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 CFANZ website the PORT CF steering committee.

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

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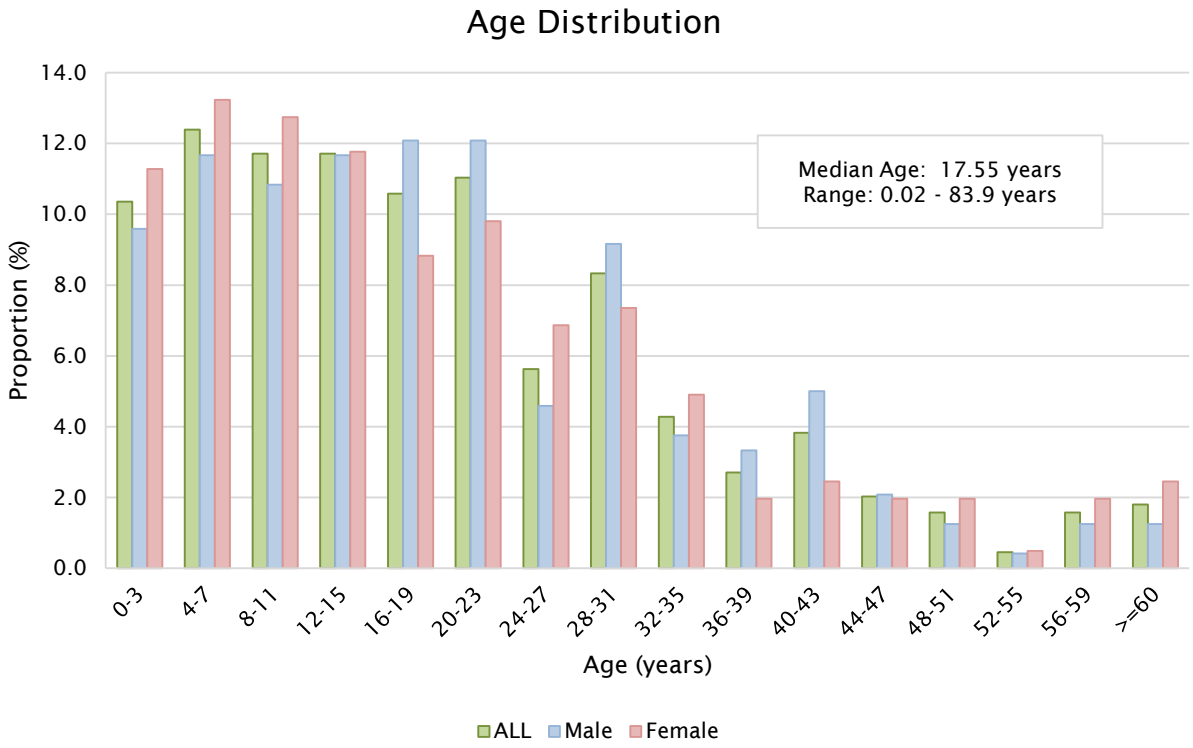
Key Indicators

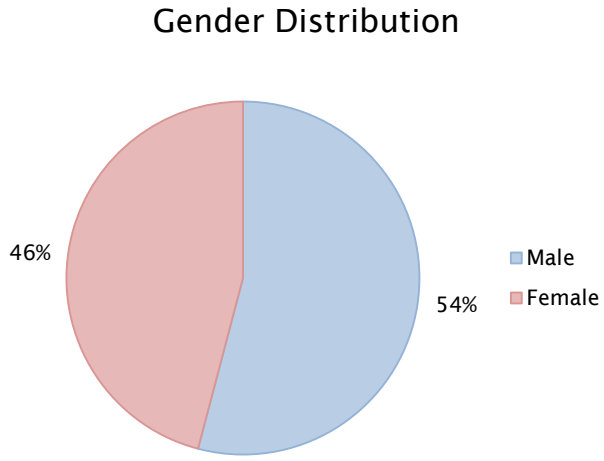
	2013	2012	2011
CF patients registered	444	423	415
Diagnosis age <1 year	5	11	11
Diagnosis age >16 years	3	2	
Age in years; median	17.55	16.15	15.71
PWCF aged >16 years	239 *53.8%*	214 *50.6%*	206 *49.6%*
Males	240 *54.1%*	228 *53.9%*	226 *54.6%*
Genotyped	426 *95.9%*	407 *96.2%*	364 *87.7%*
Median FEV1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

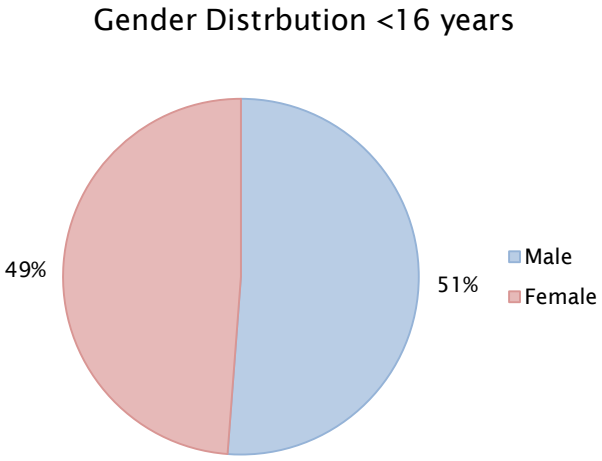
Demographics

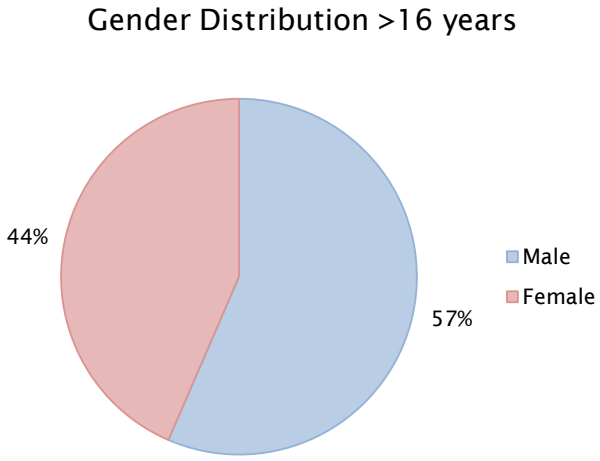


Age (yrs)	All		Male		Female	
	n	%	n	%	n	%
0-3	46	*10.4*	23	*9.6*	23	*11.3*
4-7	55	*12.4*	28	*11.7*	27	*13.2*
8-11	52	*11.7*	26	*10.8*	26	*12.7*
12-15	52	*11.7*	28	*11.7*	24	*11.8*
16-19	47	*10.6*	29	*12.1*	18	*8.8*
20-23	49	*11.0*	29	*12.1*	20	*9.8*
24-27	25	*5.6*	11	*4.6*	14	*6.9*
28-31	37	*8.3*	22	*9.2*	15	*7.4*
32-35	19	*4.3*	9	*3.8*	10	*4.9*
36-39	12	*2.7*	8	*3.3*	4	*2.0*
40-43	17	*3.8*	12	*5.0*	5	*2.5*
44-47	9	*2.0*	5	*2.1*	4	*2.0*
48-51	7	*1.6*	3	*1.3*	4	*2.0*
52-55	2	*0.5*	1	*0.4*	1	*0.5*
56-59	7	*1.6*	3	*1.3*	4	*2.0*
>=60	8	*1.8*	3	*1.3*	5	*2.5*
Total	444		240		204	
Median	17.55 years					
Range	0.02 - 83.9 years					

We have increased numbers of persons with CF contributing their health data to PORT CFNZ registry which makes it increasingly accurate and increasingly useful for both all persons with CF and for health personnel alike. It is gratifying to see the median age of PWCF increase every year over the three years we have collected this data, with an increased proportion of people in the adult age bracket.







	All		<16 years		>16 years	
	n	%	n	%	n	%
Male	240	*54.1*	105	*51.2*	135	*56.5*
Female	204	*45.9*	100	*48.8*	104	*43.5*
Total	444		205		239	

The gender distribution is even in the early years but less so in the adult years – in part we know that young women can have accelerated disease. It is difficult to compare this with other registries.

Genotype

426 (95.9%) of 444 patients have been genotyped with a recorded value.

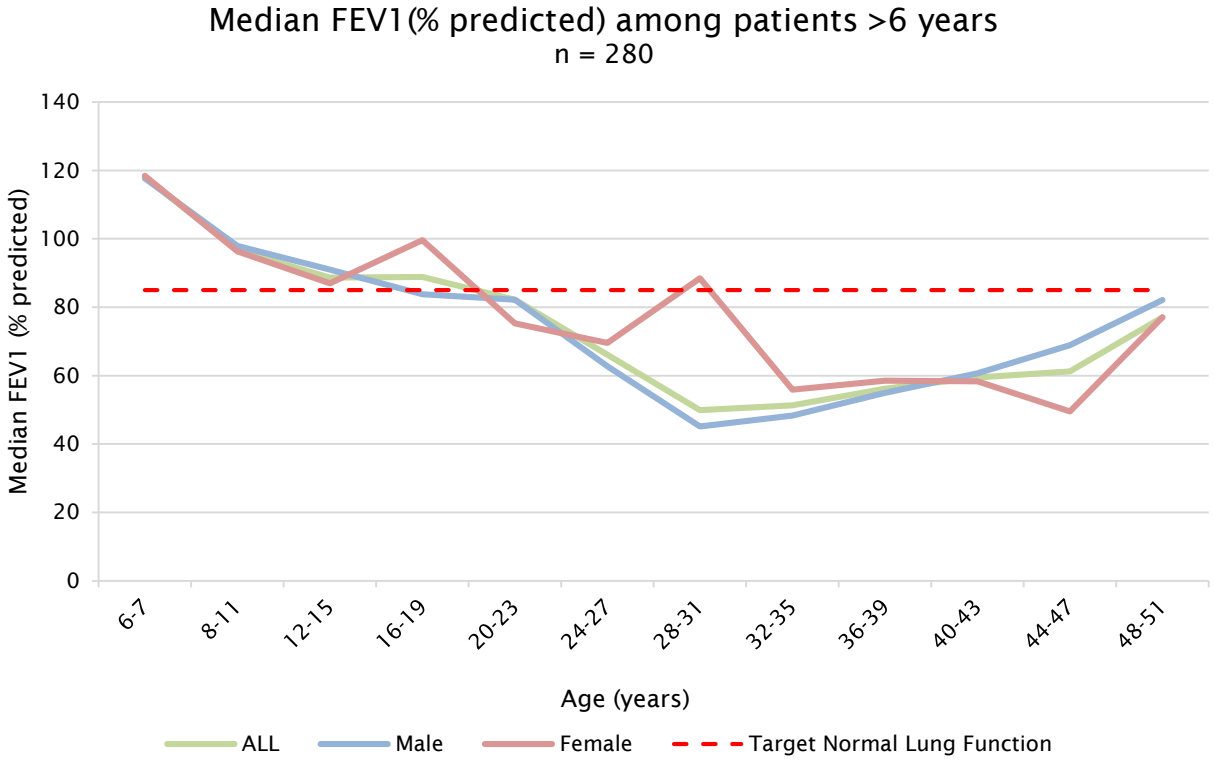
F508del Mutations	n	%
Homozygous F508del	220	51.6
Heterozygous F508del	165	38.7
No F508del or both unidentified	41	9.6
Total	426	

Mutations Identified	c.DNA Name	n	%
F508del	c.1521_1523delCTT	605	71.0
G551D	c.1652G>A	28	3.3
G542X	c.1624G>T	27	3.2
R117H	c.350G>A	18	2.1
G85E	c.254G>A	6	0.7
N1303K	c.3909c>G	6	0.7
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	0.4
2789+2insA	c.2657+2_2657+3insA	3	0.4
A455E	c.1364C>A	3	0.4
c.3718-2477C>T	c.3718-2477C>T	3	0.4
Other		86	10.1
Unidentified		43	5.0
		852	

The way that the genetic mutations are classified has been standardised such that they genes are classified by their DNA abnormalities, by the protein abnormalities, leaving behind their traditional names (legacy names).

Most of the persons on the database have had their genotype determined, which will become of increasing importance in years to come. F508 remains the dominant gene at 71%, however this is far less than in the UK (90.8%), USA (86.7%), or Australia (85.3%), suggesting that with our ethnic diversity, we have greater numbers of less common genes present in our community.

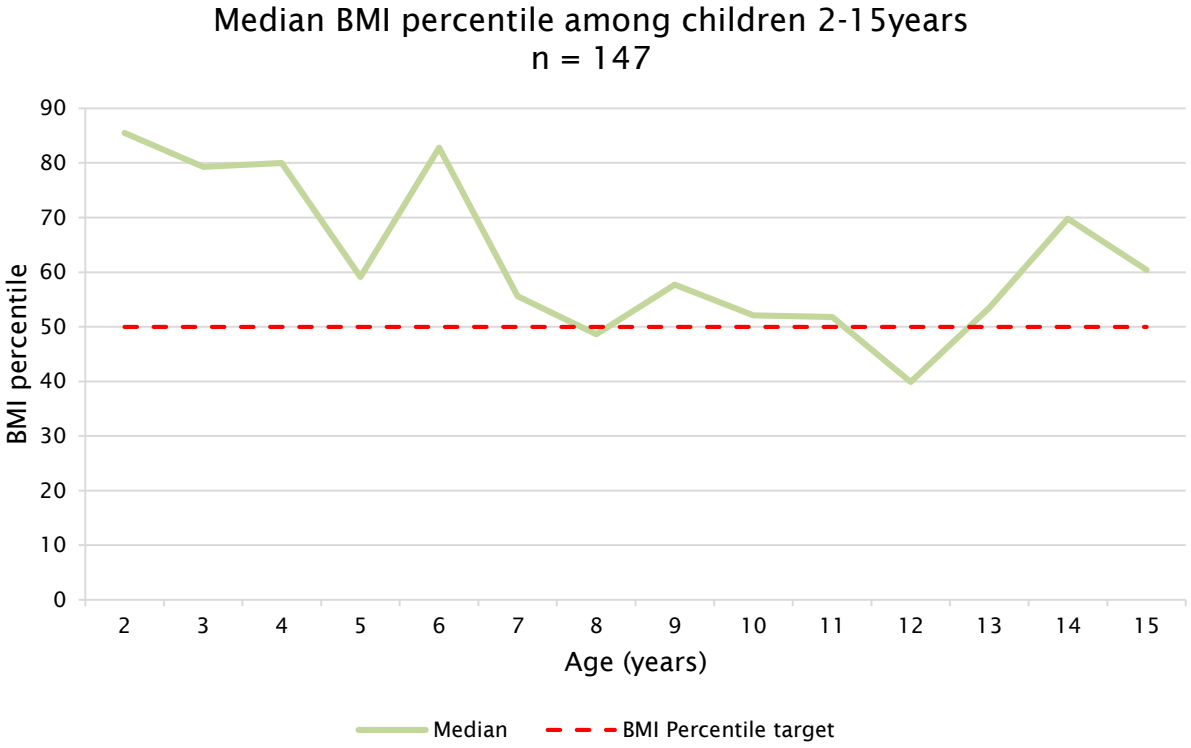
Respiratory



Age (yrs)	All		Male		Female	
	n	median	n	median	n	median
6-7	18	118.1	7	117.7	11	118.4
8-11	41	96.6	19	97.9	22	96.3
12-15	39	88.7	20	91.0	19	87.0
16-19	32	88.9	22	83.8	10	99.6
20-23	41	82.2	25	82.2	16	75.3
24-27	20	66.1	9	62.7	11	69.6
28-31	27	49.9	17	45.1	10	88.5
32-35	16	51.3	9	48.3	7	55.9
36-39	8	56.2	6	55.0	2	58.5
40-43	14	59.5	9	60.7	5	58.3
44-47	6	61.2	3	68.9	3	49.5
48-51	6	77.1	2	82.1	4	77.1
52-55	1	46.2	0		1	46.2
56-59	4	69.1	3	79.0	1	59.2
>=60	7	64.5	3	71.9	4	57.3
Total	280		154		126	

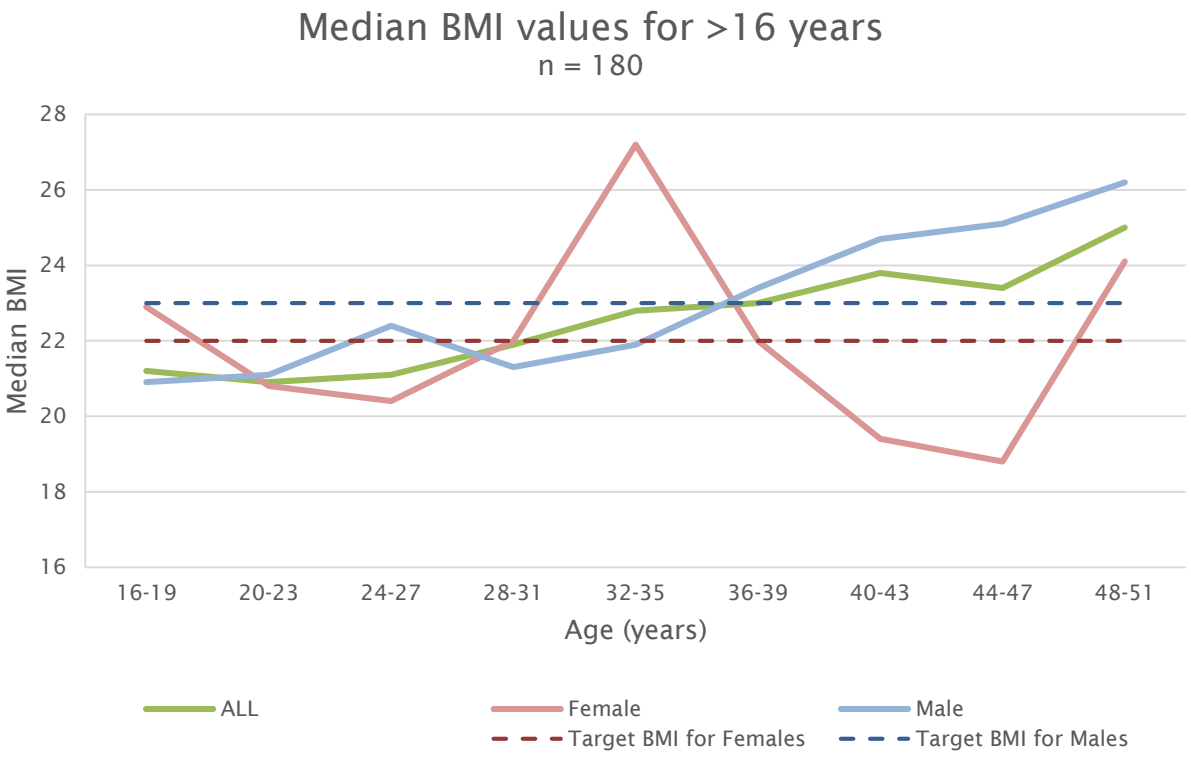
The slope of lung function (FEV1) over time is very similar to the other registries with our target lung function being greater than 85% which is in keeping with the UK registry. Our median FEV1 for those <16 years and >16 years is better than the median FEV1 in the 2012 registry report for USA.
Other registries have presented the data as the percentage of children and adults with normal, mild, moderate or severely affected lung function. We cannot compare this directly, but may be able to do this in future.

Nutrition

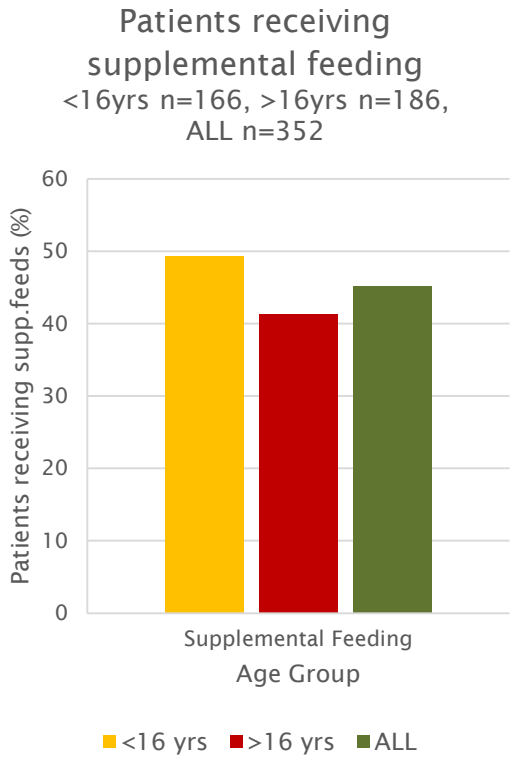


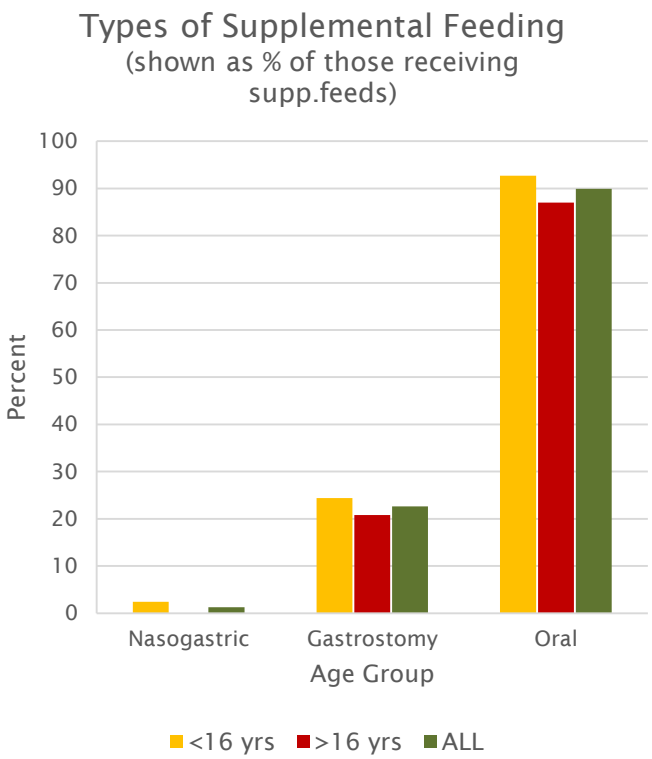
	n	median
2	7	85.5
3	15	79.3
4	15	80.0
5	8	59.1
6	15	82.8
7	9	55.6
8	13	48.6
9	5	57.7
10	12	52.1
11	13	51.8
12	8	39.9
13	8	53.5
14	10	69.8
15	14	60.4
Total	152	

The dotted line is the marker to target weight for height in children. As mentioned in the previous years' PORT CFNZ reports – our nutrition in NZ seems to be very good compared to the graphs seen in other registries.



Age (yrs)	All		Female		Male	
	n	median	n	median	n	median
16-19	30	21.2	11	22.9	19	20.9
20-23	41	20.9	16	20.8	25	21.1
24-27	20	21.1	11	20.4	9	22.4
28-31	27	21.9	10	22.0	17	21.3
32-35	16	22.8	7	27.2	9	21.9
36-39	8	23.0	2	22.0	6	23.4
40-43	14	23.8	5	19.4	9	24.7
44-47	6	23.4	3	18.8	3	25.1
48-51	6	25.0	4	24.1	2	26.2
52-55	1	28.1	1	28.1	0	
56-59	4	20.6	1	17.6	3	21.0
>=60	7	23.1	4	25.1	3	23.1
Total	180		75		105	

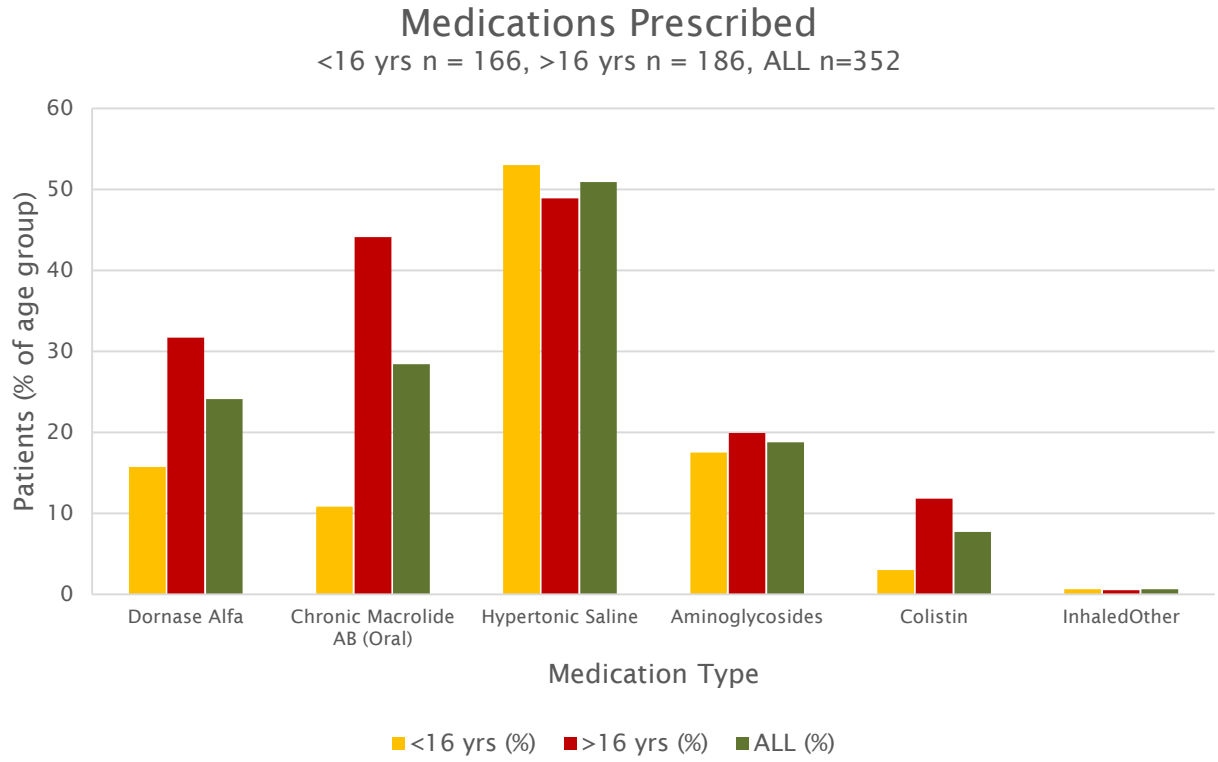




	<16 yrs, n = 166			>16 yrs, n=186			All, n = 352		
	Yes	%	% <16yrs supp.	Yes	%	% >16 yrs supp.	Yes	%	% All supp.
Supplemental Feeding	82	49.4		77	41.4		159	45.2	
Nasogastric	2	1.2	2.4	0	0.0	0.0	2	0.6	1.3
Gastrostomy	20	12.1	24.4	16	8.6	20.8	36	10.2	22.6
Oral	76	45.8	92.7	67	36.0	87.0	143	40.6	89.9

Supplemental feeding is an important part of CF nutritional management predominantly relying upon oral supplements.

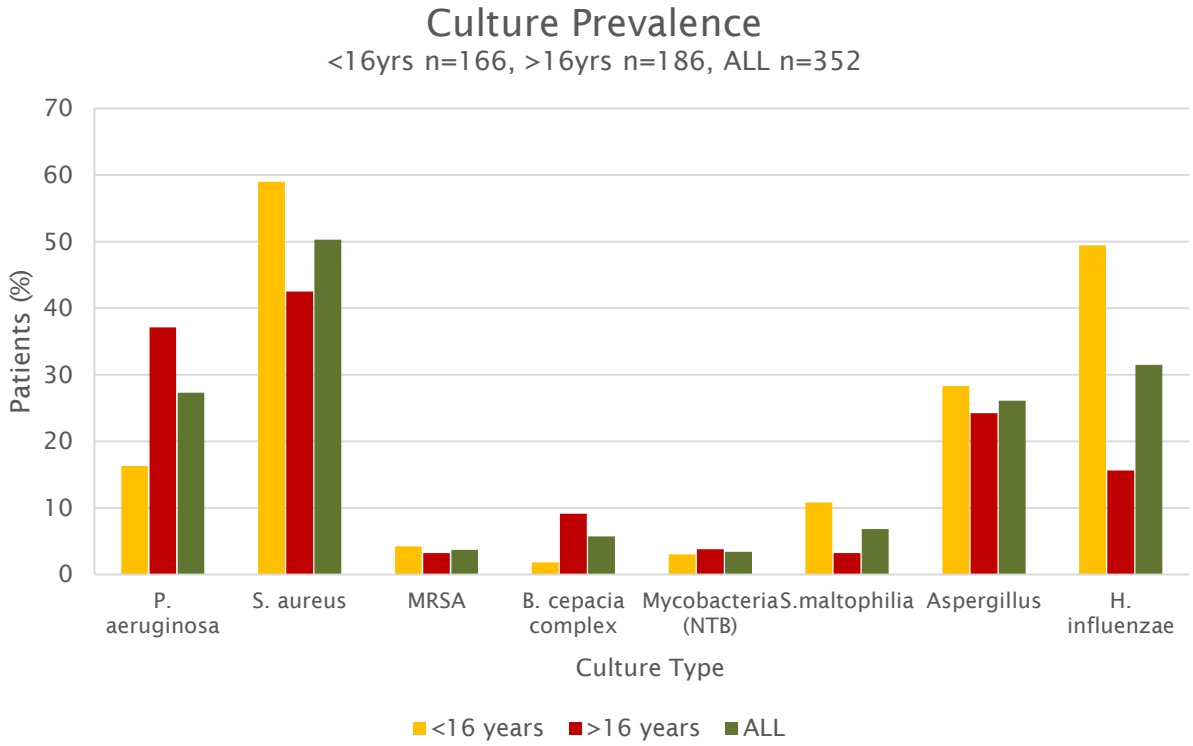
Medications



Medication	<16 yrs, n = 166		>16 yrs, n=186		All, n = 352	
	Yes	%	Yes	%	Yes	%
Dornase Alfa	26	15.7	59	31.7	85	24.1
Chronic Macrolide AB (Oral)	18	10.8	82	44.1	100	28.4
Hypertonic Saline	88	53.0	91	48.9	179	50.9
Aminoglycosides	29	17.5	37	19.9	66	18.8
Colistin	5	3.0	22	11.8	27	7.7
InhaledOther	1	0.6	1	0.5	2	0.6

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.

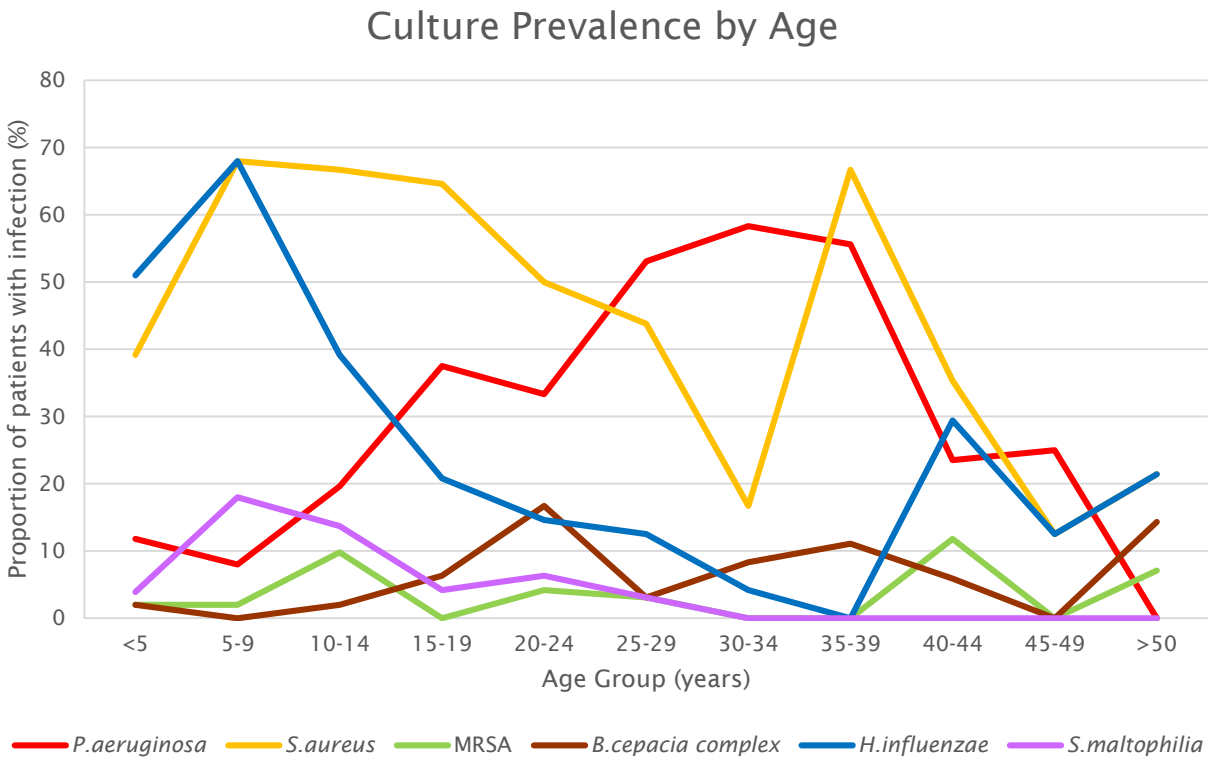
Microbiology



	<16 yrs, n = 166		>16 yrs, n=186		All, n=352	
	Yes	%	Yes	%	Yes	%
P. aeruginosa	27	16.3	69	37.1	96	27.3
S. aureus	98	59.0	79	42.5	177	50.3
MRSA	7	4.2	6	3.2	13	3.7
B. cepacia complex	3	1.8	17	9.1	20	5.7
Mycobacteria (NTB)	5	3.0	7	3.8	12	3.4
S.maltophilia	18	10.8	6	3.2	24	6.8
Aspergillus	47	28.3	45	24.2	92	26.1
H. influenzae	82	49.4	29	15.6	111	31.5

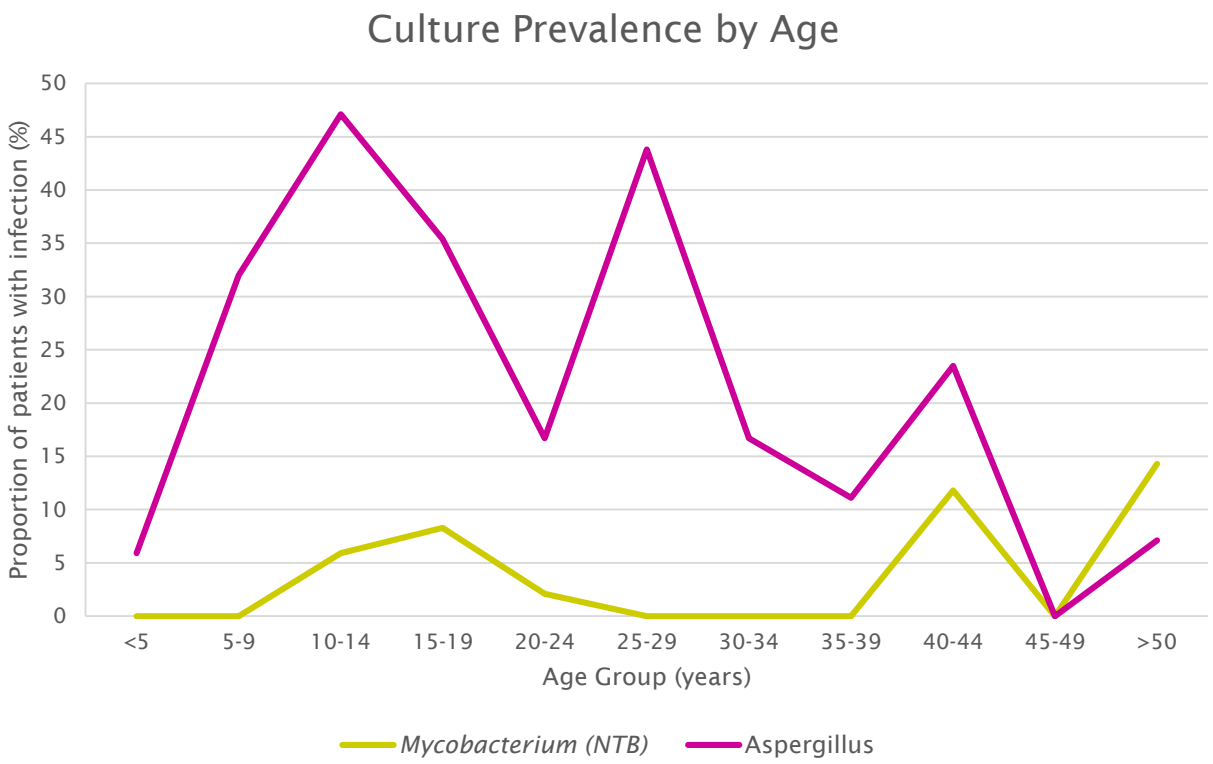
Our levels of *S.aureus* are higher that the registries that capture data as 'chronic infection', but similar to the USA which captures the data as 'ever' in the last year.
Our levels of *P.aeruginosa* infections seem lower if 'ever' or 'intermittent' infections as well as 'chronic' infection are included. 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 *B.cepacia*, and less *S.maltophilia* than elsewhere. We still have low levels of MRSA.

Compared to our last two years of Registry data here in NZ, we have a slight increases over time of *B.cepacia, S.maltophilia,* and MRSA – in part as we are likely to be looking more assiduously and identifying these organisms.



Age (yrs)		*P.aeruginosa*		*S. aureus*		MRSA		*B. cepacia* complex		*H. infuenzae*		*S. maltophilia*	
	n	n	%	n	%	n	%	n	%	n	%	n	%
<5	51	6	11.8	20	39.2	1	2.0	1	2.0	26	51.0	2	3.9
5-9	51	4	8.0	34	68.0	1	2.0	0	0.0	34	68.0	9	18.0
10-14	51	10	19.6	34	66.7	5	9.8	1	2.0	20	39.2	7	13.7
15-19	48	18	37.5	31	64.6	0	0.0	3	6.3	10	20.8	2	4.2
20-24	48	16	33.3	24	50.0	2	4.2	8	16.7	7	14.6	3	6.3
25-29	32	17	53.1	14	43.8	1	3.1	1	3.1	4	12.5	1	3.1
30-34	24	14	58.3	4	16.7	0	0.0	2	8.3	1	4.2	0	0.0
35-39	9	5	55.6	6	66.7	0	0.0	1	11.1	0	0.0	0	0.0
40-44	17	4	23.5	6	35.3	2	11.8	1	5.9	5	29.4	0	0.0
45-49	8	2	25.0	1	12.5	0	0.0	0	0.0	1	12.5	0	0.0
>50	14	0	0.0	3	21.4	1	7.1	2	14.3	3	21.4	0	0.0
Total	352	96	27.3	177	50.3	13	3.7	20	5.7	111	31.5	24	6.8

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 and 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	51	0	0.0	3	5.9
5-9	51	0	0.0	16	32.0
10-14	51	3	5.9	24	47.1
15-19	48	4	8.3	17	35.4
20-24	48	1	2.1	8	16.7
25-29	32	0	0.0	14	43.8
30-34	24	0	0.0	4	16.7
35-39	9	0	0.0	1	11.1
40-44	17	2	11.8	4	23.5
45-49	8	0	0.0	0	0.0
>50	14	2	14.3	1	7.1
Total	352	12	3.4	92	26.1

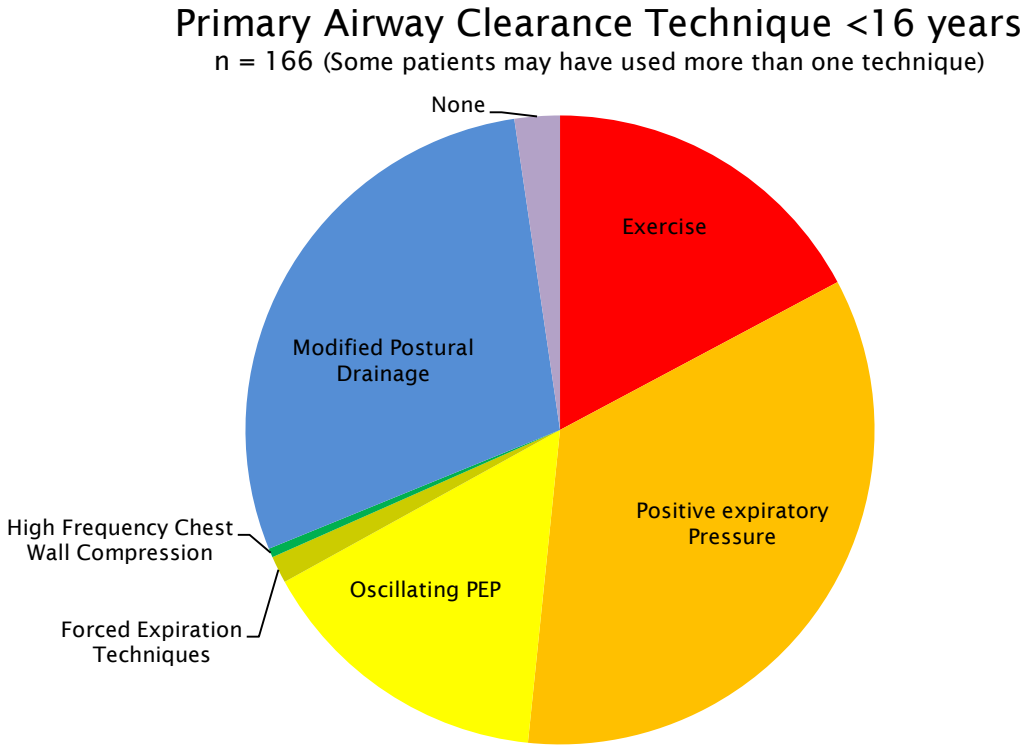
Rates of *Aspergillus* presence in respiratory sections here are similar to that reported in Australia, not all the registries have reported this indicator.
The presence of NTB seems low and we need to be sure we are looking for it 1-2 times per year especially in those considering or on chronic macrolide therapy.

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	31	1	3.2	16	16.0	8	25.8	156	20	172
4-7	38	10	26.3	108	10.8	15	39.5	220	15	328
8-11	35	9	25.7	125	13.9	13	37.1	221	17	346
12-15	37	12	32.4	443	36.9	25	67.6	680	27	1123
16-19	29	5	17.2	307	61.4	14	48.3	402	29	709
20-23	38	5	13.2	51	10.2	18	47.4	648	36	699
24-27	19	3	15.8	94	31.3	7	36.8	362	52	456
28-31	22	7	31.8	150	21.4	11	50.0	286	26	436
32-35	15	5	33.3	62	12.4	6	40.0	115	19	177
36-39	7	2	28.6	23	11.5	1	14.3	5	5	28
40-43	13	5	38.5	93	18.6	5	38.5	58	12	151
44-47	6	2	33.3	52	26.0	3	50.0	87	29	139
48-51	6	3	50.0	121	40.3	4	66.7	76	19	197
52-55	1	0	0.0	0		0	0.0	0		0
56-59	3	1	33.3	0	0.0	2	66.7	6	3	6
>=60	7	2	28.6	16		3	42.9	58	19	74
	307	72		1661		135		3380		5041

33% of intravenous antibiotic therapy was given in the home. Less in the very young age brackets appropriately, but it was available across all ages. It is presented differently in differing reports – between 12-14% of children and adults have had home IV therapy in the Australian registry.

Airway Clearance Techniques



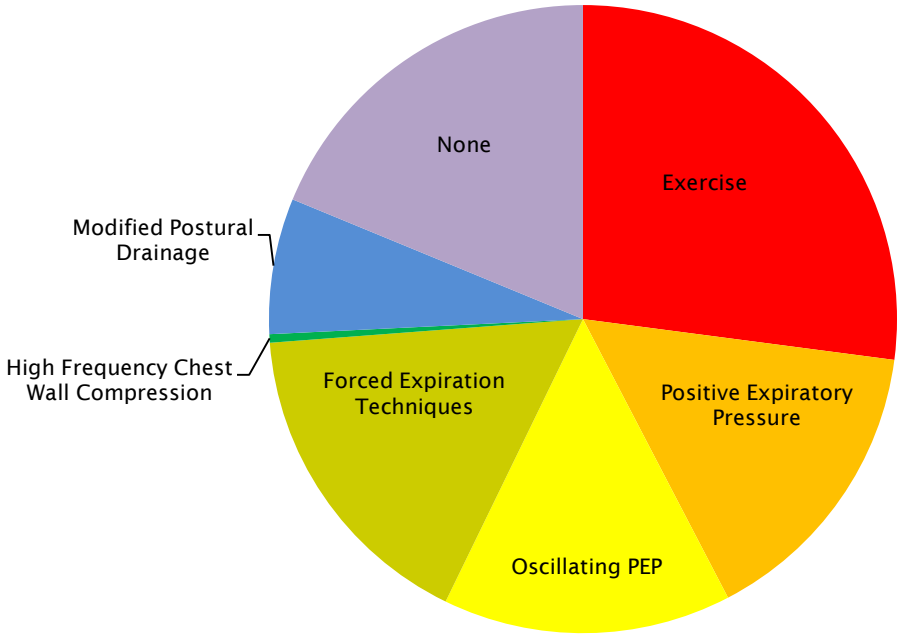
* number of individuals employing each technique at least once in the year. Data collected from 166 patients

Technique	<16 years	
Exercise	37	17.2
Positive Expiratory Pressure	74	34.4
Oscillating PEP (e.g.: Flutter, Acapella, IPV)	33	15.3
Forced Expiration Techniques: (e.g. huff cough, active cycle breathing, autogenic drainage)	3	1.4
High Frequency Chest Wall Compression: (e.g.: vest)	1	0.5
Modified Postural Drainage	62	28.8
None	5	2.3
Total	215	

There are a variety of techniques used as a first option for airway clearance, with nearly half using some airway resistance device.
It is reassuring to see that the percentage pf children and young people using no airway clearance technique has decreased considerably from 8.3% last year to 2.3% this year.

Primary Airway Clearance Technique >16 years

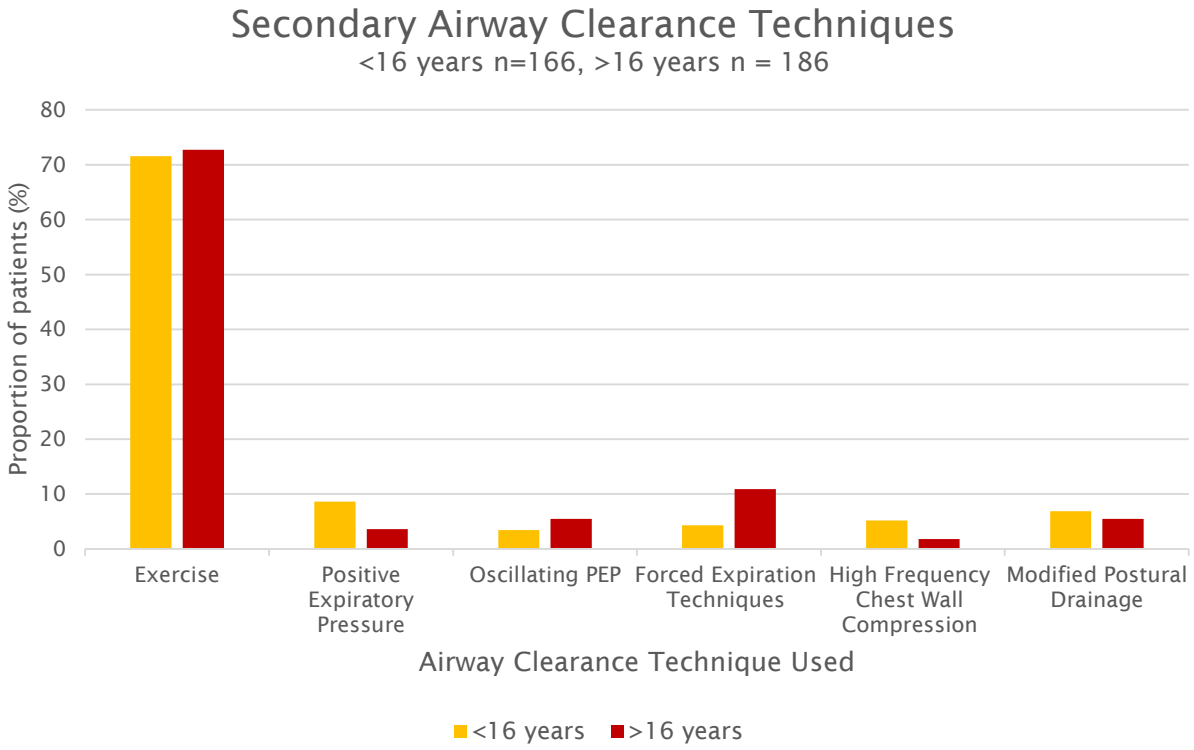
n = 186 (Some patients may have used more than one technique)



* number of individuals employing each technique at least once in the year. Data collected from 186 patients

Technique	>16 years	
Exercise	62	27.1
Positive Expiratory Pressure	35	15.3
Oscillating PEP (e.g.: Flutter, Acapella, IPV)	34	14.8
Forced Expiration Techniques: (e.g. huff cough, active cycle breathing, autogenic drainage)	38	16.6
High Frequency Chest Wall Compression: (e.g.: vest)	1	0.4
Modified Postural Drainage	16	7.0
None	43	18.8
Total	229	

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.
Of concern is the number not relying on any airway clearance to stay well.

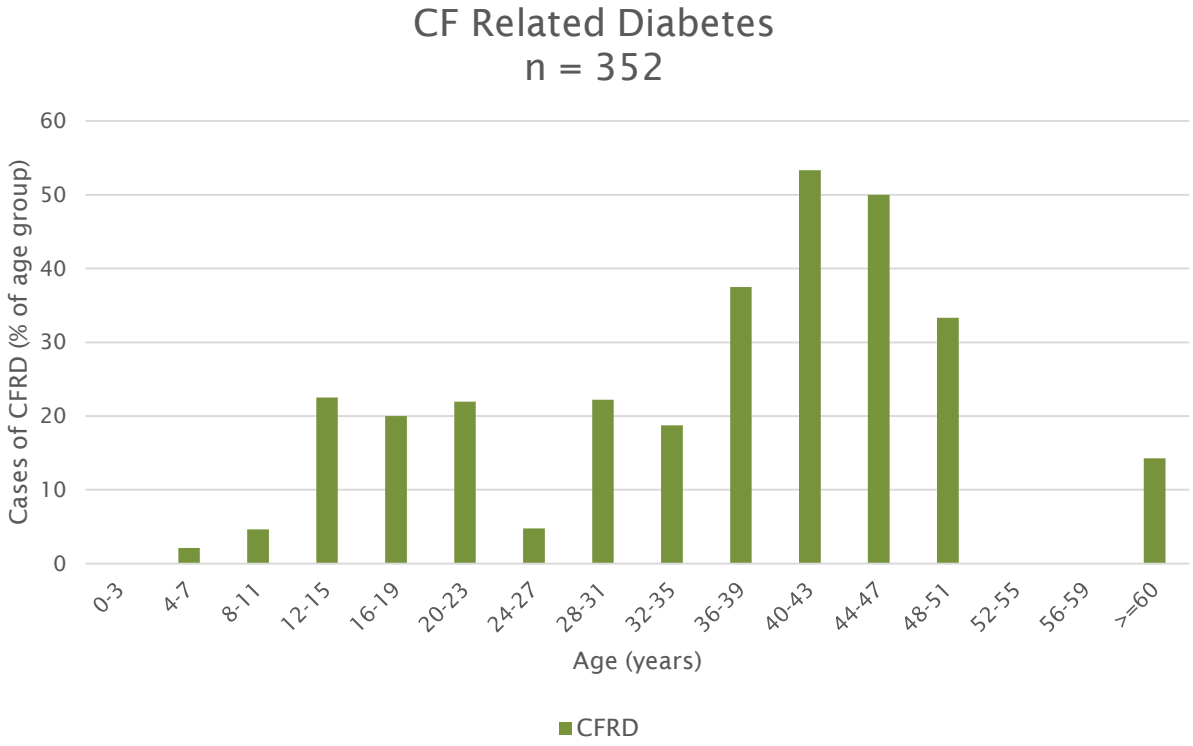


Data collected in 166 <16 years, 186 >16 years; Some patients may use more than one technique

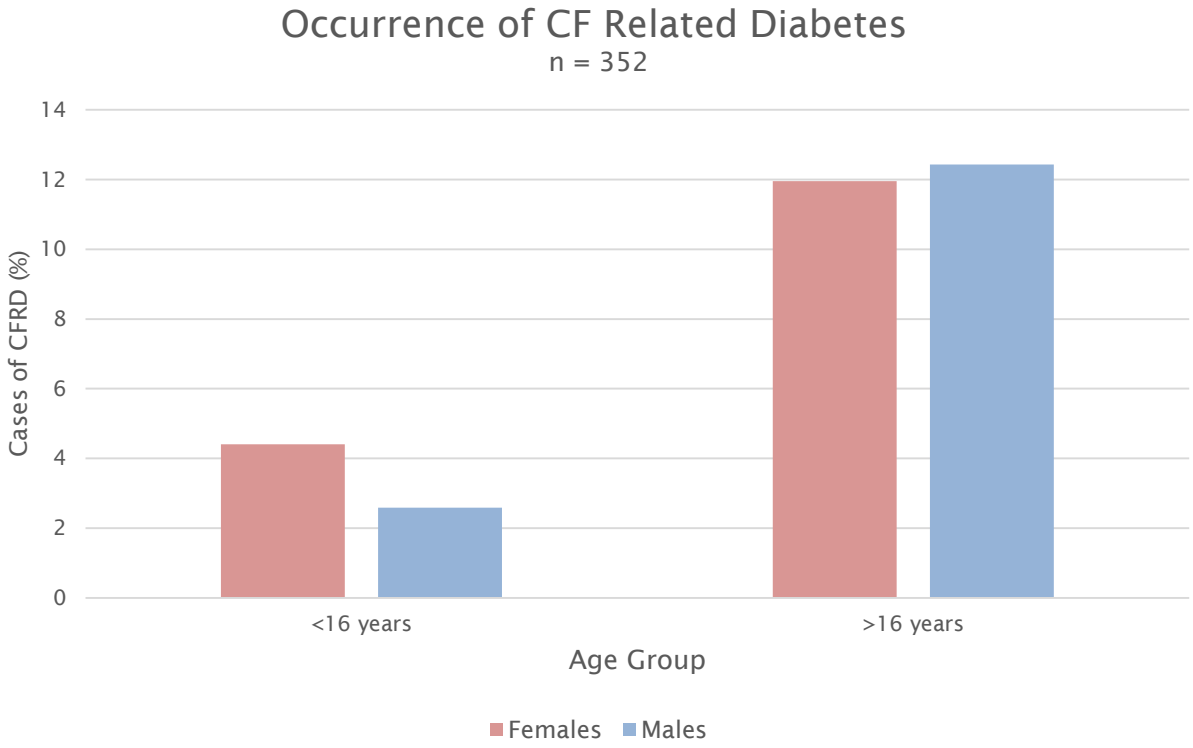
	<16 years		>16 years	
Technique	n	%	n	%
Exercise	83	71.6	40	72.7
Positive Expiratory Pressure	10	8.6	2	3.6
Oscillating PEP (eg: Flutter, Acapella, IPV)	4	3.4	3	5.5
Forced Expiration Techniques (eg:huff cough, active cycle breathing, autogenic drainage)	5	4.3	6	10.9
High Frequency Chest Wall Compression (eg: vest)	6	5.2	1	1.8
Modified Postural Drainage	8	6.9	3	5.5
Total	116		55	

Exercise remains a strong component of airway clearance – it is known to be widely beneficial and likely more fun that some other options!

CF-Related Diabetes



Age (years)	Age n	CFRD n	% Age Group	%CF Population
0-3	36	0	0.0	0.0
4-7	47	1	2.1	0.3
8-11	43	2	4.7	0.6
12-15	40	9	22.5	2.6
16-19	35	7	20.0	2.0
20-23	41	9	22.0	2.6
24-27	21	1	4.8	0.3
28-31	27	6	22.2	1.7
32-35	16	3	18.8	0.9
36-39	8	3	37.5	0.9
40-43	15	8	53.3	2.3
44-47	6	3	50.0	0.9
48-51	6	2	33.3	0.6
52-55	1	0	0.0	0.0
56-59	3	0	0.0	0.0
>=60	7	1	14.3	0.3
	352	55		15.6



	n	CFRD n	%	<16 years	%	>16 years	%
Females	159	26	16.4	7	4.4	19	11.9
Males	193	29	15.0	5	2.6	24	12.4
Total	352	55	15.6	12	3.4	43	12.2

The overall percentage of persons with CF affected by CFRD is similar to other reports, but the younger age group seems less, raising a query as to whether we are screening or acting on screening results early enough. The results have been similar for the last three years.

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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