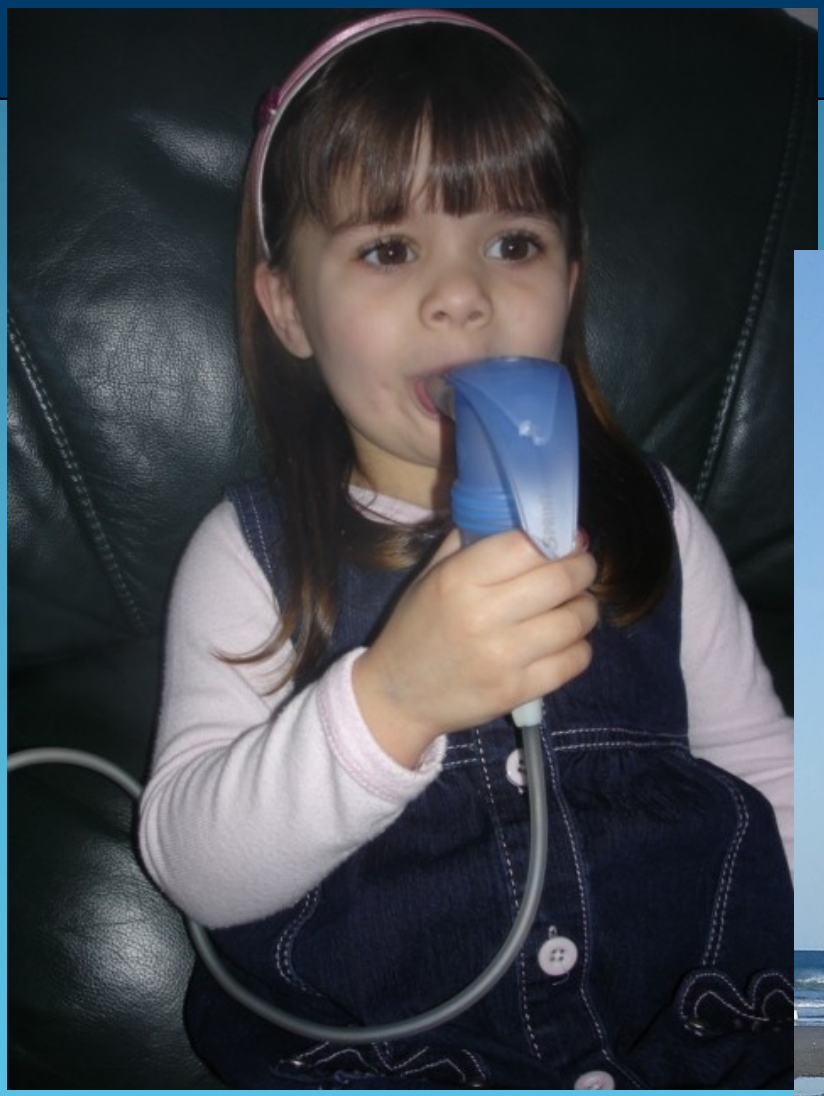
CYSTIC FIBROSIS
ASSOCIATION OF NEW ZEALAND

Port CFNZ
2011 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 was funded through conditional grants from

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Introduction and 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 2011 Report.

This first data presented using Port CFNZ will allow us to assess where New Zealand sits alongside other countries in patient outcomes. It will give us an important tool to determine where there may be gaps in CF care, to set quality improvement goals and to further our efforts to gain access to new treatments.

This inaugural report and future data collection will show where we are now and we can determine where we want to be. This will be hugely valuable to all people with cystic fibrosis and those who care for them both at home and in the clinical setting.

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. We gratefully acknowledge the assistance of the USA CF Foundation and the UK CF Trust in allowing us to use the Port CF software and providing training, mentoring, funding and advice.

Above all, thank you to the patients and their families for participating in this process – this registry should be seen as data for you not on you to aid improvement of national care.

Dr Cass Byrnes
Chair Port CFNZ Steering Committee

Dr Richard Laing
Port CFNZ Principal Investigator

Port CFNZ Steering Committee

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Dr Mark O'Carroll	Auckland Hospital and Greenlane Centre

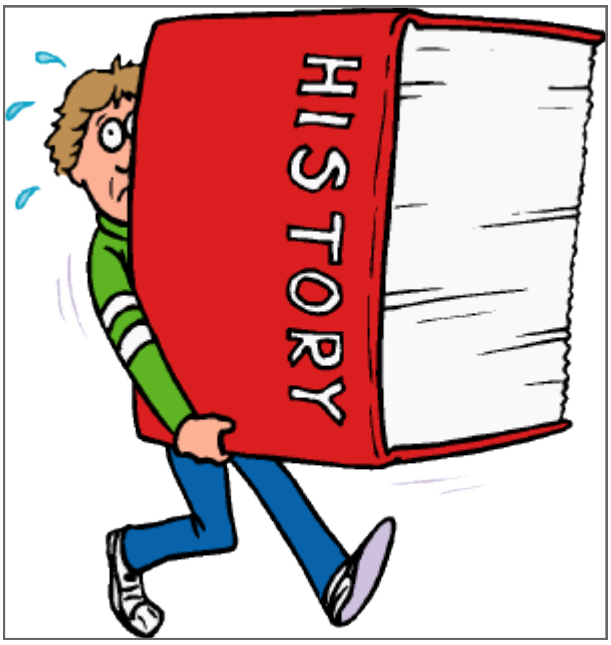
History of the Registry

Prior to 2003, New Zealand had combined its data with Australia for the Australasian Data Registry. This had been a largely manual process and as such there were serious time delays from the gathering of the data and the production of publicly available statistics.

Therefore in 2005, the Cystic Fibrosis Association of New Zealand decided to begin investigations on having their own, web-based database system, designed specifically for the New Zealand situation. In the ensuing twelve months, a number of options were explored, all of which would cost tens of thousands of dollars.

When investigating what other countries were using, the Port CF database in use in the USA and recently taken on by the UK Trust was favoured as a good model for us to pursue.

A visit to the States by CFANZ Chief Executive resulted in us being offered the use of Port CF free of charge in 2006 and by the end of 2009, when the UK had finished work on anglicising the database measurement systems, NZ was given their own copy to put into action.



In late 2010 the Association began a pilot with the Christchurch Paediatric and Adult clinics to iron out any issues and figure out what parts of the system needed further work to fit the NZ situation.

Today—we have most clinics providing us with full data with the others coming on stream as local resourcing and time allows.

This report and the ones that come after, will provide us with important data that shows us the areas we excel in and most importantly, those we need to work on to bring us up to international standards.

The power of this data to facilitate improvement in clinical care and understanding about the progression of the disease in this country cannot be overstated. This database gives us an opportunity we have not had before, to readily assess areas for new goal setting and improvement that involves not only the clinical care teams, but improvements in self-management and care in the home for patients themselves and their families.

Kua t whiti ki to t tou haerenga, ki te kore e haere tonu
He tino nui rawa a t tou mahi, kia kore e mahi nui tonu

We have come too far, not to go further
We have done too much, not to do more

CF Clinics in NZ

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)
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

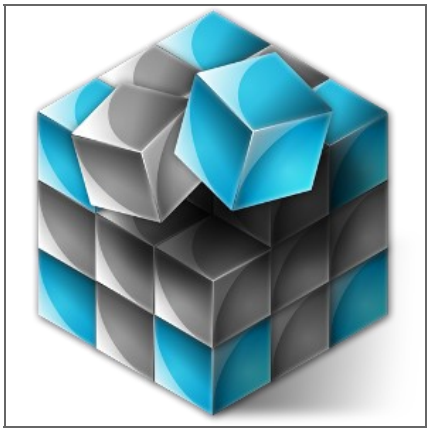
Nelson/ Marlborough (Paediatrics and Adults)
Nelson Hospital, Nelson
Wairau Hospital, Blenheim

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



Establishing a database such as this has not been without its hurdles. In particular we appreciate the time and effort needed to load individual clinical reviews into the system. In reviewing the data, it is clear that there are some questions that we are interpreting differently, some areas that we need to look into and define more clearly.

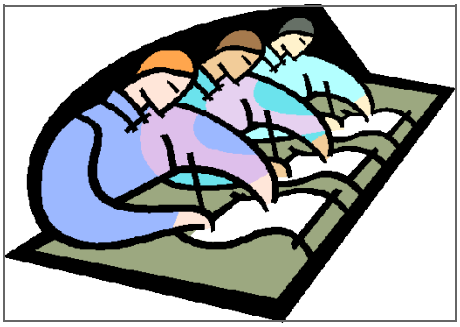
Some of the variability seen in the graphs across ages reflects our smaller numbers within any age group compared to larger population registries. It is also important to note that the 'N' number varies across the data, due to variance in the reporting from some centres that did not report across all fields in this first year. In particular, we know that a statistically significant number of adults did not have their data entered for this first registry.

The 2012 Data set will include statistics that have not been reported on for 2011, as we are still mapping what we wish to capture about such things as transplant, ethnicity etc. You will also note that we have not attempted to provide a median survival age. We will be unable to provide a statistic for New Zealand until 2015 at which time we will provide a '5-year, rolling average' only. This is due to our small population size and relatively low death rates, both of which contribute to the calculation of this statistic. That is to say, our numbers are too low to provide a meaningful statistic from one year alone.

We have made a decision to produce national statistics only. As a nation, New Zealand only 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 2013 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.

Key Results at a Glance 2011



CF patients registered	415
Diagnosis age <1 year	11
Age in years; median	15.71
PWCF aged >16yrs	206 *49.6%*
Males	226 *54.6%*
Genotyped	364 *87.7%*
Overall Median FEV_1 (% predicted) <16 years >16 years	80.5% 91.6% 70.7%

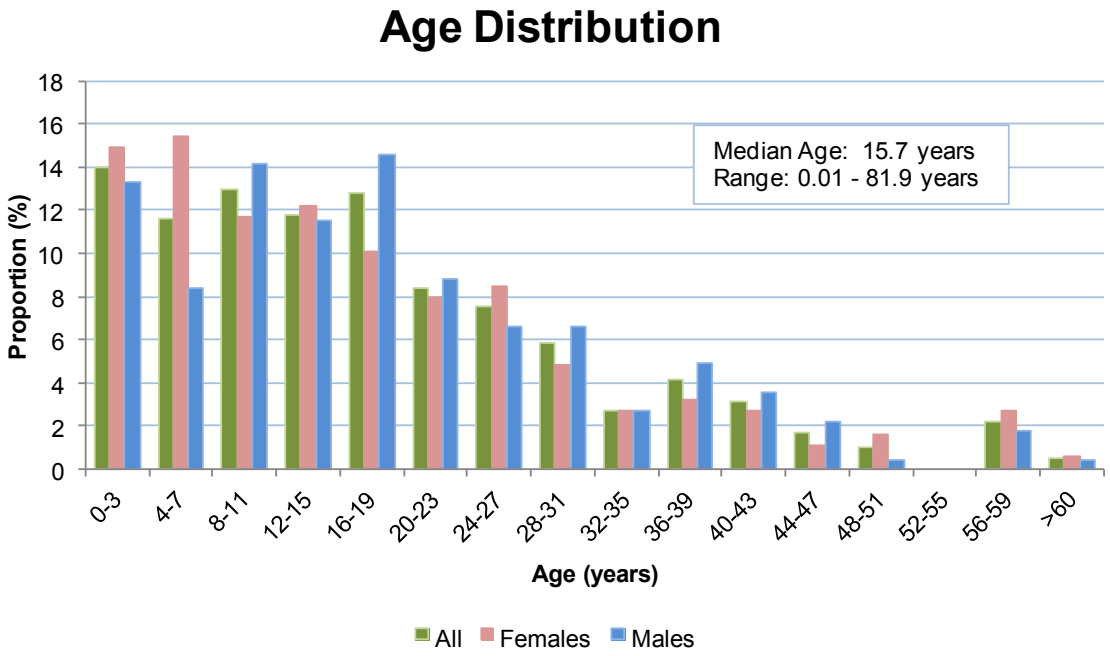
Commentary

We believe that we have captured approximately 85% of PWCF in NZ. This is excellent for the first report of a national database and far greater than the percentage captured when other country's registries started out.

The UK 2010 registry report that they have 85% after 5 years of having their similar database up and running.

We have not entered total deaths in the year on this table as reported deaths in the registry vary greatly from the number we know to be actual for the year based on CFANZ records. We will report on this from 2012 onwards.

Demographics

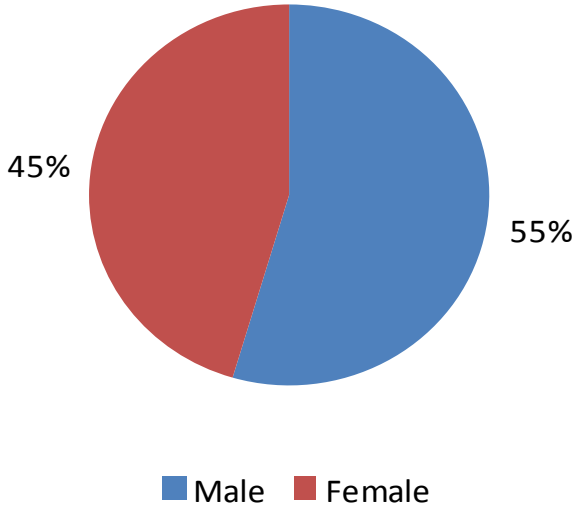


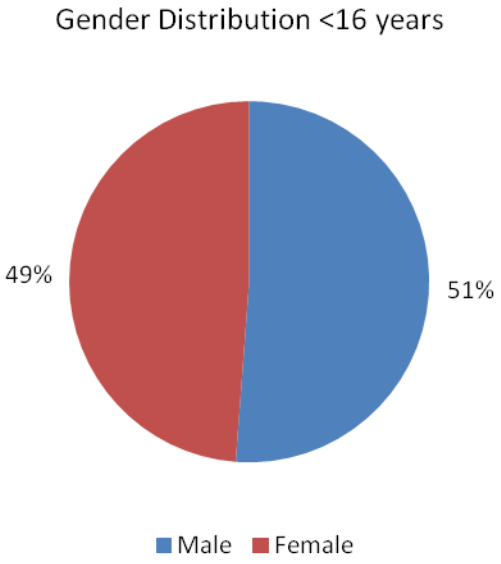
Age (years)	All	%	Female	%	Male	%
0-3	58	*14.0*	28	*14.9*	30	*13.3*
4-7	48	*11.6*	29	*15.4*	19	*8.4*
8-11	54	*13.0*	22	*11.7*	32	*14.2*
12-15	49	*11.8*	23	*12.2*	26	*11.5*
16-19	53	*12.8*	19	*10.1*	33	*14.6*
20-23	35	*8.4*	15	*8.0*	20	*8.8*
24-27	31	*7.5*	16	*8.5*	15	*6.6*
28-31	24	*5.8*	9	*4.8*	15	*6.6*
32-35	11	*2.7*	5	*2.7*	6	*2.7*
36-39	17	*4.1*	6	*3.2*	11	*4.9*
40-43	13	*3.1*	5	*2.7*	8	*3.5*
44-47	7	*1.7*	2	*1.1*	5	*2.2*
48-51	4	*1.0*	3	*1.6*	1	*0.4*
52-55				*0.0*		*0.0*
56-59	9	*2.2*	5	*2.7*	4	*1.8*
>60	2	*0.5*	1	*0.5*	1	*0.4*
	n = 415					
Median	15.71 years					
Range	0.01 - 81.9 years					

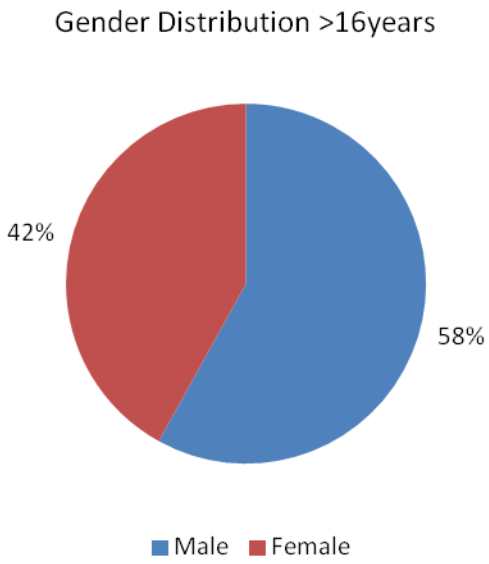
Commentary

The median age of 15.7 years should not be confused with median age of survival, which we cannot report until we are able to take a rolling average from 2015 onwards. This data is also reflective of the fact that not all adults have had their data entered for the 2011 year. Age is taken at December 31, 2011. Our current median age is 15.7 years – the median age in the USA 2011 registry is 17.5 years, in UK 2010 registry 17 years and Australian 2010 registry is 17.6 years. Ours at 15.7 years is close to, but lower, than the others. It may reflect that not all our adults with CF were entered into this first data collection, but suggests we need to look at this again critically next year.

Gender Distribution







Gender	Overall		<16 years		>16 Years	
		%		*%*		*%*
Male	226	*54.6*	107	*51.2*	119	*58.0*
Female	188	*45.4*	102	*48.8*	86	*42.0*
	414					

Genotype

364 (87.7%) of 415 patients have been genotyped with a recorded value

F508del Mutations	n	*%*
Homozygous F508del	213	*58.5*
Heterozygous F508del	128	*35.2*
No F508del or both unidentified	23	*6.3*

All mutations identified

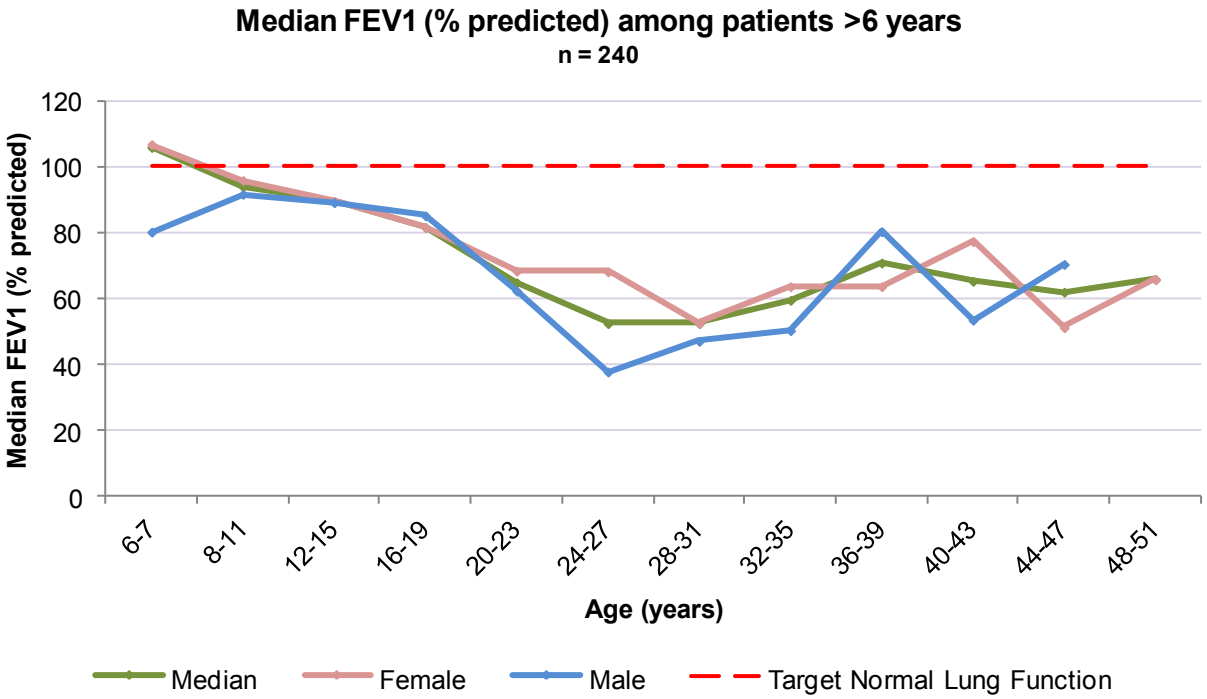
	n	*%*
F508del	554	*76.2*
G551D	24	*3.3*
G542X	24	*3.3*
R117H	17	*2.3*
3849+10kbC->T	6	*0.8*
G85E	5	*0.7*
3272-26A->G	5	*0.7*
N1303K	4	*0.5*
I507del	4	*0.5*
1898+1G->A	4	*0.5*
A455E	3	*0.4*
1717-1G->A	3	*0.4*
Y569D	3	*0.4*
Q493X	3	*0.4*
other	51	*7.0*
unknown	17	*2.3*

Commentary

It is increasingly important to ensure if possible that everyone with CF knows their own genotype.

In years to come and following on from recent findings, there is increasing research into specific treatment for specific genes such as Kalydeco® and therefore exact determination is important to ensure that treatments are targeted to specific mutation classes.

Respiratory



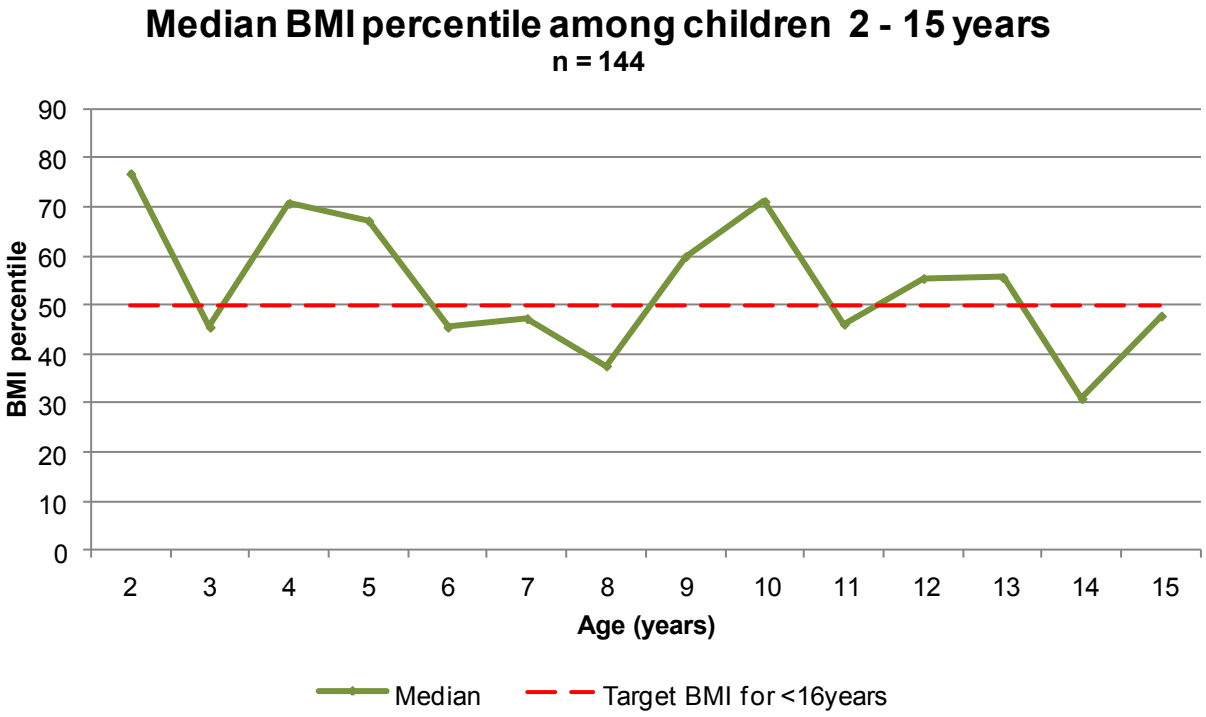
Age (years)	Female n	Female Median	Male n	Male Median
6-7	7	106.7	5	80.1
8-11	20	95.8	23	91.6
12-15	19	89.4	21	89.0
16-19	16	81.5	29	85.0
20-23	10	68.2	13	62.2
24-27	10	68.0	6	37.6
28-31	7	52.3	10	46.9
32-35	2	63.5	5	50.2
36-39	3	63.5	8	80.2
40-43	4	77.2	4	53.3
44-47	2	51.0	4	70.3
48-51	3	65.7		
52-55				
56-59	5	64.6	2	81.8
>60	1	56.9	1	127.2
	109		130	

Commentary

Our data is similar to other registries with loss of lung function most marked in the teenage and early adult years. The dotted line in this figure illustrates a target FEV% predicted. Anything near this indicates normal or near normal lung function values.

As a population our aim for good CF care is to preserve normal lung function for as long as possible for the paediatric population and maintaining stable lung function in adulthood. This is important for adults as lung function at 50% and above facilitates all the normal activities of daily living including attending work and study.

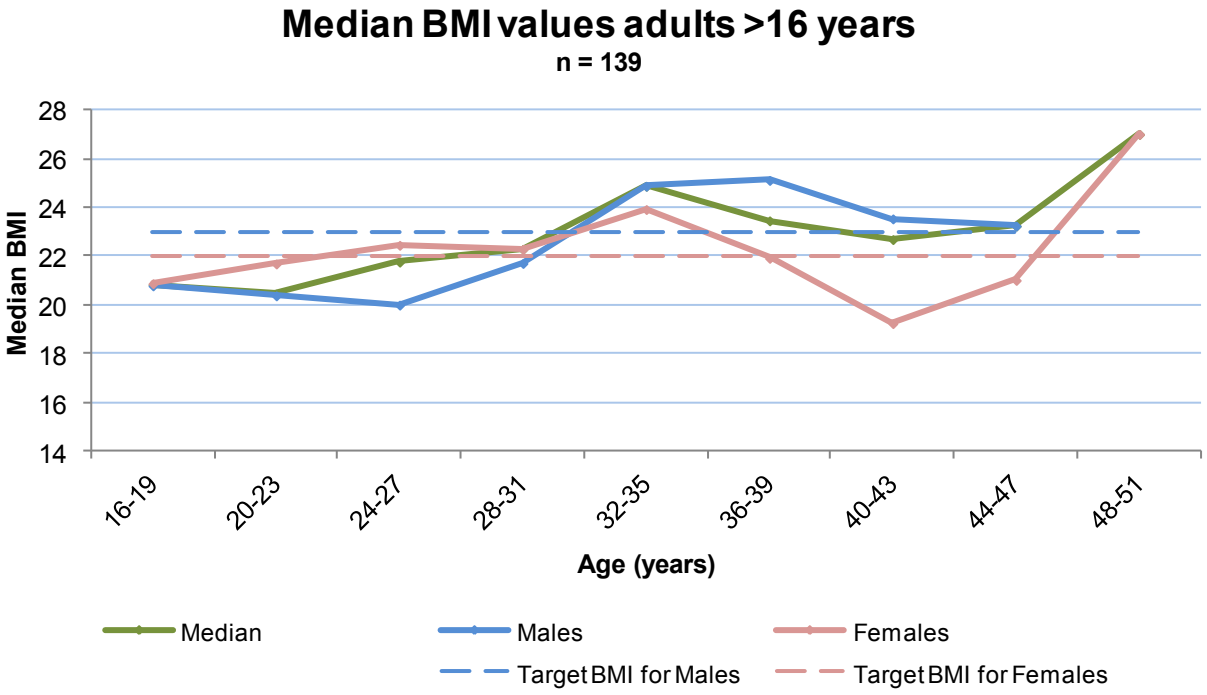
Nutrition



Age (years)	BMI Percentile n	Median
2	4	76.8
3	9	45.6
4	11	70.8
5	13	67.2
6	8	45.6
7	12	47.3
8	9	37.7
9	11	60.0
10	11	71.1
11	12	46.2
12	12	55.5
13	13	55.7
14	12	31.0
15	7	47.8
	144	

Commentary

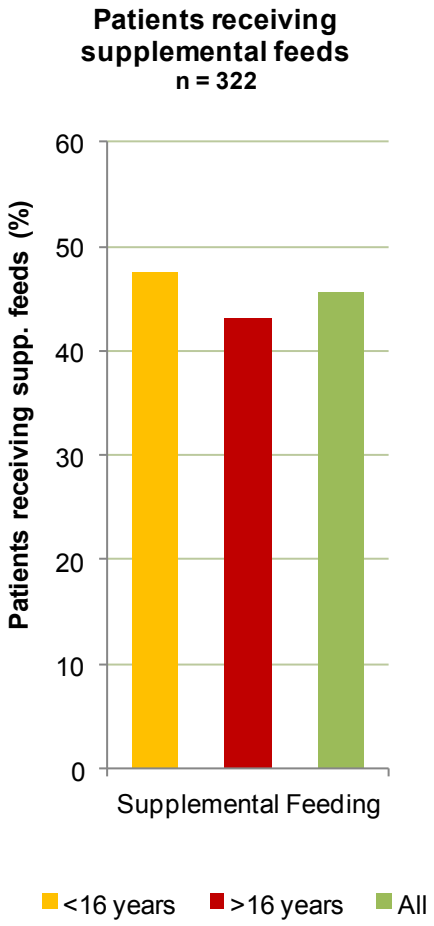
Compared to other registries our nutrition data is very encouraging. A BMI 50th Percentile or greater is recommended to assist better lung function status during childhood

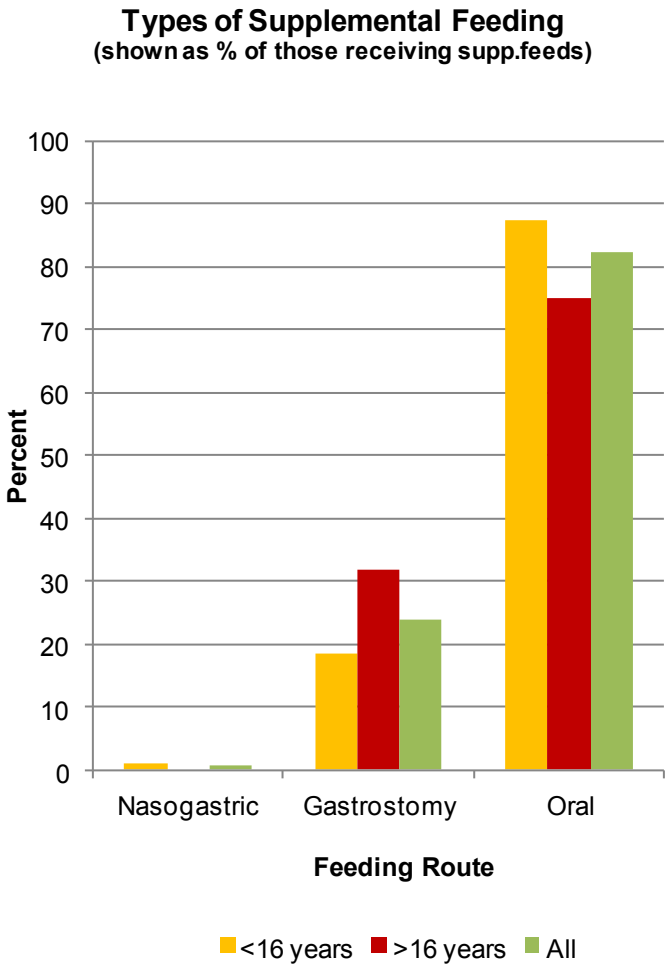


Age (years)	All n	All Median	Females n	Females Median	Males n	Males Median
16-19	39	20.8	15	20.9	24	20.8
20-23	23	20.4	10	21.7	13	20.4
24-27	16	21.8	10	22.4	6	20.0
28-31	17	22.3	7	22.3	10	21.7
32-35	7	24.9	2	23.9	5	24.9
36-39	11	23.5	3	21.9	8	25.1
40-43	8	22.7	4	19.2	4	23.5
44-47	6	23.2	2	21.0	4	23.2
48-51	3	27.0	3	27.0		
52-55						
56-59	7	27.3	5	27.0	2	29.1
>60	2	21.1	1	17.5	1	24.8
	139		62		77	

Commentary

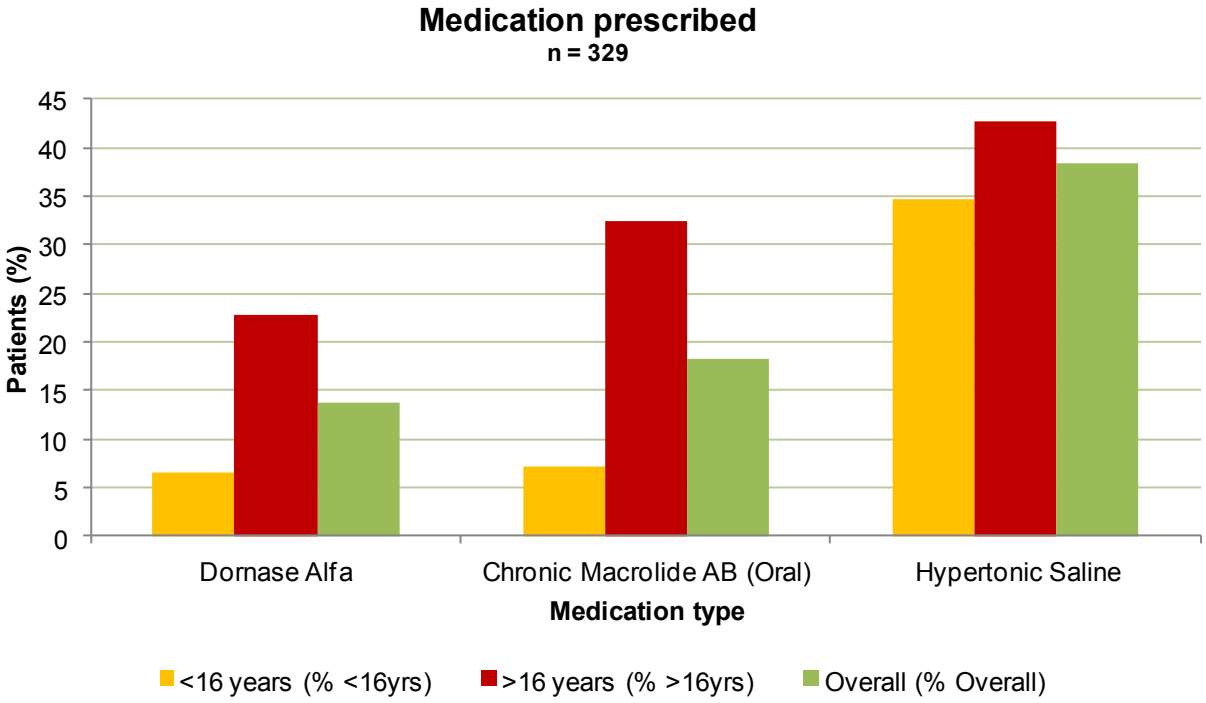
A BMI<20 at risk of malnutrition or in nutrition failure. Optimal BMI is 22 for females and 23 for males





	<16 years n = 182			>16 years n = 139			All n = 322		
	Yes	% n	% <16yrs supp.feed	*Yes*	% n	% >16yrs supp.feed	*Yes*	% n	% All supp.feed
Supp. Feeding	*87*	47.5		*60*	43.2		*147*	45.7	
Nasogastric	*1*	*0.5*	1.1	*0*	*0*	0.0	*1*	*0.3*	0.7
Gastrostomy	*16*	*8.7*	18.4	*19*	*13.7*	31.7	*35*	*10.9*	23.8
Oral	*76*	*41.5*	87.4	*45*	*32.4*	75.0	*121*	*37.6*	82.3

Medications

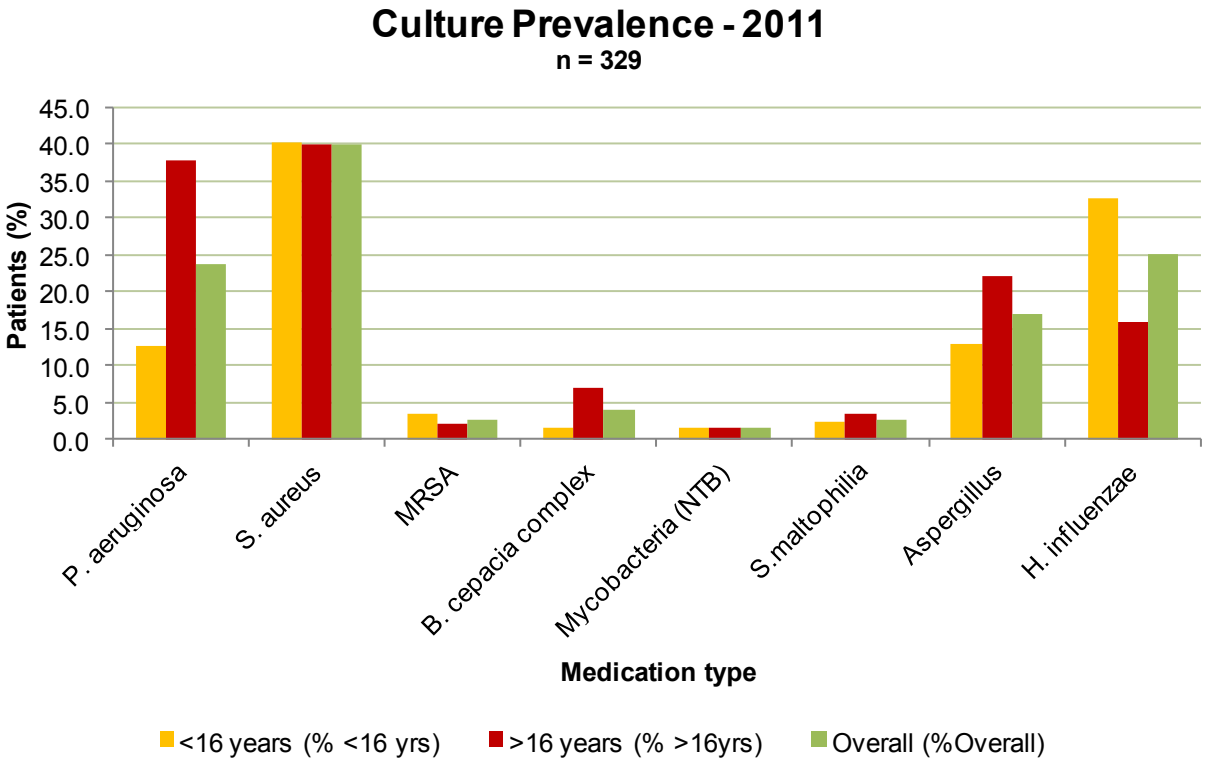


Medication	<16 years n = 184		>16 years n = 145		All n = 329	
	Yes	% n	*Yes*	% n	*Yes*	% n
Dornase Alfa	*12*	6.5	*33*	22.8	*45*	13.7
Chronic Macrolide AB (oral)	*13*	7.1	*47*	32.4	*60*	18.2
Hypertonic Saline	*64*	34.8	*62*	42.8	*126*	38.3

Commentary

Our levels of use for all three of these key medications is lower than is seen in the USA. With hypertonic saline now available in a pre-mixed solution, the relaxation of the criteria for Pulmozyme and easier access to Azithromycin, we are likely to see an increase in their use over time.

Microbiology



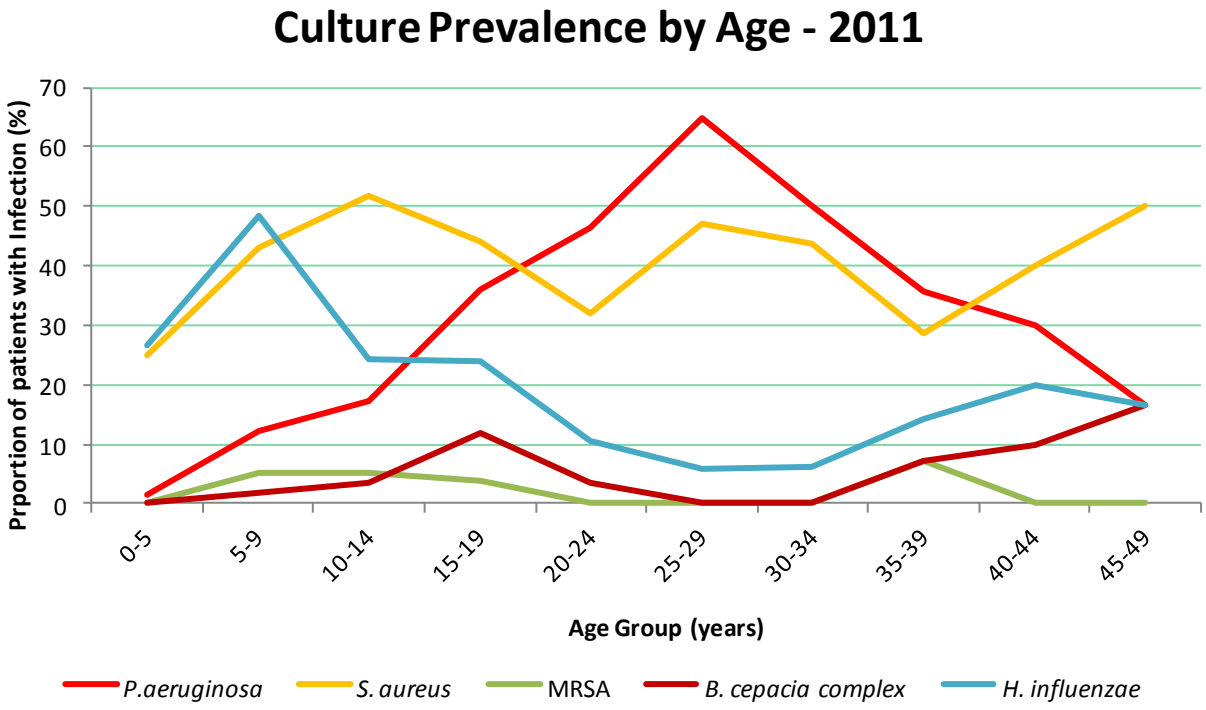
	<16 years		>16 years		All	
	n = 184		n = 145		n = 329	
	Yes	% n	*Yes*	% n	*Yes*	% n
P. aeruginosa	*23*	12.5	*55*	37.9	*78*	23.7
S. aureus	*74*	40.2	*58*	40.0	*132*	40.1
MRSA	*6*	3.3	*3*	2.1	*9*	2.7
B cepacia complex	*3*	1.6	*10*	6.9	*13*	4.0
Mycobacteria (NTB)	*3*	1.6	*2*	1.4	*5*	1.5
S. maltophilia	*4*	2.2	*5*	3.4	*9*	2.7
Aspergillus	*24*	13.0	*32*	22.1	*56*	17.0
H. influenzae	*60*	32.6	*23*	15.9	*83*	25.2

Commentary

Prevalence rates refer to any isolations of a specific organism in the 2011 year.

The culture rates of listed organisms are in keeping with that found in other countries.

Staphylococcus aureus rates have always been high in New Zealand for many respiratory and skin conditions, not just cystic fibrosis. As many of the individuals within this report have had just one annual review data entered, it is likely that the number of positive cultures over a whole year have been under-reported.



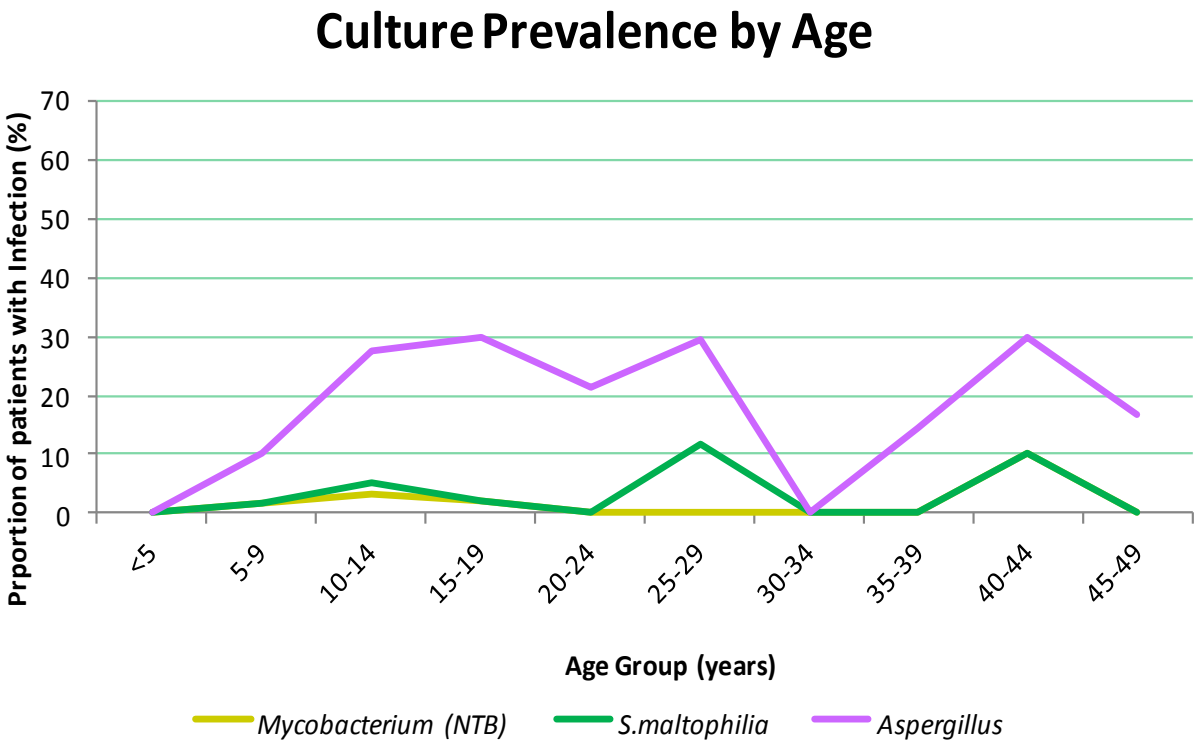
Age		*P.aeruginosa*		*S. aureus*		MRSA		*H. influenzae*		*B. cepacia complex*	
(years)	n	n	%	n	%	n	%	n	%	n	%
0-5	60	1	1.67	15	25.0	0	0.0	16	26.7	0	0.0
5-9	58	7	12.1	25	43.1	3	5.2	28	48.3	1	1.7
10-14	58	10	17.2	30	51.7	3	5.2	14	24.1	2	3.6
15-19	50	18	36.0	22	44.0	2	4.0	12	24.0	6	12.0
20-24	28	13	46.4	9	32.1	0	0.0	3	10.7	1	3.6
25-29	17	11	64.7	8	47.1	0	0.0	1	5.9	0	0.0
30-34	16	8	50.0	7	43.8	0	0.0	1	6.3	0	0.0
35-39	14	5	35.7	4	28.6	1	7.1	2	14.3	1	7.1
40-44	10	3	30.0	4	40.0	0	0.0	2	20.0	1	10.0
45-49	6	1	16.7	3	50.0	0	0.0	1	16.7	1	16.7
	317	77	24.3	127	40.1	9	2.8	80	25.2	13	4.1

Commentary

The variability in the graph reflects our fewer numbers of persons with CF at any age.

Overall, this pattern is similar to that seen overseas – with high early prevalence of *Staphylococcus aureus*, and increasing *Pseudomonas aeruginosa* with increasing age.

Of note at the current time is the data showing we have less MRSA than other countries.



Age		*Mycobacteria (NTB)*		*S. maltophilia*		*Aspergillus*	
(years)	n	n	%	n	%	n	%
0-5	60	0	0.0	0	0.0	0	0.0
5-9	58	1	1.7	1	1.7	6	10.3
10-14	58	2	3.4	3	5.2	16	27.6
15-19	50	1	2.0	1	2.0	15	30.0
20-24	28	0	0.0	0	0.0	6	31.4
25-29	17	0	0.0	2	11.8	5	29.4
30-34	16	0	0.0	0	0.0	0	0.0
35-39	14	0	0.0	0	0.0	2	14.3
40-44	10	1	10.0	1	10.0	3	30.0
45-49	6	0	0.0	0	0.0	1	16.7
	317	5	1.6	8	0.03	54	17.0

Commentary

This graph depicts numbers and presence of the less common organisms.

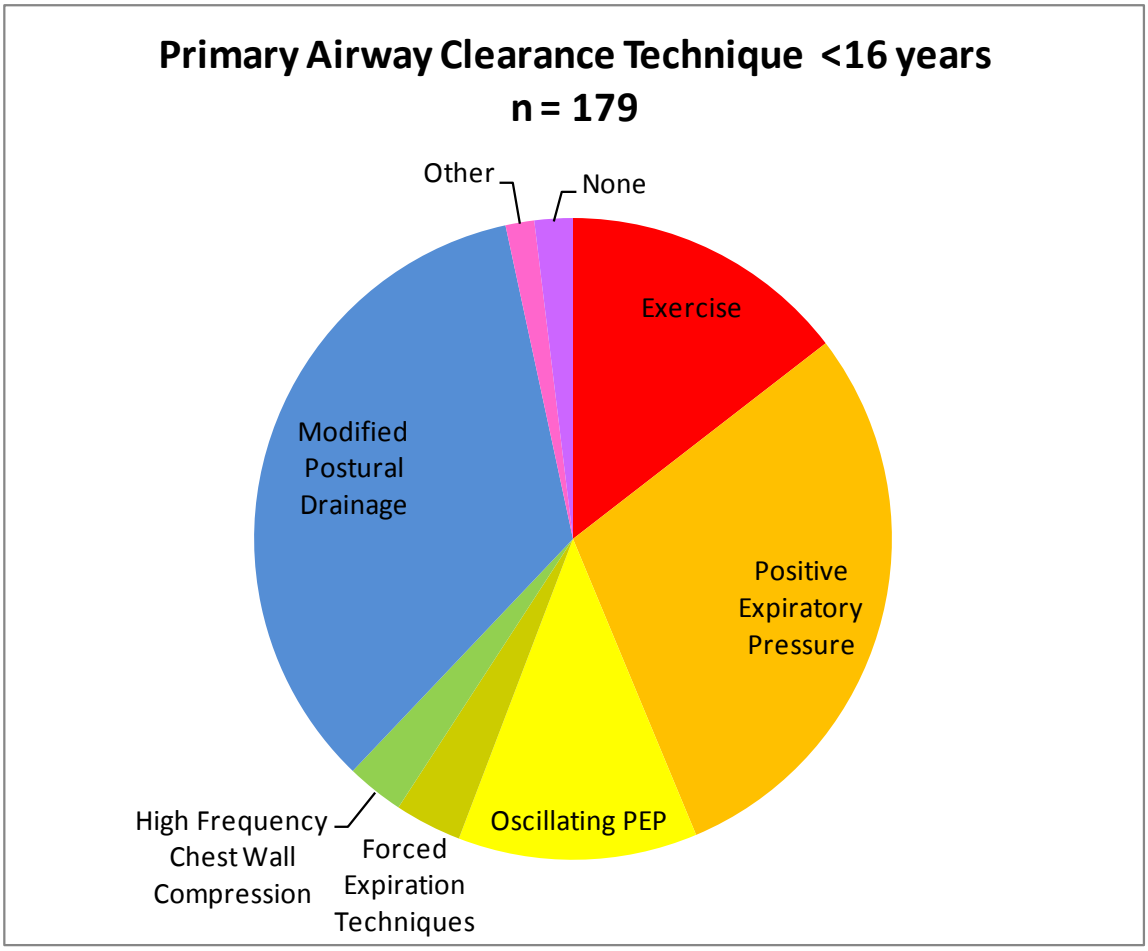
Hospital & Home IVA Days

AGE	Home IVA Days			Hospital IVA Days			Total IVA days
	n *(%)*	Total	Mean	n *(%)*	Total	Mean	
0-3	4 *(9.5)*	36	9	13 *(31.0)*	238	18	274
4-7	4 *(8.5)*	41	10	14 *(29.8)*	286	20	327
8-11	9 *(19.1)*	195	22	21 *(44.7)*	450	21	645
12-15	9 *(20.0)*	452	50	23 *(51.1)*	689	30	1141
16-19	9 *(19.6)*	155	17	20 *(43.5)*	562	28	717
20-23	7 *(31.8)*	147	21	12 *(54.5)*	445	37	592
24-27	8 *(44.4)*	199	25	13 *(72.2)*	336	26	535
28-31	5 *(27.8)*	74	15	7 *(38.9)*	135	19	209
32-35	2 *(28.6)*	23	12	2 *(28.6)*	19	9	42
36-39	5 *(45.5)*	51	10	5 *(45.5)*	145	29	196
40-43	2 (*22.2)*	26	13	3 *(33.3)*	65	22	91
44-47	2 *(28.6)*	44	22	3 *(42.9)*	47	16	91
48-51	0 *(0.0)*	0		2 *(66.7)*	32	16	32
52-55							
56-59	0 *(0.0)*	0		1 *(14.3)*	14	14	14
>60	1 *(50.0)*	10	10	1 *(50.0)*	2	2	12
		1453			3465		4918

Commentary

We have been unable to provide median or mean totals for the above data as currently there is no way within Port CF, to determine whether a course of IV's involved both a stay in hospital and time on Home IVs. This potentially means we are at risk of counting individuals twice giving an incorrect number . We hope to rectify this in 2013.

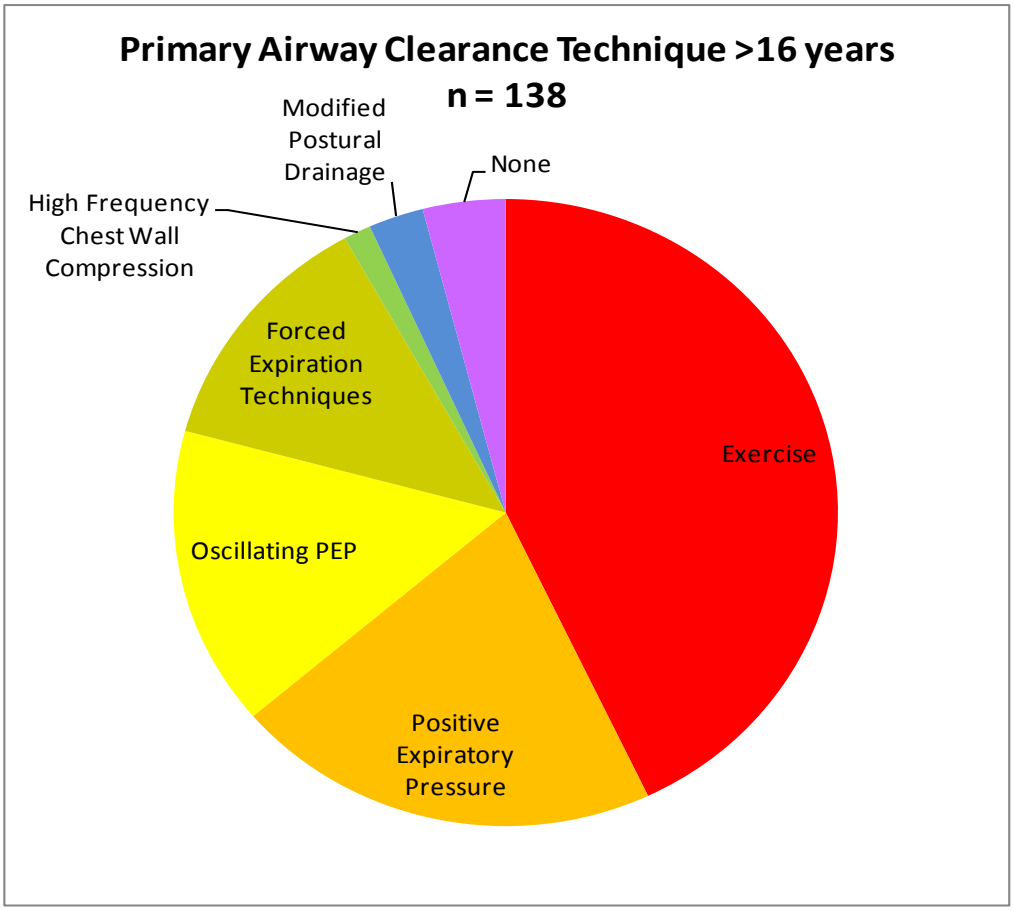
Primary Airway Clearance



Primary Airway Clearance Techniques - <16 years

*Number of individuals employing each technique at least once in the year.
Data collected on 179 patients

Technique	n	%
Exercise	30	16.8
Positive Expiratory Pressure	60	33.5
Oscillating PEP (eg: Flutter, Acapella, IPV)	25	14.0
Forced Expiration Techniques (eg: huff cough, active cycle breathing, autogenic drainage)	7	3.9
High Frequency Chest Wall Compression (eg: vest)	6	3.4
Mod. Postural Drainage	71	39.7
Other	3	1.7
None	4	2.2
	206	



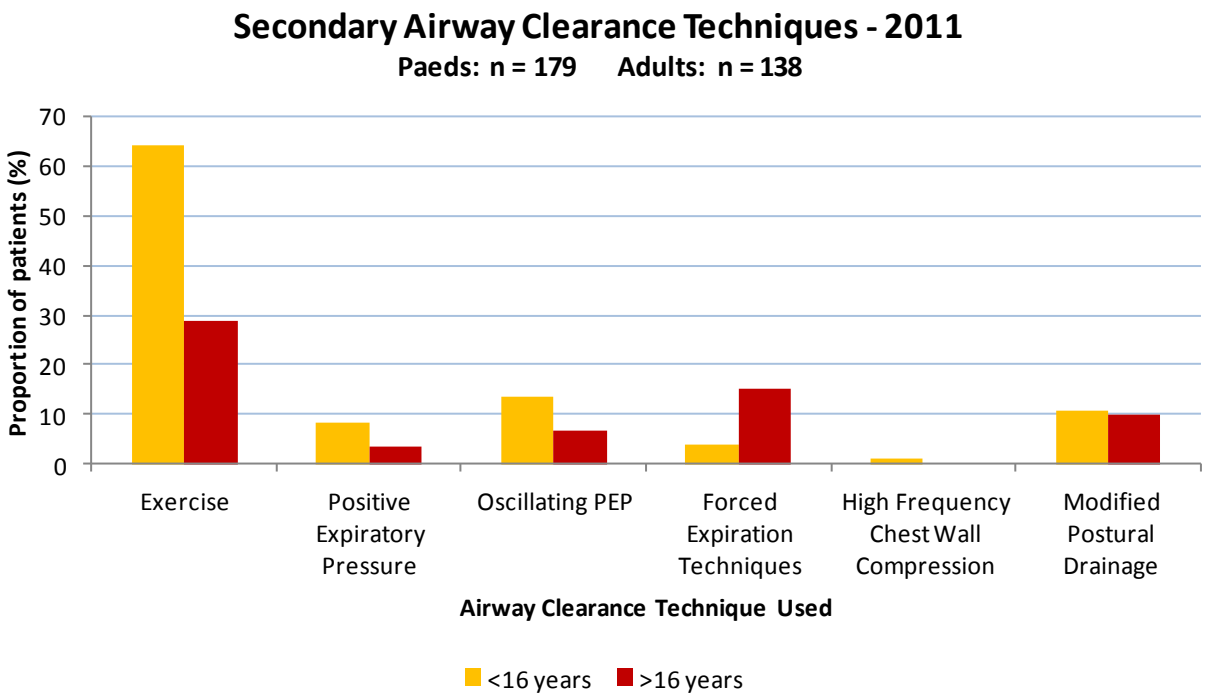
Primary Airway Clearance Techniques - >16 years

*Number of individuals employing each technique at least once in Data collected on 138 patients

Technique	n	%
Exercise	64	46.4
Positive Expiratory Pressure	31	22.5
Oscillating PEP (eg: Flutter, Acapella, IPV)	23	16.7
Forced Expiration Techniques (eg: huff cough, active cycle breathing, autogenic drainage)	19	13.8
High Frequency Chest Wall Compression (eg: vest)	2	1.5
Modified Postural Drainage	4	1.9
Other	0	0.0
None	6	4.4
	149	

Commentary

Physiotherapy techniques are difficult to compare worldwide as so many different techniques are used and the use of the 'VEST' in USA is high. While we are encouraged by how many appear to use 'exercise' as their 1st and/or 2nd line of airway clearance in both children and adults – it is hard to determine the actual implications of this as exercise means different things to different people. 16.8% of children and 46.4% of adults use it as their first line, and 64.3% of children and 29% of adults use it as their 2nd line airway clearance techniques. This compares to 4% in USA.

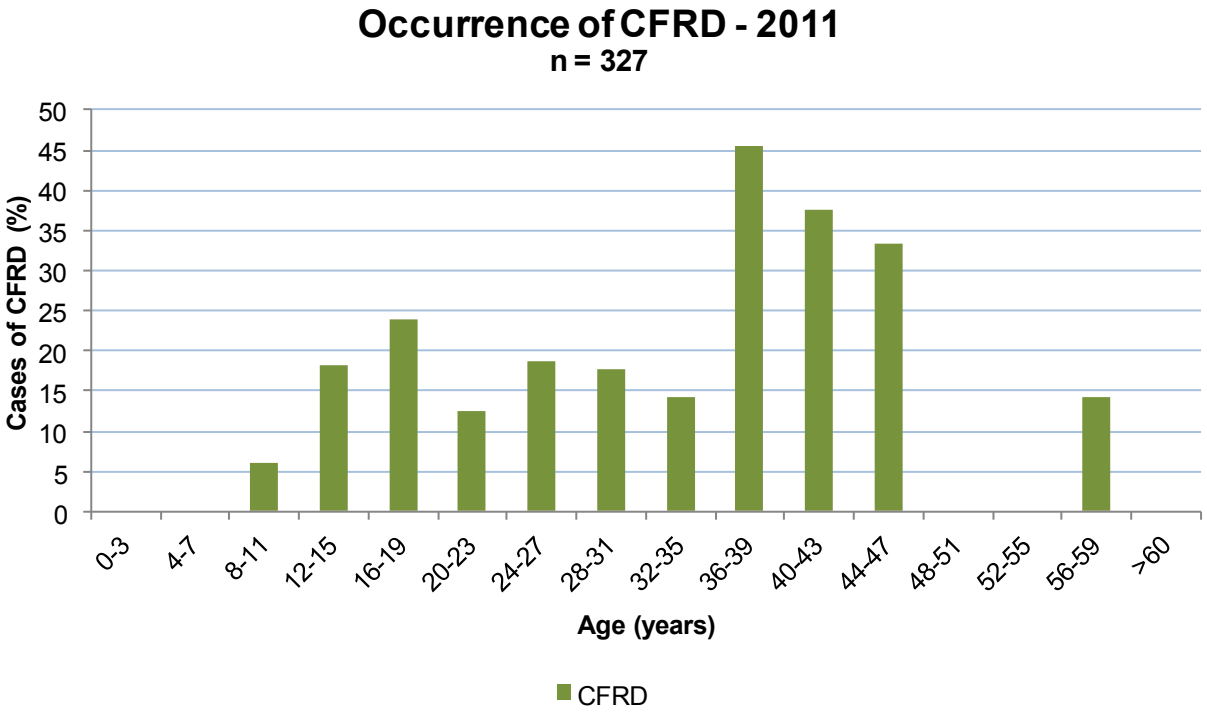


Secondary Airway Clearance Techniques

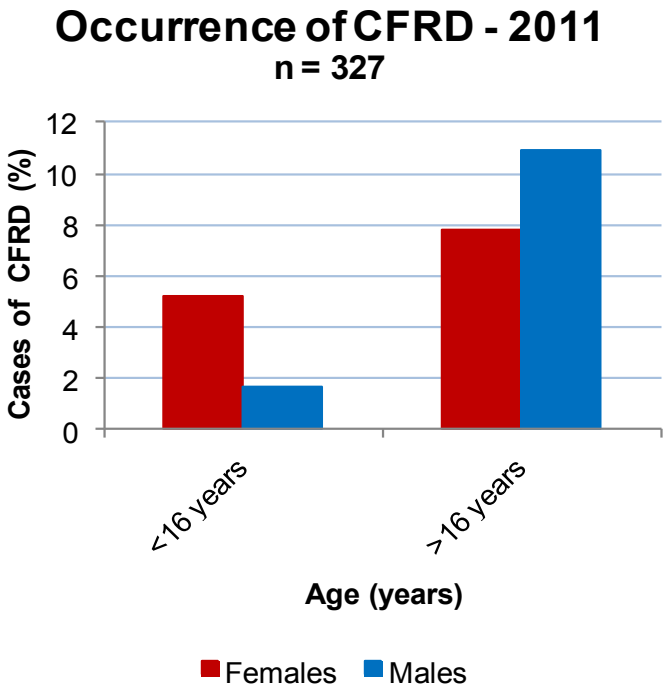
Data collected on 179 <16 years, 138 >16 years
Some patients may use mre than one technique

	<16 years		>16 years	
Technique	n	%	n	%
Exercise	115	64.3	40	29.0
Positive Expiratory Pressure	15	8.4	5	3.6
Oscillating PEP (eg: Flutter, Acapella, IPV)	24	13.4	9	6.5
Forced Expiration Techniques (eg: huff cough, active cycle breathing, autogenic drainage)	7	3.9	21	15.2
High Frequency Chest Wall Compression (eg: vest)	2	1.1	0	0.0
Modified Postural Drainage	19	10.6	10	7.3
	182		85	

CF_Related Diabetes



Age (years)	n	Occurrence of CF Related Diabetes CFRD	% age group	% CF population
0-3	43	0	0.0	0.0
4-7	48	0	0.0	0.0
8-11	49	3	6.1	0.9
12-15	44	8	18.2	2.4
16-19	42	10	23.8	3.1
20-23	24	3	12.5	0.9
24-27	16	3	18.8	0.9
28-31	17	3	17.6	0.9
32-35	7	1	14.3	0.3
36-39	11	5	45.5	1.5
40-43	8	3	37.5	0.9
44-47	6	2	33.3	0.6
48-51	3	0	0.0	0.0
52-55	0	0	0.0	0.0
56-59	7	1	14.3	0.3
>60	2	0	0.0	0.0
	n = 327	42		12.8



Occurrence of CF Related Diabetes

n = 327

				<16 years		>16 years	
	n	CFRD	%		%		%
Females	153	20	13.1	8	5.2	12	7.8
Males	174	22	12.6	3	1.7	19	10.9

Commentary

Our CF related diabetes rates are similar to overseas with the increasing prevalence noted in the early adolescent years.

Glossary of Terms

FEV1	Measurement of lung capacity as forced expired volume
BMI	Body Mass Index—measurement of weight relative to height
N	Total number of people in each dataset
Median	The middle number of a range of numbers
Range	The upper and lower values in each data set
Paediatric	0—16 years
Adult	Greater than 16 years
<	Less than
>	Greater than

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