CYSTIC FIBROSIS
ASSOCIATION OF NEW ZEALAND

Port CFNZ
2012 National Data Registry

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 was funded through a conditional grant from

Shares in Life Foundation
Putting hope into action

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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 2012 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 second registry from the Port CF database provides us with an 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 in the coming months will provide a database that is 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.

Dr Cass Byrnes
Chair Port CFNZ Steering Committee

Dr Richard Laing
Port CFNZ Principal Investigator

Port CFNZ Steering Committee

Dr Cass Byrnes (Chair)	Starship Children's Hospital & University of Auckland
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Dr Mark O'Carroll	Auckland Hospital and Greenlane Centre

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 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

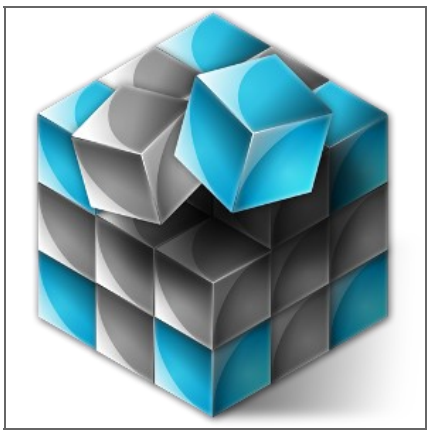
Nelson/ Marlborough (Paediatrics and Adults)
Nelson Hospital, Nelson
Wairau Hospital, Blenheim

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 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.

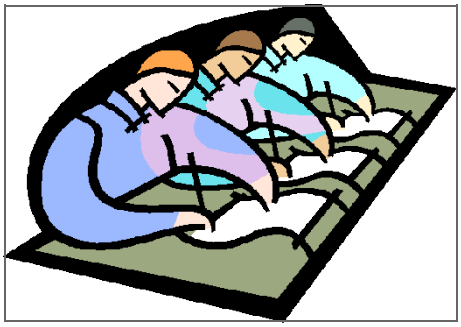
Our smaller population size provides significant challenges to our Statistician as the 'outliers' in terms of age and key markers will have a much larger impact on statistics than they would on a larger data set. Because of this, some decisions were made by the steering committee to exclude those outlier ages and statistics in order to give a more accurate picture of the overall patient outcomes for the country.

The brief commentary provided throughout this report reflects opinion based in this, our initial 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 – this will improve its use to our community and act as further checks on quality. A proposal for a project involving this national data base can be made in writing to the PORT CF steering committee.

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

Key Results at a Glance 2012



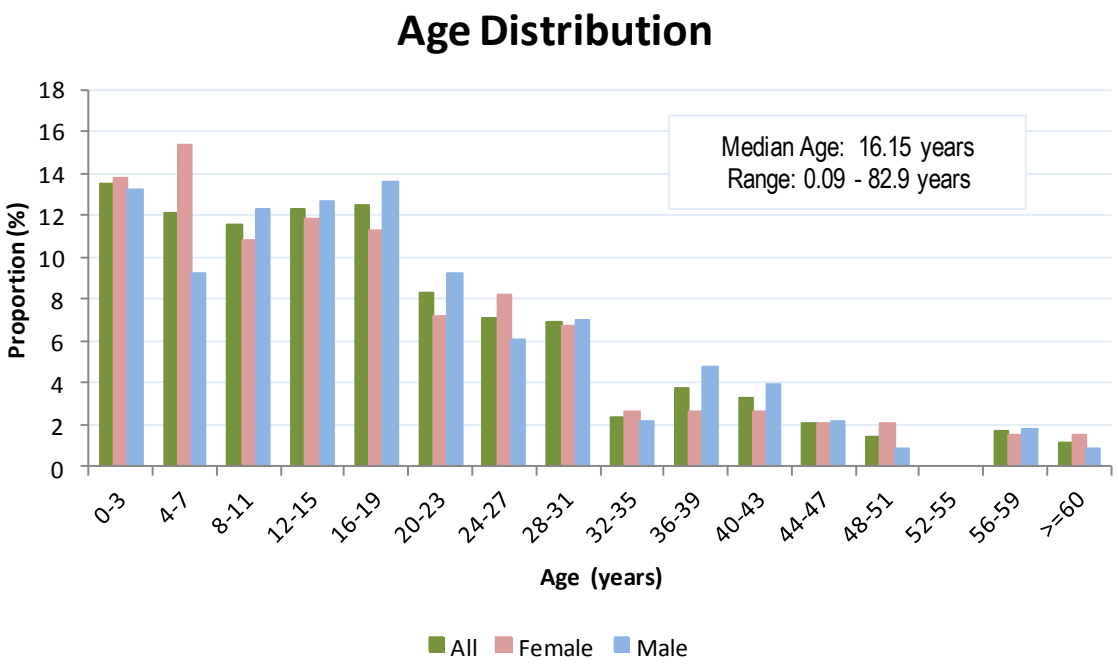
Key Indicators at a Glance

	2012	2011
CF patients registered	423	415
Diagnosis age <1 year	11	11
Age in years; median	16.15	15.71
PWCF aged >16yrs	214 *50.59%*	206 *49.6%*
Males	228 *53.9%*	226 *54.6%*
Genotyped	407 *96.2%*	364 *87.7%*
Median FEV_1 (% predicted) <16 years >16 years	84.5% 97.2% 70.6%	80.5% 91.6% 70.7%

It is encouraging to see the increase in numbers of people with CF participating and this has increased our median age and our percent predicted FEV1 in the younger population.

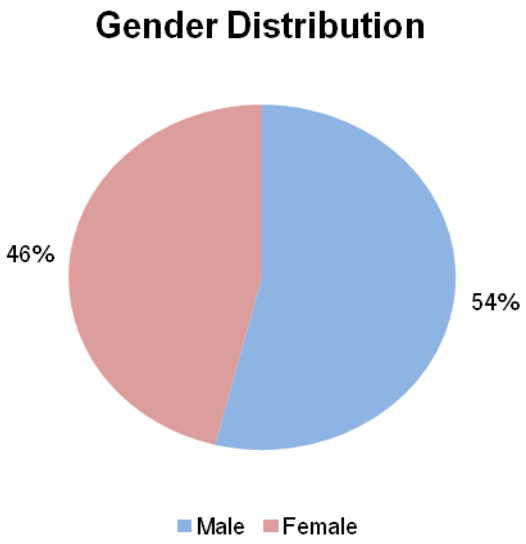
We cannot comment on life expectancy when having CF in NZ until we have at least 5 years of data at which stage we will be able to provide a rolling average.

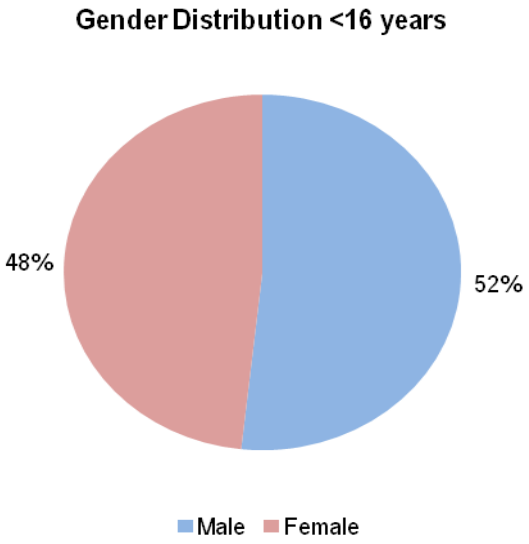
Demographics

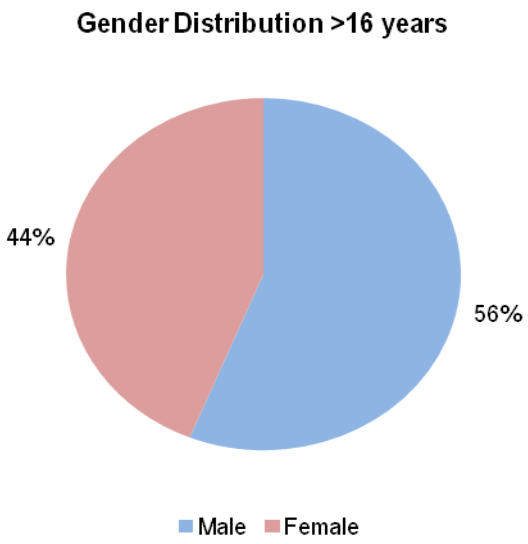


Age (yrs)	All		Male		Female	
	n	%	n	%	n	%
0-3	57	13.5	30	13.2	27	13.8
4-7	51	12.1	21	9.2	30	15.4
8-11	49	11.6	28	12.3	21	10.8
12-15	52	12.3	29	12.7	23	11.8
16-19	53	12.5	31	13.6	22	11.3
20-23	35	8.3	21	9.2	14	7.2
24-27	30	7.1	14	6.1	16	8.2
28-31	29	6.9	16	7.0	13	6.7
32-35	10	2.4	5	2.2	5	2.6
36-39	16	3.8	11	4.8	5	2.6
40-43	14	3.3	9	3.9	5	2.6
44-47	9	2.1	5	2.2	4	2.1
48-51	6	1.4	2	0.9	4	2.1
52-55	0	0.0	0	0.0	0	0.0
56-59	7	1.7	4	1.8	3	1.5
>=60	5	1.2	2	0.9	3	1.5
Total	423		228		195	
Median	16.15 years					
Range	0.09 - 82.9 years					

Our median age at 16.2 years has increased from last year which may reflect the greater capture of people within the database. It remains 12-18 months lower than the median age in other data registries such as Australia, UK and USA. Each year this comparison will become more accurate. It may be in future we will follow a trend to report age breakdowns in more appropriately grouped ages around development rather than strict 3 year cuts (e.g. 0-1 year, 2-5 years, 6-11 years, 12-17 years, 18-29 years, 30+ years)







Gender	All		<16 years		>16 years	
	n	%	n	%	n	%
Male	228	53.9	108	51.7	120	56.1
Female	195	46.1	101	48.3	94	43.9
Total	423		209		214	

Of note here is that the division of gender is very even in the younger age group but there are more males in the older age group.

This reflects the accelerated disease sometimes seen in women in late teenage and early adult years, a time we should be targeting with extra management.

Genotype

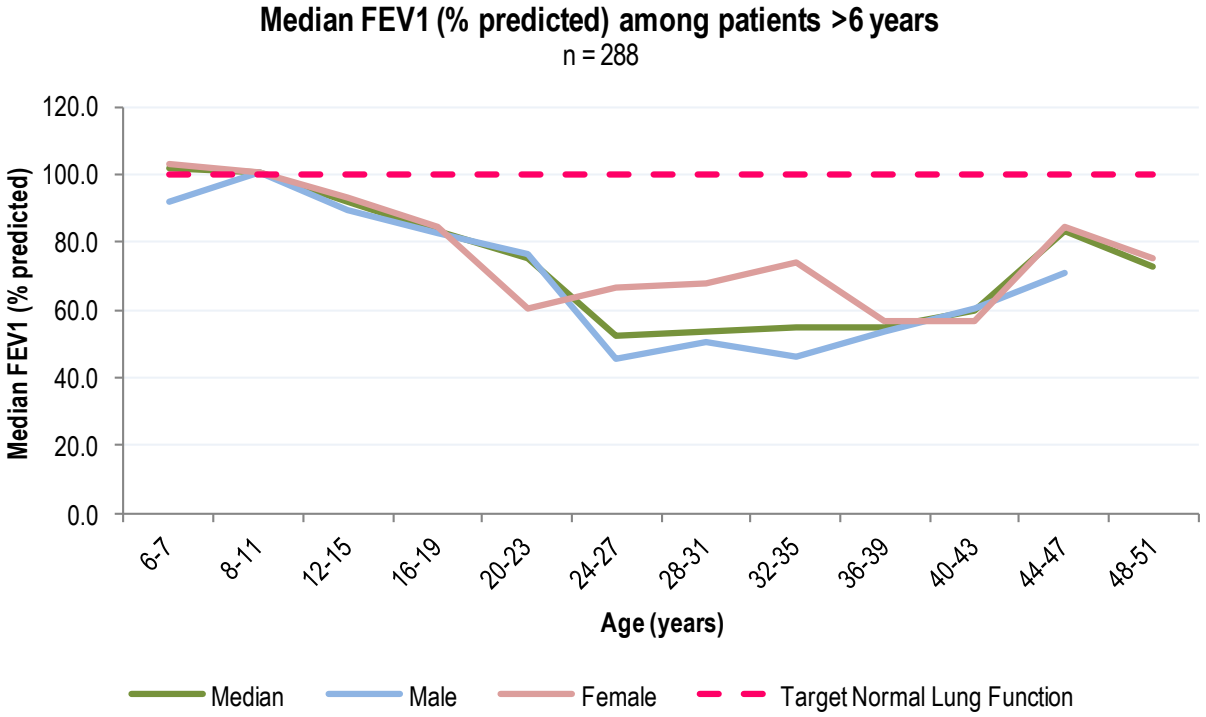
407 (96%) of 423 patients have been genotyped with a recorded value.

F508del Mutations	**n**	**%**
Homozygous F508del	215	52.8
Heterozygous F508del	154	37.8
No F508del or both unidentified	38	9.3
Total	407	

Mutations Identified	**n**	**%**
F508del	584	71.7
G551D	27	3.3
G542X	24	2.9
R117H	17	2.1
G85E	5	0.6
N1303K	5	0.6
3272-26A>G	5	0.6
I507del	4	0.5
3849+10kbC->T	4	0.5
Y563N	3	0.4
c.3718-2477C>T	3	0.4
1717-1G->A	3	0.4
1898+1G->A	3	0.4
Q493X	3	0.4
Other	86	10.6
Not Identified	38	4.7
Total	814	

With the recent development of medication specific to genotype, an area of increased research and likely further developments over the next years, it is important that all individuals know their genotype as far as possible.

Respiratory

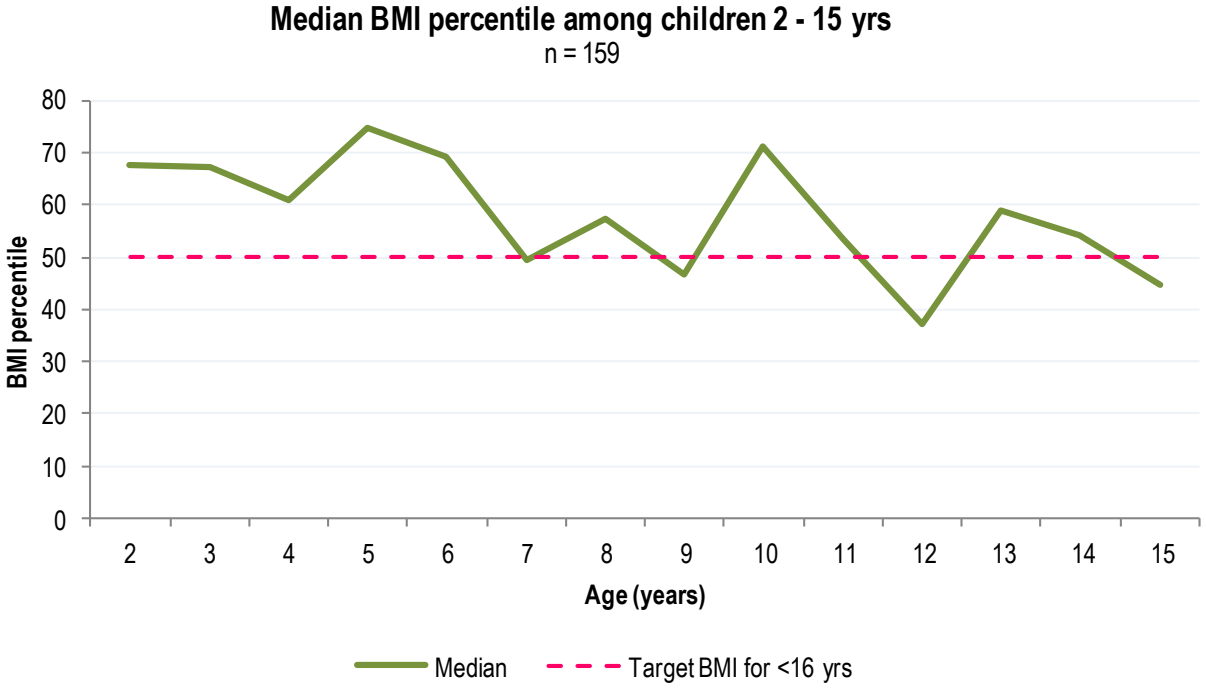


Age (yrs)	All		Male		Female	
	n	median	n	median	n	median
6-7	22	101.8	9	91.8	13	102.9
8-11	42	100.5	23	100.7	19	100.3
12-15	47	91.9	25	89.8	22	92.9
16-19	42	83.2	23	82.4	19	84.4
20-23	30	75.4	21	76.5	9	60.3
24-27	27	52.5	12	45.8	15	66.6
28-31	24	53.7	15	50.5	9	67.6
32-35	6	55.0	4	46.3	2	74.1
36-39	14	55.0	11	53.7	3	56.4
40-43	14	60.0	9	60.6	5	56.9
44-47	6	83.0	4	71.0	2	84.5
48-51	5	72.8	1		4	75.3
52-55	0		0		0	
56-59	4	74.7	2	81.2	2	63.1
>=60	5	81.8	2	89.2	3	81.8
Total	288		161		127	

The NZ trend lines for changing lung function with age is very similar to other national registries.

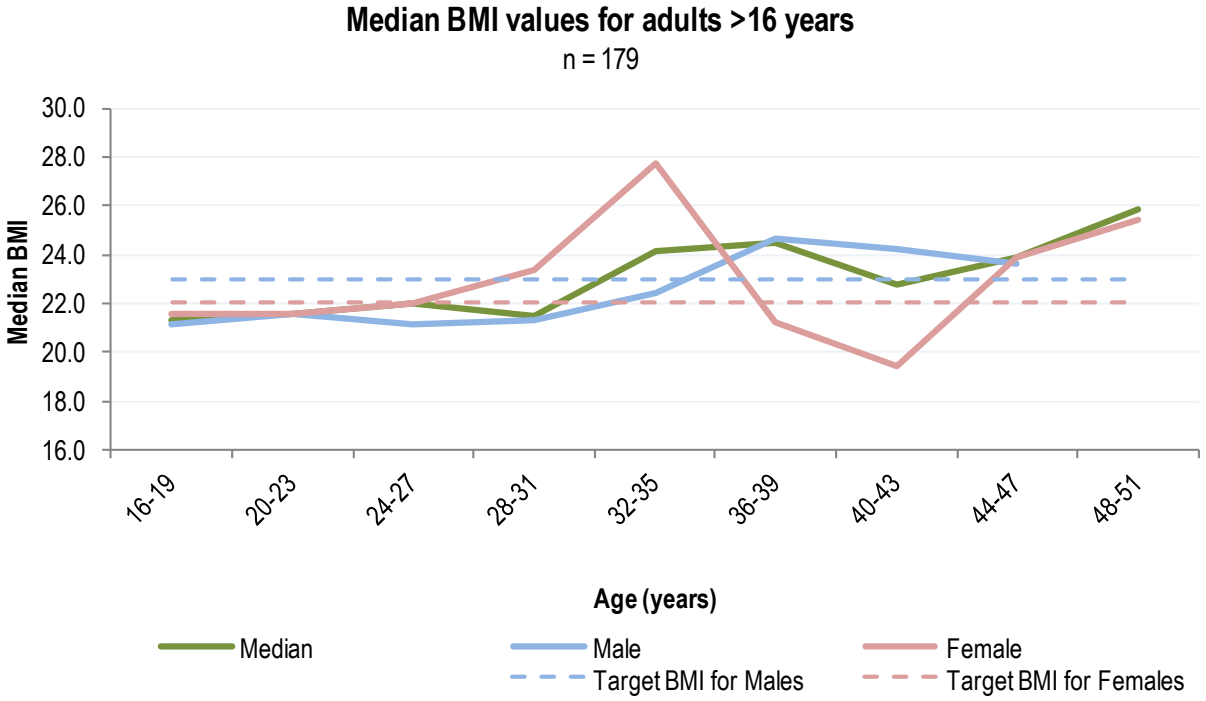
As noted last year, going into adulthood with an FEV1 equal or greater than 50% predicted facilitates normal activities of daily living such as work or study

Nutrition



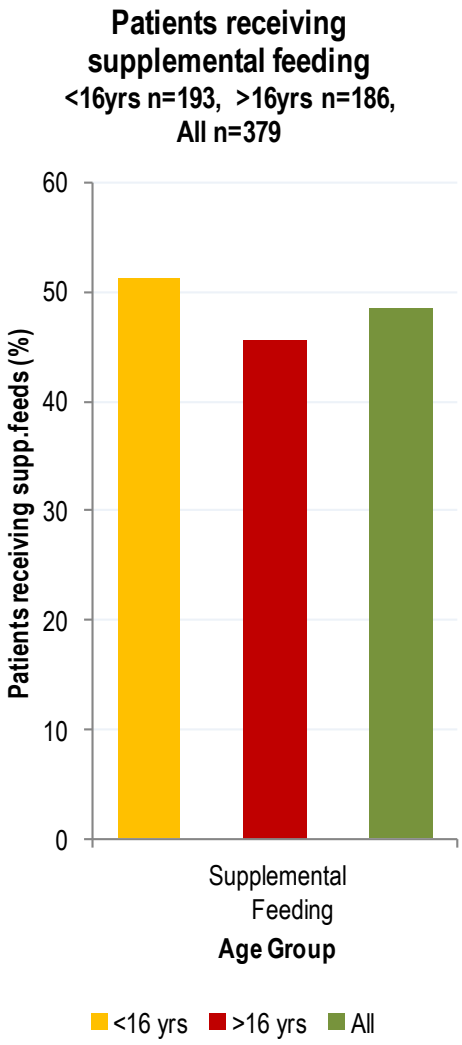
Age (yrs)	BMI Percentile	
	n	median
2	5	67.4
3	13	67.2
4	10	61.0
5	12	74.7
6	13	69.2
7	10	49.2
8	13	57.2
9	9	46.7
10	15	71.3
11	11	53.5
12	11	37.2
13	11	59.1
14	15	54.3
15	11	44.5
Total	159	

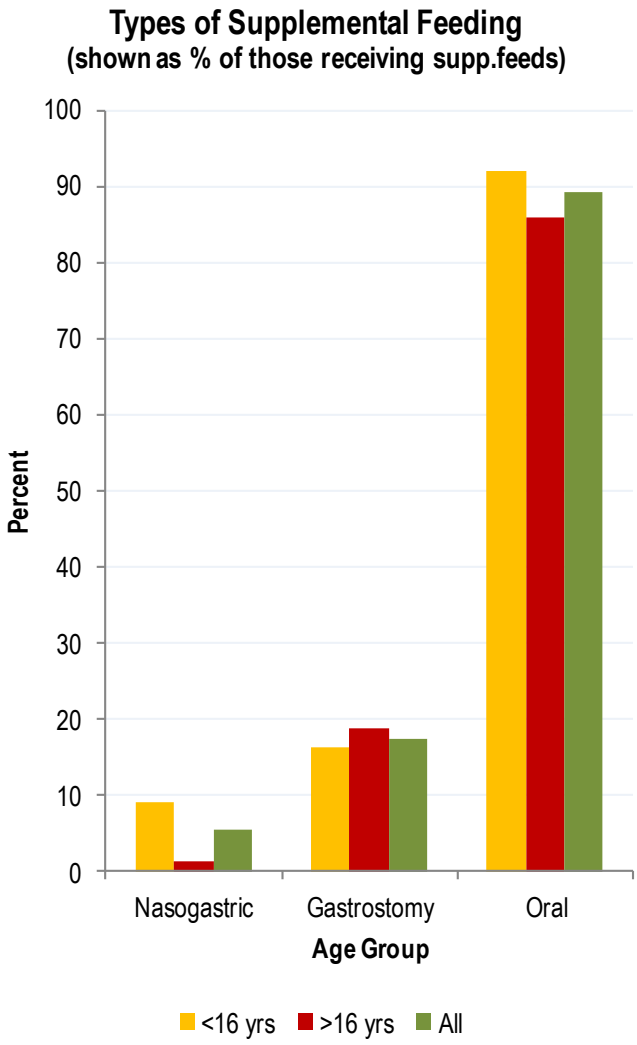
Our nutrition data is very encouraging and generally looks better than some of the comparative national registries – in part explained by early use of nutritional supplementation, newborn screening and early intervention strategies employed by the multidisciplinary teams. However, there is a falloff in late teenage years, important at the time of transition to adult services.



Age (yrs)	All		Female		Male	
	n	median	n	median	n	median
16-19	41	21.3	19	21.6	22	21.2
20-23	31	21.6	10	21.5	21	21.6
24-27	27	22.0	15	22.0	12	21.2
28-31	23	21.5	9	23.4	14	21.3
32-35	7	24.1	2	27.8	5	22.4
36-39	14	24.5	3	21.2	11	24.7
40-43	14	22.7	5	19.4	9	24.3
44-47	7	23.9	3	23.9	4	23.6
48-51	5	25.8	4	25.4	1	
52-55	0		0		0	
56-59	5	27.5	2	25.9	3	27.5
>=60	5	22.8	3	28.6	2	20.5
Total	179		75		104	

Similarly in adults, our nutrition data is encouraging in comparison to other registries; however note that the lines on the graph are the target BMI for males and females respectively and this is not always being reached.

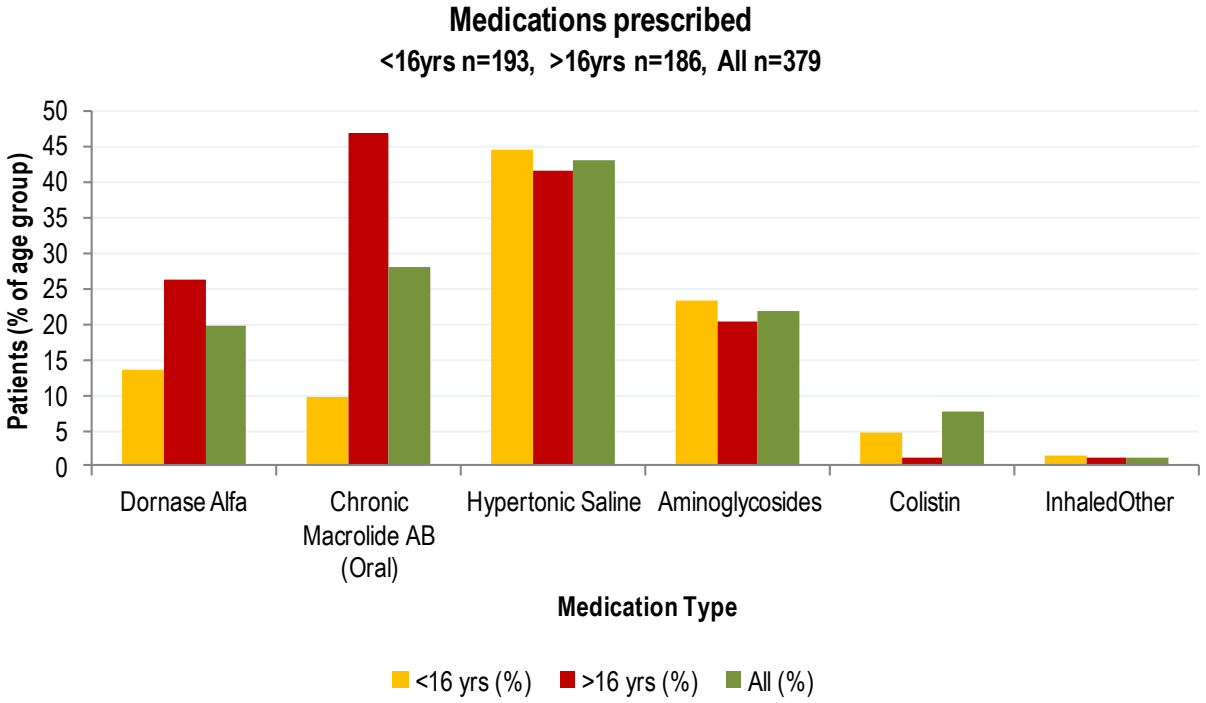




	<16 yrs, n = 193			>16 yrs, n=186			All, n = 379		
	Yes	%	% <16yrs	Yes	%	% >16 yrs	Yes	%	% All supp.
Supplemental Feeding	99	51.3		85	45.7		184	48.5	
Nasogastric	9	4.7	9.1	1	0.5	1.2	10	2.6	5.4
Gastrostomy	16	8.3	16.2	16	8.6	18.8	32	8.4	17.4
Oral	91	47.2	91.9	73	39.2	85.9	164	43.3	89.1

Note: some patients who are on enteral feeding are also on oral nutrition supplements which explains the apparent discrepancy in numbers.

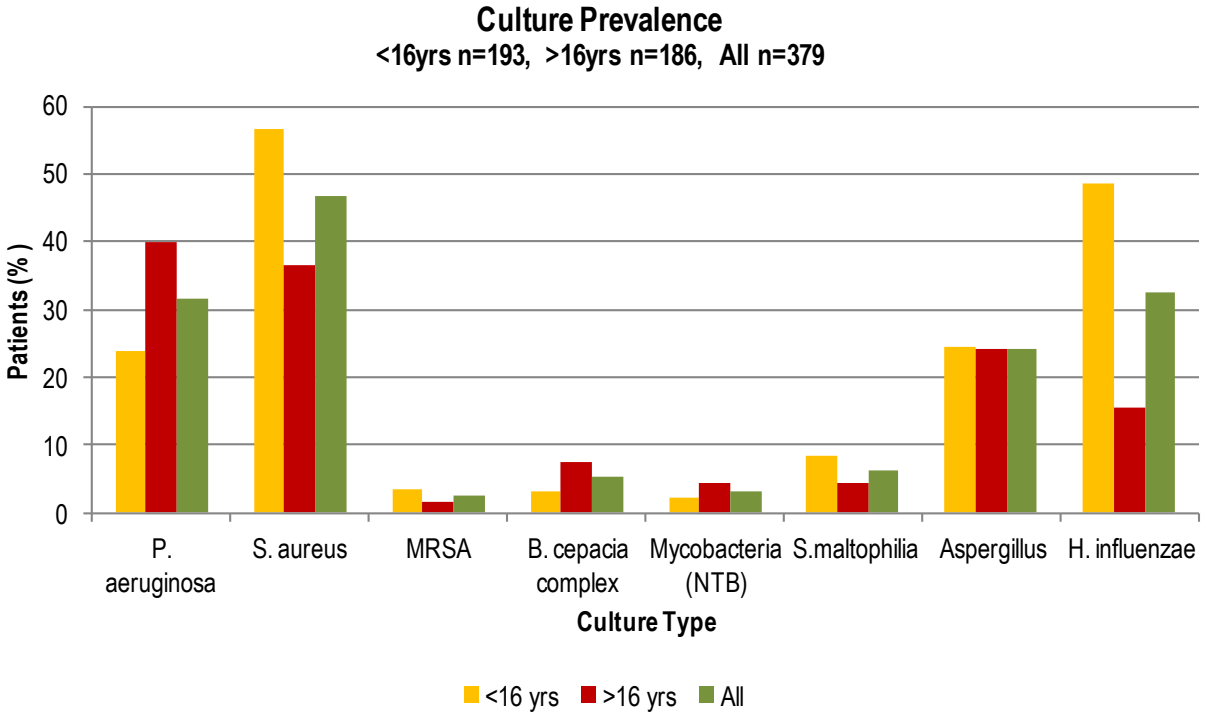
Medications



Medication	<16 yrs, n = Yes	%	>16 yrs, n=186 Yes	%	All, n = 379 Yes	%
Dornase Alfa	26	13.5	49	26.3	75	19.8
Chronic Macrolide AB (Oral)	19	9.8	87	46.8	106	28.0
Hypertonic Saline	86	44.6	77	41.4	163	43.0
Aminoglycosides	45	23.3	38	20.4	83	21.9
Colistin	9	4.7	2	1.1	29	7.7
InhaledOther	3	1.6	2	1.1	5	1.3

The use in NZ of hypertonic saline is higher than recorded in some of the comparative national registries, but our use of dornase alpha and chronic macrolide therapy is lower. With the changing criteria to obtain full funding of these medications, individuals should again be reviewed to determine if a trial of these medications could be beneficial.

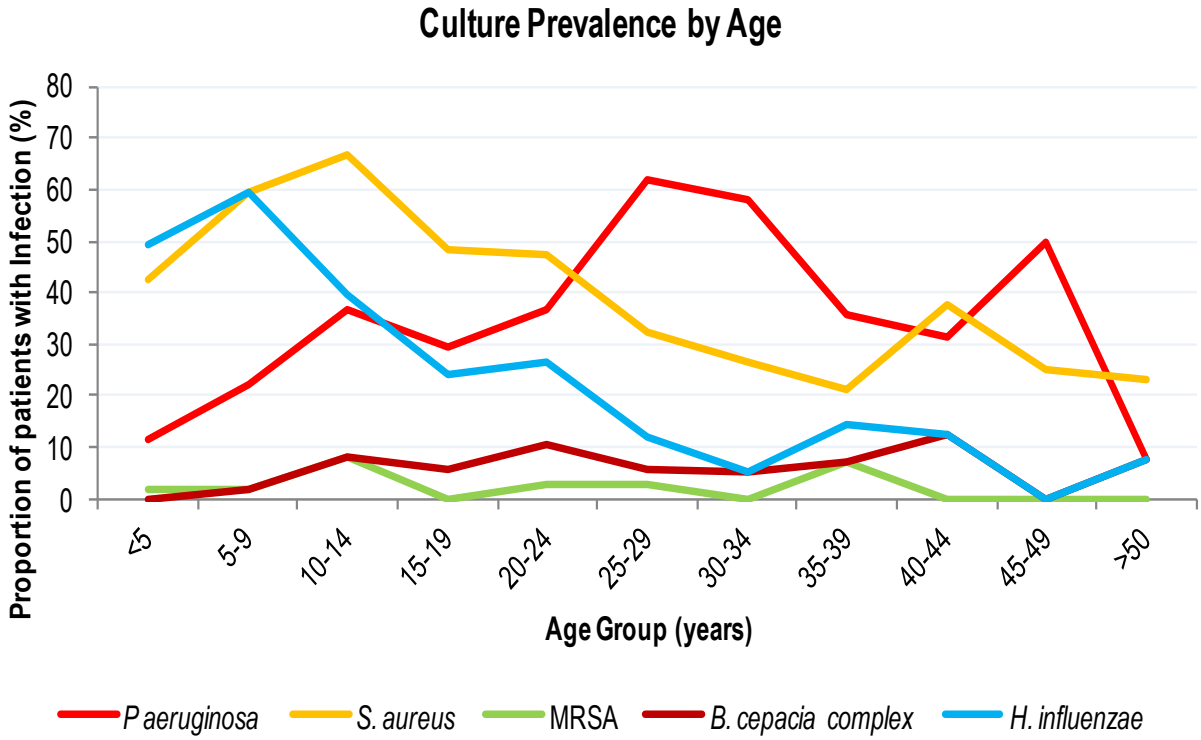
Microbiology



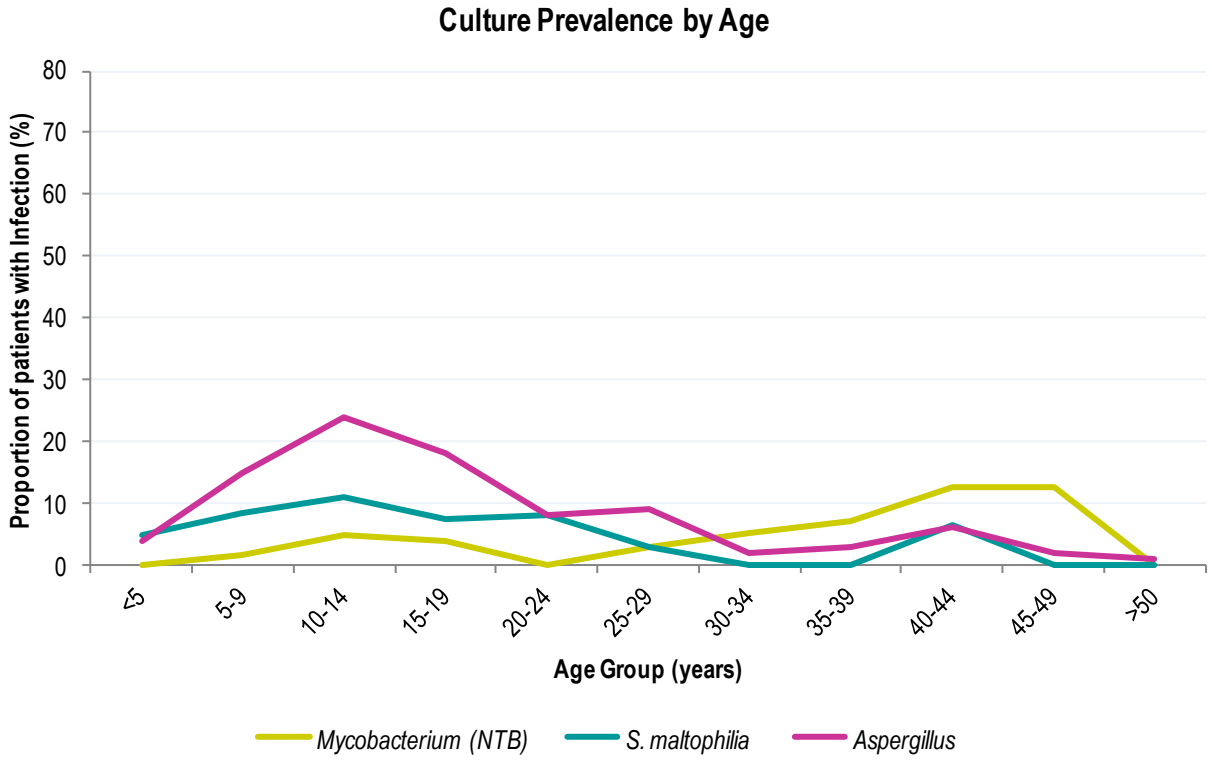
Culture	<16 yrs, n = 193		>16 yrs, n=186		All, n=379	
	Yes	%	Yes	%	Yes	%
P. aeruginosa	46	23.8	74	39.8	120	31.7
S. aureus	109	56.5	68	36.6	177	46.7
MRSA	7	3.6	3	1.6	10	2.6
B. cepacia complex	6	3.1	14	7.5	20	5.3
Mycobacteria (NTB)	4	2.1	8	4.3	12	3.2
S.maltophilia	16	8.3	8	4.3	24	6.3
Aspergillus	47	24.4	45	24.2	92	24.3
H. influenzae	94	48.7	29	15.6	123	32.5

There seems to be higher rates of Staphylococcus and Aspergillus, similar rates of Pseudomonas, and lower rates of Burkholdaria and MRSA here in New Zealand when compared to other registries.

This data may have been collected differently between clinics and needs further examination (annual review versus whole year culture results for example)



Age (yrs)		*P.aeruginosa*		*S. aureus*		*MRSA*		*B. cepacia complex*		*H. infuenzae*	
	n	n	%	n	%	n	%	n	%	n	%
<5	61	7	11.5	26	42.6	1	1.6	0	0.0	30	49.2
5-9	59	13	22.0	35	59.3	1	1.7	1	1.7	35	59.3
10-	63	23	36.5	42	66.7	5	7.9	5	7.9	25	39.7
15-	54	16	29.6	26	48.1	0	0.0	3	5.6	13	24.1
20-	38	14	36.8	18	47.4	1	2.6	4	10.5	10	26.3
25-	34	21	61.8	11	32.4	1	2.9	2	5.9	4	11.8
30-	19	11	57.9	5	26.3	0	0.0	1	5.3	1	5.3
35-	14	5	35.7	3	21.4	1	7.1	1	7.1	2	14.3
40-	16	5	31.3	6	37.5	0	0.0	2	12.5	2	12.5
45-	8	4	50.0	2	25.0	0	0.0	0	0.0	0	0.0
>50	13	1	7.7	3	23.1	0	0.0	1	7.7	1	7.7
Total	379	120	31.6	177	46.7	10	2.6	20	5.3	123	32.5



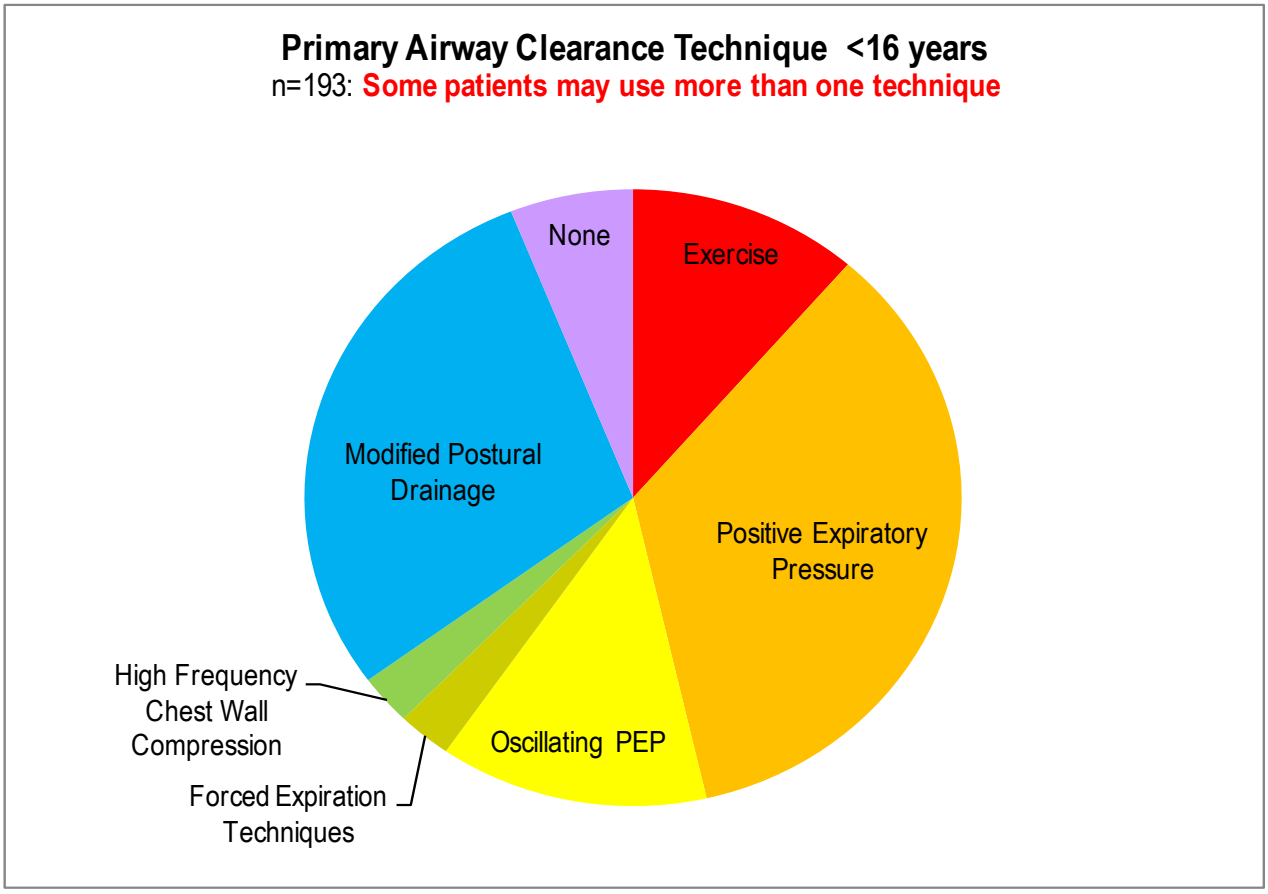
Age (yrs)		*Mycobacterium (NTB)*		*S. maltophilia*		*Aspergillus*	
	n	n	%	n	%	n	%
<5	61	0	0.0	3	4.9	4	6.6
5-9	59	1	1.7	5	8.5	15	25.4
10-14	63	3	4.8	7	11.1	24	38.1
15-19	54	2	3.7	4	7.4	18	33.3
20-24	38	0	0.0	3	7.9	8	21.1
25-29	34	1	2.9	1	2.9	9	26.5
30-34	19	1	5.3	0	0.0	2	10.5
35-39	14	1	7.1	0	0.0	3	21.4
40-44	16	2	12.5	1	6.3	6	37.5
45-49	8	1	12.5	0	0.0	2	25.0
>50	13	0	0.0	0	0.0	1	7.7
Total	379	12	3.2	24	6.3	92	24.3

Hospital & Home IVA Days

		Home IV Days				Hospital IV Days				Total
Age	n Age	n	%	Total	Mean	n	%	Total	Mean	IVA Days
0-3	39	4	10.3	51	13	13	33.3	164	13	215
4-7	41	7	17.1	75	11	16	39.0	253	16	328
8-11	41	9	22.0	193	21	17	41.5	324	19	517
12-15	41	12	29.3	198	17	24	58.5	662	28	860
16-19	42	5	11.9	181	36	19	45.2	477	25	658
20-23	25	4	16.0	56	14	11	44.0	275	25	331
24-27	22	12	54.5	279	23	13	59.1	458	35	737
28-31	21	7	33.3	186	27	9	42.9	130	14	316
32-35	8	1	12.5	26	26	2	25.0	16	8	42
36-39	11	3	27.3	50	17	3	27.3	45	15	95
40-43	13	4	30.8	93	23	9	69.2	221	25	314
44-47	7	1	14.3	14	14	2	28.6	63	32	77
48-51	5	1	20.0	30	30	2	40.0	26	13	56
52-55										
56-59	4	1	25.0	13	13	1	25.0	1	1	14
>=60	5	0	0.0	0	0	1	25.0	20	20	20
Total	325	71		1445		142		3135		4580

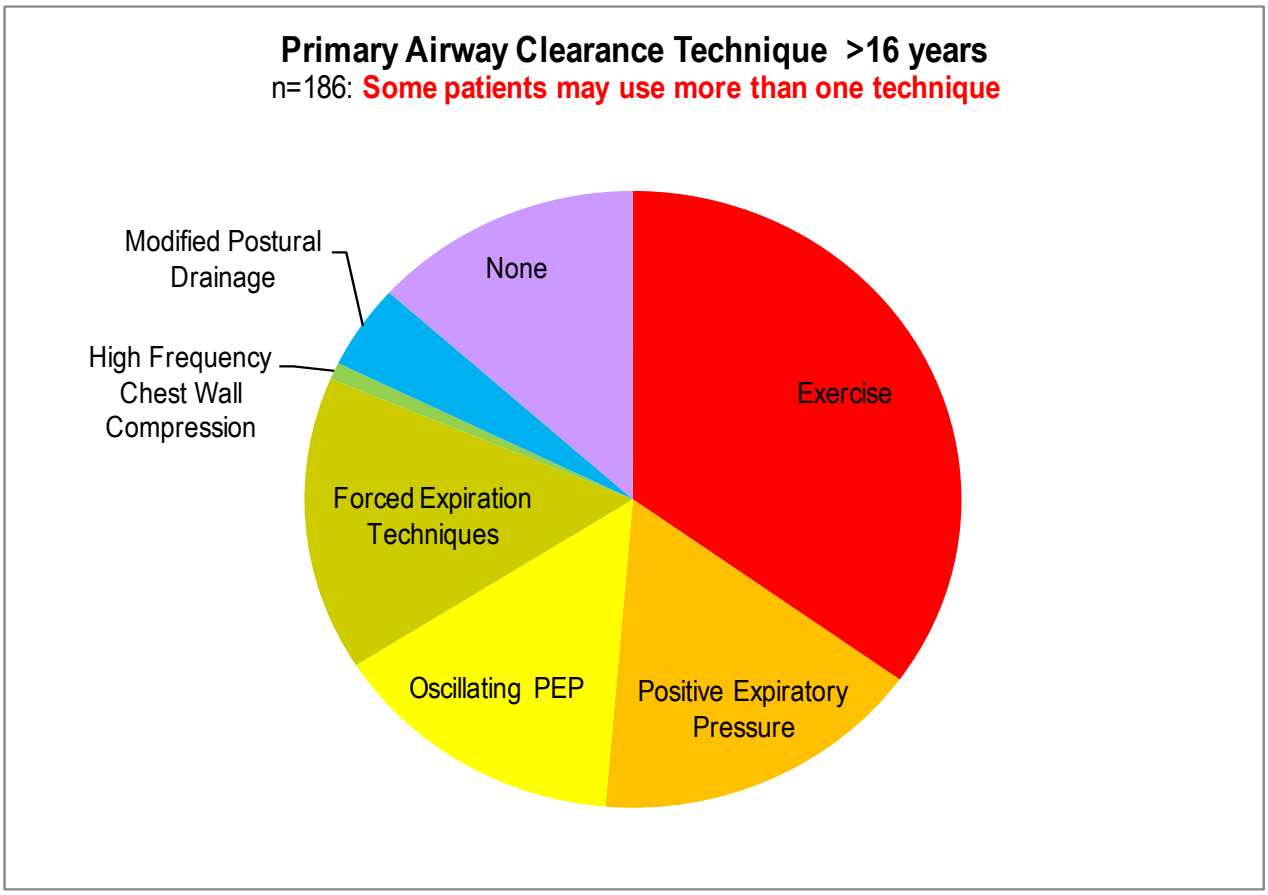
About one quarter of intravenous antibiotics are delivered in the home, with a peak prevalence occurring in early adulthood.

Primary Airway Clearance



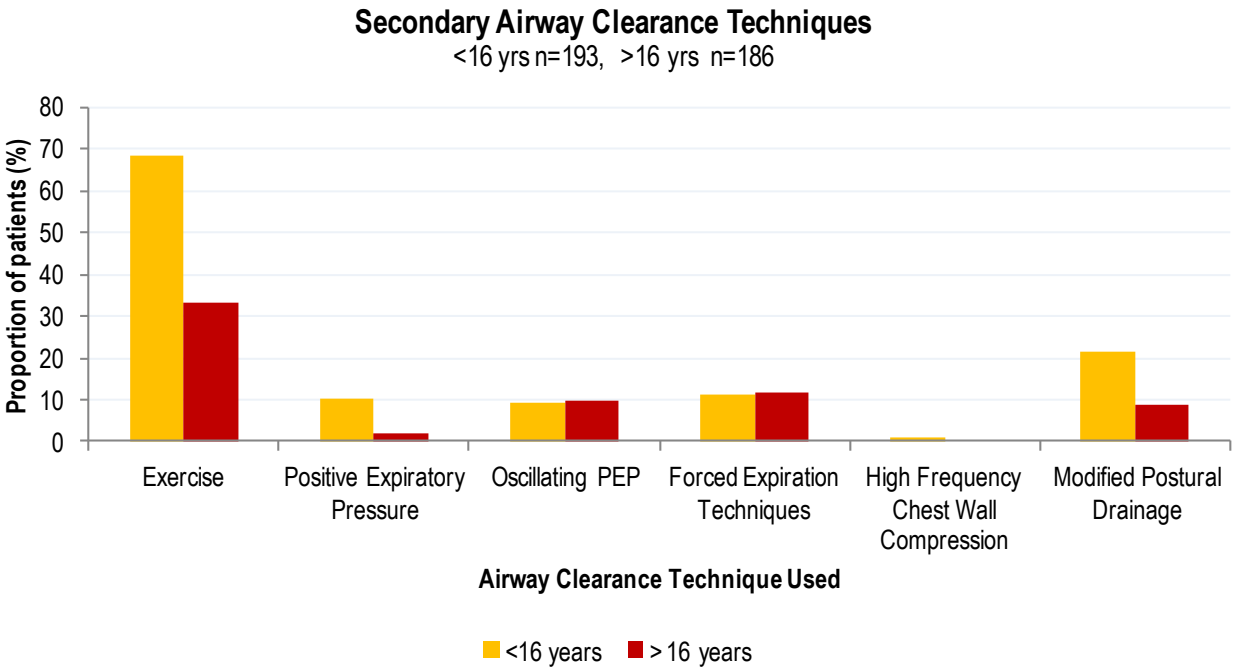
* number of individuals employing each technique at least once in the		
Technique	**<16 years**	
	n	**%**
Exercise	30	15.5
Positive Expiratory Pressure	93	48.2
Oscillating PEP (eg: Flutter, Acapella, Bubble Pep)	35	18.1
Forced Expiration Techniques (huff cough, active cycle breathing, autogenic drainage)	7	3.6
High Frequency Chest Wall Compression (eg: vest)	7	3.6
Modified Postural Drainage	77	39.9
None	16	8.3
Total	265	

In the younger children and adolescents with CF the predominant physiotherapy techniques are PEP and modified postural drainage.



* number of individuals employing each technique at least once in the year. Data collected from 186 patients		
Technique	**>16 years**	
	n	**%**
Exercise	79	42.5
Positive Expiratory Pressure	37	19.9
Oscillating PEP (eg: Flutter, Acapella, IPV)	33	17.7
Forced Expiration Techniques (eg:huff cough, active cyce breathing, autogenic drainage)	35	18.8
High Frequency Chest Wall Compression (eg: vest)	2	1.1
Modified Postural Drainage	10	5.4
None	30	16.1
Total	226	

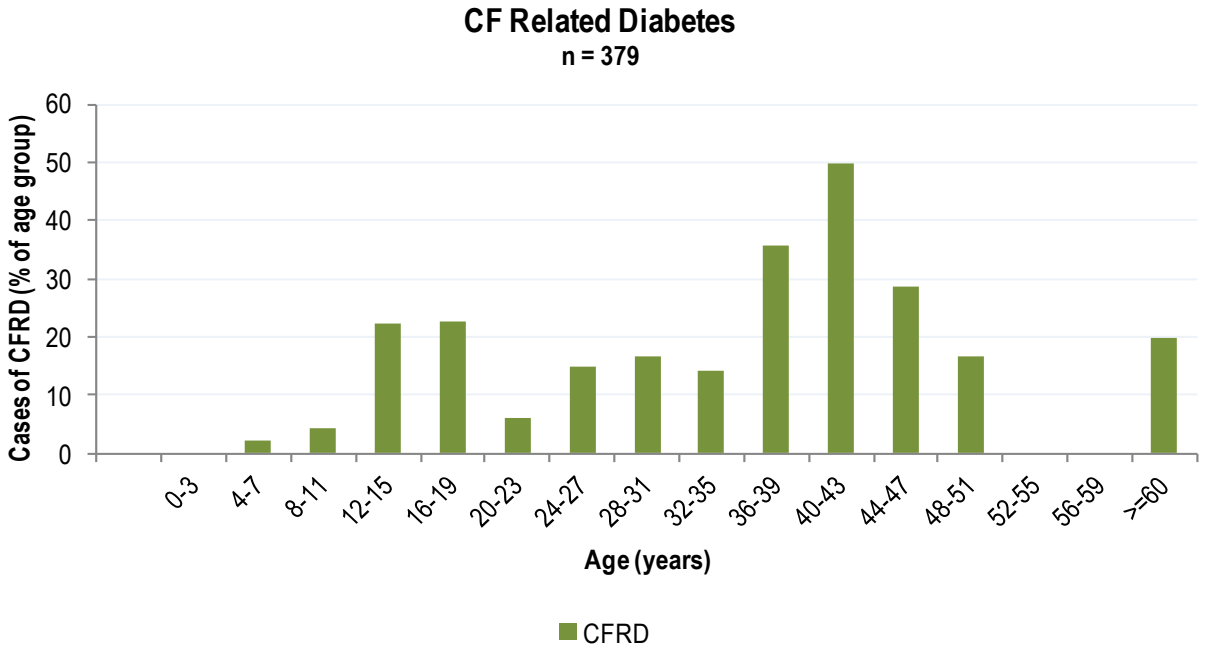
In adults with CF it appears that exercise is the primary airway clearance technique used. It would be interesting to investigate this further.



Data collected in 193 <16 years, 186 >16 years; Some patients may use more than

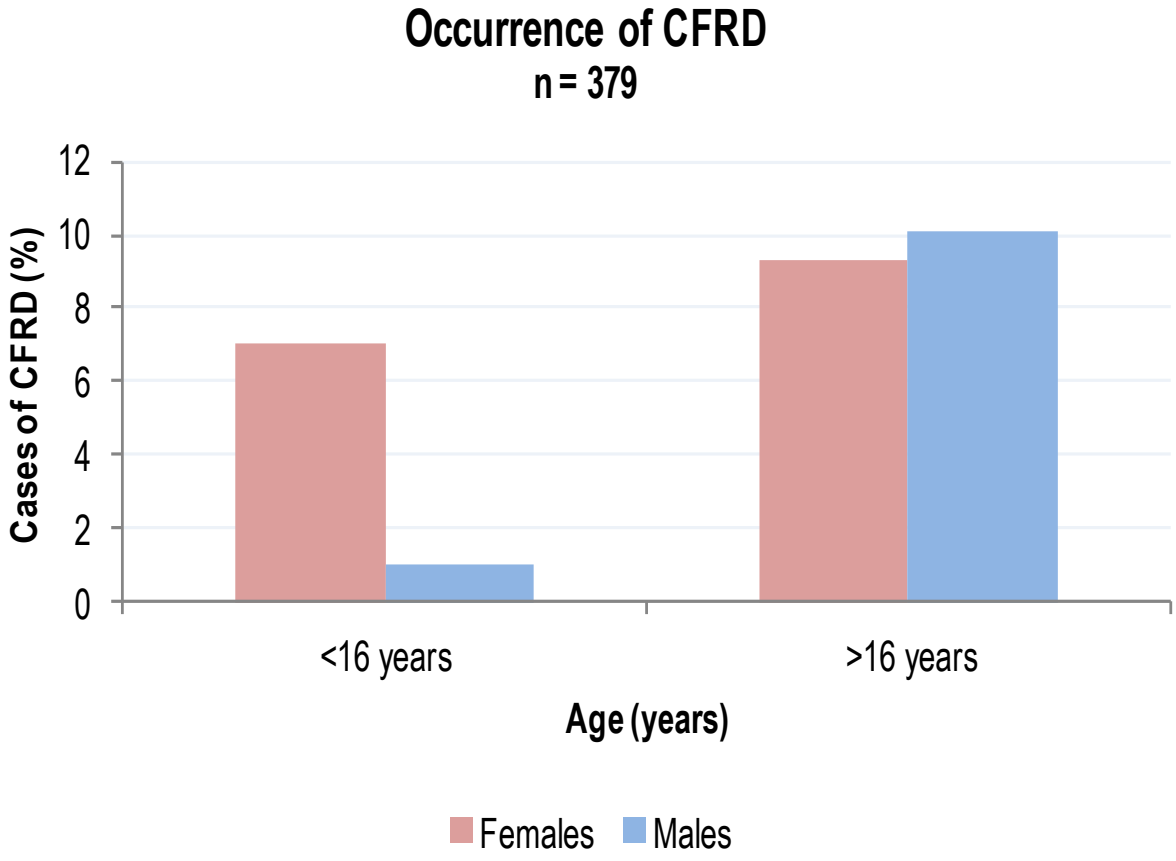
Technique	<16 years		>16 years	
	n	%	n	%
Exercise	132	68.4	62	33.3
Positive Expiratory Pressure	20	10.4	4	2.2
Oscillating PEP (eg: Flutter, Acapella,	18	9.3	18	9.7
Forced Expiration Techniques (eg:huff cough, active cyce breathing,	22	11.4	22	11.8
High Frequency Chest Wall Compression (eg: vest)	2	1.0	0	0.0
Modified Postural Drainage	41	21.2	16	8.6
Total	235		122	

CF_Related Diabetes



Age (yrs)	Age n	CFRD n	% age	% CF popula-
0-3	52	0	0.0	0.0
4-7	46	1	2.2	0.3
8-11	46	2	4.3	0.5
12-15	49	11	22.4	2.9
16-19	44	10	22.7	2.6
20-23	33	2	6.1	0.5
24-27	27	4	14.8	1.1
28-31	24	4	16.7	1.1
32-35	7	1	14.3	0.0
36-39	14	5	35.7	1.3
40-43	14	7	50.0	1.8
44-47	7	2	28.6	0.5
48-51	6	1	16.7	0.3
52-55	0	0	-	-
56-59	5	0	0.0	0.0
>=60	5	1	20.0	0.3
Total	379	51		13.5

Similar to other national registries there is a peak in the incidence of CF related diabetes in the teenage years and then again in mid adulthood.



	n	CFRD n	%	<16	%	>16	%
Females	172	28	16.3	12	7.0	16	9.3
Males	207	23	11.1	2	1.0	21	10.1
Total	379	51	13.46	14	3.7	37	9.8

There are higher numbers of girls with CF related diabetes in the younger age group but this evens out in the adult 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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