Port CF 2016

New Zealand CF Data Registry

CYSTIC FIBROSIS NZ

Contents

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

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, 2016 Registry Report, Cystic Fibrosis New Zealand. https://www.cfnz.org.nz/what-we-do/port-cf-data-registry/

Introduction

From the Chair of the Port CF Steering Committee

Associate Professor Cass Byrnes
Chair & Port CF Principal Investigator

Jane Bollard
Chief Executive, Cystic Fibrosis NZ

Cystic Fibrosis New Zealand and the Port CFNZ Steering Committee are pleased to present The National Cystic Fibrosis Data Registry 2016 Report; data collected from children, young persons and adults with cystic fibrosis (CF) in New Zealand.

We would like to thank:

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

This sixth Registry Report gives an increasingly accurate picture of people with CF and their outcomes for New Zealand with nearly 98% opting to provide this anonymous data. From 2012 – 2015 an additional 26 people were added to the registry and by 2016 another 52 people have been added.

We have developed the database further over the last year, increasing the amount of data captured with annual reviews, clinic reviews and hospital admissions. We are now part of the group working on 'harmonisation' of data registries for cystic fibrosis involving representation from all countries that have a CF registry.

The 2016 Registry Report represents the most data captured on our population with CF to inform future care and what future resources are needed.

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

Port CF Steering Committee

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A special thanks to:
Andrew Watson, Canterbury District Health Board

Prof Christopher Frampton, Christchurch

CF Clinics in New Zealand

Northland (Paediatrics)
Whangarei Hospital, Whangarei

Auckland (Paediatrics and Adults)
Starship Child Health
Greenlane Clinical Centre

Waikato (Paediatrics and Adults)
Waikato Hospital, Hamilton

Taranaki (Paediatrics and Adults)
Taranaki Base Hospital, New Plymouth

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

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

Canterbury (Paediatrics and Adults)
Christchurch Hospital, Christchurch

Otago (Paediatrics and Adults)
Dunedin Hospital, Dunedin

Southland (Paediatrics)
Kew Hospital, Invercargill

Notes to the Registry

New Zealand has a total CF population comparable to a single clinic in the USA/UK and this data gives our national statistics. Our smaller population provides significant challenges to statistical interpretation as 'outliers' in terms of late diagnoses and key markers will have an impact on outcomes reported.

The brief commentary provided throughout this report reflects opinions based on our data, and when cited as compared to other registries these are from Australia, UK and USA.

Although we have a total of 501 registered in Port CFNZ, not all individuals had an input for all questions. While the total is 501 (233 children <16 year years, 268 adults > 16 years) at the top of each table or figure is the total number that had a response to the question. For example, on pancreatic enzymes a total response was obtained from 407 patients (205 children and 202 adults) on page 20. The rest of the data for the remaining individuals is missing.

The NZ Registry data is becoming more robust and accurate; we welcome its use in audit and research projects for researchers from reputable institutions.

Enquiries regarding the use of data can be made either to Jane Bollard, CE, CFNZ or to the Project Co-ordinator Jan Tate.

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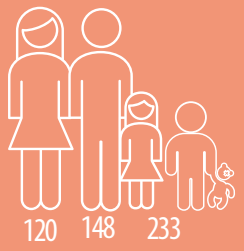
Glossary of Terms

CF	Cystic Fibrosis
CFNZ	Cystic Fibrosis New Zealand
PWCF	People with cystic fibrosis
FEV_1	Lung function measurement as forced expiratory volume in one second
BMI	Body Mass Index: measurement of weight relative to height
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
Port CFNZ	Data registry

CF at a glance

97-98% of New Zealanders with cystic fibrosis are in the Registry = **501 PATIENTS**



BONE DENSITY by DEXA Scan

13.6%	9.2%	77.1%
NORMAL	ABNORMAL	NOT DONE

CF RELATED DIABETES

11.1% Paediatrics

28.7% Adults

MEDICATION

53.7%	Hypertonic Saline
27.5%	Dornase Alfa
20%	TOBI
22.8%	Chronic Macrolide

MICROBIOLOGY

2.6% Mycobacterium Non-tuberculosis

4.4% Burkholderia Cepacia Complex

42.1% Staphylococcus Aureus

24.4% Pseudomonas Aeruginosa

LIVER FUNCTION

22.4%	7.8%	69.8%
NORMAL	ABNORMAL	NOT DONE

MEDIAN AGE

of patients in the Registry

17.38 YEARS

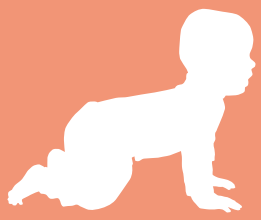
IV DAYS

144 Patients in hospital

87 Patients at home

GENOTYPE

52.8%	F508del / F508del
16.6%	F508del / Other
9.5%	No F508del
3.77%	F508del / G551D
5.1%	F508del / G542X



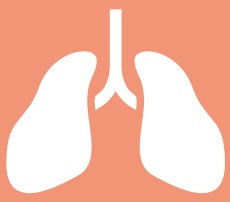
6 NEW DIAGNOSES for babies under one year of age

435 Annual Reviews

AIRWAY CLEARANCE

26% of adults and **18%** of paediatrics list excercise as their primary form of airway clearance

LUNG FUNCTION



77.3	77.4	99.4
MEN	WOMEN	CHILDREN

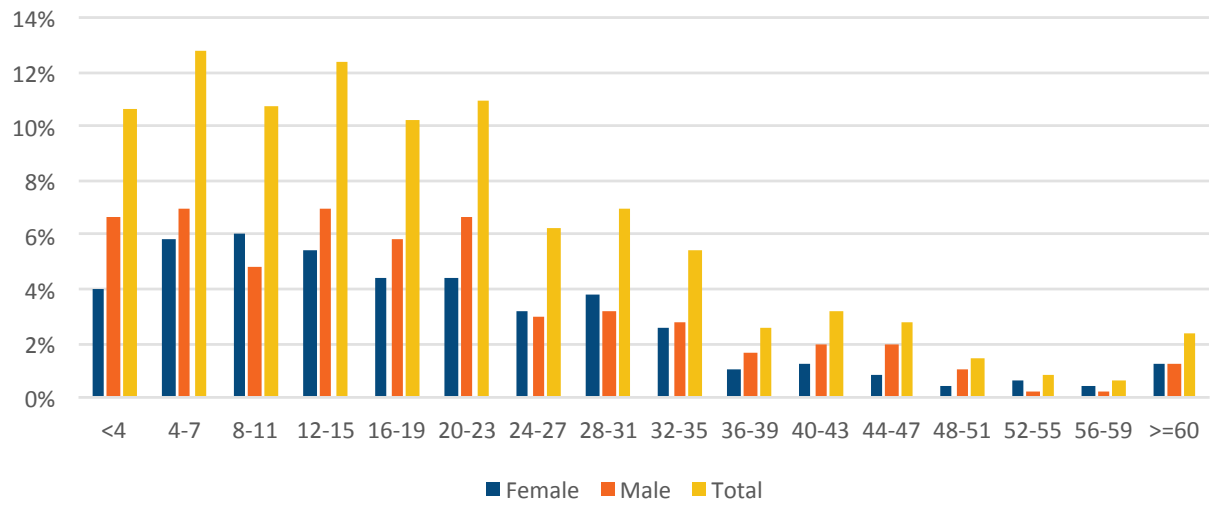
Summary of Key Indicators					
	2016	**2015**	**2014**	**2013**	**2012**
CF Patients Registered	501	449	443	444	423
Diagnosis					
Diagnosis age <1 year	6	5	7	5	11
Diagnosis age 1- 15 years	3				
Diagnosis age >16 years	2	0	2	3	2
Age					
Median Age (in years)	17.38	18.25	18.11	17.55	16.15
Mean Age (in years)	20.04				
PWCF <16 years					
Number	233	192	196	205	209
Percent	46.5%	42.8%	44.2%	46.2%	49.4%
PWCF >16 years					
Number	268	257	247	239	214
Percent	53.5%	57.2%	55.8%	53.8%	50.6%
Gender					
Males					
Number	275	247	240	240	228
Percent	54.9%	55.0%	54.2%	54.1%	53.0%
Females					
Number	226	202	203	204	195
Percent	45.1%	45.0%	45.8%	45.9%	46.1%
Genotyped					
Number	450	400	429	426	407
Percent	90.0%	89.1%	96.8%	95.9%	96.2%
FEV1 (% predicted)					
Mean	85.0%				
Median	88.4%	85.6%	85.1%	84.3%	84.5%
FEV1 < 16 Years					
Mean	97.3%				
Median	99.3%	98.9%	97.7%	96.6%	97.2%
FEV1 > 16 Years					
Mean	72.6%				
Median	77.4%	77.0%	78.0%	70.7%	70.6%
FEV1 < 18 Years					
Mean	95.0%				
Median	98.0%				
FEV1 > 18 Years					
Mean	71.2%				
Median	75.1%				

1. Demographics

501 Patients

Age Group	All		Male		Female	
	Number in age group	**Percent** of all Patients	**Number** in age group	**Percent** of all patients	**Number** in age group	**Percent** of all patients
0-3	53	10.6%	33	6.60%	20	3.99%
4-7	64	12.8%	35	6.99%	29	5.79%
8-11	54	10.8%	24	4.79%	30	5.99%
12-15	62	12.4%	35	6.99%	27	5.39%
16-19	51	10.2%	29	5.79%	22	4.39%
20-23	55	11.0%	33	6.60%	22	4.39%
24-27	31	6.2%	15	3.00%	16	3.19%
28-31	35	7.0%	16	3.19%	19	3.79%
32-35	27	5.4%	14	2.79%	13	2.59%
36-39	13	2.6%	8	1.60%	5	1.00%
40-43	16	3.2%	10	2.00%	6	1.20%
44-47	14	2.8%	10	2.00%	4	0.80%
48-51	7	1.4%	5	1.00%	2	0.40%
52-55	4	0.8%	1	0.20%	3	0.60%
56-59	3	0.6%	1	0.20%	2	0.40%
>=60	12	2.4%	6	1.20%	6	1.20%
Total	**501**	**100.0%**	**275**	**54.9%**	**226**	**45.1%**
Median	**17.4**					
Range	**0 - 73**					

1.1 Age distribution



The median age of persons with CF in New Zealand has increased from 16 to 17.4 years over the five years that we have had national registry data. 'Children' in international registries are defined as either up to 16 years or up to 18 years of age. In New Zealand if we include children as being up to 16 years we have 233 children (46.5% of total) and 268 adults (53.5% of total).

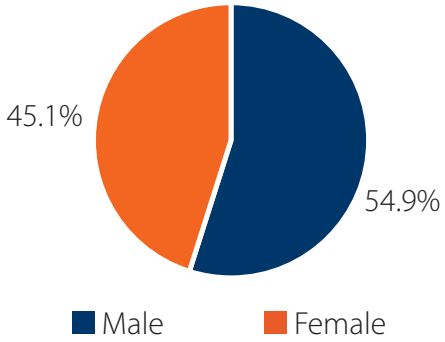
1.2 Gender Distribution

501 Patients

	All, n = 501		<16, n = 233		>16, n = 268	
	Number in age group	**Percent** of all Patients	**Number** in age group	**Percent** of all patients	**Number** in age group	**Percent** of all patients
Male	275	54.9%	127	23.4%	148	29.5%
Female	226	45.1%	106	21.2%	120	24.0%
Totals	**501**	**100%**	**233**	**46.5%**	**268**	**53.4%**

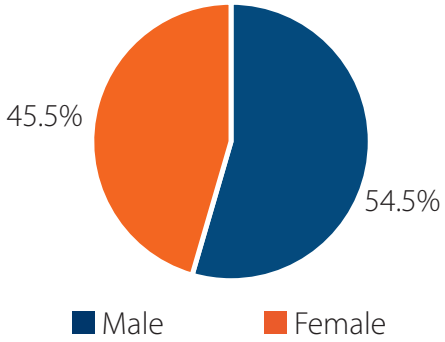
Gender Distribution of All Patients

501 Patients



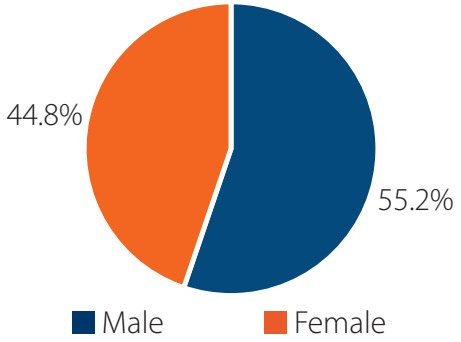
Gender Distribution <16 Years

233 Patients



Gender Distribution >16 Years

268 Patients



"The median age of persons with CF in New Zealand has increased from 15.70 to 17.4 years over the six years that we have had national registry data."

2. Genotypes

450 Patients

Mutations	Number of Patients Genotyped	Percentage of Patients Genotyped
Homozygous F508del	238	52.8%
Heterozygous F508del	170	37.7%
No F508 or both unidentified	42	9.5%
Total	**450**	

2.1 Second Allele of Heterozygous F508del

170 Patients

Second Allele	c.DNA Name	Number of Patients	Percent of Patients
G542X	c.1624G>T	23	5.10%
G551D	c.1652G>A	17	3.77%
R117H	c.350G>A	16	3.55%
G85E	c.254G>A	5	1.10%
^1507	c.1519_1521delATC	4	0.88%
621+1G->T	c.489+1G>T	3	0.67%
N1303K	c.3909c>G	3	0.67%
3849+10kbC->T	c.3717+12191C>T	2	0.44%
1717-1G->A	c.1585-1G>A	2	0.44%
1898+1G->A	c.1766+1G>A	2	0.44%
A455E	c.1364C>A	2	0.44%
1078delT	c.948delT	2	0.44%
D1152H	c.3454G>C	2	0.44%
R334W	c.1000C>T	2	0.44%
Q493X	c.1477C>T	1	0.22%
2789+5G->A	c.2657+5G>A	1	0.22%
3120+1G->A	c.2988+1G>A	1	0.22%
3659delC	c.3528delC	1	0.22%
712-1G->T	c.580-1G>T	1	0.22%
R347H	c.1040G>A	1	0.22%
R347P	c.1040G>C	1	0.22%
R560T	c.1679G>C	1	0.22%
W1282X	c.3846G>A	1	0.22%
R1158X	c.3472C>T	1	0.22%
R553X	c.1657C>T	0	0.00%
Other genetic mutation		75	16.60%

2.2 No F508 del Mutations — 42 Patients

	G542X	G551D	G85E	Other	R1162X	R117H	1717-1G->A
G542X	1			2			
G551D	1	1		3		4	1
N1303K		1		1			
Other			1	19	1		
Q493X		1				1	
R553X		1					
3849+10kbC->T							1
R117H						1	
W1282X				1			

2.3 Genotype Major Categories

Mutations	Number Patients Identified	Percentage Patients Identified
F508	408	90.5%
G551D	31	6.9%
G542X	27	5.9%
R117H	21	4.7%
G85E	6	1.3%

Note: Because people have two genes patients are counted twice, once for each gene. The total number of patients is 450.

Our high percentage of F508del is in keeping with the international registries from European derived populations. In total only 42 persons in New Zealand do not have at least one F508 mutation. Looking at the gene mutations recorded in 2016 Registry, 29 persons (6%) would not be detected by our current new born screening programme.

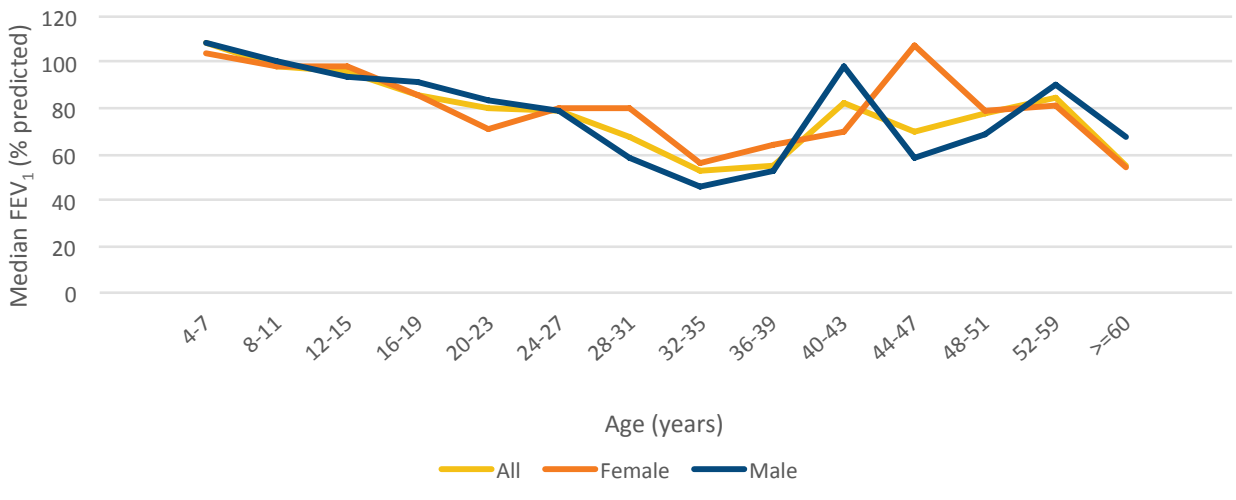
3. Respiratory

342 Patients

Age Group	All		Female		Male	
	Number in age group	Median FEV1	Number in age group	Median FEV1	Number in age group	Median FEV1
6-7	36	108.4	13	103.4	23	108.7
8-11	49	98.1	27	98.1	22	100.7
12-15	51	95.5	21	98	30	93.3
16-19	44	85.8	17	85.5	27	91.3
20-23	40	79.7	14	71.4	26	83
24-27	23	79.4	12	79.6	11	79.3
28-31	25	68	14	80	11	58
32-35	20	52.9	9	56.3	11	46
36-39	11	55.1	4	63.8	7	52.6
40-43	10	82.6	4	70.3	6	97.9
44-47	12	70	3	107.8	9	58.3
48-51	5	77.4	2	79.2	3	69.1
52-55	2	63.1	2	63.1	0	
56-59	3	86.8	2	85.4	1	90.7
>=60	11	55.2	6	54.4	5	67.1
Totals	**342**		**150**		**192**	

Median FEV1 (% predicted) >6

342 Patients



The median FEV1 of the population able to do lung function has always been >80% predicted since we started our national registry and the median this year is 88.4% (97.3% in children, 72.6% in adults). This necessarily excludes very young children who are unable to do lung function or those that find it very difficult because of technique or severity of disease. None-the-less FEV1 is an important prognostic indicator.

The trend regarding lung function with age is of gradual deterioration from early childhood to early adulthood. The late maintenance of lung function reflects those living longer with more mild disease and late diagnoses of people with milder CF phenotypes.

“The median FEV1 of the population able to do lung function has always been>80% predicted since we started our national registry.”

4. Nutrition

4.1 Paediatric BMI

189 Patients

Females <16			Males <16			All <16		
BMI Percentile			BMI percentile			BMI percentile		
Age group	Number in group	Median percentile	Age group	Number in group	Median percentile	Age group	Number in group	Median percentile
<4	15	69.2	<4	16	66.8	<4	31	68
4-7	26	66.3	4-7	32	64	4-7	58	65.2
8-11	27	59.5	8-11	24	47.8	8-11	51	53.7
12-15	17	66.8	12-15	32	52.3	12-15	49	59.6
Totals	**85**			**104**			**189**	

4.2 Adult BMI

207 Patients

Females <16			Males <16		All <16			
BMI Percentile			BMI percentile		BMI percentile			
Age group	Number in group	Median BMI	Age group	Number in group	Median BMI	Age group	Number in group	Median BMI
16-19	17	22.3	16-19	27	21.8	16-19	44	22.0
20-23	13	21.2	20-23	26	21.3	20-23	39	21.3
24-27	12	22.4	24-27	11	22.7	24-27	23	22.7
28-31	14	22.7	28-31	11	21.8	28-31	25	22.5
32-35	9	22.8	32-35	11	21.6	32-35	20	22.1
36-39	4	22.1	36-39	7	26.2	36-39	11	25.8
40-43	5	23.1	40-43	7	24.8	40-43	12	24.0
44-47	3	22.2	44-47	9	23.9	44-47	12	23.9
48-51	2	23.4	48-51	3	24.7	48-51	5	24.1
52-55	2	35.1	52-55	0		52-55	2	35.1
56-59	2	26	56-59	1	31.9	56-59	3	30.1
>=60	6	23.6	>=60	5	21.5	>=60	11	22.1
Totals	**89**			**118**			**207**	

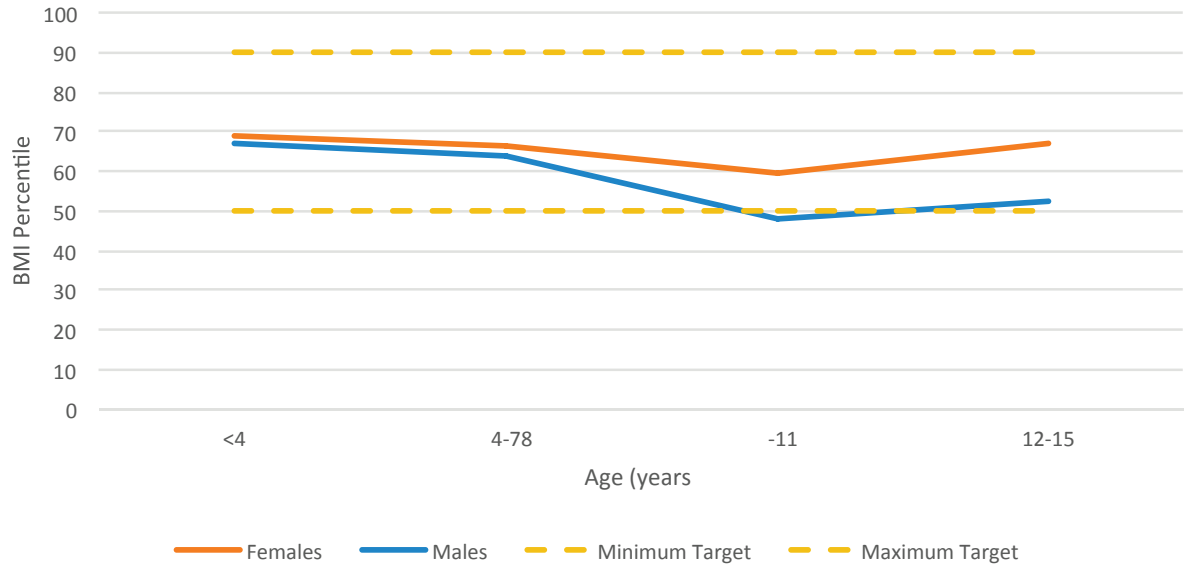
The relationship between nutrition, lung function and survival in CF is well established with normal body weight associated with better preservation of lung function. The Cystic Fibrosis Foundation (USA) suggest the following targets for optimal weight status:

1) Infants (0 to 24 months): weight-for-length ≥50th percentile using WHO growth charts
2) Children and Adolescents (2-18 years): BMI 50-85th percentile (CDC growth charts) or 50-91st percentile (WHO growth charts)
3) Adults: males BMI 23 - 27 kg/m2, females BMI 22 - 27 kg/m2

This is also reflected in the 'Nutrition Guidelines for Cystic Fibrosis in Australia and New Zealand' (https://www.thoracic.org.au/documents/item/1045)

4.3 Median BMI Percentile <16

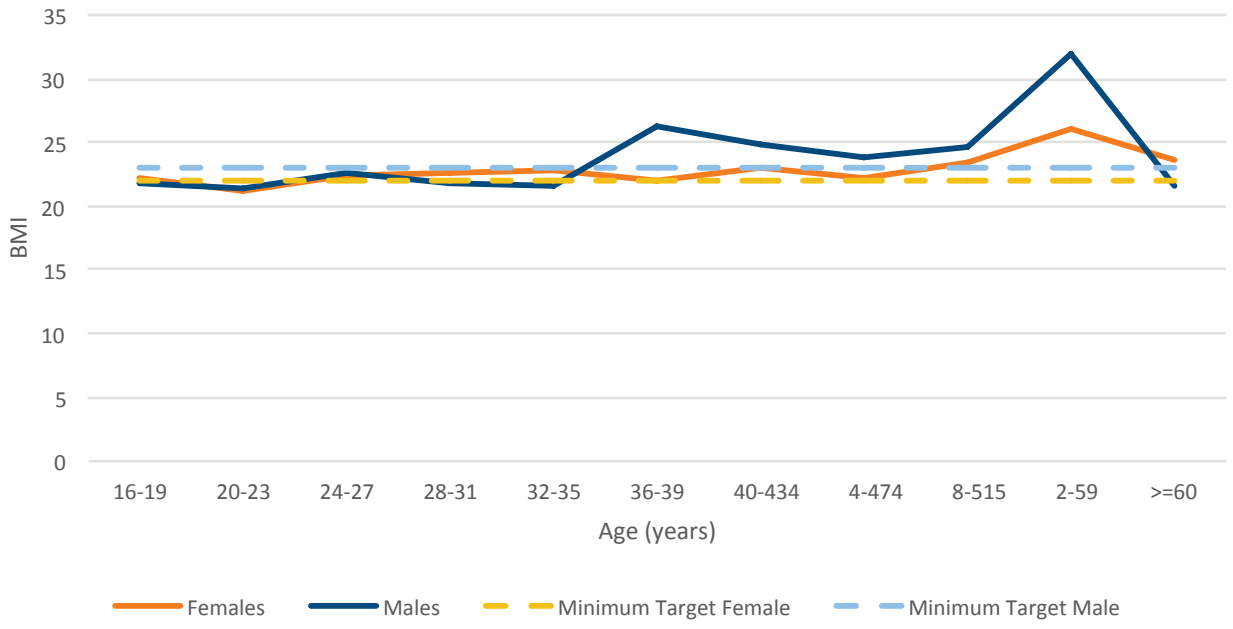
189 Patients



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

4.4 Median BMI Percentile >16

207Patients



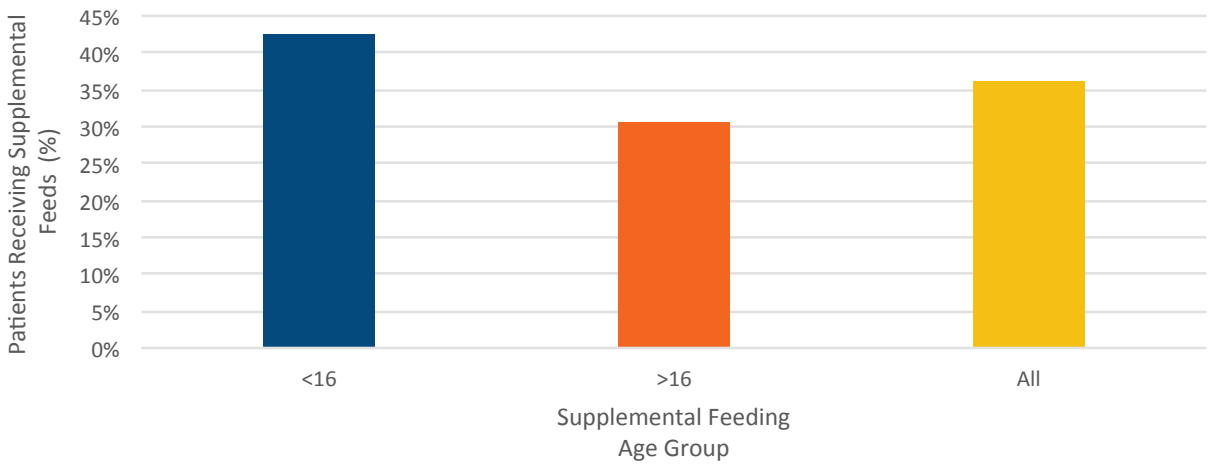
The optimal BMI for women is 22 - 27 and the dotted yellow line shows the minimum BMI for women in the target range. The optimal BMI for men is 23 - 27. The dotted blue line shows the minimum BMI for men in the target range.

4.5 Supplemental Feeding — 501 Patients

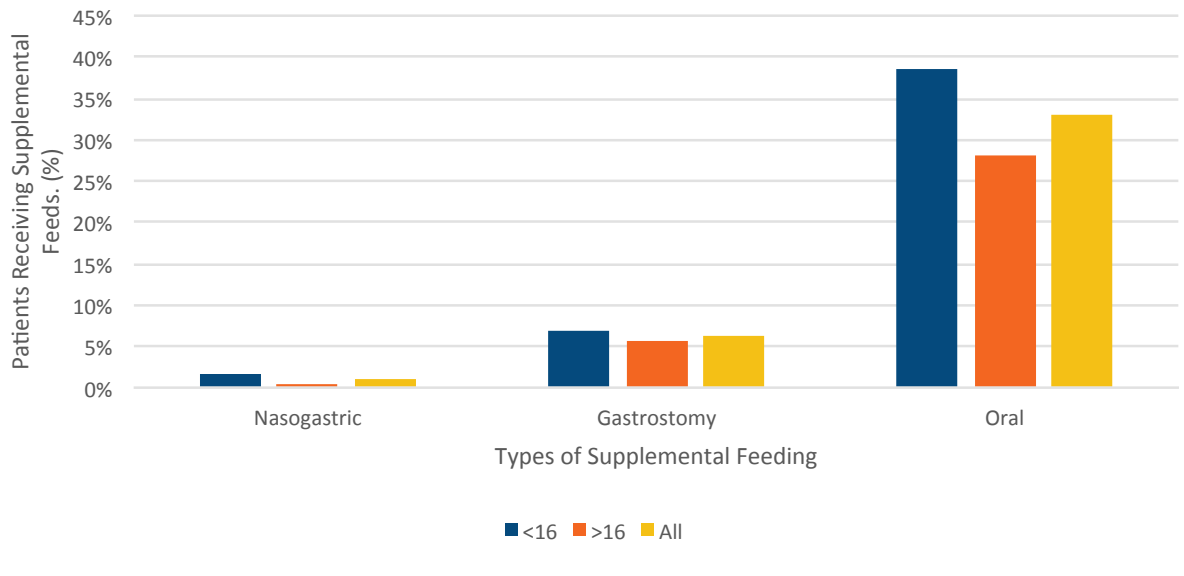
	<16 years, n =233	
	Yes	% <16 years supplemented
Supplemental Feeding	99	42.5%
Nasogastric	4	1.7%
Gastrostomy	16	6.9%
Oral	90	38.6%
	>16 years, n = 268	
	Yes	% >16 years supplemented
Supplemental Feeding	82	30.6%
Nasogastric	1	0.4%
Gastrostomy	16	6.0%
Oral	90	33.6%

NB: Some individuals maybe on more than one type of supplemental feed

4.6 Supplemental Feeding by Age Group — 398 Patients



4.7 Supplemental feeding by type — 398 Patients



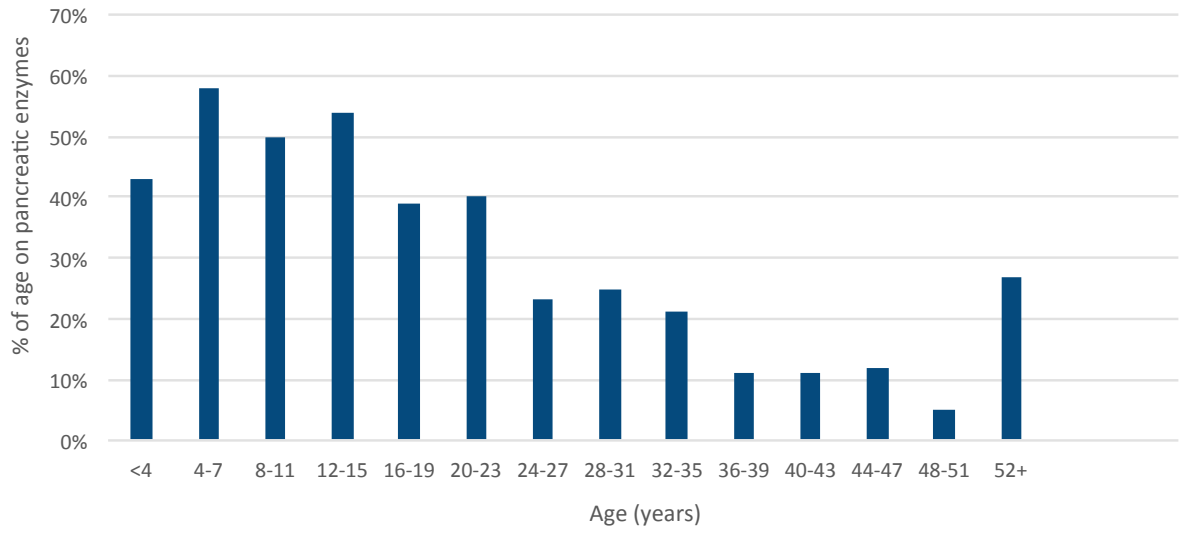
5. Pancreatic Enzymes

407 Patients

Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of CF population
<4	43	36	84%	8.8%
4-7	58	46	79%	11.3%
8-11	50	41	82%	10.1%
12-15	54	44	82%	10.8%
16-19	39	35	90%	8.6%
20-23	40	34	85%	8.4%
24-27	23	22	96%	5.4%
28-31	25	19	76%	4.7%
32-35	21	18	86%	4.4%
36-39	11	9	82%	2.2%
40-43	11	8	73%	2.0%
44-47	12	9	75%	2.2%
48-51	5	3	60%	0.7%
52-55	2	0	0%	0.0%
56-59	2	0	0%	0.0%
>=60	11	4	36%	1.0%
Totals	**407**	**328**		

Patients on Pancreatic Enzymes

407 Patients



6. Airway Clearance Techniques

6.1 Primary Airway Clearance

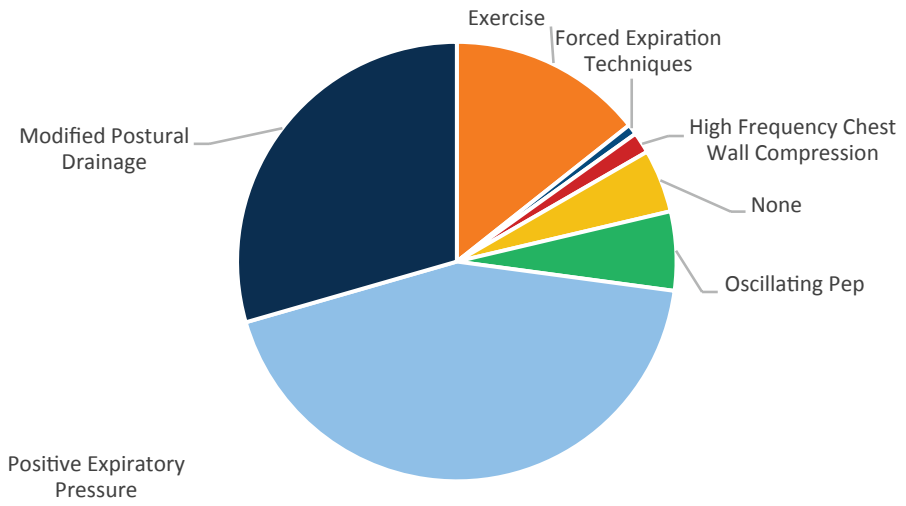
408 Patients

	<16 years, n = 206	
	Number of patients	Percent of patients
Positive Expiratory Pressure	112	54.4%
Modified Postural Drainage	76	36.9%
Exercise	37	18.0%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	15	7.3%
Forced Expiration Techniques	2	1.0%
(e.g. huff cough, active cycle breathing, autogenic drainage)		
High Frequency Chest Wall Compression (e.g. vest)	4	1.9%
None	12	5.8%

	>16 years, n = 202	
	Number of patients	Percent of patients
Positive Expiratory Pressure	74	37%
Modified Postural Drainage	9	5%
Exercise	52	26%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	32	15.8%
Forced Expiration Techniques	39	19.3%
(e.g. huff cough, active cycle breathing, autogenic drainage)		
High Frequency Chest Wall Compression (e.g. vest)	1	0.5%
None	27	13.4%

6.2 Paediatric Primary Airway Clearance

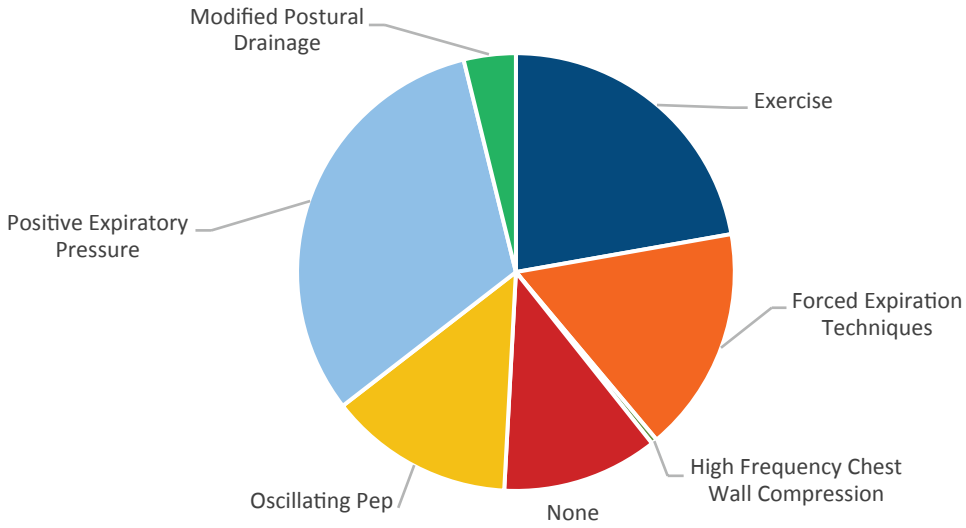
206 Patients



NOTE: Some patients may have used more than one technique as their primary airway clearance technique over the course of a year

6.3 Adult Primary Airway Clearance

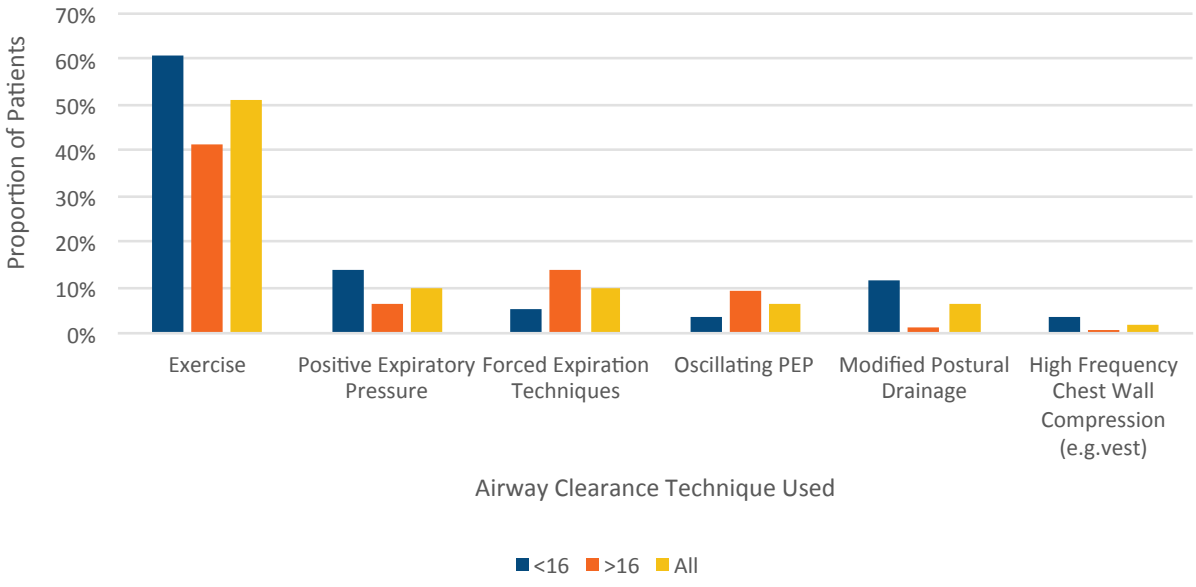
202 Patients



NOTE: Some patients may have used more than one technique as their primary airway clearance technique over the course of a year

6.4 Secondary Airway Clearance

408 Patients

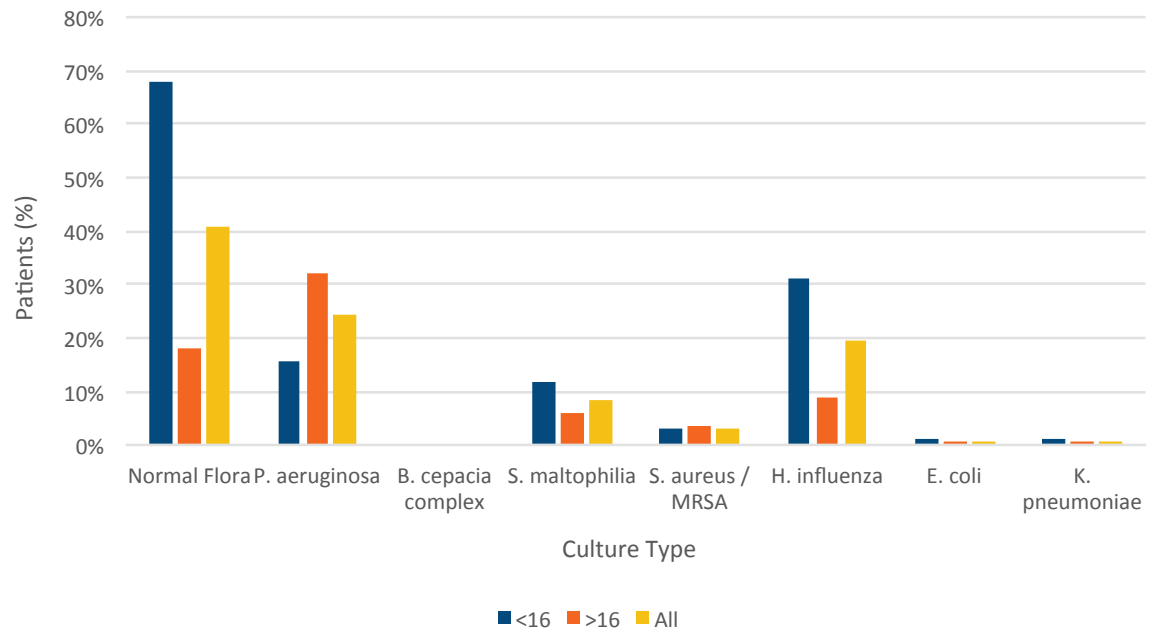


7. Microbiology

Microbiology					501Patients	
	<16 years 233 Patients		>16 years 268 Patients		All 501 Patients	
	Number	Percent	Number	Percent	Number	Percent
Normal Flora	159	68.0%	47	18.0%	206	41.0%
Haemophilus Influenza	73	31.3%	24	9.0%	97	19.4%
E. Coli	3	1.3%	1	0.4%	4	0.8%
Klebsiella Pneumoniae	2	0.9%	1	0.4%	3	0.6%
Stenotrophomonas Maltophilia	27	11.6%	16	6.0%	43	8.6%
Pseudomonas Aeruginosa	**36**	**15.5%**	**86**	**32.1%**	**122**	**24.4%**
Mucoid	11	4.7%	54	20.1%	65	13.0%
Non Mucoid	29	12.4%	54	20.1%	83	16.6%
Staphylococcus Aureus	**114**	**48.9%**	**97**	**36.2%**	**211**	**42.1%**
MSSA	107	45.9%	88	3280.0%	195	38.9%
MRSA	7	3.0%	9	3.4%	16	3.2%
Burkholderia Cepacia Complex	**2**					
Cenocepacia	0	0.0%	3	1.1%	3	0.6%
Multivorans	2	0.9%	8	3.0%	10	2.0%
Other	0	0.0%	2	0.7%	2	0.4%

7.1 Bacterial Culture Prevalence

501 Patients



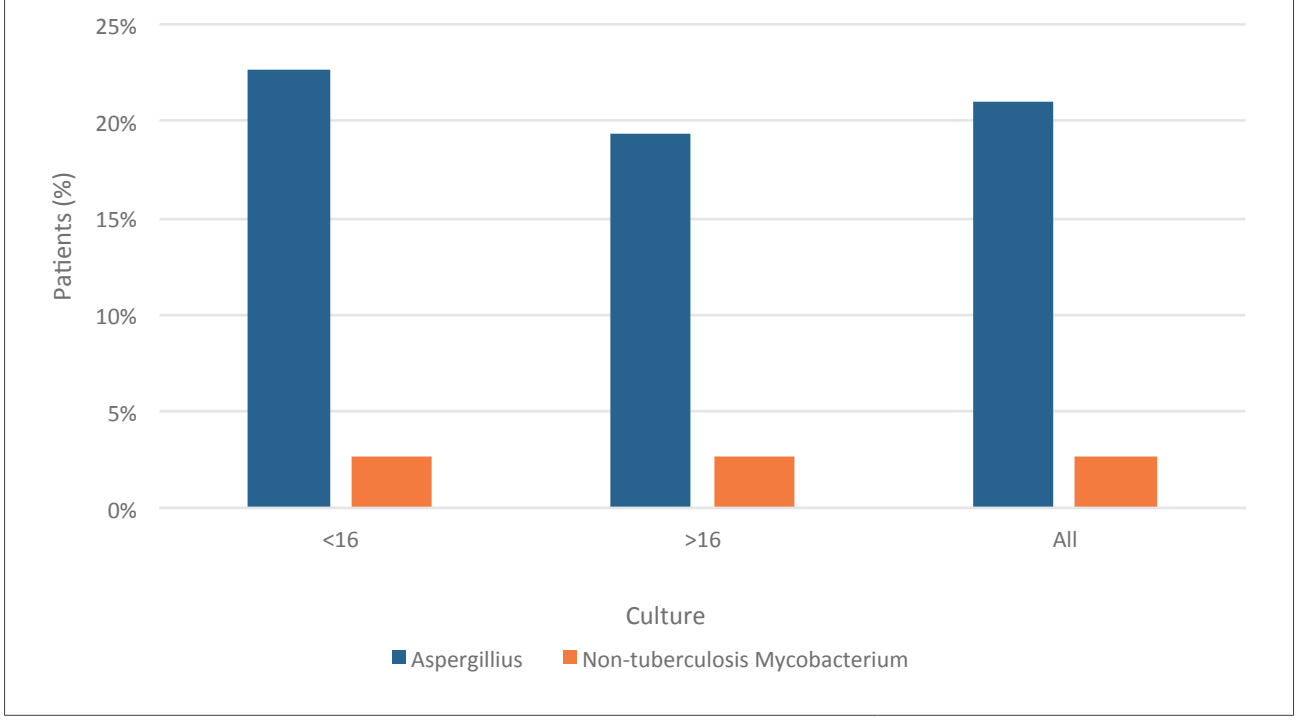
NOTE: The percentages of population with CF having had specific respiratory pathogens identified such as Staphylococcus aureus, Pseudomonas aeruginosa etc are very similar to the percentages presented in other registries.

Pseudomonas aeruginosa is found in 15.5% of the children and increases to 32.1% in adults. Our MRSA rates are relatively low at 3.2% overall.

(Australian data registry https://www.cysticfibrosis.org.au/dataregistry)

7.2 Nonbacterial/Fungal Prevalence

501 Patients

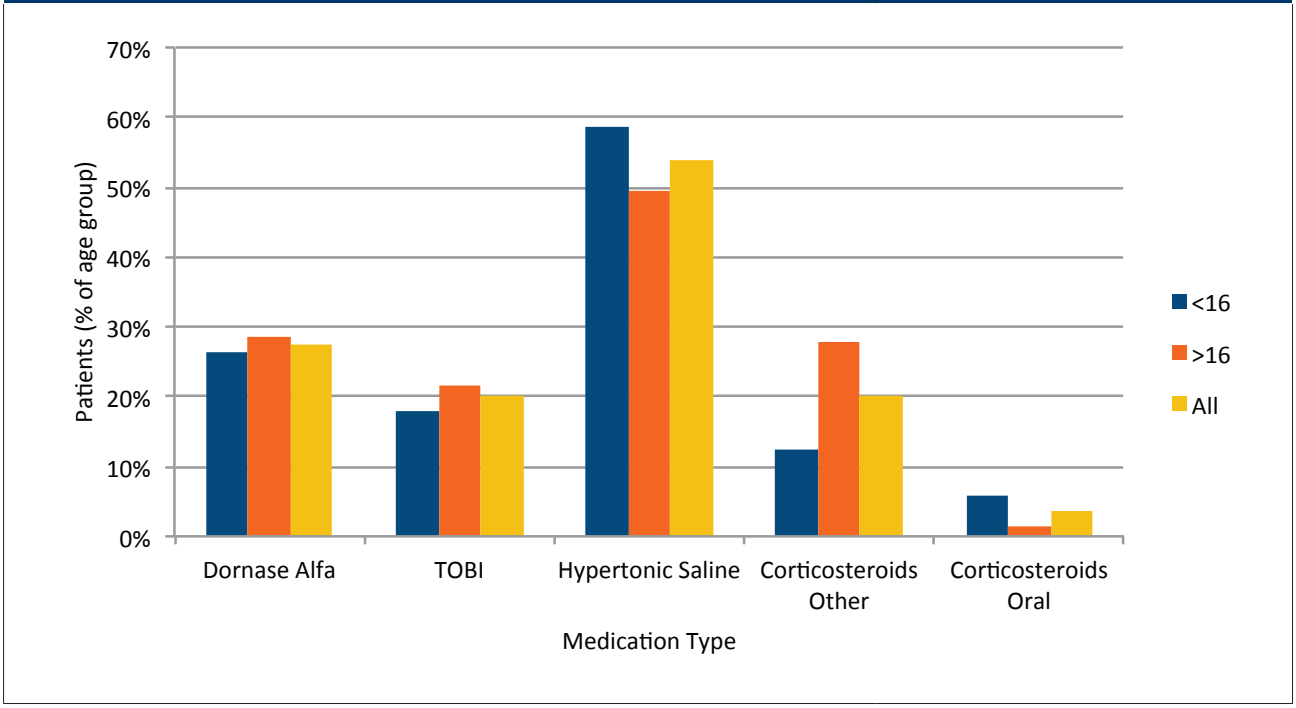


8. Medications

Medications			501 Patients
Medication	**<16**	**>16**	**All**
Hypertonic Saline	58.8%	49.3%	53.7%
Dornase alfa	26.2%	28.7%	27.5%
TOBI	18.0%	21.6%	20.0%
Inhaled Other Antibiotics	0.0%	0.0%	0.0%
Chronic Macrolide	9.9%	34.0%	22.8%
"Corticosteroids Other (Inhaled and combination treatments)"	12.3%	28.0%	20.1%
Corticosteroids Oral	5.7%	1.4%	3.6%
Antifungals	4.7%	3.8%	4.3%
Influenza Vaccine	72.5%	53.90%	63.30%

8.1 Medications Prescribed

501 Patients



NOTE: Our use of inhaled antibiotics, nebulised dornase alfa, and oral chronic macrolide therapy is lower than other international registries, but we are high users of nebulised hypertonic saline.

9. Intravenous Antibiotic Treatment

9.1 Home IV Days

408 Patients

Age	Number In Age Group	Number Patients Who Had IV Days	Percent Patients Who Had IV Days	Mean Days For People Who Have Had IV Days	Mean Days For All Patients
<4	43	2	4.7%	35	1.6
4-7	58	10	17.2%	10.5	1.8
8-11	50	7	14.0%	14.3	2
12-15	54	10	18.5%	26.8	5
16-19	40	6	15.0%	21.8	3.3
20-23	40	11	27.5%	8.7	2.4
24-27	23	10	43.5%	41.7	18.1
28-31	25	13	52.0%	26	13.5
32-35	21	7	33.3%	21.9	7.3
36-39	11	4	36.4%	26.3	9.5
40-43	11	2	18.2%	18	3.3
44-47	12	2	16.7%	9	1.5
48-51	5	1	20.0%	19	3.8
52-55	2	0	0.0%	0	0
56-59	2	0	0.0%	0	0
>=60	11	2	18.2%	8	1.5
Totals	**408**	**87**	**21.3%**	**21.5**	**4.6**

9.2 Hospital IV Days

408 Patients

Age	Number In Age Group	Number Patients Who Had IV Days	Percent Patients Who Had IV Days	Mean Days For People Who Have Had IV Days	Mean Days For All Patients
<4	43	9	20.9%	18.2	3.8
4-7	58	20	34.5%	9.1	3.1
8-15	50	17	34.0%	21.1	7.2
16-18	54	22	40.7%	26.3	10.7
16-19	40	12	30.0%	21.5	6.5
20-23	40	18	45.0%	27.7	12.5
24-27	23	10	43.5%	24.9	10.8
28-31	25	13	52.0%	23.9	12.4
32-35	21	10	47.6%	14.4	6.9
36-39	11	4	36.4%	20.3	7.4
40-43	11	2	18.2%	3	0.5
44-47	12	3	25.0%	25	6.3
48-51	5	1	20.0%	2	0.4
52-55	2	0	0.0%	0	0
56-59	2	0	0.0%	0	0
>=60	11	3	27.3%	12	3.3
Totals	**408**	**144**	**35.3%**	**20.4**	**7.2**

10. Complications

417 Patients

10.1 CF Related Diabetes

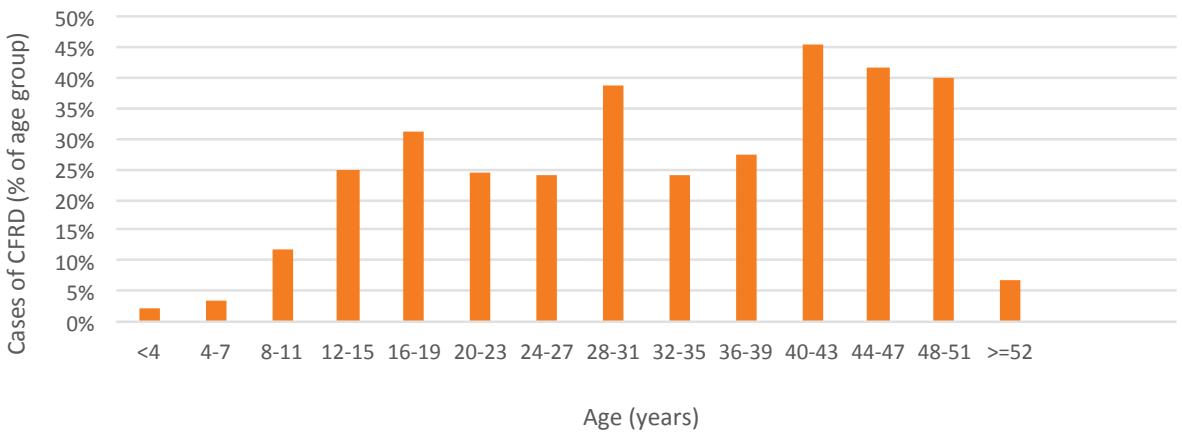
417 Patients

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of CF Population
<4	43	1	2.3%	0.2%
4-7	59	2	3.4%	0.5%
8-11	50	6	12.0%	1.4%
12-15	56	14	25.0%	3.4%
16-19	42	13	31.0%	3.1%
20-23	41	10	24.4%	9.8%
24-27	25	6	24.0%	6.0%
28-31	26	10	38.5%	6.2%
32-35	21	5	23.8%	5.0%
36-39	11	3	27.3%	2.6%
40-43	11	5	45.5%	2.6%
44-47	12	5	41.7%	2.9%
48-51	5	2	40.0%	1.2%
52-55	2	0	0.0%	0.5%
56-59	2	0	0.0%	0.5%
>=60	11	1	9.1%	2.6%

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of CF Population
<16	208	23	11.1%	5.5%
>16	209	60	28.7%	14.4%
Total	**417**	**83**	**19.9%**	

10.2 CF Related Diabetes by Age

417 Patients



The prevalence of CFRD has increased over time in the data registry data from 13.5% in 2012 to 19.9% this year. This may reflect; (i) the use of more sensitive measurement of abnormal glucose abnormalities with continuous glucose monitoring; (ii) a recognition that instituting insulin therapy earlier when abnormalities first occur is associated with better intermediate outcomes; and (iii) the increasing capture of data from adults with CF over the time the Registry has been in place.

10.3 Liver Function by Ultra Sound

411 Patients

		Normal		Abnormal		Not Done	
	Number in age group	Number of patients	Percent of patients	Number of patients	Percent of patients	Number of patients	Percent of patients
Paediatrics	207	70	33.8%	26	12.6%	111	53.6%
Adults	204	22	10.8%	6	2.9%	176	86.3%
Total	**411**	**92**	**22.4%**	**32**	**7.8%**	**287**	**69.8%**

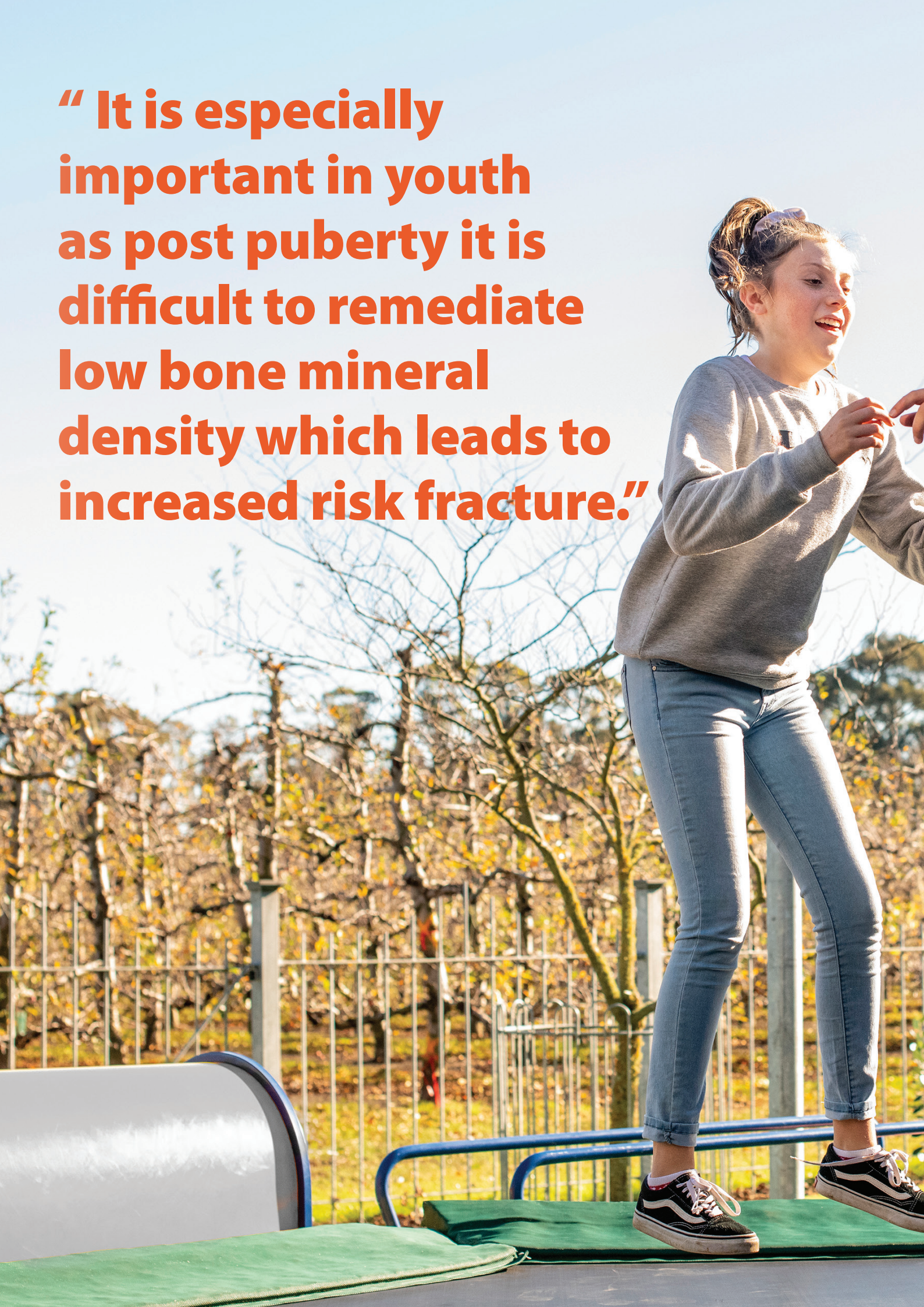
This is the first time we are presenting this data in the report. The 'not done' is because abdominal ultrasound has not been done in the current year. In children the recommended protocol is to do an abdominal ultrasound at 3, 6, 9 and 12 years of age, unless there is additional concern. From 12 years on the recommendation is annually. In adults abdominal ultrasounds are done far less often and usually in response to a new noted event.

10.4 Bone Density by DEXA Scans

411 Patients

		Normal		Abnormal		Not Done	
	Number in age group	Number of patients	Percent of patients	Number of patients	Percent of patients	Number of patients	Percent of patients
Paediatrics	207	20	9.7%	9	4.3%	178	86.0%
Adults	204	36	17.6%	29	14.2%	139	68.1%
Total	**411**	**56**	**13.6%**	**38**	**9.2%**	**317**	**77.1%**

This is the first time we are presenting this data in the report. The current recommendation is to do this at age 10 years and then every two years unless there are other concerns (a previously low bone mineral density result, development of CF related diabetes, poor nutrition, prolonged use of oral steroids, a fracture sustained, persistently low vitamin D) all of which are risk factors for poorer bone mineral density. It is concerning that so many have been listed as 'not done' and this may lend itself to a future audit to determine reasons, or that they are not being recorded. It is especially important in youth as post puberty it is difficult to remediate low bone mineral density which leads to increased fracture risk.



“ It is especially important in youth as post puberty it is difficult to remediate low bone mineral density which leads to increased risk fracture.”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