Port CF 2017

New Zealand CF Data Registry

CYSTIC FIBROSIS NZ

Contents

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 registry.

Suggested citation:
Port CF Data Registry, 2017 Registry Report, Cystic Fibrosis NZ. www.cfnz.org.nz/what-we-do/port-cf-data-registry

The Port CF Data Registry is a research project of Cystic Fibrosis NZ. For further information visit www.cfnz.org.nz.

Introduction

From the Chair of the Port CF Steering Commmittee

Associate Professor Cass Byrnes
Chair & Port CF Principal Investigator

Jane Bollard
Chief Executive, Cystic Fibrosis NZ

Cystic Fibrosis NZ and the Port CF Steering Committee are pleased to present the Port CF 2017 report - the New Zealand CF data registry with data collected from children, young persons and adults with cystic fibrosis 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 database and data entry.
- The Nurses, Specialists and Administrators who have worked to enter data, enabling a detailed analysis for NZ.
- Canterbury District Health Board for its ongoing information technology service to maintain the registry.

This seventh registry report gives an ever 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 more people had 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 2017 registry represents the most data captured on our population with cystic fibrosis, 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

Assoc. Prof Cass Byrnes (Chair)
Starship Child Health &
University of Auckland

Dr Richard Laing
Christchurch Hospital, Christchurch

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A special thanks to:
Andrew Watson - Canterbury District Health Board and 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 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 opinion 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 498 registered in Port CFNZ, not all individuals had an input for all questions. While the total is 498 (219 children <16 year years, 279 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 supplemental feeding a total response was obtained from 399 patients (198 children and 201 adults) on page 18 (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 of Cystic Fibrosis NZ or to the Project Co-ordinator Jan Tate.

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Glossary of terms

CFNZ	Cystic Fibrosis New Zealand
FEV1	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 CF	New Zealand's CF data registry

CF at a glance

- **There are 501 patients in the registry, around 97-98% of all New Zealanders with CF.**

  153 MEN **125** WOMEN **224** CHILDREN

- **Of the 501 patients in the registry, 68% are in the North Island and 32% in the South Island.**

  35% NI PAEDS **33%** NI ADULTS **13%** SI PAEDS **19%** SI ADULTS

- **Paediatric median BMI percentile**

  60.0 MALES **43.9** FEMALES

- **Adult median BMI percentile**

  22.8 MALES **22.5** FEMALES

- **Median FEV1**

  79.8 MEN **76.6** WOMEN **99.7** CHILDREN

97-98%

New Zealanders with CF are in the registry

49.2%

of people with CF have the F508del / F508del genotype

64%

of people with CF are using Hypertonic Saline

347

annual reviews recorded

25%

of people with CF received the influenza vaccine

18.27

is the median age of patients in the registry

15

new diagnoses for babies under one year of age

53.5%

of people with CF recorded as having Staphylococcus Aureus

30%

of adults list exercise as their primary form of airway clearance

31.4%

of adults have CF related diabetes, with 11.2% for paediatrics

5203

total hospital and home IV days across 161 patients

Summary of key indicators

Summary of Key Indicators						
	2017	**2016**	**2015**	**2014**	**2013**	**2012**
CF Patients Registered	498	501	449	443	444	423
Diagnosis						
Diagnosis age <1 year	15	6	5	7	5	11
Diagnosis age 1- 15 years	2	3				
Diagnosis age >=16 years	1	2	0	2	3	2
Age						
Median Age (in years)	18.27	17.38	18.25	18.11	17.55	16.15
Mean Age (in years)	20.79	20.04				
PWCF <16 years						
Number	279	233	192	196	205	209
Percent	56.0%	46.5%	42.8%	44.2%	46.2%	49.4%
PWCF >16 years						
Number	219	268	257	247	239	214
Percent	44.0%	53.5%	57.2%	55.8%	53.8%	50.6%
Gender						
Males						
Number	273	275	247	240	240	228
Percent	54.9%	54.9%	55.0%	54.2%	54.1%	53.0%
Females						
Number	224	226	202	203	204	195
Percent	45.1%	45.1%	45.0%	45.8%	45.9%	46.1%
Genotyped						
Number	484	451	400	429	426	407
Percent	97.4%	90.0%	89.1%	96.8%	95.9%	96.2%
FEV1 (% predicted)						
Mean	85.1%	85.0%				
Median	86.5%	88.4%	85.6%	85.1%	84.3%	84.5%
FEV1 < 16 Years						
Mean	96.8%	97.3%				
Median	99.3%	99.3%	98.9%	97.7%	96.6%	97.2%
FEV1 > 16 Years						
Mean	72.6%	72.6%				
Median	77.4%	77.4%	77.0%	78.0%	70.7%	70.6%
FEV1 < 18 Years						
Mean	95.1%	95.0%				
Median	98.3%	98.0%				
FEV1 > 18 Years						
Mean	72.2%	71.2%				
Median	75.6%	75.1%				

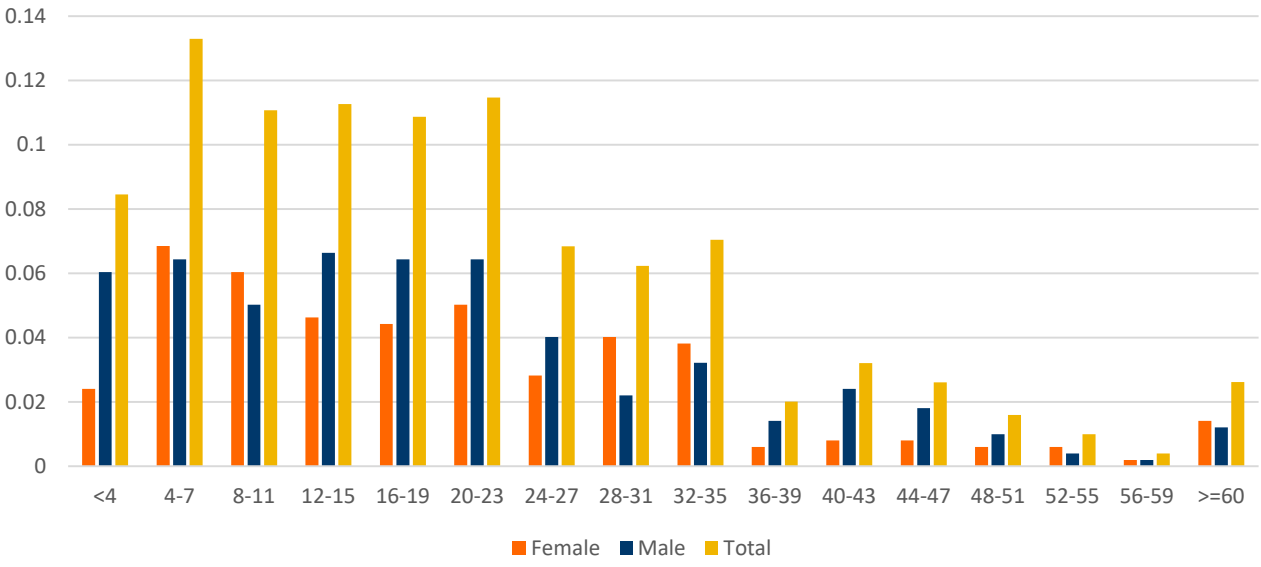
1. Demographics

398 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	42	8.5%	30	6.0%	12	2.4%
4-7	66	13.3%	32	6.4%	34	6.8%
8-11	55	11.1%	25	5.0%	30	6.0%
12-15	56	11.3%	33	6.6%	23	4.6%
16-19	55	10.9%	32	6.4%	22	4.4%
20-23	57	11.5%	32	6.4%	25	5.0%
24-27	34	6.8%	20	4.0%	14	2.8%
28-31	31	6.2%	11	2.2%	20	4.0%
32-35	35	7.0%	16	3.2%	19	3.8%
36-39	10	2.0%	7	1.4%	3	0.6%
40-43	16	3.2%	12	2.4%	4	0.8%
44-47	13	2.6%	9	1.8%	4	0.8%
48-51	8	1.6%	5	1.0%	3	0.6%
52-55	5	1.0%	2	0.4%	3	0.6%
56-59	2	0.4%	1	0.2%	1	0.2%
>=60	13	2.6%	6	1.2%	7	1.4%
Total	**498**	**100.0%**	**273**	**54.9%**	**224**	**45.1%**
Median	**18.27**					
Range	**0.09 - 74**					

1.1 Age distribution

398 patients



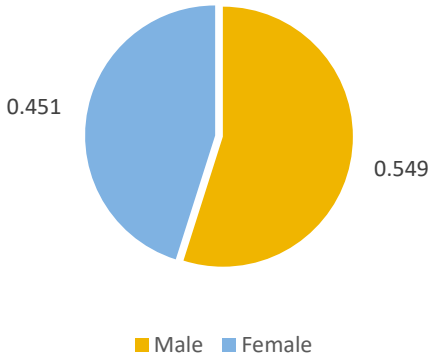
The median age of persons with CF in New Zealand has increased from 16 to 18 years over the six years that we have had national registry data. 'Children' in international registries are defined as either up to 16 years or up to 18 years of age. In New Zealand if we include children as being up to 16 years we have 219 children (44.1% of total) and 278 adults (55.9% of total), if we include children as being up to 18 years, which is our more usual clinical practice, we have 247 children (49.7% of total) and 250 adults (50.3% of total).

1.2 Gender distribution

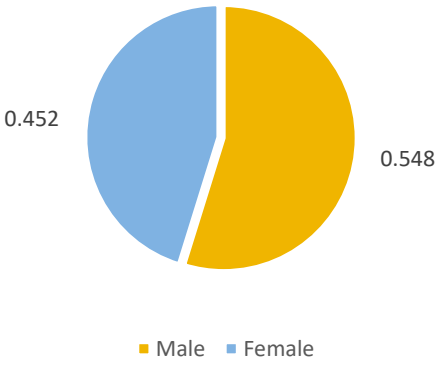
497 patients

	All		<16		>16	
	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	273	54.9%	120	54.8%	153	55.0%
Female	224	45.1%	99	45.2%	125	45.0%
Totals	**497**		**219**		**278**	

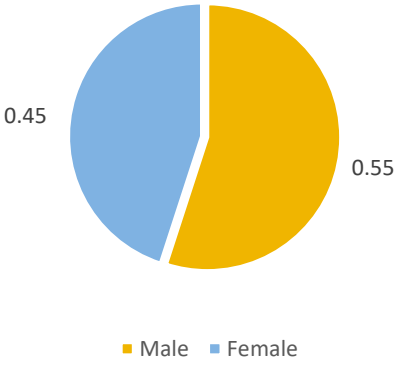
Gender Distribution of All Patients **497 Patients**



Gender Distribution <16 Years **219 Patients**



Gender Distribution >16 Years **278 Patients**



“The median age of persons with CF in New Zealand has increased from 16 to 18 years over the six years that we have had national registry data.”

2. Genotypes

484 patients

Mutations	Number of Patients Genotyped	Percentage of Patients Genotyped	
Homozygous F508del	238	49%	
Heterozygous F508del	185	38%	
No F508del or both unidentified	61	13%	
Total	**484**		

2.1 Second Allele of Heterozygous F508del

185 patients

Second Allele	c.DNA Name	Number of Patients	Percent of Patients
G542X	c.1624G>T	22	4.5%
G551D	c.1652G>A	17	3.5%
R117H	c.350G>A	15	3.1%
G85E	c.254G>A	5	1.0%
^1507	c.1519_1521delATC	4	0.8%
N1303K	c.3909c>G	3	0.6%
621+1G->T	c.489+1G>T	3	0.6%
1717-1G->A	c.1585-1G>A	2	0.4%
3849+10kbC->T	c.3717+12191C>T	2	0.4%
1898+1G->A	c.1766+1G>A	2	0.4%
A455E	c.1364C>A	2	0.4%
1078delT	c.948delT	2	0.4%
D1152H	c.3454G>C	2	0.4%
R334W	c.1000C>T	2	0.4%
Q493X	c.1477C>T	1	0.2%
2789+5G->A	c.2657+5G>A	1	0.2%
3120+1G->A	c.2988+1G>A	1	0.2%
3659delC	c.3528delC	1	0.2%
712-1G->T	c.580-1G>T	1	0.2%
R347H	c.1040G>A	1	0.2%
R347P	c.1040G>C	1	0.2%
R560T	c.1679G>C	1	0.2%
W1282X	c.3846G>A	1	0.2%
R1158X	c.3472C>T	1	0.2%
R553X	c.1657C>T	0	0.0%
Other genetic mutation		75	15.5%
Second mutation unknown		17	3.5%

2.2 No F508del Mutations 61 patients

	G542X	G551D	G85E	Other	R1162X	R117H	1717-1G->A	Not Identified
G542X	1			2				2
G551D	1	1		3		4	1	1
N1303K		1		1				
Other			1	19	1			2
Q493X		2				1		
R553X		1						
3849+10kbC->T							1	
W1282X				1				1
Not Identified		1				2		10

2.3 Genotype Major Categories 484 patients

Mutation	Patients Identified	Percentage of Patients
F508del	423	87.4%
G551D	33	6.8%
G542X	28	5.8%
R117H	22	4.5%
G85E	6	1.2%

Note: Because people have two genes, patients are counted twice, once for each gene.

The total number of patients is 484.

Our high percentage of F508del is in keeping with the international registries from European derived populations. In total only 61 persons in New Zealand do not have at least one F508 mutation. Our newborn screening programme looks for 3 genes (following an initial high immunoreactive trypsin). Looking at the gene mutations' recorded in the 2017 registry, 45 persons (9%) would not be detected by our current newborn screening programme. Although some of the 'unidentified' or 'unknown' may be in older adults who have not had genetic screening or had it many years ago when one, or a few, genes were tested. From this data we can also see that 28 (nearly 6%) would be amenable for ivacaftor treatment.

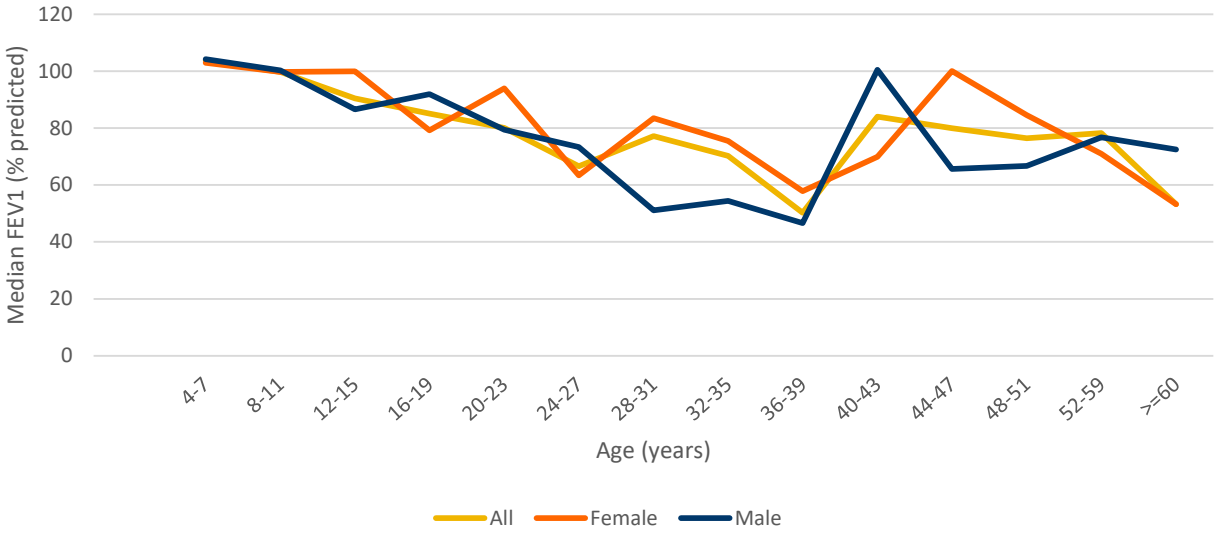
3. Respiratory

357 patients

Age Group	All		Female		Male	
	Number in age group	Median FEV1	Number in age group	Median FEV1	Number in age group	Median FEV1
4-7	44	103.7	19	102.9	25	104.2
8-11	51	99.7	28	99.7	23	100.3
12-15	47	90.4	19	99.9	28	86.6
16-19	42	85.1	14	79.2	28	91.9
20-23	41	80.1	14	94	27	79.4
24-27	25	66.6	9	63.4	16	73.3
28-31	24	77.2	18	83.5	6	51.1
32-35	28	70.2	15	75.5	13	54.4
36-39	8	50.3	2	57.9	6	46.6
40-43	14	84	4	69.9	10	100.5
44-47	12	80	4	100.1	8	65.7
48-51	6	76.4	2	84.6	4	66.7
52-55	4	67.9	3	71	1	64.9
56-59	1	88.6	0	-	1	88.6
>=60	10	53.2	6	53.2	4	72.5
Totals	**357**		**157**		**200**	

3.1 Median FEV1 (% predicted) >6

357 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 86.5% (99.3% in children, 77.4% 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 phenotype.

“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

197 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	10	55.9	<4	24	64.2	<4	34	64.2
4-7	31	53.3	4-7	31	67.8	4-7	62	68.9
8-11	28	32	8-11	24	56.6	8-11	52	57.4
12-15	19	34.5	12-15	30	51.4	12-15	49	55.1
Totals	**88**			**109**			**197**	

4.2 Adult BMI

218 patients

Females >16			Males >16			All >16		
	BMI			BMI			BMI	
Age group	Number in group	Median BMI	Age group	Number in group	Median BMI	Age group	Number in group	Median BMI
16-19	15	23.4	16-19	29	21.4	16-19	44	22.4
20-23	14	23.9	20-23	27	21.4	20-23	41	21.9
24-27	9	20.7	24-27	16	22.8	24-27	25	21.8
28-31	18	21.5	28-31	6	25.1	28-31	24	21.6
32-35	15	22.1	32-35	13	22.1	32-35	28	22.1
36-39	2	29.4	36-39	6	26	36-39	8	26
40-43	4	22.5	40-43	10	25.7	40-43	14	25.4
44-47	4	24.8	44-47	8	24.1	44-47	12	24.1
48-51	2	26.5	48-51	4	25.4	48-51	6	25.5
52-55	3	26.3	52-55	1	26.5	52-55	4	26.4
56-59	0		56-59	1	32.6	56-59	1	32.6
>=60	7	28.5	>=60	4	21	>=60	11	21.8
Totals	**93**			**125**			**218**	

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:

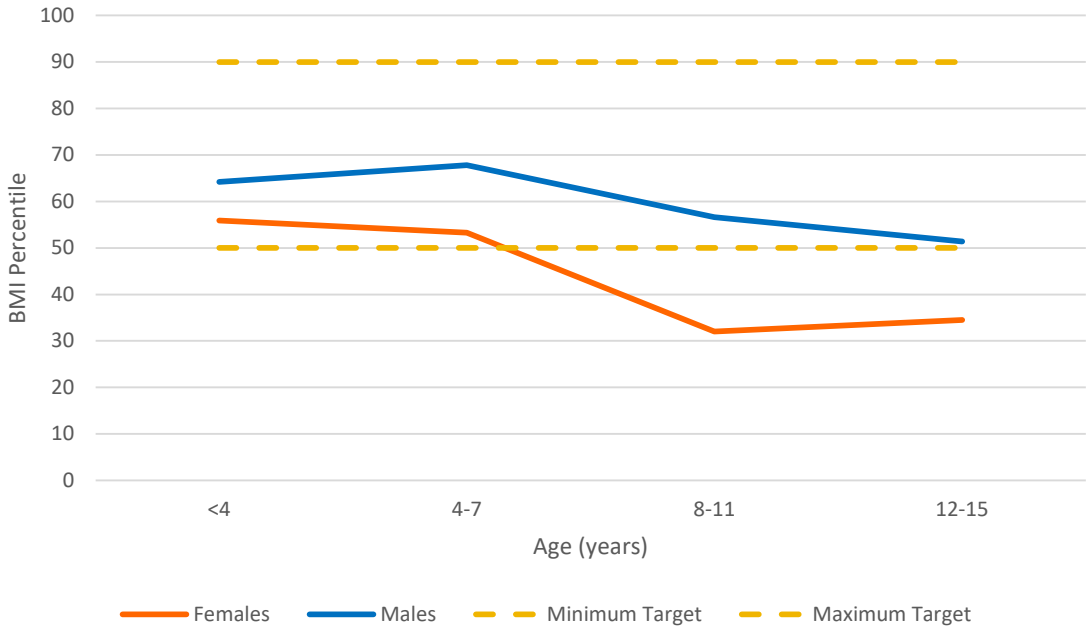
1) Infants (0 to 24 months): weight-for-length ≥50th percentile using WHO growth charts
2) Children and Adolescents (2-18 years): BMI 50-85th percentile (CDC growth charts) or 50-91st percentile (WHO growth charts)
3) Adults: males BMI 23 - 27 kg/m2, females BMI 22 - 27 kg/m2

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

For infants <4 years of age the median BMI is 64.2 percentile. For children and adolescents the median BMI is 63.2 percentile. For adults 48% of males and 54.8% of females are above the minimum target range.

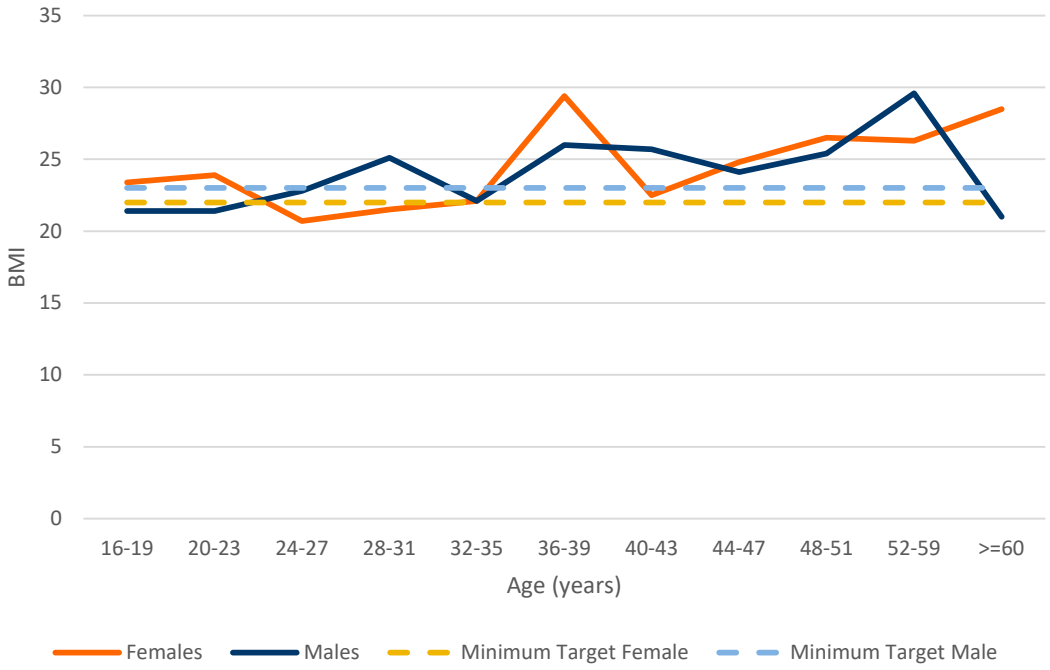

Creamy
Rice

4.3 Median BMI Percentile <16 197 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 >16 218 patients

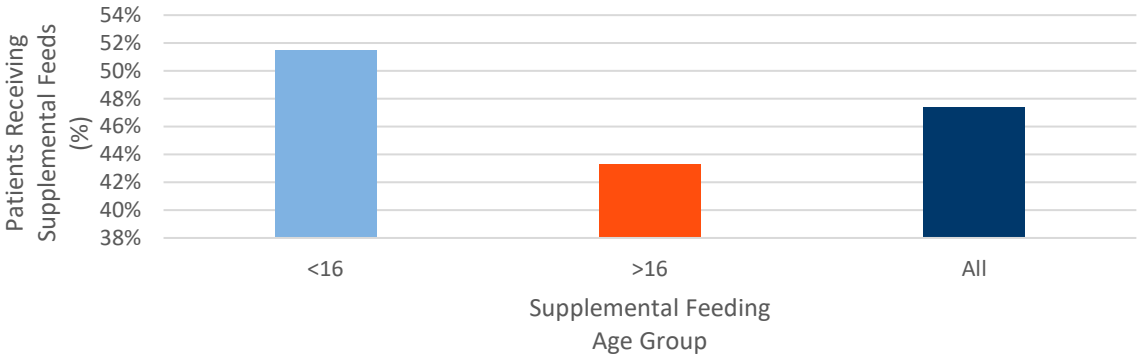


The Optimal BMI for women is 22 - 27 and the dotted yellow line shows the minimum BMI for women in the target range. The optimal BMI for men is 23 - 27. The dotted blue line shows the minimum BMI for men in the target range.

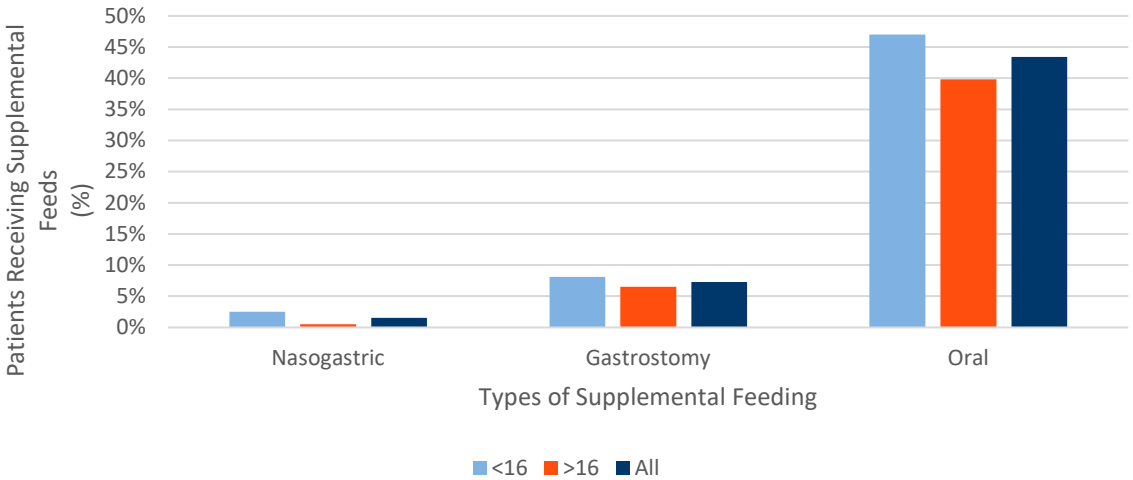
4.5 Supplemental feeding 399 patients

	<16 years, n = 198	
	Yes	**% <16 years supplemented**
Supplemental Feeding	102	51.5%
Nasogastric	5	2.5%
Gastrostomy	16	8.1%
Oral	93	47.0%
	>16 years, n = 201	
	Yes	**% >16 years supplemented**
Supplemental Feeding	87	43.3%
Nasogastric	1	0.5%
Gastrostomy	13	6.5%
Oral	80	39.8%

4.6 Supplemental feeding by age group 399 patients



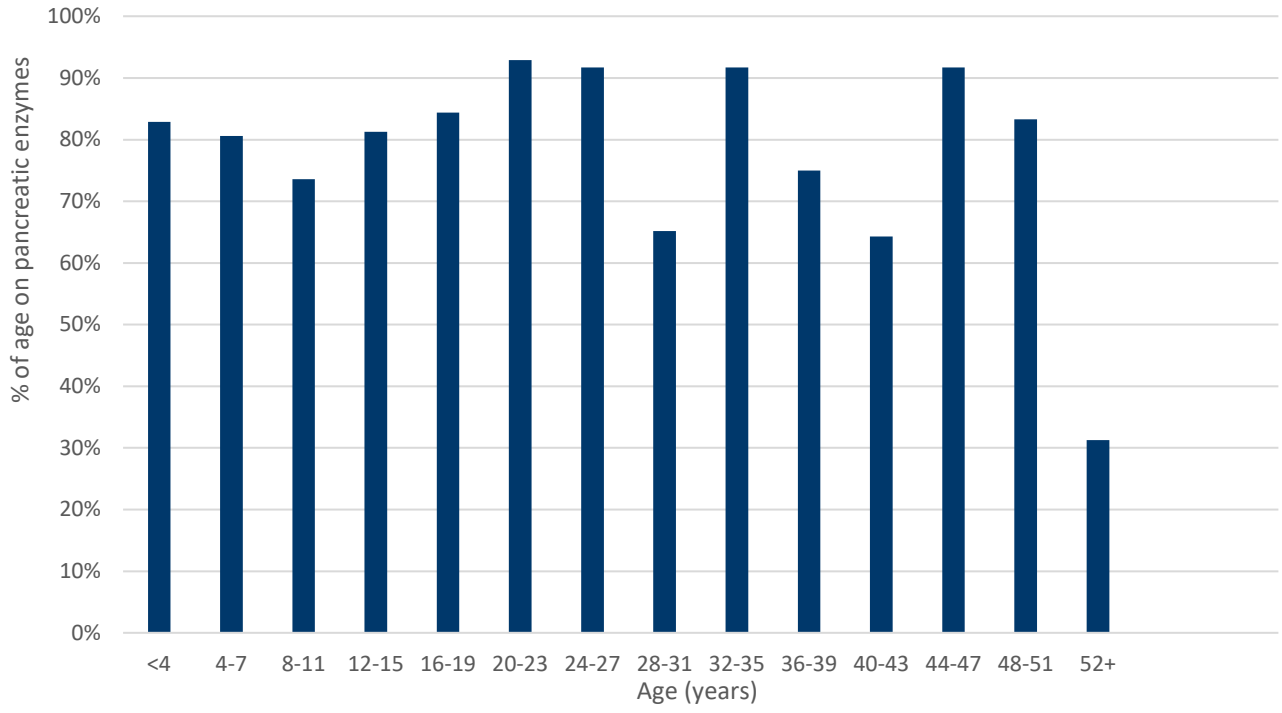
4.7 Supplemental feeding by type 399 patients

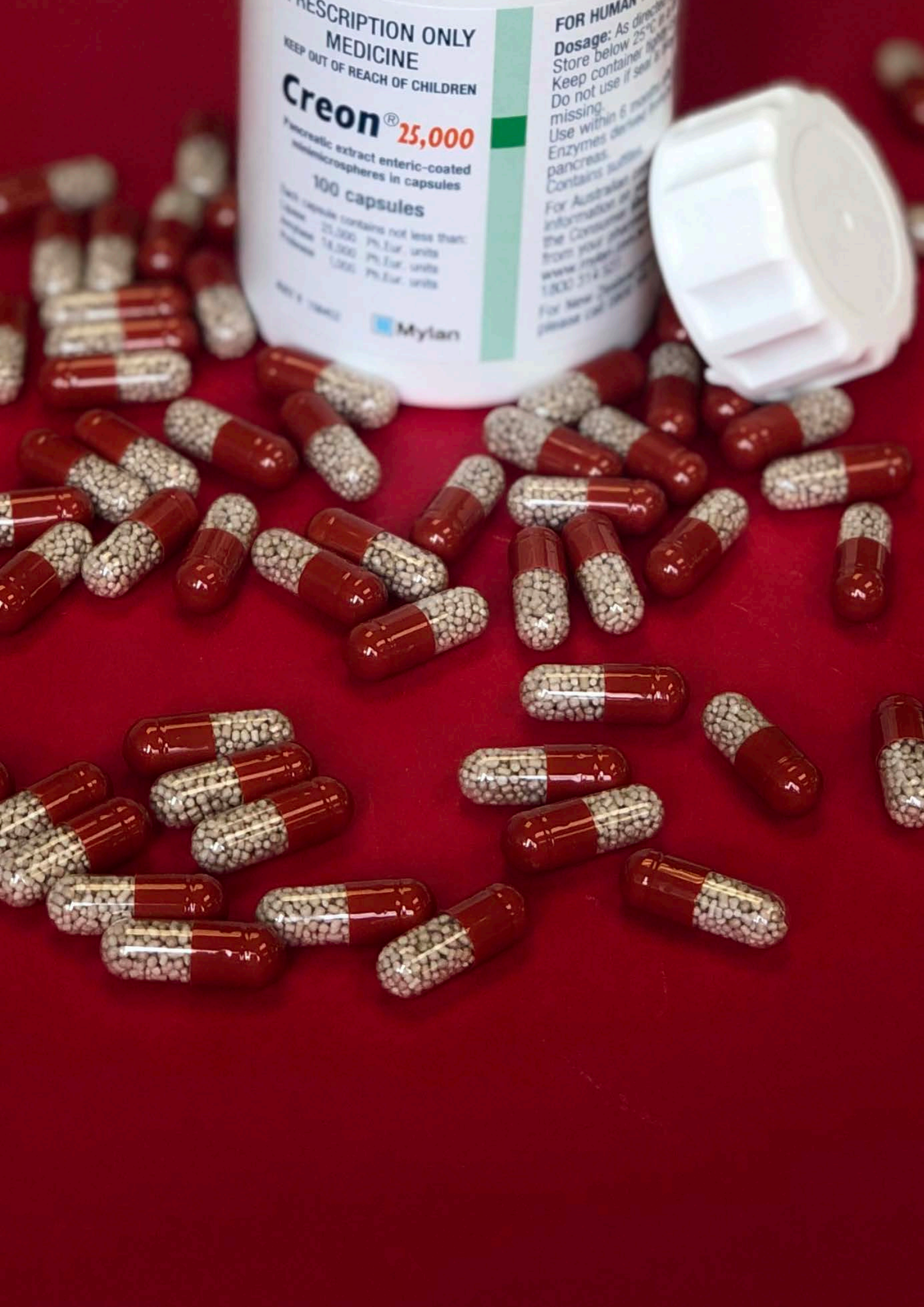


5. Pancreatic enzymes 418 patients

Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of CF population
<4	41	34	82.9%	8.1%
4-7	62	50	80.6%	12.0%
8-11	53	39	73.6%	9.3%
12-15	48	39	81.3%	9.3%
16-19	45	38	84.4%	9.1%
20-23	42	39	92.9%	9.3%
24-27	24	22	91.7%	5.3%
28-31	23	15	65.2%	3.6%
32-35	24	22	91.7%	5.3%
36-39	8	6	75.0%	1.4%
40-43	14	9	64.3%	2.2%
44-47	12	11	91.7%	2.6%
48-51	6	5	83.3%	1.2%
52-55	4	0	0.0%	0.0%
56-59	1	0	0.0%	0.0%
>=60	11	5	45.5%	1.2%
Totals	**418**	**334**		**80%**

5.1 Patients on pancreatic enzymes 418 patients

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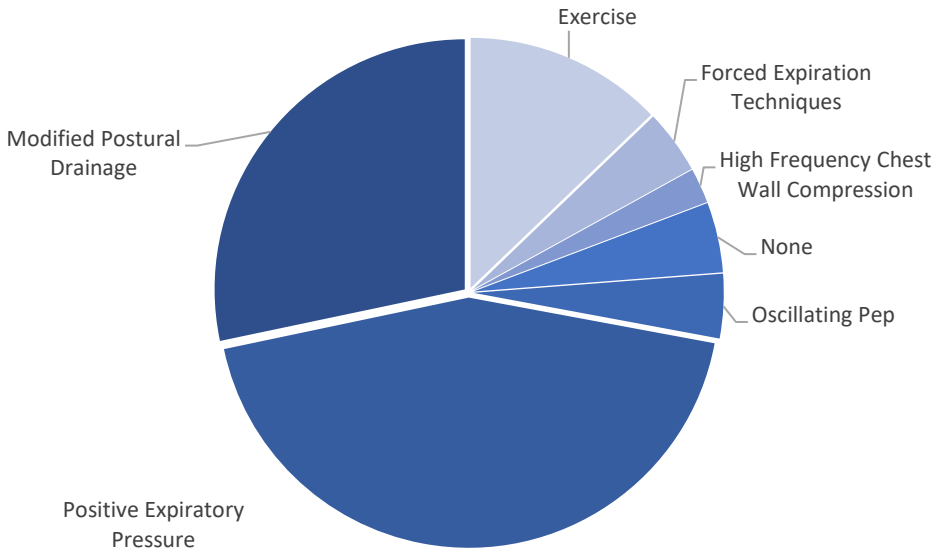
Pancreatic extract enteric-coated
minimicrospheres in capsules
100 capsules
Mylan

6. Airway clearance techniques

6.1 Primary airway clearance — 428 patients

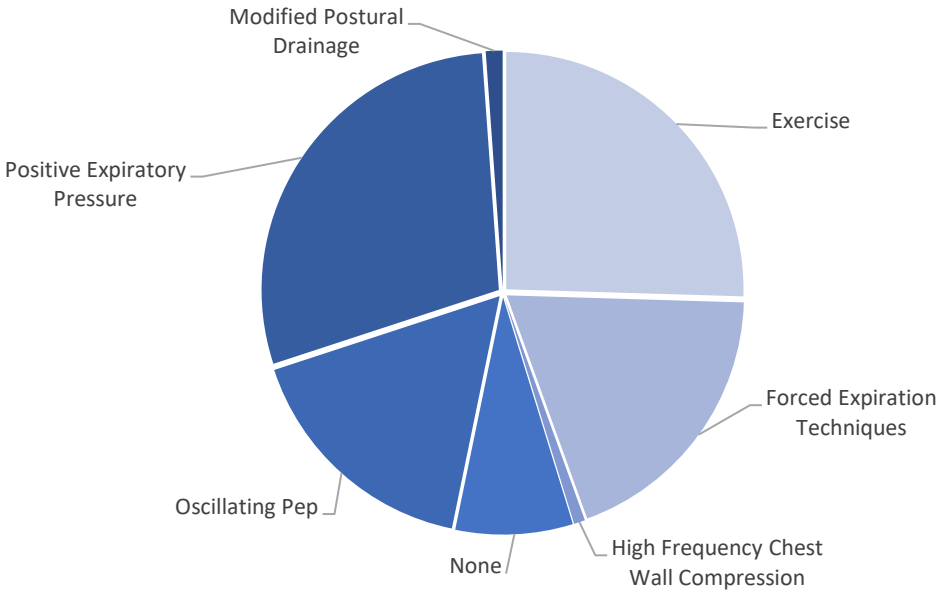
	<16 years, n = 205	
	Number of patients	Percent of patients
Positive Expiratory Pressure	116	56.6%
Modified Postural Drainage	75	36.6%
Exercise	34	16.6%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	11	5.4%
Forced Expiration Techniques	11	5.4%
(e.g. huff cough, active cycle breathing, autogenic drainage)		
High Frequency Chest Wall Compression (e.g. vest)	6	2.9%
None	12	5.9%
	>16 years, n = 223	
	Number of patients	Percent of patients
Positive Expiratory Pressure	76	34%
Modified Postural Drainage	3	1%
Exercise	67	30%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	44	19.7%
Forced Expiration Techniques	50	22.4%
(e.g. huff cough, active cycle breathing, autogenic drainage)		
High Frequency Chest Wall Compression (e.g. vest)	2	0.9%
None	21	9.4%

6.2 Paediatric primary airway clearance — 205 patients



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

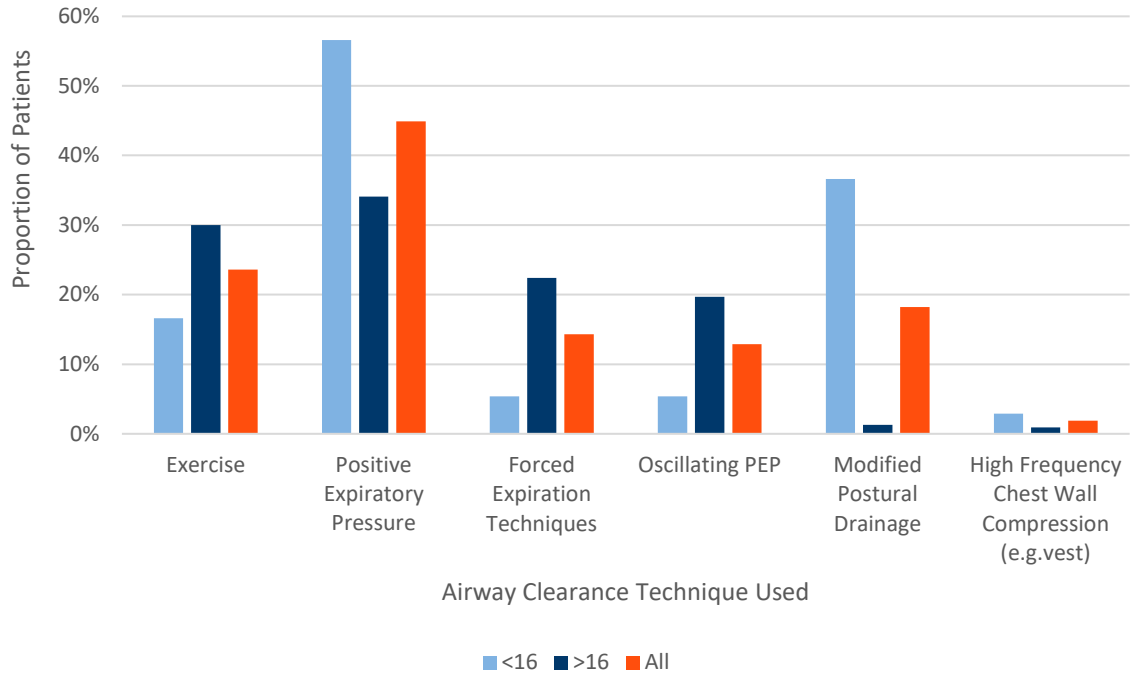
6.3 Adult primary airway clearance 223 patients



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

The nominated primary airway clearance technique adds up to greater than 100%. This is because over the clinic visits for a year an individual may use differing techniques as his/her main option at different times. Over the years of the registry reports there is a trend for an increased percentage of children to have nominated 'none' as their primary airway clearance (from 2.3% in 2013 to 5.9% in 2017) while the trend is the opposite for the adults (18.8% in 2013 to 9.4% in 2017). There has been an increasing percentage in both children and adults that now nominate 'Positive Expiratory Pressure' and 'Exercise' as their preferred option.

6.4 Secondary airway clearance 428 patients



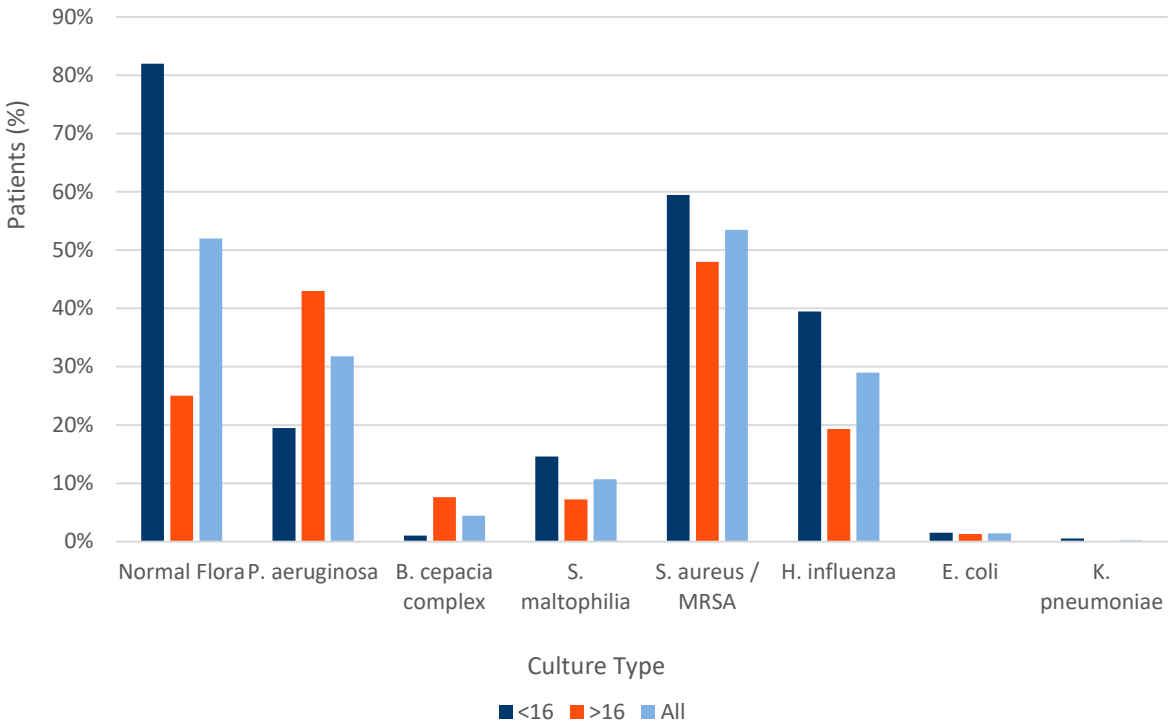
7. Microbiology

428 patients

	<16 years, n = 205		>16 years, n = 223		All, n = 428	
	Number	Percent	Number	Percent	Number	Percent
Normal Flora	168	82.0%	56	25.0%	224	52.0%
H. Influenza	81	39.5%	43	19.3%	124	29.0%
E. Coli	3	1.5%	3	1.3%	6	1.4%
K. Pneumoniae	1	0.5%	0	0.0%	1	0.2%
Stenotrophomonas Maltophilia	30	14.6%	16	7.2%	46	10.7%
Pseudomonas Aeruginosa	40	19.5%	96	43.0%	136	31.8%
Mucoid	11	5.4%	72	32.3%	83	19.4%
Non Mucoid	28	13.7%	57	25.6%	85	19.9%
Staphylococcus Aureus	122	59.5%	107	48.0%	229	53.5%
MSSA	114	55.6%	97	43.5%	211	49.3%
MRSA	8	3.9%	10	4.5%	18	4.2%
Burkholderia Cepacia Complex	2	1.0%	17	7.6%	19	4.4%
Cenocepacia	0	0.0%	2	0.9%	2	0.5%
Multivorans	2	1.0%	7	3.1%	9	2.1%
Other	2	1.0%	0	0.0%	2	0.5%

7.1 Culture prevalence

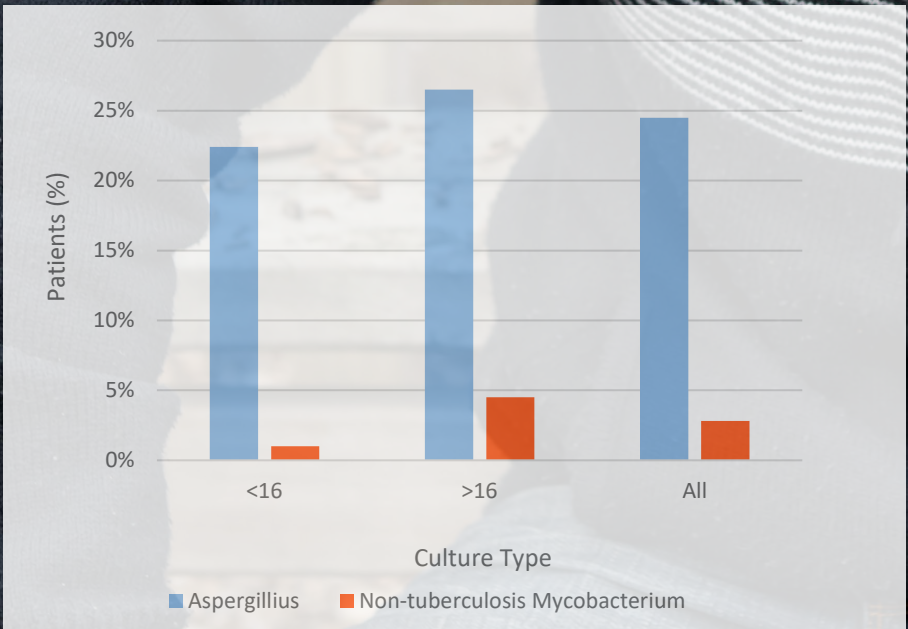
428 patients



The percentages of population with CF having had specific respiratory pathogens identified such as Staphylococcal aureus, Pseudomonas aeruginosa etc are very similar to the percentages presented in the Australian 2017 registry, with the exception of much higher percentages of Haemophilus influenza here. This pathogen is also higher in our young children and lower in our adults which is in keeping with the usual age that Haemophilus influenza causes infection in the general population. Pseudomonas aeruginosa is found in 19.5% of the children and increases to 43% in adults. Our MRSA rates are relatively low at 4.2% overall.

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

7.2 Nonbacterial/fungal prevalence 428 patients



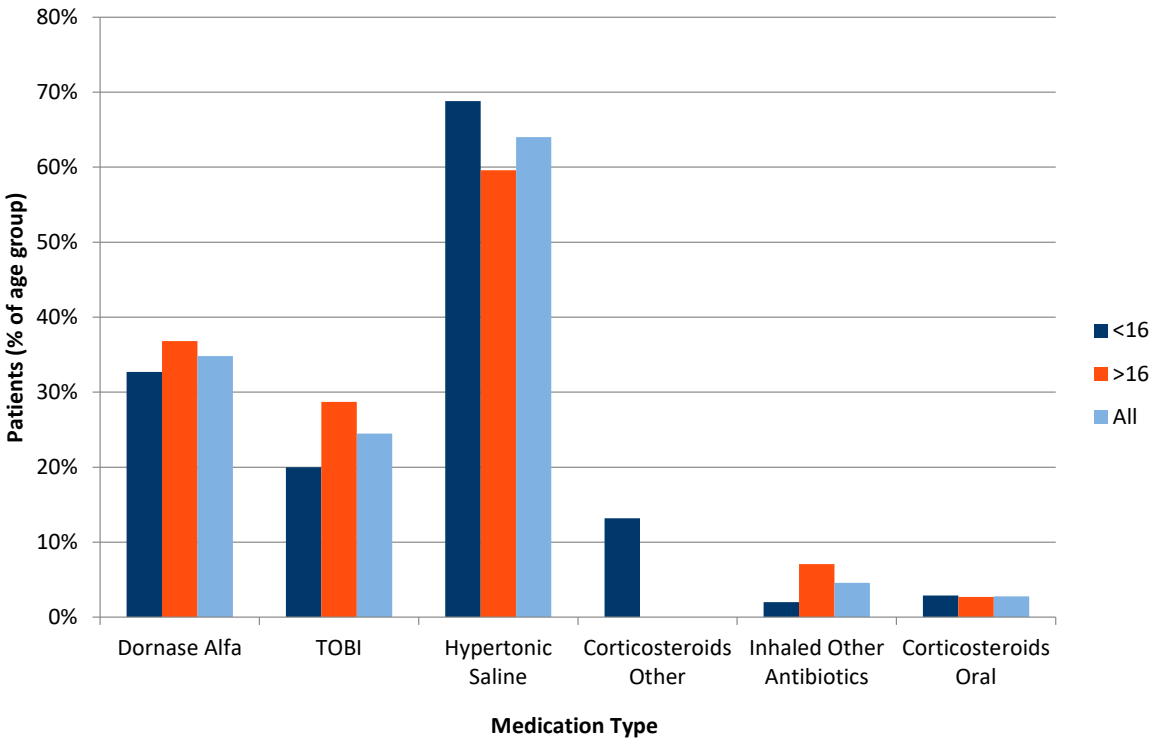
8. Medications

428 patients

Medication	<16	>16	All
Hypertonic Saline	68.8%	59.6%	64.0%
Dornase alfa	32.7%	36.8%	34.8%
TOBI	20.0%	28.7%	24.5%
Inhaled Other Antibiotics	2.0%	7.1%	4.6%
Chronic Macrolide	8.3%	39.9%	24.8%
Corticosteroids Other (Inhaled and combination treatments)	26.3%	55.2%	41.4%
Corticosteroids Oral	2.9%	2.7%	2.8%
Antifungals	4.4%	2.2%	3.3%
Influenza Vaccine	77.1%	51.10%	63.60%

8.1 Medications prescribed

428 patients



Inhaled other - This reflects nebulised Colistin, Tobramycin (intravenous solution), Gentamicin and Aztreonam.

We have listed more medication types here than we have in previous reports. 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. We also have no access to some newer medications except on research programmes – notably the gene modulater agent.

9. Intravenous antibiotic treatment

9.1 Home

418 patients

Age	Number In Age Group	Number 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	41	4	10%	8	0.8
4-7	62	8	13%	12.8	1.6
8-11	53	11	21%	15.7	3.3
12-15	48	12	25%	26.5	6.6
16-19	45	8	18%	25.5	4.5
20-23	42	7	17%	16.4	2.7
24-27	24	7	29%	39.7	11.6
28-31	23	8	35%	20.5	7.1
32-35	24	5	21%	23.2	4.8
36-39	8	2	25%	13.5	3.4
40-43	14	3	21%	16.7	3.6
44-47	12	4	33%	8.8	2.9
48-51	6	1	17%	9	1.5
52-55	4	0	0%	0	0
56-59	1	0	0%	0	0
>=60	11	0	0%	0	0
Totals	**418**	**80**	**19%**	**18.2**	**3.9**

9.2 Hospital IV stays

418 patients

Age	Number In Age Group	Number 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	41	9	22%	19.9	4.4
4-7	62	17	27%	12.9	3.5
8-15	53	18	34%	19.4	6.6
16-18	48	20	42%	26.4	11
16-19	45	19	42%	28.9	12.2
20-23	42	14	33%	22.9	7.6
24-27	24	13	54%	43.5	23.5
28-31	23	10	43%	20.6	9
32-35	24	9	38%	17.1	6.4
36-39	8	3	38%	20.3	7.6
40-43	14	3	21%	11.3	2.4
44-47	12	5	42%	9.8	4.1
48-51	6	1	17%	5	0.8
52-55	4	0	0%	0	0
56-59	1	0	0%	0	0
>=60	11	2	18%	24	4.4
Totals	**418**	**143**	**34%**	**20.1**	**7.4**

10. Complications

10.1 CF related diabetes

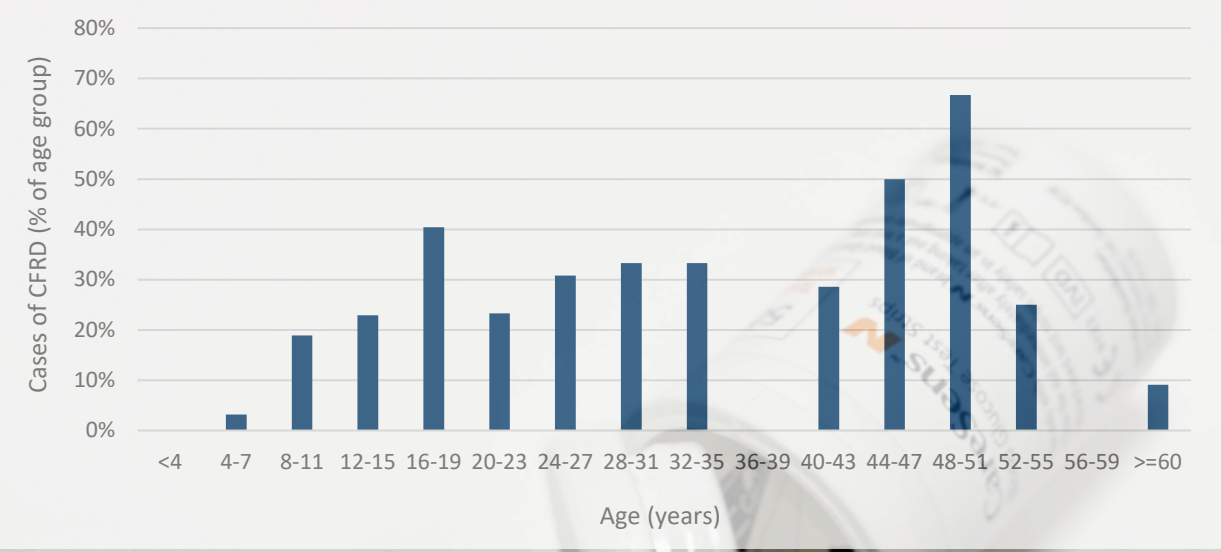
426 patients

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of CF Population
<4	42	0	0.0%	0.0%
4-7	63	2	3.2%	0.5%
8-11	53	10	18.9%	2.3%
12-15	48	11	22.9%	2.6%
16-19	47	19	40.4%	4.5%
20-23	43	10	23.3%	2.3%
24-27	26	8	30.8%	1.9%
28-31	24	8	33.3%	1.9%
32-35	24	8	33.3%	1.9%
36-39	8	0	0.0%	0.0%
40-43	14	4	28.6%	0.9%
44-47	12	6	50.0%	1.4%
48-51	6	4	66.7%	0.9%
52-55	4	1	25.0%	0.2%
56-59	1	0	0.0%	0.0%
>=60	11	1	9.1%	0.2%
Age Group	**Number in group**	**Number with CFRD**	**Percent of age group**	**Percent of CF Population**
<16	206	23	11.2%	5.4%
>16	220	69	31.4%	16.1%
Total	**426**	**92**	**22%**	**21.5%**

The prevalence of CFRD has increased over time in the data registry data from 13.5% in 2012 to 21.5% this year. This may reflect; (i) the use of more sensitive measurement of abnormal glucose abnormalities with continuing 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.

10.2 CF related diabetes by age

426 patients



10.3 Liver function by ultra sound 428 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	205	81	39.5%	21	10.2%	103	50.2%
Adults	223	12	5.4%	10	4.5%	201	90.1%
Total	**428**	**93**		**31**		**304**	

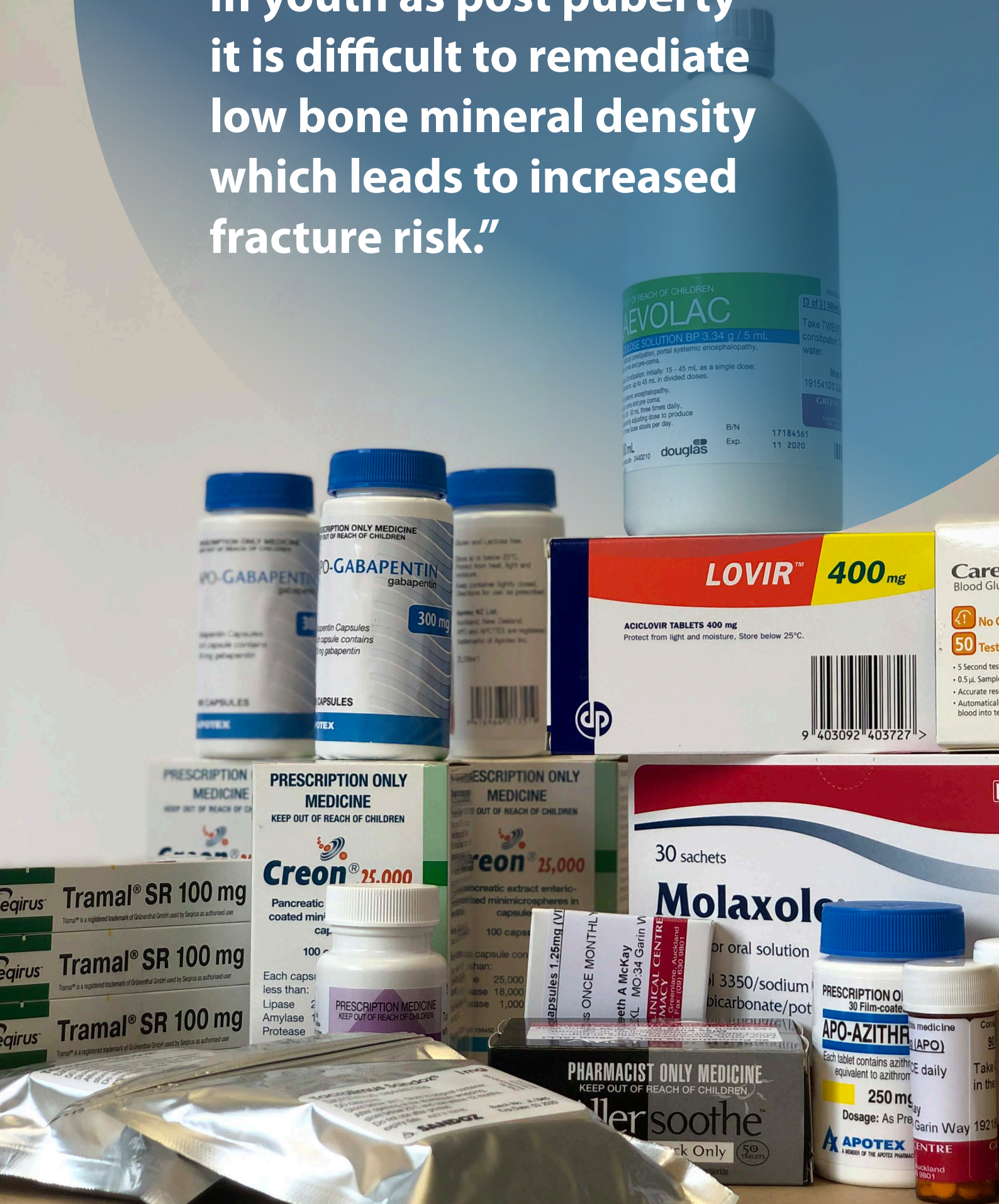
This is the first time we are presenting this data in the report. The 'not done' is because abdominal ultrasound has not been done in the current year. In children the recommended protocol is to do an abdominal ultrasound at 3, 6, 9 and 12 years of age, unless there is additional concern. From 12 years on the recommendation is annualy. 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 428 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	205	27	13.2%	4	2.0%	174	84.90%
Adults	223	28	12.6%	27	12.1%	168	75.3%
Total	**428**	**55**	**12.9%**	**31**	**7.2%**	**342**	**79.9%**

This is the first time we are presenting this data in the report. The current recommendtion is to do this at age 10 years and then do every two years unless there are other concerns (a previously low bone mineral denisty 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 fracture risk.”



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