Port CF 2019

New Zealand National Data Registry

CYSTIC FIBROSIS NZ

Contents

The Port CF National Data Registry is a research project of Cystic Fibrosis NZ. For further information about Cystic Fibrosis NZ 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 Data Registry.

Suggested Citation:
Port CFNZ National Data Registry, 2019 Registry Report, Cystic Fibrosis NZ. http://cfnz.org.nz/

Introduction

From the Chair of the Port CF Steering Commmittee

Cystic Fibrosis NZ and the Port CFNZ Steering Committee are pleased to present the National Data Registry 2019 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.
- Cystic Fibrosis NZ 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 New Zealand presented in this report.
- Canterbury District Health Board for their on-going information technology service to maintain the National Data Registry (Registry).

This ninth Registry Report gives an increasingly accurate picture of people with CF and their outcomes for New Zealand with 97-98% opting to provide this anonymised data. From 2012 – 2015, an additional 26 people were added to the Registry and by 2016 another 52 people had been added.

We have developed the Registry database further over the last four years, 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 CF involving representation from all countries that have a CF registry.

The 2019 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.

Associate Professor Cass Byrnes
Chair Port CFNZ
Port CF Principal Investigator (2017 - 2020)

Jane Bollard
CFNZ Chief Executive
(until September 2021)

Report completed by:
Cass Byrnes, Jan Tate, Emma Ellis , Alexia Searchfield

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

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

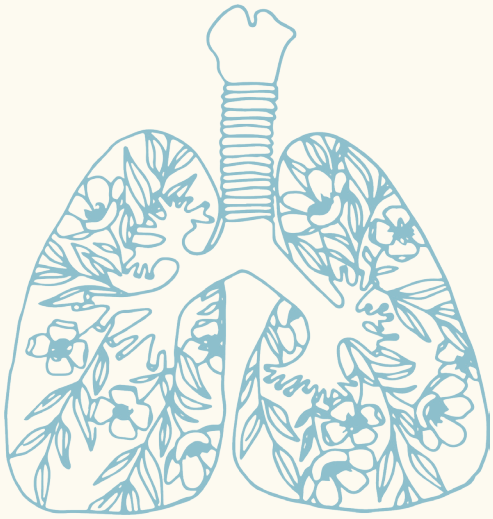
Canterbury (Paediatrics and Adults)
Christchurch Hospital, Christchurch

Otago (Paediatrics and Adults)
Dunedin Hospital, Dunedin

Southland (Paediatrics)
Kew Hospital, Invercargill

Glossary of Terms

CFNZ	Cystic Fibrosis New Zealand
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 and over
PWCF	Person with CF



Notes to the Registry

New Zealand has a total CF population comparative 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, the UK and the USA. Although we have a total of 531 registered in the Registry database, not all individuals had an input for all questions. While the total is 531 (223 children under 16 years and 308 adults16 years and over), 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 427 patients (205 children and 222 adults) on page 17. The data for the remaining individuals is missing.

New Zealand Registry data is becoming more robust and accurate and **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 the Chief Executive, Cystic Fibrosis NZ, or to the Project Co-ordinator Jan Tate.

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PO Box 110 067,
Grafton,
Auckland, 1148

OR

Chief Executive:
Lisa Burns
ceo@cfnz.org.nz

Key Indicators							531 PWCF
	2019	2018	2017	2016	2015	2014	2013
CF Patients Registered	531	514	498	501	449	443	444
Diagnosis							
Diagnosis age <1 year	11	15	15	6	5	7	5
Diagnosis age <16 years	1	0	2	3			
Diagnosis age >=16 years	2	0	1	2	0	2	3
Age							
Median Age (in years)	18.87	18.54	18.27	17.38	18.25	18.11	17.55
Mean Age (in years)	21.49	21.2	20.79	20.04			
PWCF <16 years							
Number	223	224	279	233	192	196	205
Percent	42%	43.6%	56.0%	46.5%	42.8%	44.2%	46.2%
PWCF >=16 years							
Number	308	290	219	268	257	247	239
Percent	58%	56.4%	44.0%	53.5%	57.2%	55.8%	53.8%
Gender							
Males							
Number	297	285	273	275	247	240	240
Percent	56%	55.4%	54.9%	54.9%	55.0%	54.2%	54.1%
Females							
Number	233	229	224	226	202	203	204
Percent	44%	44.6%	45.1%	45.1%	45.0%	45.8%	45.9%
Genotyped							
Number	495	466	484	450	400	429	426
Percent	93.2%	90.7%	97.4%	90.0%	89.1%	96.8%	95.9%
FEV1 (% predicted)							
Mean	76.6%	81.8	85.1%	85.0%			
Median	79%	86.2	86.5%	88.4%	85.6%	85.1%	84.3%
FEV1 < 16 Years							
Mean	95.8%	96.70%	96.8%	97.3%			
Median	97.9%	98.80%	99.3	99.3%	98.9%	97.7%	96.6%
FEV1 >=16 Years							
Mean	74.7%	75.30%	72.60%	72.6%			
Median	76.8%	79.20%	77.4	77.4%	77.0%	78.0%	70.7%
FEV1 < 18 Years							
Mean	94.5%	95.40%	95.1%	95.0%			
Median	97.2%	98.30%	98.3%	98.0%			
FEV1 >=18 Years							
Mean	73.1%	73.7%	72.2%	71.2%			
Median	74.7%	77.6%	75.6%	75.1%			

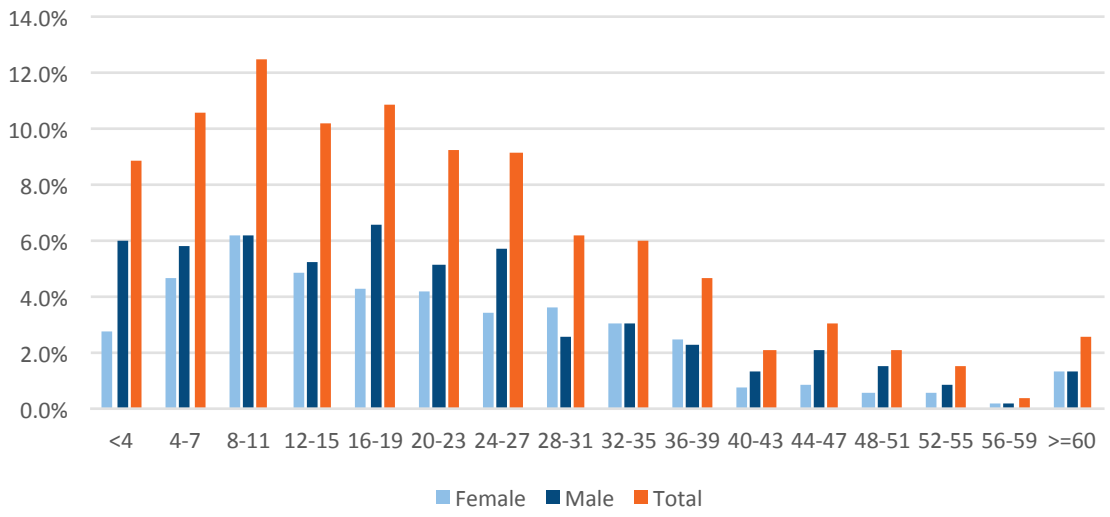
1. Demographics

530 PWCF

Age Group	All		Male		Female	
	Number in age group	**Percent** of all PWCF	**Number** in age group	**Percent** of all PWCF	**Number** in age group	**Percent** of all PWCF
0-3	47	8.9%	32	6.0%	15	2.8%
4-7	56	10.6%	31	5.8%	25	4.7%
8-11	66	12.5%	33	6.2%	33	6.2%
12-15	54	10.2%	28	5.2%	26	4.9%
16-19	58	10.9%	35	6.6%	23	4.3%
20-23	49	9.2%	27	5.1%	22	4.2%
24-27	48	9.1%	30	5.7%	18	3.4%
28-31	33	6.2%	14	2.6%	19	3.6%
32-35	32	6.0%	16	3.0%	16	3.0%
36-39	25	4.7%	12	2.3%	13	2.5%
40-43	11	2.1%	7	1.3%	4	0.8%
44-47	16	3.0%	11	2.1%	5	0.9%
48-51	11	2.1%	8	1.5%	3	0.6%
52-55	8	1.5%	5	0.9%	3	0.6%
56-59	2	0.4%	1	0.2%	1	0.2%
>=60	14	2.6%	7	1.3%	7	1.3%
Total	**530**	**100.0%**	**297**	**56%**	**233**	**44.0%**
Median	**18.87**					
Range	**0.10 - 76**					

1.1 Age distribution

530 PWCF



The median age of persons with CF in New Zealand has increased from 15.7 to 18.9 years over the nine years that we have had 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 223 children (42% total) and 308 adults (58% total), and if we include children as being up to 18 years, which is our more usual clinical practice, we have 254 children (48% total) and 276 adults (52% total).

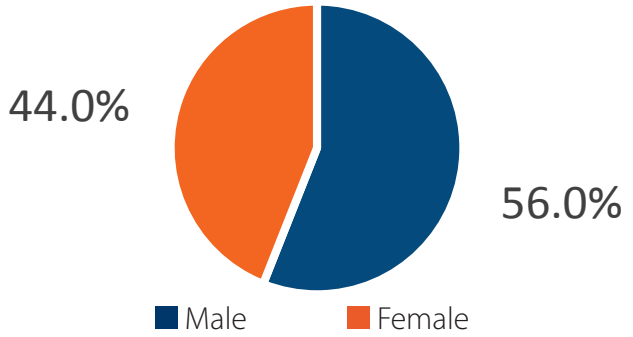
1.2 Gender Distribution

530 PWCF

	All		<16		>=16	
	Number in age group	**Percent** of all PWCF	**Number** in age group	**Percent** of all PWCF	**Number** in age group	**Percent** of all PWCF
Male	297	56.0%	124	55.6%	173	56.4%
Female	233	44.0%	99	44.4%	134	43.7%
Totals	**530**		**223**		**307**	

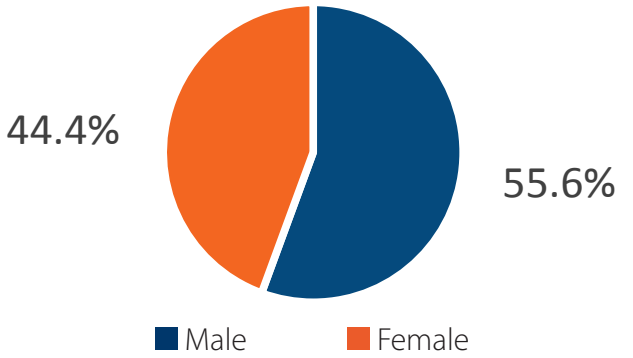
Gender Distribution of All PWCF

530 PWCF



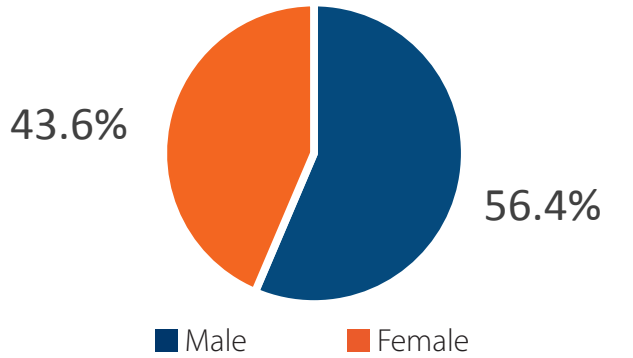
Gender Distribution <16 Years

223 PWCF



Gender Distribution >=16 Years

307 PWCF



“The median age of persons with CF in New Zealand has increased from 15.7 to 18.9 years over the nine years that we have had National Registry data.”

2. Genotypes

495 PWCF

Mutations	Number of PWCF Genotyped	Percentage of PWCF Genotyped
Homozygous F508del	252	50.9%
Heterozygous F508del	192	38.8%
No F508del	51	10.3%
Total	**495**	

2.1 Second Allele of Heterozygous F508del

192 PWCF

Second Allele	c.DNA Name	Number of PWCF	Percent of PWCF
G542X	c.1624G>T	21	4.2%
G551D	c.1652G>A	20	4.0%
R117H	c.350G>A	17	3.4%
G85E	c.254G>A	4	0.8%
^1507	c.1519_1521delATC	3	0.6%
621+1G->T	c.489+1G>T	3	0.6%
N1303K	c.3909c>G	3	0.6%
3849+10kbC->T	c.3717+12191C>T	2	0.4%
1717-1G->A	c.1585-1G>A	2	0.4%
1898+1G->A	c.1766+1G>A	2	0.4%
A455E	c.1364C>A	2	0.4%
R560T	c.1679G>C	2	0.4%
D1152H	c.3454G>C	2	0.4%
R334W	c.1000C>T	2	0.4%
Q493X	c.1477C>T	2	0.4%
1078delT	c.948delT	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%
W1282X	c.3846G>A	1	0.2%
R1158X	c.3472C>T	1	0.2%
R1162X	c.3484C>T	1	0.2%
3659delC	c.3528delC	1	0.2%
Other genetic mutation		95	19.2%

Note: Because people have two genes, they are counted twice, once for each gene. The total number of people is 495.

Our high percentage of F508del is in keeping with the international registries from European derived populations. In total only 51 people in New Zealand do not have at least one F508 mutation. Looking at the gene mutations recorded in the 2019 Registry, 27 people of the 495 who have been genotyped (5.5%) would not be detected by the current new born screening programme.

2.2 No F508del Mutations

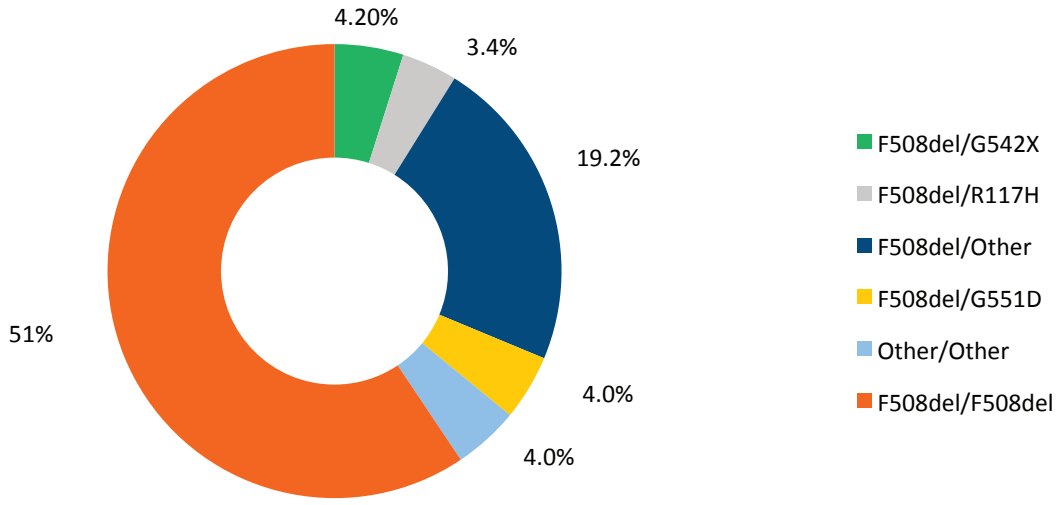
51 PWCF

	1717-1G->A	G542X	G551D	Other	Q493X	R117H
3849+10kbC->T	1	0	0	0	0	0
G542X	0	1	0	3	0	1
G551D	1	1	1	5	3	5
G85E	0	0	0	1	0	0
N1303K	0	0	1	1	0	0
Other	0	0	0	20	0	0
R1162X	0	0	0	1	0	0
R117H	0	0	0	0	1	1
R553X	0	0	1	1	0	0
W1282X	0	0	0	1	0	0

2.3 Genotype Major Categories

Mutations	Number PWCF Identified	Percentage PWCF Identified
F508del	444	89.7%
G551D	39	7.9%
G542X	28	5.7%
R117H	26	5.3%
G85E	5	1.0%

2.4 Common Genotype Combinations



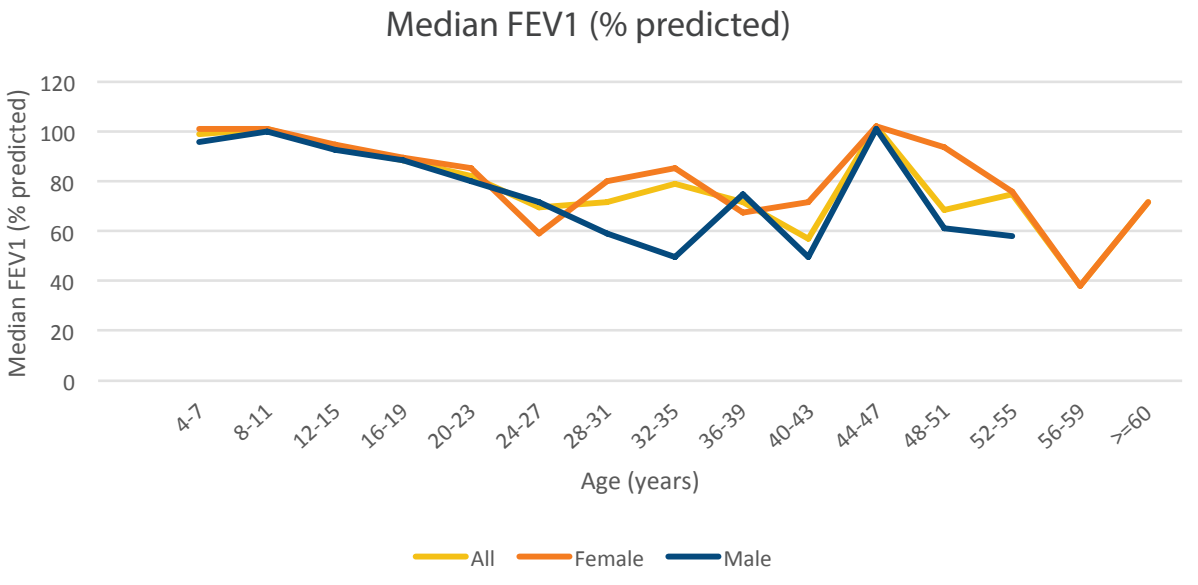
3. Respiratory

376 PWCF

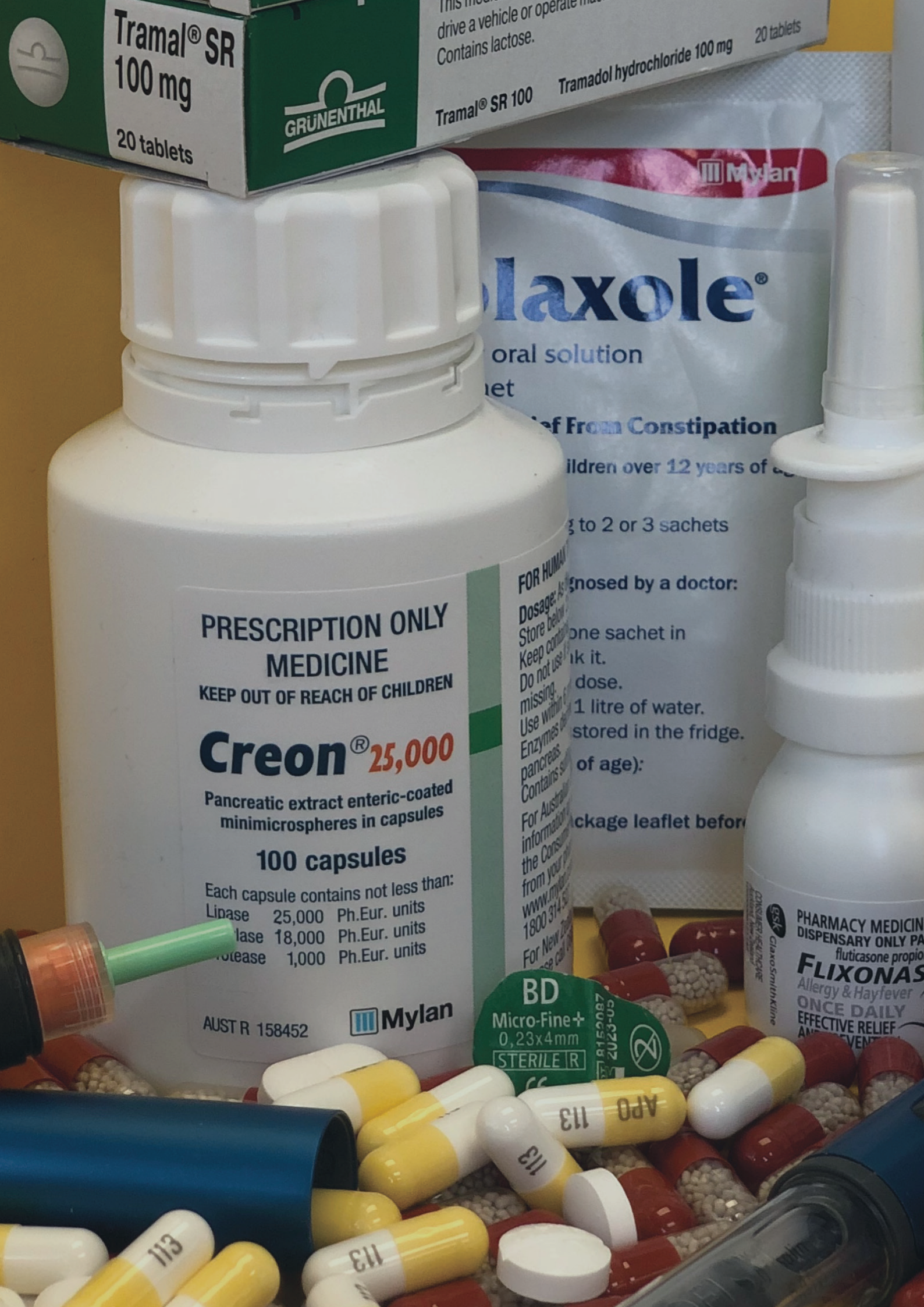
Age Group	All		Female		Male	
	Number in age group	Median FEV1	Number in age group	Median FEV1	Number in age group	Median FEV1
4-7	30	98.7	16	100.9	14	95.8
8-11	62	101	30	101	32	100
12-15	49	93.7	24	94.2	25	92.1
16-19	49	88.6	19	89.5	30	87.9
20-23	39	82.5	15	85	24	79.80
24-27	31	69.5	8	59.1	23	71.1
28-31	25	71.1	16	79.8	9	58.90
32-35	21	78.7	10	85.1	11	50.1
36-39	20	71.5	10	67.7	10	75.10
40-43	10	56.5	4	72.1	6	49.6
44-47	14	101.6	5	102.2	9	100.9
48-51	8	68.2	2	93.3	6	60.6
52-55	8	74.50	3	75.5	5	57.6
56-59	1	37.7	1	37.7	0	
>=60	9	71.5	3	71.5	6	72.4
Totals	**376**	**80.1**	**166**	**89.5**	**210**	**86**

3.1 Median FEV_1 (% predicted)

376 PWCF



The median FEV1 of the population able to do lung function has always been 80% predicted since we started our Registry while the median this year is 79.2% (97.9% in children, 76.8% 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. 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.


Tramal® SR
100 mg
20 tablets
GRÜNENTHAL
Tramal® SR 100
Tramadol hydrochloride 100 mg
20 tablets
Contains lactose.
Mylan
laxole®
oral solution
ef From Constipation
ildren over 12 years of
g to 2 or 3 sachets
gnosed by a doctor:
one sachet in
k it.
dose.
1 litre of water.
stored in the fridge.
of age):
ckage leaflet befor
PRESCRIPTION ONLY
MEDICINE
KEEP OUT OF REACH OF CHILDREN
Creon® 25,000
Pancreatic extract enteric-coated
minimicrospheres in capsules
100 capsules
Each capsule contains not less than:
Lipase 25,000 Ph.Eur. units
18,000 Ph.Eur. units
1,000 Ph.Eur. units
AUST R 158452
Mylan
BD
Micro-Fine+
0,23x4mm
STERILE R
PHARMACY MEDICIN
DISPENSARY ONLY PA
fluticasone propio
FLIXONAS
Allergy & Hayfever
ONCE DAILY
EFFECTIVE RELIEF
APO 113
113
113
113

4. Nutrition

422 PWCF

4.1 Paediatric BMI

191 PWCF

All <16 Years			Female <16 Years			Male <16 Years		
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	26	83.5	<4	8	75.2	<4	18	85.4
4-7	53	60.5	4-7	23	66.8	4-7	30	59.1
8-11	63	60.9	8-11	31	54.4	8-11	32	67.0
12-15	49	49	12-15	24	53.6	12-15	25	42.3
Totals	**191**			**86**			**105**	

4.2 Adult BMI

231 PWCF

All >=16 Years			Female >=16 Years			Male >=16 Years		
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	49	22.0	16-19	19	22.3	16-19	30	21.1
20-23	39	22.2	20-23	15	24.8	20-23	24	21.5
24-27	31	21.4	24-27	8	21	24-27	23	21.5
28-31	25	22.8	28-31	16	22.7	28-31	9	22.9
32-35	21	22.6	32-35	10	22.6	32-35	11	22.5
36-39	20	23.3	36-39	10	21.8	36-39	10	24.2
40-43	10	25	40-43	4	22.5	40-43	6	25.7
44-47	14	25.8	44-47	5	23.5	44-47	9	26.5
48-51	8	24	48-51	2	24.1	48-51	6	24
52-55	8	25.4	52-55	3	24.5	52-55	5	26.2
56-59	1	24.7	56-59	1	24.7	56-59	0	22.6
60+	5	22.1	60+	4	22.1	60+	1	23.6
Totals	**231**			**97**			**134**	

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:

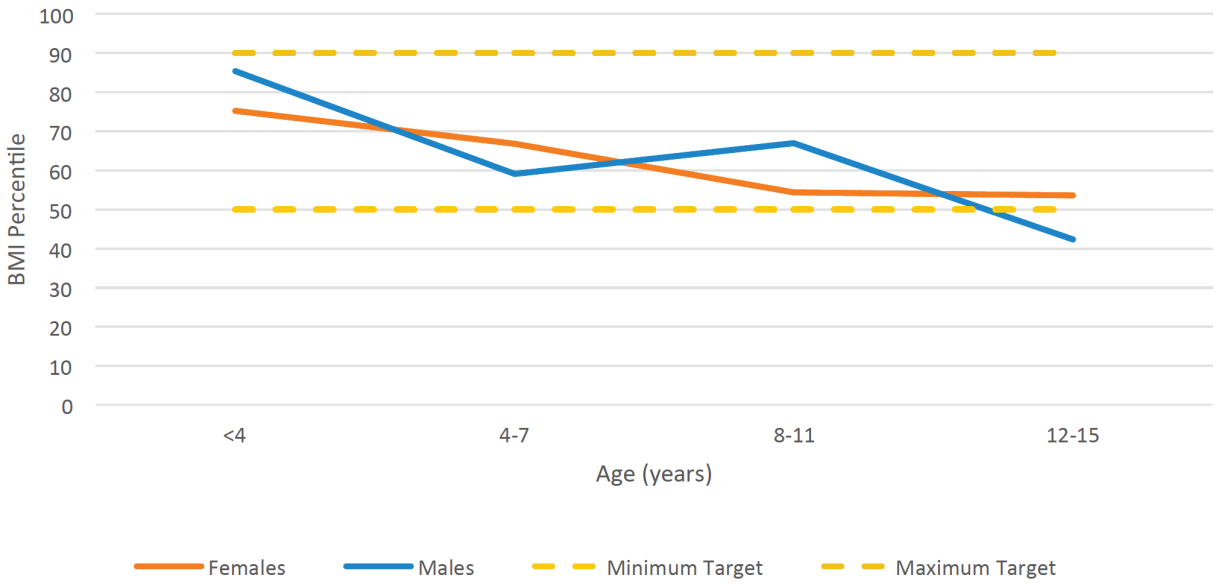
1) Infants (0 to 24 months): weight-for-length ≥50th percentile using WHO growth charts
2) Children and Adolescents (2-18 years): BMI 50-85th percentile (CDC growth charts) or 50- 91st percentile (WHO growth charts)
3) Adults: males BMI 23 - 27 kg/m2, females BMI 22 - 27 kg/m2

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 under 4 years of age the median BMI is 83.5 percentile. For children and adolescents the median BMI is 56.5 percentile. For adults 42.9% of males and 51.3% of females are above the minimum target range.

4.3 Median BMI Percentile <16 Years

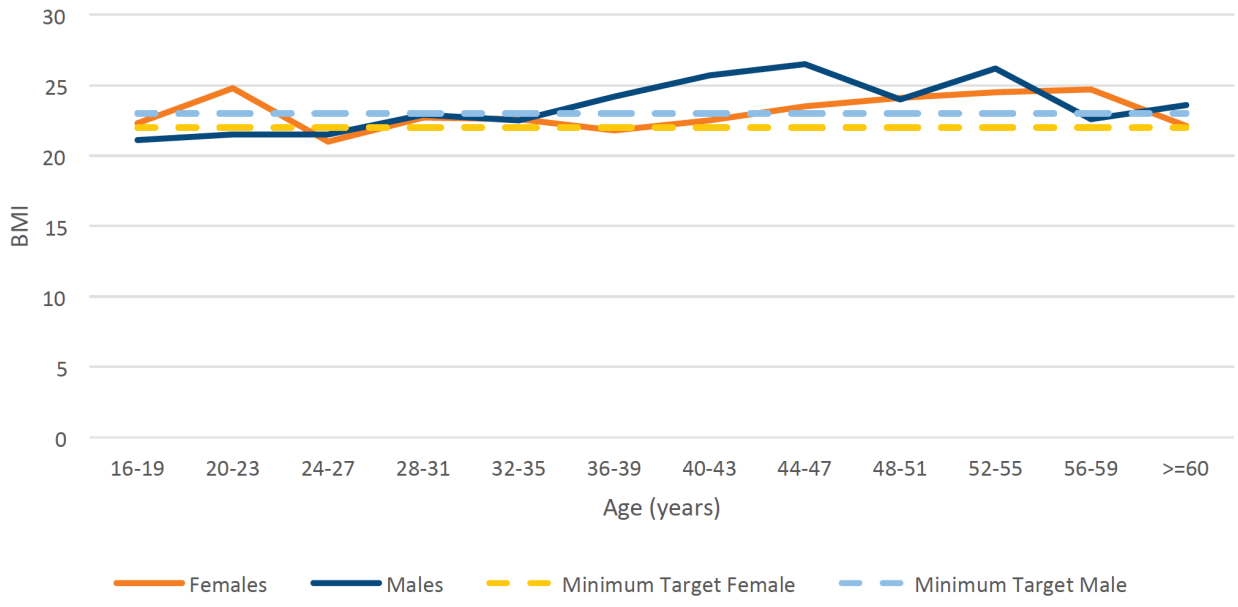
191 PWCF



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 Years

231 PWCF



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.

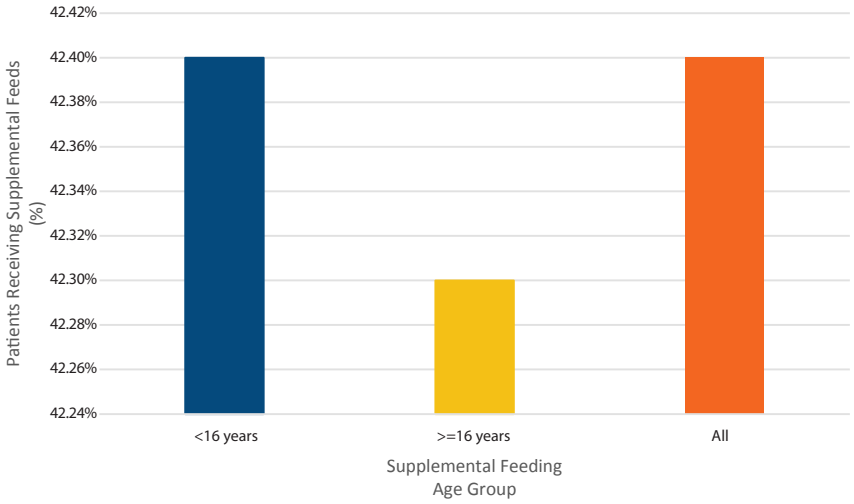
4.5 Supplemental Feeding

427 PWCF

	<16 years, n =205	
	Yes	**% <16 years supplemented**
Supplemental Feeding	**87**	**42.4%**
Nasogastric	3	1.4%
Gastrostomy	14	6.6%
Oral	80	37.7%
	>=16 years, n = 222	
	Yes	**% >=16 years supplemented**
Supplemental Feeding	**94**	**42.3%**
Nasogastric	2	0.8%
Gastrostomy	17	7.0%
Oral	84	34.4%

4.6 Supplemental Feeding by Age Group

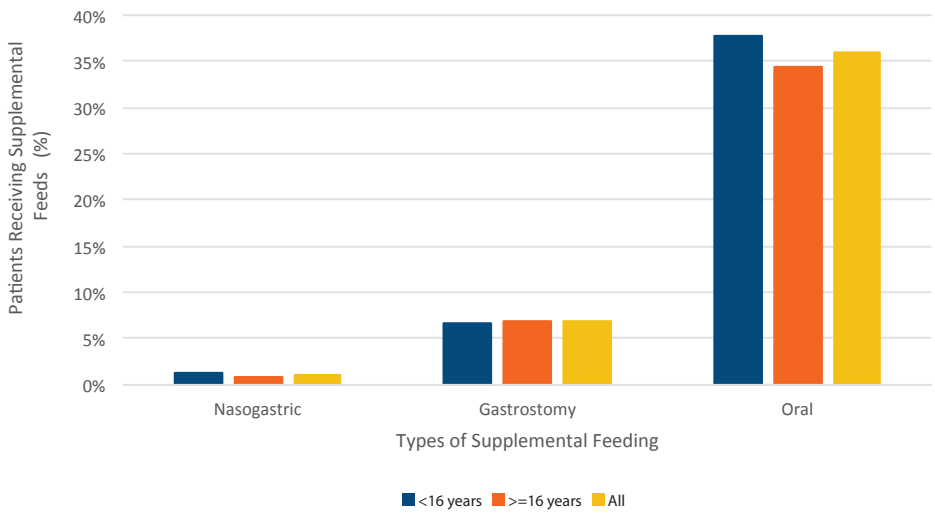
427 PWCF



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

4.7 Supplemental Feeding by Type

427 PWCF



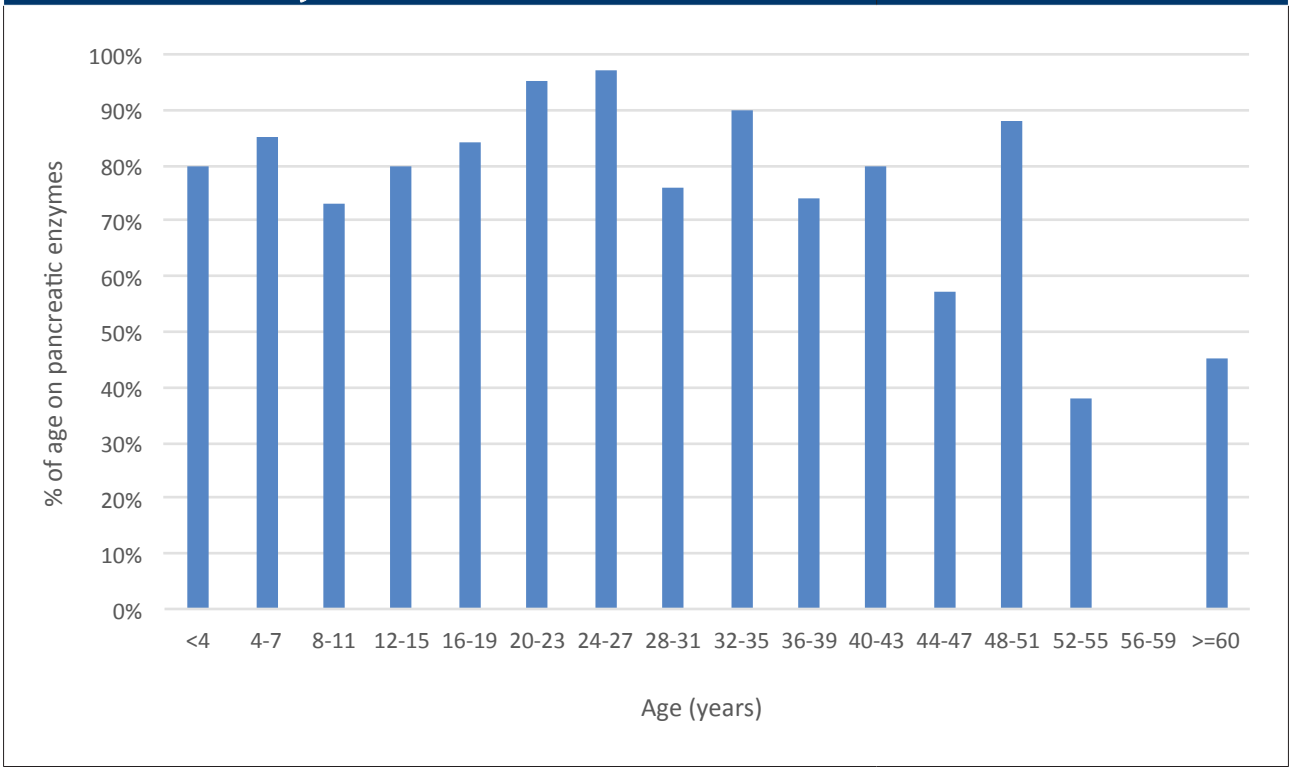
5. Pancreatic Enzymes

447 PWCF

Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of CF population
<4	45	36	80.0%	8.1%
4-7	53	45	85.0%	10.1%
8-11	62	45	73.0%	10.1%
12-15	50	40	80.0%	8.9%
16-19	49	41	84.0%	9.2%
20-23	40	38	95.0%	8.5%
24-27	32	31	97.0%	6.9%
28-31	25	19	76.0%	4.3%
32-35	20	18	90.0%	4.0%
36-39	19	14	74.0%	3.1%
40-43	10	8	80.0%	1.8%
44-47	14	8	57.0%	1.8%
48-51	8	7	88.0%	1.6%
52-55	8	3	38.0%	0.7%
56-59	1	0	0.0%	0.0%
>=60	11	5	45.0%	1.1%
Totals	**447**	**358**		**80.1%**

Pancreatic Enzymes

358 PWCF



6. Airway Clearance Techniques

6.1 Primary Airway Clearance

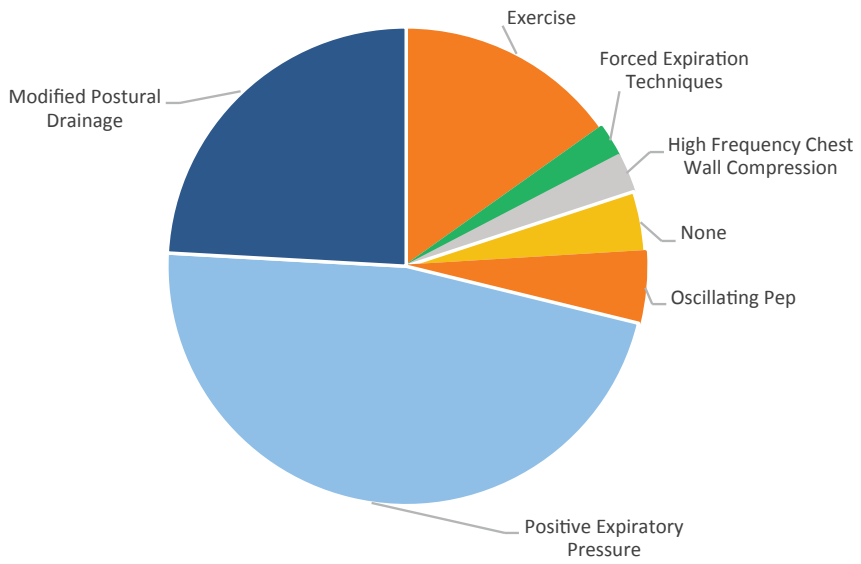
456 PWCF

	<16 years	
	Number	Percent
Positive Expiratory Pressure	127	59.9%
Modified Postural Drainage	65	30.7%
Exercise	41	19.3%
Oscillating Pep (e.g. Flutter, Acapella, IPV)	13	6.1%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	6	2.8%
High Frequency Chest Wall Compression (e.g. vest)	7	3.3%
None	11	5.2%

	>= 16 years	
	Number	Percent
Positive Expiratory Pressure	103	42.2%
Modified Postural Drainage	2	0.8%
Exercise	74	30.3%
Oscillating Pep (e.g. Flutter, Acapella, IPV)	41	16.8%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	33	13.5%
High Frequency Chest Wall Compression (e.g. vest)	0	0.0%
None	27	11.1%

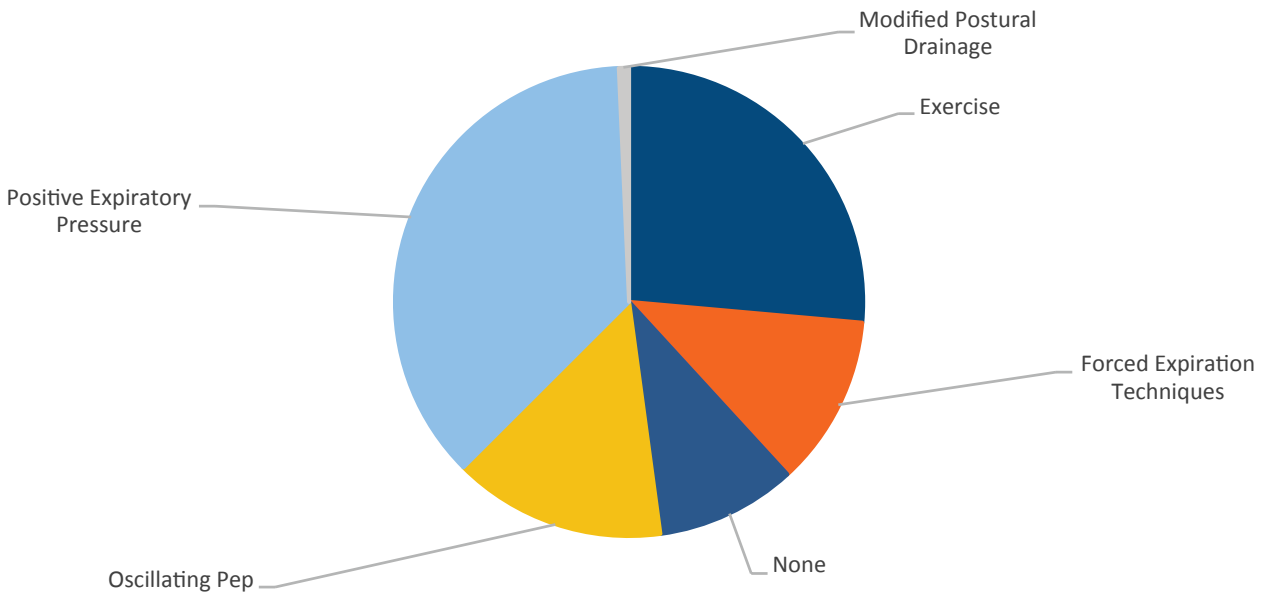
6.2 Primary Airway Clearance <16 years

212 PWCF



6.3 Primary Airway Clearance >=16 years

244 PWCF

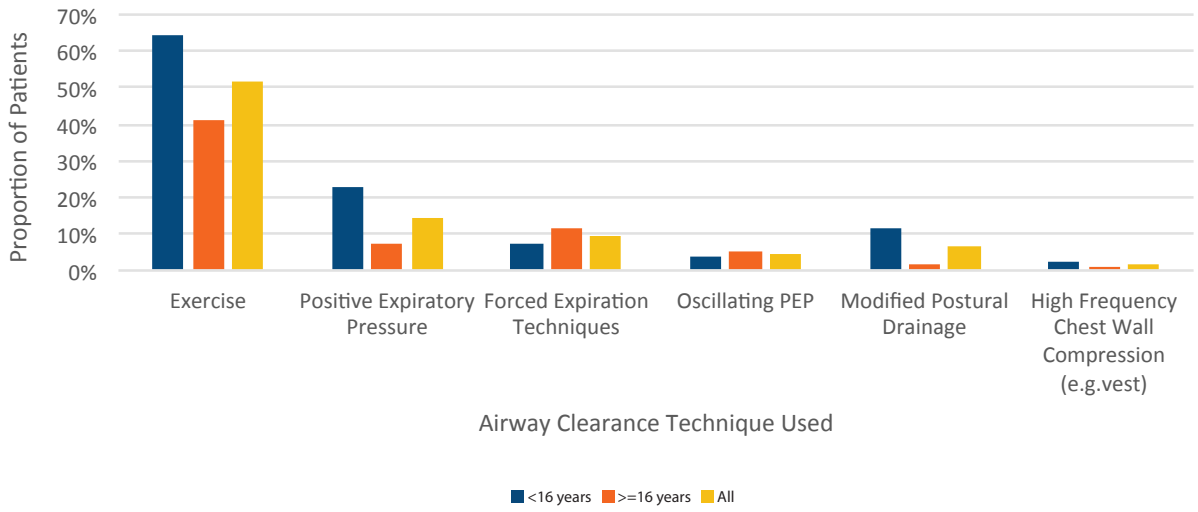


NOTE: Some patients may have used more than one technique as their primary airway clearance technique over the course of a year.

The nominated primary airway clearance technique adds up to greater than 100% 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 5.2% in 2019) while the trend is the opposite for the adults (18.8% in 2013 to 11.1% in 2019). 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

PWCF



7. Microbiology

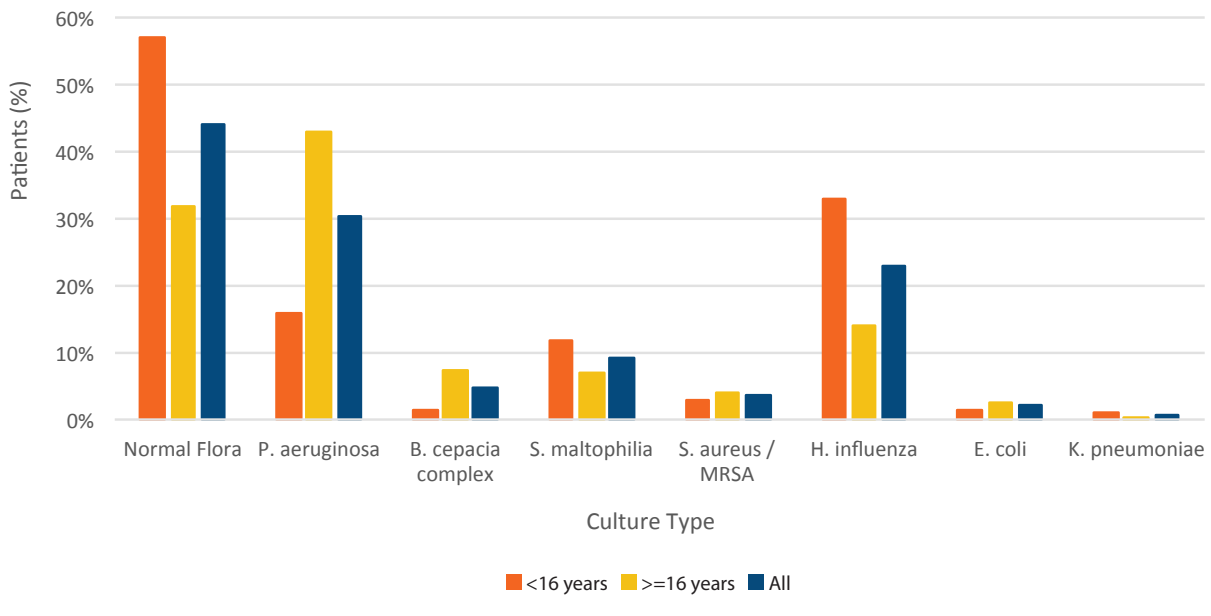
Microbiology

456 PWCF

	<16 years		>=16 years		All	
	Number	Percent	Number	Percent	Number	Percent
Normal Flora	121	57.0%	79	32.0%	200	44.0%
Haemophilus Influenza	70	33.0%	34	13.9%	104	22.8%
E.Coli	3	1.4%	6	2.5%	9	2.0%
Klebsiella Pneumoniae	2	0.9%	1	0.4%	3	0.7%
Stenotrophomonas Maltophilia	25	11.8%	17	7.0%	42	9.2%
Pseudomonas Aeruginosa	**34**	**16.0%**	**105**	**43.0%**	**139**	**30.5%**
Mucoid	9	4.2%	72	29.5%	81	17.8%
Non Mucoid	26	12.3%	71	29.1%	97	21.3%
Staphylococcus Aureus	**122**	**57.5%**	**114**	**46.7%**	**236**	**51.8%**
MSSA	116	54.0%	104	42.6%	220	48.2%
MRSA	6	2.8%	10	4.1%	16	3.5%
Burkholderia Cepacia Complex	**3**	**1.4%**	**18**	**7.4%**	**21**	**4.6%**
Cenocepacia	0	0.0%	3	1.2%	3	0.7%
Multivorans	2	0.9%	8	3.3%	10	2.2%
Other	1	0.5%	1	0.4%	2	0.4%

7.1 Bacterial Culture Prevalence

456 PWCF

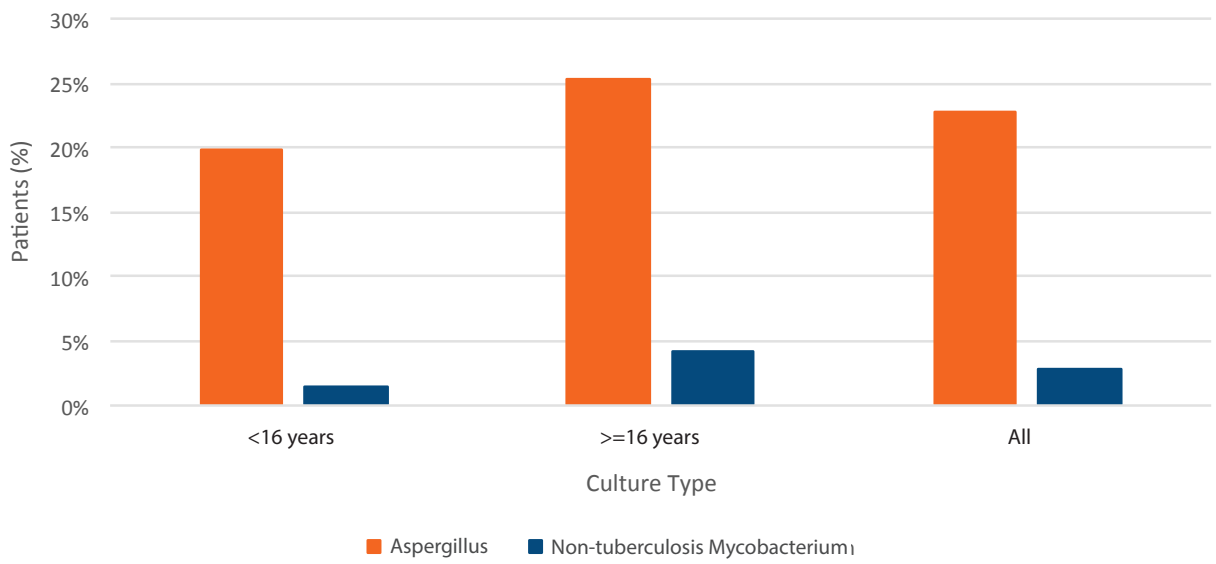


NOTE: The percentages of population with CF having had specific respiratory pathogens identified such as Staphylococcal aureus, Pseudomonas aeruginosa etc. are very similar to the percentages presented in the Australian 2017 registry, with the exception of much higher percentages of Haemophilus influenza here. This pathogen is also higher in our young children and lower in our adults. Pseudomonas aeruginosa is found in 16% of the children and increases to 43% in adults. Our MRSA rates are relatively low at 3.5% overall.

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

7.2 Nonbacterial/Fungal Prevalence

456 PWCF

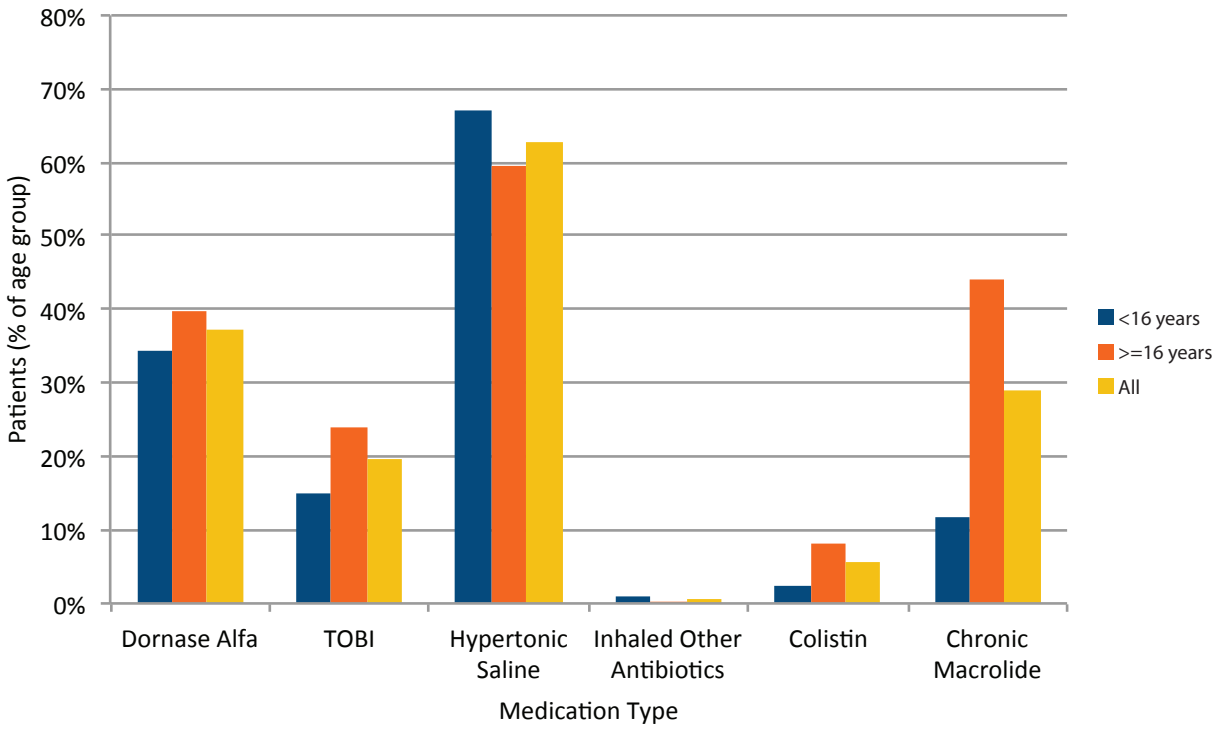


8. Medications

Medications			456 PWCF
Medication	**<16 Years**	**>=16 Years**	**All**
Hypertonic Saline	67.0%	59.4%	62.9%
Dornase alfa	34.4%	39.8%	37.3%
TOBI	15.1%	23.8%	19.7%
Inhaled Other Antibiotics	0.9%	0.4%	0.7%
Chronic Macrolide	11.8%	43.9%	28.9%
Corticosteroids Inhaled	11.3%	13.1%	12.3%
Corticosteroids Oral	4.7%	5.7%	5.3%
Antifungals	2.8%	2.5%	2.6%
Influenza Vaccine	82.1%	56.70%	68.60%

8.1 Medications Prescribed

456 PWCF



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

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. We also had no access to some newer medications in 2019 except on research programmes - notably the modulator drugs.

9. Intravenous Antibiotic Treatment

9.1 Home IV Days

449 PWCF

Age	Number In Age Group	Number Who Had IV Days	Percent Who Had IV Days	Mean Days For Those Who Had IV Days	Mean Days For All PWCF
<4	45	6	13%	8.7	1.2
4-7	53	9	17%	11.1	1.9
8-11	62	7	11%	21.6	2.4
12-15	51	9	18%	19.7	3.5
16-19	49	11	22%	19.5	4.4
20-23	41	13	32%	15.1	4.8
24-27	31	9	29%	12.6	3.6
28-31	25	12	48%	12.8	6.2
32-35	21	8	38%	21.8	8.3
36-39	19	6	32%	20.2	6.4
40-43	10	2	20%	15.5	3.1
44-47	14	4	29%	18.3	5.2
48-51	8	2	25%	7.5	1.9
52-55	8	0	0%	-	-
56-59	1	0	0%	-	-
>=60	11	3	27%	9.3	9.3
Totals	**449**	**101**	**22%**	**15.8**	**3.6**

9.2 Hospital IV Days

449 PWCF

Age	Number In Age Group	Number Who Had IV Days	Percent Who Had IV Days	Mean Days For Those Who Had IV Days	Mean Days For All PWCF
<4	45	7	16%	12.1	1.9
4-7	53	15	28%	19.5	5.5
8-15	62	15	24%	12.3	3
16-18	51	21	41%	26.0	10.7
16-19	49	21	43%	32.2	13.8
20-23	41	16	39%	22.6	8.8
24-27	31	12	39%	20.9	8.1
28-31	25	15	60%	11.1	6.6
32-35	21	11	52%	20.9	11
36-39	19	8	42%	14.4	6.1
40-43	10	3	30%	21.3	6.4
44-47	14	5	36%	18.4	6.6
48-51	8	3	38%	13.7	5.1
52-55	8	1	13%	7.0	0.9
56-59	1	1	100%	14	14
>=60	11	6	55%	15.3	8.4
Totals	**449**	**160**	**36%**	**20.1**	**7.2**

10. Complications

10.1 CF Related Diabetes

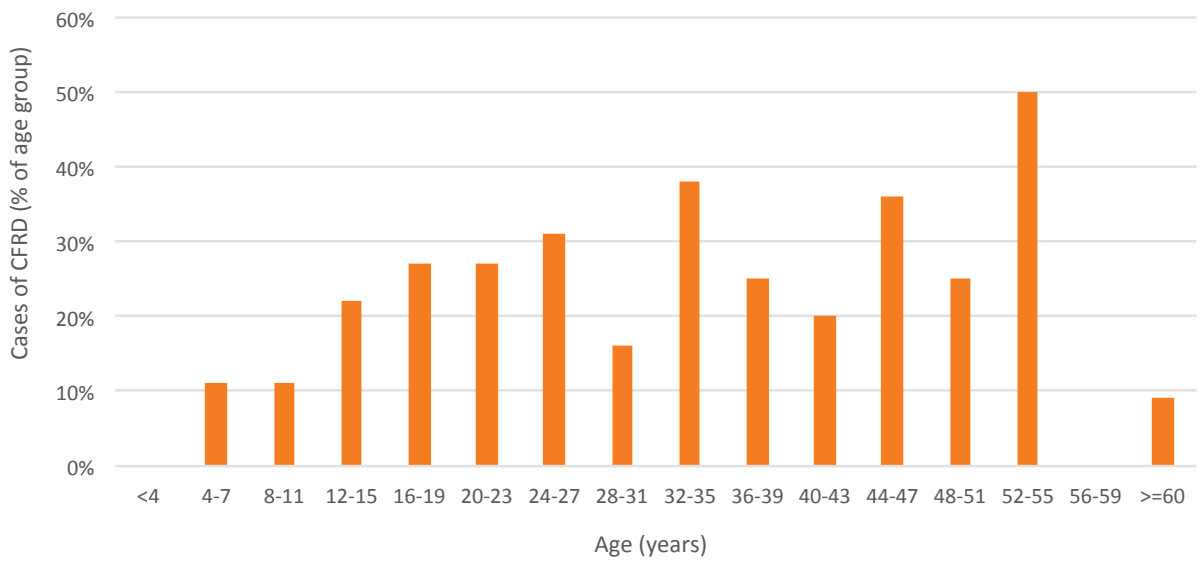
452 PWCF

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of CF Population
<4	45	0	0%	0.0%
4-7	54	6	11%	1.3%
8-11	62	7	11%	1.5%
12-15	51	11	22%	2.4%
16-19	49	13	27%	2.9%
20-23	41	11	27%	2.4%
24-27	32	10	31%	2.2%
28-31	25	4	16%	0.9%
32-35	21	8	38%	1.8%
36-39	20	5	25%	1.1%
40-43	10	2	20%	0.4%
44-47	14	5	36%	1.1%
48-51	8	2	25%	0.4%
52-55	8	4	50%	0.9%
56-59	1	0	0%	0.0%
>=60	11	1	9%	0.2%
Total	**452**	**89**		**19.7%%**

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of CF Population
<16	212	24	11.0%	5.3%
>=16	240	65	27.0%	14.2%
Total	**452**	**90**	**20%**	**19.5%**

10.2 CF Related Diabetes by Age

452 PWCF



10.3 Liver Function by Ultra Sound

452 PWCF

		Normal		Abnormal		Unknown	
	Number in age group	Number	Percent	Number	Percent	Number	Percent
Paediatrics	212	65	30.7%	19	9.0%	128	60.40%
Adults	240	21	8.8%	11	4.6%	208	86.7%
Total	**452**	**86**	**19.0%**	**30**	**6.6%**	**336**	**74.3%**

10.4 Bone Density by DEXA Scans

452 PWCF

		Normal		Abnormal		Unknown	
	Number in age group	Number	Percent	Number	Percent	Number	Percent
Paediatrics	212	32	15.1%	5	2.4%	175	82.5%
Adults	240	40	16.7%	22	9.2%	178	74.2%
Total	**452**	**72**	**15.9%**	**27**	**6.0%**	**353**	**78.1%**

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