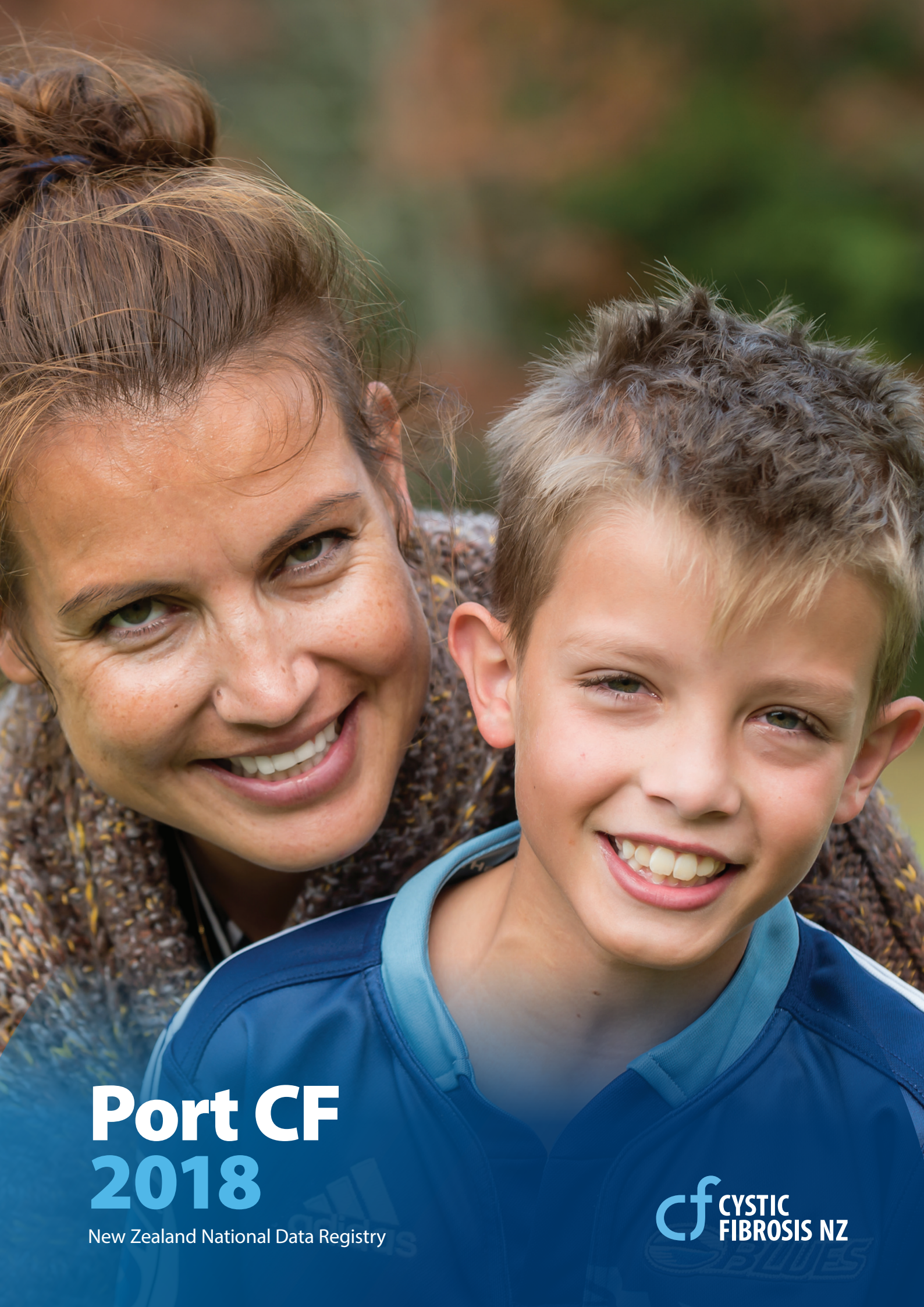

Port CF 2018

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 in New Zealand who have consented to have their data recorded as part of this National Data Registry.

Suggested Citation:
Port CF National Data Registry, 2018 Registry Report, Cystic Fibrosis NZ. http://cfnz.org.nz/

Introduction

From the Chair of the Port CF Steering Commmittee

Cystic Fibrosis NZ (CFNZ) and the Port CF Steering Committee are pleased to present the National Data Registry 2018 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 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 eighth 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 three 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 2018 Registry Report presents 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
Port CF Principal Investigator (2017 - 2020)

Jane Bollard
CFNZ Chief Executive (until September 2021)

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

A special thanks to:
Andrew Watson, Chris Frampton, 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 NZ
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	0-15 years of age
Adult	16 years and over
PWCF	Person with CF

Notes to the Registry

New Zealand has a total CF population comparable to a single clinic in the USA or the UK and this data provides 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 514 registered in Port CF, not all individuals had a response for all questions. While the total is 514 (224 children <16 years, 290 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 458 patients (212 children and 246 adults) on page 17. The data for the remaining individuals is missing.

NZ 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 of CFNZ or to the Project Co-ordinator Jan Tate.

Port CF Steering Committee
Chief Executive
ceo@cfnz.org.nz

OR

Port CF Steering Committee
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Grafton,
Auckland, 1148

Project Co-ordinator: JanT@adhb.govt.nz

Key Indicators							514 PWCF
	2018	2017	2016	2015	2014	2013	2012
CF Patients Registered	514	498	501	449	443	444	423
Diagnosis							
Diagnosis age <1 year	15	15	6	5	7	5	11
Diagnosis age <16 years	0	2	3				
Diagnosis age >=16 years	0	1	2	0	2	3	2
Age							
Median Age (in years)	18.54	18.27	17.38	18.25	18.11	17.55	16.15
Mean Age (in years)	21.2	20.79	20.04				
PWCF <16 years							
Number	224	279	233	192	196	205	209
Percent	43.6%	56.0%	46.5%	42.8%	44.2%	46.2%	49.4%
PWCF >=16 years							
Number	290	219	268	257	247	239	214
Percent	56.4%	44.0%	53.5%	57.2%	55.8%	53.8%	50.6%
Gender							
Males							
Number	285	273	275	247	240	240	228
Percent	55.4%	54.9%	54.9%	55.0%	54.2%	54.1%	53.0%
Females							
Number	229	224	226	202	203	204	195
Percent	44.6%	45.1%	45.1%	45.0%	45.8%	45.9%	46.1%
Genotyped							
Number	466	484	450	400	429	426	407
Percent	90.7%	97.4%	90.0%	89.1%	96.8%	95.9%	96.2%
FEV1 (% predicted)							
Mean	81.8	85.1%	85.0%				
Median	86.2	86.5%	88.4%	85.6%	85.1%	84.3%	84.5%
FEV1 < 16 Years							
Mean	96.70%	96.8%	97.3%				
Median	98.80%	99.3	99.3%	98.9%	97.7%	96.6%	97.2%
FEV1 >=16 Years							
Mean	75.30%	72.60%	72.6%				
Median	79.20%	77.4	77.4%	77.0%	78.0%	70.7%	70.6%
FEV1 < 18 Years							
Mean	95.40%	95.1%	95.0%				
Median	98.30%	98.3%	98.0%				
FEV1 >=18 Years							
Mean	73.7%	72.2%	71.2%				
Median	77.6%	75.6%	75.1%				

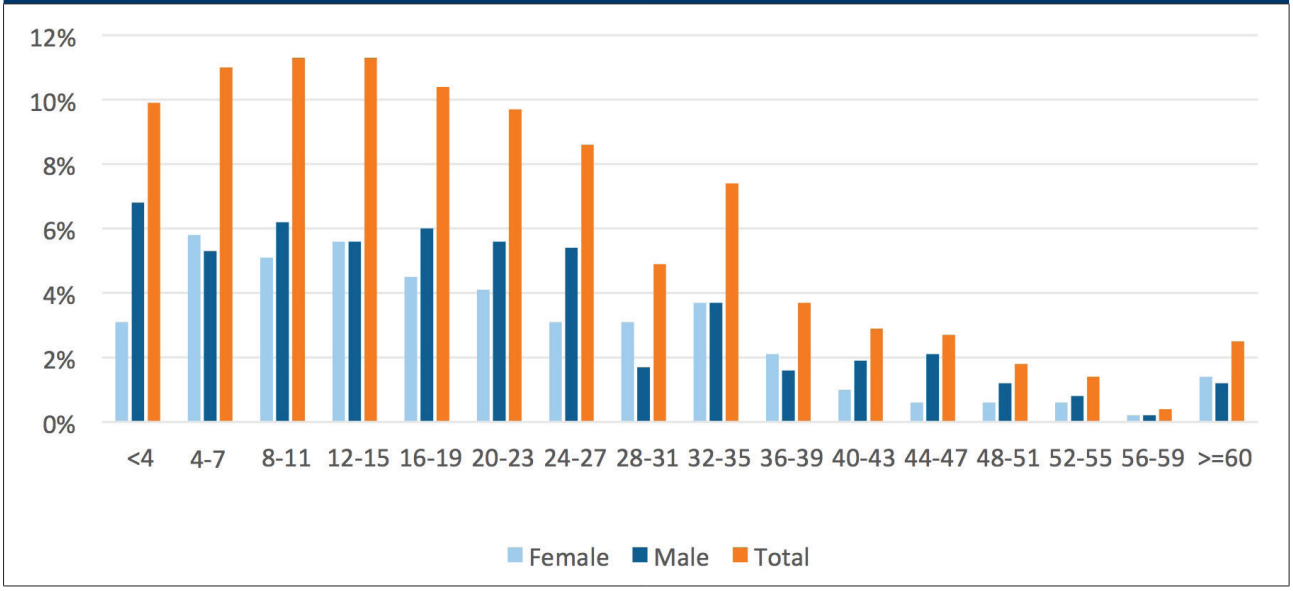
1. Demographics

514 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	51	9.9%	35	6.8%	16	3.1%
4-7	57	11.0%	27	5.3%	30	5.8%
8-11	58	11.3%	32	6.2%	26	5.1%
12-15	58	11.3%	29	5.6%	29	5.6%
16-19	54	10.4%	31	6.0%	23	4.5%
20-23	50	9.7%	29	5.6%	21	4.1%
24-27	44	8.6%	28	5.4%	16	3.1%
28-31	25	4.9%	9	1.7%	16	3.1%
32-35	38	7.4%	19	3.7%	19	3.7%
36-39	19	3.7%	8	1.6%	11	2.1%
40-43	15	2.9%	10	1.9%	5	1.0%
44-47	14	2.7%	11	2.1%	3	0.6%
48-51	9	1.8%	6	1.2%	3	0.6%
52-55	7	1.4%	4	0.8%	3	0.6%
56-59	2	0.4%	1	0.2%	1	0.2%
>=60	13	2.5%	6	1.2%	7	1.4%
Total	**514**	**100.0%**	**285**	**55.4%**	**229**	**44.6%**
Median	**18.54**					
Range	**0.13 - 75**					

1.1 Age distribution

514 PWCF



The median age of persons with CF in New Zealand has increased from 16 to 18 years over the eight years that we have had Registry data. 'Children' in international registries are defined as either under 16 years of age or under 18 years of age. In New Zealand if we include children as being those under 16 years, we have 224 children (43.6% total) and 290 adults (56.4% total). If we include children as being those under 18 years, which is our more usual clinical practice, we have 251 children (48.8% total) and 263 adults (51.2% total).

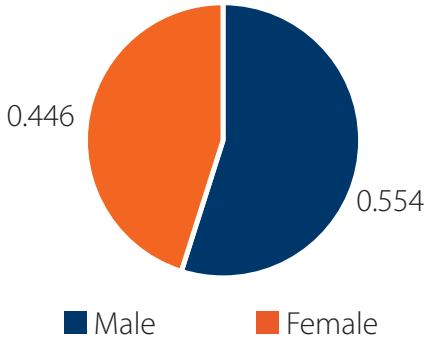
1.2 Gender Distribution

514 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	285	55.4%	123	23.9%	162	31.5%
Female	229	44.6%	101	19.6%	128	24.9%
Totals	**514**	**100%**	**224**	**43.6%**	**290**	**56.4%**

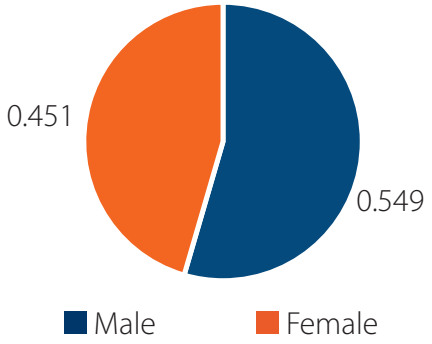
Gender Distribution of All Patients

514 PWCF



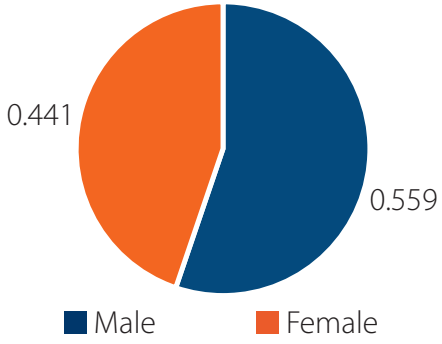
Gender Distribution <16 Years

224 PWCF



Gender Distribution >=16 Years

290 PWCF



“The median age of persons with CF in New Zealand has increased from 15.7 to 18.5 years over the eight years that we have had National Registry data.”

2. Genotypes

466 PWCF

Mutations	Number of Patients Genotyped	Percentage of Patients Genotyped
Homozygous F508del	243	52.8%
Heterozygous F508del	177	38.0%
No F508del	46	9.9%
Total	**466**	

2.1 Second Allele of Heterozygous F508del

177 PWCF

Second Allele	c.DNA Name	Number of PWCF	Percent of PWCF
G542X	c.1624G>T	21	4.5%
G551D	c.1652G>A	20	4.3%
R117H	c.350G>A	16	3.4%
G85E	c.254G>A	5	1.1%
^1507	c.1519_1521delATC	4	0.9%
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%
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%
Other genetic mutation		81	17.4%

2.2 No F508del Mutations

	1717-1G->A	G542X	G551D	Other	Q493X	R117H
3849+10kbC->T	1	0	0	0	0	0
G542X	0	1	0	2	0	0
G551D	1	1	1	3	2	5
G85E	0	0	0	1	1	0
N1303K	0	0	1	1	0	0
Other	0	0	0	19	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

466 PWCF

Mutations	Number Patients Identified	Percentage Patients Identified
F508del	420	90.1%
G551D	36	7.7%
G542X	25	5.4%
R117H	23	4.9%
G85E	6	1.3%

Note: Because people have two genes, patients are counted twice, once for each gene. The total number of patients is 466.

Our high percentage of F508del is in keeping with the international registries from European derived populations. In total, only 46 persons in New Zealand do not have at least one F508del mutation. Looking at the gene mutations recorded in 2018 Registry, 28 PWCF of the 466 PWCF who have been genotyped (6%) would not be detected by our current newborn screening programme.

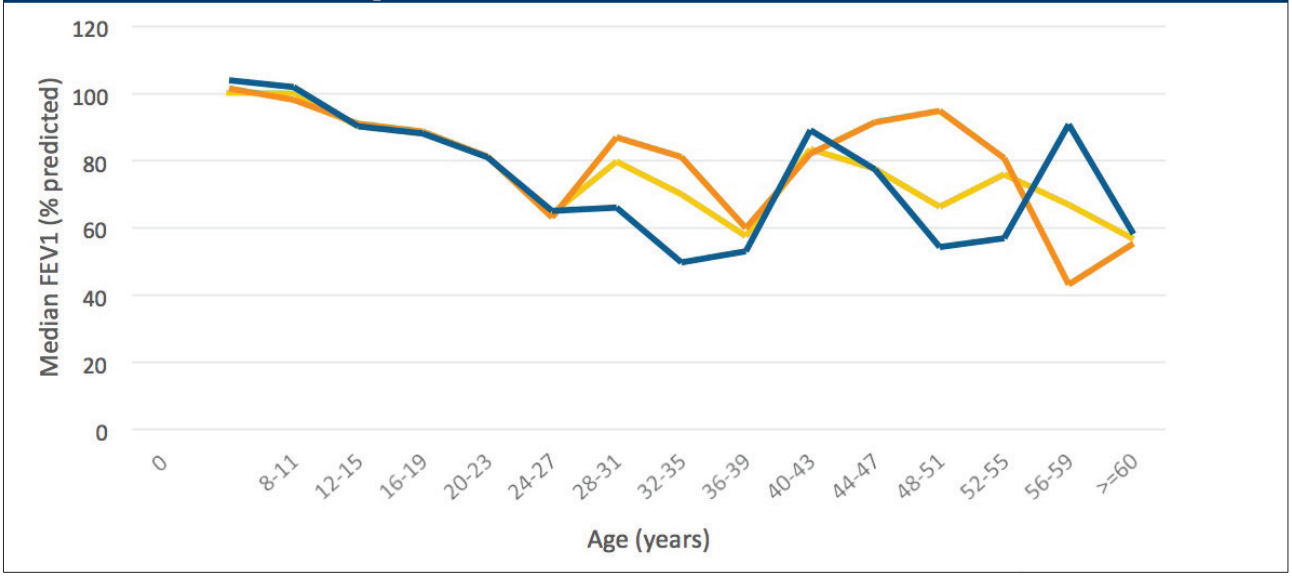
3. Respiratory

385 PWCF

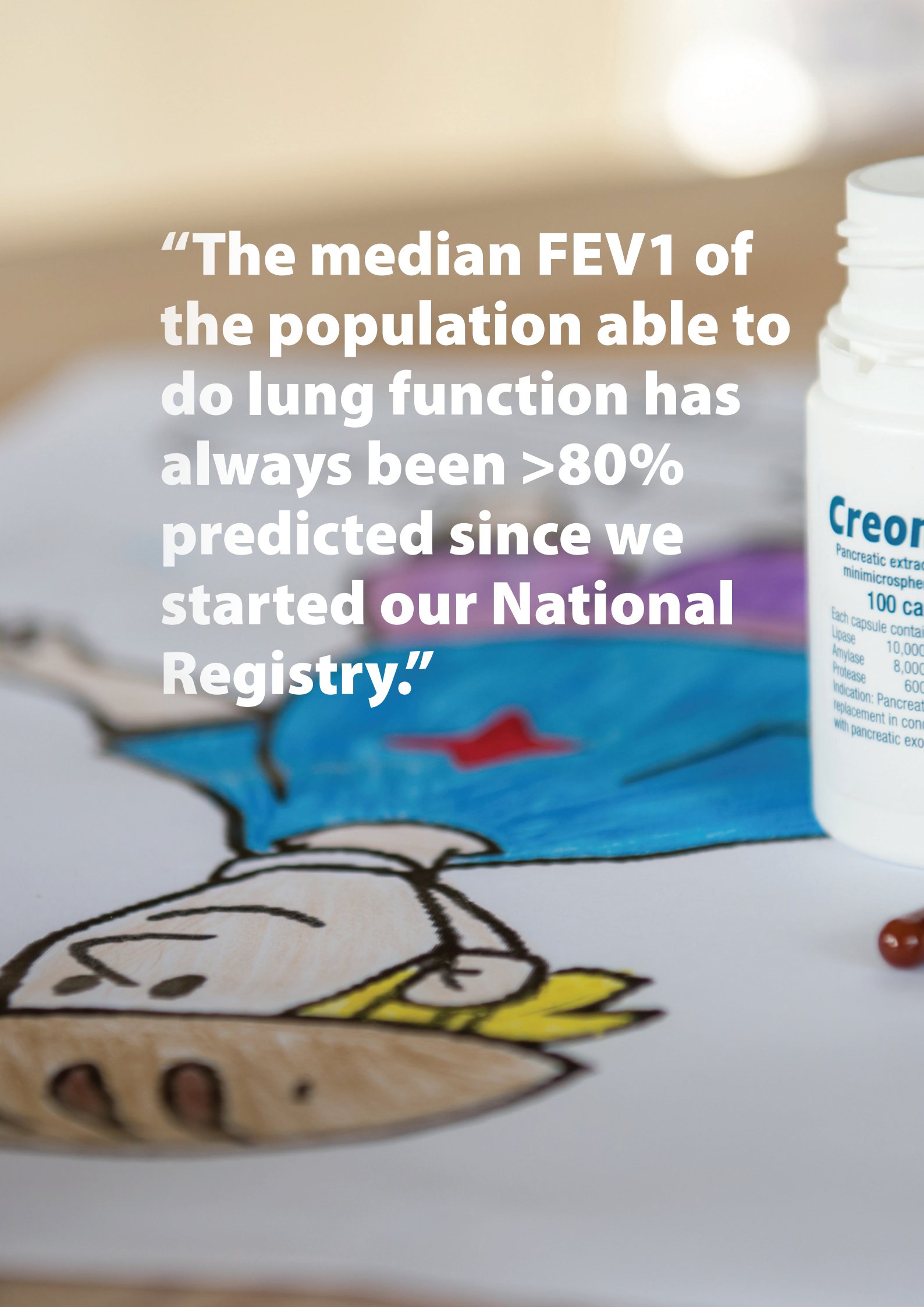
Age Group	All		Female		Male	
	Number in age group	Median FEV1	Number in age group	Median FEV1	Number in age group	Median FEV1
6-7	44	102.3	23	101.5	21	104.0
8-11	54	100.3	24	98.1	30	101.9
12-15	50	90.3	24	91.0	26	90.3
16-19	50	88.2	21	88.7	29	88.1
20-23	43	81.2	15	81.2	28	81.0
24-27	34	64.2	11	63.1	23	65.1
28-31	21	79.8	14	87.0	7	66.00
32-35	24	70.0	11	81.1	13	49.7
36-39	15	57.6	9	59.9	6	53.00
40-43	13	83.5	5	82.1	8	89.1
44-47	12	77.6	3	91.4	9	77.4
48-51	7	66.3	2	94.8	5	54.3
52-55	6	76.00	3	80.7	3	56.9
56-59	2	66.9	1	43.1	1	90.8
>=60	10	56.7	5	55.3	5	58.2
Totals	**385**		**171**		**214**	

Median FEV1 (% predicted)

385 PWCF



The median FEV_1 of the CF population able to do lung function has always been 80% predicted since we started our 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. FEV_1 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

434 PWCF

4.1 Paediatric BMI

194 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	36	87.8	<4	10	80.7	<4	26	88.6
4-7	55	65.5	4-7	28	65.7	4-7	27	62.3
8-11	55	61.00	8-11	24	65.9	8-11	31	49.2
12-15	48	48.7	12-15	22	53.00	12-15	26	44.7
Totals	**194**			**84**			**110**	

4.2 Adult BMI

240 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	50	22.0	16-19	21	22.7	16-19	29	21.0
20-23	43	22.4	20-23	15	23.1	20-23	28	21.9
24-27	34	21.9	24-27	11	21.2	24-27	23	22.3
28-31	21	22.5	28-31	14	22.3	28-31	7	24.8
32-35	25	22.1	32-35	12	21.4	32-35	13	22.5
36-39	15	23.2	36-39	9	21.8	36-39	6	23.8
40-43	13	25.5	40-43	5	22.4	40-43	8	26.1
44-47	13	24.3	44-47	3	24.3	44-47	10	24.4
48-51	7	23.5	48-51	2	25.6	48-51	5	23.5
52-55	6	26.6	52-55	3	24.6	52-55	3	26.9
56-59	2	28.3	56-59	1	24.3	56-59	1	32.2
>=60	11	22.1	>=60	6	23.9	>=60	5	22.1
Totals	**240**			**102**			**138**	

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

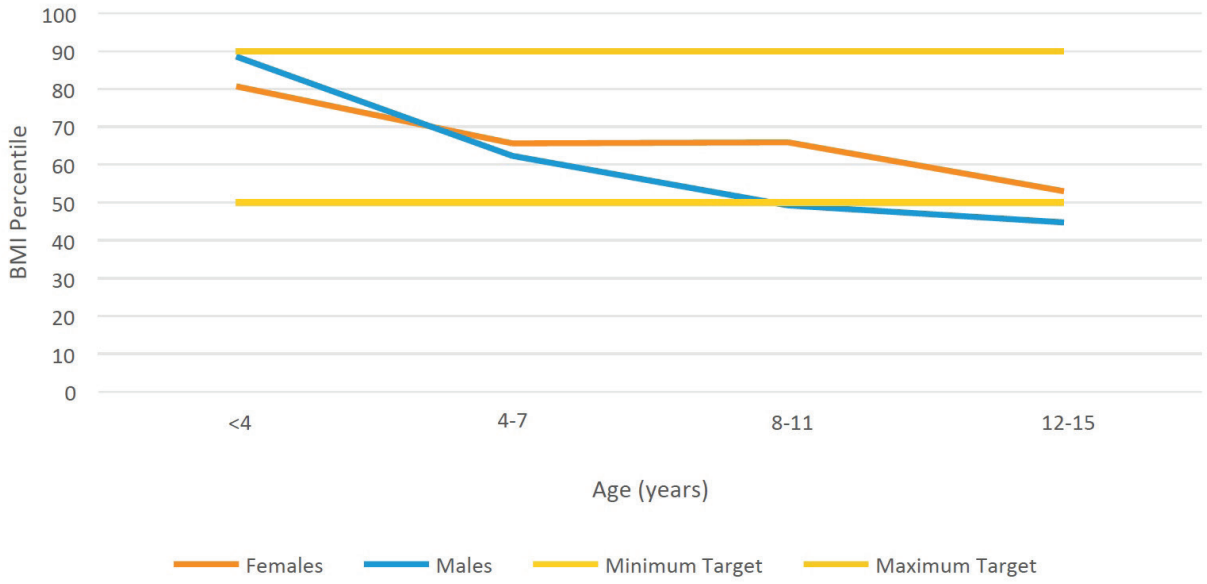
This is also reflected in the *'Nutrition Guidelines for Cystic Fibrosis in Australia and New Zealand'* (https://www.thoracic.org.au/documents/item/1045)

For infants under 4 years of age, the median BMI is 87.8 percentile. For children and adolescents, the median BMI is 62.4 percentile. For adults, 47.1 % of males and 58.8 % of females are above the minimum target range.


LIVE

4.3 Median BMI Percentile <16 Years

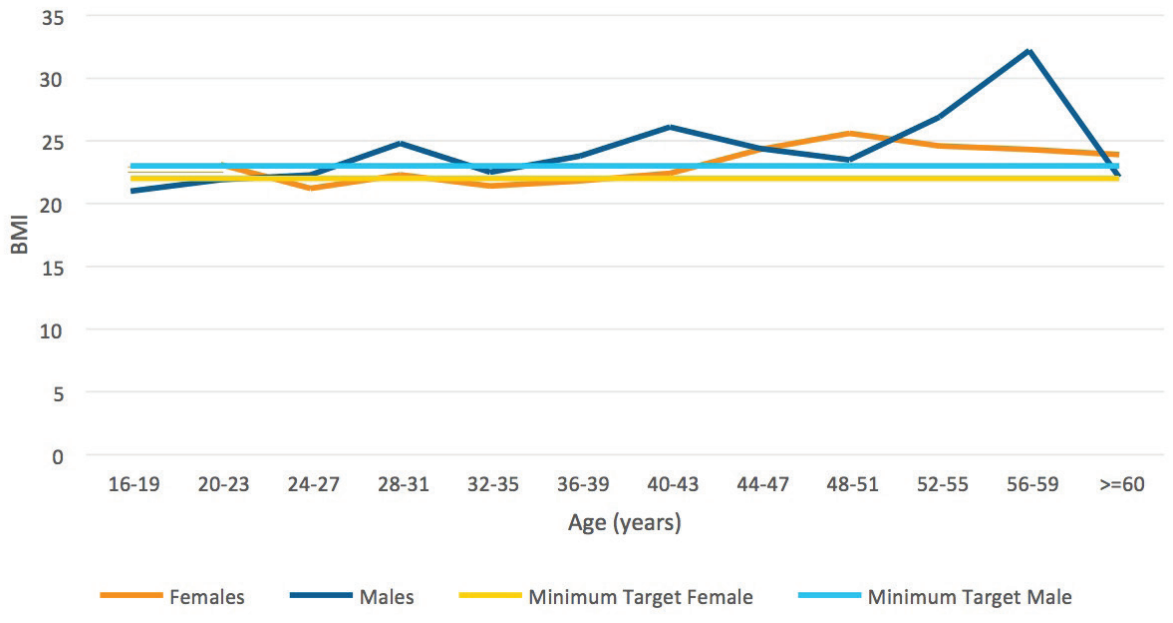
194 PWCF



The optimal BMI for children 2 – 15 years is 50 - 91 percentile using the WHO-NZ growth chart.

4.4 Median BMI Percentile >=16 Years

240 PWCF



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

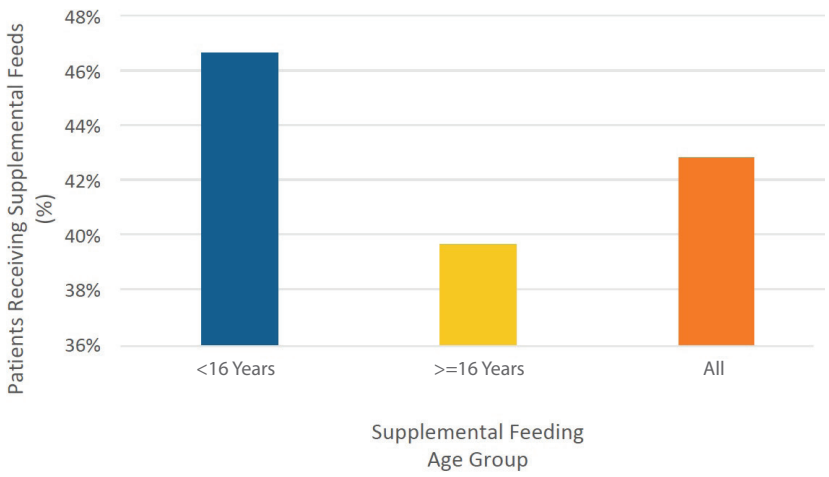
4.5 Supplemental Feeding

458 PWCF

	<16 years, n =212	
	Yes	**% <16 years supplemented**
Supplemental Feeding	**96**	**46.6%**
Nasogastric	7	3.3%
Gastrostomy	19	9.0%
Oral	87	41.0%
	>16 =years, n = 246	
	Yes	**% >=16 years supplemented**
Supplemental Feeding	**89**	**39.6%**
Nasogastric	1	0.4%
Gastrostomy	17	6.9%
Oral	78	31.7%

4.6 Supplemental Feeding by Age Group

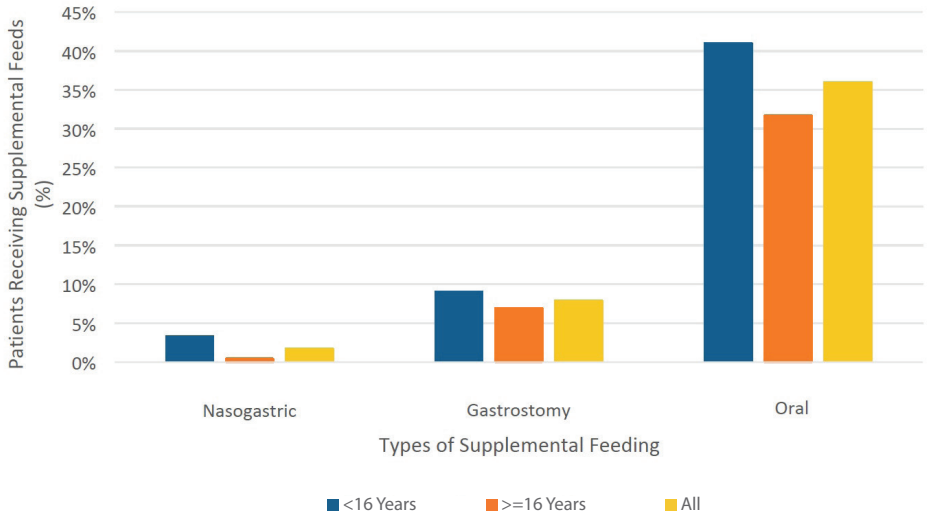
458 PWCF



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

4.7 Supplemental Feeding by Type

458 PWCF



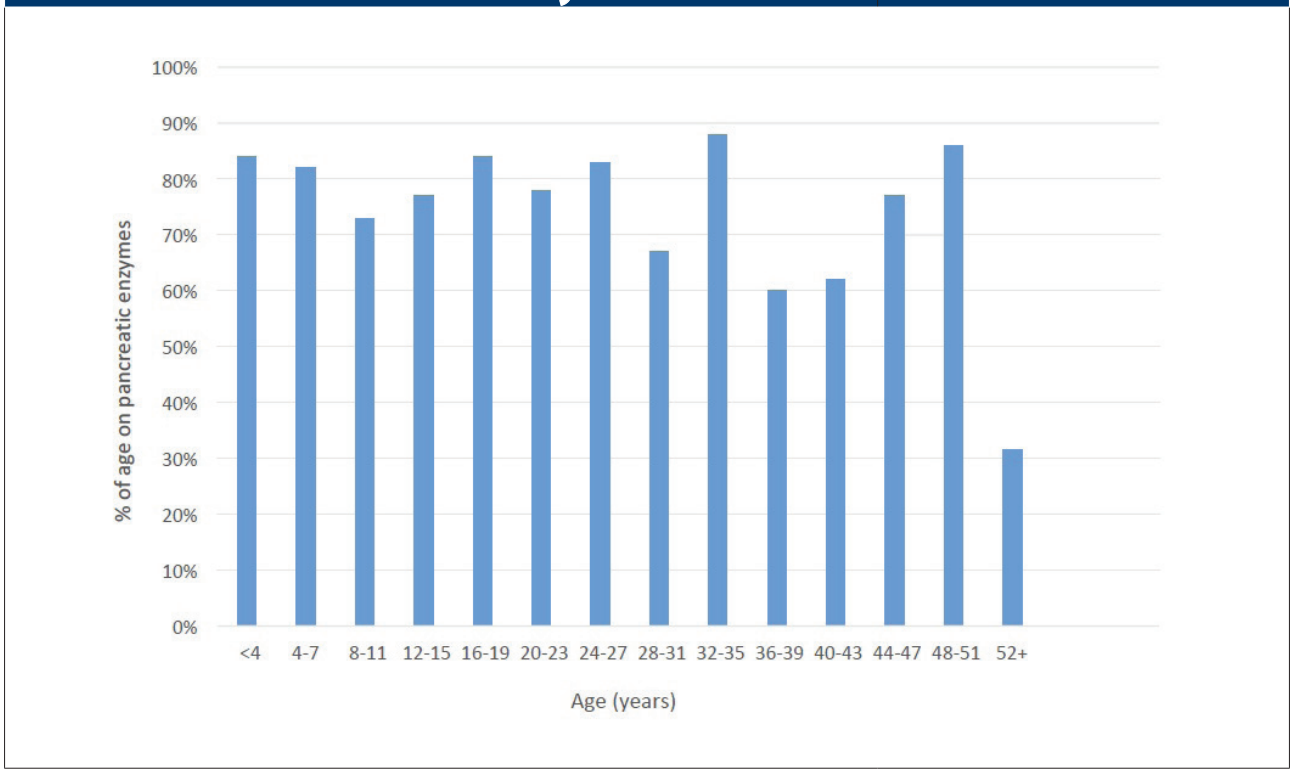
5. Pancreatic Enzymes

458 PWCF

Age Group	Number in age group	On Pancreatic Enzymes	Percent of age group	Percent of CF population
<4	50	42	84.0%	9.2%
4-7	55	45	82.0%	9.8%
8-11	55	40	73.0%	8.7%
12-15	52	40	77.0%	8.7%
16-19	51	43	84.0%	9.4%
20-23	46	36	78.0%	7.9%
24-27	35	29	83.0%	6.3%
28-31	21	14	67.0%	3.1%
32-35	26	23	88.0%	5.0%
36-39	15	9	60.0%	2.0%
40-43	13	8	62.0%	1.7%
44-47	13	10	77.0%	2.2%
48-51	7	6	86.0%	1.3%
52-55	6	1	17.0%	0.2%
56-59	2	0	0%	0.0%
>=60	11	5	45%	1.1%
Totals	**407**	**328**		**77%**

Patients on Pancreatic Enzymes

458 PWCF

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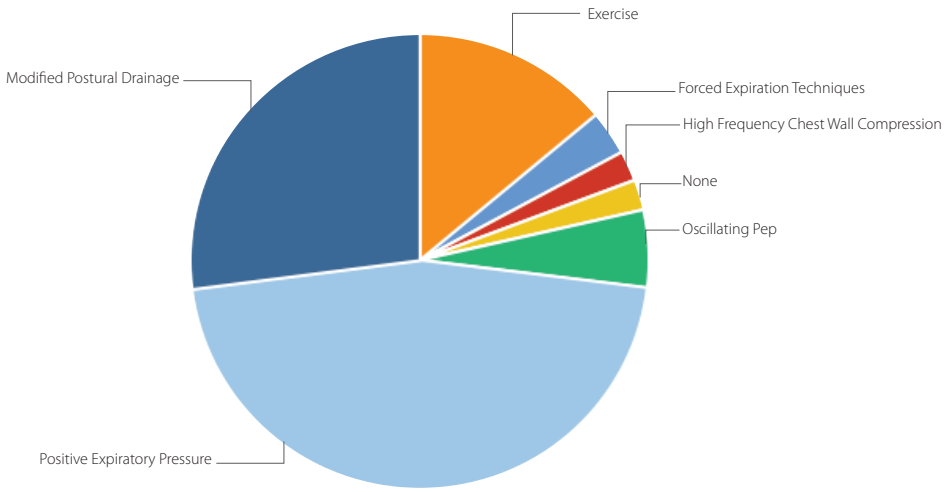
6. Airway Clearance Techniques

6.1 Primary Airway Clearance — 458 PWCF

	<16 years	
	Number of PWCF	Percent of PWCF
Positive Expiratory Pressure	128	60.4%
Modified Postural Drainage	75	35.4%
Exercise	39	18.4%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	15	7.1%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	9	4.2%
High Frequency Chest Wall Compression (e.g. vest)	6	2.8%
None	6	2.8%

	>= 16 years	
	Number of PWCF	Percent of PWCF
Positive Expiratory Pressure	92	37%
Modified Postural Drainage	1	0.4%
Exercise	59	24%
Oscillating Pep (e.g.Flutter, Acapella, IPV)	38	15.4%
Forced Expiration Techniques (e.g. huff cough, active cycle breathing, autogenic drainage)	48	19.5%
High Frequency Chest Wall Compression (e.g. vest)	1	0.4%
None	30	12.2%

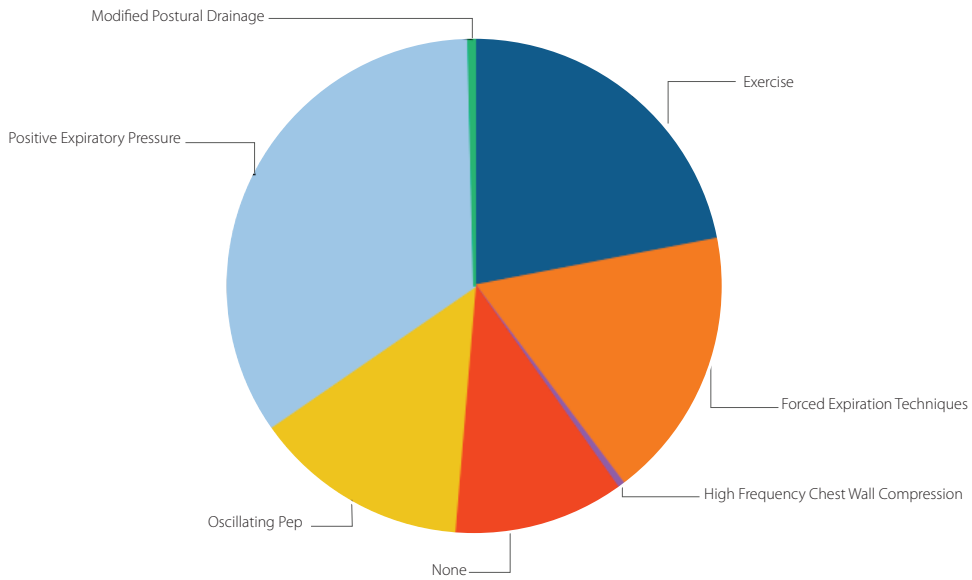
6.2 Primary Airway Clearance <16 Years — 212 PWCF



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

6.3 Primary Airway Clearance >=16 Years

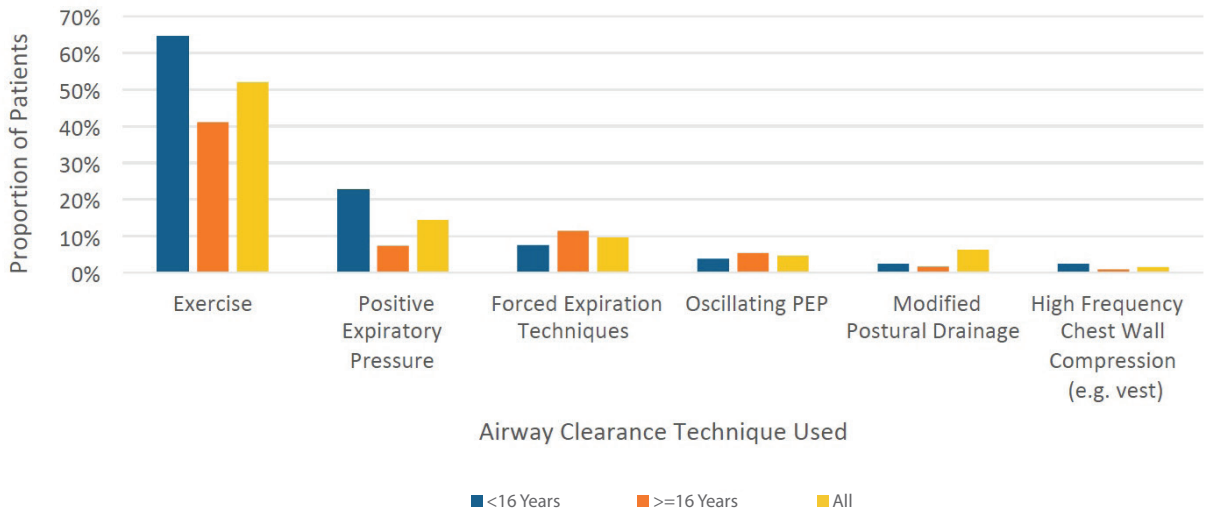
246 PWCF



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

458 PWCF

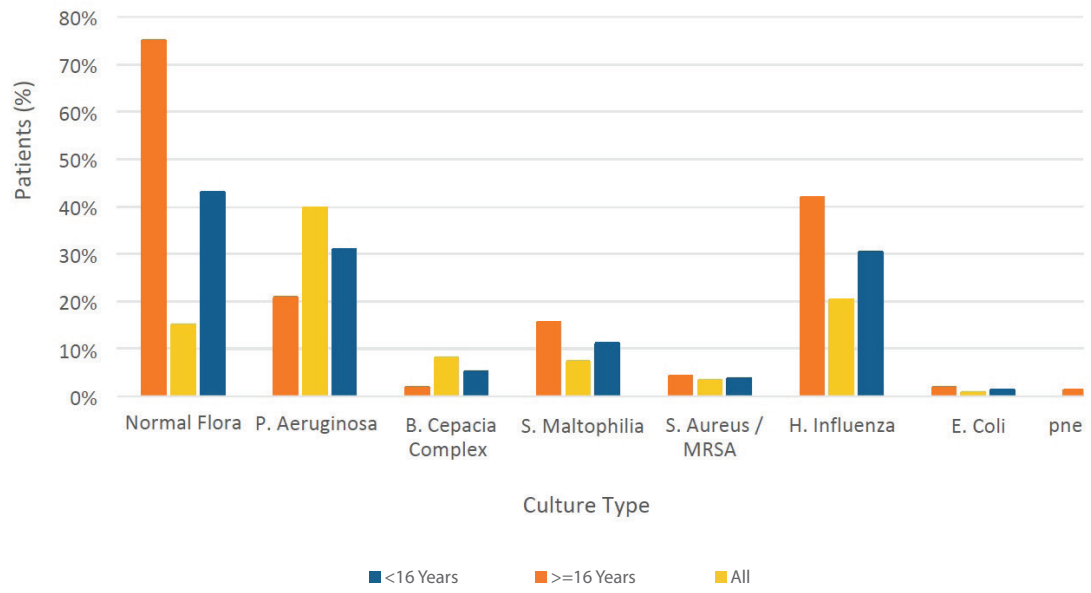


7. Microbiology

Microbiology — 458 PWCF

	<16 years PWCF		>=16 years PWCF		All
	Number	Percent	Number	Percent	Number
Normal Flora	158	75.0%	37	15.0%	195
Haemophilus Influenza	89	42.0%	50	20.3%	139
E. Coli	4	1.9%	2	0.8%	6
Klebsiella Pneumoniae	3	1.4%	3	1.2%	6
Stenotrophomonas Maltophilia	33	15.6%	18	7.3%	51
Pseudomonas Aeruginosa	**44**	**20.8%**	**98**	**39.8%**	**142**
Mucoid	10	4.7%	68	27.6%	78
Non Mucoid	28	13.2%	52	21.1%	80
Staphylococcus Aureus	**129**	**60.8%**	**124**	**50.4%**	**253**
MSSA	120	56.6%	116	47.2%	236
MRSA	8	4.2%	8	3.3%	17
Burkholderia Cepacia Complex	**4**	**1.9%**	**20**	**8.1%**	**24**
Cenocepacia	1	0.5%	3	1.2%	4
Multivorans	3	1.4%	9	3.7%	12
Other	0	0.0%	3	1.2%	3

7.1 Bacterial Culture Prevalence — 458 PWCF

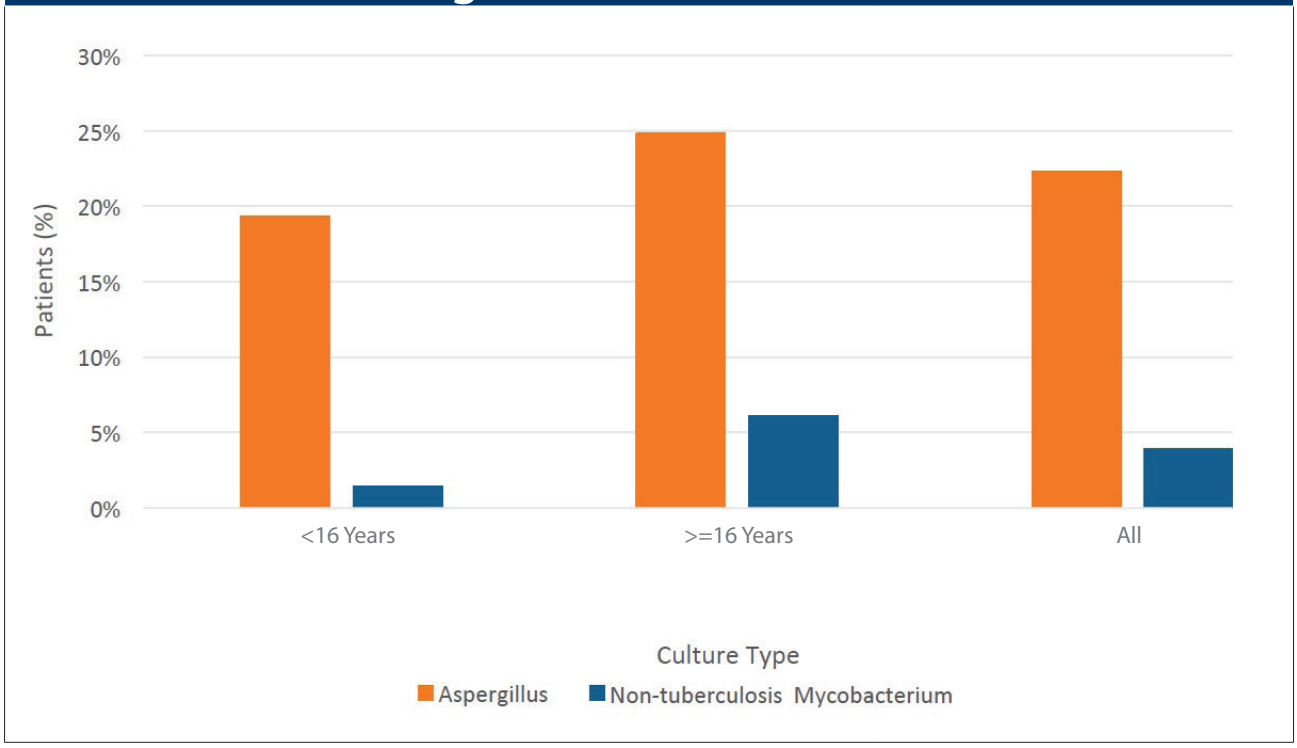


NOTE: The percentages of population with CF having had specific respiratory pathogens identified such as Staphylococcus Aureus, Pseudomonas Aeruginosa etc. are very similar to the percentages presented in the Australian 2017 Registry, with the exception of much higher percentages of Haemophilus Influenza in New Zealand. This pathogen is also higher in our young children and lower in our adults. Pseudomonas Aeruginosa is found in 20.8% of the children and increases to 39.8% in adults. Our MRSA rates are relatively low at 3.7% overall.

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

7.2 Nonbacterial/Fungal Prevalence

501 PWCF



8. Medications

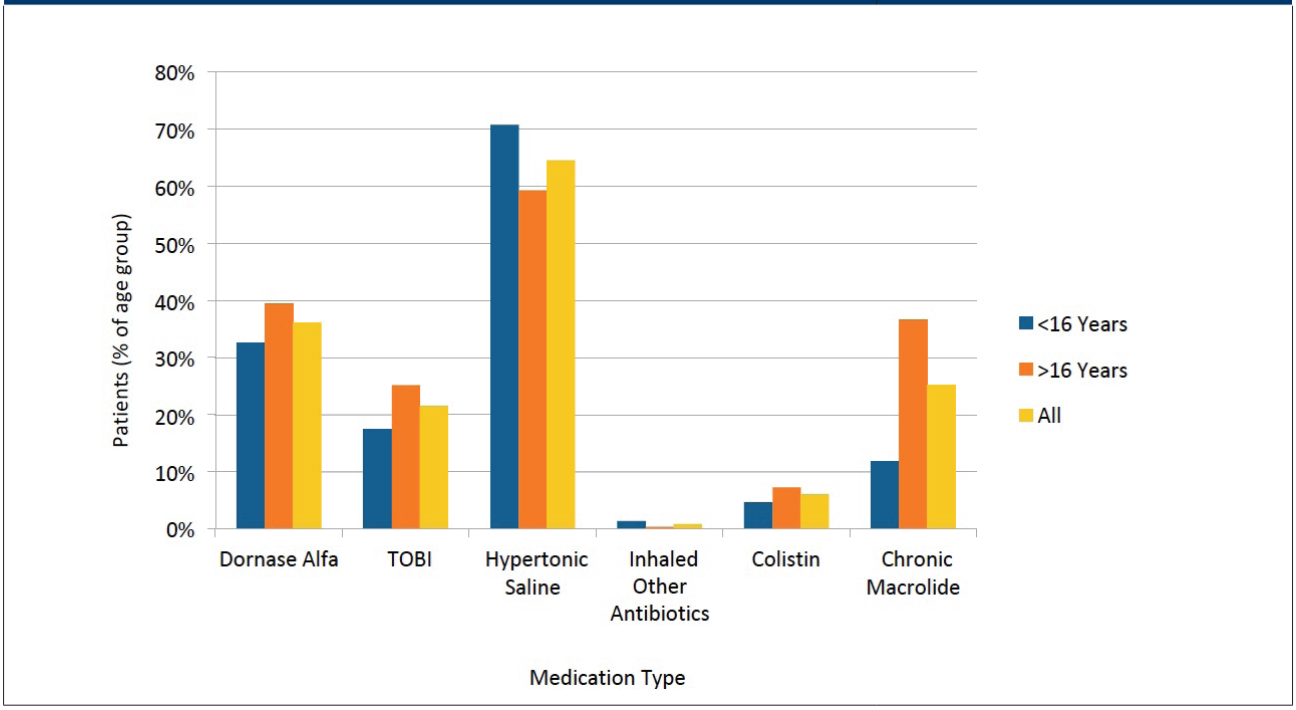
Medications

458 PWCF

Medication	<16 Years	>=16 Years	All
Hypertonic Saline	70.8%	59.3%	64.6%
Dornase alfa	32.5%	39.4%	36.2%
TOBI	17.5%	25.2%	21.6%
Inhaled Other Antibiotics	1.4%	0.4%	0.9%
Chronic Macrolide	11.8%	36.6%	25.1%
"Corticosteroids Other (Inhaled and combination treatments)"	15.6%	12.2%	13.8%
Corticosteroids Oral	4.2%	3.3%	3.7%
Influenza Vaccine	79.2%	55.7%	66.6%

8.1 Medications Prescribed

458 PWCF



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

Note: We have listed more medication types here than 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 had no access to some newer medications except on research programmes.

9. Intravenous Antibiotic Treatment

9.1 Home IV Days

459 PWCF

Age	Number In Age Group	Number Who Had IV Days	Percent PWCF Who Had IV Days	Mean Days For PWCF Who Have Had IV Days	Mean Days For All
<4	49	3	6%	17.3	1.1
4-7	56	10	18%	13.8	2.5
8-11	55	10	18%	17.9	3.3
12-15	52	14	27%	26.7	7.2
16-19	51	13	25%	16.8	4.3
20-23	45	12	27%	19	5.1
24-27	36	10	28%	12.6	3.5
28-31	21	7	33%	23.4	7.8
32-35	26	6	23%	21.7	5
36-39	16	6	38%	23.2	8.7
40-43	13	1	8%	12	0.9
44-47	13	4	31%	14.8	4.5
48-51	7	0	0%	0.0	0
52-55	6	0	0%	0.0	0
56-59	2	0	0%	0.0	0
>=60	11	3	27%	11.3	3.1
Totals	**459**	**99**	**22%**	**18.7**	**4**

9.2 Hospital IV Days

Age	Number In Age Group	Number Who Had IV Days	Percent PWCF Who Had IV	Mean Days For PWCF Who Have Had IV Days	Mean Days For All
<4	49	12	24%	15.8	3.9
4-7	56	15	27%	12.9	3.5
8-15	55	18	33%	11.0	3.6
16-18	52	22	42%	32.2	13.6
16-19	51	24	47%	25.3	11.9
20-23	45	19	42%	13.3	5.6
24-27	36	18	50%	26.9	13.4
28-31	21	8	38%	16.4	6.2
32-35	26	9	35%	28.6	9.9
36-39	16	7	44%	21.6	9.4
40-43	13	2	15%	8.0	1.2
44-47	13	4	31%	12.8	3.9
48-51	7	1	14%	14	2
52-55	6	0	0%	0.0	0
56-59	2	1	50%	14	7
>=60	11	5	45%	26.2	11.9
Totals	**459**	**165**	**36%**	**20.6**	**7.4**

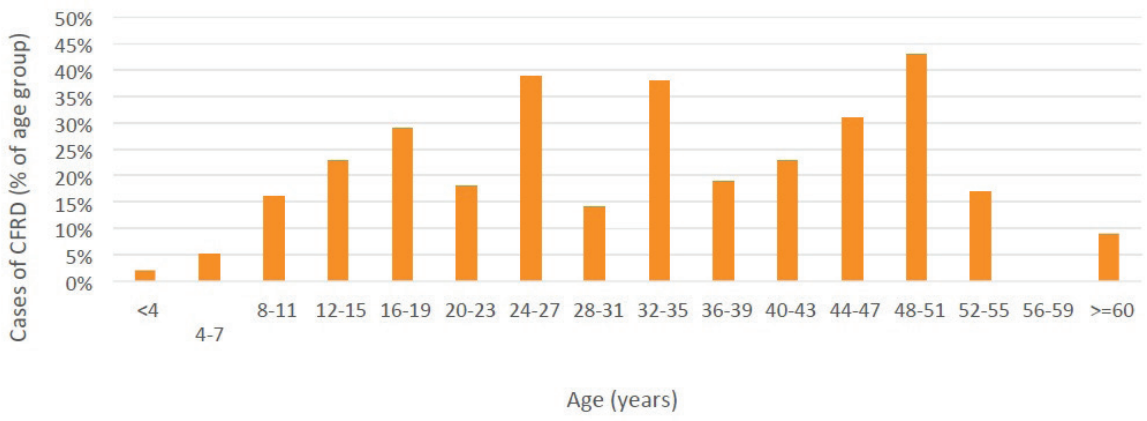
10. Complications

10.1 CF Related Diabetes (CFRD) 459 PWCF

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<4	49	1	2%	0.2%
4-7	56	3	5%	0.7%
8-11	55	9	16%	2.0%
12-15	52	12	23%	2.6%
16-19	51	15	29%	3.3%
20-23	45	8	18%	3.3%
24-27	36	14	39%	3.0%
28-31	21	3	14%	0.7%
32-35	26	10	38%	2.2%
36-39	16	3	19%	0.7%
40-43	13	3	23%	0.7%
44-47	13	4	31%	0.9%
48-51	7	3	43%	0.7%
52-55	6	1	17%	0.2%
56-59	2	0	0%	0.0%
>=60	11	1	9%	0.2%

Age Group	Number in group	Number with CFRD	Percent of age group	Percent of PWCF
<16	212	25	12.0%	5.4%
>=16	247	65	26.0%	14.2%
Total	**459**	**90**		**19.6%**

10.2 CF Related Diabetes by Age 459 PWCF



The prevalence of CFRD has increased over time in the Registry data from 13.5% in 2012 to 19.6% this year. This may reflect; the use of more sensitive measurement of abnormal glucose abnormalities with continuous glucose monitoring; a recognition that instituting insulin therapy earlier when abnormalities first occur is associated with better intermediate outcomes; and 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

428 PWCF

		Normal		Abnormal		Unknown	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	212	73	34.4%	19	9.0%	120	56.6%
Adults	246	10	41.1%	15	6.1%	221	89.8%
Total	**458**	**83**	**19.4%**	**34**	**7.9%**	**341**	**79.7%**

The 'unknown' 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

458 PWCF

		Normal		Abnormal		Unknown	
	Number in age group	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF	Number of PWCF	Percent of PWCF
Paediatrics	212	27	12.7%	8	3.8%	177	83.5%
Adults	246	49	19.9%	28	11.4%	169	68.7%
Total	**458**	**76**	**17.8%**	**36**	**8.4%**	**346**	**80.8%**

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