



Cystic Fibrosis Registry of Ireland

2012 Annual Report

Preface

Over the past decade, the Cystic Fibrosis Registry of Ireland has taken proactive steps towards proving its value to the health service and to the research community. In 2012, we succeeded in securing additional funding from industry and from Cystic Fibrosis Ireland that will contribute towards the funding of day to day operations over the next three years. In addition we secured Lotto funding in December 2012 that will allow us to update our now obsolete registry technology. It is hoped by end of 2014 we will have built a state of the art, encounter based technology platform on top of that currently being developed by the European Cystic Fibrosis Registry. This is a very important step for us as it brings us closer to the international research community while maintaining ownership and security of our Irish data sets. To substantiate true comparisons between countries there needs to be an international standardisation of definitions and data quality across the global CF community which is currently underway.

The only way to achieve recognition of a patient registries' worth is to prove it. In 2012 we have set the foundation. 2013 will see the building blocks being put in place and 2014 will hopefully see the implementation of new technologies and processes that will make a significant contribution towards improving the quality of information about the current and future status of cystic fibrosis in Ireland. The current Report shows that the Irish cystic fibrosis population remains stable at fifty two percent (52%) of the population being either 18 years or older. Survivorship in successive birth cohorts continues to improve and this should indicate future increases in the adult population. The registry intends to be positioned by 2014 to be a true example of a patient registry that is an invaluable resource providing key data to support the planning process for future services, as well as the implementation and monitoring of new treatments, and the continued support of the research community.



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Glossary

AA	Annual Assessment
ABPA	Allergic Bronchopulmonary Aspergillosis
Adult	Aged 18 years or older (≥ 18)
BAL	Bronchoalveolar lavage
BMI	Body Mass Index
CF	Cystic Fibrosis
CFI	Cystic Fibrosis Ireland
CFRI	Cystic Fibrosis Registry of Ireland
CRA	Clinical Research Associate
DIOS	Distal Intestinal Obstruction Syndrome
FH	Family history
FEV ₁	Forced Expiratory Volume in one second
FVC	Forced expiratory Vital Capacity
GORD	Gastro-Oesophageal Reflux Disease
GI	Gastrointestinal symptoms
H2RA	H2-receptor antagonists
IV	Intravenous
MI	Meconium ileus
MRSA	Methicillin Resistant <i>Staphylococcus aureus</i>
NBS	Newborn screening
NSAID	Non-steroidal anti-inflammatory drug
Paediatric	Aged under 18 years (< 18)
PEP mask	Positive expiratory pressure mask
PPI	Proton pump inhibitors
PWCF	Persons with cystic fibrosis
RESP	Respiratory symptoms

Summary of the 2012 CFRI dataset

The number of persons living with cystic fibrosis (CF) in the Republic of Ireland continues to increase. On the last day of 2012, there were 1,140 individuals registered with the Cystic Fibrosis Registry of Ireland (CFRI). This represents 92.7% of persons with CF (PWCF) known to be alive on the 31st of December 2012. The total number of known PWCF is established by the CFRI annually, as a result of undertaking a nationwide census of Specialist CF Centres and Shared Care Centres.

Over half of CFRI-registered PWCF were aged 18 years or older (52.2%). Between 2008 and 2010, the number of children (<18 years of age) with CF was largely unchanged, but consecutive increases have been noted in 2011 and 2012. The adult population continues to increase on an annual basis; in 2012 there were 37 more adults attending Specialist CF Centres than in 2011. In 2012, the median age of adults was 27.9 years.

Four double lung transplants were reported in 2012. Eighteen deaths were recorded in PWCF registered with the CFRI in 2012. This is similar to the number of deaths reported in other years. The median age at which these 18 individuals died was 25.2 years (range 5-42 years).

Table 1: Summary of 2012 CFRI data

	2008	2009	2010	2011	2012
CF persons registered (n)¹					
Children²	496	496	494	516	545
Adults³	508	531	550	558	595
Total	1004	1027	1044	1074	1140
(% census)	(87.9%)	(89.2%)	(90.0%)	(90.9%)	(92.7%)
Median age (years)					
Children²	9.5	9.9	10.2	9.9	9.6
Adults³	25.5	26.0	26.6	27.2	27.9
Overall	17.9	18.6	18.8	18.8	19.1
Gender (%)					
Female	43.9%	42.4%	42.9%	43.0%	42.7%
Male	56.1%	57.6%	57.1%	57.0%	57.3%
Deaths in CFRI-registered persons (n)					
	17	17	16	26	18
Median age at death (n)					
	23.5	25.6	28.1	23.5	25.2
Recorded annual clinical data (%)					
Children²	438	454	454	468	476
(% all children)	(86.2%)	(91.5%)	(91.9%)	(91.1%)	(87.3%)
Adults³	375	411	412	388	405
(% all adults)	(75.6%)	(77.4%)	(74.9%)	(69.4%)	(72.5%)
Overall	813	865	866	856	881
	(81.0%)	(84.2%)	(83.0%)	(79.7%)	(77.3%)

¹ Person with CF registered with the CFRI, who have no recorded date of death on/before 31st December 2008/2009/2010/2011/2012 respectively. ² Children (<18 years). ³ Adults (≥18 years).

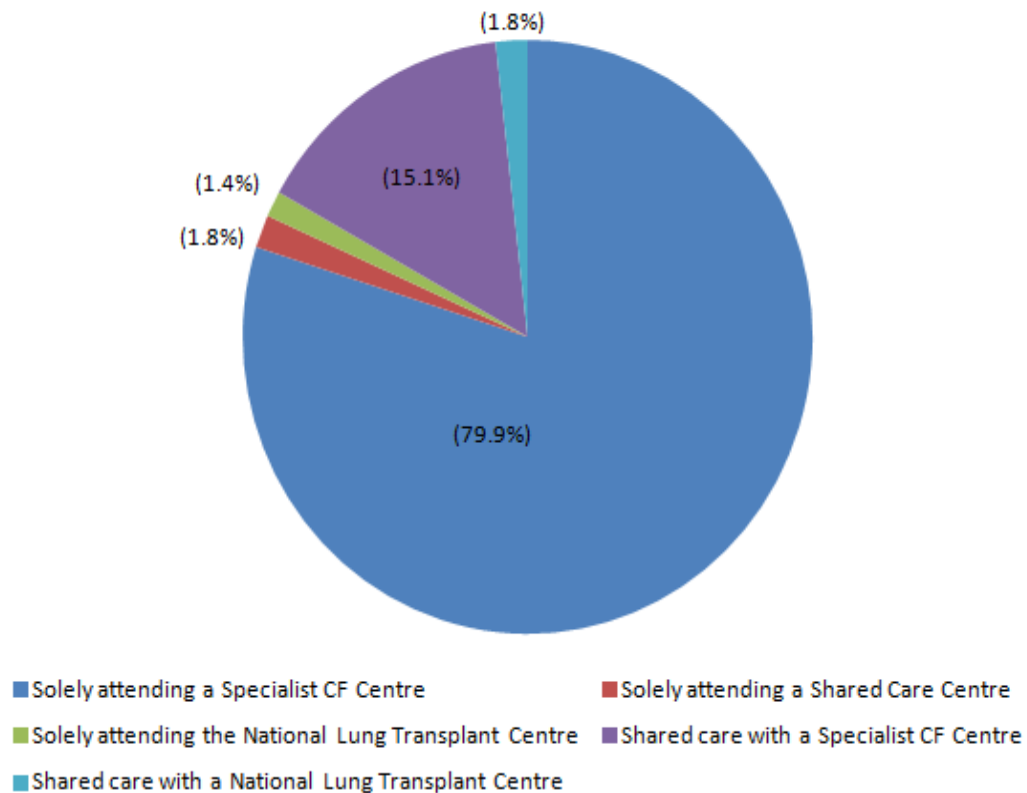
Specialist CF Centres

In response to the 'Pollock Report' published in 2005 by the Cystic Fibrosis Association of Ireland (CFAI), the Health Services Executive (HSE) published a report entitled 'Services for People with Cystic Fibrosis in Ireland – Conclusions of a Working Group established by the Health Services Executive' in 2009. This report recommended a configuration of CF services that involved National Referral Centres (adult and paediatric), Specialist CF Centres, and Shared Care Centres (offering shared care to PWCF under the supervision of a Specialist CF Centre).

In 2012, 948 PWCF were registered at a single location for CF care (83.2%). The remainder (192, 16.8%) opted to receive CF care that was shared between two or more locations.

Figure 1 shows the breakdown in attendance at Specialist CF Centres and Shared Care Centres in 2012. Four out of every five PWCF solely attended one Specialist CF Centre.

Figure 1: Proportions of patients attending Specialist CF Centres and Shared Care Centres in 2012.



The names and designation of hospitals attended by PWCF registered at a single location for CF care are provided in Table 2.

Table 2: Hospitals attended by PWCF availing of CF care in a single hospital in 2012 (n=948)

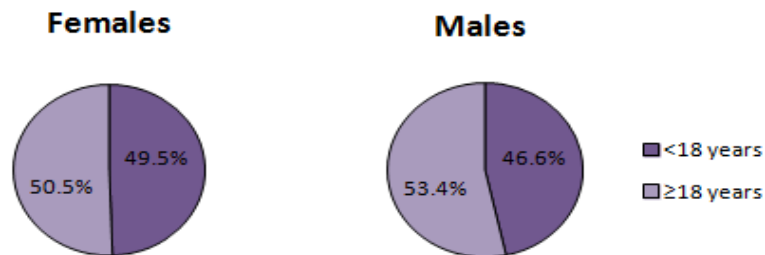
		Adults	Children	Specialist CF Centre	National Referral Centre
1	Beaumont Hospital	Y		Y	
2	Cavan General Hospital	Y			
3	Cork University Hospital	Y	Y	Y	
4	Mayo General Hospital	Y	Y		
5	Midwestern Regional Hospital	Y	Y	Y	
6	Adelaide and Meath Hospital Dublin, Incorporating the National Children's Hospital		Y	Y	
7	Our Lady of Lourdes Hospital, Drogheda		Y		
8	Our Lady's Children's Hospital, Crumlin		Y	Y	Y
9	Sligo General Hospital		Y		
10	St Vincent's University Hospital	Y		Y	Y
11	Temple Street Children's University Hospital		Y	Y	
12	University College Hospital, Galway	Y	Y	Y	
13	Waterford Regional Hospital		Y		
	National Lung Transplant Centre:				
14	Mater Misericordiae University Hospital*	Y			

*The CFRI do not collect clinical data for PWCF attending this hospital.

Age distribution by gender

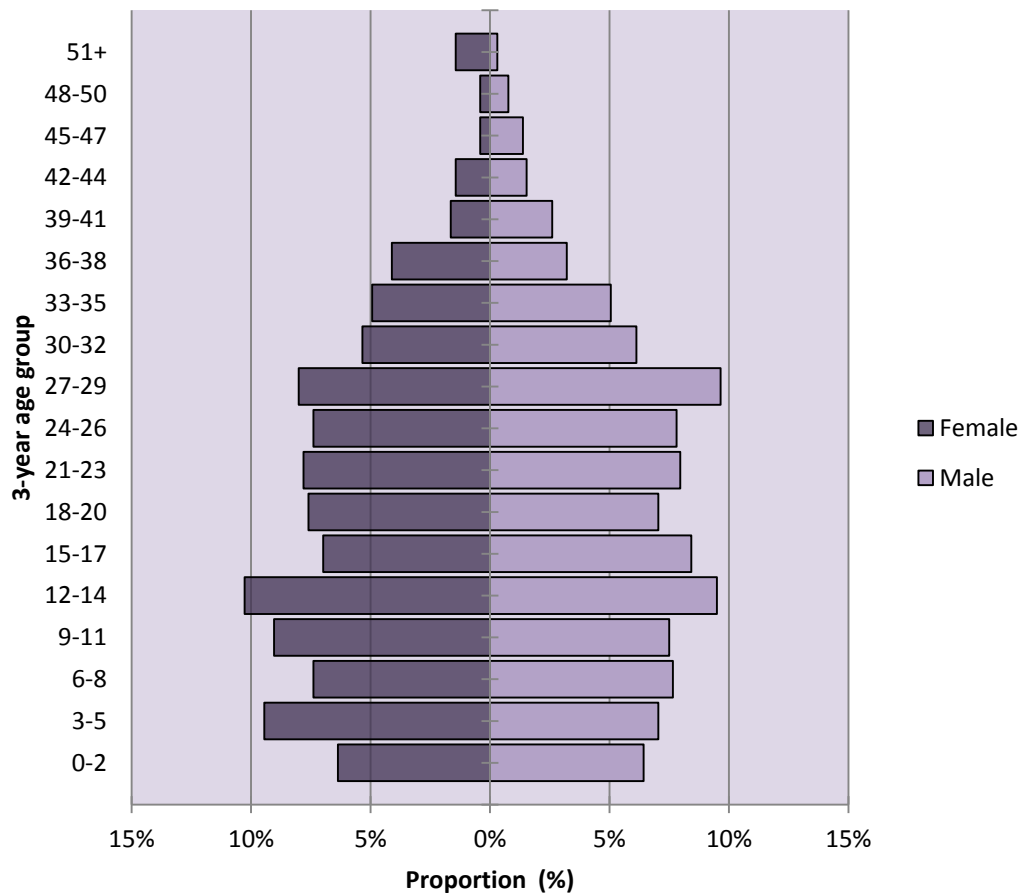
In Figure 2, age is calculated on the last day of 2012. A slightly greater proportion of males are aged ≥ 18 years than < 18 years, whereas the ratio of female children (< 18 years) to female adults (≥ 18 years) is 1:1.

Figure 2: Age distribution by gender



Of the 1,140 PWCF on the registry in 2012, 79.5% were under 30 years of age. Figure 3 shows the age distribution of PWCF by three-year age band. The three-year age bands with the largest proportions of PWCF were the 12-14 and 27-29 year olds. For females, the largest proportion of PWCF was in the 12-14 age band and for males, the 27-29 age band.

Figure 3: Age and gender distribution



Diagnosis

Just over two-thirds of PWCF on the registry in 2012 (1,140) were diagnosed before age one. Comparing age at diagnosis for PWCF who were <18 years and ≥18 years in 2012, we find that 70.5% of <18s and 58.2% of ≥18 year olds were diagnosed before the age of one. One in five PWCF aged ≥18 years in 2012 were diagnosed at age five or older.

Table 3: Age at diagnosis

	Overall (n=1140)	<18 years in 2012 (n=545)	≥18 years in 2012 (n=595)
Birth - 2 months	41.5%	47.7%	35.8%
3 - 5 months	11.9%	11.6%	12.3%
6 - 11 months	10.6%	11.2%	10.1%
1 year	8.3%	10.8%	6.1%
2 years	6.1%	5.9%	6.2%
3 years	3.7%	4.8%	2.7%
4 years	2.7%	2.2%	3.2%
5+ years	12.5%	4.8%	19.7%
Not known	2.6%	1.1%	4.0%

Table 4 summarises the symptoms experienced by PWCF that led to a diagnosis of CF. Gastrointestinal and/or respiratory symptoms, or both, were reported by 57.5% of PWCF. PWCF had a median age of diagnosis of 4.2 months. Patients with respiratory symptoms had a median age at diagnosis of nearly 2 years. Eighteen PWCF on the registry in 2012 were diagnosed as a result of the neonatal cystic fibrosis screening programme in 2012.

Table 4: Symptoms at diagnosis

	% (n=1140)	Median age at diagnosis (months)
Gastrointestinal symptoms	20.5%	6.9
Respiratory symptoms	19.7%	22.7
Respiratory and gastrointestinal symptoms	17.3%	8.5
Family history	14.5%	1.4
Meconium ileus	13.8%	0.5
Family history and ≥1 symptom	7.2%	2.3
Unknown	3.2%	9.6
Neonatal & antenatal screening	3.2%	0.5
Other	0.5%	9.6
Total		4.2

Twenty-two PWCF registered with the CFRI were diagnosed in 2012 as a result of the National Newborn Bloodspot Screening Programme (Table 5). This is less than that reported by Temple Street Children's University Hospital, who manage the programme (n=27). The CFRI can occasionally experience a time lag in offering parents of newborns the opportunity to register.

Table 5: Age of PWCF diagnosed in 2012 and factors leading to a CF diagnosis

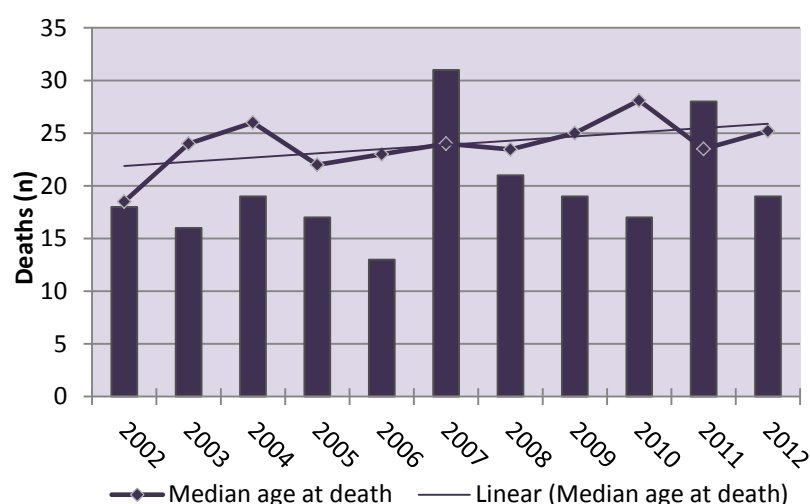
	N	Reason leading to diagnosis of CF
Birth - 2 months	22	All newborn screening
3 - 11 months	0	-
1 - 4 years	7	3 family history, 3 symptoms of CF, 1 unknown
5 - 17 years	5	1 family history, 3 symptoms of CF, 1 unknown
18+ years	4	1 family history, 3 symptoms of CF
Total	38	

Deaths

The CFRI captures information on deaths in PWCF on the registry, but also those who are not. Since 2002, 218 deaths have occurred in PWCF. In the past three years (2010-2012), 60 out of 64 deaths in PWCF reported to the CFRI occurred in registered PWCF (93.8%).

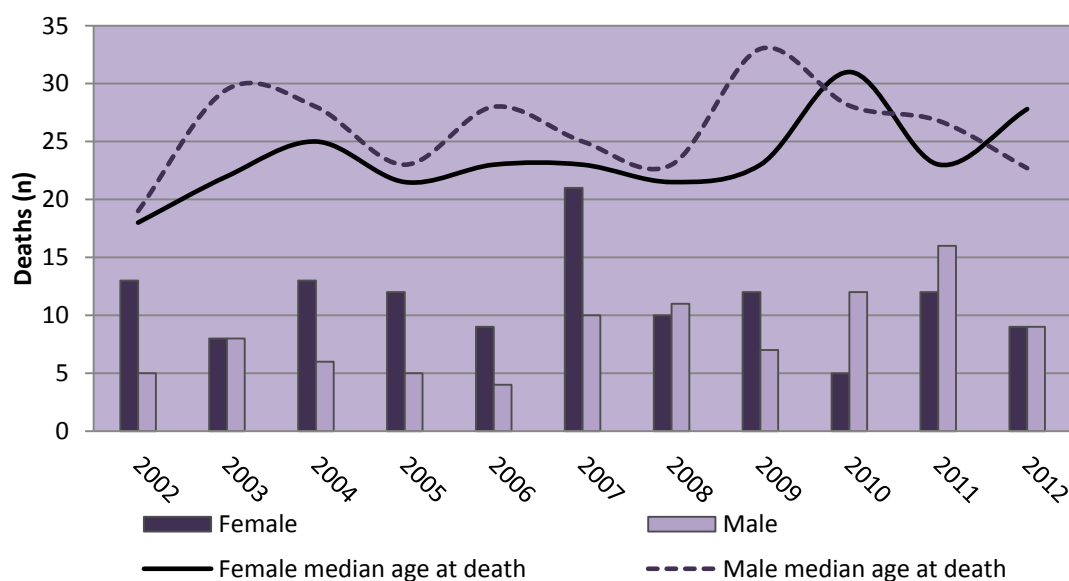
Eighteen deaths occurred in CFRI-registered PWCF in 2012 (9 male, 9 female). The primary cause of death for 11 of these PWCF was respiratory or cardiac failure. The median age at death for the 18 decedents in 2012 was 25.2 years (range: 5-42 years). The unusually young age (5 years) at which one PWCF died is notable; however the cause of death was not CF-related.

Figure 4: Deaths and median age



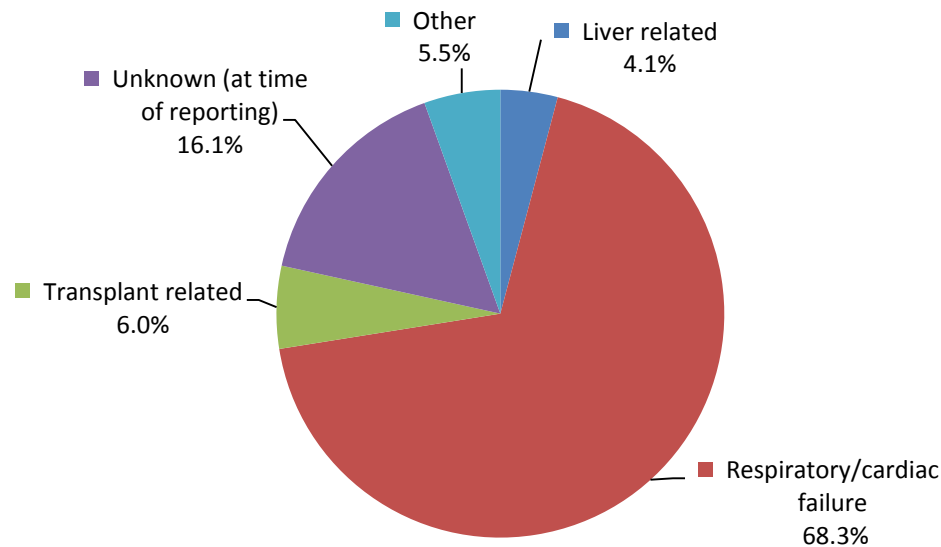
In CF, there is a recognised disparity in survival between males and females. Figure 5 shows the number and median age at death for males and females. Throughout the earlier part of the 2000's greater numbers of females were dying. Though proportionately more males than females died in 2010 and 2011, equal numbers of deaths were reported for males and females in 2012.

Figure 5: Deaths and median age at death by gender



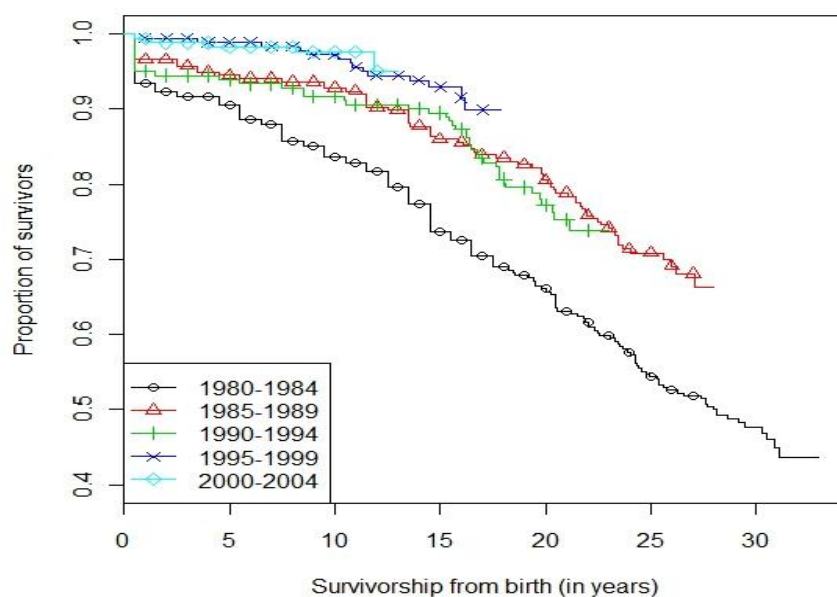
The cause of death for all known PWCF (registered and non-registered, n=218) who died between 2002 and 2012 is shown in Figure 6. Cause of death is unknown for 16.1% because information for PWCF not registered by the CFRI can often be incomplete. By excluding PWCF with an unknown cause of death, it can be determined that 81.4% of PWCF with a known cause of death died as a result of respiratory/cardiac failure.

Figure 6: Cause of death



Attrition (as a result of death) from birth cohorts (i.e. a grouping according to the year a PWCF was born) between 1980 and 2004 are shown by way of Kaplan-Meier survivorship curves in Figure 7. Over half of patients born between 1980 and 1984 are now deceased. Approximately two-thirds of PWCF born between 1985 and 1989 were alive at the end of 2012. Survivorship in successive birth cohorts continues to improve at all ages.

Figure 7: CF survival by birth cohort



Genotype

Genotyping is performed by the National Centre for Medical Genetics in Our Lady's Children's Hospital, Crumlin. Of the 1,140 PWCF on the registry in 2012, genotyping was reported to have been undertaken for 1,113, and 1,095 had a known genotype at the time of reporting (96.1%). Ninety percent of PWCF have ≥ 1 p.F508del mutation and 13.9% (n=159) have ≥ 1 p.G551D mutation.

Table 6: p.F508del mutations

Mutation	PWCF (%)
p.F508del	1028 (90.18%)
p.F508del homozygous	640 (56.14%)
p.F508del heterozygous	388 (34.04%)

Table 7: All CF mutations

Mutation	PWCF	%
p.F508del homozygous	640	56.1%
p.F508del heterozygous		
p.F508del, p.G551D	126	11.1%
p.F508del, p.R117H	43	3.8%
p.F508del, p.R560 T/K	31	2.7%
p.F508del, c.621+1 G-->T	16	1.4%
p.F508del, c.1717-1 G-->A	15	1.3%
p.F508del, p.G542X	15	1.3%
All other p.F508del heterozygote mutations	142	12.5%
Homozygous p.G551D	11	1.0%
All other mutations	56	4.9%
Mutation not recorded	27	2.4%
Mutation identification pending	18	1.6%
Total	1140	

Annual clinical data collection

Clinical data is extracted annually from medical records by the CFRI for PWCF who consent to share their medical information. Clinical information was gathered for 881 PWCF in 2012, representing 77.3% of the 1,140 PWCF on the CFRI in 2012 (Table 8). Clinical data is collected for on average 90% of PWCF <18 years, and 74% of PWCF ≥18 years.

Table 8: Annual clinical data collection

	2008	2009	2010	2011	2012
<18 years (% children)	438 (86.2%)	454 (91.5%)	454 (91.9%)	468 (91.1%)	476 (87.3%)
≥18 years (% adults)	375 (75.6%)	411 (77.4%)	412 (74.9%)	388 (69.4%)	405 (72.5%)
Overall (% all registered)	813 (81.0%)	865 (84.2%)	866 (83.0%)	856 (79.7%)	881 (77.3%)

CFRI staff aim to capture annual clinical data for each CFRI PWCF on an annual basis. For PWCF sharing care between two or more hospitals, information is sought from all hospitals.

Summary of clinical event data

For each PWCF, the number of hospitalisations since the last registry update is recorded. In 2012, PWCF were hospitalised once on average. Reasons for admission to hospital can vary.

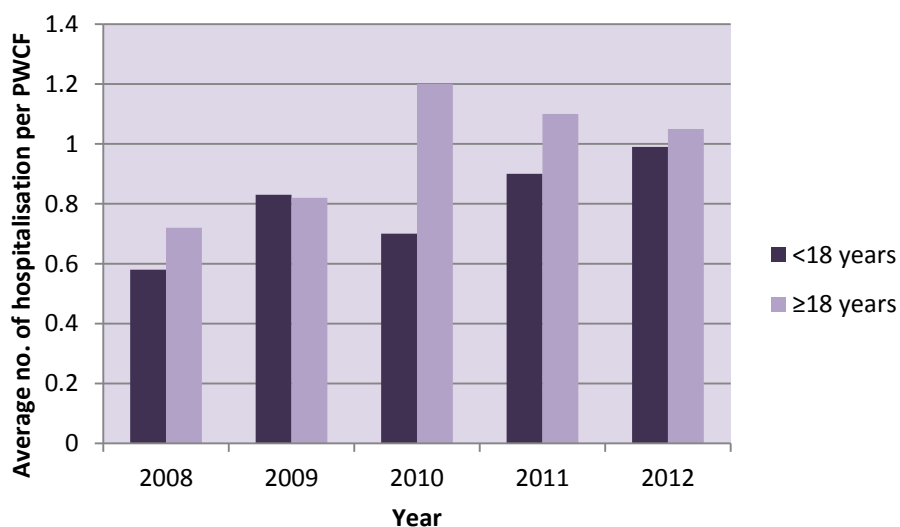
PWCF <18 years experienced one respiratory exacerbation requiring IV antibiotics on average in 2012, compared with 1.7 respiratory exacerbations for PWCF ≥18 years.

Table 9: Summary of clinical events

	<18 years		≥18 years	
	n	Average per PWCF	n	Average per PWCF
Hospitalisations	472	0.99	427	1.1
Respiratory exacerbations requiring IV antibiotics	451	0.95	680	1.7

Each year, greater numbers of hospitalisation, respiratory exacerbations requiring IV antibiotics and complications are recorded by the CFRI. The actual number of recorded hospitalisations increased from 256 and 271 for children and adults respectively in 2008, to 472 and 427 in 2012. Figure 8 shows that the average number of hospitalisations per child increased between 2008 and 2012, while average hospitalisations recorded for adults have declined slightly in the past two years.

Figure 8: Average number of hospitalisations per PWCF



Complications

The frequency of cardiorespiratory, gastrointestinal and other complications are recorded each year. A total of 1,444 complications were experienced by children (3.0 on average per child), and 1,661 (4.1 on average per adult) in 2012. For children, chronic *Staphylococcus aureus* infection (57.6%) and for adults, chronic *Pseudomonas aeruginosa* (65.4%), were the most commonly reported cardiorespiratory complications.

Chronic *P. aeruginosa*/*Staphylococcus aureus* infection is defined here as >2 isolates per year. Other complications such as Allergic Bronchopulmonary Aspergillosis (ABPA), liver disease etc. are physician-reported.

Table 10: Cardiorespiratory complications

	<18 years n (%)	≥18 years n (%)
Chronic <i>Pseudomonas aeruginosa</i> infection	116 (24.4%)	265 (65.4%)
Chronic <i>Staphylococcus aureus</i> infection	274 (57.6%)	145 (35.8%)
<i>Burkholderia cepacia</i> infection	6 (1.3%)	12 (3.0%)
MRSA	51 (10.7%)	36 (8.9%)
Mycobacteria species (nontuberculous)	10 (2.1%)	16 (4.0%)
Nasal polyps	12 (2.5%)	8 (2.0%)
Allergic Bronchopulmonary Aspergillosis	15 (3.2%)	19 (4.7%)
Asthma	5 (1.1%)	9 (2.2%)
All other cardiorespiratory complications*	2 (0.4%)	2 (0.5%)

*Cor pulmonale, pneumothorax, haemoptysis

Pancreatic insufficiency (defined as that which requires pancreatic enzyme use) affects nearly all children (95.7%) and adults (91.9%). Half of all adults (51.5%) and a third of children (31.1%) had gastro-oesophageal reflux disease. Liver disease was reported for 103 PWCF. Slightly more adults were found to have liver disease in 2012 (n=72) than in 2011 (n=59).

Table 11: Gastrointestinal complications

	<18 years n (%)	≥18 years n (%)
Distal intestinal obstructive syndrome (physician-defined)	4 (0.8%)	16 (4.0%)
Pancreatic Insufficiency	456 (95.8%)	372 (91.9%)
Abnormal liver function tests	4 (0.8%)	4 (1.0%)
Liver disease	31 (6.5%)	72 (17.8%)
Gastro-oesophageal reflux disease	148 (31.1%)	207 (51.1%)
All other gastrointestinal complications**	4 (0.8%)	3 (0.7%)

**Haematemesis, colonic stricture, gallbladder disease

Osteopenia/osteoporosis was documented for 38.5% of adults in 2012, fewer than that reported in 2011 (43.8%) and 2010 (49.8%).

Table 12: Other complications

Other	<18 years n (%)	≥18 years n (%)
Diabetes requiring insulin	10 (2.1%)	104 (25.7%)
Clubbing	203 (42.6%)	148 (36.5%)
Osteopenia/osteoporosis	14 (2.9%)	156 (38.5%)
Other non-CF morbidities	89 (18.7%)	83 (20.5%)

Pulmonary function

An important indicator of lung function is FEV₁, which is the maximum amount of air that can be exhaled in one second. The FEV₁ value is typically presented as a percentage of the predicted/reference value for a healthy peer (FEV₁% predicted). Lung function is presented here for children from age six years and for adults.

In 2012, the results of 1,611 lung function tests were captured for PWCF aged 6 and older (Table 13). The median FEV₁% predicted value for PWCF aged 6-17 years was 81% (interquartile range: 65-96) and 58% (interquartile range: 39-80) for PWCF aged ≥18 years. FEV₁% predicted is expected to decline over time, as the condition of the lungs worsens with age.

Table 13: Median FEV₁% predicted by age

Age (years)	PFTs (n)	Median FEV ₁ % predicted	Mean FVC % predicted
6-8	159	89%	93%
9-11	159	82%	90%
12-14	181	80%	87%
15-17	158	75%	91%
18-19	100	73.5%	86.5%
20-24	296	54%	76%
25-29	284	57%	76%
30-34	134	55%	79.5%
35-39	70	51.5%	72%
≥40	70	69%	92.5%
Total	1611	69%	85%

Figure 9 shows the median FEV₁% predicted for males and females by age grouping.

Figure 9: Median FEV₁% predicted by age and gender

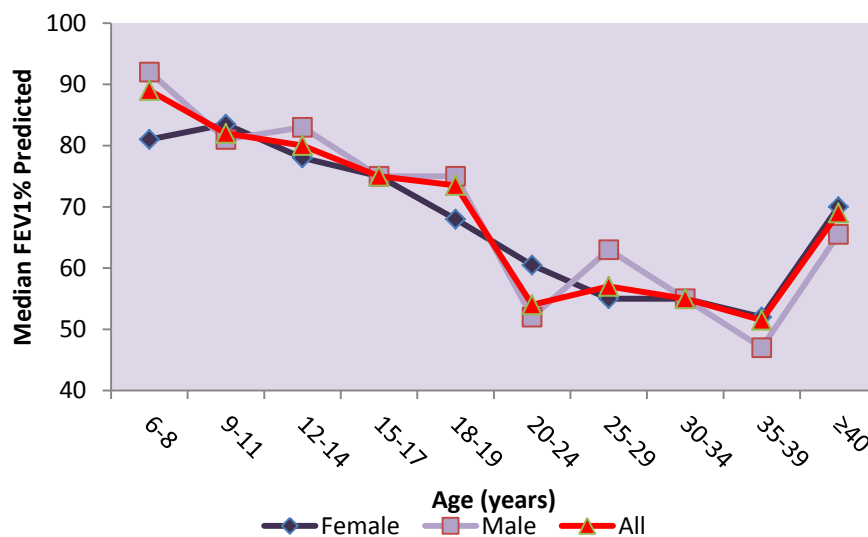
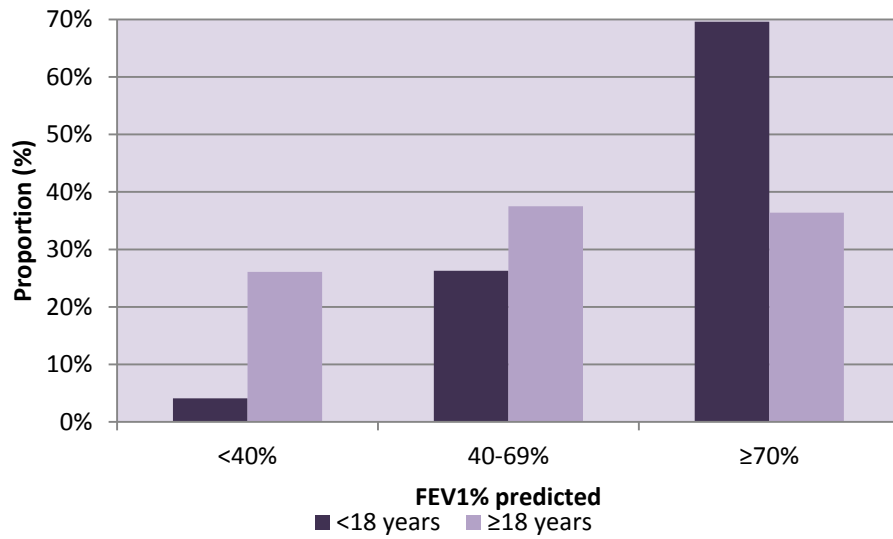


Figure 10 shows that in 2012, 69.6% of PWCF <18 years had a FEV1% predicted of 70% or more compared to 36.4% of PWCF ≥18 years.

Figure 10: Proportion of FEV1% predicted by age category

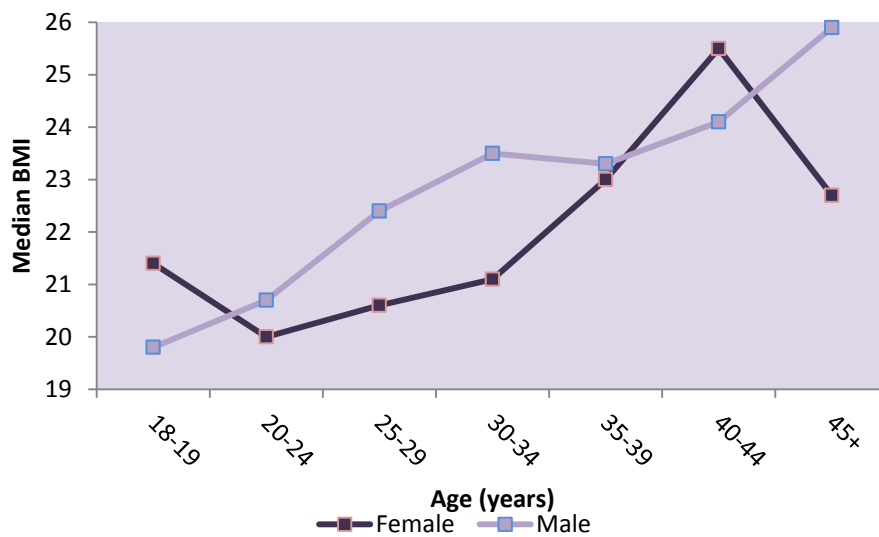


Body Mass Index

Body mass index (BMI) is a measure often used to assess how much an individual's body weight departs from what is considered normal for a person of a given height. It is calculated by taking a person's body weight (in kilograms) and dividing it by the square of their height (in metres). BMIs can then be categorised into underweight, normal, overweight and obese subclasses. Nearly 1,000 BMI values ($n=989$) were recorded for PWCF ≥ 18 years in 2012, which equates to an average of 2.4 recorded BMI measurements per adult ($n=405$).

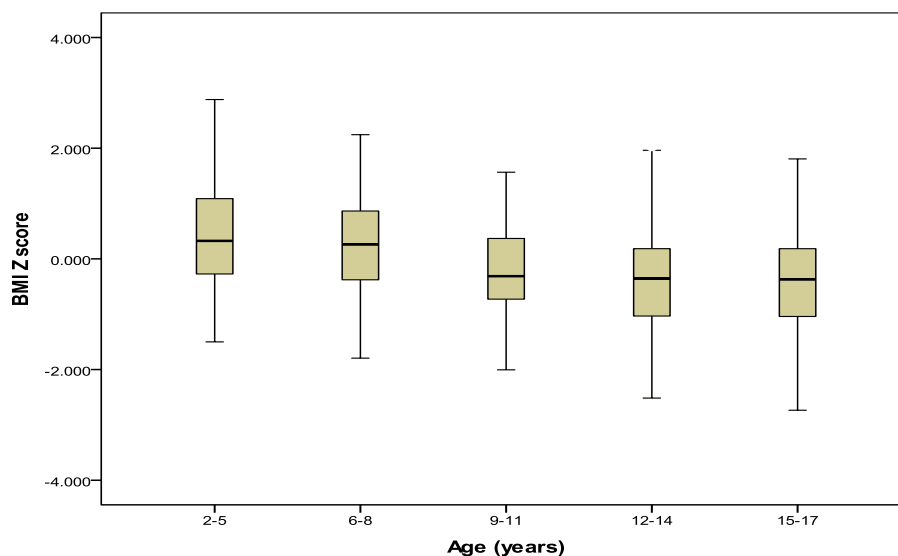
Male and female adults had median BMI values of 21.8 and 20.9 respectively. Median BMI values among adults are shown in figure 11.

Figure 11: Median BMI values in PWCF ≥ 18 years



Over 1,000 BMI values were recorded for PWCF aged 2-17 years, giving an average number of BMI measurements of 2.1 per PWCF. BMI Z scores among children are shown in Figure 12.

Figure 12: BMI z-scores in PWCF aged 2-17 years



Cultures

In 2012, the results of 7,226 culture swabs were captured by the CFRI. For PWCF ≥ 18 years, most cultures were grown from sputum (97.3%), with an average of 7 sputum specimen cultures being reported per PWCF. Children's samples came from sputum (53.0%), throat (22.4%) and cough swabs (20.2%).

Table 14: Culture type by age

	<18 years		≥ 18 years	
	n	Average per PWCF	n	Average per PWCF
Sputum samples	2281	4.8	2846	7.0
Throat swabs	962	2.0	10	<0.1
Cough swabs	868	1.8	47	0.1
BAL sample	59	0.1	9	<0.1
Nasal swabs	48	0.1	3	<0.1

The results of 5,127 sputum cultures were recorded by the CFRI in 2012. *Pseudomonas aeruginosa* was detected in 27.8% of sputum cultures. *Candida* and *Staphylococcus aureus* were the second and third most commonly detected micro-organisms in sputum (17.5% and 17.1% respectively).

Table 15: Micro-organisms detected in sputum culture

	n	%
Pseudomonas aeruginosa	1423	27.8%
P. aeruginosa (mucoid status not reported)	553	10.8%
P. aeruginosa (mucoid)	529	10.3%
P. aeruginosa (non-mucoid)	341	6.7%
All Candida species	897	17.5%
Staphylococcus aureus	879	17.1%
Normal flora	530	10.3%
Aspergillus fumigatus	304	5.9%
MRSA	176	3.4%
Haemophilus influenza	155	3.0%
Stenotrophomonas maltophilia	129	2.5%
Gram positive cocci	87	1.7%
Gram negative bacilli	61	1.2%
Haemophilus parainfluenza	31	0.6%
Burkholderia cepacia complex*	38	0.7%
Other	417	8.1%
Total	5,127	

*Contains 2 *Burkholderia cepacia* complex genomovar I, 14 genomovar II, 8 genomovar III and 11 genomovar unspecified.

Antibiotics

The CFRI recorded information on 3,196 courses of antibiotics with a 2012 start date. Just over half (54.8%) of antibiotics were given intravenously (IV - either at home or in hospital), 40.3% were given in an oral form, and 4.8% were inhaled. The three most frequently recorded courses of antibiotic were tobramycin (14.2%) ciprofloxacin (13.5%) and ceftazidime (11.2%).

There were 1,738 courses of IV antibiotics commenced in 2012, 55.1% of which were administered in hospital. The cumulative number of days spent on IV antibiotics totalled 21,579 days (9,751 home IV and 11,828 hospital IV administration). The median number of days on a course of IV antibiotics was 14 days (median of 14 days for home and 13 days for hospital IV). Figure 13 shows that compared with <18s, PWCF ≥ 18 years had twice as many hospital IV days and three times as many home IV days in 2012.

Figure 13: Home and hospital IV antibiotics

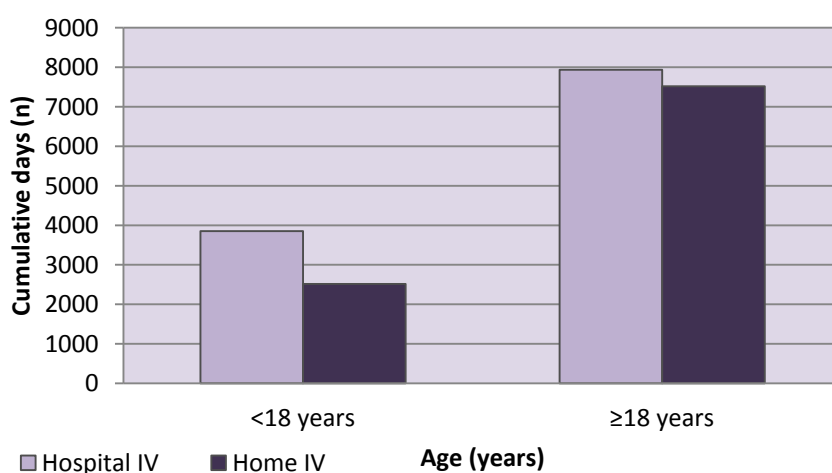
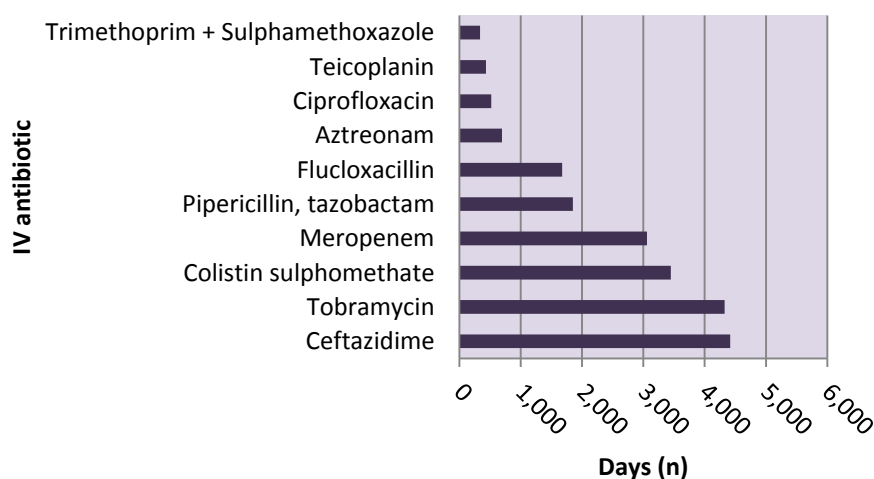


Figure 14 shows the cumulative number of days PWCF spent taking IV antibiotics. As in other years, ceftazidime, tobramycin and colistin sulphomethate have the highest cumulative number of antibiotic days.

Figure 14: Cumulative number of days on IV antibiotics



Long-term medications

Long-term medications are defined as medications taken regularly for a minimum of 3 months. In 2012, two out of every three PWCF were prescribed beta-agonists to open the airways and improve lung function (Table 16). Over half of children (55.9%) were treated with nebulised hypertonic saline for the purposes of improving mucociliary clearance in the lungs. Use of rhDNase (to improve clearance of lung secretions) was reported for 46% of PWCF.

Table 16: Respiratory medications

	<18 years n=476 (%)	≥18 years n=405 (%)
Inhaled		
Beta-agonist	322 (67.6%)	261 (64.4%)
Steroid	148 (31.1%)	180 (44.4%)
Anti-cholinergic	3 (0.6%)	29 (7.2%)
Nebulised		
Recombinant Human DNase (rhDNase)	220 (46.2%)	185 (45.7%)
Hypertonic saline*	266 (55.9%)	137 (33.8%)
Oxygen		
Continuous	2 (0.4%)	11 (2.7%)
Night-time	11 (2.3%)	9 (2.2%)
Oral steroid	28 (6.9%)	33 (8.1%)

*Continuous and intermittent use

Over half of adults (51.9%) and 30.5% of children used H2RA or PPIs to treat gastro-oesophageal reflux disease (Table 17).

Table 17: Gastro-intestinal medications

	<18 years n=476 (%)	≥18 years n=405 (%)
Urso-deoxycholic acid	73 (15.3%)	70 (17.3%)
H2RA/PPI	145 (30.5%)	210 (51.9%)
Lactulose/movicol	70 (14.7%)	48 (11.9%)

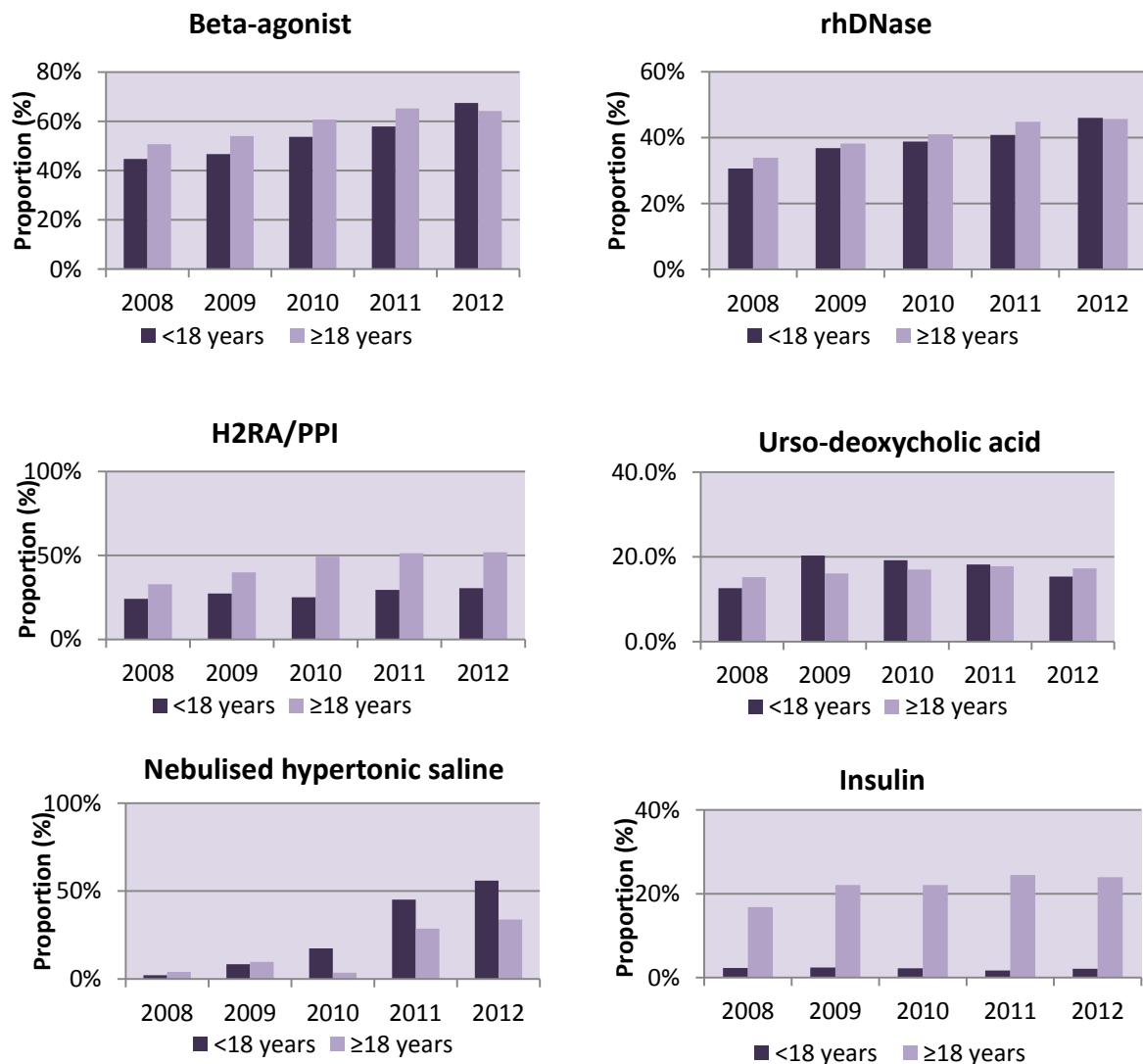
Nearly one in four adults required insulin for CF-related diabetes, while use of osteoporosis treatment was reported for 43.2% of adults (Table 18).

Table 18: Other long-term medications

	<18 years n=476 (%)	≥18 years n=405 (%)
Osteoporosis treatment(s)	73 (15.3%)	175 (43.2%)
Insulin	10 (2.1%)	99 (24.4%)

Figure 15 shows trends in usage of long-term medications over a five-year period (2008-2012). Beta-agonists and rhDNase use has increased. The use of nebulised hypertonic saline in PWCF ≥18 years increased from 4% in 2008 to 55.9% in 2012. The proportion of <18 years olds using urso-deoxycholic acid has increased slightly, whereas use by adults decreased from 2009 onwards. Use of H2RA/PPI by adults increased over the five-year period, and to a lesser extent in children. Insulin use by ≥18s has increased from 16.8% in 2008 to 24.0% in 2012.

Figure 15: Trends in long-term medications



Nutrition

Nearly all PWCF were reviewed by a dietician in 2012. Most supplemented their diet with vitamins. One in ten PWCF <18, and one in 4 PWCF ≥18 years took calorie supplements.

Table 19: Nutrition

	<18 years n=476 (%)	≥18 years n=405 (%)
Reviewed by dietician	469 (98.5%)	391 (96.5%)
Calorie supplements	46 (9.7%)	105 (25.9%)
Vitamins	460 (96.6%)	376 (92.8%)
Minerals	25 (5.3%)	21 (5.2%)
Gastrostomy feeds	36 (7.6%)	40 (9.9%)

Nearly all PWCF in 2012 reported using pancreatic enzymes. Supplemental feeding was also reported for a third of PWCF. The proportion of PWCF using oral supplements remains on a par with that reported in previous years.

Table 20: Supplemental feeding

	<18 years n=476 (%)	≥18 years n=405 (%)
Oral supplements	107 (22.5%)	105 (25.9%)
Nasogastric tube insertion	1 (0.2%)	1 (0.2%)
Gastrostomy tube insertion	45 (9.5%)	43 (10.6%)

Airway clearance

Each year, greater proportions of children and adults are reviewed by a physiotherapist. Over 85% of PWCF ≥ 18 years and 93.5% of PWCF < 18 years attended for physiotherapy services in 2012. The Table below (Table 21) summarises the airway clearance techniques adopted by PWCF. These techniques are not mutually exclusive, as PWCF may adopt multiple techniques at any given time.

The airway clearance techniques most commonly cited as being performed by children include acapella and use of the PEP mask. For adults, autogenic drainage (55.5%) and the PEP mask (21.5%) were most frequently used.

While nearly all PWCF indicated participating in exercise activities, no modalities of physiotherapy were recorded for 15.5% of children and 25.9% of adults in 2012.

Table 21: Airway clearance techniques in children and adults

Children	<18 years n=476 (%)	Adults	≥ 18 years n=405 (%)
Reviewed by physiotherapist	445 (93.5%)	Reviewed by physiotherapist	345 (85.2%)
Acapella	158 (33.2%)	Autogenic drainage	223 (55.1%)
PEP Mask	142 (29.8%)	PEP Mask	87 (21.5%)
Other*	131 (27.5%)	Acapella	46 (11.4%)
Percussion	50 (10.5%)	Other*	23 (5.7%)
Autogenic drainage	43 (9.0%)	Flutter	13 (3.2%)
Active cycle breathing	18 (3.8%)	Active cycle breathing	10 (2.5%)
Flutter	13 (2.7%)	Percussion	2 (0.5%)
Vest	13 (2.7%)	Vest	1 (0.2%)
Postural drainage	4 (0.8%)		

*Other: Therapep, trampolining, age appropriate activities

Financial information

The financial summary below lists the Income and Expenses for the CFRI in 2012.

Table 22: Financial summary

Income & Expenses		2012 €
<u>Income</u>		
Grant income		140,000
Sundry income		8,488
	Total income	148,488
<u>Expenses</u>		
Wages & salary		107,485
Employer's PRSI		8,101
Rent payable		6,006
Service Charges		662
Insurance		458
Computer network & server costs		4,190
Database costs		252
Printing, postage and stationery		245
Travelling & subsistence		4,193
Legal & professional fees		4,000
Audit		984
Bank charges		230
Subscriptions		120
Depreciation on equipment		636
	Total expenses	137,562
	(Deficit)/Surplus	10,926

The full audited accounts were prepared Hayden Brown, Chartered Accountants, Grafton Buildings, 34 Grafton Street, Dublin 2 and copies are available upon written request to CFRI.

Acknowledgements

There are many individuals and groups that have contributed to and supported the work of the CFRI in this reporting year.

First we would like to thank the HSE for providing financial support to the CFRI since its inception in 2002. In addition we would like to thank our industry partners who have provided unconditional grants to the registry. Without this support we would not be able to maintain operations.

Each PWCF and/or their guardian who kindly agreed to share medical information with this registry. By consenting in such large numbers, the information reported by the registry each year has become an important research tool.

The management committee of the CFRI have provided great support during a period of growth and development within the registry.

Cystic Fibrosis Ireland was integral in the initiation of this registry and continues to support the work that is undertaken by the registry.

Each CF centre and clinic provides immense assistance to CFRI staff in the collection of this important information. In particular, we thank the CF teams in each of the hospitals for their continuing co-operation.

The CFRI researchers who have worked tirelessly collecting the required data from hospital based paper records under challenging conditions.

The UCD School of Public Health, Physiotherapy and Population Science particularly Prof C Kelleher and her staff who have made an invaluable contribution to the CFRI research programme.

We thank the HSE's Health Intelligence Unit particularly Dr D Beaton, Dr H Johnson, Dr F Donohue and Ms. Imelda Crone for their support and encouragement during the year.

We would also like to thank Prof Philip Mayne, Children's University Hospital confirming the numbers of infants detected in 2012 with two CFTR mutations.

Finally, we thank Dr Abaigeal Jackson and Larry Ungar for their hard work and dedication in preparing this report.

CFRI executive council membership 2012

Dr Ed McKone (Chairperson)	Consultant in Respiratory Medicine St. Vincent's University Hospital, Dublin
Dr Cedric Gunaratnam (Vice Chairperson)	Consultant in Respiratory Medicine Beaumont Hospital, Dublin
Prof Charles Gallagher (Immediate Past Chairperson)	Consultant in Respiratory Medicine St. Vincent's University Hospital, Dublin
Dr Marion Rowland (Secretary)	Lecturer UCD School of Medicine & Medical Sciences
Mr John Coleman (Treasurer)	Chairperson Cystic Fibrosis Ireland
Mr Godfrey Fletcher (CEO)	Chief Executive Cystic Fibrosis Registry Ireland
Dr Peter Greally	Consultant in Paediatric Respiratory Medicine The Adelaide and Meath Hospital Dublin, Incorporating the National Children's Hospital
Prof Gerry McElvaney	Professor of Medicine, Royal College of Surgeons in Ireland & Consultant in Respiratory Medicine Beaumont Hospital, Dublin
Dr Paul McNally	Consultant in Paediatric Respiratory Medicine Our Lady's Children's Hospital, Crumlin
Dr Barry Linnane	Consultant in Paediatric Respiratory Medicine Midwestern Regional Hospital, Limerick
Dr Barry Plant	Consultant in Respiratory Medicine Cork University Hospital, Cork
Dr Dubhfeasa Slattery	Consultant in Paediatric Respiratory Medicine Temple Street Children's University Hospital, Dublin
Mr Philip Watt	Chief Executive Cystic Fibrosis Ireland
Mr Martin Wickham	Patient Representative

Mission Statement of the CFRI

“The national Cystic Fibrosis Registry of Ireland will endeavour to collect and analyse information relating to cystic fibrosis in order to improve the quality of care for all of the people with cystic fibrosis in the Republic of Ireland.”



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