

The Cystic Fibrosis Registry of Ireland

Annual Report 2010



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Executive Summary

The Cystic Fibrosis Registry of Ireland reached a major milestone in 2010. Our annual census of centres providing care to people with cystic fibrosis (PWCF) confirms that our data now represents 90% of the known CF population in the Republic of Ireland. This has been achieved by the willingness of PWCF and their guardians to participate in the registry and an appreciation of the importance of registry data. High patient enrolment is also due with thanks to the co-operation of hospital staff (medical and administrative). Access to patient charts continues to be a major challenge. It would not be possible to capture the required data without the huge dedication of our researchers who have to comb through mountains of paper records.

Our next milestone must be to make the registry data more easily available to practitioners and researchers so that our vast data resources can be used to assist with the day to day patient management and for national quality improvement programmes. This cannot be achieved on our existing operational budget. In times of cutbacks it is more likely that we will have to appeal to the generosity of the private sector to provide urgently required technological resources.

It is very comforting to note that the Irish CF survival statistics continue to improve. We continue to report median age of death even though it is a rudimentary measure of survival in that it only describes the duration of life in those who have died. In comparison to 2009 the median age at death improved by 3 ½ years to 28 ½ years. Nearly 57% of the total population are now over 18 years of age compared to 51.7% in 2009.

Ireland's percentage of families with 2 or more members with CF remains the same at 13.5%. Our median age at diagnosis also remained the same at 4.1 months. This should start to improve with the introduction of neonatal screening for CF in 2011.

Over the last number of years there has been considerable investment in the development of Cystic Fibrosis Services. In parallel with this new treatments continue to be developed which should continue to contribute to improved patient outcomes. The Registry intends to continue to play an active role in monitoring CF patient outcomes, contributing to research, and the development of a national CF quality improvement programme.



Godfrey Fletcher

Interim CEO
The Cystic Fibrosis Registry of Ireland

Summary Statistics for 2010

	2009	2010
Live CFRI enrolees	1,027	1,044
Median age (years)	18	19
Number of patients with genotype information	96.9%	97.3%
No. of deceased CFRI enrolees	17	16
Median age at death (years)	25.0	28.1
% Males*	57.6%	57.1%
% Adults (≥18 years)*	51.7%	52.7%
Coverage of the Republic of Ireland population	89.2%	90.0%
No of enrolees for whom annual assessment data was collected	865	866

*Of those alive at the end of 2010 (n=1044)

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CFRI Data Collection Cystic Fibrosis Centres, 2010

County	Hospital	Consultant	Type of Centre
Cork	Cork University Hospital	Dr Barry Plant /	Adult
		Dr Michael Henry	
		Dr Muireann ní Chroínín/	Paediatric
		Dr David Mullane	
Dublin	Beaumont Hospital	Prof NG McElvaney /	Adult
		Dr Cedric Gunaratnam	
	St Vincent's University Hospital	Prof Charles Gallagher/	Adult
		Dr Ed McKone	
	The Children's University Hospital	Dr Dubhfeasa Slattery	Paediatric
	The Adelaide and Meath Hospital Dublin, Incorporating the National Children's Hospital	Dr Peter Greally /	Paediatric
		Dr Basil Elnazir	
	Our Lady's Children's Hospital	Dr Gerry Canny /	Paediatric
		Dr Paul McNally*	
	Mater Misericordiae University Hospital	Prof Jim Egan	Heart/lung transplant
Galway	University College Hospital Galway	Dr Mary Herzig	Paediatric
	Merlin Park Hospital, Galway	Dr JJ Gilmartin	Adult
Kerry	Kerry General Hospital	Dr Fergus Leahy	Paediatric
Limerick	Midwestern Regional Hospital	Dr Michael J Mahony /	Paediatric
		Dr Barry Linnane*	
		Dr Brian Casserly	Adult
Louth	Our Lady of Lourdes Hospital	Dr Amjad Altaf	Paediatric
Mayo	Mayo General Hospital	Dr Michael O'Neill	Paediatric
Sligo	Sligo General Hospital	Dr Rohininath Tummaluru	Paediatric
Waterford	Waterford Regional Hospital	Dr Animitra Das	Paediatric
		Dr Mark Rogan	Adult

*Dr Linnane moved from Our Lady's Hospital for Sick Children to the MidWestern Regional in mid-2010 and was replaced by Dr P McNally.

CFRI Executive Council 2010

Prof C Gallagher	Consultant in Respiratory Medicine, St. Vincent's University Hospital, Dublin	Chairperson
Dr G Canny	Consultant in Paediatrics, Our Lady's Children's Hospital, Dublin	Vice-Chairperson
Dr P Greally	Consultant in Paediatric Respiratory Medicine, Adelaide and Meath National Children's Hospital, Dublin	Honorary Secretary
Mr G Fletcher	Chief Executive (interim), CFRI	Council Member (non voting)
Prof NG McElvaney	Professor of Medicine, Royal College of Surgeons in Ireland & Consultant in Respiratory Medicine Beaumont Hospital, Dublin	Immediate Past Chairman, ex-officio
Dr B Linnane	Consultant in Paediatrics, Our Lady's Children's Hospital, Dublin	Council Member
Dr E McKone	Consultant in Respiratory Medicine, St. Vincent's University Hospital, Dublin	Council Member
Dr B Plant	Consultant in Respiratory Medicine, Cork University Hospital, Cork	Council Member
Dr M Rowland	UCD School of Medicine, Medical Sciences, Children's Research Centre, Crumlin, Dublin	Council Member
Dr D Slattery	Consultant in Paediatric Respiratory Medicine, Children's University Hospital, Dublin	Council Member
Mr. M Wickham	Non medic council member	Council Member

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
CFAI	Cystic Fibrosis Association of Ireland
CFRI	Cystic Fibrosis Registry of Ireland
CRA	Clinical Research Associate
CSO	Central Statistics Office of Ireland
DEXA	Dual Energy X-ray Absorptiometry
DIOS	Distal Intestinal Obstruction Syndrome
FH	Family history
FEV ₁	Forced Expiratory Volume in one second
FVC	Forced expiratory Vital Capacity
GI	Gastrointestinal symptoms
HRB	Health Research Board
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

Hospital Abbreviations

AMNCH	The Adelaide and Meath Hospital Dublin, Incorporating the National Children's Hospital, Tallaght, Dublin 24
BMT	Beaumont Hospital, Dublin 9
Cavan GH	Cavan General Hospital, Cavan
CUH	Cork University Hospital, Wilton, Cork
CUHTS	Children's University Hospital, Temple Street, Dublin 1
Mayo GH	Mayo General Hospital, Castlebar
MWRH	Midwest Regional Hospital, Limerick
MMUH	Mater Misericordiae University Hospital, Dublin 7
OLCH	Our Lady's Children's Hospital, Crumlin, Dublin 12
OLLH	Our Lady of Lourdes Hospital, Drogheda
Sligo GH	Sligo General Hospital, Sligo
SVUH	St Vincent's University Hospital, Dublin 4
UCHG	University College Hospital Galway (including Merlin Park Hospital)
WRH	Waterford Regional Hospital, Waterford

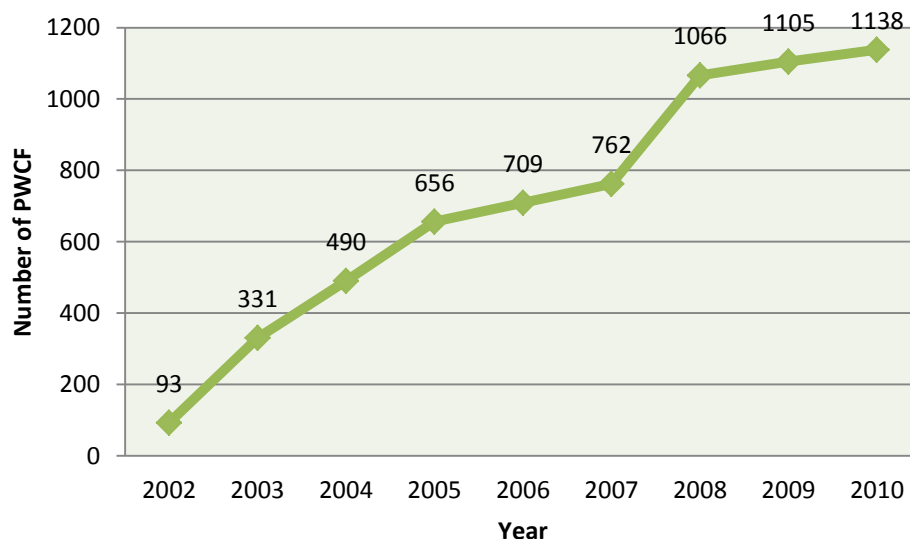
CFRI Enrolment

All persons with CF (PWCF) receiving medical care at CF centres or clinics in the Republic of Ireland are eligible to have their information included on the Cystic Fibrosis Registry of Ireland (CFRI). The collection of each PWCF's medical information by the CFRI has been approved by all CF centre Research Ethics Committees, with the proviso that each PWCF must provide the CFRI with written confirmation of their consent to participate.

Annual growth in enrolment figures has slowed in recent years, as is expected as enrolment reaches saturation point. Thirty-three PWCF enrolled in 2010, a figure similar to the previous year (40 in 2009). The 33 enrolees comprise mostly of PWCF born between 2008 and 2010. The total number of PWCF enrolled by the end of 2010 was 1,138 (Figure 1).

On the last day of 2010, 1,044 of the 1,138 enrolees were alive (91.7%). Of those alive, 57.1% were male, and 52.7% were aged 18 years and older. There were 16 deaths in CFRI enrolees in 2010, a figure in keeping with other years.

Figure 1: CFRI enrolment 2002 – 2010

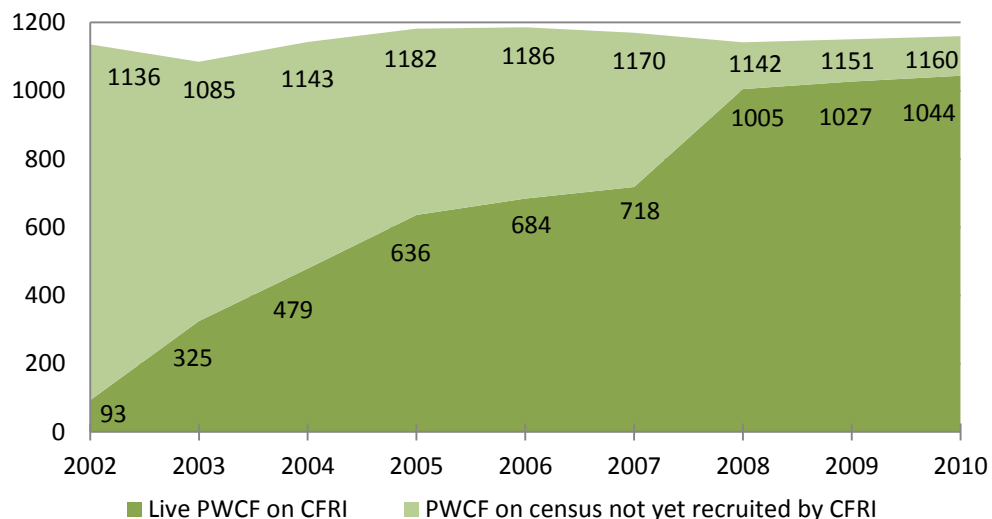


CF Centre & Clinic Census 2010

A census recording the numbers of live PWCF registered at CF centres and clinics in the Republic of Ireland was performed by the CFRI in early 2010. A total of 1,160 PWCF were identified. One-thousand and forty-four of the 1,160 census-identified PWCF had enrolled with the CFRI by the end of that year (Figure 2). This suggests that 90% of the known CF population in the Republic of Ireland has enrolled on the CFRI. This level of coverage of the CF population is similar to that of the larger CF registries.

This is the third year in which a more accurate census of the CF population was undertaken. Since 2008, the identities of PWCF have been recorded as part of the annual census. This ensures double-counting of patients attending more than one CF centre does not occur, thereby enhancing the accuracy of the census. The use of this improved census methodology gives confidence that the observed increase in the size of the CF population since 2008 (in the order of approximately 10 PWCF per annum) is unlikely to be artefactual.

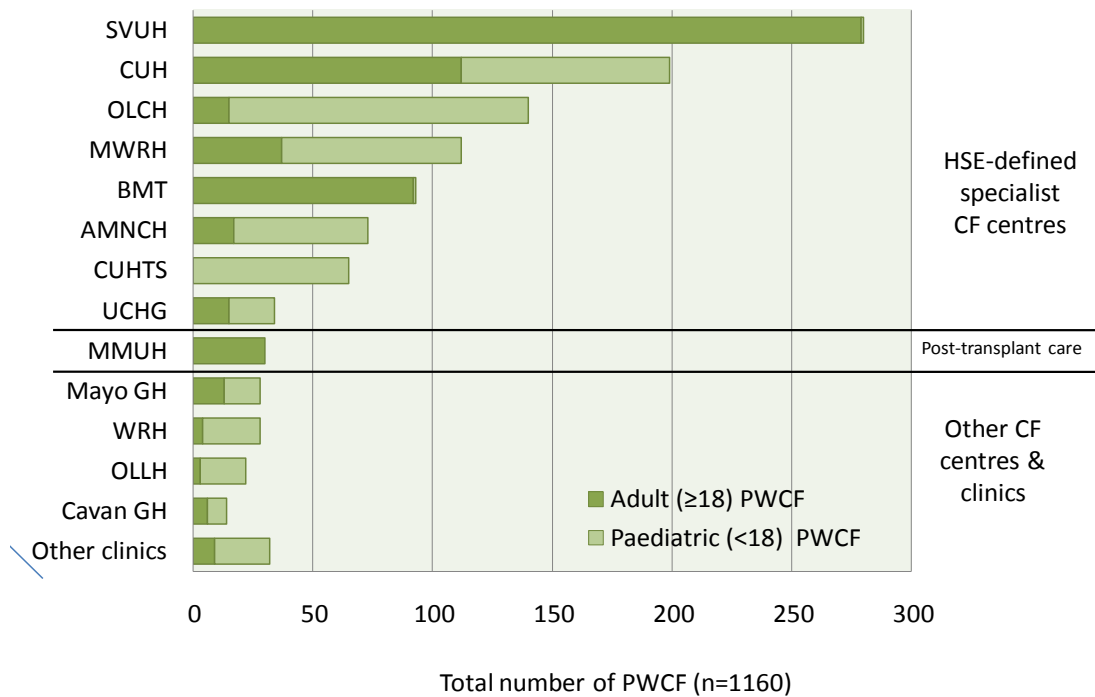
Figure 2: Live CFRI enrollee numbers and CF centre census, 2010



Census estimates of PWCF attending CF specialist centres and clinics are shown in Figure 3. As patterns of shared CF care are complex, these are not easily summarily described. For the purposes of presentation, PWCF attending two or more hospitals for CF care are attributed to just one of those sites, to ensure the individual is counted just once. Such PWCF are attributed to the CF specialist centre, as the team at the specialist centre are deemed responsible for the care of a PWCF receiving shared care (according to the 2009 HSE Report 'Services for people with cystic fibrosis in Ireland'). As such, the number of PWCF attending non-specialist CF clinics may be an underestimate.

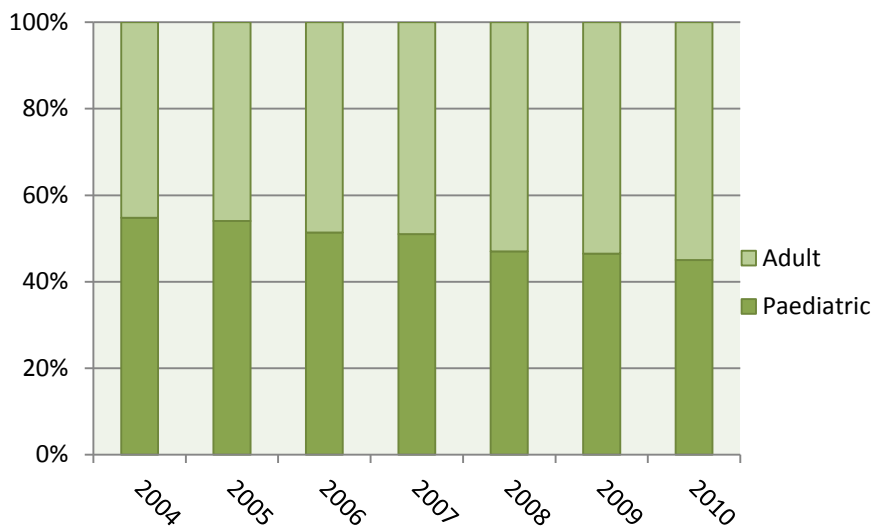
The numbers attending HSE-defined centres are as follows. For adult CF services, St Vincents University Hospital had the largest number of adults attending for CF services (n= 279). This was followed by Cork University Hospital (n=112) and Beaumont Hospital (n=92), the MidWestern Regional Hospital (n=37) and University College Hospital, Galway (n=15). For paediatric services, the largest number of paediatric PWCF attended Our Lady's Children's Hospital (n=125), followed by Cork University Hospital (n=87), the Mid-Western Regional Hospital (n=75), the Children's University Hospital, Temple Street (n=65) and the Meath and Adelaide Hospital Dublin, Incorporating The Children's University Hospital (n=56) and University College Hospital, Galway (n=19). The numbers of PWCF receiving care at nearly all of these centres increased slightly in the last year.

Figure 3: CFRI census of PWCF by CF specialist centre/clinic, 2010



The proportion of the CF population aged 18 years and older continues to grow. In 2010, adults represented 54.5% of census-identified individuals (cf. 53.0% in 2008 and 53.2% in 2009). The average annual increase in census-reported adult PWCF since 2002 is approximately 2% per annum. It is difficult to predict whether this rate of growth will continue in the coming years.

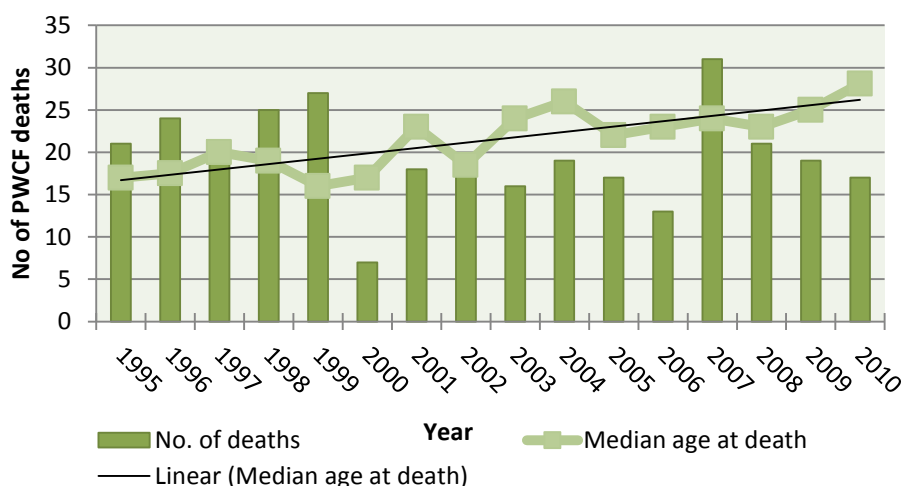
Figure 4: Ratio of adult to paediatric census-identified PWCF, 2004-2010



CF Survival

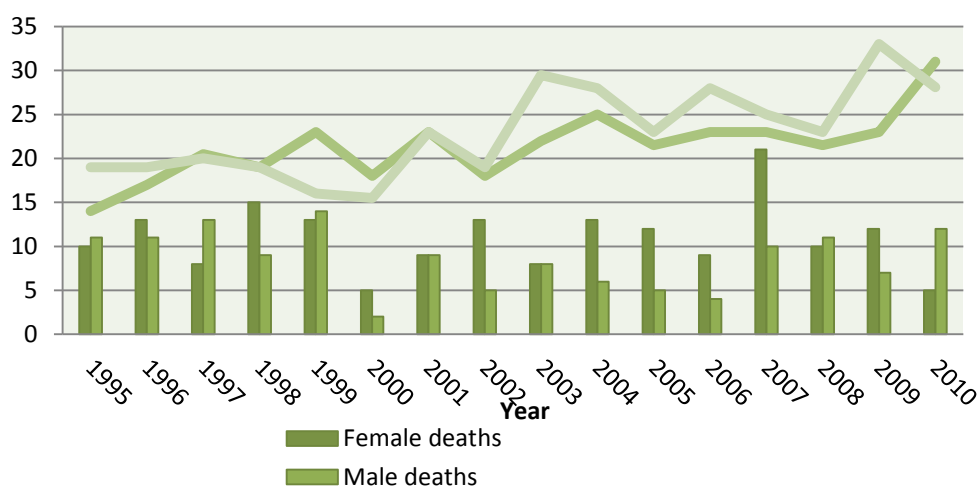
Less than 2% of the CF population die each year. The absolute annual number of deaths remains small overall, and fluctuations can occur from year to year. In 2010, the CFRI learned of a total of 17 deaths (Figure 5); 16 deaths were identified during the course of data collection by the CFRI, and one CF death previously unknown to the CFRI was reported by the CFAI. This is the third consecutive decrease since 2007, when the number of deaths peaked at 33. It is unlikely however, that this downward trajectory will continue indefinitely.

Figure 5: Total number of deaths and median age at death of PWCF, 1995-2010



There were no deaths in PWCF under 16 years of age in the Republic of Ireland in 2010. Age at death ranged from 16 to 51 years. Although there is an observable trend whereby more deaths occur in females each year, more male deaths (n=12) were reported than females (n=5) in 2010 (Figure 6).

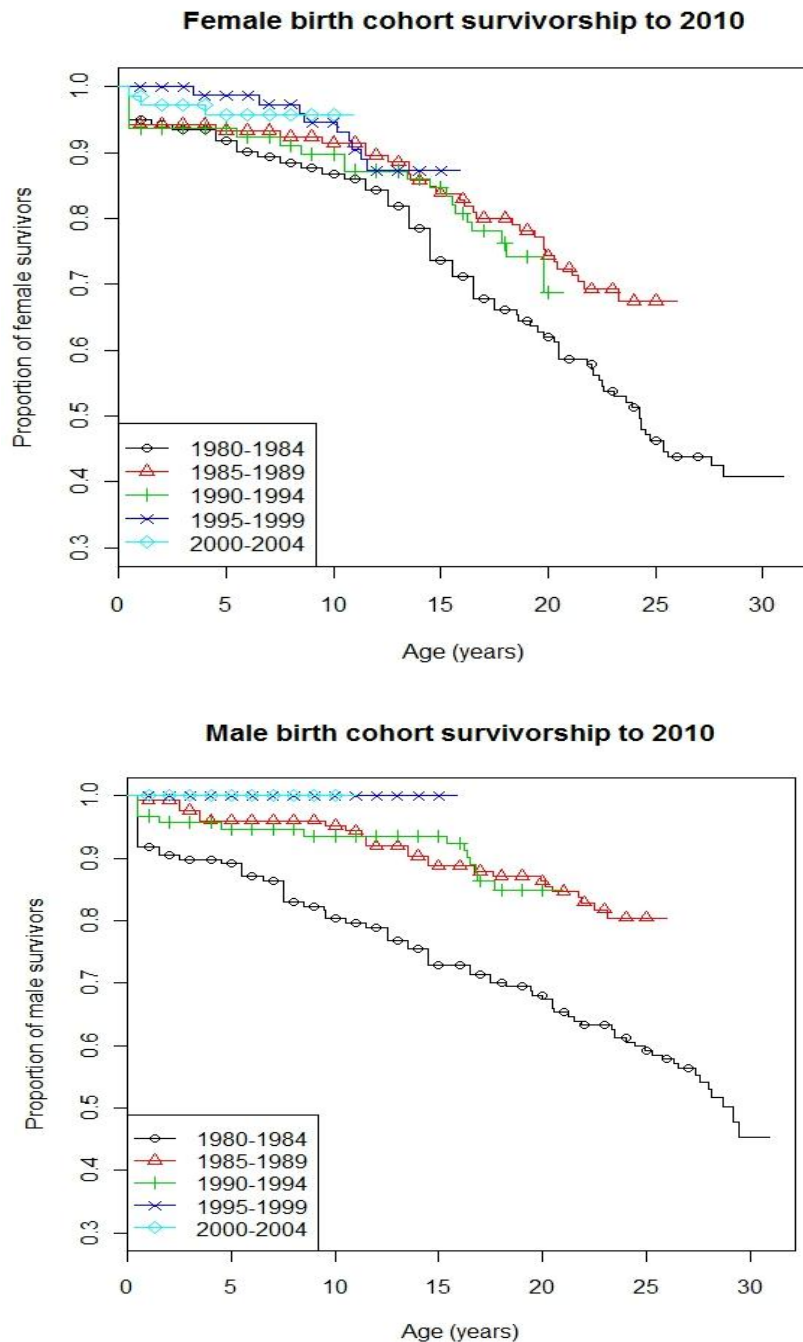
Figure 6: Number of CF deaths and median age at death by gender, 1995-2010



The median age at death describes the duration of life only in those who have died, not those who remain alive, and as such, is a rudimentary measure of CF survival. Median age at death increased to 28.5 years from 25 years in 2009. This sizeable increase from 2009 most likely reflects the unusually older age death of females in 2010; median age at death in females was 23 years in 2009 (n=12) compared with 31 years in 2010 (n=5). A gender gap has long been recognised in CF, with females dying sooner than males for reasons that are poorly understood.

Median survival is the preferred statistic to describe duration of life in the entire CF population, as it takes an entire CF population and follows them until half have died. Figure 7 shows birth cohort lifetable (Kaplan-Meier) survivorship curves to 2010 for males and females born between 1980 and 2004. More than half of PWCF born 1980-1984 died by the end of 2010, making it possible to observe median survival for this cohort. The median survival estimate for PWCF born 1980-1984 was 24.2 years for females and 28.7 years for males. More than half of PWCF born 1985-2004 were alive at the end of 2010, therefore it was not possible to derive a median survival estimate for those birth cohorts at this time.

Figure 7: Birth cohort survivorship to 2010

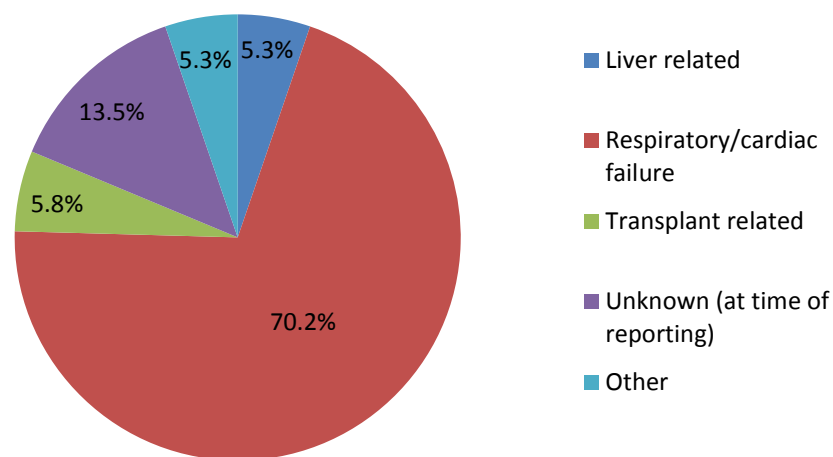


Cause of Death

Of the 17 deaths that occurred in 2010, 13 were due to respiratory/cardiac failure, 2 were transplant-related and the remainder were due to other unspecified reasons. In total, 171 deaths occurred between 2002 and 2010. The causes of death are shown in Figure 8.

Respiratory/ cardiac failure remains the most frequently cited cause of death (70.2%). This is less than the figure of 80% reported internationally and is likely due to the large proportion of deaths reported to the CFRI without a cause of death (13.5%). Transplant-related deaths account for 5.8% of deaths (n=10) over the 9-year period. "Other" causes of death (5.3%) include distal intestinal obstruction, metastatic carcinoma, septic shock, septicaemia and suicide.

Figure 8: Principal cause of death, 2002-2010



Demographics of the CFRI

Demographic data for the 1044 PWCF alive at the end of 2010 is shown in Table 1. Twelve individuals were diagnosed with CF in 2010. However, as it is unlikely that all new diagnoses will have enrolled on the CFRI by the end of 2010, this number will increase as CFRI enrolment continues.

The oldest person on the registry is 60 years of age. The average age of PWCF on the CFRI database continues to increase and on the last day of 2010, the average age was 19.6 years. In 2010, 52.7% of PWCF on the CFRI were aged 18 years and older. This shows the demographic make-up of PWCF is changing from a predominantly paediatric to a growing adult population. A greater proportion of enrolees are male (57.1%), a finding mirrored by other registries. In the Republic of Ireland, this may be due to the disproportionate number of deaths in CF females.

Table 1: Demographic data from the CFRI, 2010

Year	2010	%
PWCF consented	33	
Age range*	<1-60	
Mean age (yrs)*	19.6	
Median age (yrs)*	19	
Number diagnosed during year	12	
Number of males*	596	57.1%
Number of females*	448	42.9%
Number <18 yrs*	494	47.3%
Number ≥18 yrs*	550	52.7%
Number males ≥18 yrs*	321	30.7%
Number females ≥18yrs*	229	21.9%
Irish ethnicity*	1017	97.4%
Deaths during year†	16	
Total PWCF on CFRI who are alive at end of year	1044	

* of the 1044 PWCF on the registry who are alive at the end of 2010

† of those on the registry (i.e. excludes those who died and were not enrolled on the registry)

Figure 9 shows the age distribution of CFRI enrolees by gender. At 168, the 10-14 age band contains the largest number of PWCF, followed by the 20-24 year age band (n=157), 25-29 (n=151) and the 5-9 age band (n=149). In 2010, there were 45 PWCF aged 40 years and over, 60% of which were male.

The county of primary residence for PWCF is reported in Table 2. Dublin PWCF account for 27% of the CF population, followed by Cork (13.2%), Limerick (5.6%) and Tipperary (5.0%). Specialist paediatric and adult CF services are available in the Dublin, Cork and Limerick.

Figure 9: Age and gender distribution by age band, 2010

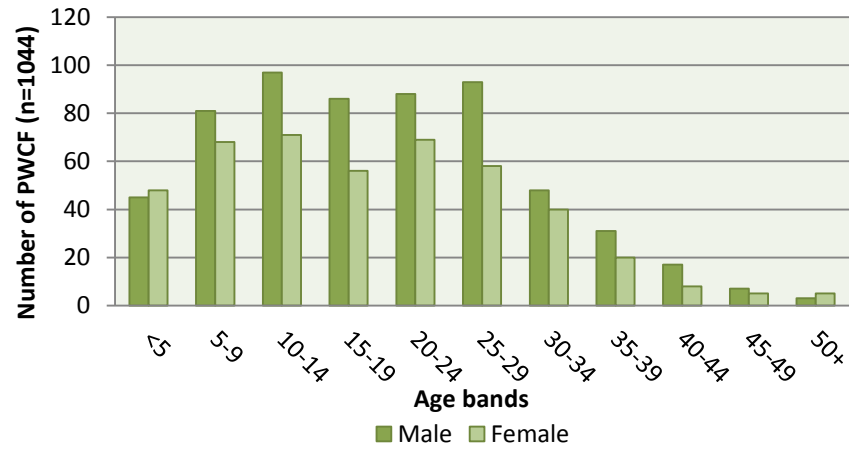


Table 2: PWCF by county of residence, 2010

	Number of PWCF	%
Dublin	282	27.0%
Cork	138	13.2%
Limerick	58	5.6%
Tipperary	52	5.0%
Kildare	50	4.8%
Galway	44	4.2%
Kerry	43	4.1%
Wicklow	40	3.8%
Meath	37	3.5%
Clare	35	3.4%
Mayo	31	3.0%
Wexford	21	2.0%
Kilkenny	20	1.9%
Louth	20	1.9%
Cavan	19	1.8%
Laois	19	1.8%
Westmeath	19	1.8%
Carlow	16	1.5%
Offaly	16	1.5%
Monaghan	14	1.3%
Donegal	13	1.2%
Sligo	13	1.2%
4 counties with <10 patients	23	2.2%
Total	1044	100.0%

Siblings

Due to the small absolute numbers of PWCF in the country (relative to registries in larger countries), it is possible, using data gathered by the CFRI, to monitor the number of CF families and CF siblings. Nine-hundred and ninety-four families with at least one member with CF were represented on the CFRI by the end of 2010. Eight hundred and sixty families had one family member enrolled with the CFRI, 126 had two family members, and 8 families had three or more family members enrolled with the CFRI. These figures do not take into account CF siblings from these families who may have died before the CFRI was introduced, or siblings who have yet to be invited to enrol. These figures may therefore underestimate the number of CF siblings and families in the Republic of Ireland.

Table 3: CFRI families and siblings, 2010

Irish CF families	
994	Families represented
13.5%	134 of 994 families had 2 or more members with CF
126	Number of families with 2 members with CF
8	Number of families with 3 or more members with CF

Diagnosis

2010 marked the last full calendar year before the introduction of a universal programme of newborn screening for cystic fibrosis in the Republic of Ireland (introduced in July 2011). Patterns in CF diagnosis described in future CFRI annual reports will likely change to reflect this new practice.

Of all those enrolled on the CFRI by 2010, gastrointestinal symptoms alone were the most common reason PWCF sought medical care leading to a CF diagnosis (22.2%), followed by respiratory symptoms alone (19.5%), and both respiratory and gastrointestinal symptoms (17.3%) (Figure 10). Nearly 22% (21.7%) had a medical investigation leading to a CF diagnosis as a result of having a family history of CF (some of which also had symptoms). Nearly 15% (14.9%) had symptoms of meconium ileus at birth; just over half of which were treated surgically. Those with “other symptoms” include those who were diagnosed through newborn screening programmes in other countries, those who had some form of screening in Ireland (i.e. meconium test), and others who initially had a diagnosis of asthma.

Figure 10: Symptoms leading to a diagnosis of CF, 2010

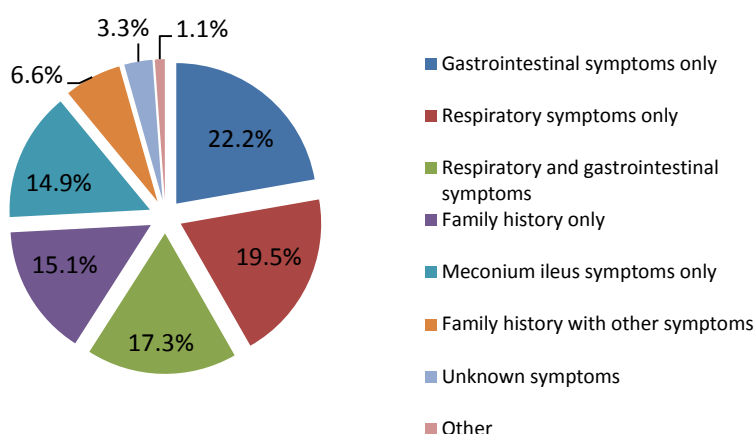
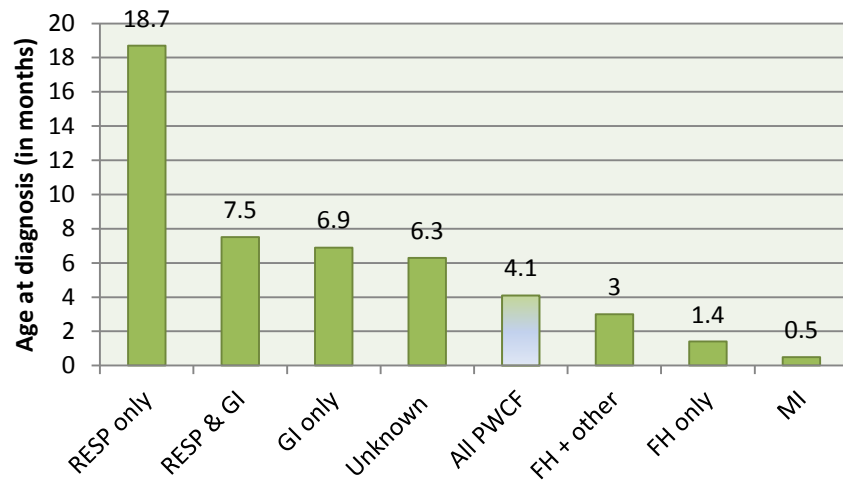


Figure 11 shows the median age at diagnosis in months by symptom type. The figures remain unchanged from 2009. The median age at diagnosis was 4.1 months, with the oldest person diagnosed with CF aged in their late forties. PWCF with respiratory (RESP) symptoms had the oldest median age at diagnosis (one and a half years), followed by PWCF with both respiratory and gastrointestinal (RESP & GI) symptoms (7 and a half months) and PWCF with gastrointestinal (GI) symptoms only (just under seven months). PWCF with meconium ileus (MI) were detected shortly after birth (median age at diagnosis of approximately 2 weeks old). Second to PWCF with meconium ileus, individuals with a family history (FH) of CF were the youngest at diagnosis (1.4 months for family history only and 3 months for PWCF with a family history and symptoms).

Figure 11: Median age at diagnosis in months by symptom category, 2010



Genotype

Genotyping is routinely reported by the National Centre for Medical Genetics in Our Lady's Children's Hospital in Dublin. By 2010, 97.5% of all live CFRI-enrolled PWCF (n=1,044) had genotyping performed. This rate is higher than many leading CF registries. The most commonly detected genotype is delta F508 homozygous (57.3%). Delta F508 G551D (10.8%) and delta F508 R117H (3.6%) are the next most frequently identified genotypes. Ninety percent (90.3%) of CFRI enrolees have at least one delta F508 allele, and such PWCF tend to have more severe disease.

Table 4: Frequencies of the most common CF mutations in living CFRI enrolees, 2010

	Number of PWCF	%
ΔF508 homozygous	598	57.3%
ΔF508 G551D	113	10.8%
ΔF508 R117H	38	3.6%
ΔF508 R560 T/K	25	2.4%
ΔF508 1717-1 G-->A	14	1.3%
ΔF508 621+1 G-->T	13	1.2%
ΔF508 G542X	14	1.3%
All other ΔF508 heterozygotes	108	10.3%
ΔF508 allele 2 unknown	20	1.9%
G551D G551D	8	0.8%
Other genotypes	49	4.7%
Not genotyped	26	2.5%
Pending	18	1.7%
Total	1044	

Hospitalisations and Complications

Each year, the two CFRI Clinical Research Associates (CRA's) retrospectively gather clinical information referred to as "annual assessment data" from CFRI enrollees' medical records at CF centres and clinics across the country. This is currently a paper-based exercise, with large amounts of data collected for each enrollee. This can lead to delays in the completion and reporting of annual datasets.

Annual assessment data was gathered for 866 PWCF in 2010, a similar number to the previous year (n=865). A greater proportion of annual assessment data was gathered for paediatric patients (91.1%) than adults (74.9%) (Table 5). Each hospital has different practices for recording, storing and accessing medical record files, and this presents a challenge to CRA's when attempting to gather a complete annual assessment record for each CFRI enrollee. Data on hospitalizations and respiratory exacerbations can often be incomplete for adults in particular.

Table 5: Annual assessment data collected, 2010

2010 Annual Assessments (AAs)		
	Paediatric	Adult
PWCF with 2010 AA data collected	454	412
Proportion of live PWCF with completed 2010 AAs (N=1044)	91.9%	74.9%

As PWCF age, their condition becomes more complicated. Adult PWCFs therefore have on average a greater number of hospitalisations, respiratory exacerbations and complications than their paediatric counterparts. The average number of hospitalisations and respiratory exacerbations requiring IV antibiotics in paediatric PWCF is slightly lower in 2010 than in 2009, though the number of complications is slightly higher. For adult PWCF, hospitalisations, respiratory exacerbations and complications have increased in 2010.

Table 6: Hospitalisations, exacerbations and complications by age-band, 2010

2010 Annual Assessments (AAs)				
	Paediatric		Adult	
	Number	Average per PWCF	Number	Average per PWCF
Number of hospitalisations	311	0.7	493	1.2
Number of respiratory exacerbations requiring IV antibiotics	321	0.7	741	1.8
Number of complications	1364	3.0	1637	4.0

Very few PWCF reported having no complications in 2010; paediatric PWCF had on average of 3 complications, while adults had 4 on average (Table 7). Gastrointestinal complications were reported more commonly than respiratory or other (miscellaneous) complications.

All paediatric PWCF and nearly all adults (98.1%) were described as pancreatic insufficient and requiring pancreatic enzymes. The number of paediatric PWCF with liver disease continues to increase from 13 in 2008, to 26 in 2009 and 38 in 2010, as does the number of adults with liver disease: 46 in 2008, 58 in 2009 and 64 in 2010.

Chronic *Staphylococcus aureus* infection (≥ 3 isolates in a year) was reported in 54.0% of paediatric and 24.2% of adult PWCF, while chronic *Pseudomonas aeruginosa* (≥ 3 isolates in a year) affected 24.2% of

paediatric and 65.5% of adults. The proportion of PWCF with these infections in both paediatric and adults has increased slightly since 2008. The proportion of CFRI enrollees with *Burkholderia cepacia* has also increased, with nearly twice as many affected in 2010 than in 2009 (12 in 2009 and 23 in 2010). MRSA however, has been reported less frequently in 2010, though more MRSA continues to be detected in paediatric than adult PWCF. The proportion of adults with insulin-dependent diabetes and osteopenia/osteoporosis has continued to increase, affecting 30.1% and 43.3% respectively in 2010.

Table 7: Complication rates by system by age-band, 2010

2010 Annual Assessment				
Paediatric			Adult	
No of PWCF with completed AAs	454		412	
PWCF with no complications	5		4	
Total number of complications	1364		1637	
Cardiac/Pulmonary Complications				
	Number of paediatric PWCF	% of group	Number of adult PWCF	% of group
Chronic <i>Pseudomonas</i>	110	24.2%	270	65.5%
Chronic <i>Staphylococcus</i>	245	54.0%	113	27.4%
<i>Burkholderia</i>	6	1.3%	17	4.1%
MRSA	57	12.6%	32	7.8%
Nasal polyps	20	4.4%	3	0.7%
ABPA	16	3.5%	14	3.4%
Asthma	5	1.1%	6	1.5%
All other cardiac/pulmonary complications*	4	0.9%	6	1.5%
<i>Total card/pulm complications</i>	463		461	
Gastrointestinal Complications				
DIOS	5	1.1%	15	3.6%
Rectal Prolapse	0	0%	0	0%
Pancreatic Insufficiency	454	100%	404	98.1%
Abnormal LFTs	5	1.1%	8	1.9%
Liver disease	38	8.4%	64	15.5%
GI Reflux	102	22.5%	146	35.4%
All other gastro complications**	0	0%	7	1.7%
<i>Total gastro complications</i>	604		6440	
Miscellaneous Complications				
Diabetes requiring insulin	11	2.4%	124	30.1%
Clubbing	197	43.4%	139	33.7%
Osteopenia/osteoporosis	19	4.2%	205	49.8%
Other non-CF morbidities	70	15.4%	64	15.5%
<i>Total misc complications</i>	297		532	

*All other respiratory/cardiac complications: cor pulmonale, pneumothorax, haemoptysis

**All other gastrointestinal complications: haematemesis, colonic stricture, gallbladder disease, pancreatitis

Cultures

In 2010, the results of 11,243 culture swabs were recorded by the CFRI. Of those, 7,189 were sputum samples, a figure similar to that reported in 2009 (n=7,103). Sputum samples are the most frequently tested specimen type; on average 9.4 sputum cultures were tested for paediatric PWCF compared with 9.0 for adults in 2010 (Table 8). In paediatric PWCF, throat and cough swabs are also commonly taken for examination.

Table 8: Culture types by age-band, 2010

Sample type	Paediatric		Adult	
	Number	Average number of cultures per paediatric PWCF	Number	Average number of cultures per adult PWCF
Sputum samples	4071	9.4	3118	9.0
Cough swabs	1681	3.9	81	0.2
Throat swabs	1696	3.9	32	0.1
BAL swabs	124	0.3	13	<0.1
Nasal swabs	222	0.5	26	0.1

Organisms frequently detected in sputum specimens are presented in Table 9. *P aeruginosa* is the most commonly detected organism in sputum (24.7%). Species of *Candida* were the next most frequently recorded organism in sputum cultures (15.1%), followed by *S aureus* (16.0%).

Table 9: Frequently detected organisms in sputum cultures, 2010

	Number of positive sputum cultures	% of positive sputum cultures
<i>Staphylococcus aureus</i>	1149	16.0%
All <i>Candida</i> species	1083	15.1%
<i>Pseudomonas aeruginosa</i> (Muroid status not reported)	869	12.1%
Normal flora	793	11.0%
<i>Pseudomonas aeruginosa</i> (Muroid)	641	8.9%
<i>Aspergillus fumigatus</i>	388	5.4%
Gram positive cocci	315	4.4%
MRSA	289	4.0%
<i>Pseudomonas aeruginosa</i> (Non-muroid)	267	3.7%
<i>Haemophilus influenza</i>	254	3.5%
<i>Stenotrophomonas maltophilia</i>	209	2.9%
Gram negative bacilli	191	2.7%
<i>Burkholderia cepacia</i> complex*	97	1.3%
Other	644	9.0%
Total	7189	

*Contains 37 unspecified *Burkholderia cepacia* sputum cultures, 36 *B multivorans*, 15 *B cenocepacia* and 9 *B cepacia*.

Antibiotics

The cumulative number of days PWCF spent taking IV antibiotics in 2010 is shown in Figure 12. In total, PWCF spent over 29,000 days taking the IV antibiotics listed in Figure 12 alone. This equates to an average of 33.8 days per PWCF with annual assessment data in 2010 (n=866). Since 2009, the cumulative number of IV antibiotic days has increased by approximately 8,000 days. This increase is most notable for IV antibiotics flucloxacillin and meropenem, where the number of cumulative IV antibiotic days rose by 1,000 days for each antibiotic respectively since 2009. In 2010, tobramycin (n=6,489 days), ceftazidime (n=6,288), meropenem (n=4,587) and colistin sulphomethate (4,072) were the four IV antibiotics with the greatest cumulative number of antibiotic days.

Figure 12: Rank of order of IV antibiotics, 2010

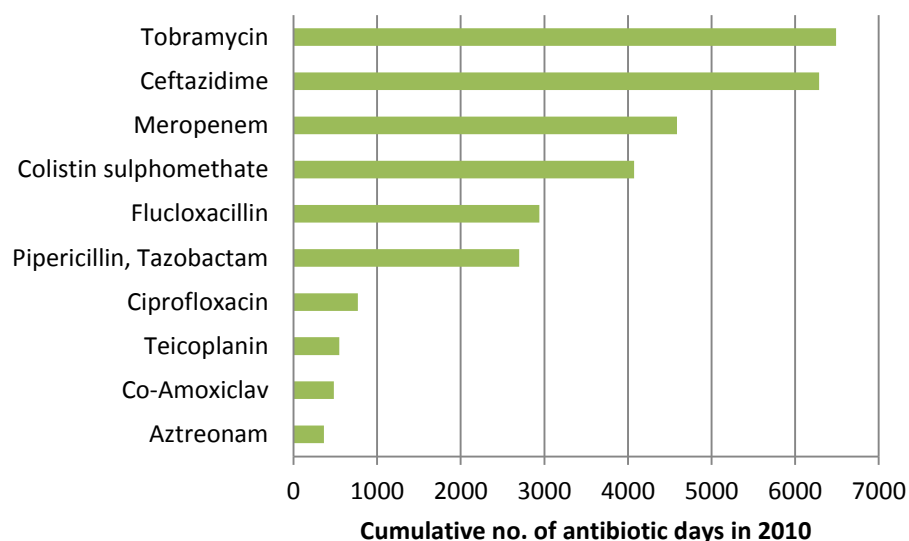
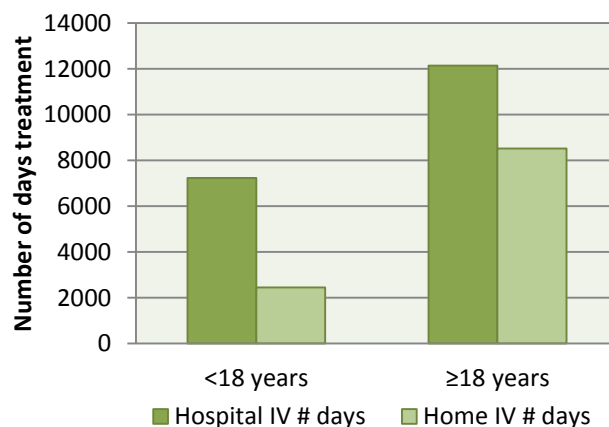


Figure 13 shows the cumulative number of days paediatric and adult PWCF received IV antibiotics according to where they were administered (hospital/home). Overall, the administration of IV antibiotics in hospital continues to be more popular than at home. For adult PWCF in 2009, home IV administration of antibiotics occurred nearly as frequently as hospitalisation for IV antibiotic treatment. This trend did not continue in 2010; adults had 12,130 hospital IV antibiotic days compared to 8509 home IV antibiotic days.

Figure 13: Cumulative number of hospital and home IV antibiotic days, 2010



Pulmonary Function

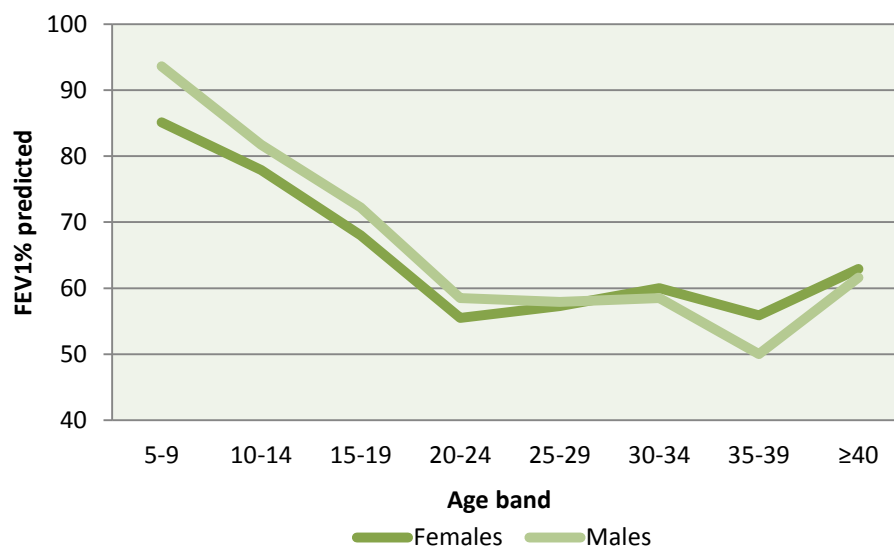
The results of 2,686 pulmonary function tests (PFTs) were recorded in PWCF aged 5 years and older in 2010 (Table 10). Table 10 shows that in 2010 mean FEV₁% predicted and FVC% predicted was best in those aged 5-9 years and that lung function was poorer in older age cohorts. This is expected, as CF is a disease which causes the functioning of the lungs to deteriorate with age. Compared with 2009, mean FEV₁% predicted and FVC% predicted in 2010 was slightly better in nearly every age band.

Table 10: Pulmonary function test summary, 2010

Age bands	Number of PFTs	Mean FEV ₁ % predicted	Mean FVC % predicted
5 - <10	444	89.5	89.6
10 - <15	567	80.1	86.0
15 - <20	424	70.3	82.3
20 - <25	464	57.3	75.7
25 - <30	372	57.6	75.7
30 - <35	211	59.2	81.9
35 - <40	101	53.0	74.1
≥40	103	62.2	84.9
Total	2686		

Figure 14 shows that in 2010, mean FEV₁% predicted was better in males than in females in each age band from 5 to 24 years. Mean female FEV₁% predicted was worst in those aged 20-24, but generally improved in successive age bands thereafter. Mean male FEV₁% predicted remained constant in the 20-34 age bracket and fluctuated in successive age bands. The trend shown in Figure 14 is broadly similar to that reported in previous years, as males had better mean FEV₁% predicted in younger age bands, and females aged 30 years and older had better values than males.

Figure 14: FEV₁% predicted mean values by age band and gender, 2010



Body Mass Index

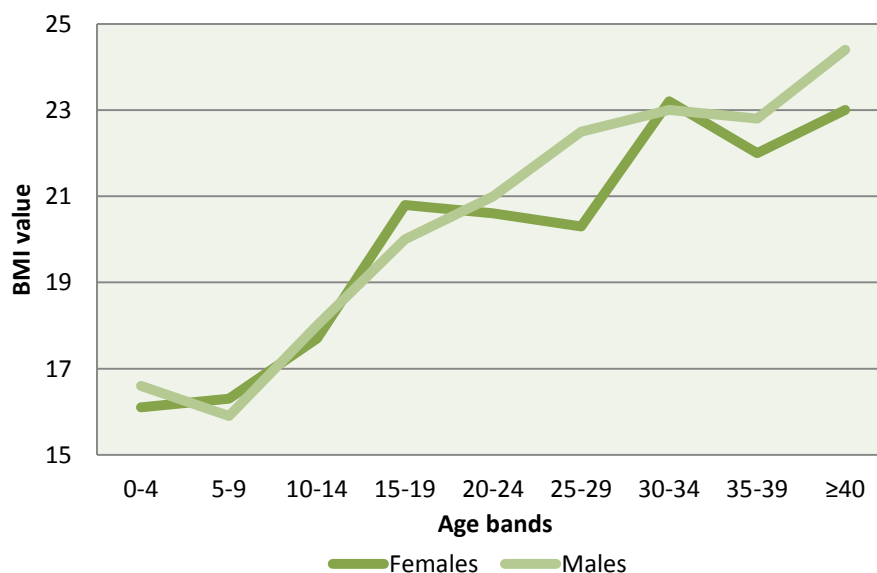
Nearly 3,000 body mass index (BMI) scores recorded in 2010 were retrieved by the CFRI (Table 11). This is similar to that recorded in 2009 (n=2820) for reason previously described. BMI values across all age bands remain similar to those reported in previous years. BMI is a useful measure of nutrition and growth for individuals aged ≥ 20 years but are not entirely appropriate for measuring growth and nutrition in childhood.

Table 11: BMI summary, 2010

Age bands	N	Mean BMI
2 - <5	270	16.4
5 - <10	548	16.0
10 - <15	600	17.9
15 - <20	406	20.3
20 - <25	427	20.8
25 - <30	344	21.5
30 - <35	164	23.0
35 - <40	88	22.4
≥ 40	82	23.7
No of PWCF	2929	

Stratifying mean BMI by gender (Figure 14), we find that mean BMI is relatively similar for both sexes across most age bands (with the exception of PWCF aged 25-29 years where males and females have a mean BMI of 22.5 and 20.3 respectively). BMI generally increased in each consecutive age band. As in previous years, there was a distinct pattern in females whereby mean BMI worsened between the 20-24 and 25-29 age bands.

Figure 15: BMI mean values by age band and gender, 2010



Nutrition

Nutritional therapies are important for improving growth and nutrition in PWCF, as better nutrition is associated with improved life expectancy. Most PWCF received at least one form of nutritional treatment in 2010 (Table 12); vitamin supplements being the most popular in both paediatric (94.3%) and adult patients (85.0%).

Table 12: Nutritional treatments, 2010

2010 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		412	
	Number of paediatric PWCF	% of group	Number of adult PWCF	% of group
Calorie supplements	85	18.7%	90	21.8%
Vitamins	428	94.3%	350	85.0%
Minerals	34	7.5%	33	8.0%
Gastrostomy feeds	12	2.6%	39	9.5%

Nearly one third of PWCF (31.5%) received some form of supplemental feeding in 2010, and the strategies adopted are described in Table 13. Oral supplements were used by approximately one in five PWCF. Ten percent of adults and paediatric PWCF respectively had a gastrostomy performed in 2010.

Table 13: Supplemental feeding, 2010

2010 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		412	
	Number of paediatric PWCF	% of group	Number of adult PWCF	% of group
Oral supplements	103	22.7%	83	20.1%
Nasogastric tube insertion	5	1.1%	2	0.5%
Gastrostomy tube insertion	45	9.9%	42	10.2%

Physiotherapy

More paediatric PWCF are reviewed by physiotherapists than adult PWCF each year (Table 14). Nearly 88% of paediatric PWCF were reviewed in 2010, an increase of approximately 8% since 2009. Fewer adults, however, were reviewed in 2010 (57.5%) than in 2009 (62.5%).

The number of PWCF using each physiotherapy modality is shown in Table 14 (patients often use more than one physiotherapy modality). For paediatric patients, the positive expiratory pressure mask was reported most frequently with one in three paediatric PWCF using this form of physiotherapy. Acapella (30.6%) and percussion (16.3%) was the next most common. For adult PWCF, the most popular modality was autogenic drainage, and was used by approximately one in three (35.4%), followed by the PEP mask (17.7%).

Table 14: Physiotherapy summary, 2010

2010 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		412	
PWCF seen by Physio at AA	399		237	
% seen by Physio	87.9%		57.5%	
	Number of paediatric PWCF	% of group	Number of adult PWCF	% of group
PEP Mask	153	33.7%	73	17.7%
Acapella	139	30.6%	25	6.1%
Postural drainage	21	4.6%	2	0.5%
Percussion	74	16.3%	0	-
Active cycle breathing	16	3.5%	12	2.9%
Autogenic drainage	24	5.3%	146	35.4%
Flutter	22	4.8%	12	2.9%
Vest	15	3.3%	5	1.2%
Other*	35	7.7%	13	3.2%

*Other: positioning, Therapep, trampolining, blowing bubbles, chest compressions, BiPAP, running.

Long-Term Medications

Long-term medications used by PWCF are described in Table 15. Beta-agonists remain the most frequently administered therapy, and the proportion of adults and paediatric PWCF receiving this treatment has increase since 2009 (from 46.7% to 53.7% in paediatric, and 54% to 60.7% in adult PWCF). More adults were receiving osteoporosis treatment in 2010 than in the previous year (53.6% and 47.9% respectively). More adults were also being treated with H2RA/proton pump inhibitors (PPI) (39.9% in 2009 and 49.3% in 2010). Just over one in five PWCF are taking insulin for CF-related diabetes. For paediatric patients, the proportion using nebulised saline in 2010 increased by nearly 9% since 2009. Beta-agonists rhDNAse and inhaled steroids are the most common long-term treatments administered to under 18s.

Table 15: Long-term medication summary, 2010

2010 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		412	
	Number of paediatric PWCF	% of group	Number of adult PWCF	% of group
Beta agonist	244	53.7%	250	60.7%
Inhaled steroid	143	31.5%	215	52.2%
rhDNAse	176	38.8%	169	41.0%
Urso-deoxycholic acid	87	19.2%	70	17.0%
Osteoporosis treatment	103	22.7%	221	53.6%
H2RA/PPI	114	25.1%	203	49.3%
Anti-cholinergic	12	1.6%	12	2.9%
Lactulose	24	5.3%	16	3.9%
Oral steroid	18	4.0%	31	7.5%
Insulin	10	2.2%	91	22.1%
Night-time oxygen	12	2.6%	15	3.6%
Nebulised saline	79	17.4%	15	3.6%
Aminophylline/theophylline	1	0.2%	6	1.5%

Financial Information

The financial summary in Table 16 lists the Income and Expenses for the CFRI in 2010.

Table 16: Income & Expenses for 2010

Income & Expenses		2010 €
Income		
Grant income		132,000
Sundry income		504
Bank deposit interest		
	Total income	132,504
Expenses		
Salaries & interim management		123,334
Employer's PRSI		8,969
Rent payable		1,379
Property provision		(17,813)
License fee		267
Insurance		489
Computer network & server costs		4,142
Database costs		0
Heat & light		221
Repairs & maintenance		0
Printing, postage and stationery		492
Computer costs		409
Travelling & subsistence		7,797
Audit		968
Bank charges		241
Sundry expenses		0
Depreciation on equipment		1,519
	Total expenses	132,414
	(Deficit)/Surplus	90

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. We would also like to thank the Health Research Board for funding a 3-year research programme (2008-2010).

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.

The Cystic Fibrosis Association of 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 nursing staff at each site for their continuing co-operation.

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 De La Harpe and Dr H Johnson for their support and encouragement during the year.

Finally, we thank the National Centre for Medical Genetics in Our Lady's Children's Hospital in Crumlin for providing CF genotyping information.

Published Papers and Conference Contributions, 2010

Published Papers

1. Jackson AD, Harrington M, Zhou S, Daly L, Kelleher C, Fitzpatrick P, Foley L. 'Delayed cystic fibrosis presentation in children in the absence of newborn screening' *Irish Medical Journal*, 2010, 103(4) p113-116.

Conference Contributions

2. Jackson AD, Daly L, Kelleher C, Fletcher G, Harrington M, Zhou S, Fitzpatrick P. 'Cystic fibrosis service provision and usage patterns: an analysis of Cystic Fibrosis Registry of Ireland data' *Royal College of Physicians Faculty of Public Health Medicine Ireland Winter Meeting*, 2010, *Irish Journal of Medical Science*, 180(Suppl 6).
3. Jackson AD, Daly L, Kelleher C, Fletcher G, Harrington M, Zhou S, Fitzpatrick P. 'Factors associated with poor pulmonary function in cystic fibrosis adults in the Republic of Ireland: a cross-sectional analysis of registry data' *The Irish Thoracic Society Annual Scientific Meeting*, 2010, *Irish Journal of Medical Science*, 179(Suppl 12), pS464.
4. Jackson AD, Kelleher C, Fitzpatrick P, Fletcher G, Harrington M, Zhou S, Daly L. 'How representative are cystic fibrosis (CF) registries of the CF population' *24th Annual North American Cystic Fibrosis Conference*, 2010. *Pediatric Pulmonology*, 45(Suppl 33) p392.
5. Jackson AD, Daly L, Kelleher C, Jackson AL, Marshall BC, Quinton HB, Harrington M, Zhou S, Foley L, Fitzpatrick P. 'A novel method of projecting cystic fibrosis birth cohort survival estimates that overcomes short duration of follow-up' *33rd European Cystic Fibrosis Society Conference*, 2010. *Journal of Cystic Fibrosis*, 9(Suppl 1) pS112. (Winner of 33rd European Cystic Fibrosis Society Conference 2010 Best Oral Epidemiology Poster Award.)
6. Jackson AD, Daly L, Jackson AL, Kelleher C, Harrington M, Zhou S, Foley L, Fitzpatrick P. 'CF Registry Ireland data shows improved survival in the Republic of Ireland' *10th Irish National Cystic Fibrosis Conference*, Killarney, 2010.

“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.”

Mission Statement of the CFRI

Cystic Fibrosis is an inherited condition that affects many body functions such as breathing, digestion, and reproduction. This lifelong condition usually becomes more severe with age and affects both males and females in equal proportions. The symptoms and severity of cystic fibrosis vary from person to person. The majority of people have both respiratory and digestive problems. There is no cure for cystic fibrosis. Life expectancy has increased steadily over the past 20 years, and today cystic fibrosis is no longer exclusive to childhood.

Better treatment strategies help to improve the length and quality of life of people with CF by controlling their symptoms. Improved treatments can be developed using patient registries. Cystic fibrosis registries gather information on all aspects of a patient's condition. They act as information storehouses for infection and treatment statistics. Detailed analysis of this information can yield significant findings about the most effective treatments for CF. It is through these analyses that better management of CF may be achieved.

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Charity Number: CHY17566

