Cystic Fibrosis Registry of Ireland Annual Report 2009

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Executive Summary

The Cystic Fibrosis Registry of Ireland (CFRI) collects and analyses 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.

Adequate representation of the CF population is required to ensure statistics generated by analysis of registry datasets are valid. By comparing CFRI's annual census of all CF centres with PWCF's enrolled we have been able to estimate that by the end of 2009, the registry had achieved an ascertainment level 89.2%. This level of coverage of the CF population by a CF registry is on par with leading international CF patient registries and ensures that statistics generated are adequately representative of the CF population in the Republic of Ireland. With this high ascertainment level a huge effort had to be put in by registry staff during 2009 to collect the required data from patient charts. This is a very manual and tedious process and requires considerable collection and interpretation skills. We have made huge strides in getting our patient data current and 2010 data collection will be completed by June/July of 2011 with earlier targets for 2011 data.

Quality data that is current is essential to support research and decision making processes that will improve the health of the population, effectively manage scarce resources and assist in the monitoring of the quality of care provided to our patient population.

In October 2009 the HSE released the findings of the CF Working Group and made a specific reference to the registry by stating that the registry should continue to be supported by the HSE and be developed further to address the following needs:

- Clinical information for each individual unit to support local service audit and accreditation.
- Timely activity data for planning and service monitoring purposes.
- Costs of treatment to allow for banding packages of care.
- Information downloads to the CFRI Register capable of providing outcome information on survival, lung function (FEV 1), nutritional status, number of infections per year, quality of life measures (e.g. fertility in adults, employment) and symptom control.

It is critically important that the registry is properly resourced as the data that is collected is an essential tool in supporting clinical governance and research.

The registry's IT technology has supported us very well since we were established. Five years ago our platform was innovative and incorporated the latest technology being the first patient registry to use secure internet technology to access patient data. Due to a lack of funding, investment in our technology has fallen behind. The capture of patient data currently is manual, time consuming and expensive. We need to improve the data capture process and move to data capture at source and be able to interface to and pull relevant data from other national databases thereby increasing efficiencies. We need to improve our technology in order to make data more readily available to health carers in their clinics so that they have appropriate tools to assist in the management of their patients.

In conclusion, I would particularly like to thank Ms Mary Harrington, Dr Shijun Zhou and Dr Abaigeal Jackson for their support and the huge volume of work that they put in during the year.

Godfrey Fletcher
Interim CEO

Summary Statistics for 2009

	2007	2008	2009
Number of patients consented	762	1065	1105
Median age (years)	17	17.9	18
Number diagnosed during the year	32	19	9
Number of patients with genotype information	97%	96%	96.9%
Number of enrolled patients deceased during the year	17	17	19
Median age at death (years)	24	23	25
Number of consented individuals alive at end of year	718	1004	1027
% alive at end of year who were male	53.5%	57.4%	57.6%
% adults (≥18 years) alive at end of year	45.5%	49.4%	51.7%
Number of live patients identified by annual census	1170	1142	1151
% ascertainment of the live CF population by the CFRI	61.4%	87.9%	89.2%
Number of patients for whom annual clinical data was collected	143	813	865
% of patients alive at end of year for whom annual clinical data was collected	20.6%	81.0%	84.2%

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Cystic Fibrosis Specialist Centres, 2009

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
	National Children's Hospital	Dr Peter Greally /	Paediatric
		Dr Basil Elnazir	
	Our Lady's Children's Hospital	Dr Gerry Canny /	Paediatric
		Dr Barry Linnane	
	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 MJ Mahony	Paediatric
		Dr Eithne Mulloy /	Adult
		Dr TH Peirce	
Louth	Our Lady of Lourdes Hospital	Dr David Vaughan	Paediatric
		Dr John Kiely	Adult
Mayo	Mayo General Hospital	Dr Michael O'Neill	Paediatric
Sligo	Sligo General Hospital	Dr R Tummaluru	Paediatric
Waterford	Waterford Regional Hospital	Dr A Das	Paediatric
		Dr S Foley	Adult

CFRI Executive Council: 2008-2011

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
CFTR	Cystic Fibrosis Transmembrane conductance Regulator
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 *Staphylococcus aureus*
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

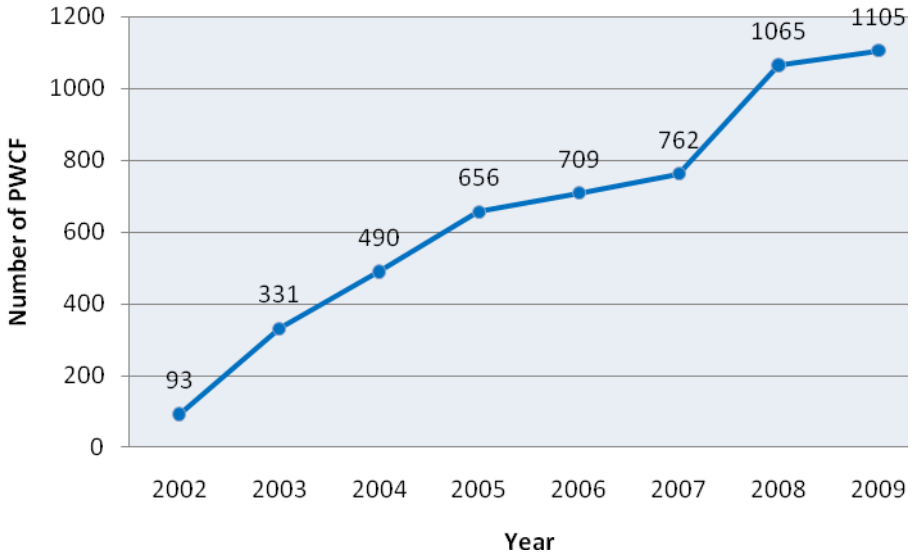
AMNCH	National Children's Hospital at the Adelaide & Meath Hospital, Tallaght, Dublin 24
BMT	Beaumont Hospital, Dublin 9
Cavan GH	Cavan General Hospital, Cavan
CUH	Cork University Hospital, Wilton, Cork
CUHTS	Children's University Hospital, Temple Street, Dublin 1
Kerry GH	Kerry General Hospital, Tralee
Letterkenny GH	Letterkenny General Hospital
Mayo GH	Mayo General Hospital, Castlebar
MWRH	Midwest Regional Hospital, Limerick
MMUH	Mater Misericordiae University Hospital, Dublin 7
Mullingar GH	Mullingar General Hospital
OLCH	Our Lady's Children's Hospital, Crumlin, Dublin 12
OLLH	Our Lady of Lourdes Hospital, Drogheda
Portiuncula GH	Portiuncula General Hospital
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

By the end of 2009, 1,105 persons with CF (PWCF) had enrolled with the CFRI (Figure 1). The number of PWCF (n=40) enrolled in 2009 was down by comparison with the previous year. 2008 was an exceptional year as a second CRA was added to the CFRI team. The growth in CFRI enrolees has led to a corresponding increase in the total number of PWCF for whom annual clinical assessment data must be collected on an ongoing basis. In 2009, both CRAs dedicated a large portion of their time to the collection of 2008 and 2009 annual assessment data.

Of the 1,105 PWCF enrolled by the end of 2009, 56.1% were male. Nineteen enrolled PWCF were known to have died in 2009. Of the 1,027 enrolled PWCF alive at the end of 2009, 51.7% were aged 18 years or older. This is the first year in which adults outnumbered paediatric patients on the CFRI database.

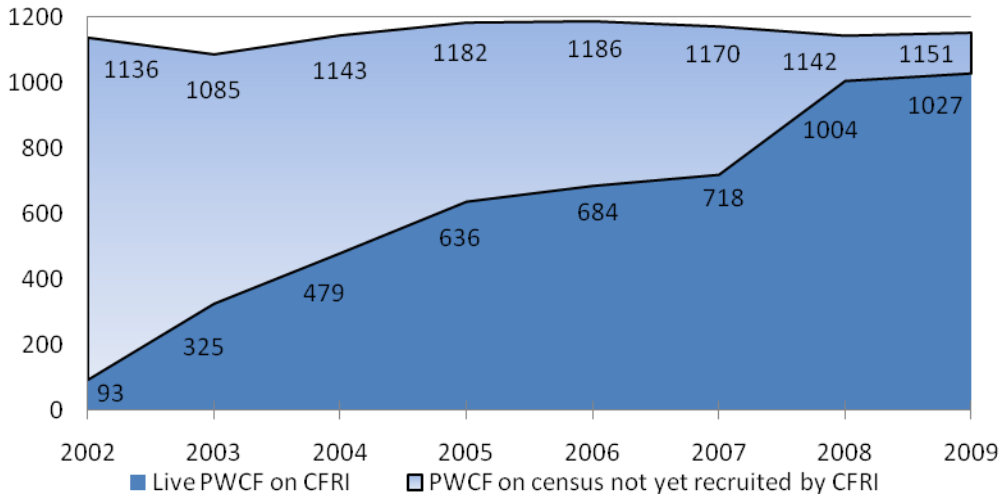
Figure 1: CFRI enrolment 2002 – 2009



CF Centre & Clinic Census 2009

The CFRI's annual census of living PWCF indicates that there were 1,151 persons with a known diagnosis of CF attending CF centres and clinics in the Republic of Ireland in 2009. By the end of 2009, an estimated 89.2% had enrolled with the CFRI (Figure 2). This level of coverage of the CF population by a CF registry is on par with leading international CF patient registries and ensures that statistics generated are adequately representative of the CF population in the Republic of Ireland. The CFRI will continue to enrol new and existing PWCF, though the rate of enrolment may have peaked and further increases in coverage of the CF population will prove a challenge in the years ahead.

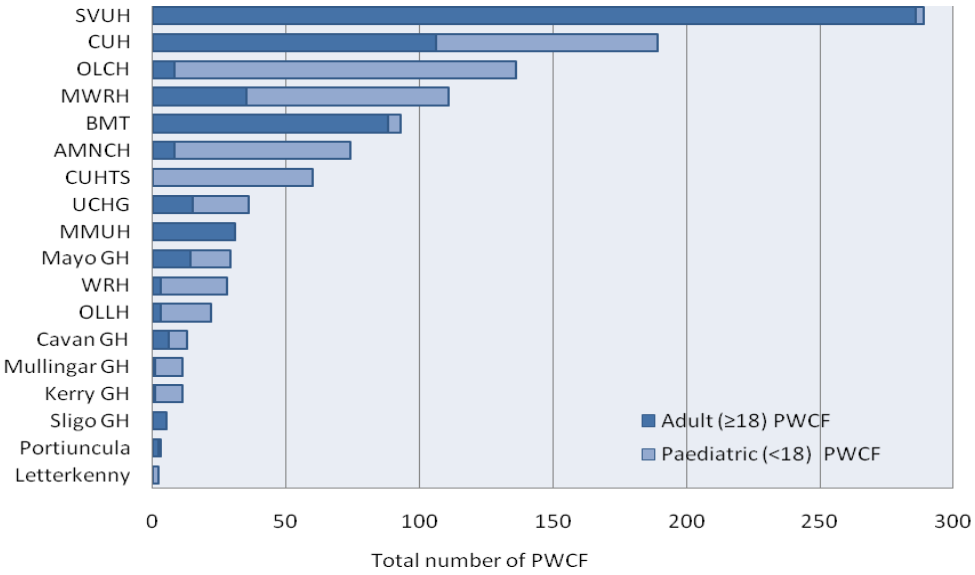
Figure 2: Gap in CFRI enrolment and CF centre census of living PWCF



Census estimates are presented by CF centre/clinic and age band in Figure 3. The accuracy of census reporting has been improved in recent years as duplicate records have been identified and removed from the census. While the total census estimate for the Republic of Ireland (n=1151) is likely to be accurate, the enumeration of PWCF attending specific hospitals is more difficult because PWCF often share care between specialist CF centres or specialist and satellite centres. Census estimates for specific hospitals in 2009 may not be entirely accurate.

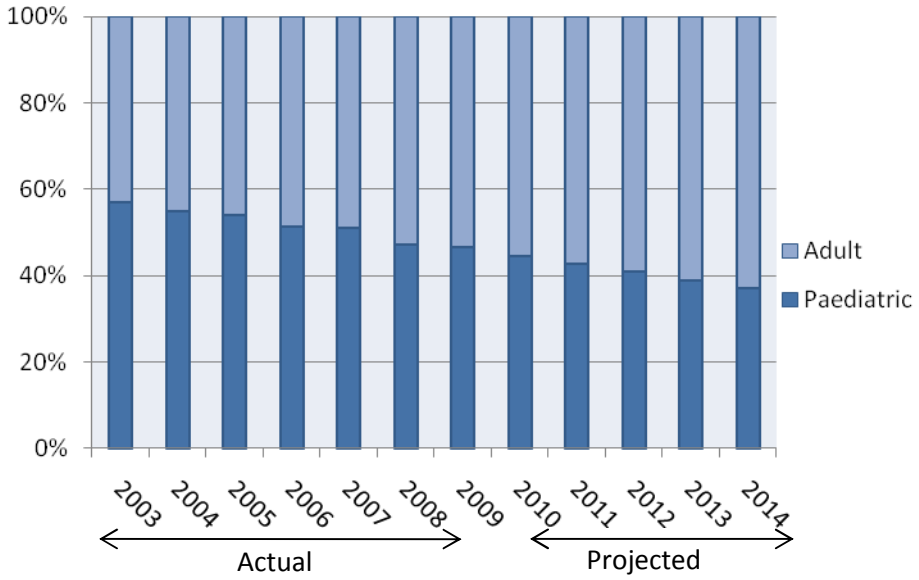
As in previous years, St Vincents University Hospital (n=286), Cork University Hospital (n=106) and Beaumont Hospital (n=88) had the largest number of adults attending for CF services in 2009. Our Lady's Children's Hospital (n=128), Cork University Hospital (n=83) and the Mid-Western Regional Hospital (n=76) had the largest number of paediatric patients availing of CF services in the same year.

Figure 3: CF centre census of live patients in 2009, n=1151



In 2009, 53.2% of census-identified PWCF were aged 18 years or older. Census estimates indicate that the adult CF population has grown. More accurate reporting of census-identified PWCF in 2008 and 2009 may be a reason that the growth in the proportion of adult PWCF was less marked between 2008 and 2009 (an increase from 53% to 53.2% respectively). The linear projection of growth in the adult population to 2014 shown in Figure 4 is based on the average annual increase in census-reported adult PWCF since 2002. While it is anticipated that the adult CF population will continue to grow in the coming years, actual observed year-on-year increases in the adult CF population may be more conservative than that projected in Figure 4.

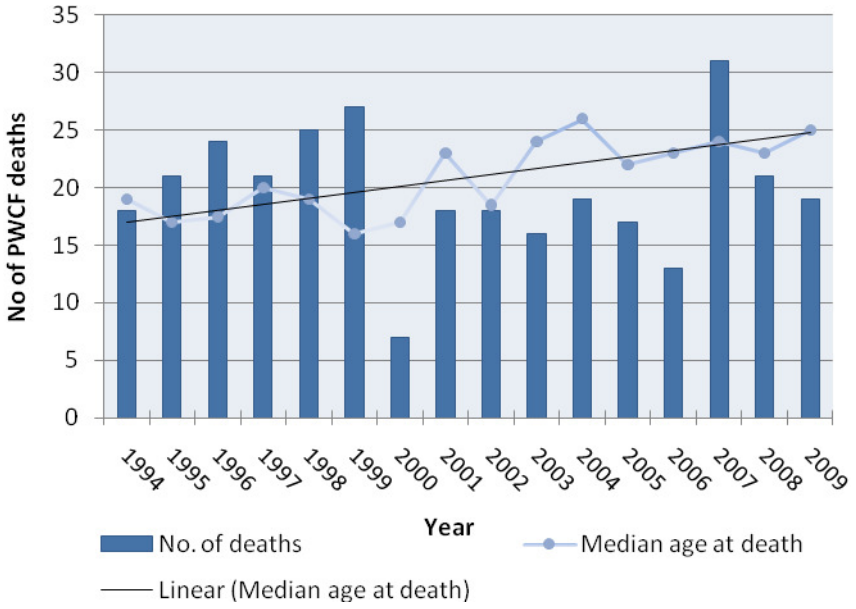
Figure 4: Projected ratio of adult to paediatric CF patients to 2014



CF Survival

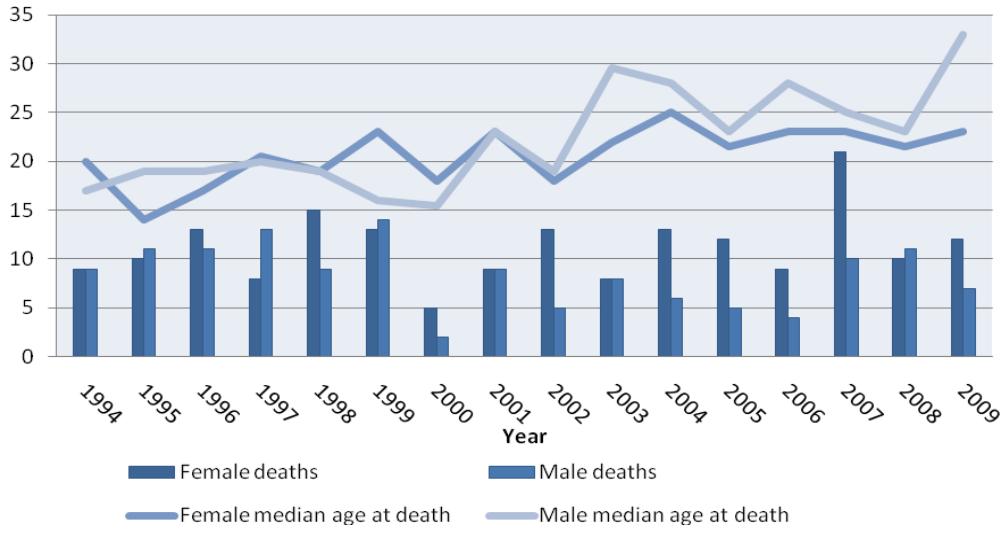
In addition to reporting on CF death information from the CFRI alone, information is now routinely sourced from the Central Statistics Office and the Cystic Fibrosis Association of Ireland. In total, 19 deaths were reported in 2009. This is within the normal range of deaths observed in any given year in the Republic of Ireland (Figure 5). The median age at death in 2009 was 25 years, indicating that median age at death continues to increase. The ages at which PWCF died in 2009 ranged from 10 to 39 years.

Figure 5: Total number of deaths and median age at death of PWCF, 1994-2009



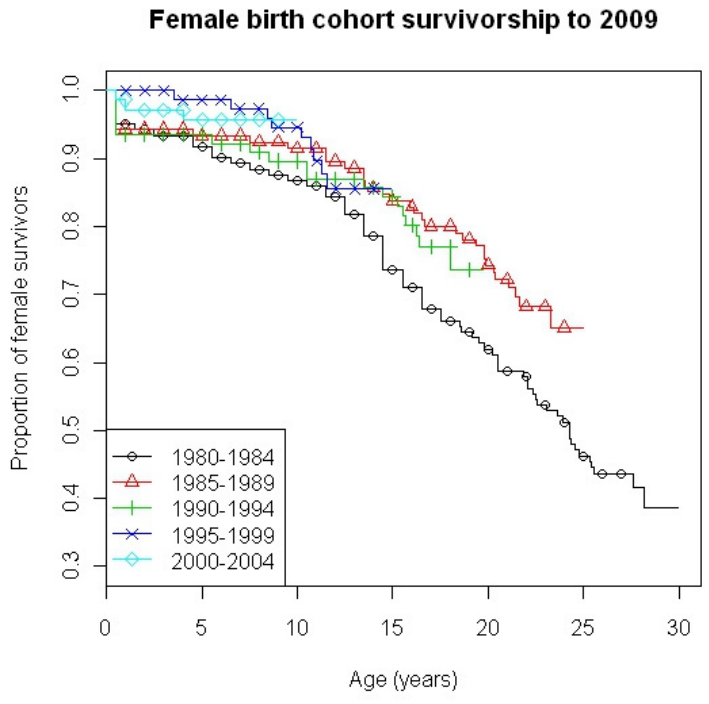
More deaths occurred in females (n=12) than in males (n=7) in 2009, a trend that has prevailed for much of the last decade. Furtheremore, median age at death is older in males than in females (Figure 6). While female median age at death in 2009 was 23 years (and has remained around 23 years for the last five years or so), median age at death for males in 2009 was unusually large in 2009 (33 years). It should be noted, however, that the male estimate was based on a very small number of deaths.

Figure 6: Number of CF deaths and median age at death by gender, 1994-2009



Birth cohort lifetable (Kaplan-Meier) survivorship curves to 2009 are shown in Figure 7 for males and females born between 1980 and 2004. More deaths occurred in infancy (unde 5 years) for PWCF born 1980-1994 than for PWCF born 1995-2004. As death in infancy has become increasingly rare, the average age to which PWCF can expect to live has improved, particularly for PWCF born from 1995 onwards. A gender gap in CF survival has long been recognised both at home and abroad, although reasons for this disparity are not entirely clear.

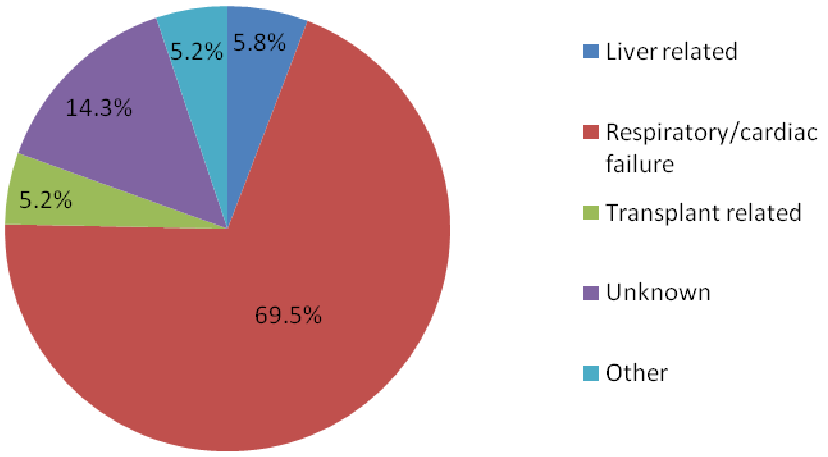
Figure 7: Birth cohort survivorship to 2009 for PWCF born 1980-2004 by gender



Cause of Death

One hundred and fifty-four CF deaths occurred between 2002 and 2009 (Figure 8). Respiratory/cardiac failure was cited as the principle cause of death for approximately 70% of deceased PWCF. This compares with a figure of 80% internationally. The proportion of deaths due to respiratory and cardiac failure in the Republic of Ireland has declined from 81% in 2007 to 69.5% in 2009. However, there has been a concurrent increase in the number of deaths with an unknown cause of death (from 5% in 2007 to 14.3% in 2009). This is a direct result of the inclusion of CSO registered death information in 2008 and 2009 CFRI Annual Reports. Cause of death information taken by the CSO from death certificates may on occasion record death as being due to cystic fibrosis, but information about the factors precipitating death are not always provided.

Figure 8: Principle cause of death, 2002-9, n=154



Demographics of the CFRI

Table 1 shows some summary statistics relating to PWCF alive on the last day of 2009 (n=1027). The age of PWCF ranged from less than one year to 59 years, with an average age of 18.7 years. The vast majority were of Irish origin (97.5%). Nine patients were reported as being diagnosed with CF in 2009, however this is an underestimate because many PWCF diagnosed in 2009 will be invited to enrol with the CFRI in 2010. There is a lag period from the date of diagnosis to recruitment as families are given time to come to terms with the diagnosis. The numbers diagnosed in a particular year will be more accurate once neonatal screening comes into effect.

Table 1: Demographic data from the Cystic Fibrosis Registry of Ireland (CFRI)

Year	2004	2005	2006	2007	2008	2009	Total	%
PWCF consented	159	166	53	53	303	40	1105	
Age range*						<1-59		
Mean age (yrs)*						18.7		
Median age (yrs)*						18		
Number diagnosed during year						9		
Number of males*						592		57.6%
Number of females*						435		42.4%
Number <18 yrs*						496		48.3%
Number ≥18 yrs*						531		51.7%
Number males ≥18 yrs*						312		30.4%
Number females ≥18yrs*						219		21.3%
Irish ethnicity*						1001		97.5%
Deaths during year	5	9	5	19	17	19	80	
Total PWCF on CFRI who are alive at end of year							**1027**	**92.9%**

* of the 1027 PWCF on the registry who are alive at the end of 2009

The age distribution of PWCF alive at the end of 2009 is shown by gender in Figure 8. Males outnumber females in nearly all five-year age bands and this may be due to the disproportionate number of deaths that have occurred in females in the last decade. Males and females aged 20-24 years comprised the largest age band.

Figure 9: Age and gender distribution by age band, 2009 (n=1027)

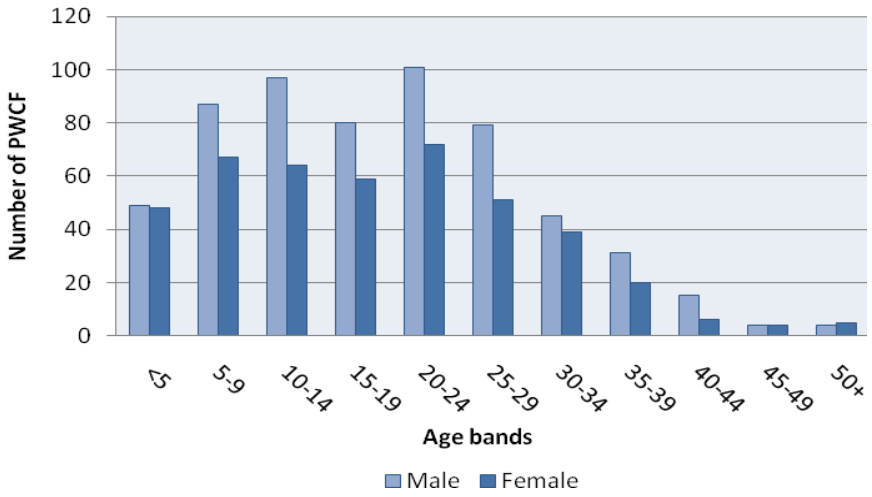


Table 2 shows that the largest proportion of PWCF live in Dublin (27.4%), followed by those living in Cork (13%) and Limerick (5.5%). The distribution of county of residence remains largely unchanged from previous years.

Table 2: PWCF by county of residence

	Number of PWCF	%
Dublin	281	27.4%
Cork	134	13.0%
Limerick	56	5.5%
Kildare	51	5.0%
Tipperary	51	5.0%
Galway	45	4.4%
Kerry	40	3.9%
Wicklow	38	3.7%
Meath	35	3.4%
Clare	34	3.3%
Mayo	31	3.0%
Wexford	23	2.2%
Waterford	21	2.0%
Cavan	20	1.9%
Louth	20	1.9%
Westmeath	20	1.9%
Kilkenny	19	1.9%
Laois	18	1.8%
Carlow	15	1.5%
Offaly	15	1.5%
Donegal	13	1.3%
Monaghan	13	1.3%
Sligo	11	1.1%
4 counties with <10 patients	23	2.3%
Total	**1004**	**100.0%**

Siblings

There were 965 families represented on the CFRI in 2009. Of those families, 835 had only one family member enrolled with the CFRI, 124 had two family members, and 7 families had three or more family members enrolled with the CFRI.

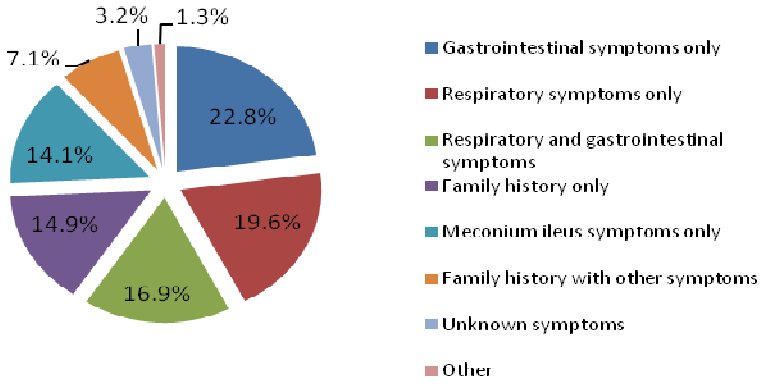
Table 3: CFRI families and siblings, 2009

Irish CF families	
1105	Total number of PWCF enrolled
965	Families represented
13.5%	130 of 965 families had 2 or more members with CF
123	Number of families with 2 members with CF
7	Number of families with 3 or more members with CF

Diagnosis

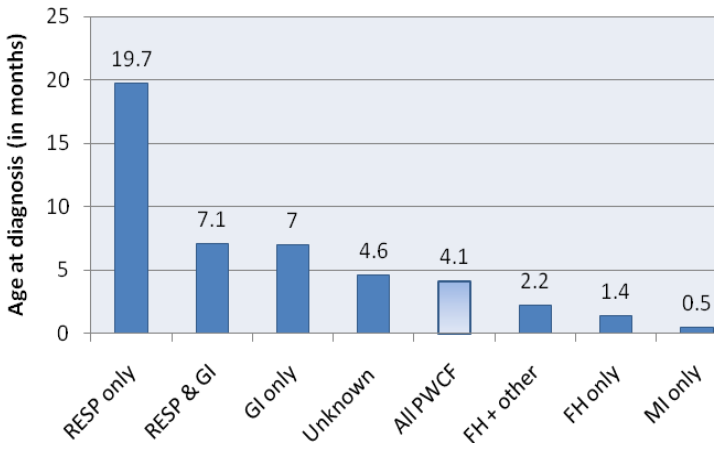
Figure 9 shows the modes of presentation for PWCF enrolled with the CFRI by 2009. As in previous reports, gastrointestinal symptoms were the most frequent symptom type reported which preceded a CF diagnosis (22.8%), followed by respiratory symptoms alone (19.6%), and both respiratory and gastrointestinal symptoms (16.9%). Approximately 15% of PWCF underwent investigation for CF due to an existing family history alone. Just over 150 PWCF presented with symptoms of meconium ileus (14.1%), with a median age at diagnosis of 0.5 months.

Figure 10: Symptoms leading to a diagnosis of CF, n=1105



The median age at diagnosis for PWCF enrolled with the CFRI by the end of 2009 was 4.1 months (range: 1 day to late forties). Diagnosis occurred earlier, on average, for PWCF presenting with symptoms of meconium ileus (0.5 months), and those with a family history with or without other symptoms (2.2 and 1.4 months respectively). PWCF presenting with respiratory symptoms took longest on average to be diagnosed with CF (median age at diagnosis: 19.7 months).

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



Genotype

Genotyping was performed for 96.9% of PWCF enrolled by the end of 2009. Delta F508 homozygous, which is one of the more severe CF genotypes, continues to be the most commonly detected (57.8%). PWCF with at least one ΔF508 alleles account for 90.1% of the CF population. Such individuals tend to have more severe disease.

Table 4: Frequencies of the most common CF mutations

	Number of PWCF	%
ΔF508 homozygous	639	57.8%
ΔF508 G551D	119	10.8%
ΔF508 R117H	38	3.4%
ΔF508 R560 T/K	24	2.2%
ΔF508 1717-1 G-->A	16	1.4%
ΔF508 621+1 G-->T	15	1.4%
ΔF508 G542X	14	1.3%
All other ΔF508 heterozygotes	110	10.0%
ΔF508 allele 2 unknown	21	1.9%
G551D G551D	8	0.7%
Other genotypes	48	4.3%
Not genotyped	34	3.1%
Pending	19	1.7%
Total	**1105**	

Hospitalisations and Complications

Annual assessment (AA) data, which refers to information on hospitalisations, number and type of exacerbations, complications, antibiotic and long-term medication usage etc., are gathered each year for each individual CFRI-enrolee. In 2009, AA data were gathered for more PWCF than ever before (n=865). AA data were gathered for a greater proportion of paediatric (91.5%) than adult (77.4%) PWCF (Table 5). However, due to difficulties experienced in accessing medical records in certain adult hospitals, partial AA data may have been gathered for individual adult PWCFs. This was unavoidable and 2009 data on hospitalizations and respiratory exacerbations in particular may be incomplete for those adults.

Table 5: Annual assessment data collected, 2009

2009 Annual Assessments (AAs)		
	Paediatric	**Adult**
PWCF with 2009 AA data collected	454	411
Proportion of live PWCF with completed 2009 AAs (N=1027)	91.5%	77.4%

Table 6 shows that respiratory exacerbations and complications occur with greater frequency in the adult population than in the paediatric population, while the number of hospitalisations on average for adult and paediatric PWCF appears to be similar. A greater number of hospitalisations, exacerbations and complications were reported in 2009 than in 2008. This is likely due to the capture of more PWCF's AA data than ever before, rather than an actual increase in frequency of observation between 2008 and 2009.

Table 6: Number of hospitalisations, exacerbations and complications, paediatric vs adult groups

2009 Annual Assessments (AAs)				
	Paediatric		**Adult**	
	Number	**Average per PWCF**	**Number**	**Average per PWCF**
Number of hospitalisations	375	0.83	338	0.82
Number of respiratory exacerbations	366	0.81	560	1.36
Number of other exacerbations	33	0.07	39	0.09
Number of complications	1206	2.66	1370	3.33

Table 7 examines CF-related complications in greater detail. Chronic *Pseudomonas aeruginosa* infection (defined as having ≥3 positive cultures in a year) was reported for 62.8% of adult PWCF for whom 2009 AA data was gathered. This is similar to the rate observed in adults in 2008 (63.2%). *Burkholderia cepacia* syndrome was observed in 12 adults in 2009, four more than in the 2008. MRSA rates remain similar to those reported in 2008, with more detected in paediatric than adult PWCF. Forty-six percent of paediatric patients had chronic *Staphylococcus aureus* infection in 2009. Reported annual rates of *S. aureus* have increased consecutively since 2007 (from 28%) and may reflect improved collection of laboratory information for paediatric patients.

The majority of adult and paediatric PWCF are pancreatic insufficient and required pancreatic enzymes in 2009. The number of paediatric PWCF with liver disease increased from 13 to 26 between 2008 and 2009, as did the number of adult PWCF with insulin-dependent diabetes (from 84 to 104 PWCF).

Osteopenia/osteoporosis was reported for 43.3% of adults compared with 5.5% of paediatric PWCF in 2009.

Table 7: Complication rates by system; paediatric vs. adult groups

2009 Annual Assessment				
	Paediatric		Adult	
No of PWCF with completed AAs	454		411	
PWCF with no complications	6		14	
Total number of complications	1206		1370	
Cardiac/Pulmonary Complications				
	Number of paediatric PWCF	**% of group**	**Number of adult PWCF**	**% of group**
Chronic *Pseudomonas*	105	23.1%	258	62.8%
Chronic *Staphylococcus*	207	45.6%	107	26.0%
Burkholderia	3	0.7%	12	2.9%
MRSA	61	13.4%	40	9.7%
Nasal polyps	16	3.5%	1	0.2%
ABPA	12	2.6%	13	3.2%
Asthma	1	0.2%	3	0.7%
All other cardiac/pulmonary complications	1	0.2%	4	1.0%
Total card/pulm complications	*406*		*434*	
Gastrointestinal Complications				
DIOS	8	1.8%	14	3.4%
Rectal Prolapse	0	0%	0	0%
Pancreatic Insufficiency	429	94.5%	341	83%
Abnormal LFTs	4	0.9%	9	2.2%
Liver disease	26	5.9%	58	11.4%
All other gastro complications	61	13.4%	44	10.7%
Total gastro complications	*528*		*458*	2.7%
Miscellaneous Complications				
Diabetes requiring insulin	13	2.9%	108	26.3%
Clubbing	182	40.1%	121	29.4%
Osteopenia/osteoporosis	25	5.5%	178	43.3%
Other morbidity	52	11.5%	67	16.3%
Total misc complications	*272*		*474*	

Cultures

CFRI staff collected data on approximately 7,100 sputum swabs taken from PWCF in 2009. With the exception of sputum and throat samples in paediatric PWCF, similar numbers of all sample types were recorded in 2008 and 2009. On average, 7.8 positive sputum cultures were reported for paediatric PWCF compared with 8.3 for adults in 2009 (Table 8). The average number of sputum culture positive samples in adults declined between 2008 (9.0) and 2009 (8.3), although this may reflects difficulties encountered with accessing medical records from certain adult hospitals for the collection of 2009 AA data.

Table 8: Culture types, paediatric vs. adults

	Paediatric		Adult	
Sample type	**Number**	**Average number of positive cultures per paediatric PWCF**	**Number**	**Average number of positive cultures per adult PWCF**
Sputum samples	3739	7.8	3364	8.3
Cough swabs	1518	3.2	125	0.3
Throat swabs	1474	3.1	49	0.1
BAL swabs	85	0.2	26	0.1
Nasal swabs	137	0.3	21	0.1

Table 9 shows the 12 most commonly detected organisms in sputum cultures from PWCF of all ages in 2009. *S. aureus*, *P. aeruginosa* and *Candida sp.* were the most frequently detected. For the first time, *S. aureus* positive sputum cultures were detected more frequently than *P. aeruginos*a (perhaps due to incomplete collection of microbiological information for adult PWCF, in whom *P. aeruginosa* is commonly detected.)

Table 9: Twelve most frequently detected organisms in sputum cultures

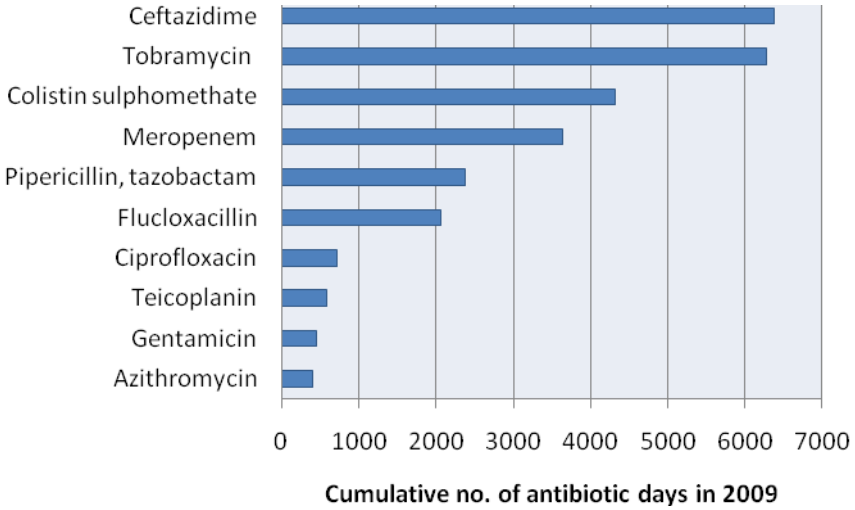
	Number of positive sputum cultures	% of positive sputum cultures
Staphylococcus aureus	1063	15.0%
All *Candida* species	1058	14.9%
***Pseudomonas aeruginosa* (Mucoid status not reported)**	1051	14.8%
***Pseudomonas aeruginosa* (Mucoid)**	728	10.2%
Normal flora	656	9.2%
Aspergillus fumagatus	415	5.8%
MRSA	329	4.6%
Gram positive cocci	310	4.4%
Haemophilius influenza	248	3.5%
***Pseudomonas aeruginosa* (Non-mucoid)**	241	3.4%
Gram negative bacilli	210	3.0%
Stenotrophomonas maltophilia	172	2.4%
Other*	622	8.8%
Total	7103	

*Contains 35 *Burkholderia cepacia* complex positive sputum cultures; 22 *cenocepacia*, 17 *multivorans*, and 1 cepacia.

Antibiotics

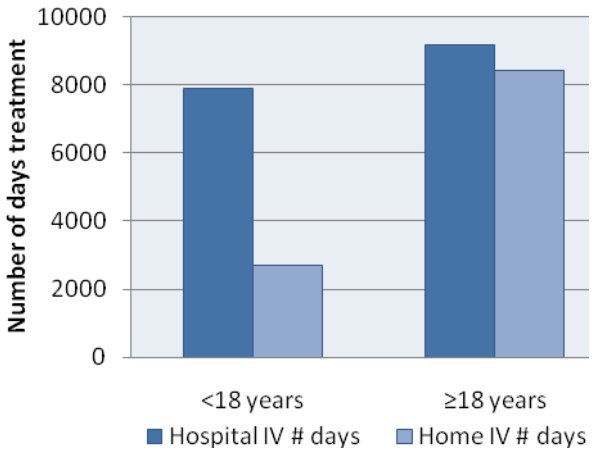
The number of days PWCF spent taking the more commonly prescribed IV antibiotics in 2009 is shown in Figure 11. The ten most commonly prescribed IV antibiotics in 2009 alone accounted for over 21,000 antibiotic days (cumulative number of days in which PWCF were administered IV antibiotics). Ceftazidime, tobramycin, colistin sulpohmethate and meropenem are the four most commonly prescribed IV antibiotics. In 2009, ceftazidime (6,386 days) and tobramycin (6,276 days) accounted for approximately 60% of the IV antibiotic days presented in Figure 12.

Figure 12: Rank of order of IV antibiotics, 2009



For paediatric PWCF, administration of antibiotic IVs in hospital is more common than at home (Figure 13). In the adult CF population, home IV administration of antibiotics occurs nearly as frequently as hospitalisation for IV antibiotic treatment.

Figure 13: Days of treatment by hospital and home IV antibiotics, adults vs. paediatrics in 2009



Pulmonary Function

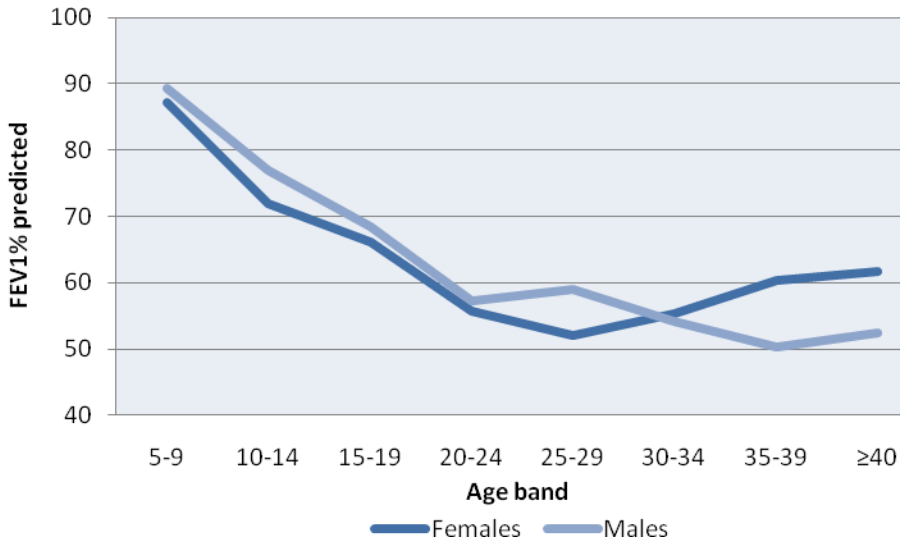
Approximately 2,500 pulmonary function test results were recorded by the CFRI in 2009. Slightly fewer test results were captured in 2009 than in 2008 for reasons of access to medical records which were previously described. Table 10 provides a snapshot of mean pulmonary function values in 2009 across all age bands from 5 years of age onwards (regardless of whether PWCF had undergone lung transplantation). Mean FEV_1% and FVC% predicted decline rapidly in the early age bands (5-19 years) and appear to stabilise from the age of twenty.

Table 10: Pulmonary function test summary, 2009

Age bands	Number of PFTs	Mean FEV_1 % predicted	Mean FVC % predicted
5-9	407	88.4	92.0
10-14	480	75.0	83.5
15-19	460	67.5	79.0
20-24	496	56.7	72.6
25-29	284	56.1	74.2
30-34	172	54.7	75.2
35-39	85	54.7	74.4
≥40	62	57.3	80.5
Total	**2446**		

Stratifying mean FEV_1% predicted data by gender we find that in 2009, values for males were higher than females between the age of 5 and 29 years. Mean FEV_1% predicted values appear to improve for females in their thirties. This may be because those with poorer FEV_1% predicted values died before reaching their thirties, so those reaching their thirties are in relatively good health and correspondingly mean FEV_1% predicted values are good. Lung transplantation may also play a role in improved pulmonary function in adulthood however, the collection of transplantation data is difficult for the CFRI to gather and is not reported on here.

Figure 14: FEV_1% predicted mean values by age band and gender, 2009



Body Mass Index

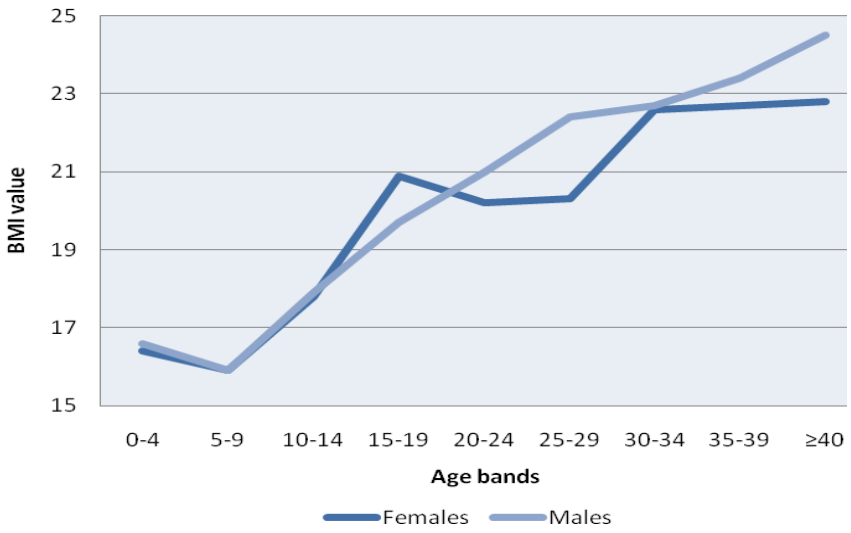
Table 11 shows mean body mass index (BMI) scores reported in 2009 by age band. Measurement of nutrition and growth using the BMI score is recommended for those aged ≥20 years, and BMI centile-for-age for those aged 2 to 19 years. However, BMI centile-for-age is not routinely recorded by the CFRI, rather height and weight measurements are recorded. The CFRI will be working to improve reporting of nutritional information for paediatric PWCF in the coming year. In the interim, BMI values are reported here for paediatric patients, although it is recognised this may not be an entirely useful measure of growth and nutrition in childhood.

Table 11: BMI summary, 2009

Age bands	N	Mean BMI
0-4	314	16.4
5-9	509	15.9
10-14	494	17.8
15-19	456	20.3
20-24	482	20.7
25-29	266	21.5
30-34	162	22.7
35-39	83	23.1
≥40	54	23.6
No of PWCF	2820	

Figure 14 shows mean BMI values in 2009 by age band and gender. Mean BMI from the age of 20 onwards is greater in males than females.

Figure 15: BMI mean values by age band and gender, 2009



Nutrition

Table 12 provides information about nutrition and feeding for paediatric and adult PWCF in 2009. The majority were prescribed vitamins and calorie supplements were prescribed for one in five PWCF. Compared with 2008, more paediatric and adult PWCF reported supplemental feeding in 2009 (20% compared with approximately 30% in 2009).

Table 12: Nutrition summary, 2009

2009 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		411	
	Number of paediatric PWCF	**% of group**	**Number of adult PWCF**	**% of group**
Calorie supplements	92	20.3%	84	20.4%
Vitamins	429	94.5%	330	80.3%
Minerals	29	6.4%	44	10.7%
Gastrostomy feeds	7	1.5%	29	7.1%
Supplemental feeding	141	31.1%	122	29.7%
Oral supplements	100	22%	80	19.5%
Nasogastric	5	1.1%	2	0.5%

Physiotherapy

Table 13 shows that 80% of paediatric PWCF were reviewed by a physiotherapist in 2009. Sixty-three percent of adults were reviewed by a physiotherapist in 2009, which is an improvement on 2008 (45.6%). While the 2009 figure for adults is likely to be an underestimate, the capture of physiotherapy information for adults in particular appears to be improving.

For paediatric PWCF, use of a PEP mask was most frequently reported in 2008 (28.5%) and 2009 (31.9%). Acapella (21.4%) and percussion (14.1%) were the next most commonly reported physiotherapy manoeuvres in 2009. For adults, autogenic drainage was used by one quarter of PWCF and was the most common manoeuvre in 2009, followed by use of the PEP mask (17.8%). By contrast, the PEP mask was the most common physiotherapy modality used in 2008 (12.5%). The discrepancy between the most common modality reported in 2008 and 2009 may be due to the small amount of physiotherapy data recorded in hospital medical charts for adult PWCF in 2008.

Table 13: Physiotherapy summary, 2009

2009 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		411	
PWCF seen by Physio at AA	363		257	
% seen by Physio	80.0%		62.5%	
	Number of paediatric PWCF	**% of group**	**Number of adult PWCF**	**% of group**
PEP Mask	145	31.9%	73	17.8%
Acapella	97	21.4%	37	9.0%
Postural drainage	22	4.8%	0	-
Percussion	64	14.1%	1	0.2%
Active cycle breathing	21	4.6%	22	5.4%
Autogenic drainage	22	4.8%	103	25.1%
Flutter	19	4.2%	16	3.9%

Long-Term Medications

Table 14 shows the long-term medications used by PWCF in 2009. Beta-agonists are the most frequently reported long-term medication in both paediatric (46.7%) and adult (54.0%) PWCF. Osteoporosis treatment (47.9%) and inhaled steroids (46.2%) were the second and third most common medications used by adults in 2009. RhDNAse and Inhaled steroids were ranked second and third most common medications for paediatric PWCF.

The number of paediatric PWCF receiving osteoporosis treatment doubled (from 38 to 76) between 2008 and 2009, but may reflect improvements in the capture of such information. Fewer paediatric PWCF were reported as using urso-deoxycholic acid in 2009 (n=92) compared to 2008 (n=55). An increase in use of nebulised saline was observed in 2009 (8.4% of paediatric and 9.7% of adult PWCF). For adult PWCF, slightly more adults were receiving insulin in 2009 (n=91) than in 2008 (n=63).

Table 14: Long-term medication summary, 2009

2009 Annual Assessment				
	Paediatric		Adult	
No of PWCF	454		411	
	Number of paediatric PWCF	**% of group**	**Number of adult PWCF**	**% of group**
Beta agonist	212	46.7%	222	54.0%
Inhaled steroid	149	32.8%	190	46.2%
rhDNAse	167	36.8%	157	38.2%
Urso-deoxycholic acid	92	20.3%	66	16.1%
Osteoporosis treatment	76	16.7%	197	47.9%
H2RA/PPI	124	27.3%	164	39.9%
Anti-cholinergic	28	6.2%	20	4.9%
Lactulose	19	4.2%	13	3.2%
Oral steroid	22	4.8%	19	4.6%
Insulin	11	2.4%	91	22.1%
Night-time oxygen	10	2.2%	17	4.1%
Nebulised saline	38	8.4%	40	9.7%
Aminophylline/theophylline	0	-	3	0.7%

Financial Information

The financial summary in Table 15 lists the Income and Expenses for the CFRI in 2008 and 2009.

The following points should be noted:

This is the first full financial year which CFRI has reported on since our change in governance to an independent organisation.

The primary source of income for CFRI is the Department of Health & Children; through the Health Service Executive in the form of an annual grant. The amount of this grant for the past two years was €272,000. This was drastically cut back to €132,000 in 2009 and has put CFRI under severe financial strain. This cutback in core funding has to be addressed as a matter of urgency.

A deficit of €78,746 is reported for 2009 and this has to be addressed during 2010.

Table 15: Income & Expenses for 2009

Income & Expenses	2009 €	2008 €
Income		
Grant income	132,000	272,000
Sundry income	134	1,118
Bank deposit interest	-	17
Total income	132,134	273,135
Expenses		
Wages & salary	142,448	149,049
Employer's PRSI	15,313	16,023
Rent payable	4,830	4,830
Insurance	489	485
Computer network & server costs	4,151	3,153
Database costs	30,910	12,520
Heat & light	662	662
Repairs & maintenance	-	431
Printing, postage and stationery	4,024	13,086
Computing	383	-
Travelling & subsistence	4,701	13,547
Audit	968	979
Bank charges	136	142
Sundry expenses	346	554
Depreciation on equipment	1,519	1,112
Pre entry expenses		(30,200)
Total expenses	210,880	186,373
(Deficit)/Surplus	(78,746)	86,762

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 Central Statistics Office in Cork for providing information on CF decedents, and the National Centre for Medical Genetics in Our Lady's Children's Hospital in Crumlin for providing CF genotyping information.

Publications from CFRI, 2009

Jackson AD, Daly L, Jackson AL, Kelleher C, Harrington M, Zhou S, Foley L, Fitzpatrick P. 'Using new techniques to estimate cystic fibrosis survival in the Republic of Ireland' *Royal College of Physicians Faculty of Public Health Medicine Ireland Winter Meeting*, 2009, Irish Journal of Medical Science, 179(Suppl 11), pS445.

"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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CFRI
The Cystic Fibrosis Registry of Ireland