



Cystic Fibrosis Registry of Ireland

2015 Annual Report


Preface

2015 was the year we began the migration from our old technological platform to our new platform. This was an essential move and we are fortunate that we were able to raise the necessary funds from generous donations from the Lotto Fund and from corporate donations. Thank you to everyone who supported this project as without your assistance we would not have had the correct tools to support both external and internal cystic fibrosis research. We also moved from annual assessment reporting to an encounter based reporting system. An encounter based system is more useful in the clinical environment and we are pleased that the lung transplant unit at the Mater Hospital are now entering their own data. As our technology continues to evolve we would hope that more centres will be able to enter their own data.

Our new technology was one of the pieces of the puzzle that contributed towards CFRI winning an open competition for a thirty-month research grant from the VIA programme, which was signed on 15th September 2015. The aim of our study is to examine the effect of patient access to a CF Registry Patient Portal on a range of clinical outcomes, health service usage, health literacy, patient reported outcomes, and costs of care. This study will include numerous Irish sites as well as two European CF centres in Denmark and Slovenia. This was made possible by the fact that we all share a common registry technology platform at a European level. This is the start of a new era where CFRI can play a proactive role in pan-European and international CF research without impinging upon our domestic commitments. This project has allowed us to add an experienced statistician to the CFRI team. This role is essential going forward to analyse and quality control our wealth of information as well as offering contracted statistical services to our CF researchers. This will also assist us in getting more of our internal research work published and presented at international meetings.

CFRI continues to play an active role as a member of the Working Group of the National Clinical Programme for Cystic Fibrosis and will be co-chairing the Data Sub Committee.

We look forward to 2016 and to play our part in improving outcomes for people with cystic fibrosis.



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Summary registry data, 2009-2015

The table below provides general information about people with cystic fibrosis (CF) in the Republic of Ireland in 2015.

Table 1: Summary of the CF Registry of Ireland, 2011-2015

	2011	2012	2013	2014	2015
CFRI-registered individuals¹	1025	1098	1137	1176	1,219
Children (<18 years)	501	535	534	540	558
%	(48.9%)	(48.7%)	(47%)	(45.9%)	(45.8%)
Adults (≥18 years)	524	563	603	636	661
%	(51.1%)	(51.3%)	(53%)	(54.1%)	(54.2%)
Males	597	639	664	682	698
%	(58.2%)	(58.2%)	(58.4%)	(58%)	(57.3%)
Females	428	459	473	494	521
%	(41.8%)	(41.8%)	(41.6%)	(42%)	(42.7%)
Median age (years)	18	18	19	19	19
(IQR)²	(9-27)	(9-28)	(10-29)	(10-29)	(10-30)
Newly diagnosed persons³	45	47	43	31	15
Detected by newborn screening⁴	14	27	34	28	12
Number of deaths	26	18	8	20	15

¹Calculated from all CFRI-registered individuals alive on the last day of each reporting year.

²Interquartile range (IQR) is a measure of the spread of data. It shows the mid-spread or middle fifty percent of the data, or the difference between the upper (Q3) and lower quartiles (Q1).

³Calculated from all CFRI-registrants (alive and dead) on the database. Some diagnosis data are added after the reporting year has ended, therefore figures from previous years have been updated.

⁴The number of patients born in the year that were diagnosed with CF as a result of the National Newborn Bloodspot Screening Programme (commenced July 2011).

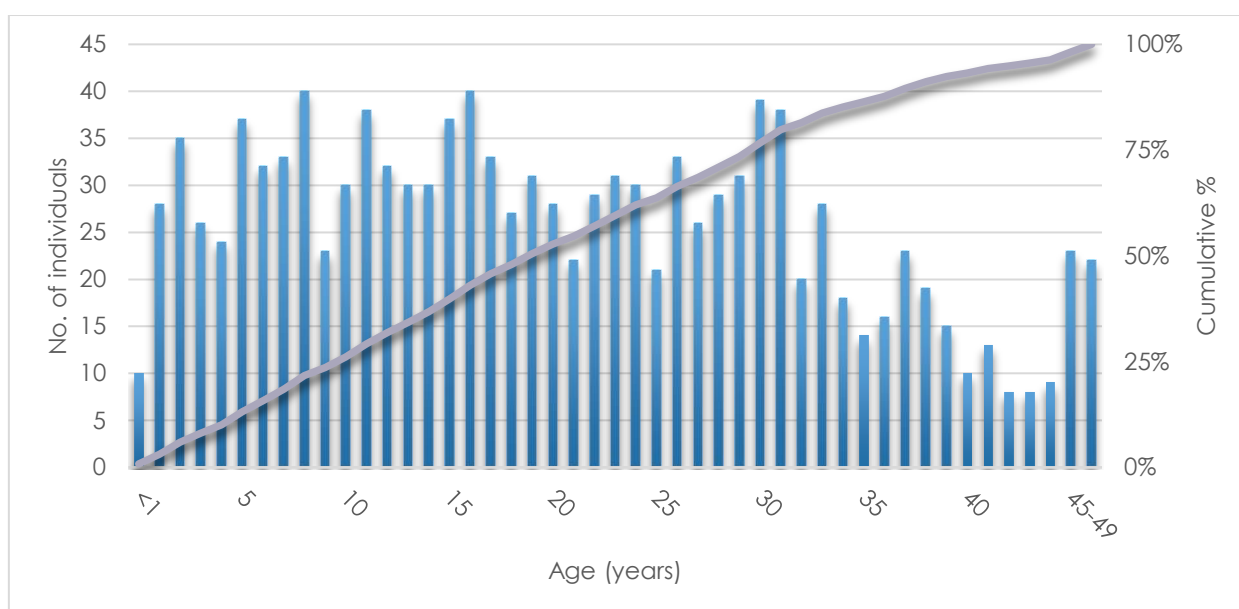
Demography

The CF Registry of Ireland (the 'registry') has gathered information on people with CF since 2002. This section provides information on the age, gender, and geographic distribution of people living with CF in 2015.

The number of people living with CF increases annually. The number of adults with CF outnumber children with CF. In 2015, 1,219 people were in the registry; 558 children and 661 adults.

In 2015, the median age of a person with CF was 19 years. In other words, half of the 1,219 individuals were under 19 years of age, and the other half were older than 19 years. The age range is from birth to over 80 years, however the majority of people with CF are in the younger age groups.

Figure 1: Age distribution of people living with CF in 2015



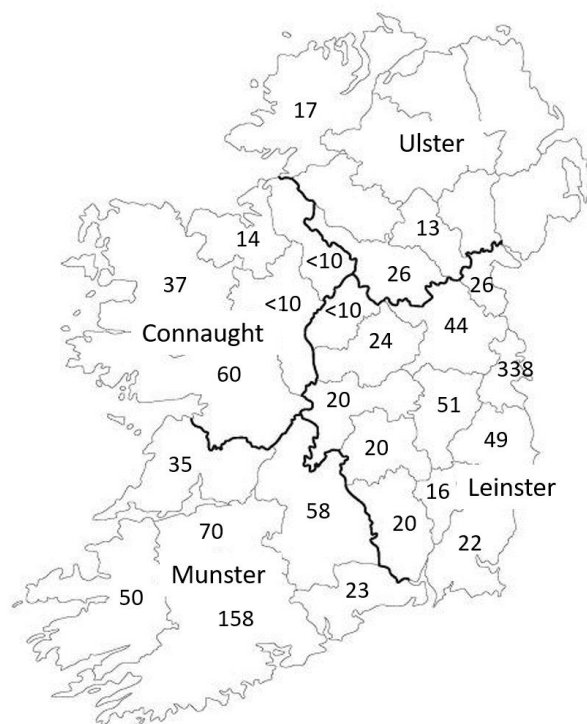
Individuals with CF are distributed across the Republic of Ireland. Half were located in Leinster, a third in Munster, and the remainder (15%) in Ulster and Connaught.

Table 2: Distribution of people living with CF by province in 2015

	Number	%
Connaught	127	10.4
Leinster	639	52.4
Munster	394	32.3
Ulster	57	4.7
Unknown	2	<1%

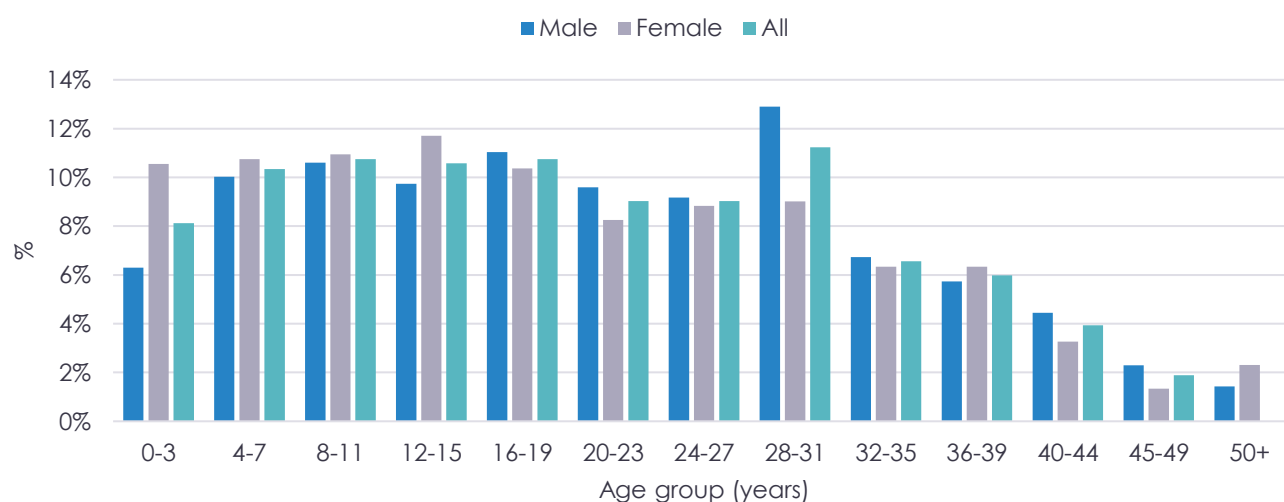
The counties with the largest numbers of individuals include Dublin (n=338), Cork (n=158), Limerick (n=70), Galway (n=60) and Tipperary (n=58). These figures reflect the county of residence of people with CF on the registry.

Figure 2: Distribution of people living with CF by county in 2015



As reported in other years, and as found in other countries, the gender mix in the Irish CF population in 2015 was imbalanced (females: 43%, males: 57%). This is likely due to historically poorer survival in females with CF. Median age for females was 18 years (IQR 8-29) and 20 years for males (IQR 11-30) (independent samples median test of age in 2015 found this difference was statistically significant: $p < 0.026$).

Figure 3: Age distribution by gender of people living with CF in 2015



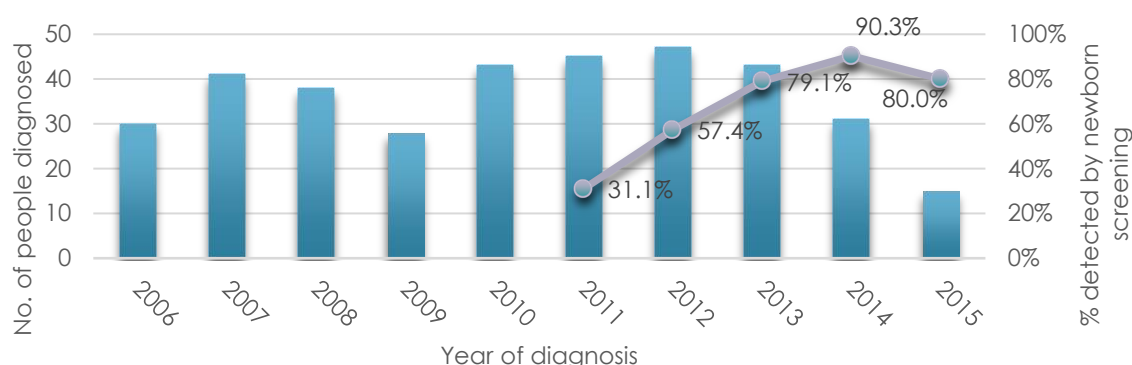
Diagnosis

Early diagnosis of CF provides opportunities for earlier medical intervention. Providing infants with the best possible care may result in better nutritional and lung function outcomes later in life. In this section, we examine how and when patients are being diagnosed with CF in 2015, and how these trends compare to previous years.

In the ten years between 2006 and 2015, an average of 36 individuals were diagnosed with CF annually (range 15-47). The number of people reported here as being diagnosed in 2015 (n=15) and 2014 (n=31) is an underestimate, which results from a delay in joining the registry. Future reports are adjusted to include new registry participants.

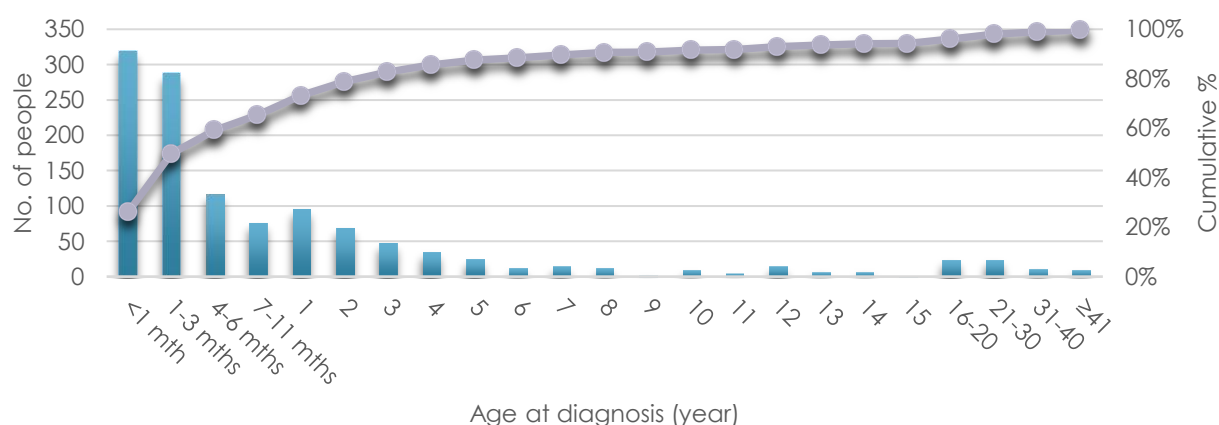
Since the introduction of newborn screening for CF in July 2011, the proportion of new CF diagnoses made as a result of newborn screening has gradually increased. In 2015, 12 of the 15 new CF diagnoses (80%) were made following newborn screening.

Figure 4: Number of new CF diagnoses, 2006-2015



The majority of people living with CF in 2015 were diagnosed following symptomatic presentation to medical services. One quarter of individuals were diagnosed by 1 month of age, another quarter by 6 months, and another quarter by one year of age. The remainder were diagnosed at one year or older.

Figure 5: Age at diagnosis of people living with CF in 2015



The introduction of newborn screening has resulted in earlier diagnosis of people with CF. In 2015, all one year olds had received their CF diagnosis by 3 months of age. Compare this with all those aged ten years in 2015; less than half (46.7%) had been diagnosed by 3 months.

Table 3: Age at diagnosis of people with CF aged one year and ten years in 2015

Age at diagnosis	All ages in 2015		1 year	10 years
	N=1,219	%	%	%
<1 month	319	26.2%	96.4%	30.0%
1-3 months	287	23.5%	3.6%	16.7%
4-6 months	116	9.5%	-	10.0%
7-11 months	76	6.2%	-	10.0%
1 year	95	7.8%	-	10.0%
2 years	69	5.7%	-	6.7%
3 years	47	3.9%	-	10.0%
4 years	34	2.8%	-	3.3%
≥5 years	172	14.1%	-	3.3%
Unknown	4	0.3%	-	

Of those living with CF in 2015, 37.7% experienced respiratory symptoms prior to diagnosis, 24.2% failed to thrive due to malnutrition in infancy and 21.9% reported having a family member with a CF diagnosis.

All children aged one year in 2015 were detected as a result of newborn screening, whereas one in three 10 year olds reported failure to thrive/malnutrition and almost half experienced respiratory symptoms.

Table 4: Modes of diagnosis of people with CF aged one year and ten years in 2015

	All ages in 2015		Aged 1	Aged 10
	N=1,219	%	%	%
Meconium ileus:				
treated surgically	99	8.1%	14.3%	10%
medically managed	64	5.3%	-	10%
management unknown	13	1.1%	-	-
Other neonatal bowel/intestinal obstruction	13	1.1%	-	-
Steatorrhea +/- abnormal stools +/- malnutrition	265	21.7%	-	30%
Failure to thrive/malnutrition	295	24.2%	-	33.3%
Rectal prolapse	27	2.2%	-	-
Respiratory	460	37.7%	-	46.7%
Sinus disease/nasal polyps	16	1.3%	-	3.33%
Newborn screening (in Ireland/other country)	144	11.8%	100%	13.3%
Family history	267	21.9%	10.7%	20%
Other	108	8.9%	-	6.7%

*Diagnostic categories are not mutually exclusive, i.e. multiple diagnosis categories may apply to an individual.

The National Newborn Bloodspot Screening Programme (NNBSP) reported that 21 newborns in 2015 were diagnosed with CF (12 registered with the CF Registry by the end of 2015). The statistics from the NNBSP are as follows:

Table 5: National Newborn Bloodspot Screening Programme outcomes, 2015

	N (%)
Number of newborns* screened in 2015	65,802
Number of samples with raised IRT sent to National Centre for Medical Genetics (NCMG)	784 (1.19%)
Number of newborns with one mutation identified	60
Number of newborns with two mutations identified	17
Number of newborns referred to a CF Specialist Centre	77
Number of initial sweat tests performed	75
Number of sweat test failures	17
Number of newborns with positive sweat test	13
Number of newborns with negative sweat test	40
Number of newborns with borderline sweat test	5
Number of newborns requiring repeat sweat test	4
Number of newborns diagnosed with CF	21
Number of newborns diagnosed with variant CF	1
Number of newborns diagnosed as carriers	54
Number of newborns without a definitive diagnosis**	1

*Figures reported here are based on baby's date of birth. **Remains under the care of a CF Consultant for further investigation.

Since the introduction of the NNBSP in July 2011, 131 individuals have been diagnosed as a result of the National Newborn Bloodspot Screening Programme.

Table 6: Summary of National Newborn Screening Programme outcomes, July 2011-2015

Number of screened newborns diagnosed with CF	
2011 (Jul-Dec)	16
2012	28
2013	34
2014	32
2015	21
Total	131

*Source: Prof P Mayne, Director of the National Newborn Bloodspot Screening Laboratory, Temple Street.

CFTR gene mutations

CF is a genetic condition, and people with CF inherit one copy of a faulty CF gene from each parent. About 2,000 CFTR gene mutations have been identified to date. Genetic testing provides important information to assist the diagnosis and treatment of CF. This section examines the most common CF causing CF mutations.

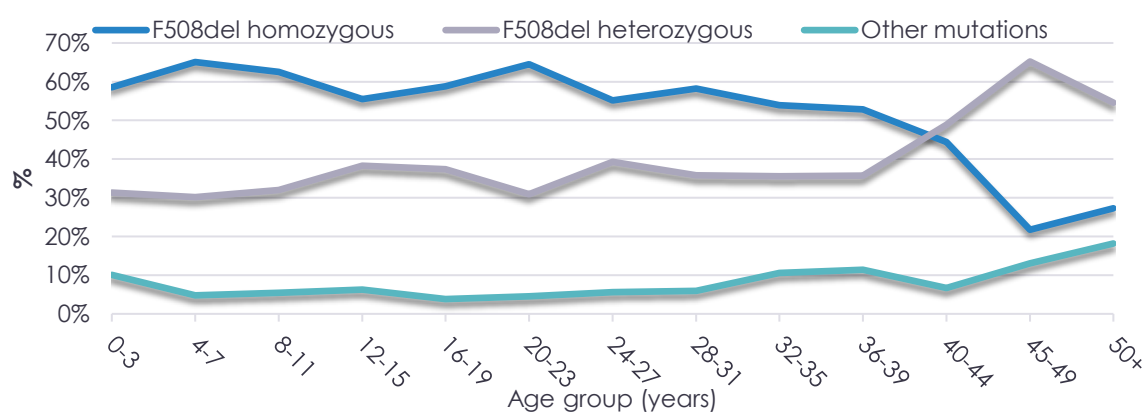
F508del is the most commonly detected CFTR mutation in people living with CF in 2015; 91.7% have at least one copy, 14.8% have at least one copy of G551D, and 5.2% have at least one copy of R117H. All other CFTR mutations affect less than 5% of the CF population. CFTR allele frequencies are shown below.

Table 7: CFTR allele frequency, 2015

Legacy name	cDNA name	Protein name	% Allele frequency N=2,362
F508del	.1521_1523delCTT	p.Phe508del	76.2%
G551D	c.1652G>A	p.Gly551Asp	8.3%
R117H	-	p.Arg117His	2.8%
R560T	c.1679G>C	p.Arg560Thr	1.7%
621+1G->T	c.489+1G>T	-	1.1%
1717-1G->A	c.1585-1G>A	-	1%
G542X	c.1624G>T	p.Gly542X	0.8%
V520F	c.1558G>T	p.Val520Phe	0.5%
I507del	c.1519_1521delATC	p.Ile507del	0.4%
Other			7.2%
Total			100%

Fifty-six percent of patients living with CF in 2015 had two copies of the F508del mutation (F508del homozygous). This compares with 46.1% in the United States and 50.3% in the United Kingdom. Thirty-six percent (35.7%) of people had just one copy of the F508del mutation, (F508del heterozygous). The F508del homozygous mutation is the most common CFTR gene mutation in people under 40 years. In those over 40, F508del heterozygous mutations are most common.

Figure 6: Prevalence of F508del mutations by age, 2015



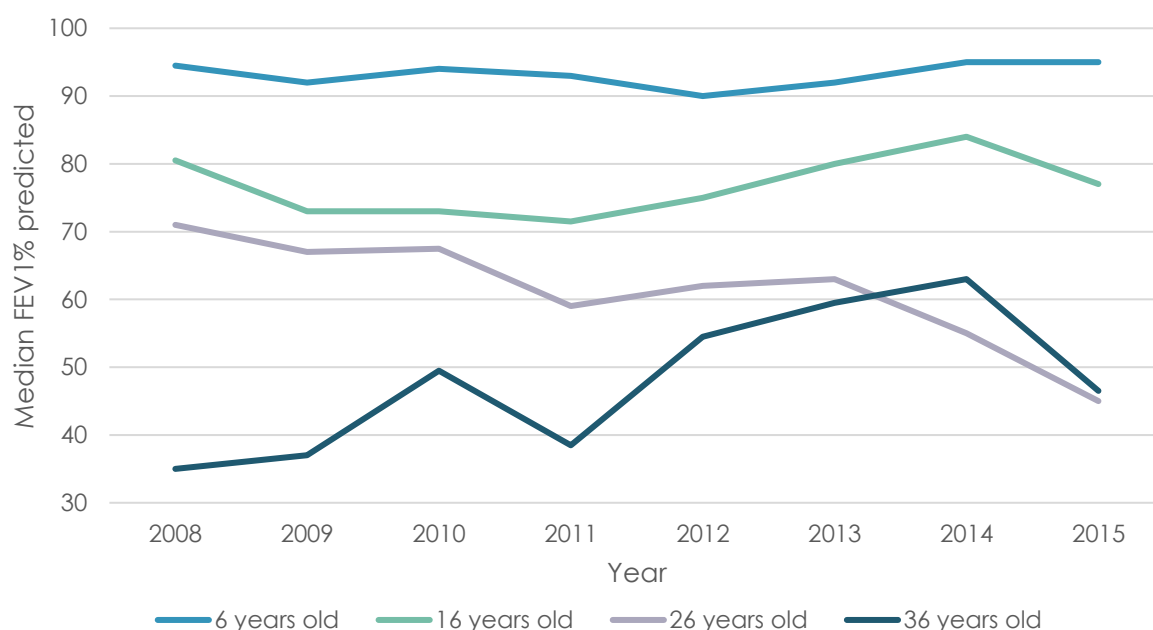
Lung function

The health of the lungs is an important indicator of the overall condition of a person with CF. It is measured using FEV1; the Forced Expiratory Volume of air in the first second of an exhaled breath. Although this is measured in litres, it is expressed as a percentage of the expected value from people without cystic fibrosis of the same age, gender, height, and ethnicity (hence percent predicted). An FEV1% predicted (FEV1pp) of 100 indicates that the lung function is the same as the mean lung function of people without CF.

The annual median FEV1 percent predicted (FEV1pp) estimated using all CFRI-recorded, centre-reported FEV1pp values in 2015 was 72% (IQR: 50-91%). By comparison, the annual median FEV1% predicted between 2008 and 2014 ranged from 70 to 75%.

Median FEV1pp estimated in children aged 6 years in 2015 remains largely unchanged from previous years, with near normal lung health. Overall, median FEV1pp in 16 and 36 year olds improved since 2008. Year-on-year, median FEV1pp in those aged 26 years has declined, possibly reflect the fact that patients are now living longer with a lower level of lung function than ever before.

Figure 7: Median FEV1pp at age 6, 16, 26 and 36 years, 2008-2015



To examine the profile of lung health in 2015, the best FEV1 percent predicted (FEV1pp) recorded for each person aged 6 years and older was used (patients who received a lung transplant by the end of 2015 were excluded). Nearly 90% of children had either normal lung health or mild lung disease, compared with 48.3% of adults. Fifteen percent of adults had severe lung disease.

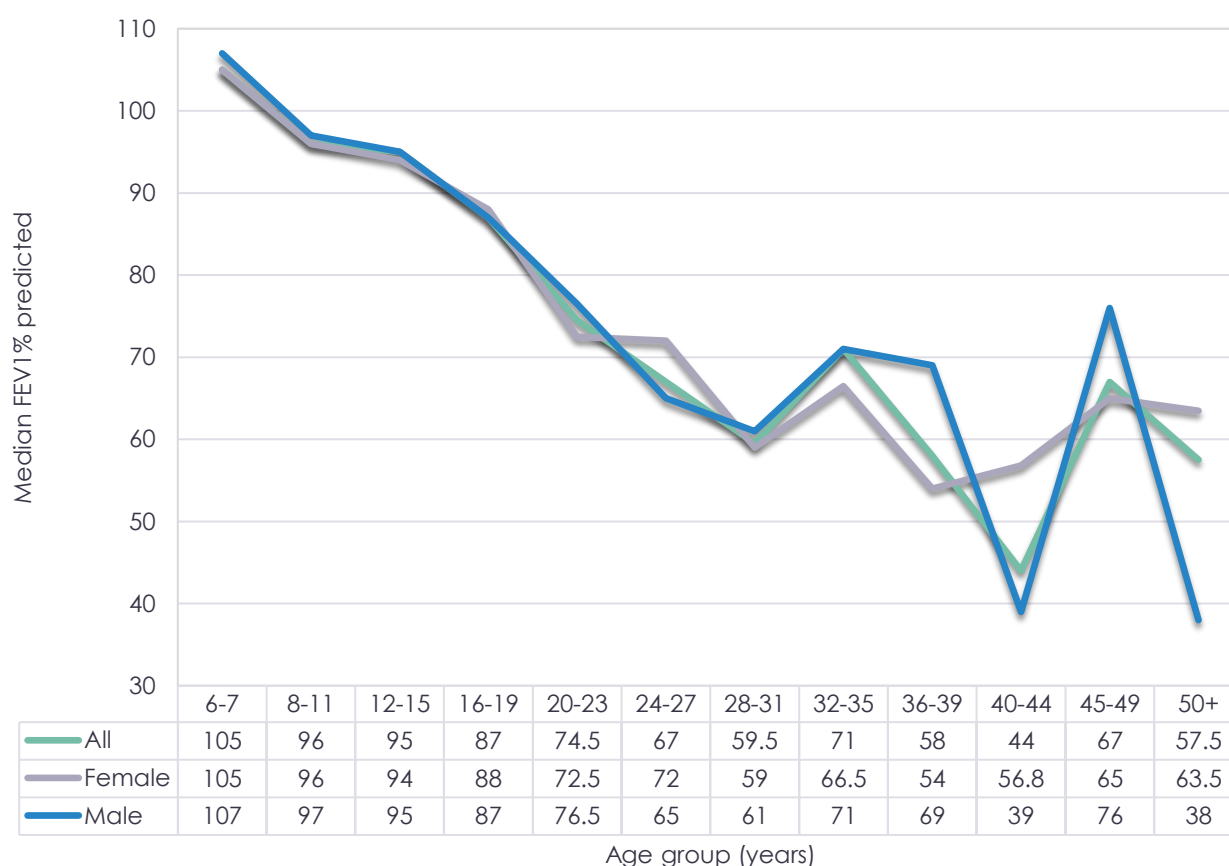
Table 8: Severity of lung disease, 2015

	Severe FEV1pp <40%		Moderate FEV1pp 40-69%		Mild/normal FEV1pp ≥70%	
Children (6-17 years)	<10	<1%	36	9.8%	328	89.4%
Adult (≥18 years)	73	15.4%	172	36.3%	229	48.3%
All	76	9.0%	208	24.7%	557	66.2%

In 2015, the median FEV1% predicted estimated using patient's best FEV1% in the year was 83% (IQR: 55-99). For females, median FEV1% predicted was 84% (IQR: 59-100%) and 82% (IQR: 52-99%) for males.

Median FEV1pp values were similar in males and females between the ages of 6 and 23 years. Males and females in their thirties had a higher median FEV1pp values than those in aged in their late twenties in 2015. Higher median FEV1pp values in males and females in their forties is likely due to the small number of patients (less than 50), many with a mild CFTR mutation, who have milder lung disease.

Figure 8: Median FEV1pp* by gender, 2015



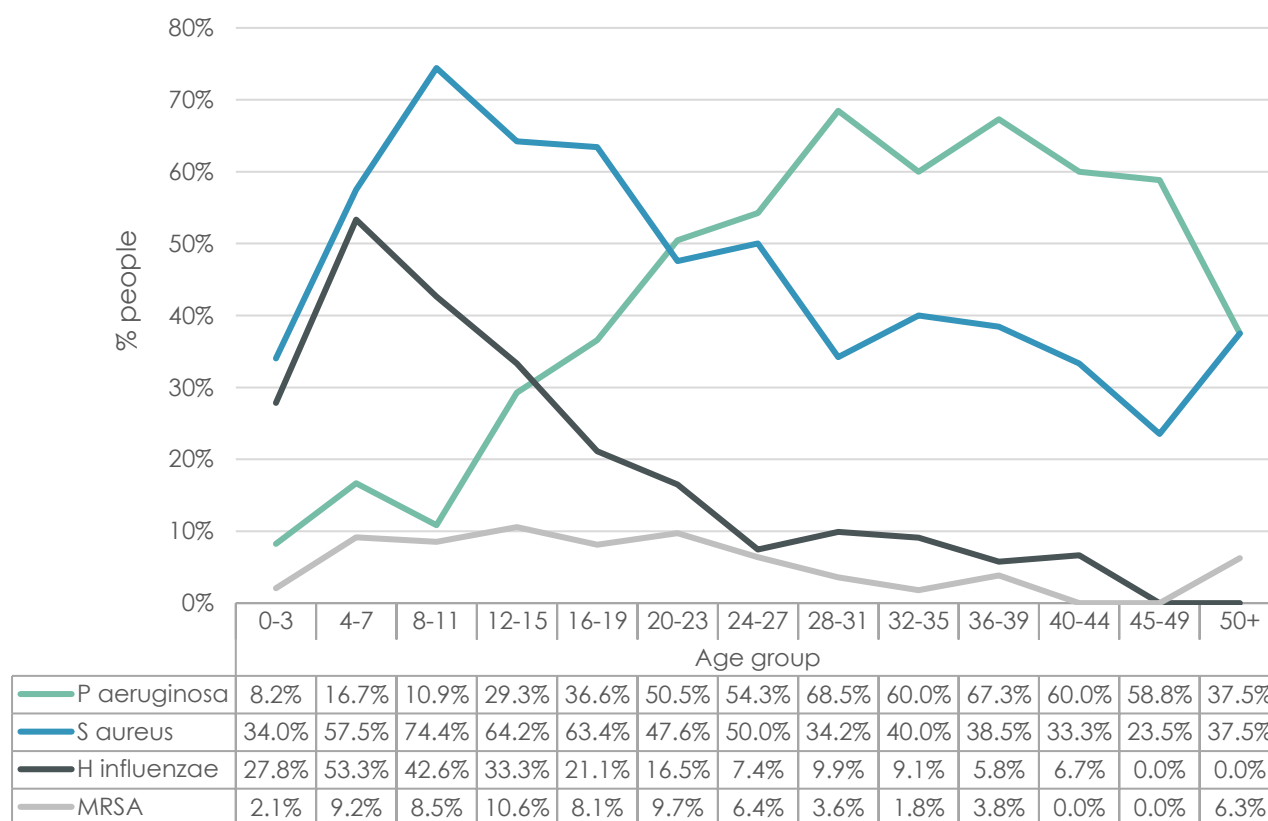
*People who received a lung transplant by the end of 2015 were excluded.

Individuals with CF may develop lung infections because bacteria may not be cleared easily from the thickened mucus of the lung. Infections can result in reduced lung function.

Infections commonly detected in people with CF are shown below. In children, *Staphylococcus aureus* (59.7%) and *Haemophilus influenza* (37.6%) were the most prevalent organism cultured from respiratory specimens. In adults, *Pseudomonas aeruginosa* (57.9%) was most prevalent.

Twenty-six people had a reported *Burkholderia cepacia* complex infection in 2015; 17 of which were confirmed as being genomovar II *B. multivorans*. Twenty-six patients had non-tubercular mycobacterium infections (*M. abscessus*, *M. avium* complex, *M. chelonae*) in 2015, compared with 11 in 2014.

Figure 9: Prevalence of *P aeruginosa*, *S aureus*, *H influenzae* and MRSA by age group, 2015



Lung infections caused by *Pseudomonas aeruginosa* and *Staphylococcus aureus* can become 'chronic', which means they can't be removed from the lung and can cause an irreversible reduction in lung function. In this report, chronic status is defined as three or more positive reports in a 12-month period.

In 2015, 236 adults (44.4%) and 59 children (11.0%) had chronic *P. aeruginosa* infection. Chronic *S. aureus* infection was reported in 138 adults (25.9%) and 242 children (45.0%).

Figure 10: Chronic *P. aeruginosa* and *S. aureus* infection, 2015

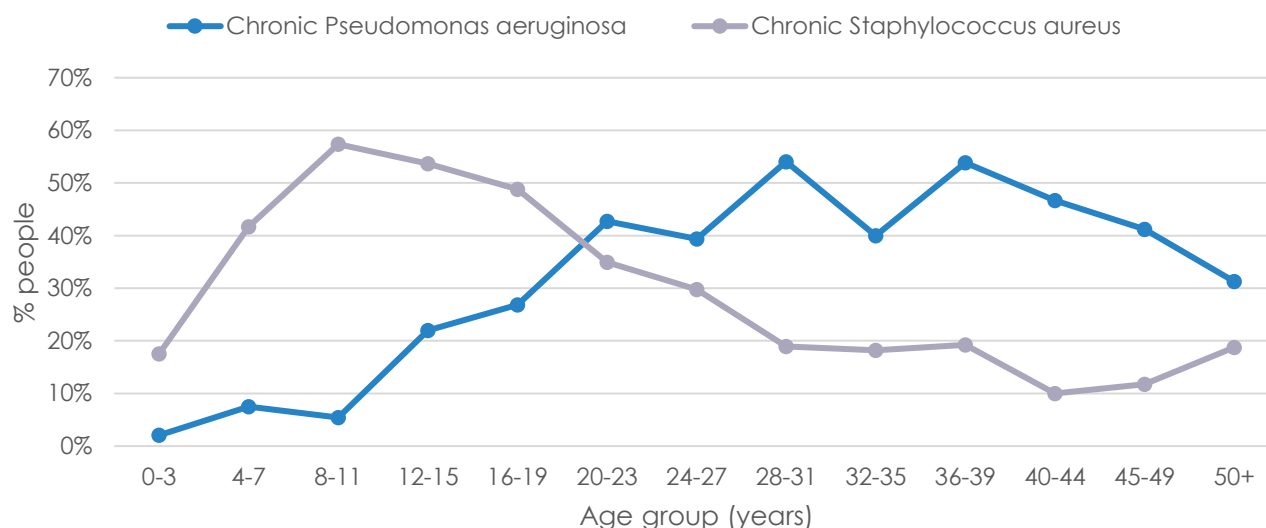
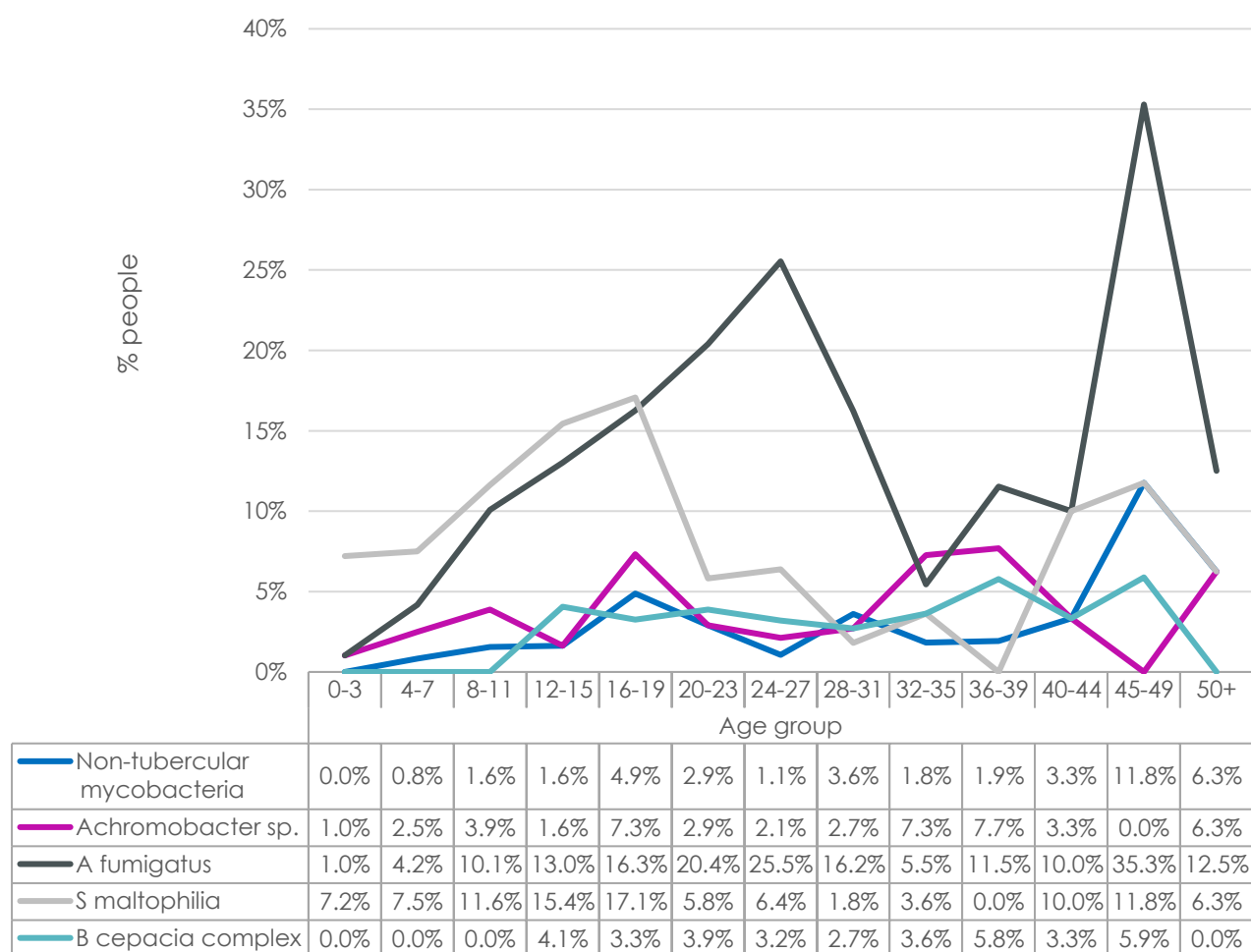


Figure 11: Prevalence of non-tubercular mycobacteria, *Achromobacter* species, *A. fumigatus*, *S. maltophilia* and *B. cepacia* complex by age group, 2015

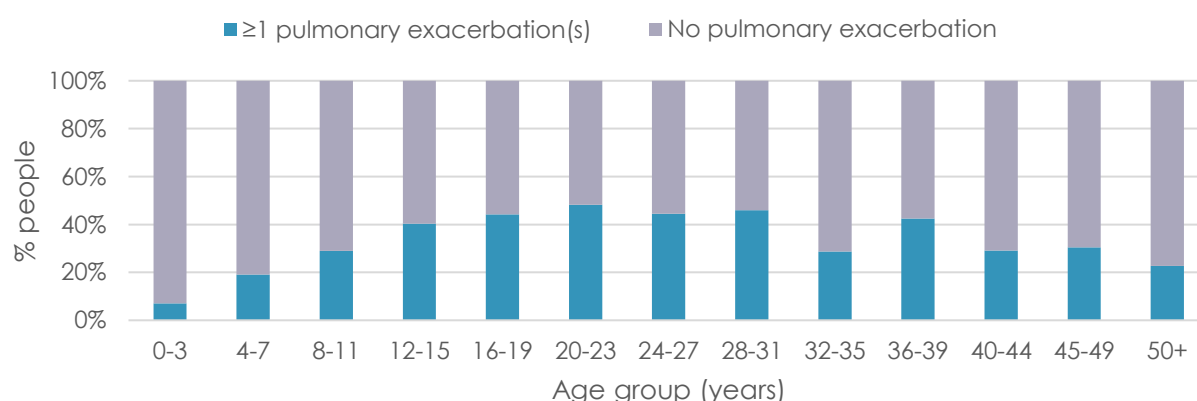


Pulmonary exacerbations

A pulmonary exacerbation (PE_x) is the development of a new symptom(s) or worsening of existing symptoms, which requires the treatment of intravenous (IV) antibiotics at home or in hospital.

In 2015, 424 (34.8%) people had at least one pulmonary exacerbation, for which they received IV antibiotics; 27.2% of children (n=152) and 41.1% of adults (n=272).

Figure 12: Pulmonary exacerbations, 2015



Of those who experienced pulmonary exacerbation(s) in 2015, 55.7% (n=236) had two or more.

Table 9: Recurrence of pulmonary exacerbations, 2015

Courses of IV antibiotics	Children N=558	Adults N=661	All N=1,219
0	72.8%	58.8%	65.2%
1	14.5%	16.2%	15.4%
2	4.1%	11.8%	8.3%
≥3	8.6%	13.2%	11.1%

In 2015, the number of days spent on IV antibiotics for a pulmonary exacerbation(s) summed for all people with CF came to a total of 15,116. The average (mean) duration of pulmonary exacerbation treatment was 35.7 days (SD ±32.5) and median duration was 29 days (IQR: 15-45 days).

Table 10: Duration of pulmonary exacerbation (PE_x), 2015

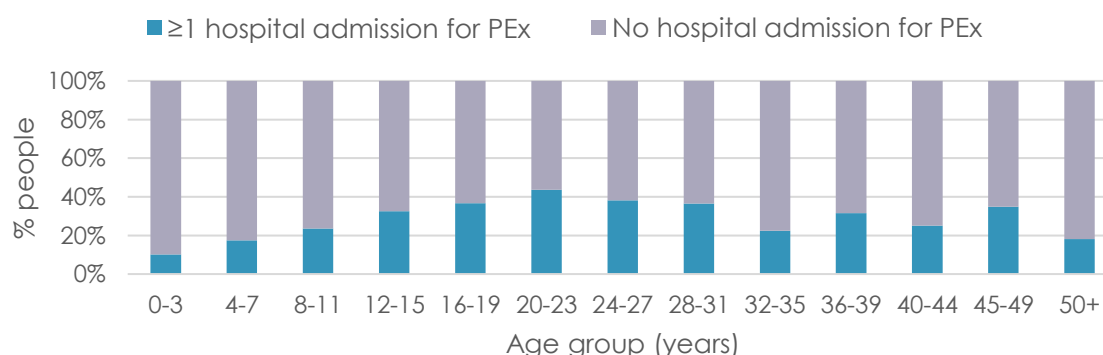
	Children N=152	Adults N=272	All N=424
Cumulative days on IV antibiotics	5,024	10,092	15,116
Mean duration of PE _x treatment (SD)	33.1 (±30.6)	37.1 (±33.5)	35.7 (±32.5)
Median duration of PE _x treatment (IQR)	19.5 (15-44)	30 (15-46)	29 (15-45)

Hospitalisations

Hospitalisations reported here reflect admission to a hospital for treatment of pulmonary exacerbation.¹

Nearly 30% (n=358) of people were hospitalised for a pulmonary exacerbation (PE_x) in 2015; 23.1% of children (n=129) and 34.6% of adults (n=229). While 424 people received treatment for PE_x in 2015, treatment can be delivered at home and/or hospital. For this reason, more people are treated than are hospitalised for PE_x.

Table 11: Hospitalisation for treatment of pulmonary exacerbation (PE_x), 2015



Of the 358 people who were hospitalised in 2015, 47.2% (n=169) were admitted to hospital for a pulmonary exacerbation two or more times in the year.

Table 12: Frequency of hospitalisation for pulmonary exacerbation, 2015

Number of hospitalisations	Children N=558	Adults N=661	All N=1,219
0	76.9%	65.4%	70.6%
1	13.4%	17.2%	15.5%
2	4.5%	8.6%	6.7%
≥3	5.2%	8.8%	7.1%

In 2015, the number of days spent in hospital for treatment of a pulmonary exacerbation(s) summed for all people with CF came to a total of 9,716 days. The average (mean) length of stay was 27.1 days (SD ±25.8) and median was 17 days (IQR: 11-36.25).

Table 13: Duration of hospital stay for pulmonary exacerbation treatment

	Children N=558	Adults N=661	All N=1,219
Cumulative days in hospital	2,665	7,051	9,716
Mean duration of hospitalisation (SD)	20.7 (±22.6)	30.8 (±26.8)	27.1 (±25.8)
Median (IQR)	15 (7.5-24.5)	22(14.5-42)	17 (11-36.3)

¹ Elective admission to hospital on a 3 monthly basis for IV antibiotics are excluded.

Ambulatory day care

In 2015, the registry moved from annual assessment reporting to an encounter based reporting system. This resulted in the collection of more detailed information from each hospital visit.

Over six and a half thousand outpatient visits to CF specialist centres and CF clinics were attended by people with CF in 2015.

Table 14: Frequency of hospital visit by type, 2015

	Number	%
Outpatient appointment	2,635	40.0
CF day-unit review	1,592	24.2
Other*	1,280	19.4
Annual assessment**	843	12.8
Drop-in	180	2.7
Unknown	62	0.9
Total	6,592	100.0

*Includes amongst other reasons: review of annual assessment results, first dose or level-check of IV or nebulised medication, gastrointestinal issues, pulmonary function testing, weight check, physiotherapy review, port-a-cath flush, non-CF related visit. **815 patients had one annual review in 2015, 14 patients had two.

No hospital visit in 2015 was recorded for 5.2% of children, and 21% of adults. This likely reflects the fact that patients may have left the country (or emigrated), may have not attended CF services in that year, and the unavailability of hospital records at some adult hospitals.

Table 15: Number of people with a recorded hospital visit in 2015

	No recorded hospital visit	≥1 recorded hospital visit
Children (<18 years)	5.2%	94.8%
Adult (≥18 years)	21.0%	79.0%
All	13.8%	86.2%

Long-term medicines

Maintenance therapies are drug treatments used to maintain health for more than 3 months.

Table 16: Maintenance therapies, 2015

	Children <6 years N=147	Children 6-17 yrs N=377	Adult N=509	All N=1,033
Pulmonary				
Oral antibiotic	53.1%	39.0%	70.7%	56.6%
Inhaled antibiotic	6.1%	26.3%	73.5%	46.7%
Mucolytic	56.5%	87.3%	64.2%	71.5%
Bronchodilator	44.9%	65.0%	85.5%	72.2%
Inhaled steroid	10.2%	20.4%	31.2%	24.3%
Nasal medication	4.8%	26.0%	44.6%	32.1%
Gastrointestinal				
H2 receptor antagonist	3.4%	1.1%	6.5%	4.1%
Proton pump inhibitors	17.7%	31.3%	58.5%	42.8%
CF-related liver disease				
Ursodeoxycholic acid	1.4%	6.6%	17.3%	11.1%
CF-related diabetes				
Insulin	-	5.3%	22.2%	12.9%
CFTR modulator				
Kalydeco™	-	16.2%	15.7%	13.6%

Airway clearance

Airway clearance aids the clearance of mucus from the lungs.

Positive expiratory pressure (PEP) is the most commonly used airway clearance technique and usually involves using a PEP mask to provide resistance when breathing out. This helps to keep small airways open and loosen secretions. Treatment consists of breathing through a face mask (or sometimes a combination PEP-nebuliser system) with different sizes of resistors, then removing the mask to perform the forced expiration technique (huff and breathing control).

Techniques reported here are not mutually exclusive and represent primary and secondary forms of physiotherapy. In children, PEP and acapella are used most frequently, and in adults, autogenic drainage and PEP.

Table 17: Airway clearance techniques, 2015

	Children N=483	Adult N=420	All N=903
Any form of PEP	46.4%	38.6%	42.7%
Autogenic drainage	12.4%	71.0%	39.6%
Acapella	38.7%	19.3%	29.7%
Active cycle of breathing techniques	4.6%	5.2%	4.9%
Age appropriate activity	17.0%	0.2%	9.2%
Flutter	1.7%	2.6%	2.1%
Vest	3.3%	1.2%	2.3%

Table 18: Other therapies, 2015

	Child N=524	Adult N=518	All N=1,042
Home oxygen therapy	1.7%	7.1%	4.4%
Non-invasive positive pressure ventilation	2.5%	4.2%	3.4%

Transplants

Lung transplantation is an option for some people with severe lung disease.

The Irish National Lung Transplant Programme operates out of the Mater Misericordiae University Hospital. In 2014, the Irish National Lung Transplant Programme took over the full adult lung transplant service, meaning that Irish adults with CF were no longer required to travel to the United Kingdom to receive a transplant.

Twenty-two people with CF were on or were added to the Mater Misericordiae University Hospital's lung transplant waiting list in 2015. Outcomes for these individuals by the end of 2015 included; removed from the waiting list, remained on the waiting list, or received a lung transplant.

Bilateral lung transplant is the most common type of transplant in people in CF. The table below reports on the number of people with CF who received a bilateral lung transplant performed either at the Mater Misericordiae University Hospital or in the United Kingdom. At the end of 2015, no deaths had been recorded for people who had a bilateral lung transplant between 2010 and 2015.

People with CF underwent other organ transplant procedures between 2010-2015, e.g. liver and kidney. These are not reported here due to small numbers (<10).

Table 19: Transplant summary data, 2015

	No. of people receiving a bilateral lung transplant
2010	11
2011	6
2012	7
2013	10
2014	20
2015	10
Total	64

Nutrition

Nutritional outcomes height, weight and BMI are an important measure of health in people with CF and can be expressed using a Z-score. The Z score is measurement of the relationship between an individual's height/weight/BMI, and that of the average of a group of 'healthy' individuals.

Table 20: Nutritional measurements, 2015

Age (years)	Height Z score Median (IQR) N=1,029		Weight Z score Median (IQR) N=948		BMI Z score Median (IQR) N=498	
0-3	0.03	(-0.75, 0.8)	0.08	(-0.65, 0.88)	0.5	(-0.41, 1.1)
4-7	-0.1	(-0.84, 0.74)	0.02	(-0.58, 0.79)	0.22	(-0.37, 0.69)
8-11	-0.23	(-0.74, 0.38)	0.03	(-0.54, 0.56)	0.19	(-0.21, 0.83)
12-15	-0.51	(-1.18, 0.12)	-0.26	(-1.07, 0.39)	-0.04	(-0.64, 0.53)
16-19	-0.56	(-1.02, 0.19)	-0.41	(-1.31, 0.24)	-0.29	(-0.98, 0.36)
20-23	-0.21	(-0.72, 0.51)	-0.57	(-1.09, 0.09)	-	-
24-27	-0.52	(-1.1, 0.1)	-0.47	(-1.01, 0.1)	-	-
28-31	-0.49	(-1.13, 0.16)	-0.26	(-0.74, 0.24)	-	-
32-35	-0.26	(-1.01, 0.3)	-0.19	(-0.61, 0.64)	-	-
36-39	-0.52	(-1.37, 0.57)	-0.02	(-0.8, 1.0)	-	-
40-44	-0.26	(-0.75, 0.51)	0.06	(-0.54, 0.78)	-	-
45-49	-0.82	(-1.75, 0.63)	0.25	(-0.27, 1.14)	-	-
50+	-0.68	(-1.03, 0.29)	0.07	(-0.61, 0.82)	-	-
ALL	-0.36	(-0.96, 0.4)	-0.21	(-0.84, 0.5)	-	-

IQR=interquartile range.

BMI describes the weight/height relationship and is considered a good measure of nutritional status in adults. The ECFS Standards of Care² recommend BMI of greater than 20 kg/(m²), and the World Health Organisation consider a BMI of 25.0 to 29.9 as overweight.

Table 21: BMI in adults, 2015

Age (years)	BMI Median (IQR) N=393	
20-23	21.0	(19.5, 22.4)
24-27	22.0	(20.2, 24.2)
28-31	22.2	(20.7, 23.9)
32-35	23.2	(20.3, 24.4)
36-39	23.2	(21.2, 26.5)
40-44	23.3	(20.8, 24.8)
45-49	24.7	(23.0, 28.0)
50+	23.3	(21.2, 27.3)

² A.R. Smyth et al, ECFS Standards of Care: Best Practice guidelines. Journal of CF 2014;13, S23–S42.

Pancreatic enzyme replacement therapy (PERT) is required by almost all people, due to pancreatic insufficiency. When the exocrine pancreas no longer functions adequately, the production of pancreatic enzymes is reduced, leading to fat malabsorption. The consequences of this include steatorrhea, malnutrition, and fat-soluble vitamin deficiencies.

Table 22: Pancreatic enzyme replacement therapy, 2015

	Child N=524	Adult N=518	All N=1,042
Pancreatic enzyme replacement therapy for pancreatic insufficiency	93.3%	94.8%	92.1%

Supplementary feeding can be required due to continued weight loss.

Table 23: Supplemental feeding, 2015

	Children N=523	Adult N=501	All N=1,024
Any supplemental feeding	40.7%	30.9%	36.9%
Oral supplementation	29.1%	22.1%	25.5%
Gastrostomy tube/button	8.0%	6.1%	8.3%
Multiple approaches	<1%	<1%	<1%
Nasogastric tube	-	<1%	<1%
Not known	2.8%	2.1%	2.4%

Complications

Complications of CF affect many organ systems, and can interfere with a person's health and quality of life.

The prevalence of complications in 2015 is shown below. Sinus disease, CF-related diabetes and gastro-oesophageal reflux disease are common in adults.

Table 24: Complications, 2015

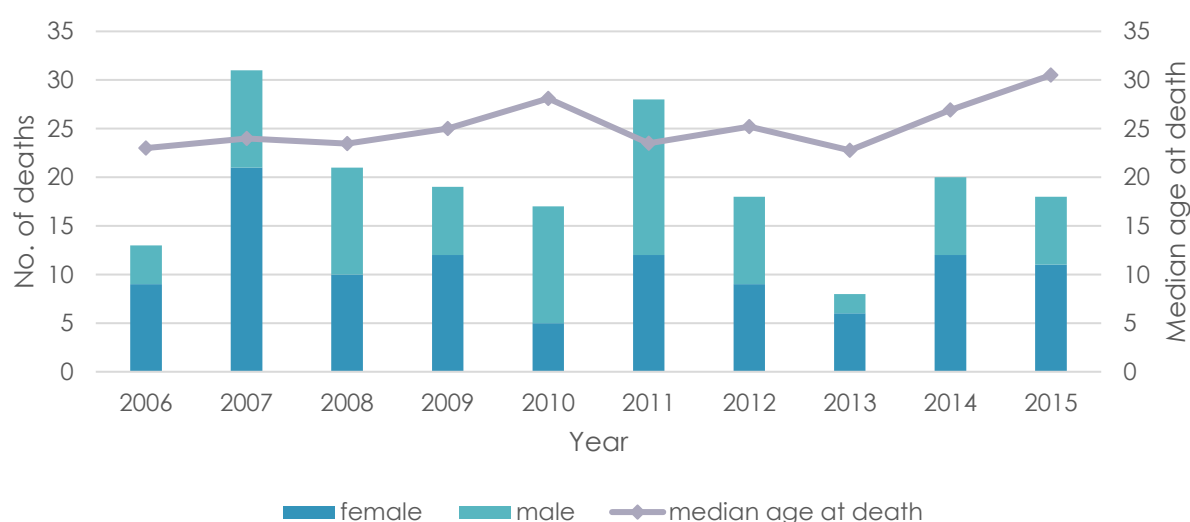
	Child N=558	Adult N=661	All N=1,219
Respiratory related			
Allergic Bronchopulmonary Aspergillosis	3.2%	4.2%	3.8%
Nasal polyps	1.8%	2.7%	2.3%
Asthma	1.8%	5.9%	4.0%
Symptomatic sinus disease	4.3%	30.1%	18.3%
Other respiratory-related complications	0.9%	3.5%	2.3%
Hepatobiliary & pancreas			
Elevated liver enzymes	1.6%	1.1%	1.3%
Liver disease other than cirrhosis	7.3%	8.2%	7.8%
Cirrhosis with portal hypertension	1.4%	4.1%	2.9%
Cirrhosis without portal hypertension /hypertension status unknown	0.4%	1.1%	0.7%
Impaired OGT	1.8%	4.1%	3.0%
CF related diabetes (CFRD)	4.5%	21.8%	13.9%
Diet-controlled CFRD	0.5%	3.3%	2.1%
Diet & insulin-controlled CFRD	3.9%	18.5%	11.8%
Gastrointestinal			
Gastro-oesophageal reflux disease	27.1%	45.2%	36.9%
Distal intestinal obstructive syndrome	1.3%	2.4%	1.9%
Musculo-skeletal			
Osteopenia	1.6%	11.5%	7.0%
Osteoporosis	0.4%	19.7%	10.8%
Arthritis/arthropathy	0.7%	1.8%	1.3%
Other			
Depression	-	5.4%	3.0%

Survival

The deaths of eighteen people with CF occurred in 2015 (11 female, 7 male), though 3 of these deaths occurred in people not registered with the CF registry. These individuals died between the ages of 16 and 70 years, with a *median age of death* of 30.5 years. As *median age of death* is based only on the length of life in people with CF who died during the year, it is an underestimate of the length of life in all people (and importantly those still living) with CF.

Of the 15 deaths in CF registry participants, deaths were due to respiratory/cardio-pulmonary causes (n=10), transplant-related issues (n=3), and other causes (n=2).

Figure 13: Deaths, 2015



Survival rates continue to improve. Nearly eighty-four percent (83.9%) of people with CF born between 1980 and 1984 survived their tenth birthday, compared with 97.7% of those born 2005-2009. One in two people born 1980-1984 survived their thirtieth birthday, compared to people born 1985-1989, where two in three survived.

Table 25: Birth cohort survival to 2015

Birth cohort	No. of births	Survived 1 st birthday	Survived 10 th birthday	Survived 20 th birthday	Survived 30 th birthday	Birth cohort survived to 2015
1980-84	279	261 93.5%	234 83.9%	186 66.7%	138 49.5%	122 43.7%
1985-89	241	233 96.7%	224 92.9%	195 80.9%	162* 67.2%*	161 66.8%
1990-94	185	176 95.1%	170 91.9%	144 77.8%	n/a** n/a**	136 73.5%
1995-99	185	184 99.5%	180 97.3%	164* 88.6%*	n/a** n/a**	163 88.1%
2000-04	174	173 99.4%	170 97.7%	n/a** n/a**	n/a** n/a**	167 96.0%
2005-09	164	164 100.0%	162* 98.8%*	n/a** n/a**	n/a** n/a**	162 98.8%
Total	1,228	1,191 97.0%	1,140 92.8%	1,020 83.1%	1,008 82.1%	911 74.2%

*not all people had reached the age indicated. **no person in the birth cohort had reached the age indicated (i.e. age 20 or 30 years) in 2015.

Applications for CFRI data, 2015

One of the CF registry's founding principles is to promote and facilitate the use of clinical data in approved research projects.

Requests for information held by the registry are assessed by the registry's scientific committee. If approved, data provided to third parties are anonymised (all identifiable data such as name, date of birth, home address, is removed). This means that individuals in the registry cannot be identified from the data shared with third parties.

The number of data applications continues to increase year-on-year; 9 in 2013, 11 in 2014 and 13 in 2015.

Table 26: Summary of application for CFRI data, 2015

	N=13
Approved data applications	8
CFRI-affiliated research team	5
Other independent researchers	2
Other	1
Applications for published data (data application not required)	5

Data applications were approved for the following projects:

Table 27: CFRI data applications, 2015

Applicant	Co-investigator	Institution	Study title
Dr A Dudina	Dr E McKone	SVUH	Obesity in cystic fibrosis patients
E Reece	Dr P Grealley	AMNCH/TCD	Co-infecting microbes: implications for cystic fibrosis airways disease
M Bourke	Dr B McCarthy	UHG	Development of a Transition Readiness Scale for use in clinical practice
P Savage		Quintiles GmbH	A phase IIA randomised double-blind study of multiple doses of GLP1837 in subjects with CF and the G551D mutation
Dr S Carter	Dr E McKone	SVUH	Understanding CF pulmonary exacerbations
Dr B Treston	Prof P McNally	OLCH	Atopy in cystic fibrosis
Dr D Hughes	Prof NG McElvaney	RCSI	Clinical correlation between different bacterial/fungal infections and CF genotype in regards to lung function, exacerbation frequency and CF-ABLE score
Dr H Veber Olesen		Aarhus University Denmark	Prevalence of liver disease study in patients with cystic fibrosis in European countries

In 2015, the registry continued to collaborate with Irish researchers on a number of ongoing CF projects.

Table 28: CFRI collaboration on ongoing research studies, 2015

	Principal Investigators	Project title	Start date	End date
1	Prof P Fitzpatrick, UCD Dr B Linnane, UHL	Early evaluation of the clinical and economic effects of the cystic fibrosis newborn screening programme*	2013	2016
2	Dr M Rowland, UCD	Cystic fibrosis liver disease*	2005	Ongoing
3	Ms E Reece & Dr P Greally, AMNCH & TCD	Co-infecting microbes: implications for cystic fibrosis airways disease**	2013	2016
4	Dr K Schaffer, SVUH Dr A-R Prior, BMT, on behalf of the Irish CF microbiology group	Epidemiology of <i>Burkholderia cepacia</i> in Ireland**	2015	2016

*Study participants authorised the release of their CFRI record to the study investigators.

**Anonymised datasets provided

The Irish CF registry contributes annually to the European CF Society Patient Registry (ECFSR). The 2014 ECFSR Annual Report has recently been published:

https://www.ecfs.eu/sites/default/files/images/ECFSR_Annual%20Report%202014_Nov2016.pdf.

In addition, research conducted using European registry data has resulted in the publication of a scientific article in 2015, entitled 'Future trends in Cystic Fibrosis demography in 34 European countries'³.

³ Burgel P-R; Bellis G, Olesen HV, Viviani L, Zolin A, Blasi F and Elborn JS. on behalf of the ERS/ECFS Task Force on The Provision of Care for Adults with Cystic Fibrosis in Europe 2015. European Respiratory Journal 2015 Mar 18. pii: ERJ-01963-2014.

CFRI research

CF Registry of Ireland research projects underway in 2015 were as follows:

Table 29: CFRI research projects, 2015

Funding body		Project title
2014-2017	Vertex Pharmaceuticals Inc	Ivacaftor Long-Term Safety Study
2015-2017	VIA	Evaluating outcomes in European cystic fibrosis patients with access to their health records: a randomised control trial of a Registry Patient Portal

In 2015, the CFRI were awarded research funding through the VIA programme. The proposed project set out to develop and evaluate a patient registry portal, which would allow participants to view their own registry data through a mobile App. Participants were recruited to the study in 2016, and the study is scheduled for completion in 2017.

The CFRI participates alongside other European countries in a long-term safety study of CF therapy ivacaftor. Summarised, anonymous information is provided for the purposes of ongoing medicine safety monitoring, which is required as per Vertex Pharmaceutical's licensing agreement with the European Medicines Agency.

Financial information

Financial information for the Cystic Fibrosis Registry of Ireland for 2015 is shown below.

Table 30: Financial information, 2015

Income & Expenses	2015 €
Income	
Core Funding	140,000
Unrestricted Grants	15,960
ECFS VIA Research Award	55,440
Commissioned Research	43,494
Deferred income released – Vertex Grant	24,500
Deferred income released – Dept of Health	17,768
Sundry income	101
Total income	297,263
Expenses	
Wages & salary	197,809
Employer's PRSI	16,318
Rent payable	6,072
Staff training	2,400
Insurance	464
Computer network & server costs	20,470
Registry software development cost	36,900
Software development cost for patient portal	13,530
Telephone & fax	218
Printing, postage and stationery	1,600
Travelling & subsistence	8,029
Legal & professional fees	0
Audit	984
Bank charges	334
Subscriptions	240
Depreciation on equipment	527
Sundry expenses	100
Total expenses	305,995
(Deficit)/Surplus	(8,732)

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 Cystic Fibrosis Registry of Ireland during this reporting year.

First we would like to thank all those who have financially supported the registry. This includes the HSE (through our Service Level Agreement), Cystic Fibrosis Ireland, our industry partners who have provided us with unconditional grants, and other funded researchers who have included us in their research grant applications. Without your support CFRI could not survive.

Most importantly we would like to thank each person with CF and/or their guardian for consenting for their medical data to be collected and used in a de-identified form to drive research into the development of new treatments and models of care for cystic fibrosis patients nationally and internationally.

We would also like to thank every member of the CF multi-disciplinary teams in every centre who assist our Research Associates in collecting data and assist in the patient consent process.

Our CFRI staff, Dr Shijun Zhou, Ms Mary Harrington, Ms Paulina Jeleniewska, Dr Laura Kirwan and Dr Abi Jackson deserves special thanks for working tirelessly in the collection and preparation of quality data that contributes so much to CF research, service and treatment development, and quality management. Particularly for the extra work that had to be done to assist with the development of the new registry technology.

Our management committee have been very supportive during the year and are always available when any assistance is required.

We would also like to thank UCD for supplying us with affordable accommodation through the sponsorship of the School of Public Health, Physiotherapy and Sport Science. We appreciate the support and mentorship of Prof Kelleher and her colleagues who have made an invaluable contribution to our own internal research programme.

The HSE's Health Intelligence Unit have been particularly supportive and special thanks go to Dr H Johnson, Mr D Beaton and Dr F Donohue.

We would to thank Prof Philip Mayne, Children's University Hospital for confirming the numbers of infants detected in 2015 by the National Newborn Bloodspot Screening Programme screening programme. We would also like to thank Prof David Barton and his team at the National Centre for Medical Genetics at Our Lady's Children's Hospital, Crumlin for assisting in the confirmation of CFTR genotyping.

Finally, we thank Dr Abi Jackson for preparing this report.

Appendix 1: Glossary

Abbreviation	
AA	Annual Assessment
ABPA	Allergic Bronchopulmonary Aspergillosis
ADULT	Aged 18 years or older (≥ 18)
AMCNH	Adelaide and Meath Hospital Inc. the Children's National Hospital
BMI	Body Mass Index
BMT	Beaumont Hospital
CFRD	Cystic fibrosis-related diabetes
CFRI	Cystic Fibrosis Registry of Ireland
CFTR	Cystic Fibrosis Transmembrane conductance Regulator (mutation)
CHILD	Aged under 18 years (< 18)
CRA	Clinical Research Associate
DIOS	Distal Intestinal Obstruction Syndrome
ECFS	European Cystic Fibrosis Society
FEV ₁	Forced Expiratory Volume in one second
FEV1pp	FEV1 percent predicted
HSE	Health Service Executive
IRT	Immunoreactive trypsinogen
IV	Intravenous
IQR	Interquartile range
MRSA	Methicillin Resistant Staphylococcus aureus
NBS	Newborn screening
NCMG	National Centre for Medical Genetics
OLCH	Our Lady's Children's Hospital, Crumlin
PAEDIATRIC	Aged under 18 years (< 18)
PEP MASK	Positive expiratory pressure mask
PERT	Pancreatic enzyme replacement therapy
PEx	Pulmonary exacerbation
RCSI	Royal College of Surgeon's Ireland
SVUH	St Vincent's University Hospital
TCD	Trinity College Dublin
UCD	University College Dublin
UHG	University Hospital Galway
UHL	University Hospital Limerick
VIA	Vertex Innovation Award

Appendix 2: Technical notes

Data collection sites

In 2015, the CFRI gathered data from the eight HSE-designated CF specialist centres (Beaumont Hospital, St Vincent's University Hospital, the Adelaide and Meath Hospital Dublin, Incorporating the National Children's Hospital, Our Lady's Children's Hospital, Crumlin, Temple Street Children's University Hospital, University Hospital Galway, Cork University Hospital and University Hospital Limerick), and five shared care CF centres (Cavan General Hospital, Mayo General Hospital, Our Lady of Lourdes Hospital, Drogheda, Sligo Regional Hospital, and Waterford Regional Hospital).

Lung function

As spirometry cannot reliably be performed until the age of 6 years, only reported values for CF individuals aged 6 and older were considered. For individuals who were reported to have had a lung transplant in or before 2014, their FEV1% predicted values in the year of transplant and post-transplant were excluded. The best value of the year was considered. The CF centre reported value of FEV1% predicted was considered. In future years, the Global Lung Function Initiative equations described by Quanjer PH *et al.*⁴ will be used.

Nutrition

Heights and weights recorded at the patient's best FEV1% predicted of the year were included in the analysis. Where no spirometry was performed, the nutrition record with the best weight measurements in the year was considered. The reference group used to estimate Z scores was the Centre for Disease Control (CDC) 2000 reference charts.⁵

Z scores are a statistical measurement of the relationship between an individual's height/weight/BMI, and the mean of a group of reference individuals. A z-score of 0 means that the measurement (e.g. height/weight/BMI) is equal to the mean measurement of individuals of the same age and sex in the reference (i.e. healthy population). A z-score of -2 means that the value is two standard deviations below the mean of people of the same age and sex in the reference population, and a score of +2 means that the value is two standard deviations. The average score for a healthy population is typically zero.

Maintenance therapies, airway clearance, complications

Treatments/complications recorded at individuals' annual assessments were considered. If an AA record was not available, data from the person's last hospital visit of 2015 was considered.

⁴ Multi-ethnic reference values for spirometry for the 3-95 year age range: the global lung function 2012 equations'. Report of the Global Lung Function Initiative (GLI), ERS Task Force to establish improved Lung Function Reference Values, Eur Respir J. 2012; 40(6): 1324–43.

⁵ Kuczmarski RJ, Ogden CL, Guo SS, et al. 2000 CDC Growth Charts for the United States.

Appendix 3: Executive council membership 2015

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	CF Ireland
Mr Godfrey Fletcher	CEO	Chief Executive Officer Cystic Fibrosis Registry Ireland
Prof Peter Grealley		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
Prof 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
Prof Barry Plant		Consultant in Respiratory Medicine Cork University Hospital, Cork
Mr Philip Watt		Chief Executive CF Ireland

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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