2012 Annual Report

The Canadian Cystic Fibrosis Registry

Cystic Fibrosis Canada

Breathing life into the future®

Cystic Fibrosis

Cystic fibrosis (CF) is the most common fatal genetic disease affecting Canadian children and young adults. It is a multi-system disease that affects mainly the lungs and the digestive system. In the lungs, where the effects are most devastating, a build-up of thick mucus causes severe respiratory problems. Mucus and protein also build up in the digestive tract, making it difficult to digest and absorb nutrients from food. As improved therapies have helped to address the malnutrition issues, ultimately most deaths related to cystic fibrosis are due to lung disease. There is no cure.

Cystic Fibrosis Canada

Cystic Fibrosis Canada is a national charitable not-for-profit corporation established in 1960, and is one of the world's top three charitable organizations committed to finding a cure for cystic fibrosis. As an internationally-recognized leader in funding cystic fibrosis research, innovation, and clinical care, we invest more funding in life-saving CF research and care than any other non-governmental agency in Canada.

Since 1960, Cystic Fibrosis Canada has invested more than $150 million in leading research, innovation and care, resulting in one of the world's highest survival rates for Canadians living with cystic fibrosis. For more information, visit www.cysticfibrosis.ca.

Our mission is to help people with cystic fibrosis by:

- Funding research towards the goal of a cure or control for cystic fibrosis;
- Supporting high quality cystic fibrosis care;
- Promoting public awareness of cystic fibrosis; and
- Raising and allocating funds for these purposes.

Published: February, 2014

This document is made from 100% recycled paper.
Join us in reducing our impact on the environment by choosing to recycle.

Table of Contents

The Canadian Cystic Fibrosis Registry

The first national *Canadian Cystic Fibrosis Registry* was created in the early 1970s with the goal of monitoring important clinical trends in the Canadian CF population. Previously named the *Canadian Patient Data Registry*, it was rebranded in 2012 to ensure the Registry featured the name of the disease.

The Registry has played an invaluable role in helping to improve the quality and length of life of people with cystic fibrosis.

Since the majority of CF patients attend one of 42 accredited CF clinics (child and adult) within Canada, it includes data on virtually all Canadians diagnosed with cystic fibrosis — giving a comprehensive picture of the CF population in this country.

The Registry is used by CF clinicians and researchers to improve their knowledge of disease patterns and care of patients with cystic fibrosis.

CF clinicians can access the Registry data to better understand their own clinic population and respond to emerging health care issues, including nutritional status, infectious pathogens, pulmonary treatment, and more.

The data collected within the Registry can be used for quality improvement efforts. Clinics can compare pulmonary and nutritional outcomes of individuals followed at their clinic to the national median value.

Quality improvement initiatives can be developed and clinical outcomes can be tracked over time using the Registry in order to show improvements. These efforts will ultimately translate into improved outcomes for people with cystic fibrosis.

Epidemiologic research examines trends and improvements in a given population over time. Since CF patient data registries study large populations of individuals with cystic fibrosis over time, the Registry is a powerful research tool.

The Registry can be used as an educational tool. The summary statistics help to graphically show important clinical outcomes over time. Incorporating these summary statistics into presentations for the public, medical and allied health care professionals, and many other groups can increase knowledge about this disease in Canada.

For the first time in the history of Cystic Fibrosis Canada, CF clinic specific data from the *Canadian CF Registry* are available to the public online on our new website at www.cysticfibrosis.ca. This data transparency promotes open communication within the CF community and can help improve the quality of CF care and treatment across Canada.

Thanks to the contributions and participation of clinical teams and Canadian CF patients, in addition to the generous support of our donors, partners and volunteers, it will be possible to ensure that data remain available and worthy of study in the future.

2012 Highlights

- Approximately **4,000 Canadians** with cystic fibrosis received care at one of the **42 specialized CF clinics** based in hospitals across Canada
- There were **105 new diagnoses made in 2012**: 60 were under 6 months of age and 16 were over 18 years of age
- Nearly **60%** of all people with cystic fibrosis in Canada **are adults**
- Cumulatively, CF patients spent almost **20,000 days in hospital** and attended over **15,000 clinic visits** in 2012
- Cumulatively, CF patients underwent **787 courses of home IV therapy** in 2012
- The **median age of survival** for Canadians with cystic fibrosis is currently estimated to be **49.7 years of age**
- Of the 43 patients who died in 2012, **half were under 32 years old**
- FEV_1 (a measure of lung function) is improving for persons with cystic fibrosis: **half of all 30 year olds** with cystic fibrosis have an **FEV_1 greater than 64.2%** predicted in 2012 compared to 46.4% two decades ago
- **86.7%** of Canadians with cystic fibrosis must take **pancreatic enzymes** to digest food and absorb nutrients
- **30.6% of female adults** with cystic fibrosis and **18.9% of male adults** with cystic fibrosis are classified as **underweight**
- **37 CF patients received transplants** in 2012
- Nearly **half of all patients** with cystic fibrosis are **infected in their lungs with harmful bacteria** including *Staphylococcus aureus* and *Pseudomonas aeruginosa*
- **22.8% of all CF patients have CF-related diabetes**, and approximately 34.7% of all individuals with cystic fibrosis-related diabetes are 35 years of age and older
- Over **1,900 different mutations** in the *CFTR* gene have been identified, however **87.5% of CF patients in Canada** carry at least one copy of the most common CF-causing mutation, **deltaF508**

Message from Maureen Adamson

President and CEO, Cystic Fibrosis Canada

What an incredible time in the history of the *Canadian CF Registry* — an outstanding national treasury of crucial CF patient data.

After more than four decades of publishing data from the *Canadian CF Registry*, we have stepped-out into the online world. For the first time in our history, clinic-specific outcomes from the *Canadian CF Registry* are now publicly available on our new website at cysticfibrosis.ca. This bold step demonstrates our enhanced focus on excelling in CF care.

We are thrilled to report that the median age of survival of Canadians with cystic fibrosis has increased to 49.7 years of age in the 2012 data. It is a clear indication that Canadians with cystic fibrosis are living longer, healthier lives than ever before and that we continue to make a positive impact through our collective efforts.

Thank you to so many who devoted their time and expertise to the production of the 2012 annual report including the clinic teams who painstakingly entered their patient data; Dr. Anne Stephenson, Director of the *Canadian CF Registry*; and the team at Cystic Fibrosis Canada. Moreover, without the consent of all those with cystic fibrosis in Canada to include their data, this report — and its important findings — would not be possible.

It is through the generosity, passion and commitment of our donors, partners and volunteers that it is possible to bring you this important national CF resource.

In the year ahead, we will invest in a brand new CF data registry that will enhance our abilities to more easily capture key data about CF patients and bring us even greater insights as we continue to lead the way in improving clinical care for Canadians with cystic fibrosis.

Together, we have set our sights squarely on finding a cure.

Message from Dr. Anne Stephenson MD, PhD

Director, CF Registry

I am pleased to present the *Canadian CF Registry 2012 Annual Report*.

The *Canadian CF Registry* continues to be an important national resource as we capture clinical data on over 4,000 people diagnosed with cystic fibrosis in Canada. There are new additions to this report that we hope will provide further insights and stimulate important dialogue among clinicians throughout Canada. Furthermore, Registry data are being used to improve CF care across the country through new quality improvement initiatives.

Researchers can access existing data within the Registry to investigate clinical questions and conduct studies in a timely and efficient way. The historical records make it possible to uncover patterns and examine trends over time.

As always, patient privacy and confidentiality is our utmost priority. We are committed to securely protecting patient information so that privacy is maintained and respected.

Thank you to my wonderful colleagues in the CF clinical community across Canada who devote hours of their time and take meticulous care to enter clinical data into the Registry. In particular, we are deeply grateful to the patients and their families who allow us to collect and publish their data year after year.

We continue our great tradition together in producing this excellent resource for the CF community.

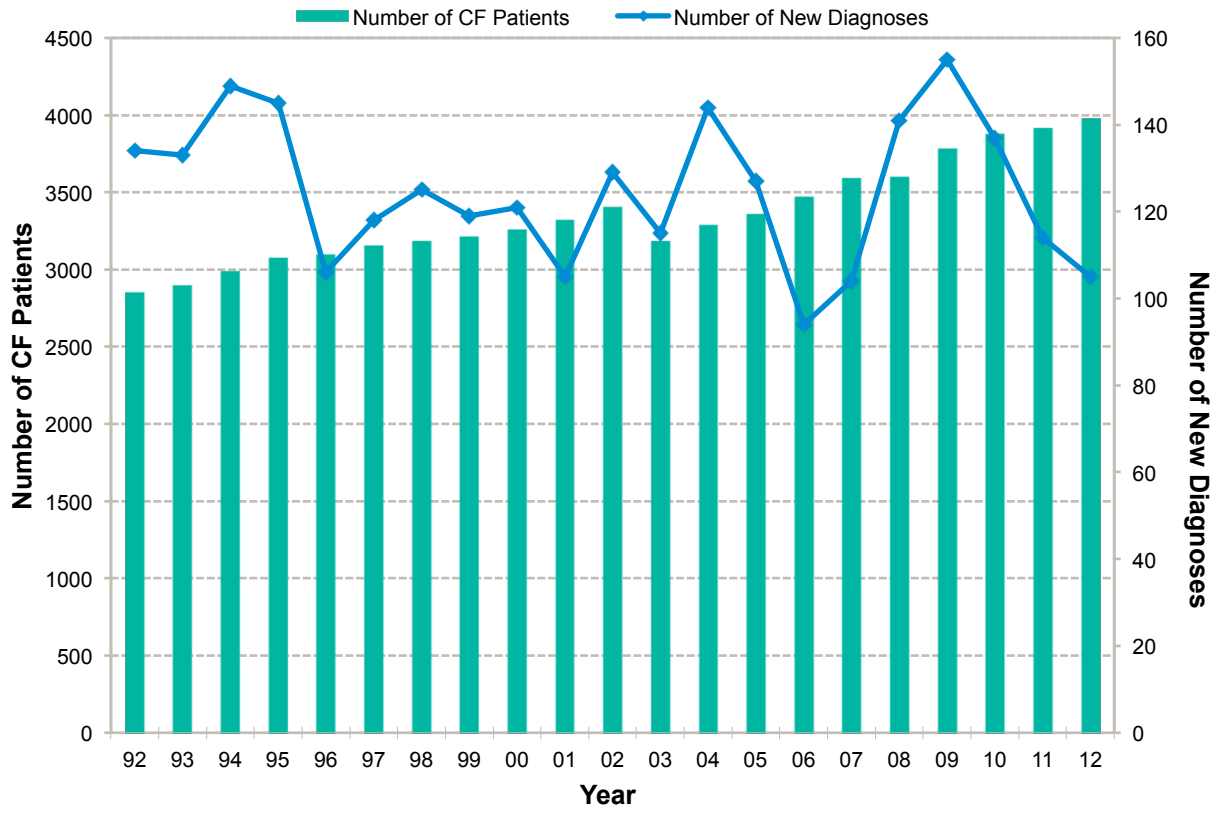
Demographic Data

Number of Canadians with Cystic Fibrosis

In 2012, a total of 3,975 individuals with cystic fibrosis had clinical records submitted by 42 CF clinics (Figure 1). When an individual was seen at multiple clinics in one year, she or he was only counted once (*i.e.* unique individuals) for the purpose of generating this graph. In 2012, 105 individuals were newly diagnosed with this disease.

Figure 1

Total number of CF patients and new diagnoses recorded in the Registry, 1992 to 2012



Demographic Data

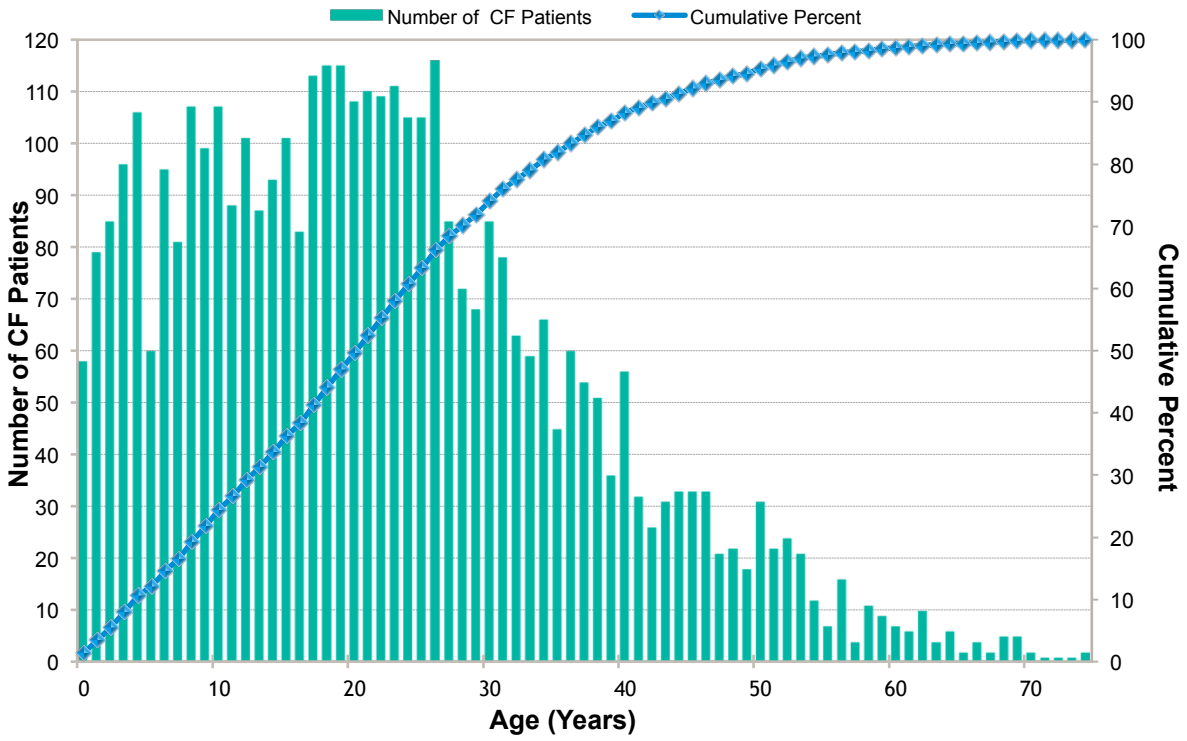
Ages of Canadians with Cystic Fibrosis

Figure 2 shows the age distribution of the Canadian CF population for 2012. The ages of individuals with cystic fibrosis range from birth to over 70 years old. The median age is 21 years.

Males account for 52.9% of individuals in the Registry in 2012, and 58.8% of individuals in the Registry are 18 years of age or older (Figure 3).

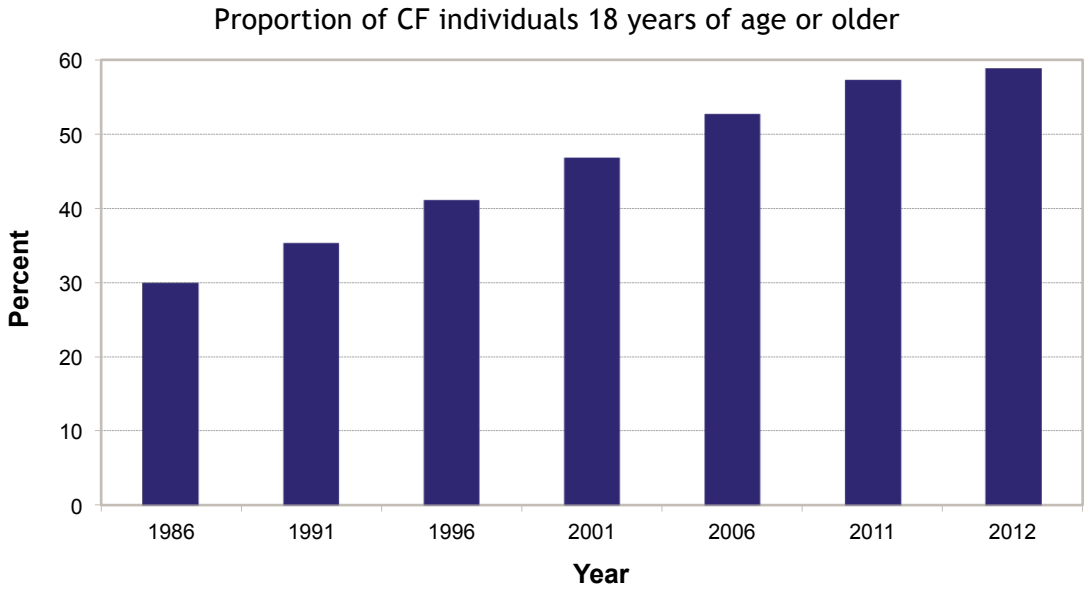
Figure 2

Age distribution of the CF population, 2012



Demographic Data

Figure 3



Demographic Data

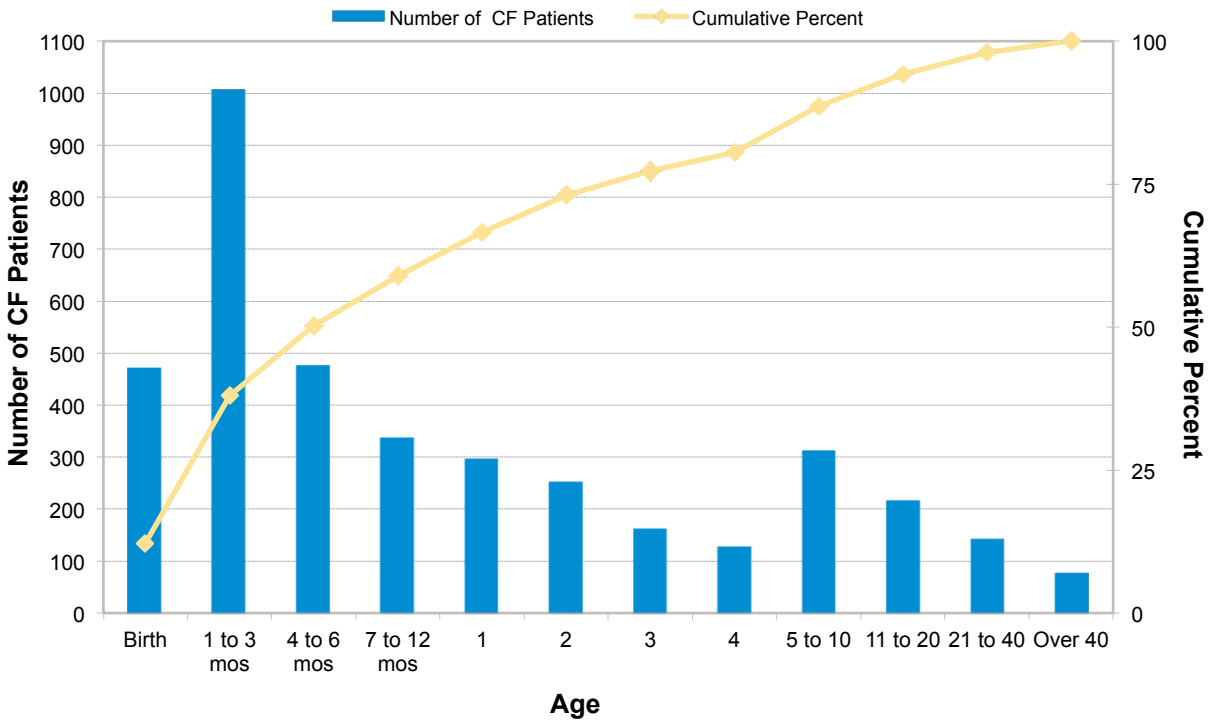
Age at Diagnosis

Fifty percent (50%) of individuals are diagnosed by six months of age, and 73% by the age of two years (Figure 4). Adults continue to be diagnosed with cystic fibrosis, with 2% diagnosed after the age of 40 years.

As newborn screening (NBS) programs for cystic fibrosis are introduced in Canadian provinces (currently available in BC, AB, SK, MB, ON and available in NS starting in 2014), the majority of individuals with cystic fibrosis will be diagnosed at birth. Figure 5 shows the percentage of babies diagnosed through the NBS program over the past decade. In 2012, 39% of all new diagnoses were made from the NBS program.

Figure 4

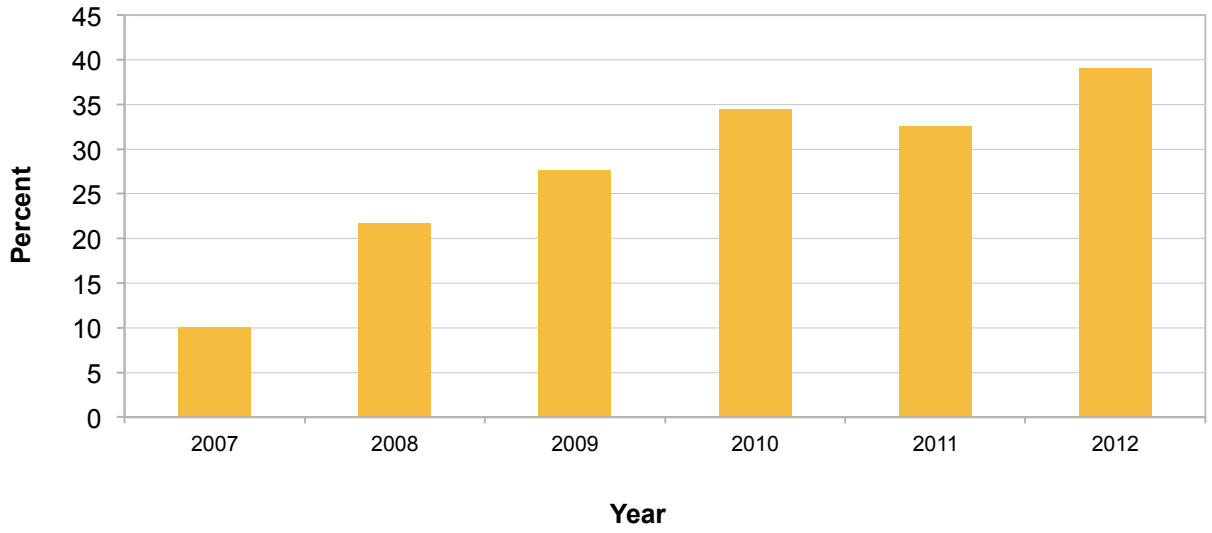
Age at diagnosis, all patients, 2012



Demographic Data

Figure 5

Proportion of newborn diagnoses made through the NBS program, 2007 to 2012



Genotype

Cystic fibrosis is caused by mutations in a single gene located on chromosome 7, termed the Cystic Fibrosis Transmembrane Regulator (*CFTR*) gene. The *CFTR* gene codes for a protein called the cystic fibrosis transmembrane conductance regulator (CFTR) which functions as a chloride channel and is involved in many cellular functions. To date, more than 1,900 different mutations in the *CFTR* gene have been identified.

The most common mutation worldwide is a three base-pair deletion in the *CFTR* gene resulting in the deletion of the phenylalanine (F) residue at position 508 in the CFTR protein, commonly referred to as **deltaF508**. Of those individuals with genetic information recorded within the Registry, 49.2% carry two deltaF508 mutations (Figure 6) and 87.5% carry at least one deltaF508 mutation (Table 1).

Figure 6

Genotype (based on N=3,871)

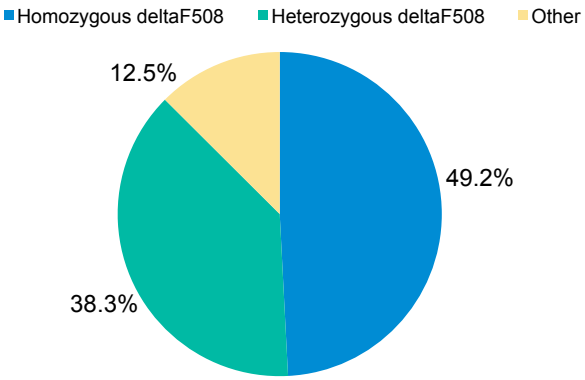


Table 1

Frequency of CF mutations on one or both alleles (top five)

Genotype	Number	Percentage
DeltaF508	3,477	87.5%
621+1G->T	229	5.8%
G542X	144	3.6%
G551D	118	3.0%
711+1G->T	98	2.5%

Respiratory

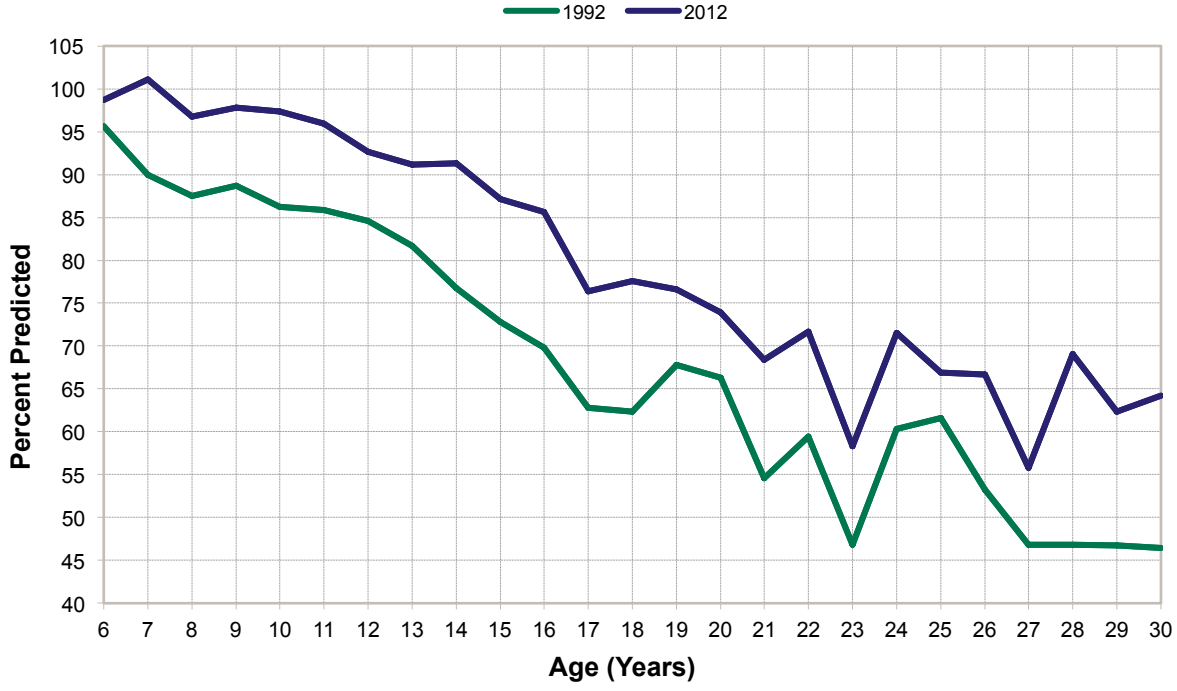
Median FEV_1 Percent Predicted

Respiratory measures are needed to evaluate lung health. FEV_1 (forced expiratory volume in one second) is the volume of air that a person can forcibly blow out in one second. FEV_1 % predicted for a patient is calculated against the average FEV_1 of a healthy population of similar age, height and sex. The national median FEV_1 percent predicted for adults is 65.5% (ages 18+) and 92.2% for children (ages 6-17).

Median FEV_1 percent predicted has improved over the last two decades but interestingly, the trend in lung function is similar between 1992 and 2012. The median FEV_1 percent predicted in 2012 at 30 years of age is 64.2% compared to 46.2% in 1992 (Figure 7). An average decline in lung function of 0.7% per year occurs from ages 6 to 11 years but from ages 11 to 23, a much larger average decline of 2.5% per year is seen. After age 23, lung function seems to have stabilized with an average decrease of 0.1% per year. This suggests that perhaps the adolescent/young adulthood years are a vulnerable time for individuals with cystic fibrosis.

Figure 7

Median FEV_1 percent predicted vs. age, 1992 and 2012



Respiratory

Respiratory Status

Lung function is measured starting at six years of age. For children ages 6 to 17 years, the majority of individuals with cystic fibrosis (58%) have normal lung function (FEV_1 greater than or equal to 90 percent predicted), compared to 55% in 2011. For adults, the majority (40%) have lung function classified as 'moderate' severity (Figure 8). These figures are similar to those in 2011.

Figure 8

Respiratory status of CF children and adults, 2012

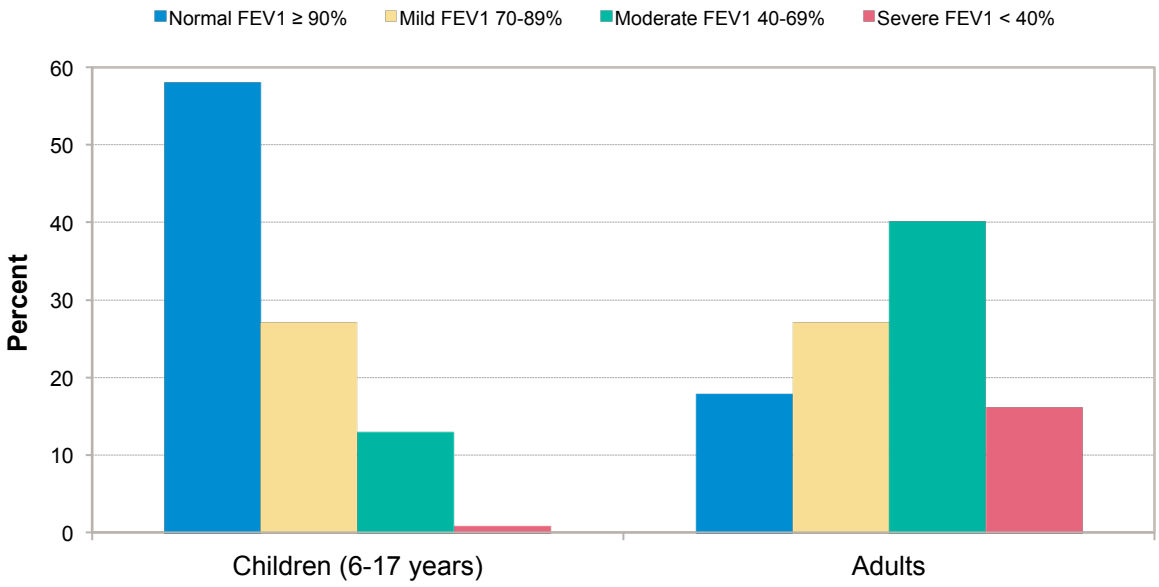


Table 2

Respiratory classification

Classification	Range
Normal FEV_1	≥90% of predicted
Mild FEV_1	70 - 89% of predicted
Moderate FEV_1	40 - 69% of predicted
Severe FEV_1	<40% of predicted

Respiratory

Respiratory Status by Sex

Figures 9 and 10 show that lung function is generally quite similar between males and females. As patients get older, a larger proportion of individuals have lung function in the moderate and severe range.

Figure 9

Respiratory status of CF children ages 6 to 17, by sex, 2012

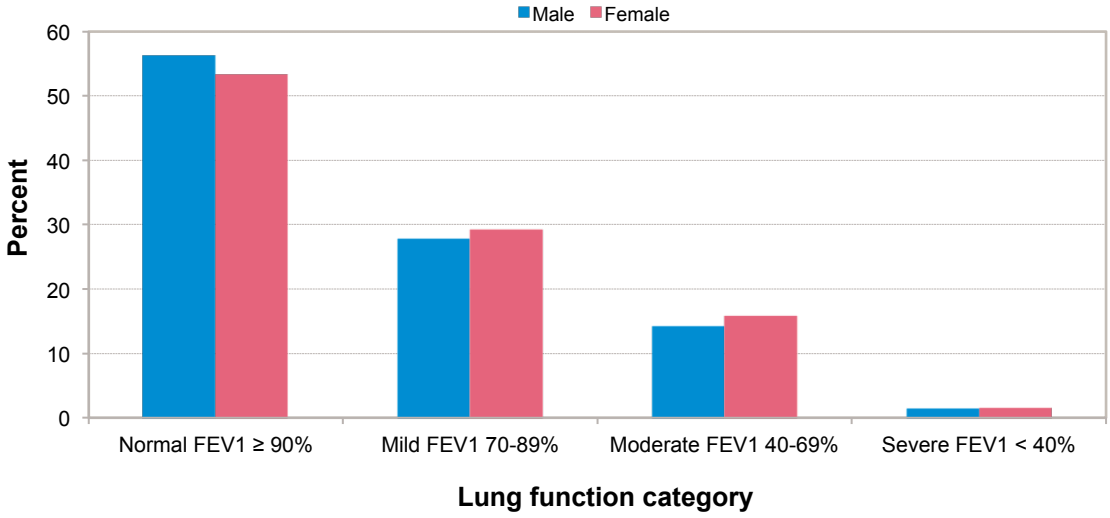
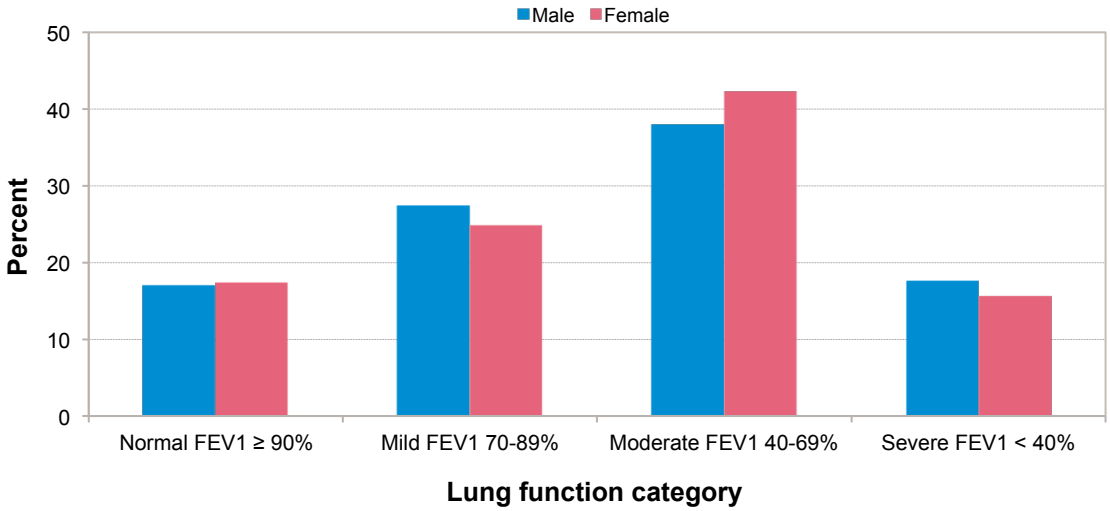


Figure 10

Respiratory status of CF adults, by sex, 2012



Nutrition

Pancreatic Status

Malnutrition is commonly seen in individuals with cystic fibrosis as a result of pancreatic insufficiency. Pancreatic enzymes are given as a supplement to help with digestion and absorption of nutrients. In 2012, 86.7% of individuals with cystic fibrosis were taking supplemental pancreatic enzymes (pancreatic insufficient), whereas 13.3% did not require oral pancreatic enzyme supplementation to digest their food (pancreatic sufficient) (Figure 11).

For those individuals 40 years of age or older, 33.7% are pancreatic sufficient (Figure 12). This is likely a reflection of the fact that individuals diagnosed with cystic fibrosis as adults are more likely to have milder mutations that are associated with pancreatic sufficiency.

Figure 11

Pancreatic sufficiency in individuals with cystic fibrosis

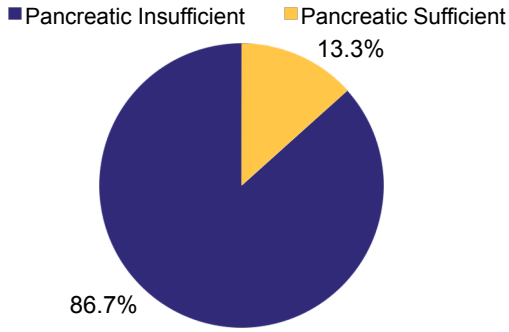
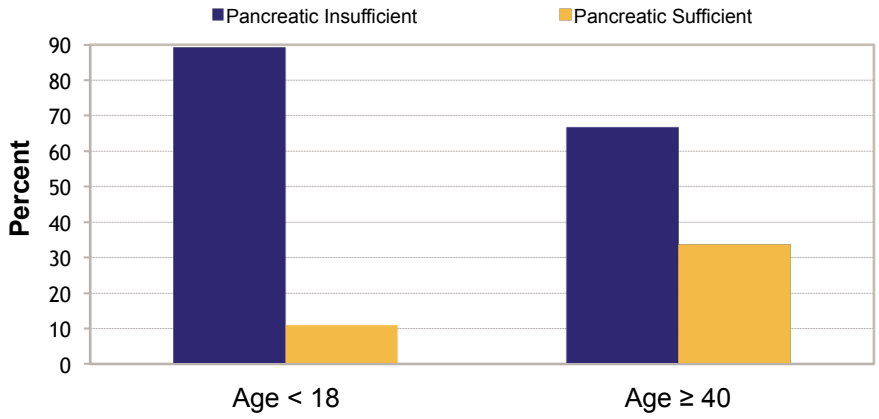


Figure 12

Pancreatic status by age group, 2012



Nutrition

Body Mass Index (BMI)

Body mass index (BMI) is a measurement of nutrition and is based on a person's weight (in kilograms) and height (in meters). Typically, we calculate this for adults only because they have attained their maximal height whereas children are rapidly growing and therefore one must consider the child's age when assessing their nutritional status. The national median BMI for adults (ages 18+) is 21.9 kg/m^2.

Table below describes the BMI classifications and their BMI ranges. In 2012, the majority of the adult CF population (59.4%) has an adequate weight while 4.7% are considered obese (Figure 13).

Figure 13

BMI classification for adults with cystic fibrosis, 2012

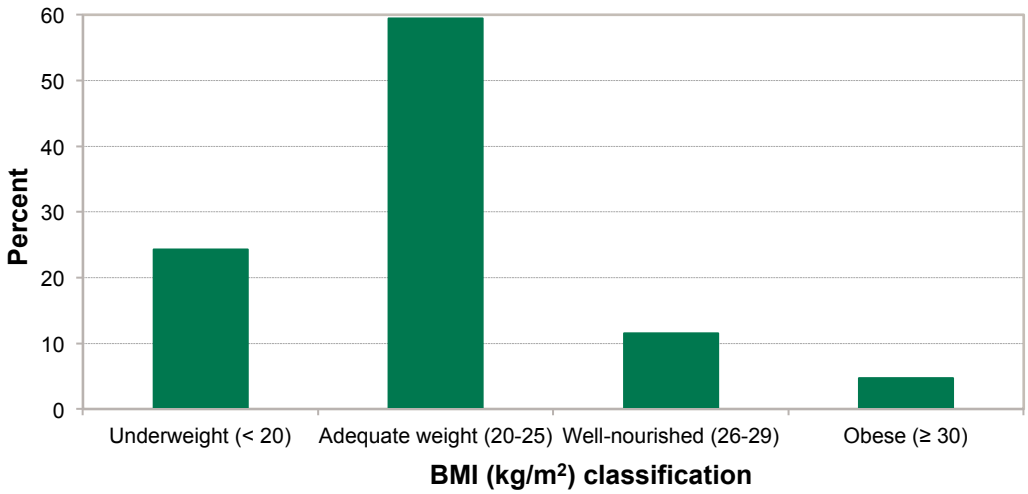


Table 3

BMI classification

Classification	Range
Underweight	<20.0 kg/m^2
Adequate weight	20.0 - 25.9 kg/m^2
Well-nourished	26 - 29.9 kg/m^2
Obese	≥30 kg/m^2

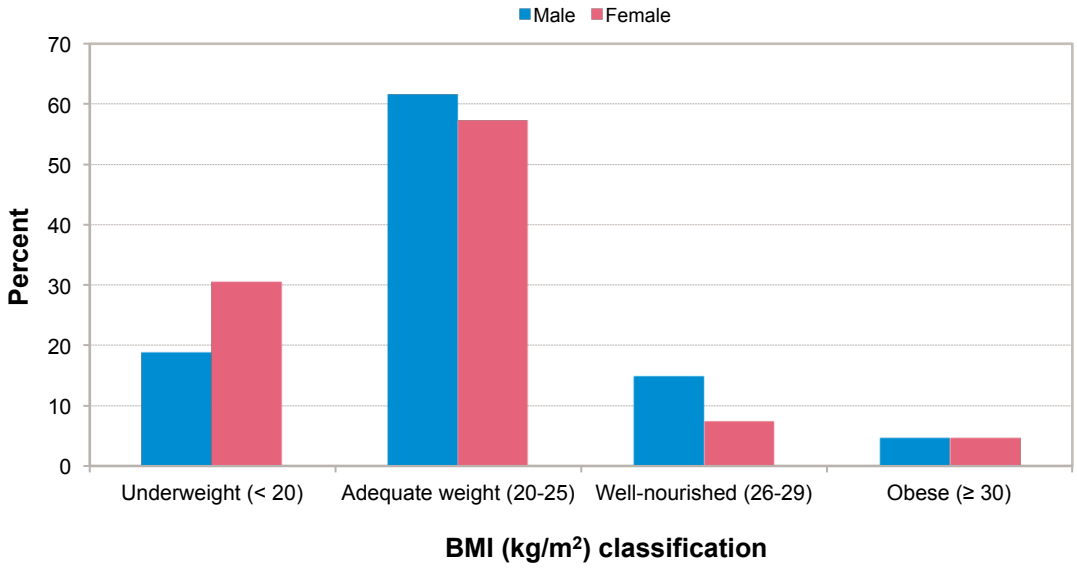
Nutrition

BMI by Sex

Figure 14 shows the breakdown of BMI categories (see previous page for categories) for adult males and females. A larger proportion of females are considered underweight (BMI < 20 kg/m^2) compared to males. Individuals who are muscular can have a BMI between 26-29 kg/m^2 due to increased weight from high muscle mass.

Figure 14

BMI classification for adults with cystic fibrosis, by sex, 2012



Nutrition

BMI Percentile

For children ages 2 to 17, BMI percentiles are calculated comparing the individual's height and weight to those of children who are the same age and sex following the Centers for Disease Control and Prevention guidelines (Figure 15). BMI percentile is not calculated for those under the age of two years. The national median BMI percentile for children is 45.4 (ages 2-17). The majority of patients (82%) have a healthy weight with a small proportion considered overweight (6.9%) and obese (2.8%).

Figure 15

BMI percentile classification for children with cystic fibrosis, 2012

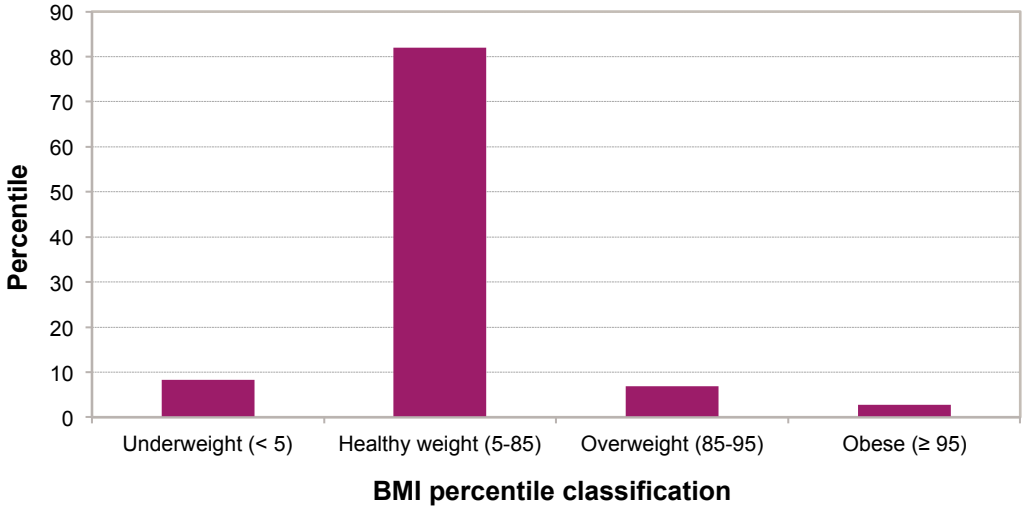


Table 4

BMI percentile classification

Classification	Percentile Range
Underweight	<5th percentile
Healthy weight	5th percentile - <85th percentile
Overweight	85th - <95th percentile
Obese	≥95th percentile

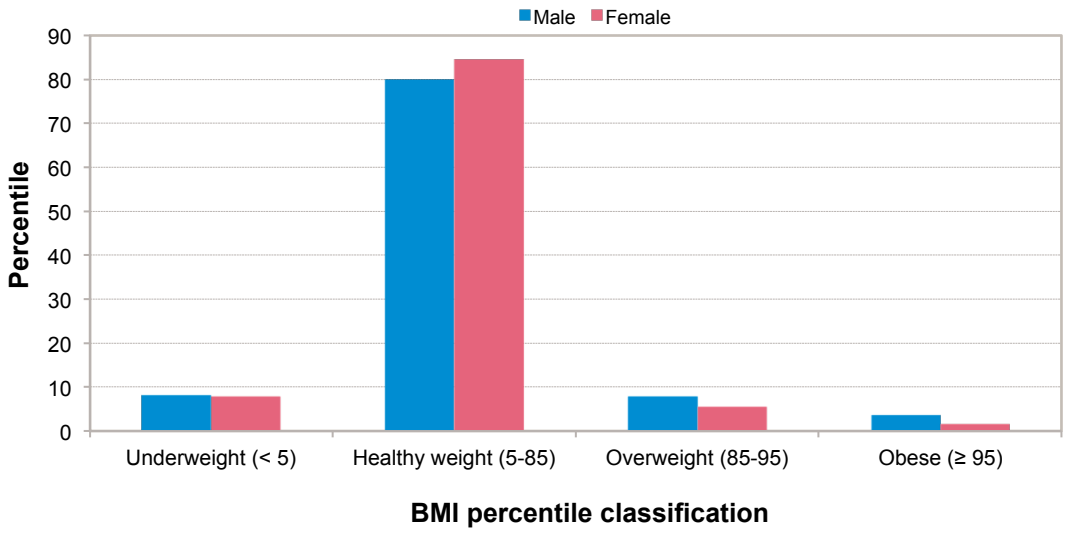
Nutrition

BMI Percentile by Sex

Figure 16 shows the breakdown of BMI percentile classifications for males and females between the ages of 2 and 17 (Table 3). While there are slightly more females with a healthy weight than males (4% difference), there are minor disparities between males and females across the remaining BMI percentile categories (difference of 0.2-2.3% across remaining categories).

Figure 16

BMI percentile classification for children with cystic fibrosis, by sex, 2012

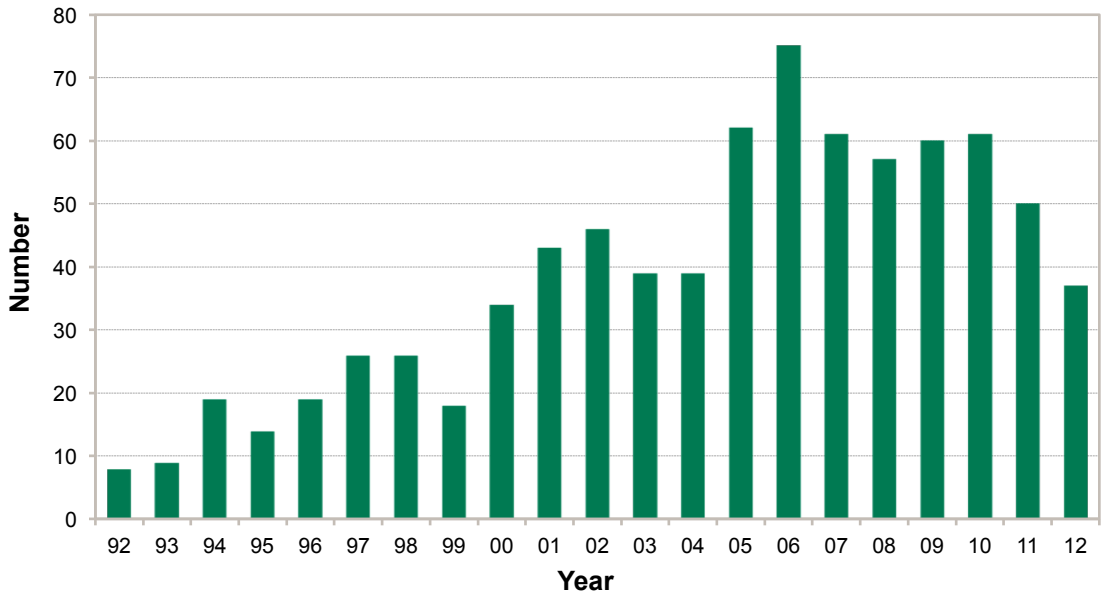


Transplants

Figure 17 shows the number of transplants carried out per year as reported in the Registry. Although the numbers provided represent primarily lung transplants, individuals who received other combinations (*e.g.* lung-liver, liver, heart-lung, etc.) are also included in the total. As of December 31, 2012, there are 602 CF patients recorded in the Registry as having received one or more transplants and of those individuals, 361 individuals are reported as being alive.

Figure 17

Number of transplants per year, 1992 to 2012



Microbiology

Bacterial Species and Respiratory Infections

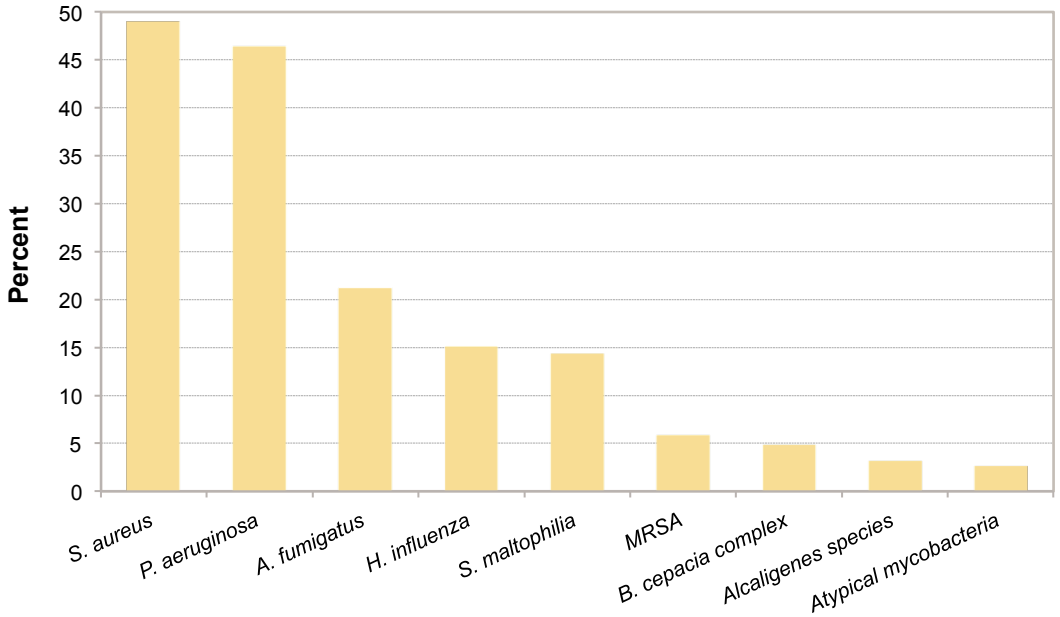
Overall, *Pseudomonas aeruginosa* and *Staphylococcus aureus* are the most common pulmonary pathogens in Canadians with cystic fibrosis (Figure 18). In 2011, clinics began to record additional microbiology data including the prevalence of Alcaligenes (achromobacter) species and Atypical mycobacteria.

The prevalence of *Aspergillus fumigatus* species, *Stenotrophomonas maltophilia*, and Methicillin-Resistant *Staphylococcus aureaus* (MRSA) has increased over the last several years (Figure 19). The largest increase was seen in *Aspergillus* species. The increased prevalence may be partly due to increase surveillance for these organisms. The prevalence of MRSA has gradually been increasing and in 2012, almost 6% of Canadians had a positive sputum sample for MRSA. MRSA data were not collected in the Registry prior to 2003. As expected, *Staphyloccocus aureus* is more common in the children CF population whereas *Pseudomonas aeruginosa* is more common in the adult CF population.

The highest prevalence of *Stenotrophomonas maltophilia* is in the teen years (11-17 years) and appears to be less prevalent in older individuals (Figure 20). *Burkholderia cepacia* complex is more commonly seen in older individuals with cystic fibrosis. New acquisition of *B. cepacia* complex in general has decreased substantially over the years, due to infection control practices, making its prevalence in children low. However, those individuals who previously acquired *B. cepacia* complex are aging, making the prevalence of this organism higher in older individuals.

Figure 18

Prevalence of bacterial species cultured in airways in CF patients (all ages), 2012



Microbiology

Figure 19

Prevalence of respiratory infections, 2008-2012

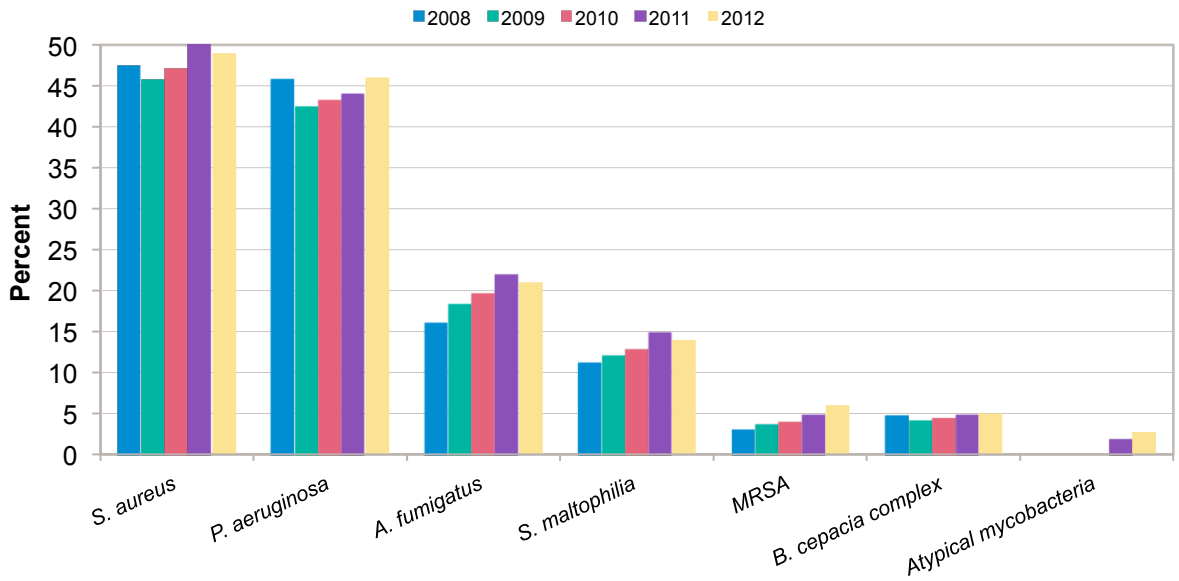
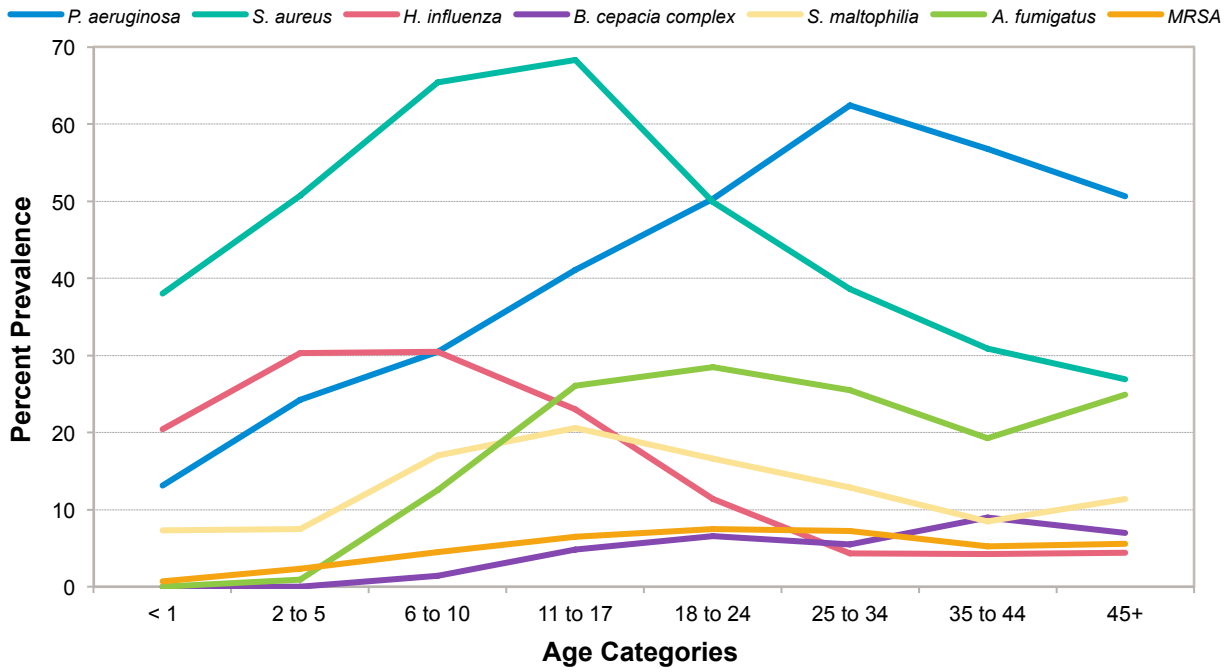


Figure 20

Age-specific prevalence of respiratory infections in CF patients, 2012



Microbiology

Burkholderia cepacia complex (BCC)

There were 195 patients with *Burkholderia cepacia* complex (BCC) species in 2012 (Figure 21). *B. cenocepacia* and *B. multivorans* were the two most common types of BCC species. Of the patients with BCC species, 82.6% are adults (Figure 22). Not all BCC bacteria have been genotyped represented by the fact that 22.1% of the BCC species in the Registry are classified as unknown. *B. gladioli* has a prevalence of 7.2% and was not included in Figure 21 because it is not officially recognized as part of the BCC species.

Figure 21

Burkholderia cepacia complex species, 2012

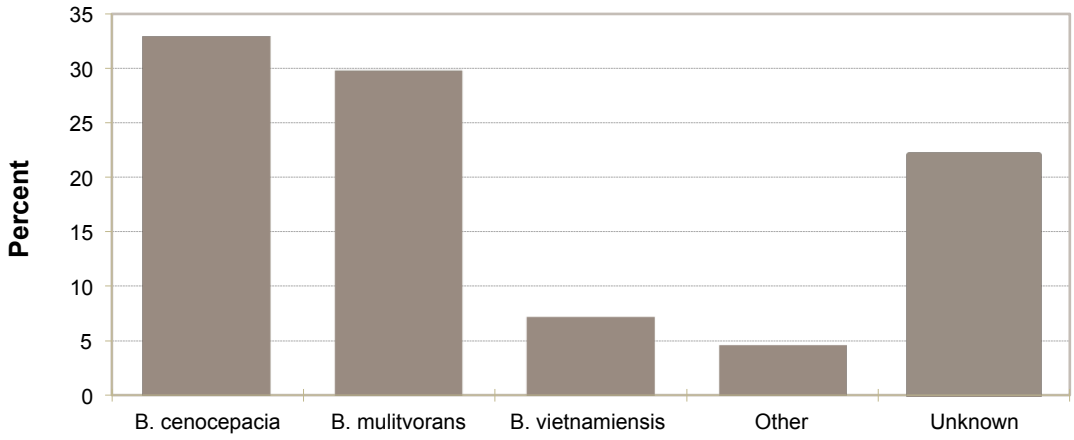
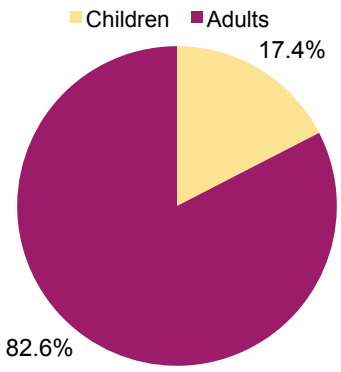


Figure 22

Burkholderia cepacia complex distribution by age group, 2012

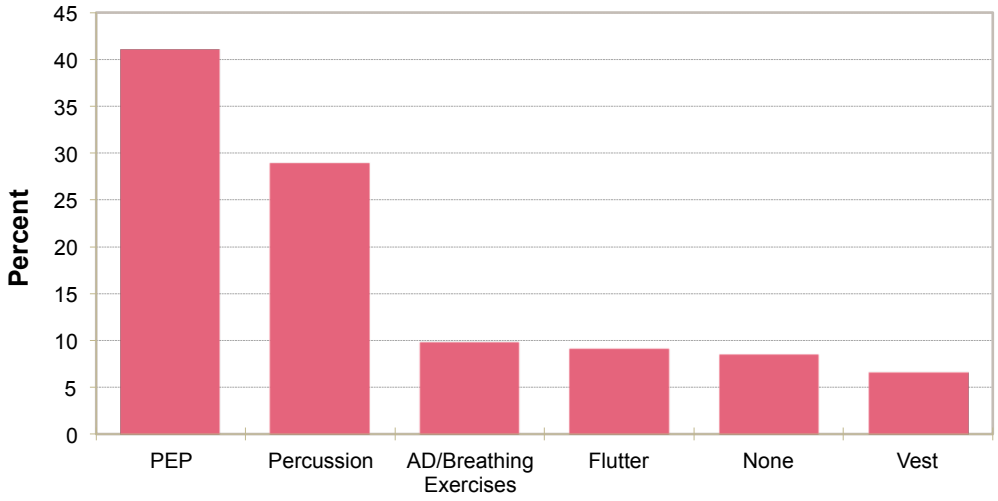


Physiotherapy

Physiotherapy is done to help clear mucus from airways using a variety of methods. Positive expiratory pressure (PEP) and percussion are the most common forms of physiotherapy used by Canadian CF patients (Figure 23). Individuals who had received a transplant were excluded from these calculations because, typically, no chest phyisotherapy is needed since transplanted lungs do not have cystic fibrosis.

Figure 23

Physiotherapy (based on N = 3,728), 2012

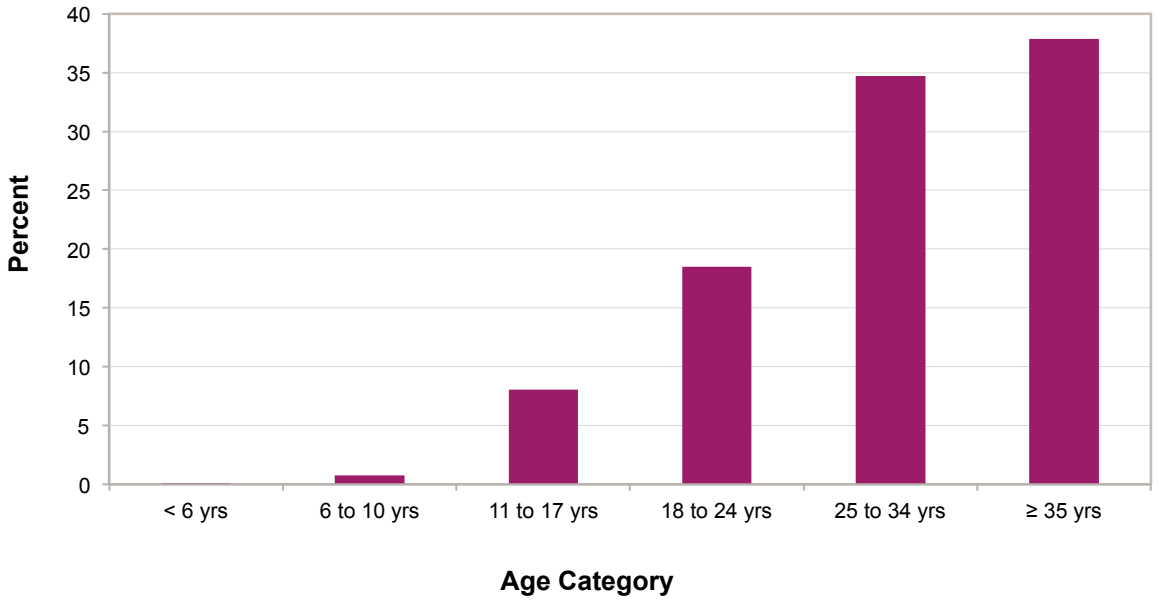


CF-Related Diabetes (CFRD)

Overall, CFRD was reported in 22.8% of individuals with cystic fibrosis in 2012. In those individuals with CFRD, approximately 37.8% are 35 years of age and older. The prevalence of CFRD increases with age (Figure 24). Of those with CFRD, 51.8% are female.

Figure 24

Percentage of patients with CFRD by age category, 2012



Hospitalization and Home IV

Pulmonary Exacerbation is the Leading Cause

In 2012, 1,540 hospitalizations were recorded in the Registry (Table). The most common recorded reason for admission was a pulmonary exacerbation. In 2012, 787 courses of home IV therapy were recorded within the Registry.

Table 5

Number of hospital days and home IV courses, 2012

	Total Number
Hospital Days	19,649
Hospitalizations	1,540
Clinic Visits	15,676
Home IV Courses	787
Home IV Days	13,174

Survival

There were 43 deaths recorded in the Registry for 2012. Since there are relatively few deaths per year, the sum of all deaths from 2008 to 2012 have been included in Figure 25. The median age at death in 2012 was **32 years of age** (Figure 26). Median age at death is calculated simply by taking into account all deaths in a given year, placing them in ascending order, and determining which age is the middle number. The median age at death is calculated using only those individuals who have died. In other words, of those who died, half died before the median age of death and half died later than the median age of death. The most common cause of death was pulmonary-related.

Figure 25

Age at death, 2008 to 2012

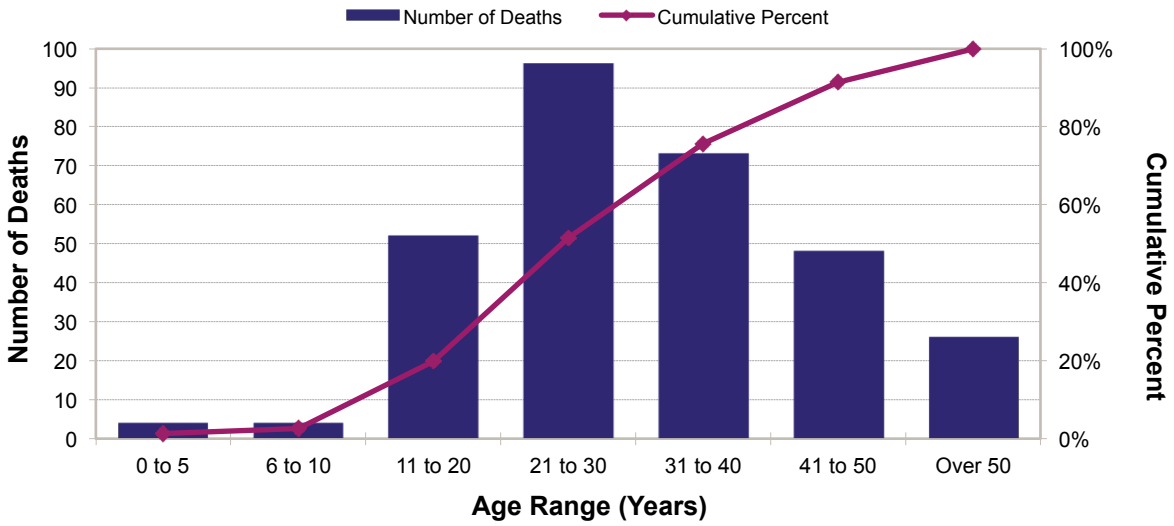
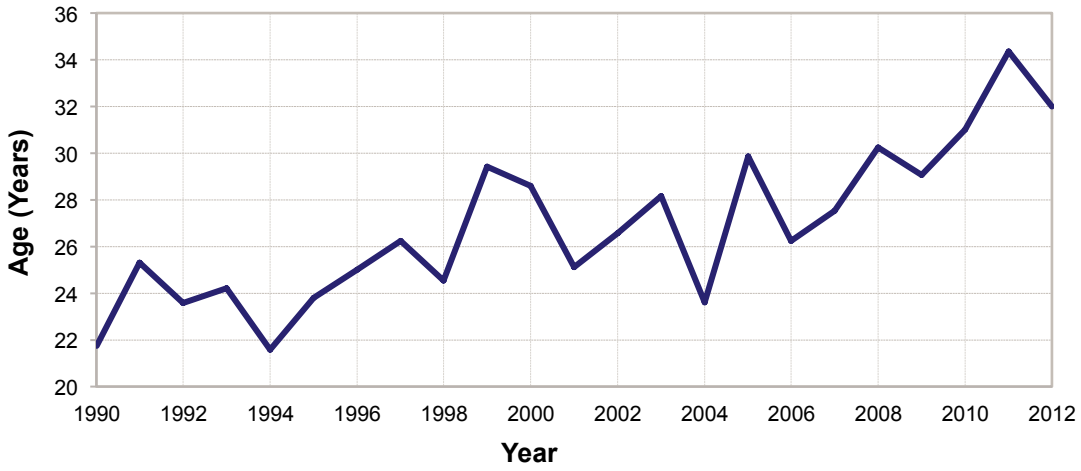


Figure 26

Median age at death, 1990 to 2012



Survival

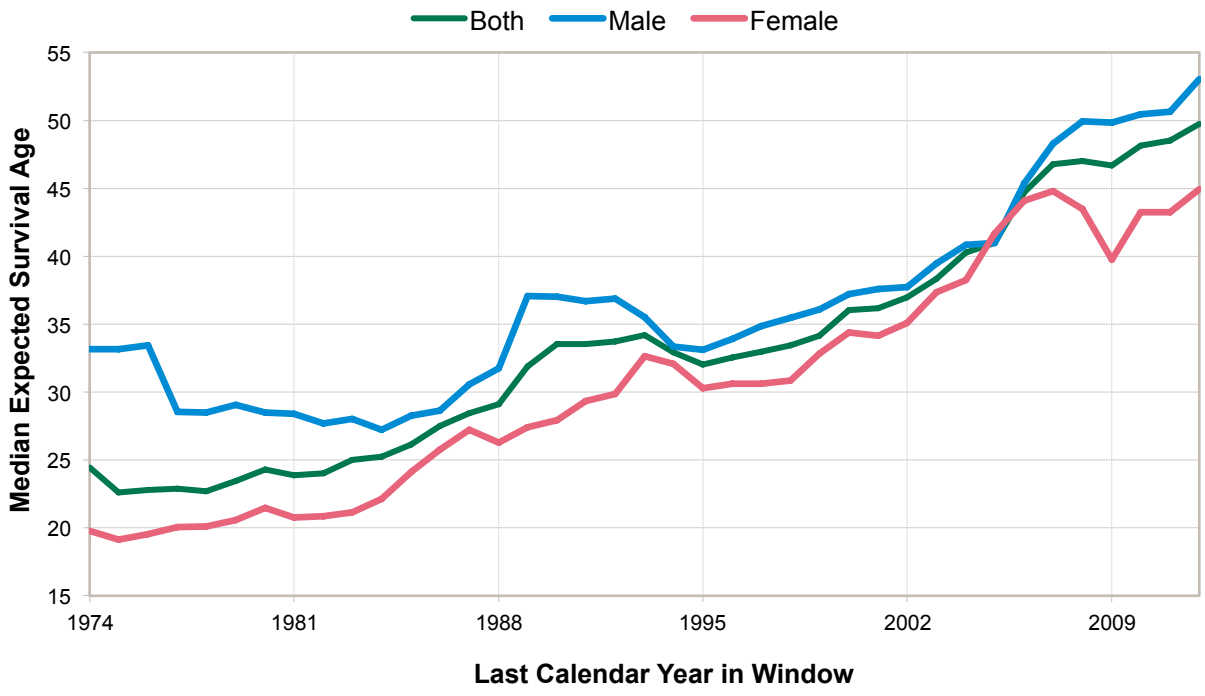
Median Age of Survival

The median age of survival for Canadians with cystic fibrosis is currently estimated to be **49.7 years of age** (Figure 27). Over the years, females have had higher mortality than males with cystic fibrosis which continues to persist in 2012. The cause of lower survival in females is not well understood but has been documented in the literature in many countries in addition to Canada. Because there are relatively few deaths per year in Canada, we use a 5-year window in order to calculate median survival age. This allows for more stable estimates over time.

It is not possible to know for certain the reason for the improvement seen in survival for Canadians living with cystic fibrosis and in truth, there are likely multiple factors. Certainly it is a statistic that would not be possible without the hard work and dedication of CF families, volunteers, partners, donors, researchers and CF clinic teams. Everyone can be very proud of this accomplishment.

Figure 27

Median age of survival for a moving 5-year window by sex



Survival

Median Age of Death

The **median age at death** is very different than the **median age of survival**. Median age at death is calculated simply by taking into account all deaths in a given year, placing them in ascending order, and determining which age is the middle number. The median age at death is calculated using only those individuals who have died. In other words, of those who died, half died before the median age of death and half died later than the median age of death.

This calculation does not provide information about the individuals who have survived. You need to know the ages of those still living to get information on median survival.

Life Expectancy

The life expectancy is the **expected average length of life** based on current age-specific mortality rates. Life expectancy at birth in Canada for example is 77.3 years. This means that a baby born now will, on average, be expected to live 77.3 years. It is not the same as median age of survival.

Median Age of Survival

Median age of survival is the **estimated duration of time until 50 percent of a given population dies**. Half of the population are still alive and half have died. The age at which this occurs is the **median survival age**.

Summary Data

Table 6

Summary data from Registry, 1987 to 2012

	1987	1992	1997	2002	2007	2012
General Profile						
Number of patients with clinical records in reporting year (n)	2,433	2,851	3,153	3,403	3,592	3,975
Male, % of total patients	53.1	53.8	53.8	53.6	53.3	52.9
Age, mean (yr)	13.4	15.2	17.0	18.7	21.0	22.3
Age, median (yr)	12.0	14.0	15.0	17.0	19.0	21.0
% over 18 yrs	31.1	36.3	42.3	47.2	55.1	58.8
Race, % Caucasian	98.1	97.4	96.8	96.2	94.3	92.5
Black	0.2	0.4	0.6	0.7	0.8	0.8
Asian	0.4	0.4	0.4	0.4	0.4	0.6
First Nations People	0.3	0.5	0.7	0.7	0.7	1.0
South Asian	---	---	---	---	---	0.1
Other	0.8	1.1	1.2	1.5	1.9	0.1
Unstated	0.2	0.3	0.3	0.1	0.1	0.1
Transplants (#)	---	8	26	46	61	37
Pancreatic insufficient (%)	82.9	91.9	92.2	90.8	89.1	86.7
% with genotype analysis	---	---	---	---	---	97
Diagnosis						
Age at diagnosis, mean (yr)	2.2	2.4	2.8	3.2	3.6	4.0
median (mo)	7.0	6.0	6.0	6.0	7.0	6.0
New diagnoses in year (n)	136	135	117	130	109	105
% with meconium ileus at birth	14.2	16.4	17.6	21.9	9.2	14.8
Survival/Mortality						
Age at death, mean (yr)	18.2	23.0	27.3	28.1	30.3	34.0
median (yr)	17.2	23.6	26.2	26.6	27.5	32.0
Total deaths (n)	49	57	58	57	49	43
Crude mortality rate (%)	2.0	2.0	1.8	1.7	1.4	1.1
Median age of survival (yr)	28.4	33.7	32.9	36.9	46.7	49.7
Male	30.5	36.8	34.8	37.7	48.2	53.0
Female	27.2	29.8	30.6	35.0	44.8	44.9
Nutritional Markers						
≥ 18 yrs of age: n (%) in BMI categories						
< 20,	334 (47%)	383 (41%)	434 (36%)	424 (29%)	479 (27%)	561 (25%)
20-25.9,	348 (49%)	480 (52%)	681 (56%)	861 (59%)	1069 (60%)	1334 (59%)
26-29.9,	22 (3%)	59 (6%)	84 (7%)	144 (10%)	171 (10%)	254 (11%)
≥ 30	3 (0%)	9 (1%)	19 (2%)	28 (2%)	60 (3%)	104 (5%)

Summary Data

Males < 23 BMI	328 (81%)	390 (74%)	467 (71%)	481 (60%)	551 (58%)	635 (53%)
Females < 22 BMI	241 (80%)	302 (75%)	385 (69%)	416 (64%)	523 (64%)	614 (58%)
2 - 17 yrs of age: n (%) in BMI categories						
< 50th BMI %ile	959 (64%)	1043 (61%)	1042 (60%)	980 (58%)	885 (57%)	844 (55%)
< 25th BMI%ile	557 (37%)	598 (35%)	588 (34%)	515 (30%)	464 (30%)	414 (27%)
Pulmonary Function						
% predicted FEV_1, mean	70.3	71.1	71.8	72.9	71.9	73.2
% predicted FEV_1, median	72.1	73.2	74.4	74.9	73.1	75.5
n (%) FEV1 % predicted categories for ≥ 18 years:						
Normal: ≥ 90%	68 (11%)	116 (13%)	161 (14%)	168 (12%)	234 (13%)	382 (18%)
Mild: 70-89%	115 (19%)	171 (20%)	247 (21%)	329 (23%)	436 (25%)	577 (27%)
Moderate: 40-69%	240 (40%)	338 (39%)	482 (41%)	600 (43%)	737 (42%)	862 (40%)
Severe: < 40%	182 (30%)	241 (28%)	290 (25%)	305 (22%)	339 (19%)	350 (16%)
n (%) FEV1 % predicted categories for ages 6 to 17:						
Normal: ≥ 90%	329 (39%)	409 (38%)	507 (43%)	611 (50%)	537 (51%)	532 (58%)
Mild: 70-89%	255 (30%)	338 (32%)	377 (32%)	363 (30%)	321 (30%)	251 (27%)
Moderate: 40-69%	207 (24%)	252 (24%)	236 (20%)	200 (16%)	167 (16%)	124 (13%)
Severe: < 40%	59 (7%)	70 (7%)	51 (4%)	45 (4%)	31 (3%)	13 (1%)
n (%) on oxygen	---	---	---	97 (3%)	83 (2%)	134 (3%)
n (%) on BiPAP	---	---	---	---	---	28 (1%)
Microbiology						
n (%) with positive culture (first culture of the year (76-96), starting in 2001 any culture in the year):						
Pseudomonas aeruginosa	1028 (42%)	1226 (43%)	1379 (44%)	1666 (49%)	1619 (45%)	1831 (46%)
Staphylococcus aureus	603 (25%)	857 (30%)	1043 (33%)	1618 (48%)	1732 (48%)	1934 (49%)
Haemophilus species	308 (13%)	398 (14%)	398 (13%)	602 (18%)	593 (17%)	597 (15%)
Stenotrophomonas maltophilia	---	---	112 (4%)	264 (8%)	400 (11%)	571 (14%)
Aspergillus	---	---	---	311 (9%)	450 (13%)	839 (21%)
MRSA	---	---	---	---	112 (3%)	235 (6%)
Alcaligenes (achromobacter) species	---	---	---	---	---	127 (3%)
Atypical mycobacteria	---	---	---	---	---	106 (3%)
Burkholderia cepacia complex	226 (9%)	275 (10%)	221 (7%)	187 (5%)	167 (5%)	191 (5%)
B. cenocepacia	---	---	---	---	---	61 (32%)
B. multivorans	---	---	---	---	---	57 (30%)
B. vietnamiensis	---	---	---	---	---	14 (7%)
B. gladioli	---	---	---	---	---	14 (7%)
B. cepacia Other	---	---	---	---	---	9 (5%)
Unknown	---	---	---	---	---	41 (21%)

Acknowledgments

Thank you to the following groups and people who made outstanding contributions to the *Canadian Cystic Fibrosis Registry* and this 2012 Annual Report.

Canadian CF Registry Review Panel

Dr. Mark Chilvers (BC Children's Hospital, Vancouver)
Dr. Peter Durie (The Hospital for Sick Children, Toronto)
Dr. Larry Lands (Montreal Children's Hospital)
Dr. Mark Montgomery (Alberta Children's Hospital, Calgary)
Dr. Hans Pasterkamp (Winnipeg Children's Hospital)
Dr. Elizabeth Tullis (St. Michael's Hospital, Toronto)
Dr. Ian Waters (Royal Jubilee Hospital, Victoria)

Cystic Fibrosis Canada

Maureen Adamson, President and CEO
Ken Chan, Vice President, Advocacy, Research and Healthcare
Dr. Anne Stephenson, Director, CF Registry
Dr. Denise Mak, Program Advisor, Healthcare
Ian McIntosh, Program Director, Healthcare
Amanda Bland, Registry Support Coordinator

Canadian CF Clinics

Victoria General Hospital	Hotel-Dieu Hospital, Kingston
Royal Jubilee Hospital, Victoria	Children's Hospital of Eastern Ontario, Ottawa
BC Children's Hospital, Vancouver	Ottawa General Hospital, Ottawa
St. Paul's Hospital, Vancouver	*Centre de santé et de services sociaux de Gatineau, Hull*
Alberta Children's Hospital, Calgary	Montreal Children's Hospital, Montreal
Foothills Hospital, Calgary	Montreal Chest Institute, Montreal
University of Alberta Hospitals, Edmonton	*Hôpital Ste-Justine, Montréal*
Royal University Hospital, Saskatoon	*Hôtel-Dieu de Montréal, Montréal*
Regina General Hospital, Regina	*Centre Universitaire de Santé de l'Estrie, Sherbrooke*
Winnipeg Children's Hospital, Winnipeg	*Centre hospitalier de l'Université Laval, Québec*
Health Sciences Centre, Winnipeg	*Institut universitaire de cardiologie et de pneumologie de Québec, Québec*
Health Sciences North/*Horizon Santé-Nord*, Sudbury	*Hôpital de Chicoutimi, Chicoutimi*
Windsor Regional Hospital, Windsor	*Centre hospitalier régional de Rimouski, Rimouski*
London Health Sciences Centre and Children's Hospital at LHSC, London	*Centre de santé et des services sociaux de Rouyn-Noranda, Rouyn-Noranda*
Grand River Hospital, Kitchener	IWK Health Centre, Halifax
St. Mary's Hospital, Kitchener	QEII Health Sciences Centre, Halifax
Hamilton Health Sciences Corporation, Hamilton	Saint John Regional Hospital, Saint John
The Hospital for Sick Children, Toronto	Janeway Children's Health Centre, St. John's
St. Michael's Hospital, Toronto	Health Sciences Centre, St. John's

Breathing life into the future®

Since 1960, Cystic Fibrosis Canada has invested more than $150 million in innovative CF research and clinical care in Canada.

Funds for Cystic Fibrosis Canada's research and clinical care programs are raised through national events like the Great Strides™ walk, and community initiatives across the country.

Cystic Fibrosis Canada's work relies on the generosity of many individual donors, corporate partners, organizations, and volunteers.

Together we are making a difference. With every stride we take, we never know which breakthrough will lead to a cure. We continue to boldly invest in promising research in our mission to find a cure or control for cystic fibrosis.

Join the fight against cystic fibrosis today.

Donate and find out more at www.cysticfibrosis.ca

Cystic Fibrosis Canada™

2323 Yonge Street, Suite 800, Toronto, Ontario, M4P 2C9
2014-02 | *Cette publication est aussi disponible en français.*
Charitable registration: 10684 5100 RR0001