

2013 Annual Report

The Canadian Cystic Fibrosis Registry



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.

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The Canadian Cystic Fibrosis Registry

The *Canadian Cystic Fibrosis Registry* is a national resource comprising clinical data on the Canadian CF population. First created in the early 1970s, the Registry was established to support the CF research and clinical community in their knowledge of disease patterns and care of patients with cystic fibrosis.

The Registry is a powerful research tool used to monitor important epidemiological trends that help guide the direction of research and clinical care - with the goal of improving the quality and length of life of people with cystic fibrosis.

The Registry is also used for educational purposes. The summary statistics help to graphically show clinical outcomes over time. These visuals are presented to the public, medical and allied healthcare professionals, and many other groups to share and increase the knowledge about this disease in Canada.

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.

CF clinicians can access the Registry data to better understand their own clinic population and respond to emerging healthcare 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. These efforts will ultimately translate into improved outcomes for people with cystic fibrosis.

CF clinic-specific data from the *Canadian CF Registry* are available to the public on our 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.

It is impossible to know for certain the reason for the improvement in survival for Canadians living with cystic fibrosis and in truth, there are multiple factors. Certainly it would not be possible without the hard work and dedication of CF families, volunteers, partners, donors, researchers, and CF clinic teams.

Thank you to the CF clinic teams that contribute their time and effort in capturing the data, to the Canadian CF patients who participate, and to the generous support of our donors, partners and volunteers. Without you all, this Registry could not exist and would not be the vital resource that it has become. Everyone can be very proud of this accomplishment.

2013 Highlights

- Over **4,000 Canadians** with cystic fibrosis received care at one of the **42 specialized CF clinics** based in hospitals across Canada
- The median age of Canadians with cystic fibrosis is **21.4 years of age**
- The **median age of survival** for Canadians with cystic fibrosis is currently estimated to be **50.9 years of age**
- There were **118 new diagnoses made in 2013**: 68 were under 6 months of age and 13 were over 18 years of age
- **59.0%** of CF patients are diagnosed within their first year of life
- **Almost 60%** of all people with cystic fibrosis in Canada are adults
- Cumulatively, CF patients spent over **24,500 days in hospital** and attended over **16,500 clinic visits** in 2013
- Cumulatively, CF patients underwent **676 courses of home IV therapy** in 2013
- Of the 40 patients who died in 2013, **half were under 35.1 years of age**
- FEV₁ percent predicted (a measure of lung function) is improving for persons with cystic fibrosis: **half of all 30 year olds** with cystic fibrosis had an FEV₁ greater than **63.7%** in 2013 compared to **46.0%** two decades ago
- **86.2%** of Canadians with cystic fibrosis must take **pancreatic enzymes** to digest food and absorb nutrients
- **29.6%** of female adults with cystic fibrosis and **18.1% of male adults** with cystic fibrosis are classified as **underweight**
- **44.4%** of female children and **43.0%** of male children with cystic fibrosis are above the national goal of 50th BMI percentile
- **44 CF patients** received transplants in 2013
- **Over 40%** of all patients with cystic fibrosis are infected in their lungs with harmful bacteria including *Staphylococcus aureus* and *Pseudomonas aeruginosa*
- **23.1%** of all CF patients have CF-related diabetes, and 39.7% of these individuals are 35 years of age and older
- Over **1,900 different mutations** in the *CFTR* gene have been identified; however **89.7%** of CF patients in Canada carry at least one copy of the most common CF-causing mutation, **F508del**

Message from Norma Beauchamp

President and CEO, Cystic Fibrosis Canada

Cystic Fibrosis Canada works tirelessly so that all Canadians with cystic fibrosis (CF) have access to specialized CF care through its network of 42 accredited CF clinics. We are proud to report that, based on the most recent patient data in the *Canadian Cystic Fibrosis Registry*, the future for CF patients has never looked brighter.

The first Canadian CF patient registry was created in the early 1970s with the goal of monitoring important clinical trends in the Canadian CF population. The data has played an indispensable role in helping to improve the quality and length of life of Canadians with cystic fibrosis.



The data collected within the Registry are used to better understand clinic populations, respond to emerging healthcare issues, develop quality improvement initiatives and track clinical outcomes over time. This translates into outstanding care for Canadian CF patients. Through our collective efforts, Canadians with cystic fibrosis are living longer, healthier lives than ever before and the predicted median age of survival has increased to 50.9 years of age according to the 2013 data.

While we have made great progress in the quest for a cure or control for cystic fibrosis, our work is far from over. Too many lives are still cut too short - of the patients who lost their battle to CF in 2013, half were under 35 years old. One of the most crucial components to continue to improve the quality of care for Canadians with cystic fibrosis is through the data collected at CF clinics across the country.

This year we invested in a brand new CF data registry that has enhanced our abilities to 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. The data collected in the *Canadian Cystic Fibrosis Registry* are more than an invaluable resource to clinicians and researchers - they benchmark the tremendous progress we've made and highlight key trends that will lead to even better CF research and treatments.

The *Canadian Cystic Fibrosis Registry* would be impossible without generous funding from our donors and partners; we are indebted for their ongoing investment and support. We are deeply grateful to the CF patients and their families who generously agree to share their data, without them this important national CF resource would also be impossible.

Together, we can make a difference in the lives of Canadians with cystic fibrosis.

Message from Dr. Anne Stephenson MD, PhD

Director, CF Registry, Cystic Fibrosis Canada



I am proud to present the *Canadian CF Registry 2013 Annual Report*. This annual report serves to inform and enhance our understanding of cystic fibrosis in Canada by examining historical clinical records and highlighting important trends. The *Canadian CF Registry* is a national resource and plays a significant role in both CF research and clinical studies, and in clinical CF settings through quality improvement initiatives.

This report was made possible because of the hard work and tireless efforts of many people. We would like to recognize the commitment and dedication of the CF clinical community across Canada. These individuals enter clinical data into the Registry and ensure that it is both comprehensive and complete.

We would like to acknowledge the individuals living with cystic fibrosis and their families who allow their clinical data to be collected and published - the Registry could certainly not exist without their support. Our top priorities, as they always have been, are patient privacy and confidentiality. We continue our commitment to safeguard, respect and maintain patient information.

Finally, we would like to thank the generous community of donors whose care and encouragement have been unwavering in our shared vision of finding a cure or control for cystic fibrosis.

We strive, each year, to improve the knowledge and outlook on the health of people living with cystic fibrosis in Canada. This report represents the accomplishments of everyone involved and we are increasingly optimistic about what the future holds.

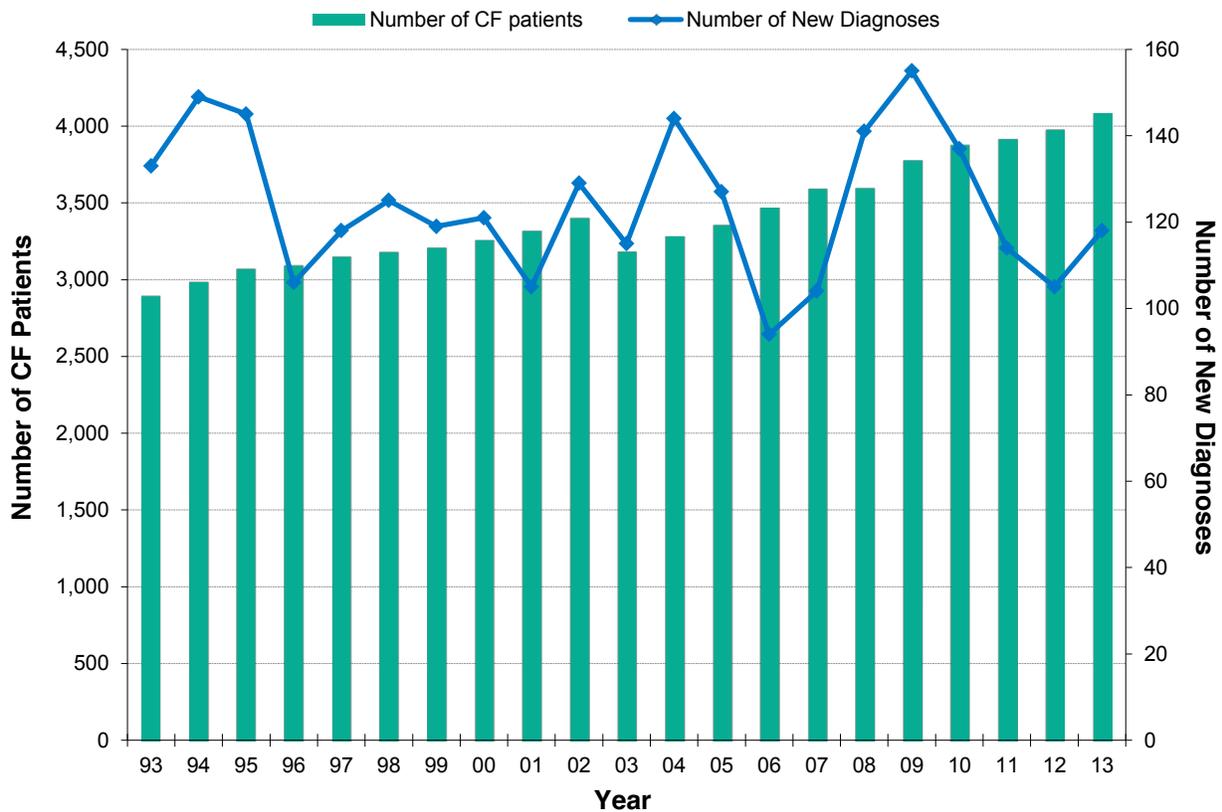
Demographic Data

Number of Canadians with Cystic Fibrosis

A total of 4,077 individuals with cystic fibrosis had clinical records submitted by 42 CF clinics in 2013 (Figure 1). When an individual was seen at multiple clinics in one year, she or he was counted only once (*i.e.* unique individuals) in the graph below. In 2013, there were 118 new diagnoses of cystic fibrosis in Canada.

Figure 1

Total number of CF patients and new diagnoses recorded in the Registry, 1993 to 2013



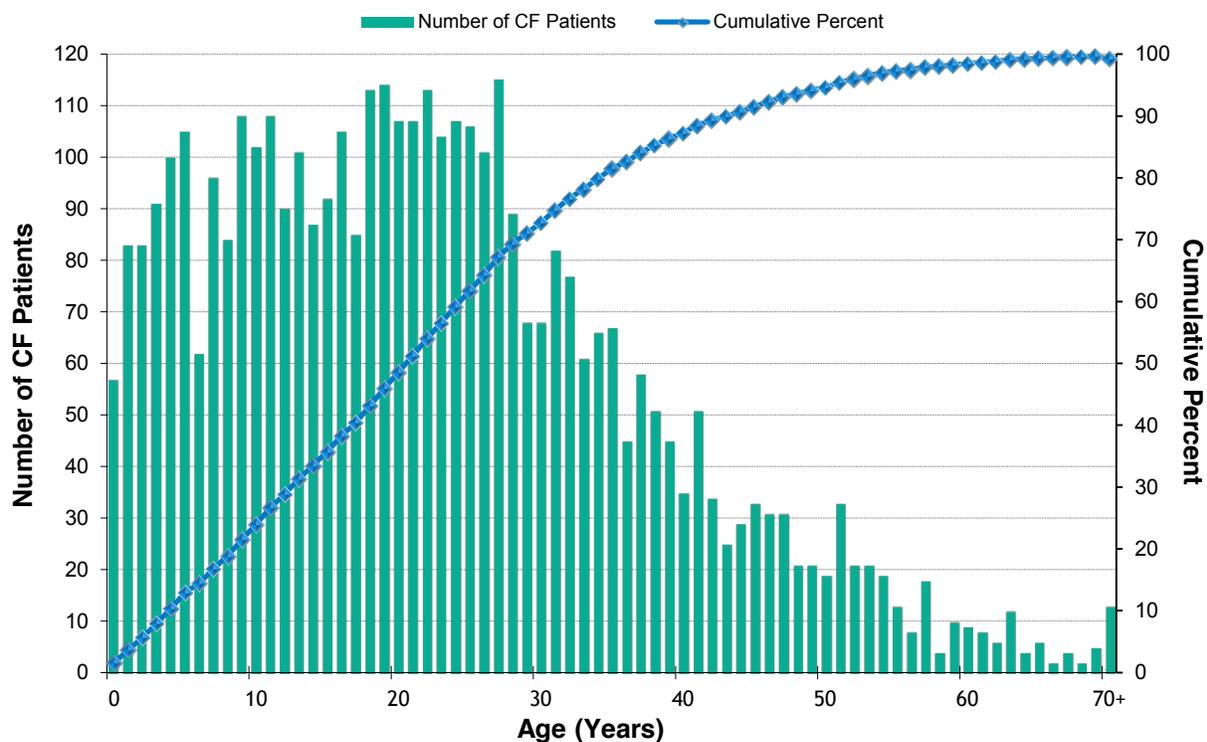
Demographic Data

Ages of Canadians with Cystic Fibrosis

Figure 2 shows the age distribution of the Canadian CF population in 2013. The ages of individuals with cystic fibrosis range from birth to almost 80 years old. The median age of all patients reported on in 2013 was 21.4 years, with 59.6% of individuals over 18 years of age (Figure 3). Males accounted for 52.9% of individuals in the Registry in 2013.

Figure 2

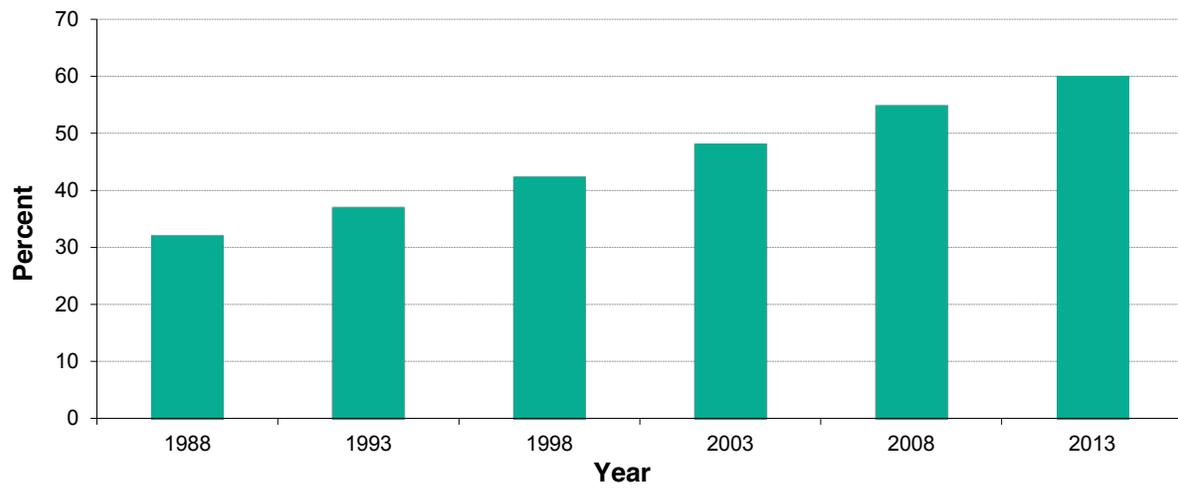
Age distribution of the CF population, 2013



Demographic Data

Figure 3

Proportion of CF individuals 18 years of age or older, 1988 to 2013



Demographic Data

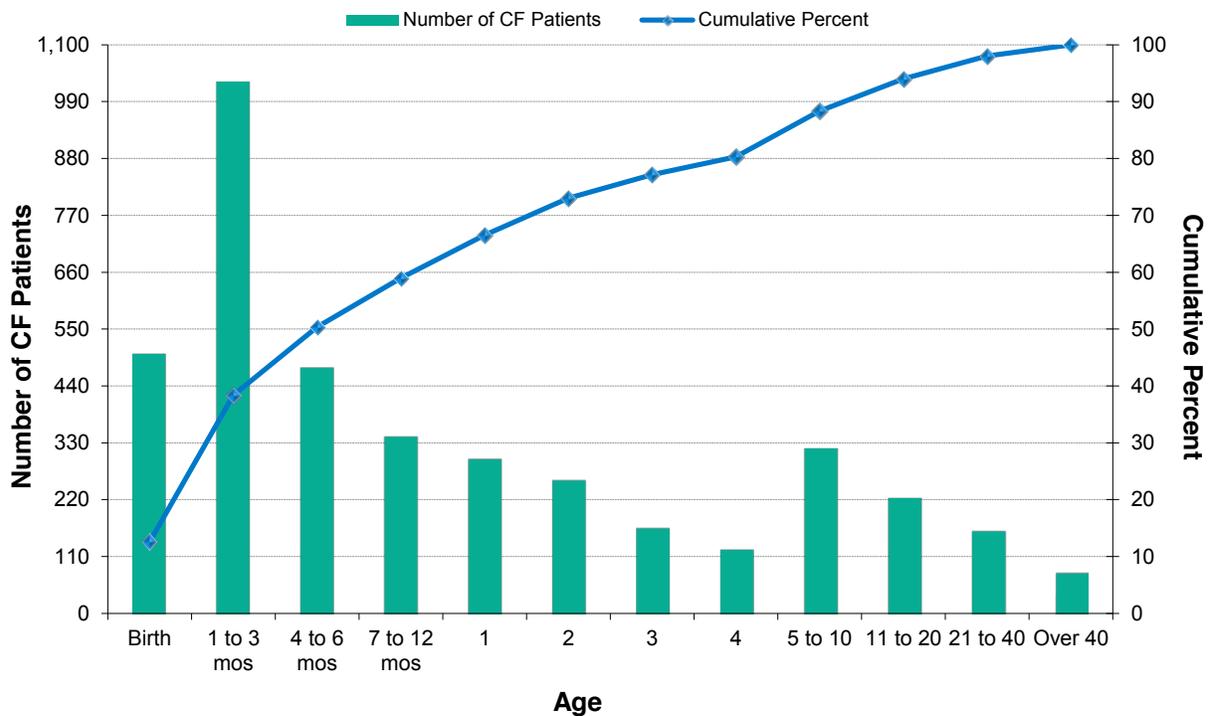
Age at Diagnosis

By one year of age, 66.5% of patients are diagnosed with CF, and 73.0% are diagnosed by the age of two years (Figure 4). Adults diagnosed later in life (over the age of 40) account for almost 2% of all diagnoses.

As newborn screening (NBS) programs for cystic fibrosis continue to be introduced in Canadian provinces (at the time of publication, available in all provinces, except Quebec), the majority of individuals with cystic fibrosis will be diagnosed at birth. Figure 5 shows the percentage of newborns diagnosed through the NBS program over the last 7 years. In 2013, 32.2% of all new diagnoses were made through the NBS program.

Figure 4

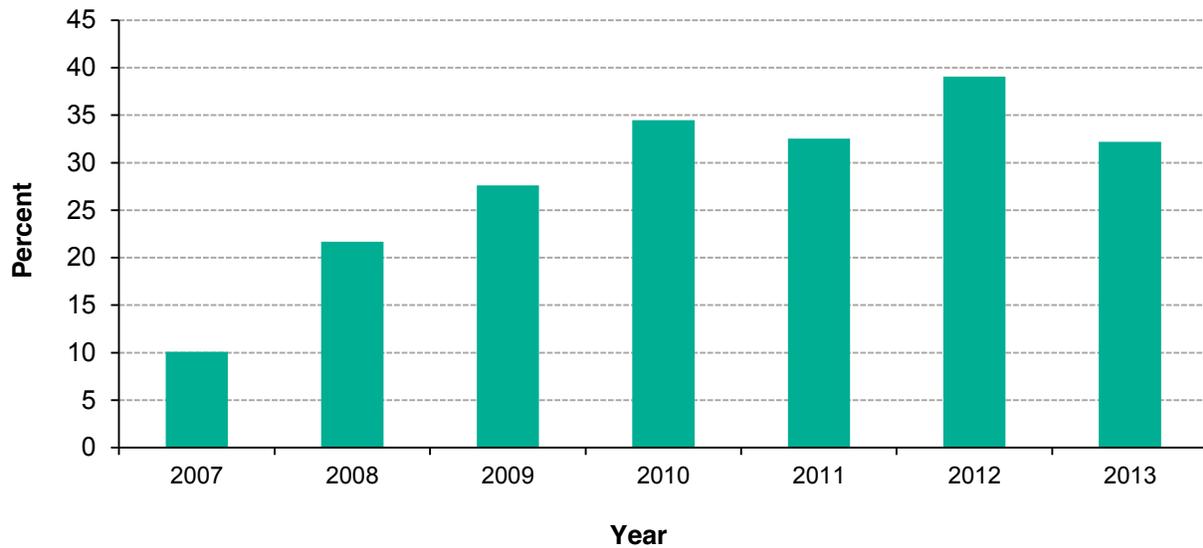
Age at diagnosis, all patients in Registry as of December 31, 2013



Demographic Data

Figure 5

Proportion of all new diagnoses made through the NBS program, 2007 to 2013



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 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 **F508del**. Of those individuals with genetic information recorded within the Registry, 50.0% carry two F508del mutations (Figure 6) and 89.7% carry at least one F508del mutation (Table 1).

Figure 6

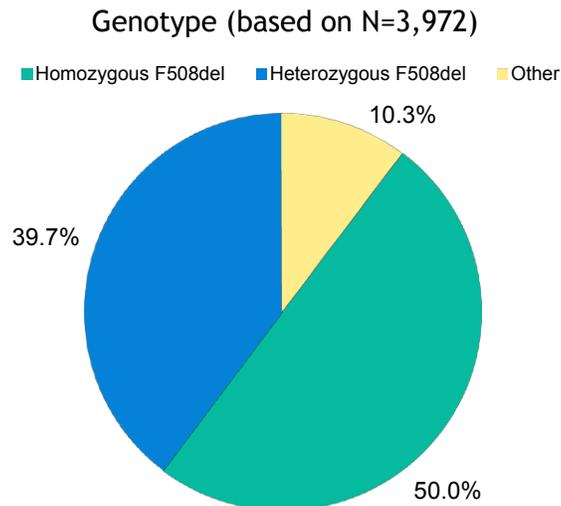


Table 1

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

Genotype	Number	Percentage
F508del	3,563	89.7%
621+1G->T	241	6.1%
G542X	138	3.5%
G551D	122	3.1%
A455E	102	2.6%

Respiratory

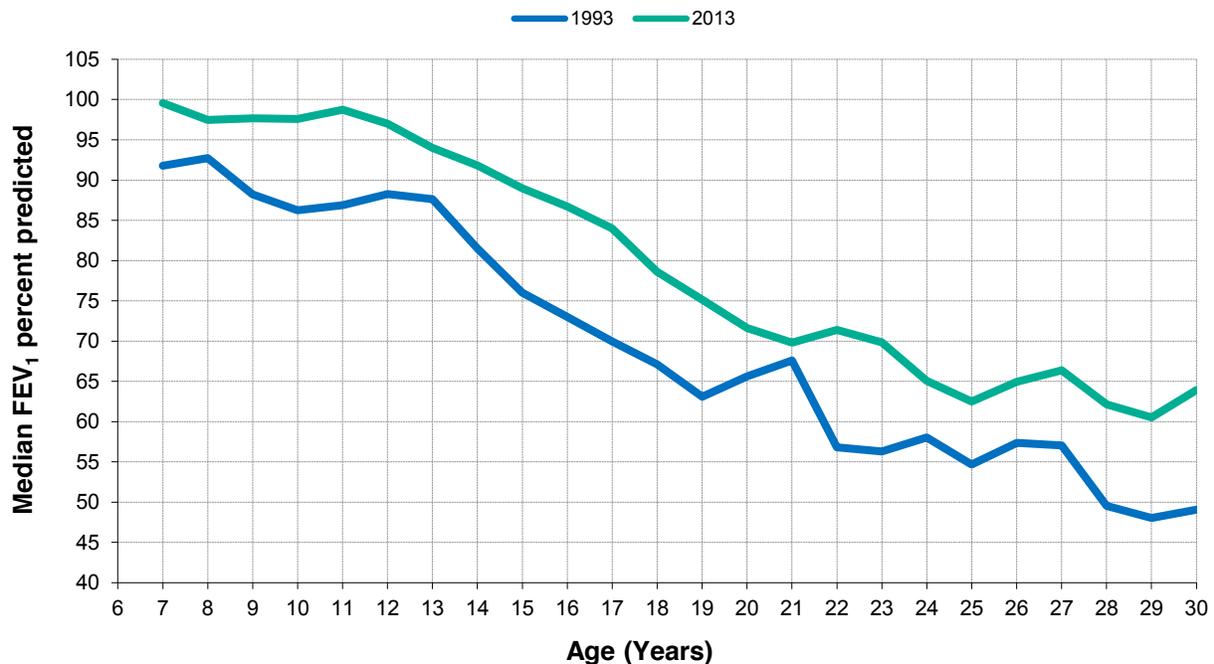
Median FEV₁ Percent Predicted

Respiratory measures are needed to evaluate lung health. FEV₁ (forced expiratory volume in one second) is the volume of air that a person can forcibly blow out in one second. FEV₁ percent predicted for a patient is calculated against the average FEV₁ of a healthy population of similar age, height and sex. The national median FEV₁ percent predicted for adults (ages 18+) is 65.2% and 94.2% for children (ages 6-17).

Median FEV₁ percent predicted has improved over the last two decades but interestingly, the trend in lung function is similar between 1993 and 2013. In 2013, the median FEV₁ percent predicted at 30 years of age was 63.7% compared to 46.0% in 1993. Figure 7 shows in a 2-year moving average window, the median FEV₁ percent predicted from the ages 6 to 30 years. The data for 2013 suggest a very modest average decline in lung function of 0.2% per year from ages 6 to 11 years, but a greater average decline of 2.6% per year is observed from ages 11 to 23. After age 23, lung function seems to stabilize once again with an average decrease of 0.7% per year. This suggests that perhaps the adolescent/young adulthood period is a vulnerable time for individuals with cystic fibrosis.

Figure 7

Median FEV₁ percent predicted vs. age (in a 2-year moving average window), 1993 and 2013



Respiratory

Respiratory Status

Lung function is measured starting at six years of age. Table 2 below summarizes the FEV₁ percent predicted classifications. For children ages 6 to 17 years, the majority (58.3%) have normal lung function (FEV₁ greater than or equal to 90% predicted). For adults, the majority (38.5%) have lung function classified as 'moderate' severity (Figure 8). These data are similar to those observed in 2012.

Figure 8

Respiratory status of children and adults with CF, 2013

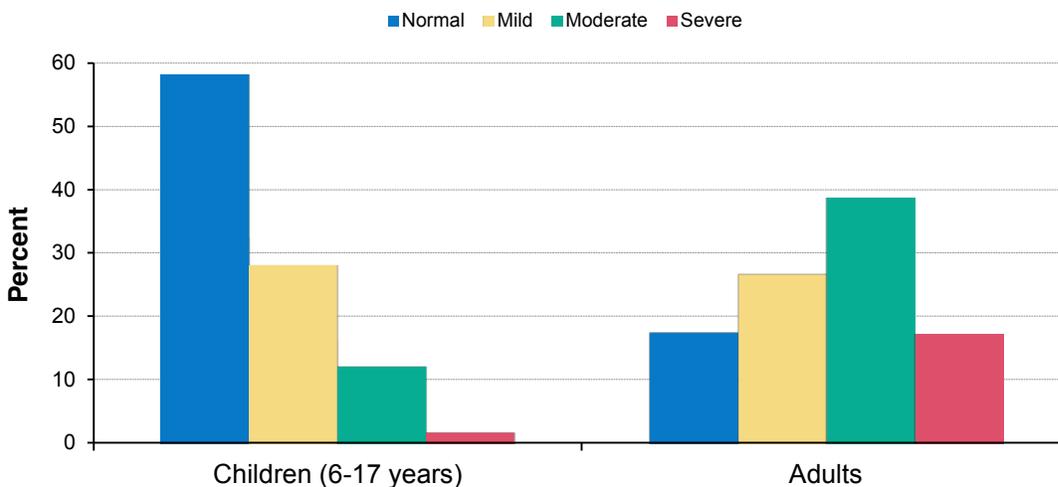


Table 2

FEV₁ percent predicted classification

Classification	Range
Normal	≥ 90 %
Mild	70 - 89%
Moderate	40 - 60 %
Severe	< 40 %

Respiratory

Respiratory Status by Sex

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

Figure 9

Respiratory status of children (6 to 17 years) with CF, by sex, 2013

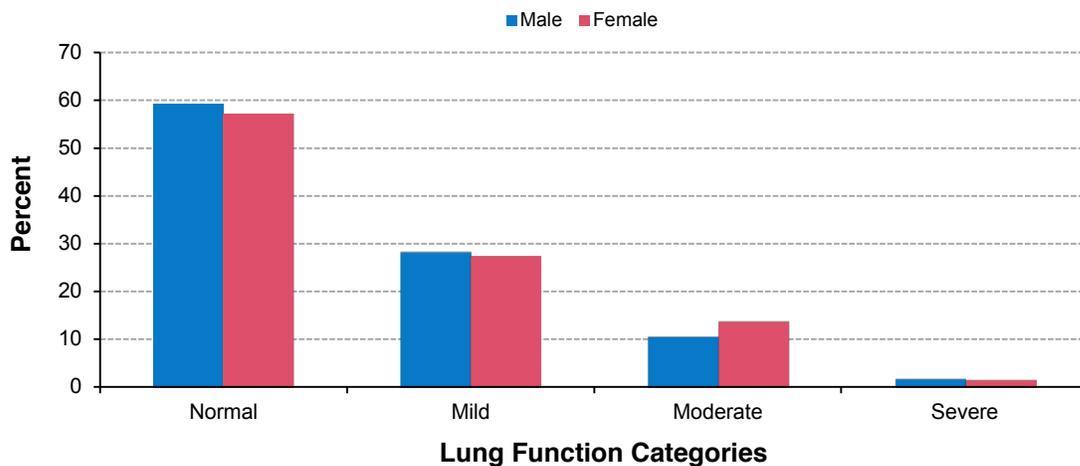
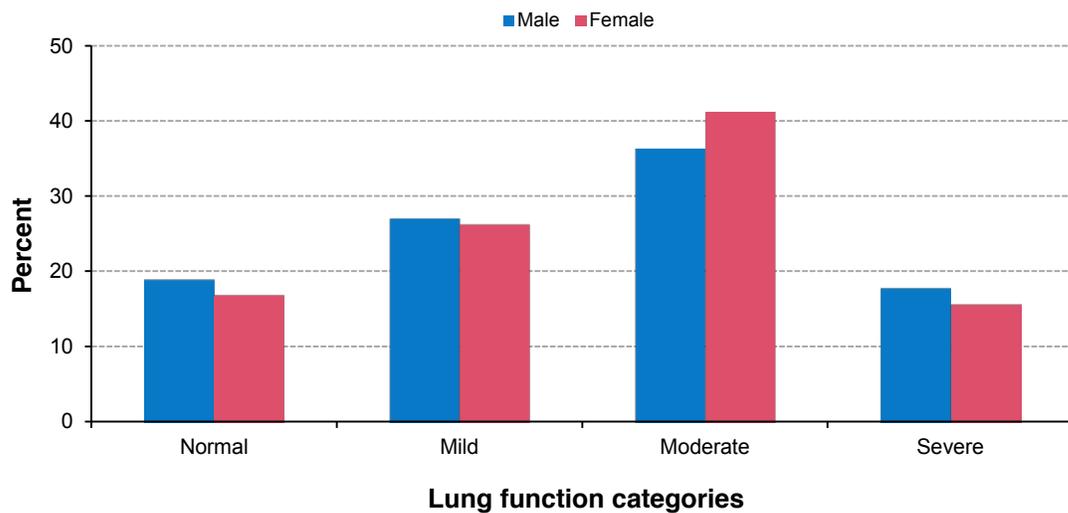


Figure 10

Respiratory status of adults with CF, by sex, 2013



Nutrition

Pancreatic Status

Malnutrition is common in individuals with cystic fibrosis as a result of pancreatic insufficiency. Pancreatic enzymes are given as supplements to help with digestion and absorption of nutrients. In 2013, 86.2% of individuals with cystic fibrosis were taking supplemental pancreatic enzymes (pancreatic insufficient), whereas 13.8% did not require oral pancreatic enzyme supplementation to digest their food (pancreatic sufficient) (Figure 11).

For those individuals 40 years of age or older, 32.6% were pancreatic sufficient (Figure 12). This is 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

■ Pancreatic Insufficient ■ Pancreatic Sufficient

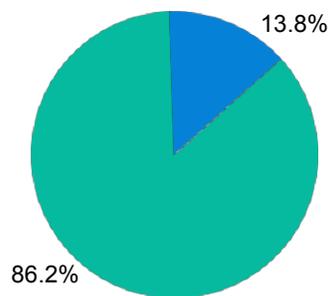
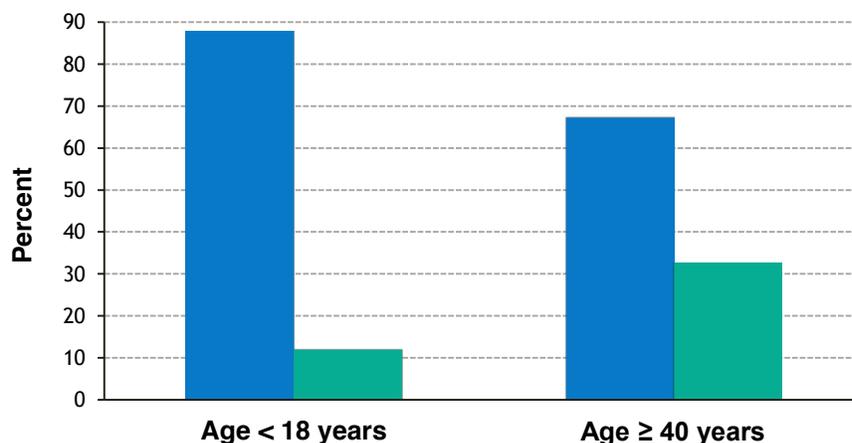


Figure 12

Pancreatic status by age group, 2013

■ Pancreatic Insufficient ■ Pancreatic Sufficient



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 metres). Typically, this is calculated for adults only because they have attained their maximal height. Children are rapidly growing and therefore, one must consider the child's age when assessing their nutritional status. The national median BMI for adults (≥ 18 years) is 22.1 kg/m^2 .

Table 3 below describes the BMI classifications and their BMI ranges. In 2013, the majority (60.0%) of the adult CF population had an adequate weight while 23.5% were considered underweight and 4.3% were considered obese (Figure 13).

Figure 13

BMI classification for adults with cystic fibrosis, 2013

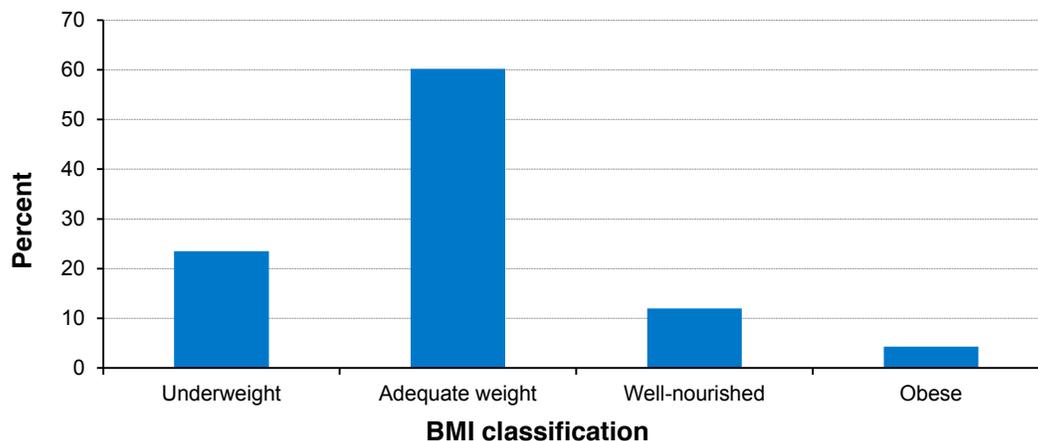


Table 3

BMI classification

Classification	Range
Underweight	$< 20.0 \text{ kg/m}^2$
Adequate weight	$20.0 - 25.9 \text{ kg/m}^2$
Well-nourished	$26 - 29.9 \text{ kg/m}^2$
Obese	$\geq 30 \text{ kg/m}^2$

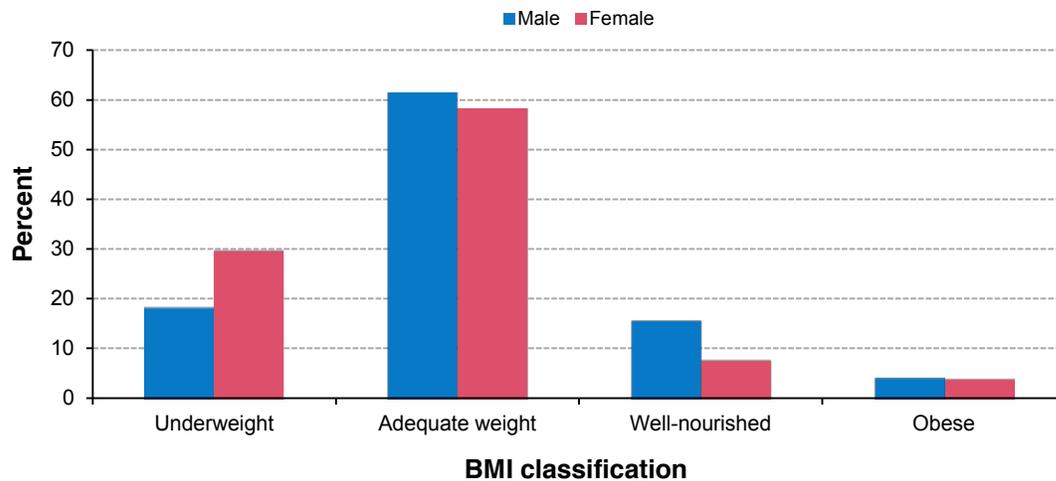
Nutrition

BMI by Sex

Figure 14 shows the breakdown of BMI categories (see previous page for definitions) for adult males and females. A larger proportion of females are considered underweight (BMI < 20 kg/m²) compared to males. Individuals who are muscular may have a BMI between 26-29 kg/m² due to increased weight from high muscle mass.

Figure 14

BMI classification for adults with cystic fibrosis, by sex, 2013



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. Table 4 summarizes the BMI percentile classification categories. The national median BMI percentile for children is 44.0 (ages 2-17). The majority (85.1%) of children with CF have a healthy weight with a small proportion considered overweight (7.0%) or obese (2.5%). The national goal for children with cystic fibrosis (ages 2-17) is 50th BMI percentile. Of all children with cystic fibrosis, 43.7% are above the national goal with 50.9% being male.

Figure 15

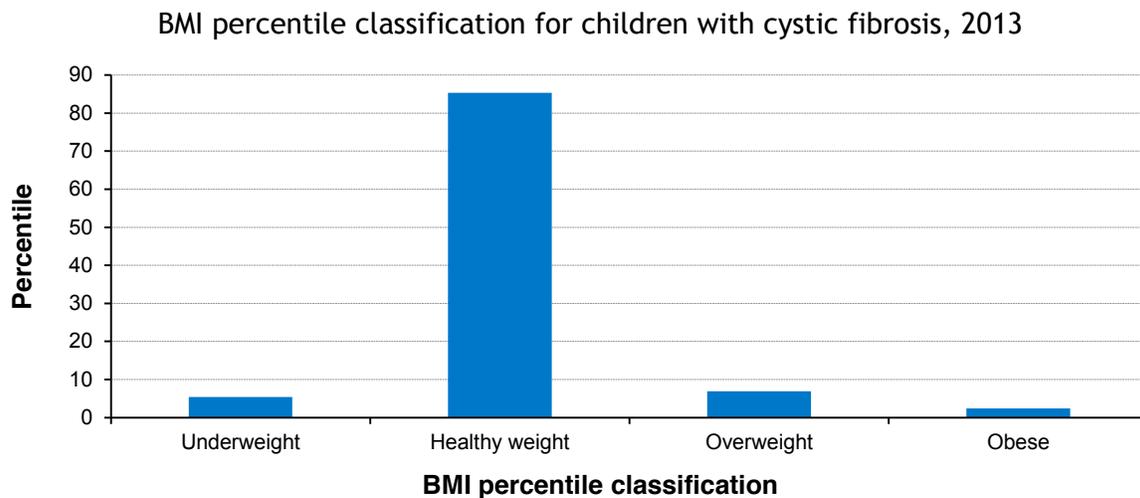


Table 4

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

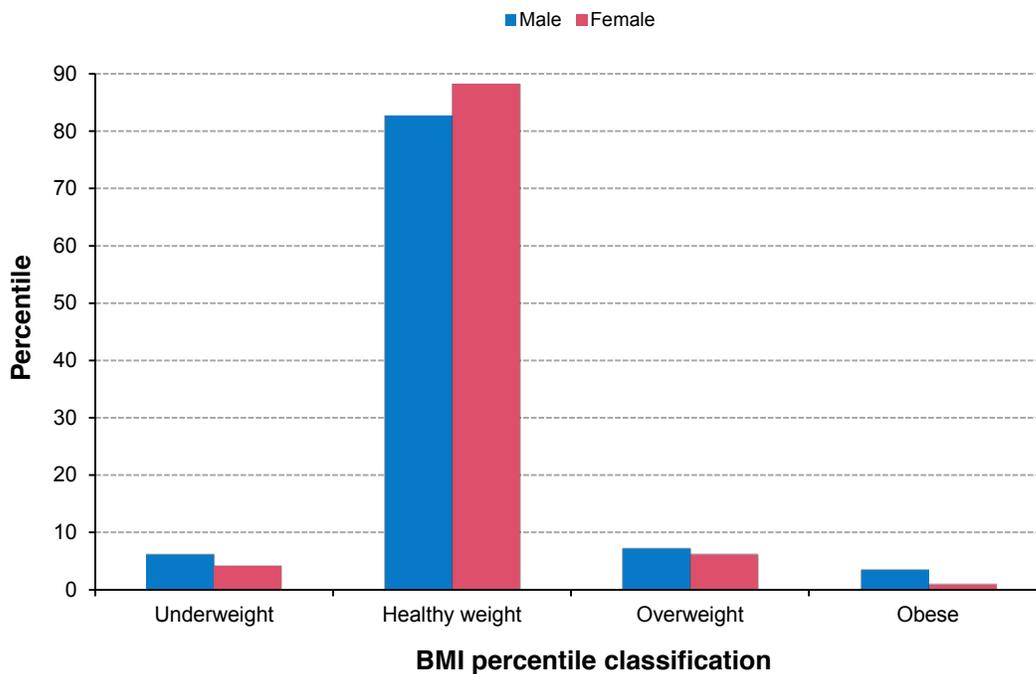
Nutrition

BMI Percentile by Sex

Figure 16 shows the breakdown of BMI percentile classifications for males and females between the ages of 2 and 17. While there are slightly more females with a healthy weight than males (4.9% difference), there are minor disparities between males and females across the remaining BMI percentile categories (range of 1.2-2.4% across the remaining categories).

Figure 16

BMI percentile classification for children with cystic fibrosis, by sex, 2013

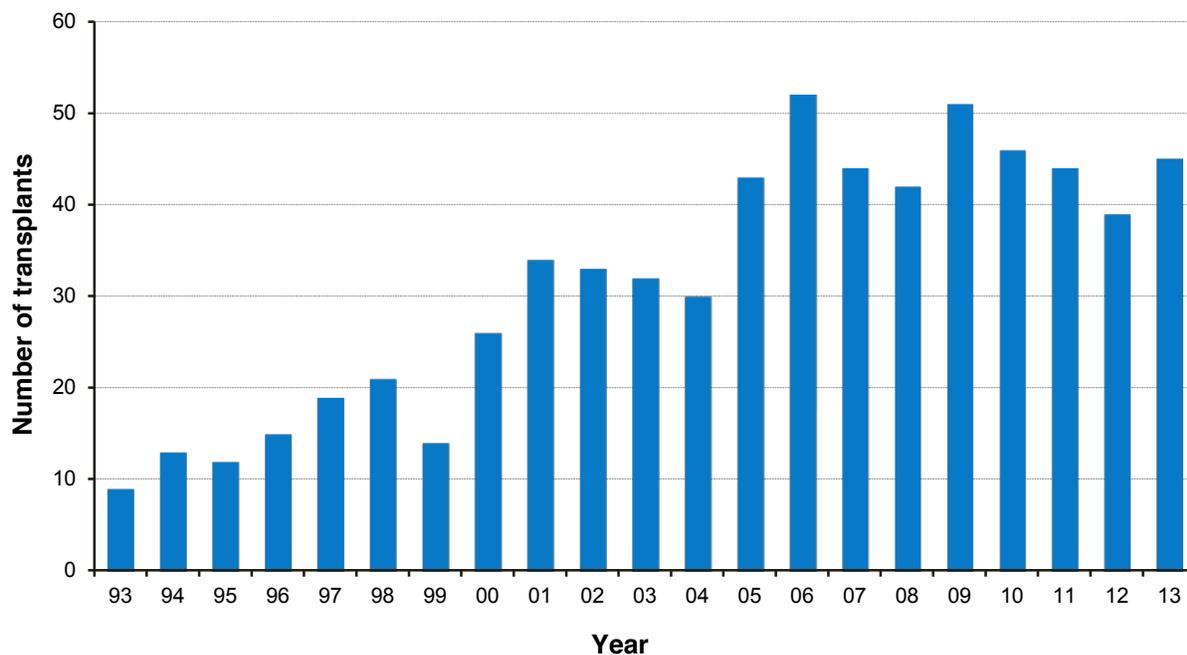


Transplants

Figure 17 shows the number of transplants carried out per year as reported in the Registry. In 2013, 44 CF patients received a transplant. 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, 2013, there were 638 CF patients recorded in the Registry as having received one or more transplants and of those individuals, 384 individuals were reported as being alive.

Figure 17

Number of transplants per year, 1993 to 2013



Microbiology

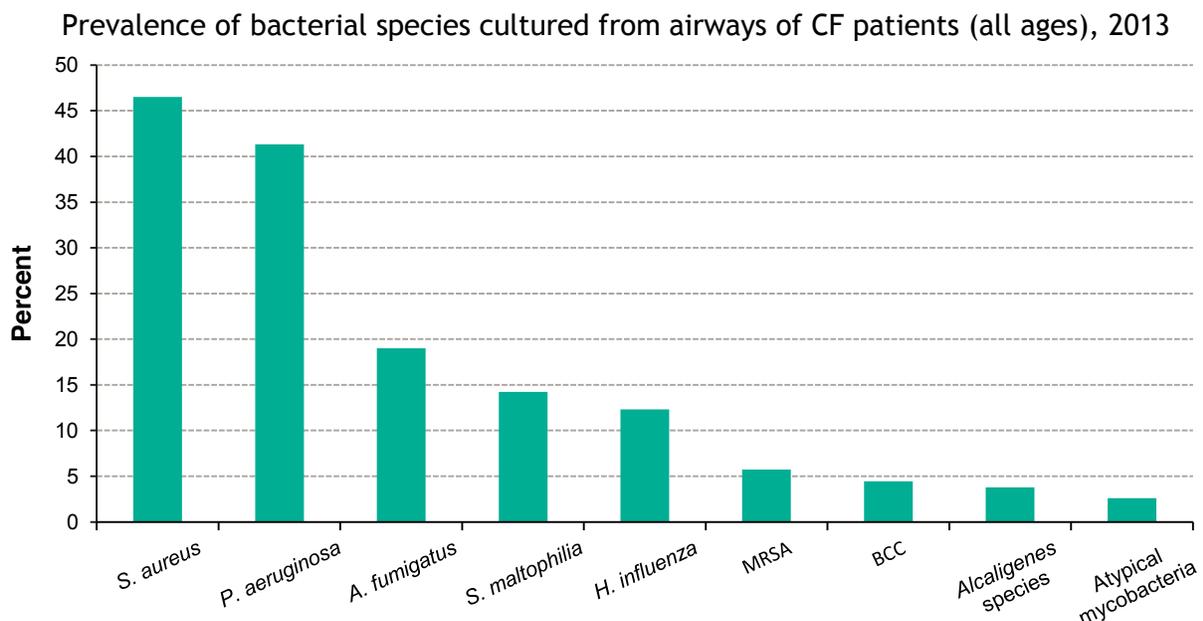
Bacterial Species and Respiratory Infections

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 aureus* (MRSA) has increased over the last several years (Figure 19). The largest increase was seen in *Aspergillus* species. The higher prevalence may be partly due to increased surveillance for these organisms. The prevalence of MRSA has gradually been increasing and in 2013, 6.0% of Canadians had a positive sputum sample for MRSA. MRSA and Atypical mycobacteria data were not collected in the Registry prior to 2003 and 2011, respectively.

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

Figure 18



Microbiology

Figure 19

Prevalence of respiratory infections, 2008-2013

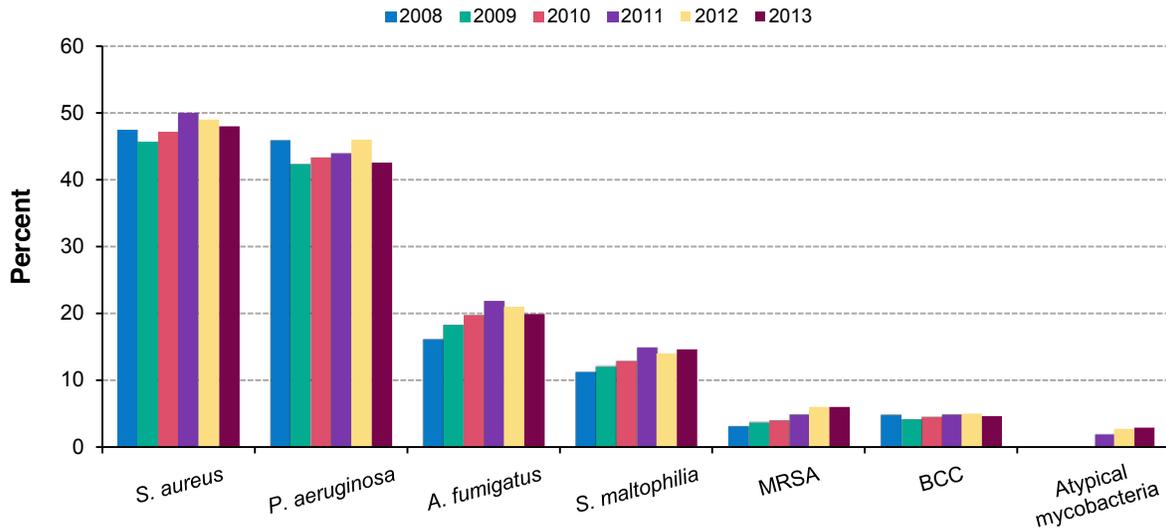
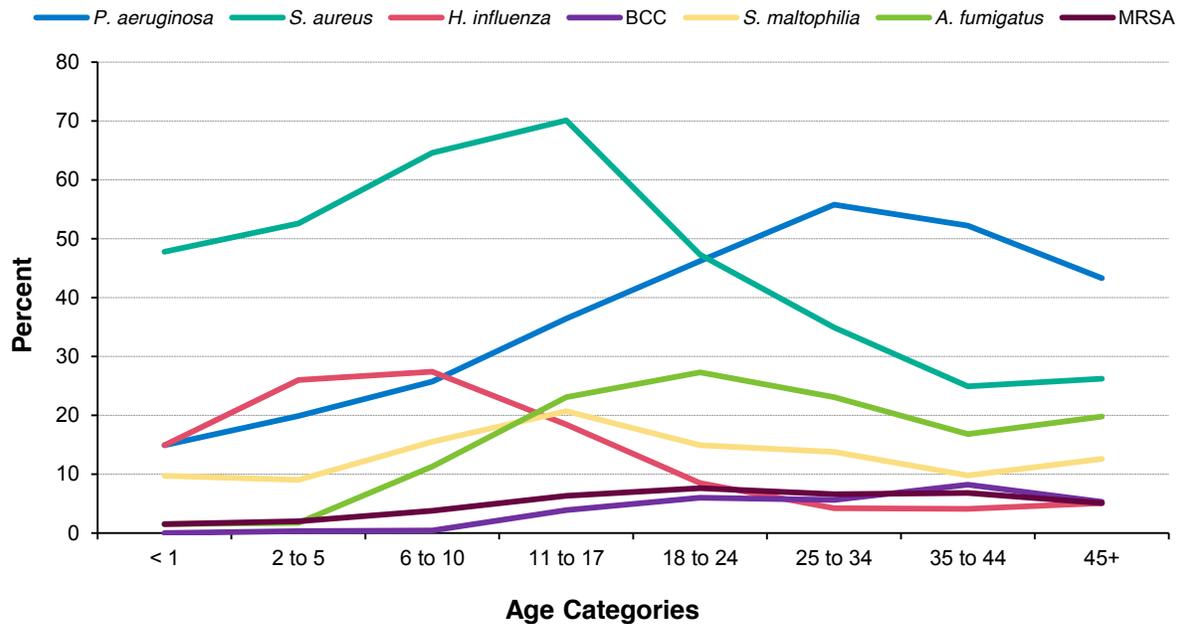


Figure 20

Age-specific prevalence of respiratory infections in CF patients, 2013



Microbiology

Burkholderia cepacia complex (BCC)

There were 177 patients with *Burkholderia cepacia* complex (BCC) species in 2013 (Figure 21). *B. cenocepacia* and *B. multivorans* were the two most common types of BCC species. Of the patients with BCC in 2013, 88.2% were adults (Figure 22). Not all BCC bacteria were genotyped and therefore, 10.7% of the BCC species in the Registry were classified as unknown. The prevalence of *B. gladioli* was 4.8%, though it was not included in Figure 21 because it is not officially recognized as part of the BCC.

Figure 21

Burkholderia cepacia complex species, 2013

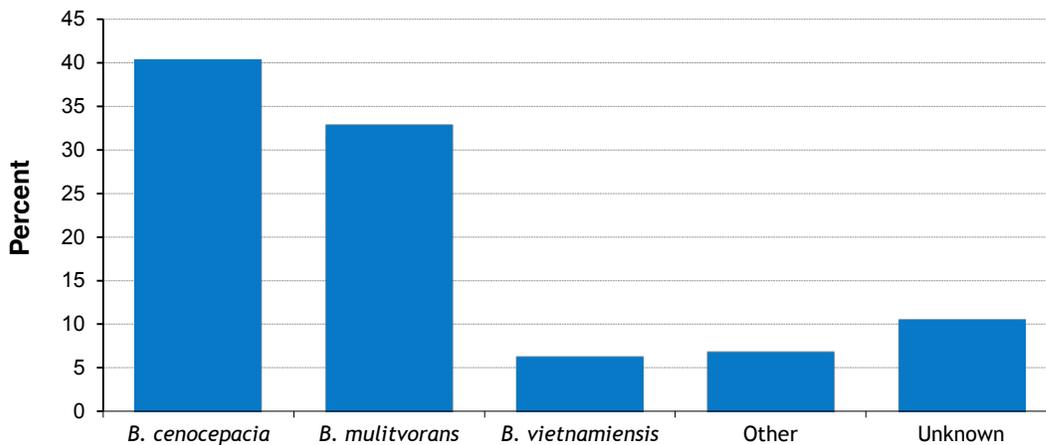
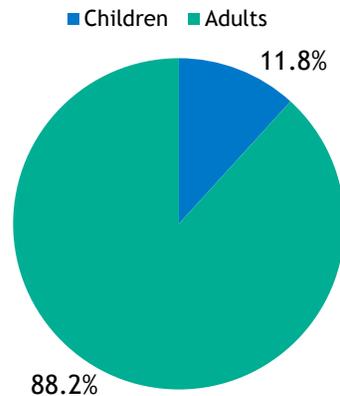


Figure 22

Burkholderia cepacia complex distribution by age, 2013

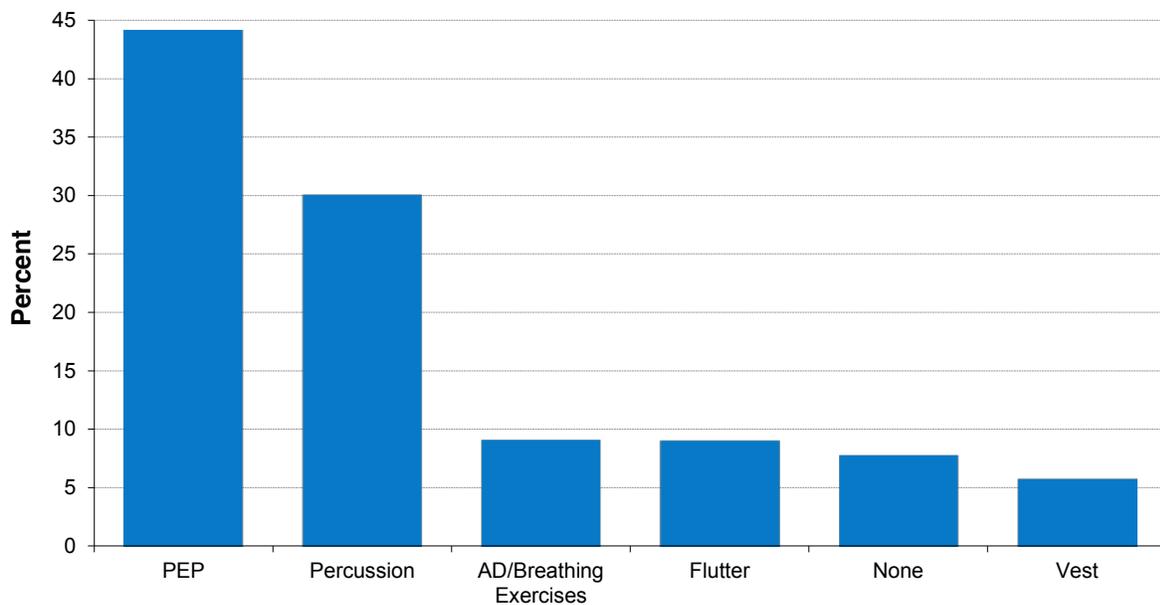


Physiotherapy

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 physiotherapy is needed since transplanted lungs do not have cystic fibrosis.

Figure 23

Physiotherapy (based on N = 3,804), 2013

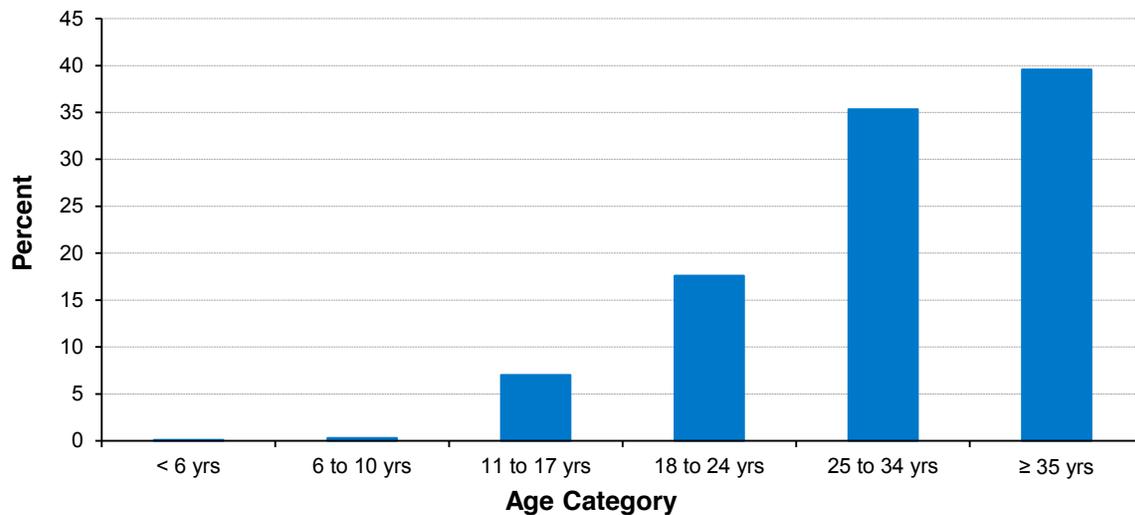


CF-Related Diabetes (CFRD)

The prevalence of CFRD increases with age (Figure 24). In 2013, CFRD was reported in 23.0% of individuals with cystic fibrosis. Of those individuals, 51.2% were female and 39.7% were 35 years of age or older.

Figure 24

Proportion of patients with CFRD by age, 2013



Hospitalization and Home IV

Pulmonary Exacerbation is the Leading Cause

In 2013, 1,858 hospitalizations were recorded in the Registry (Table 5). The most common reason for admission was a pulmonary exacerbation. In total, 676 courses of home IV therapy were recorded in the Registry.

Table 5

Number of hospital days and home IV courses, 2013

	Total Number
Hospital Days	24,619
Hospitalizations	1,858
Clinic Visits	16,687
Home IV Courses	676
Home IV Days	11,548

Survival

There were 40 deaths recorded in the Registry in 2013. Since there are relatively few deaths per year, the sum of all deaths from 2009 to 2013 has been included in Figure 25. The median age at death in 2013 was 35.1 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. It is calculated using only those individuals who have died. In other words, of those who died, half died before the median age at death and half died later than the median age at death. The most common cause of death was related to pulmonary complications.

Figure 25

Age at death, 2009 to 2013

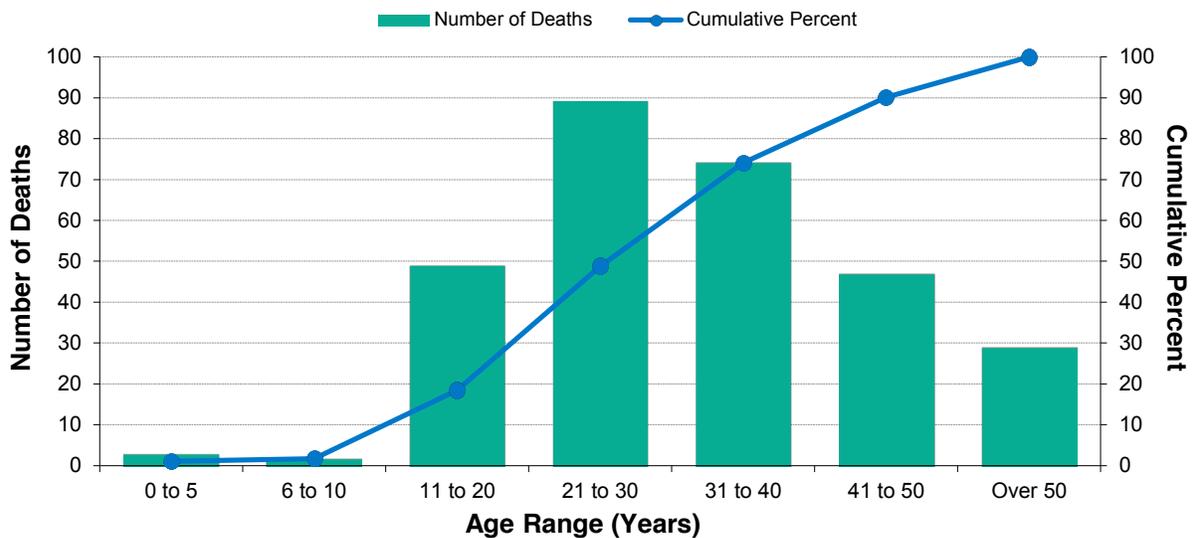
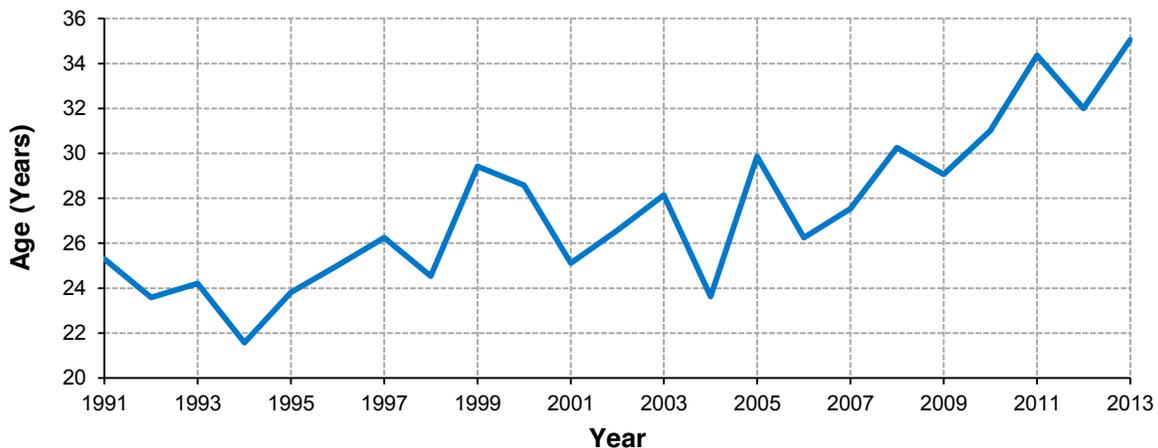


Figure 26

Median age at death, 1991 to 2013



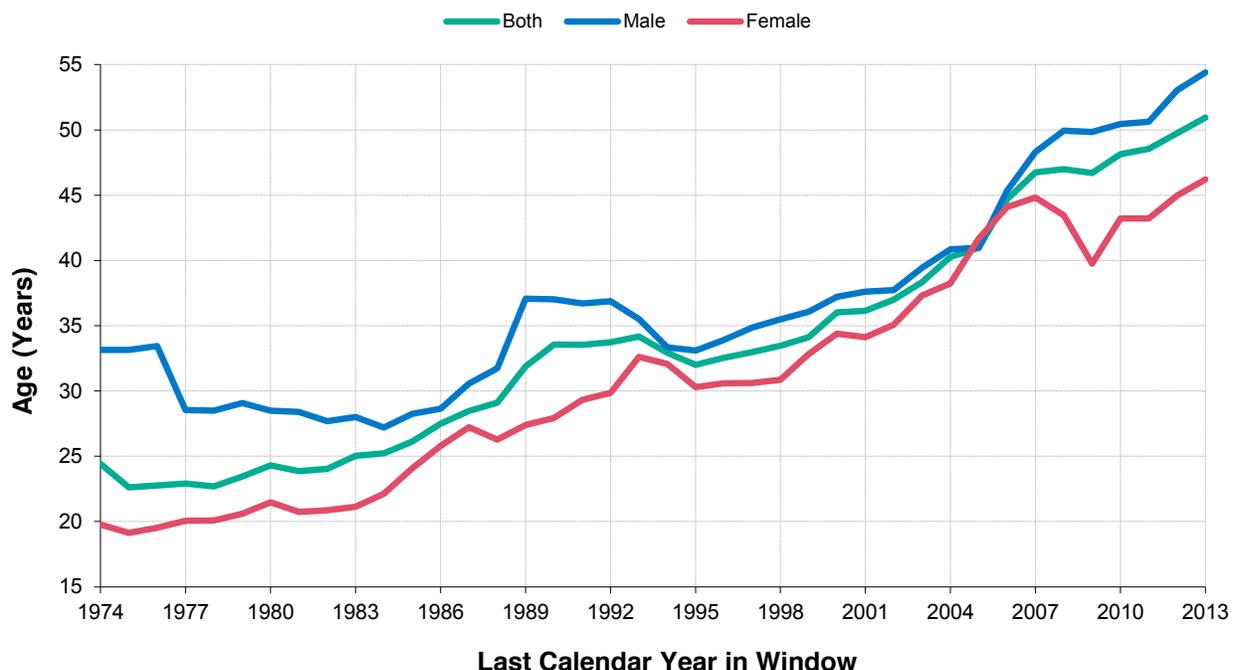
Survival

Median Age of Survival

The median age of survival for Canadians with cystic fibrosis is currently estimated to be **50.9 years of age** (Figure 27). Males continue to have a higher median age of survival compared to females. The cause of lower survival in females is not well understood but has been documented in the published CF literature. Because there are relatively few deaths per year in Canada, a 5-year window was used to calculate the median age of survival. This allows for more stable estimates over time.

Figure 27

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



Survival

Median Age at 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 a given year**. In other words, of those who died in the year, half died before the median age at death and half died later than the median age at death.

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

Median Age of Survival

Median age of survival is calculated based on cross-sectional data (i.e. data taken across different age groups) of the CF population and takes into consideration data from both individuals who have died AND those who are still alive. It is the **estimated age beyond which 50 percent of the CF population would be expected to live, assuming the mortality rate in CF remained constant**. This is NOT the age at which people with CF would be expected to die, (i.e. how long someone can expect to live, on average - see *life expectancy* below). Median age of survival is simply *one way* to evaluate survival in the CF population; however, there are other measures that provide us with additional information about how long people with CF are living (for example, median age at death and annual death rate).

When we say that the median age of survival in 2013 is 50.9 years, we are saying that if a child with CF is born in Canada in 2013, they have a 50 percent probability of living beyond 50.9 years of age based on current mortality rates. In other words, half of the CF population would be expected live to an age older than 50.9 years. Of course, mortality rates are not static and are constantly changing as new therapies and medicines for CF become available. Thus, this estimate is a reflection of the most accurate data that is available today.

Keep in mind that these survival estimates apply to a population of individuals and do not necessarily apply to any one individual.

Life Expectancy

The life expectancy is the average age to which someone can be expected to live. In other words, it is the **expected average length of life based on current age-specific mortality rates**. For the general population born today, life expectancy in Canada is 77 years for males and 82 years for females based on data from Statistics Canada. This means that, *on average*, a male baby born today will be expected to live 77 years and a female baby, *on average*, will be expected to live to 82 years of age. In comparison, the median age of survival is the estimated age beyond which 50 percent of the population will live - it is *not* an average.

Life expectancy is *not* the same as median age of survival. This measurement uses the mean (average) and not the median. For example, the average life expectancy for women in

Survival

Canada is 82 years of age. This means that a baby girl born today will be expected to live until the age of 82 years, *on average* (i.e. some girls will die before the age of 82 years and some will live beyond the age of 82 years but, on average, women will live 82 years).

It is possible to calculate the life expectancy in CF but we do not usually do it because it is influenced by extreme values more so than the median age of survival. For example, the life expectancy may change significantly if one or two people in the population lived until a very old age because it is calculating a mean age whereas the median age is less sensitive to extreme values and is a more robust measurement.

Summary Data

Table 6

Summary data from Registry, 1988 to 2013

	1988	1993	1998	2003	2008	2013
General Profile						
Number of patients with clinical records in reporting year (n)	2,512	2,895	3,181	3,180	3,594	4,077
Male, % of total patients	53.3	53.6	53.5	53.3	53.4	52.9
Age, mean (yr)	13.9	15.4	17.1	18.9	20.9	22.6
Age, median (yr)	13	14	15	17	19	21.4
% over 18 yrs	32.3	37.2	42.5	48.3	55	59.6
Race, % Caucasian	97.7	97.4	96.8	95.8	94.1	92.0
Black	0.4	0.4	0.6	0.7	0.8	0.8
Asian	0.4	0.3	0.4	0.4	0.5	0.7
First Nations People	0.4	0.6	0.7	0.7	0.8	1.0
South Asian	---	---	---	---	---	0.1
Other	1	1	1.3	1.5	1.8	0.1
Unstated	0.3	0.3	0.3	0.1	0.1	0.2
Transplants (#)	2	9	21	32	42	44
Pancreatic insufficient (%)	85	92.7	92.3	91.4	89.3	86.2
% with genotype analysis	---	---	---	---	93	97
Diagnosis						
Age at diagnosis, mean (yr)	2.2	2.4	2.7	3.2	3.7	4.0
median (mo)	7	6	6	6	7	6
New diagnoses in year (n)	128	135	125	117	146	118
% with meconium ileus at birth	15.3	19.8	16	19	9.9	17.5
Survival/Mortality						
Age at death, mean (yr)	20.3	23.4	26.5	29	30.5	35.9
median (yr)	19.5	24	24	28	30	35.1
Total deaths (n)	58	56	65	46	53	40
Crude mortality rate (%)	2.3	1.9	2	1.4	1.5	1.0
Median age of survival (yr)	29.1	34.2	33.5	38.3	47.0	50.9
Male	31.7	35.5	35.5	39.4	49.9	54.4
Female	26.3	32.6	30.9	37.3	43.4	46.2
Nutritional Markers						
≥ 18 yrs of age: n (%) in BMI categories						
< 20	333 (44%)	405 (41%)	437 (35%)	374 (28%)	483 (26%)	553 (24%)
20-25.9	388 (52%)	504 (51%)	683 (55%)	811 (60%)	1108 (60%)	1413 (60%)
26-29.9	25 (3%)	61 (6%)	92 (7%)	139 (10%)	184 (10%)	272 (12%)
≥ 30	4 (1%)	10 (1%)	19 (2%)	35 (3%)	58 (3%)	99 (4%)

Summary Data

Males < 23 BMI	338 (78%)	390 (71%)	446 (68%)	437 (60%)	582 (59%)	656 (52%)
Females < 22 BMI	240 (75%)	321 (74%)	395 (69%)	384 (61%)	502 (60%)	625 (58%)
2 - 17 yrs of age: n (%) in BMI categories						
< 50 th BMI percentile	943 (62%)	1032 (61%)	1053 (62%)	874 (58%)	846 (55%)	863 (56%)
< 25 th BMI percentile	520 (34%)	586 (35%)	600 (35%)	454 (30%)	452 (30%)	444 (29%)
<u>Pulmonary Function</u>						
% predicted FEV ₁ , mean	70.7	70.9	72.4	73.4	72.0	74.2
% predicted FEV ₁ , median	72.6	72.9	73.8	74.9	72.9	76.7
n (%) FEV ₁ % predicted categories for patients ≥ 18 years:						
Normal: ≥ 90%	80 (12%)	101 (11%)	149 (13%)	171 (13%)	247 (14%)	421 (18%)
Mild: 70-89%	136 (21%)	208 (23%)	247 (21%)	311 (24%)	449 (25%)	628 (27%)
Moderate: 40-69%	234 (37%)	352 (38%)	517 (44%)	559 (43%)	770 (43%)	878 (38%)
Severe: < 40%	191 (30%)	260 (28%)	274 (23%)	259 (20%)	323 (18%)	373 (16%)
n (%) FEV ₁ % predicted categories for ages 6 to 17:						
Normal: ≥ 90%	345 (38%)	439 (42%)	518 (44%)	526 (51%)	515 (50%)	596 (60%)
Mild: 70-89%	271 (30%)	317 (30%)	360 (31%)	296 (29%)	308 (30%)	266 (27%)
Moderate: 40-69%	227 (25%)	240 (23%)	233 (20%)	183 (18%)	192 (19%)	111 (11%)
Severe: < 40%	68 (7%)	61 (6%)	57 (5%)	25 (2%)	22 (2%)	15 (2%)
n (%) on oxygen	---	---	1 (0%)	72 (2%)	100 (3%)	136 (3%)
n (%) on BiPAP	---	---	---	---	---	29 (1%)
<u>Microbiology</u>						
n (%) with positive culture (first culture of the year [1976-1996], any culture in the year [starting in 2001]):						
<i>Pseudomonas aeruginosa</i>	1146 (46%)	1229 (42%)	1384 (44%)	1450 (46%)	1673 (47%)	1734 (43%)
<i>Staphylococcus aureus</i>	631 (25%)	861 (30%)	1036 (33%)	1414 (44%)	1752 (49%)	1954 (48%)
<i>Haemophilus</i> species	386 (15%)	401 (14%)	310 (10%)	494 (16%)	625 (17%)	511 (13%)
<i>Stenotrophomonas maltophilia</i>	---	---	102 (3%)	262 (8%)	413 (11%)	595 (15%)
<i>Aspergillus</i>	---	---	---	376 (12%)	609 (17%)	811 (20%)
MRSA	---	---	---	34 (1%)	117 (3%)	244 (6%)
<i>Alcaligenes (achromobacter)</i> species	---	---	---	---	---	159 (4%)
Atypical mycobacteria	---	---	---	---	---	116 (3%)
<i>Burkholderia cepacia</i> complex	227 (9%)	275 (9%)	209 (7%)	165 (5%)	170 (5%)	184 (5%)
<i>B. cenocepacia</i>	---	---	---	---	---	76 (40%)
<i>B. multivorans</i>	---	---	---	---	---	62 (33%)
<i>B. vietnamiensis</i>	---	---	---	---	---	12 (6%)
<i>B. gladioli</i>	---	---	---	---	---	9 (5%)
<i>B. cepacia</i> Other	---	---	---	---	---	13 (7%)
Unknown	---	---	---	---	---	20 (11%)

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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	<i>Centre de santé et de services sociaux de Gatineau, Hull</i>
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Foothills Hospital, Calgary	Montreal Chest Institute, Montreal
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Royal University Hospital, Saskatoon	<i>Hôtel-Dieu de Montréal, Montréal</i>
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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

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