The Canadian Cystic Fibrosis Registry

2011 Annual Report

Cystic Fibrosis

Cystic fibrosis 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 non-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 CF 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 $140 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 CF care;
- Promoting public awareness of cystic fibrosis; and
- Raising and allocating funds for these purposes.

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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 (paediatric and adult) within Canada, it is felt that the Canadian Registry is very complete (i.e. it includes data on virtually all Canadians with cystic fibrosis) — giving a comprehensive picture of the CF population in this country.

The Registry is used both 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.

In 2011, the data captured within the Registry were expanded to allow for more detailed information such as medication use, complications, and multiple clinical measurements per year rather than annual data. This will give a better understanding of how care is being delivered to the Canadian CF population. Further, this information can be used by advocacy groups to improve health care delivery.

With the continued cooperation and participation of clinical personnel and Canadian CF patients, along with the generous support of Cystic Fibrosis Canada's many friends and donors, it will be possible to ensure that data remain available and worthy of study in the future.

Highlights

- Approximately **4,000 Canadians** with cystic fibrosis received care at one of the **42 specialized CF clinics** based in hospitals across Canada
- At least two children per week were diagnosed with cystic fibrosis in 2011; of the **114 new diagnoses**, 55 were under 6 months of age
- Nearly **60%** of all people with cystic fibrosis in Canada **are adults**
- Cumulatively, CF patients spent over **25,000 days in hospital** and attended over **15,000 clinic visits** in 2011
- Cumulatively, CF patients underwent **792 courses of home IV therapy** in 2011
- The **median age of survival** for Canadians with cystic fibrosis is currently estimated to be **48.5 years of age**
- Of the 45 patients who died in 2011, **half were under 34 years old**
- FEV1 (a measure of lung function) is improving for persons with cystic fibrosis: **half of all 30 year olds** with cystic fibrosis have an **FEV1 of approximately 72%** predicted in 2011 compared to 46% two decades ago
- **86%** of Canadians with cystic fibrosis must take **pancreatic enzymes** to digest food and absorb nutrients
- **30% of female adults** with cystic fibrosis and **17% of male adults** with cystic fibrosis are classified as **underweight**
- **44 CF patients received transplants** in 2011
- Nearly **half of all patients** with cystic fibrosis are **infected with harmful bacteria** such as *Staphylococcus aureus* and/or *Pseudomonas aeruginosa* in their lungs
- **16% of CF patients have CF-related diabetes**, and approximately 38% 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 **91.5% 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

For more than 40 years, Cystic Fibrosis Canada has published a Canadian patient data registry — offering an outstanding resource of important CF patient data.

You may notice something different about this year's report. Known for years as the *Canadian Patient Data Registry (CPDR),* the Registry will be named the *Canadian Cystic Fibrosis Registry*, starting with this 2011 annual report publication. The disease itself is now clearly denoted.

I am also pleased to inform you that beginning in July 2013, CF clinic specific data from the *Canadian Cystic Fibrosis Registry* will become available to the public for the first time in our history. This positive step forward demonstrates our enhanced focus on leading in quality improvement and excelling in CF care, and ultimately moves us closer to our goal of establishing national standards for CF care and treatment across Canada.

Our ability to bring you the *Canadian Cystic Fibrosis Registry* would be impossible without generous funding from our donors and partners and we are tremendously grateful for their ongoing support and investment.

We hope the *Canadian Cystic Fibrosis Registry* will continue to enhance knowledge and highlight key trends that will lead to better CF research and treatments.

The data collected are more than an invaluable resource to clinicians and researchers. It benchmarks the tremendous progress we've made, together with our donors and CF community, to improve the lives of Canadians with cystic fibrosis.

Together, our sights are set squarely on finding a cure.

Sincerely,

Maureen Adamson
President and CEO, Cystic Fibrosis Canada

Message from Dr. Anne Stephenson MD, PhD

Director, CF Registry

It is my pleasure to present the *Canadian Cystic Fibrosis Registry 2011 Annual Report* — the first rebranded report that now includes the name of the disease.

As we capture clinical data on every person diagnosed with cystic fibrosis in Canada, we continue a decades-old tradition of creating an important cystic fibrosis national resource. Similar to our annual report of 2010, we continue to capture back-data on individuals with the disease. Therefore, please note there may be minor discrepancies between this report and previous ones.

There are many people who deserve acknowledgment in the production of this year's annual report — our CF clinical community, the Canadian CF Registry Working Group, the generous community of donors whose support makes this report possible, and my colleagues at Cystic Fibrosis Canada for their leadership.

And of course, we could not do any of this without the Canadians with cystic fibrosis who agree to have their data collected. Thank you.

Together, we have again created a wealth of knowledge about the health of CF patients in Canada and an outstanding tool for CF clinicians and researchers alike.

Sincerely,

Dr. Anne Stephenson. MD, PhD
Director, CF Registry
Cystic Fibrosis Canada

Summary Data

Table 1
Summary data from Registry, 1986 to 2011

	1986	1991	1996	2001	2006	2011
General Profile						
Patients with data in reporting year (n)	2,381	2,798	3,094	3,318	3,468	3,913
Male, % of total patients	53.4	54.0	53.8	53.6	53.1	52.7
Age, mean (yr)	13.2	14.9	16.5	18.5	20.3	21.8
Age, median (yr)	12.0	13.0	15.0	16.0	18.0	20.0
% over 18 yrs	30.0	35.3	41.1	46.8	52.6	57.2
Race, % Caucasian	98.1	97.4	96.8	96.2	94.5	93.1
Black	0.3	0.5	0.5	0.7	0.8	0.7
Asian	0.4	0.4	0.4	0.4	0.4	0.6
First Nations People	0.3	0.5	0.7	0.8	0.7	0.9
South Asian	---	---	---	---	---	0.0
Other	0.8	1.0	1.3	1.5	1.9	0.0
Unstated	0.2	0.3	0.3	0.2	0.1	0.2
Transplants (#)	---	9.0	15.0	32.0	51.0	44.0
Pancreatic insufficient (%)	80.7	89.8	92.4	90.5	89.4	85.8
% with genotype analysis	---	---	---	---	---	95.0
Diagnosis						
Age at diagnosis, mean (yr)	2.2	2.4	2.7	3.2	3.5	3.9
median (mo)	7.0	6.0	6.0	6.0	7.0	7.0
New diagnoses in year (n)	148.0	130.0	106.0	105.0	94.0	114.0
% with meconium ileus at birth	12.2	15.9	16.1	11.6	17.9	11.9
Survival/Mortality						
Age at death, mean (yr)	19.7	24.6	25.7	27.9	28.5	33.6
median (yr)	19.0	25.0	24.5	25.0	25.0	34.0
Total deaths (n)	52.0	53.0	56.0	64.0	45.0	45.0
Crude mortality rate (%)	2.2	1.9	1.8	1.9	1.3	1.2
Median age of survival (yr)	27.5	33.5	32.5	36.1	44.7	48.5
Male	28.6	36.7	33.9	37.6	45.3	50.6
Female	25.8	29.3	30.6	34.1	44.1	43.2
Nutritional Markers						
≥ 18 yrs of age: n (%) in BMI categories						
< 20,	298 (46%)	400 (44%)	429 (36%)	452 (32%)	462 (28%)	519 (24%)
20-25.9,	328 (51%)	450 (50%)	669 (57%)	811 (57%)	1012(60%)	1309 (60%)
26-29.9,	14 (2%)	49 (5%)	65 (6%)	133 (9%)	156 (9%)	255 (12%)
≥ 30	3 (0%)	6 (1%)	14 (1%)	32 (2%)	50 (3%)	91 (4%)
Males < 23 BMI,	303 (81%)	394 (77%)	473 (73%)	501 (64%)	520 (57%)	621 (53%)
Females < 22 BMI	221 (82%)	294 (75%)	371 (70%)	420 (65%)	475 (61%)	593 (59%)

Summary Data

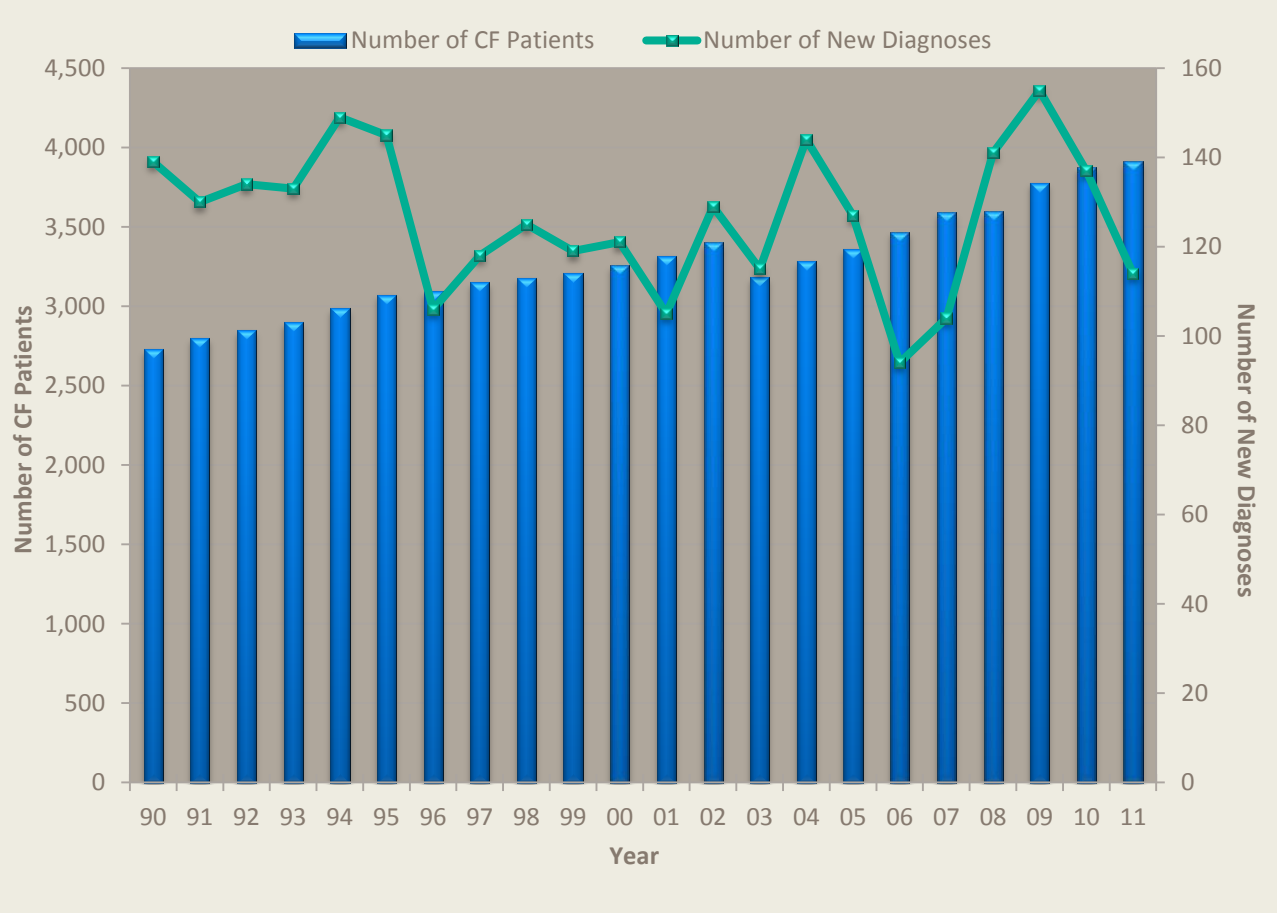
2 - 17 yrs of age: n (%) in BMI categories						
< 50th BMI %ile	---	---	---	---	---	784 (55.7%)
< 25th BMI%ile	---	---	---	---	---	408 (29.0%)
< 25th weight %ile	---	---	---	---	---	557 (39.6%)
< 10th weight %ile	---	---	---	---	---	278 (19.8%)
< 25th height %ile	---	---	---	---	---	634 (45.1%)
< 10th height %ile	---	---	---	---	---	317 (22.5%)
Pulmonary Function						
% predicted FEV_1, mean	70.4	70.7	71.8	72.2	72.3	72.6
% predicted FEV_1, median	72.5	72.9	73.8	73.0	73.6	73.7
n (%) FEV_1 % predicted categories for ≥ 18 years:						
Normal: ≥ 90%	61 (13%)	105 (12%)	133 (12%)	172 (12%)	228 (14%)	345 (16%)
Mild: 70-89%	109 (22%)	172 (20%)	236 (21%)	312 (23%)	401 (25%)	561 (26%)
Moderate: 40-69%	169 (35%)	315 (37%)	464 (41%)	587 (43%)	701 (43%)	834 (39%)
Severe: < 40%	147 (30%)	256 (30%)	293 (26%)	307 (22%)	301 (18%)	402 (19%)
n (%) FEV_1 % predicted categories for ages 6 to 17:						
Normal: ≥ 90%	284 (37%)	423 (40%)	532 (46%)	571 (48%)	519 (49%)	570 (55%)
Mild: 70-89%	214 (28%)	329 (31%)	355 (30%)	344 (29%)	329 (31%)	284 (27%)
Moderate: 40-69%	207 (27%)	241 (23%)	224 (19%)	235 (20%)	185 (17%)	163 (16%)
Severe: < 40%	57 (7%)	62 (6%)	54 (5%)	41 (3%)	32 (3%)	16 (2%)
n (%) on oxygen	---	---	---	111 (3%)	100 (3%)	146 (4%)
n (%) on BiPAP	---	---	---	---	---	18 (0%)
Microbiology						
n (%) with positive culture (first culture of the year (76-96), starting in 2001 any culture in the year):						
Pseudomonas aeruginosa	978 (41%)	1186(42%)	1337(43%)	1670(50%)	1660(48%)	1741 (44%)
Staphylococcus aureus	550 (23%)	796 (28%)	980 (32%)	1439(43%)	1641(47%)	1946 (50%)
Haemophilus species	332 (14%)	341 (12%)	415 (13%)	551 (17%)	542 (16%)	652 (17%)
Stenotrophomonas maltophilia	---	---	104 (3%)	253 (8%)	356 (10%)	572 (15%)
Aspergillus	---	---	---	272 (8%)	413 (12%)	854 (22%)
MRSA	---	---	---	---	101 (3%)	188 (5%)
Alcaligenes (achromobacter) species	---	---	---	---	---	111 (3%)
Atypical mycobacteria	---	---	---	---	---	91 (2%)
Burkholderia cepacia complex	172 (7%)	273 (10%)	226 (7%)	188 (6%)	159 (5%)	200 (5%)
B. cenocepacia	---	---	---	---	---	69 (34%)
B. multivorans	---	---	---	---	---	60 (30%)
B. vietnamiensis	---	---	---	---	---	9 (4%)
B. gladioli	---	---	---	---	---	11 (6%)
B. cepacia Other	---	---	---	---	---	18 (9%)
Unknown	---	---	---	---	---	34 (17%)

Demographic Data

Number of Canadians with Cystic Fibrosis

In 2011, a total of 3,913 individuals with cystic fibrosis had clinical records submitted by 42 CF clinics. 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 2011, 114 individuals were newly diagnosed with this disease.

Figure 1
Total number of CF patients and new diagnoses in the Registry, 1990 to 2011



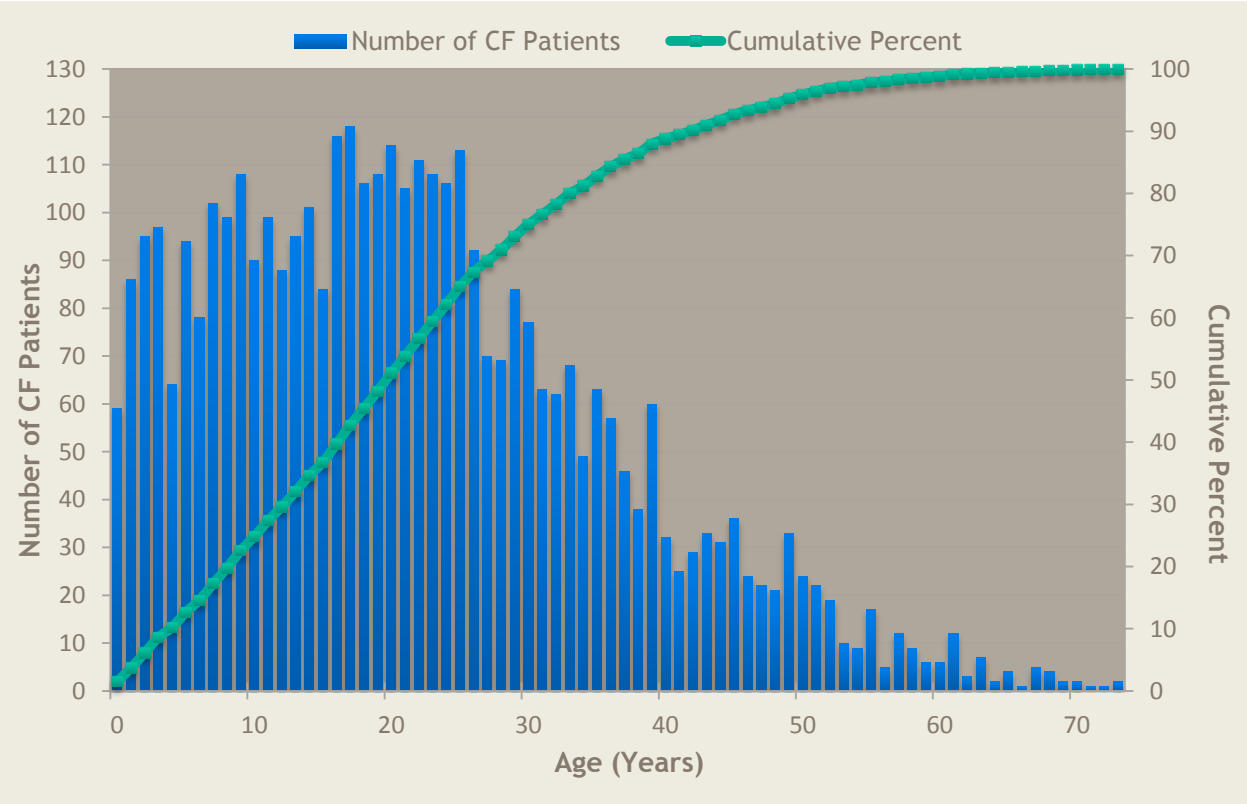
Demographic Data

Ages of Canadians with Cystic Fibrosis

Figure 2 shows the age distribution of the Canadian CF population for 2011. The ages of individuals with cystic fibrosis range from birth to 75 years old. The median age remains 20 years, the same as 2010.

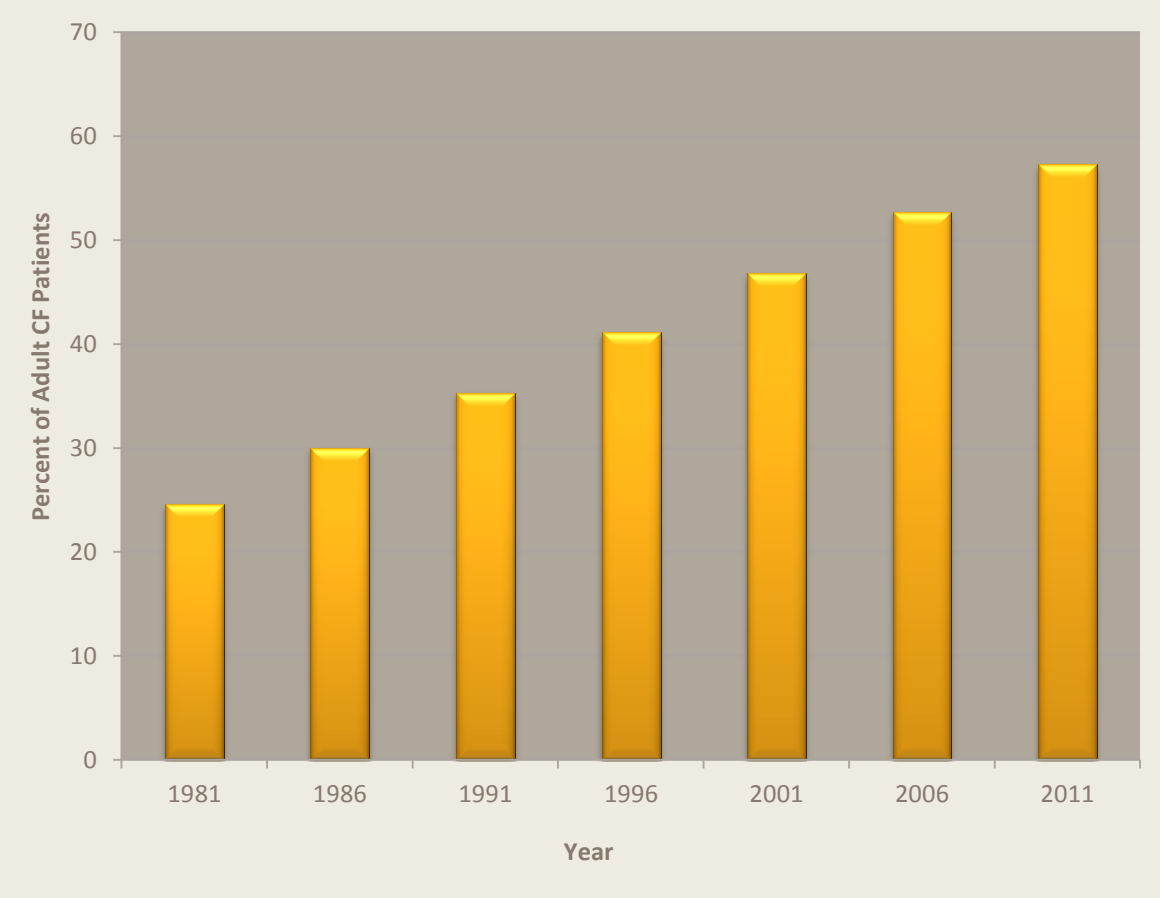
Males account for 52.7% of individuals in the Registry in 2011, and 57.2% of individuals in the Registry are 18 years of age or older (Figure 3).

Figure 2
Age distribution of the CF population, 2011



Demographic Data

Figure 3
Proportion of individuals with cystic fibrosis 18 years of age or older



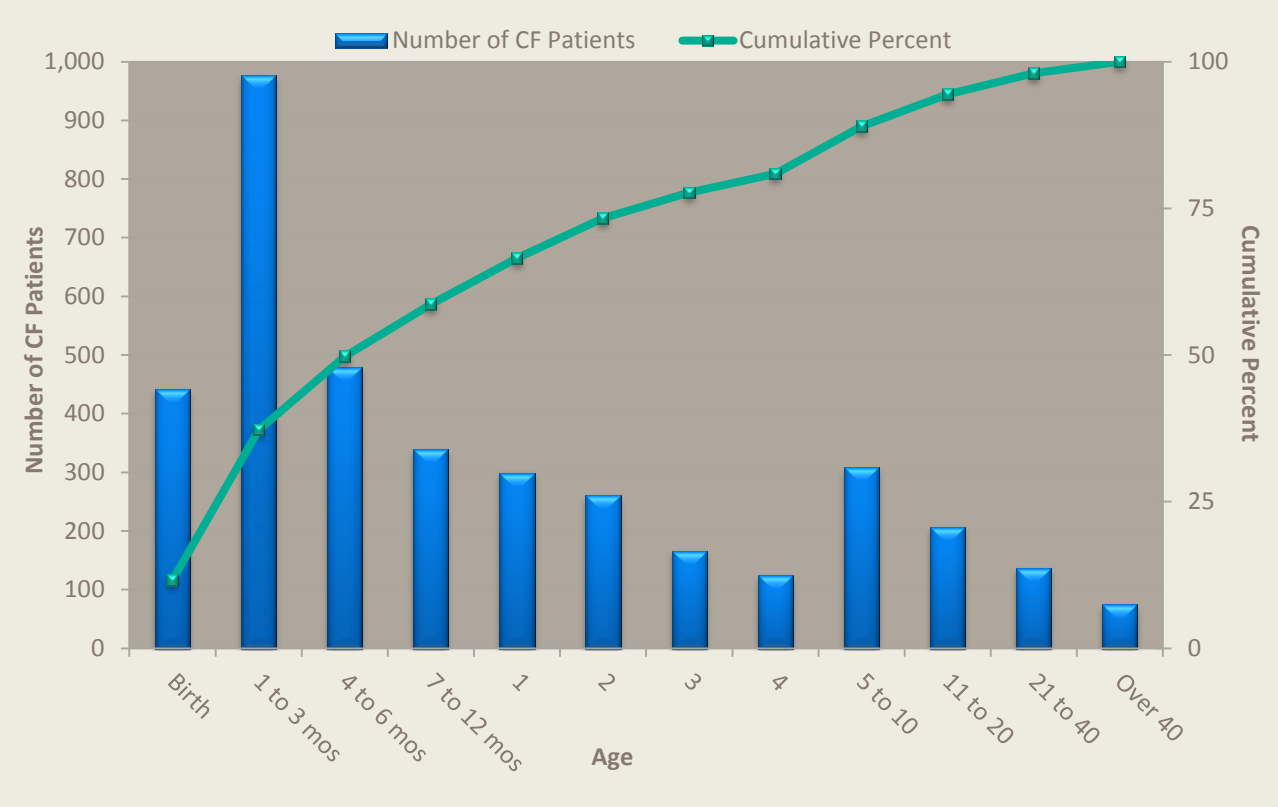
Demographic Data

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 programs for cystic fibrosis are introduced in Canadian provinces, the majority of individuals with cystic fibrosis will be diagnosed at birth.

Figure 4
Age at diagnosis, all patients, 2011



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 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 resulting in the deletion of the phenylalanine residue at amino acid position 508, commonly referred to as deltaF508. Of those individuals with genetic information recorded within the Registry, 56.3% carry two deltaF508 mutations (Figure 5) and 91.5% carry at least one deltaF508 mutation (Table 2).

Figure 5
Genotype (based on N=3,348)

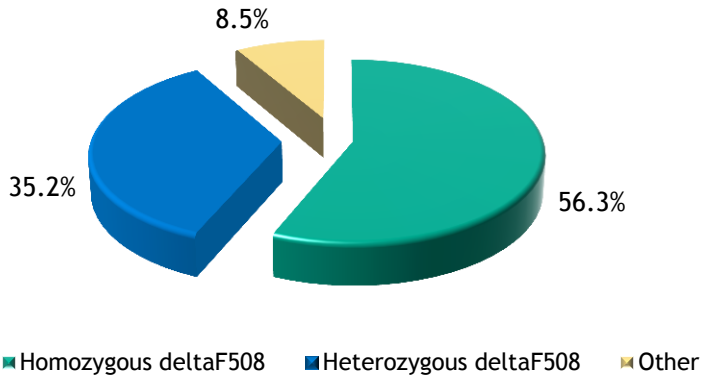


Table 2
Frequency of CF mutations on one or both alleles (top five)

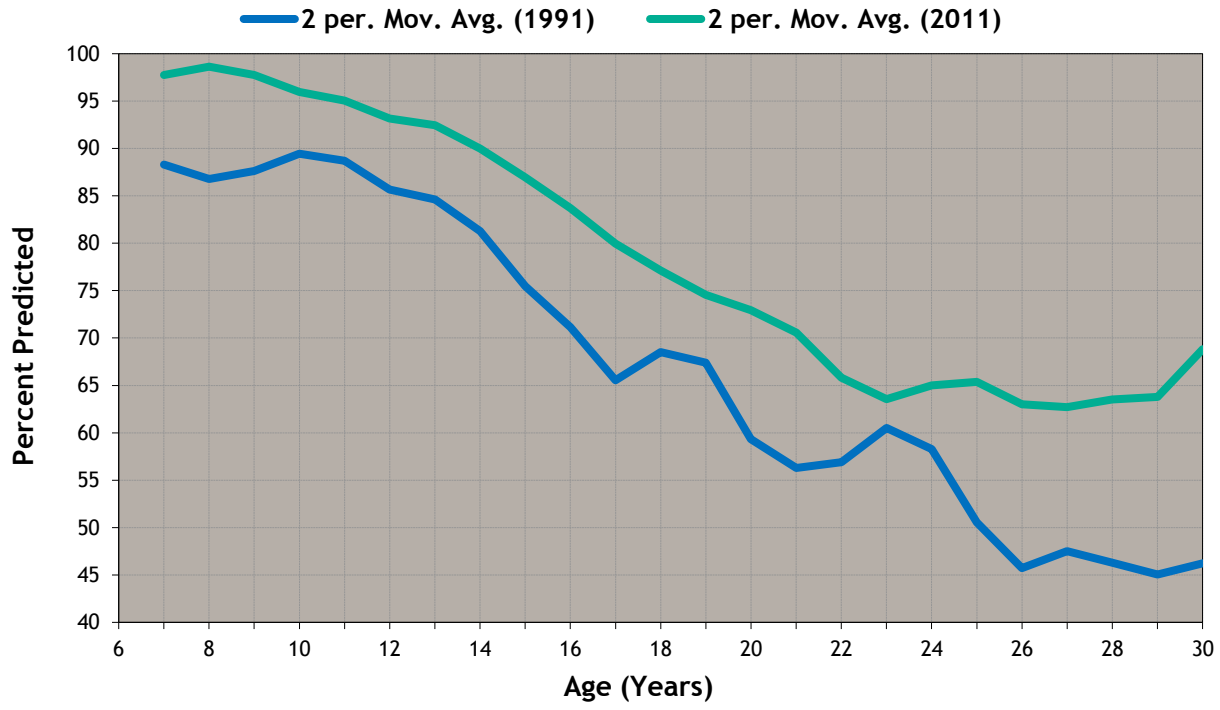
Genotype	Number	Percentage
DeltaF508	3,063	91.5
621+1G->T	217	6.5
G542X	134	4.0
G551D	115	3.4
711+1G->T	96	2.9

Respiratory

Median FEV_1: Drop in Lung Function

Median FEV_1 percent predicted has improved over the last two decades. The median FEV_1 in 2011 at 30 years of age is 72.3% predicted compared to 45.7% in 1991 (Figure 6). From Figure 6, we can see that the largest drop in lung function is between the ages of 13 and 22 years. Once an individual reaches age 22 years, lung function seems to stabilize. This suggests that perhaps the adolescent/young adulthood years are a vulnerable time for individuals with cystic fibrosis.

Figure 6
Median percent predicted FEV_1 vs. age, 1991 and 2011

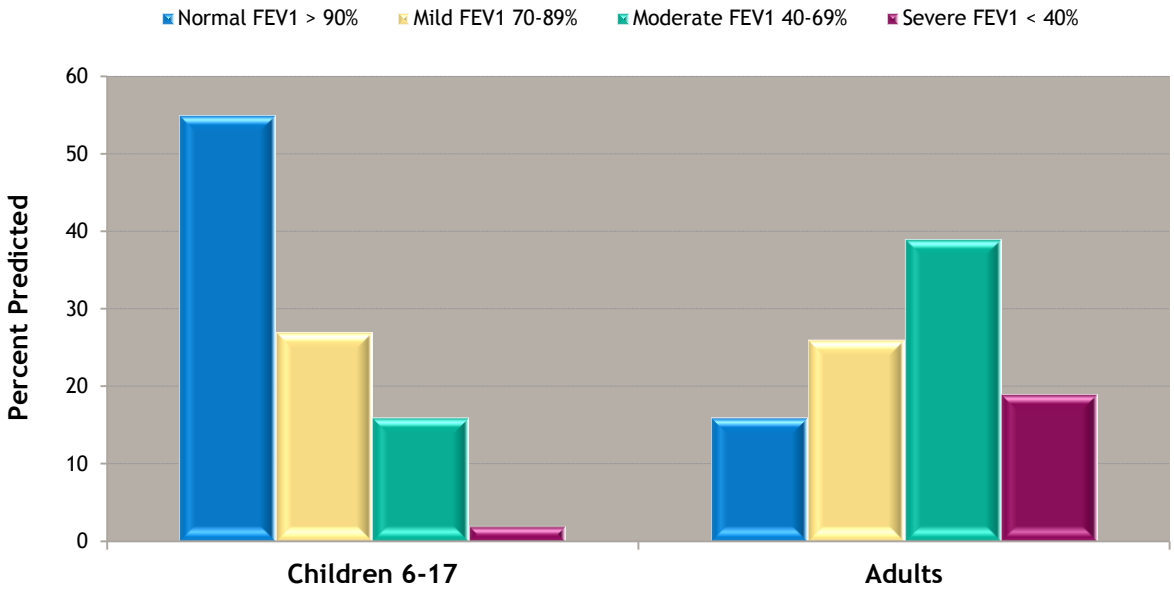


Respiratory

Respiratory Severity

Lung function is measured from age six years and older. For children ages 6 to 17, the majority of individuals with cystic fibrosis (55%) have normal lung function (FEV_1 greater than 90% predicted), compared to 50% in 2010. For adults, the majority (39%) have lung function classified as 'moderate' severity (Figure 7). These figures are similar to those in 2010.

Figure 7
Respiratory severity of CF children and adults (FEV_1 percent predicted), 2011



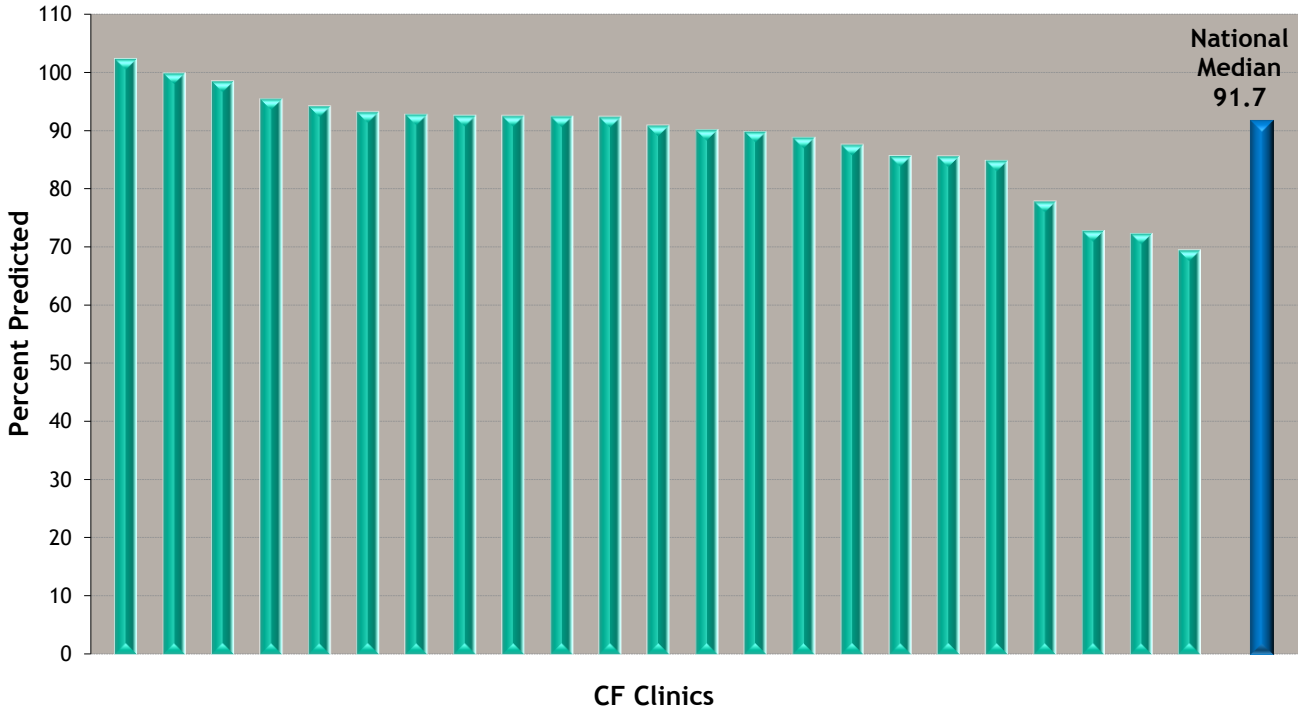
Respiratory

Median FEV_1 by Reporting CF Clinic

In order for a CF clinic to be included in Figure 8, it had to report lung function data on 10 or more CF patients. Twenty-three CF clinics are included in the graph.

The number of observations per clinic used to calculate the median FEV_1 ranges from 10 to 185. The national median FEV_1 is 91.7% predicted with a range from 69.6% to 102.5% (Figure 8).

Figure 8
Median FEV_1 percent predicted for CF patients 6-17 years of age by CF clinic, 2011



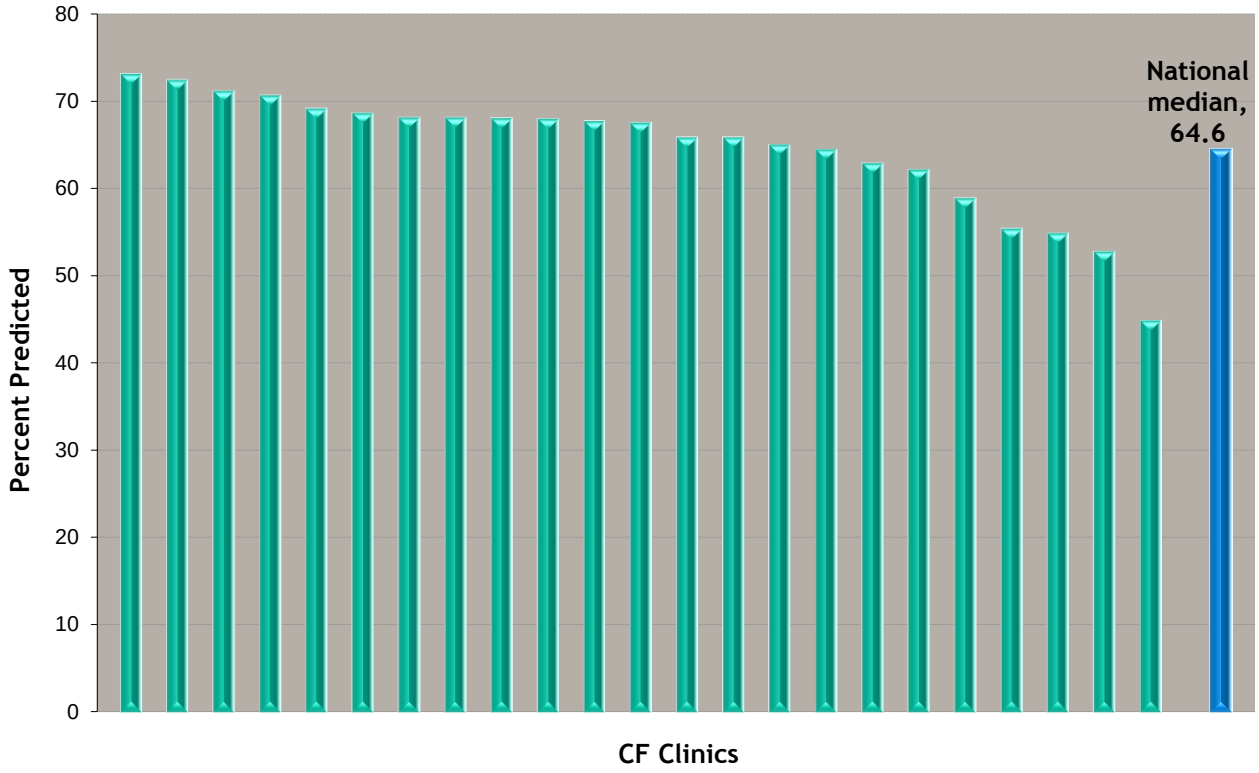
Respiratory

National Median FEV_1

Figure 9 shows the median FEV_1 percent predicted by CF clinics for those patients 18 years of age and older. In order to be included in this graph, a CF clinic had to report lung function data on at least 10 CF patients.

A total of 23 CF clinics are shown on the graph. The number of observations per clinic used to calculate the median FEV_1 ranges from 17 to 356. The national median FEV_1 is 64.6% predicted with a range from 45.0% to 73.2%.

Figure 9
Median FEV_1 percent predicted for CF patients 18 years of age and older by CF clinic, 2011



Nutrition

In 2011, 86% of individuals with cystic fibrosis were taking supplemental pancreatic enzymes (pancreatic insufficient), whereas 14% were considered pancreatic sufficient (Figure 10).

For those individuals 40 years of age or older, 65% are pancreatic insufficient and 35% are pancreatic sufficient (the same as in 2010). This is likely a reflection of the fact that older adults with cystic fibrosis are more likely to have milder mutations that are associated with pancreatic sufficiency (Figure 11).

Figure 10
Pancreatic sufficiency in CF patients

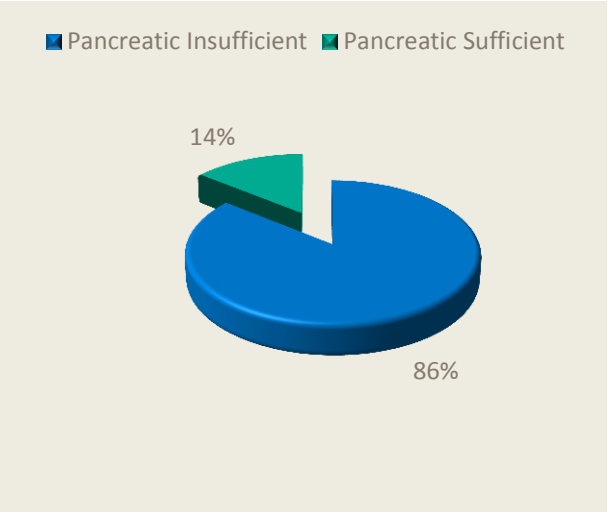
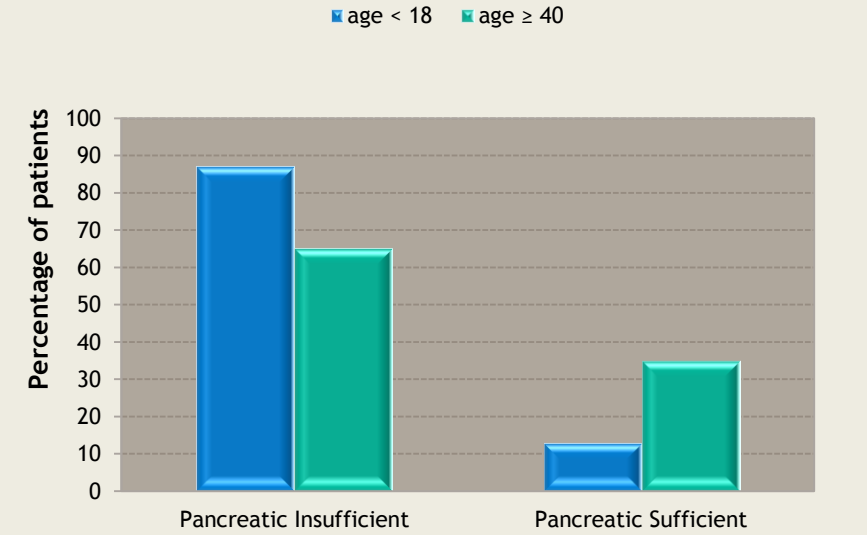


Figure 11
Pancreatic status by age group



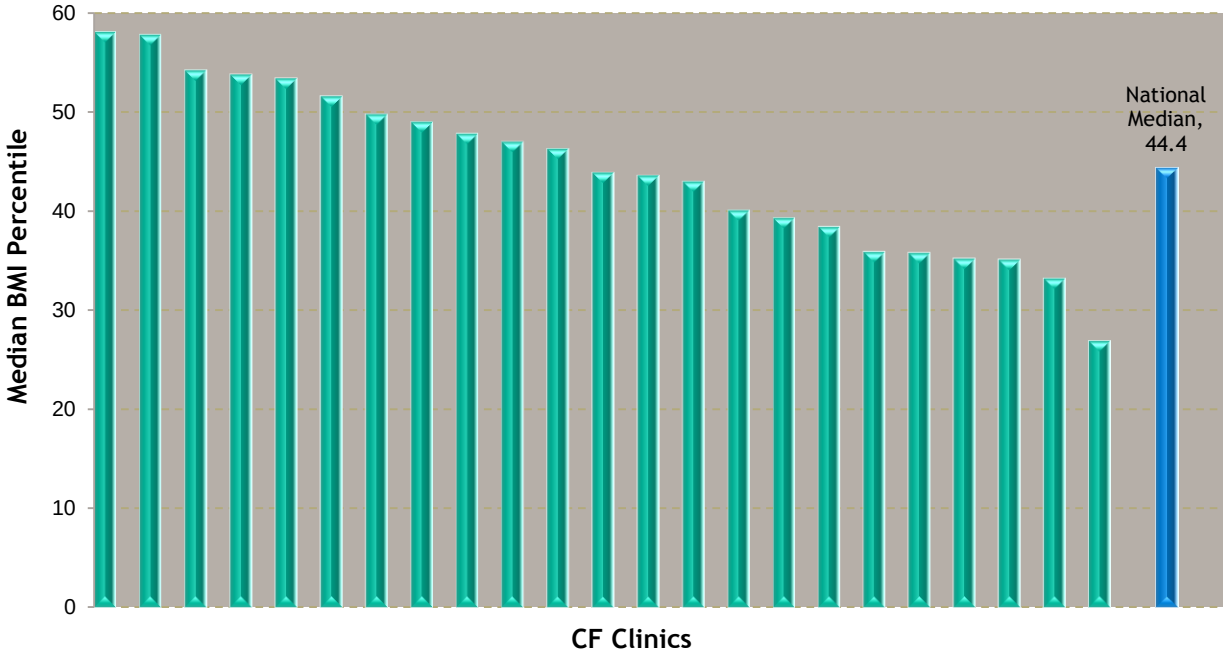
Nutrition

Body Mass Index (BMI)

A total of 23 paediatric CF clinics are included in Figure 12. In order to be included in this graph, a CF clinic had to report data on at least 10 CF patients. In CF patients 2 to 17 years of age, the national median Body Mass Index percentile, as per the Centers for Disease Control (BMI CDC), is 44.4%.

The median percentiles range from 27% to 58.2%. BMI percentile is not calculated for those under the age of two years.

Figure 12
Median BMI percentile for CF patients 2-17 years by CF clinic, 2011



Weight Status Category	Percentile Range
Underweight	Less than the 5th percentile
Healthy weight	5th percentile to less than the 85th percentile
Overweight	85th to less than the 95th percentile
Obese	Equal to or greater than the 95th percentile

Nutrition

Figure 13
Median BMI for CF patients 18 years of age and older by CF clinic, 2011

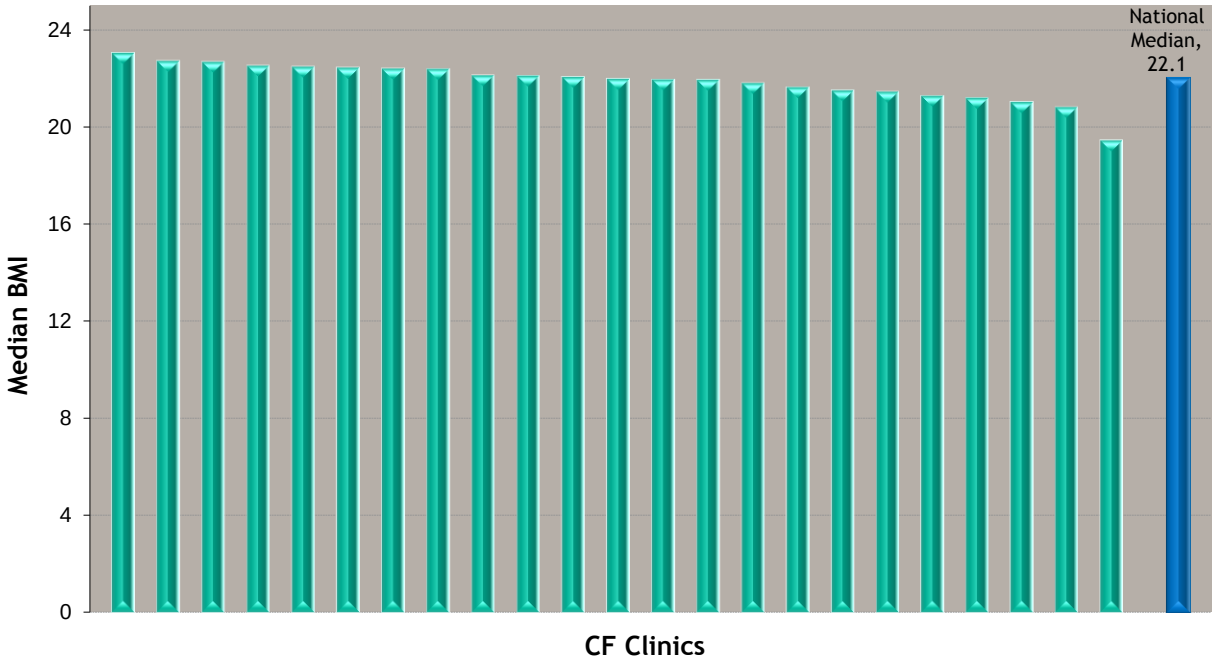


Figure 13 shows the median BMI by clinic for those CF patients 18 years of age and older. There are 23 CF clinics included in this graph. In order to be included in this graph, a CF clinic had to report data on at least 10 CF patients. The national median BMI is 22.1 kg/m^2. The median BMI ranges from 19.5 to 23.1 kg/m^2.

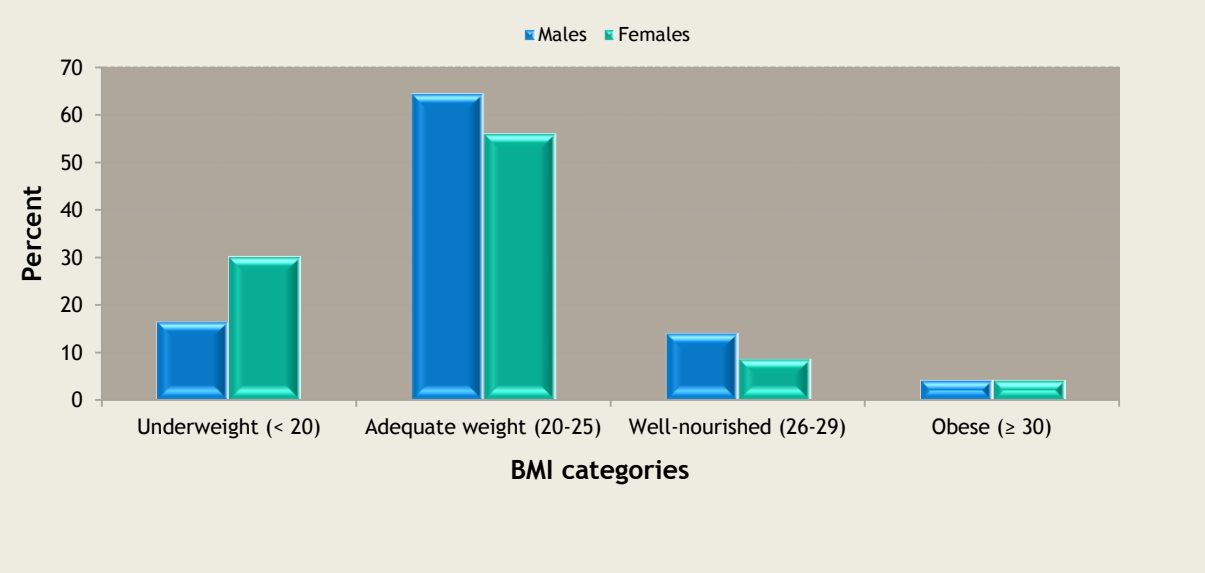
BMI Categories	*Range*
Underweight	*<20.0 kg/m^2*
Adequate weight	*20.0 - 25.9 kg/m^2*
Well-nourished	*26 - 29.9 kg/m^2*
Obese	*≥30 kg/m^2*

Nutrition

BMI by Patient 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 compared to males (BMI < 20 kg/m^2). Often young men who are muscular and fit can have a BMI between 26-29 kg/m^2 due to high muscle mass. Overall, 4.4% of the CF adult population has a BMI in the "obese" category.

Figure 14
BMI classification for CF patients, by sex, 2011



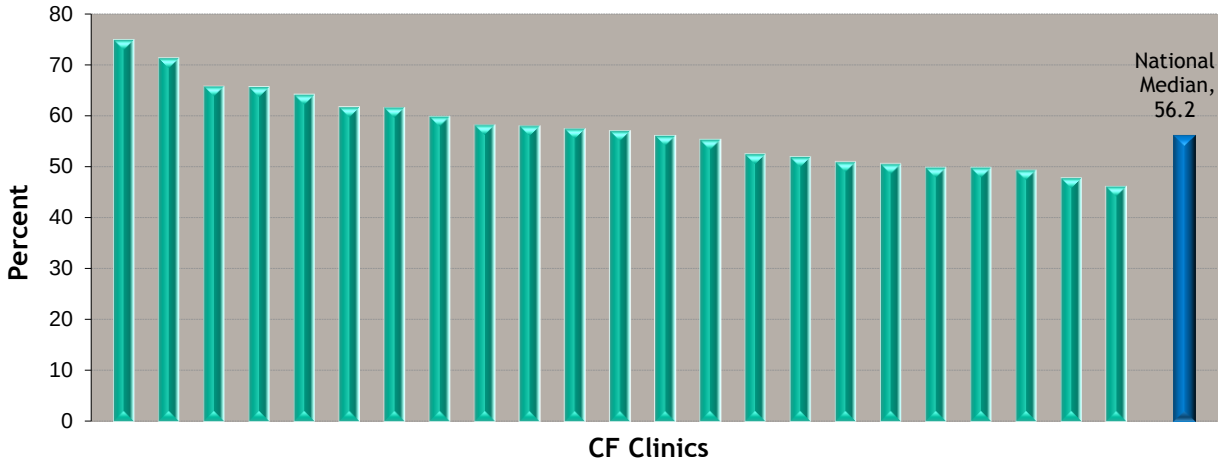
Nutrition

National Average BMI 18 years of age and older

The national average percentage of CF patients with BMI < 22 kg/m^2 for females and < 23 kg/m^2 for males in patients 18 years of age and older is 56.2% with a range from 46.3% to 75.0%. In order to be included in Figure 15, a CF clinic had to report data on at least 10 CF patients. There are 23 CF clinics included in this graph.

Figure 15

Percentage with BMI < 22 kg/m^2 for females and < 23 kg/m^2 for males for CF patients 18 years of age and older by CF clinic, 2011



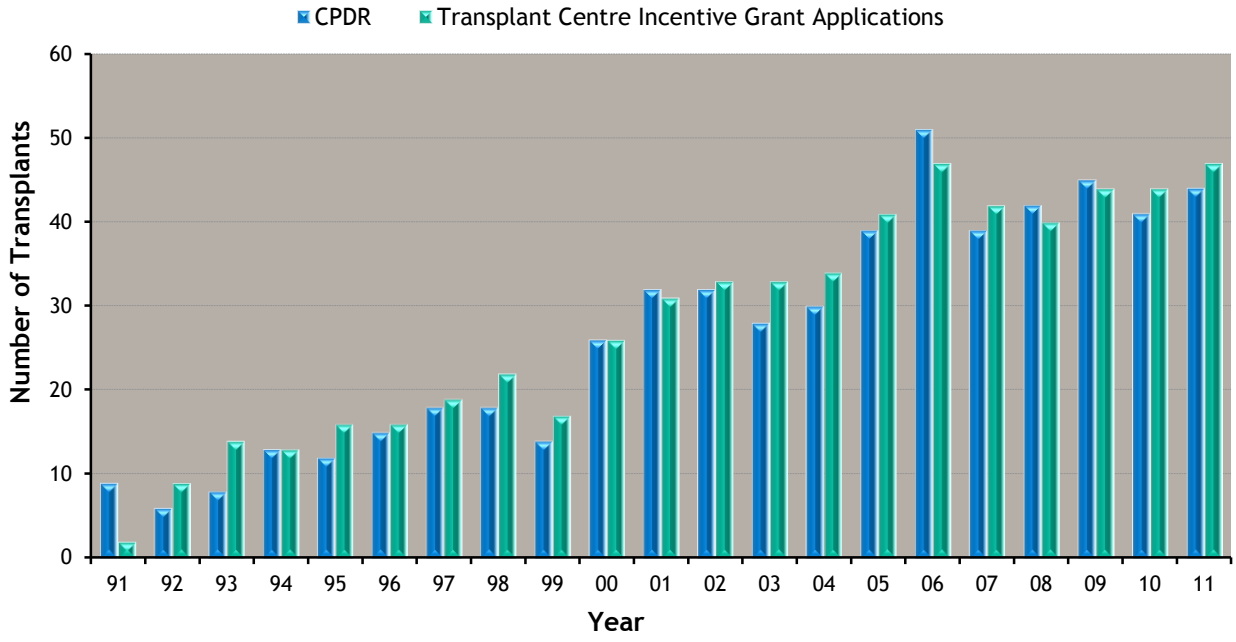
Transplantation

Figure 16 shows the number of transplants carried out per year as reported in the Registry and as reported by the transplant centres on their incentive grant applications. 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.

Based on the transplant statistics reported in the transplant centres' incentive grant applications, the number of CF patients transplanted in 2011 was 47, compared to 44 recorded in the Registry.

We are working hard with transplant centres to ensure we capture accurate data on transplants within the Registry. We hope that in the future transplant information will be entered into the Registry directly from the transplant centres.

Figure 16
Number of patients transplanted per year, 1991 to 2011



Microbiology

Bacterial Species & Respiratory Infections

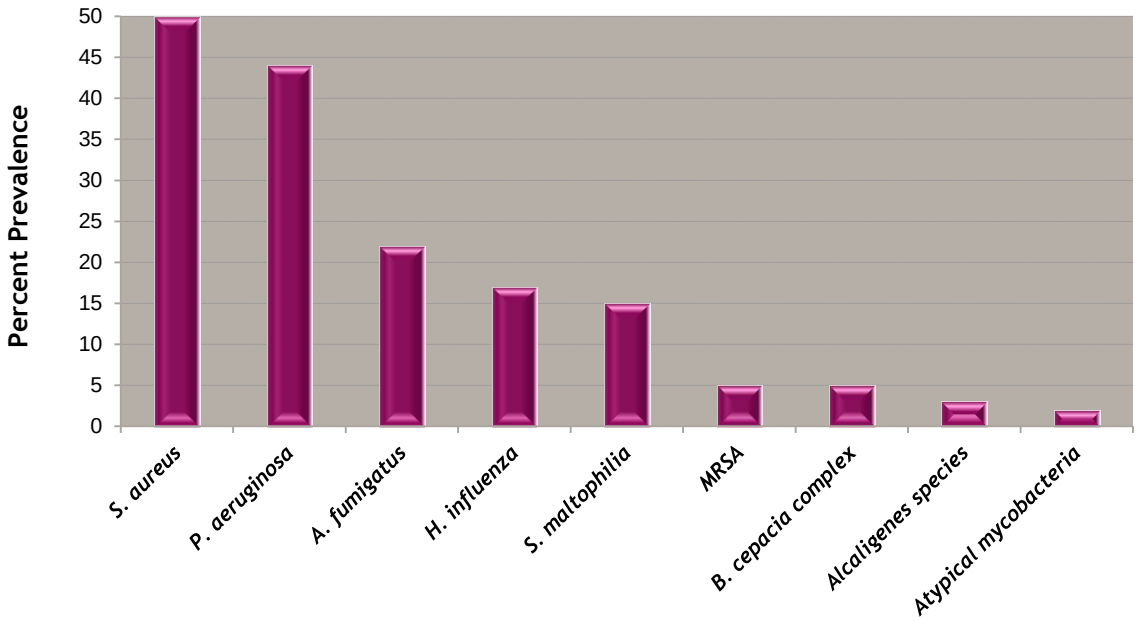
Overall, *Pseudomonas aeruginosa* and *Staphylococcus aureus* are the most common pulmonary pathogens in Canadians with cystic fibrosis (Figure 17). In 2011, clinics began to record additional microbiology data including the prevalence of Alcaligenes (achromobacter) species and Atypical mycobacteria.

The prevalence of *Aspergillus fumigatus* species, *Stenotrophomonas maltophilia*, and MRSA continues to increase since 2007 (Figure 18). 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 2011, 5.0% of Canadians carried MRSA. This information was not collected in the Registry prior to 2003. As expected, *Staphyloccocus aureus* is more common in the paediatric population whereas *Pseudomonas aeruginosa* is more common in the adult CF population.

The prevalence of *Stenotrophomonas maltophilia* increases until the teen years but then appears to stabilize (Figure 19). *Burkholderia cepacia* complex is more commonly seen in older individuals with cystic fibrosis which represents the fact that 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 17

Prevalence of bacterial species cultured in airways in CF patients (all ages), 2011



Microbiology

Figure 18
Prevalence of respiratory infections, 2007-2011

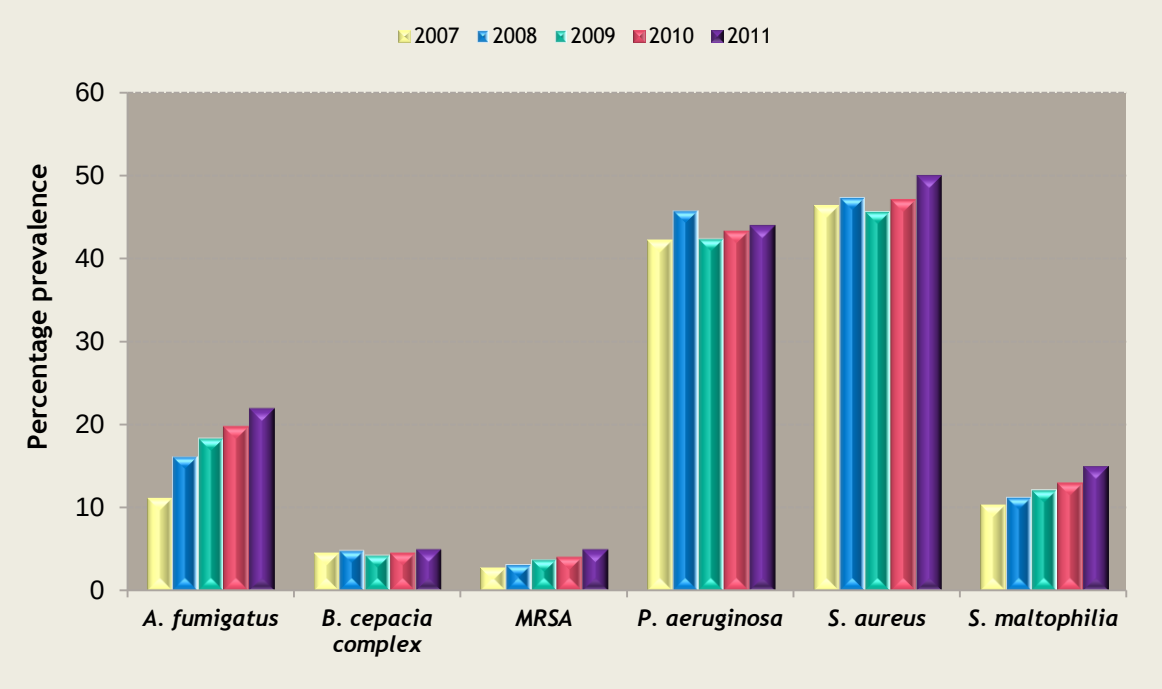
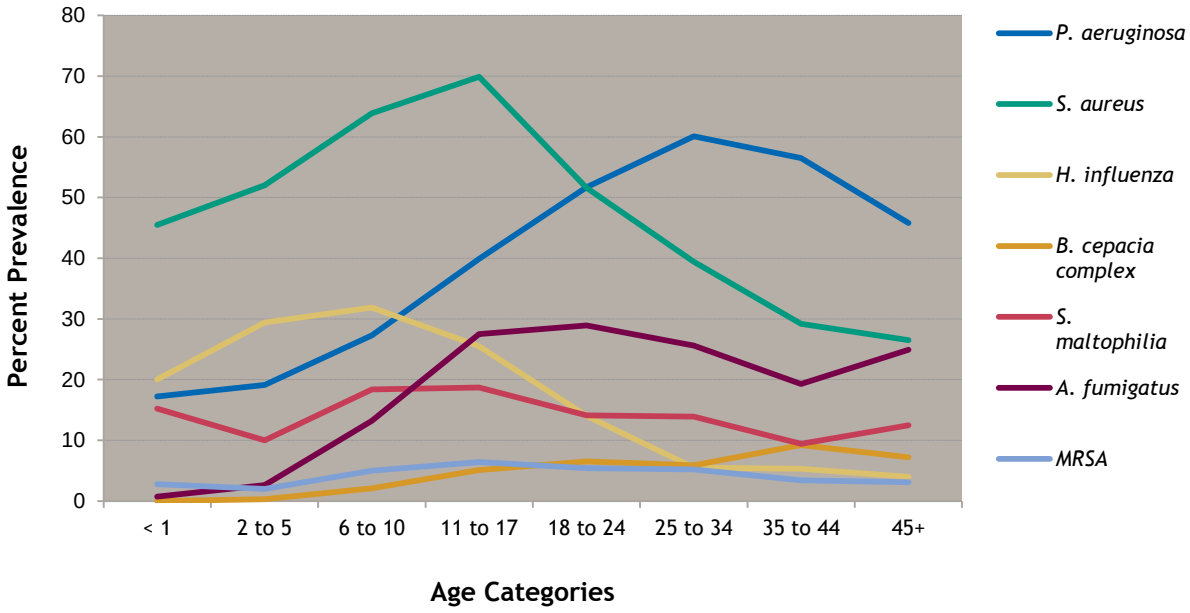


Figure 19
Age-specific prevalence of respiratory infections in CF patients, 2011



Microbiology

New Data: *Burkholderia cepacia* complex (BCC)

New for 2011, clinics submitted data on the type of *Burkholderia cepacia* complex (BCC) species (Figure 20). *B. cenocepacia* and *B. multivorans* were the two most common types of BCC species. Of the patients with BCC species, 77% are adults (Figure 21). Not all BCC bacteria has been genotyped which is represented by the fact that 17% of the BCC species in the Registry are classified as unknown.

Figure 20
Burkholderia cepacia complex species, 2011

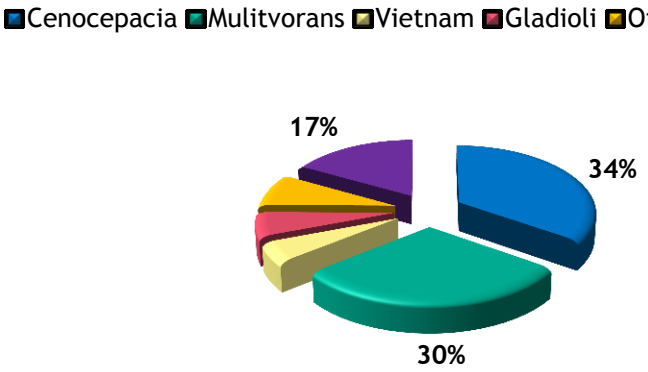
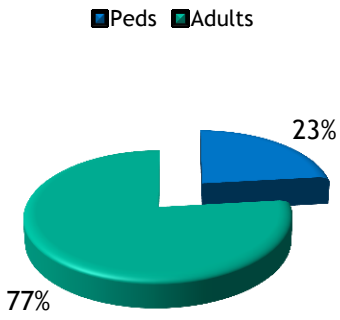


Figure 21
Burkholderia cepacia complex distribution by age, 2011

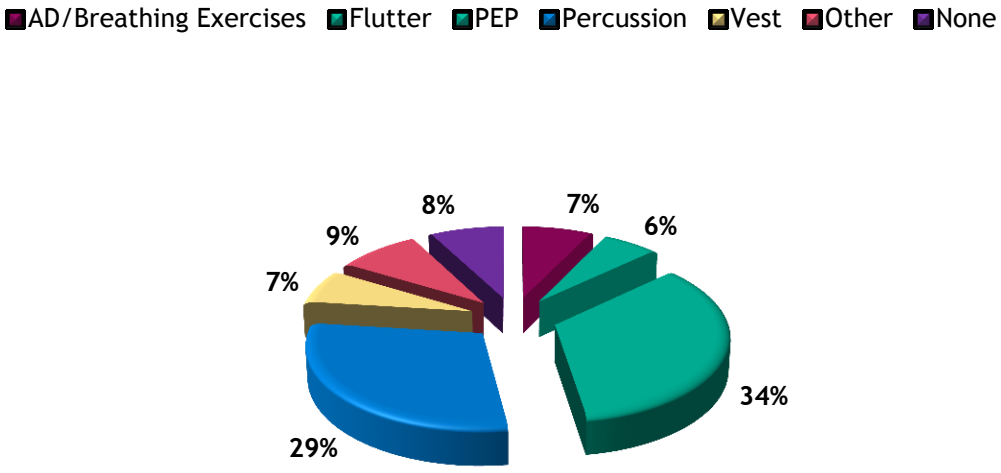


Microbiology

Physiotherapy

Positive expiratory pressure (also called, PEP) and percussion are the most common forms of physiotherapy used by Canadian CF patients. Individuals who had received a transplant were excluded from these calculations.

Figure 22
Physiotherapy, 2011

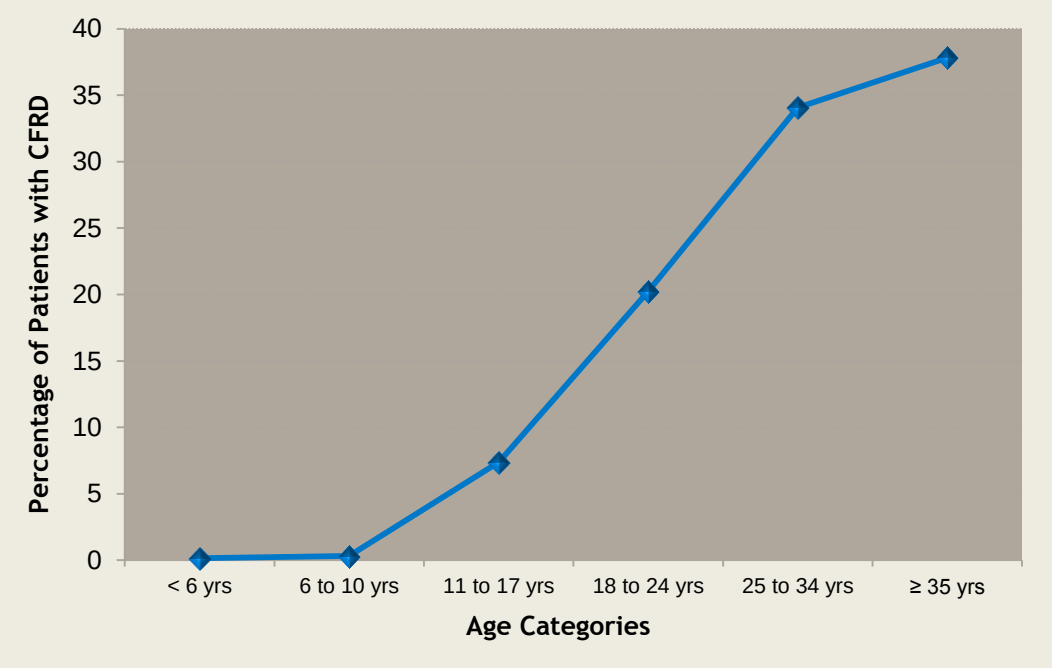


CF-Related Diabetes (CFRD)

Overall, CFRD was reported in 16.3% of individuals with CF in 2011. In those individuals with CFRD, approximately 38% are 35 years of age and older.

The prevalence of CFRD increases with age (Figure 23). Of those with CFRD, 52.7% are female.

Figure 23
Percentage of patients with CFRD by age category, 2011



Hospitalization and Home IV

Pulmonary Exacerbation Leading Cause

In 2011, 1,875 hospitalizations were recorded in the Registry.The most common recorded reason for admission was a pulmonary exacerbation. In 2011, 792 courses of home IV therapy were recorded within the Registry.

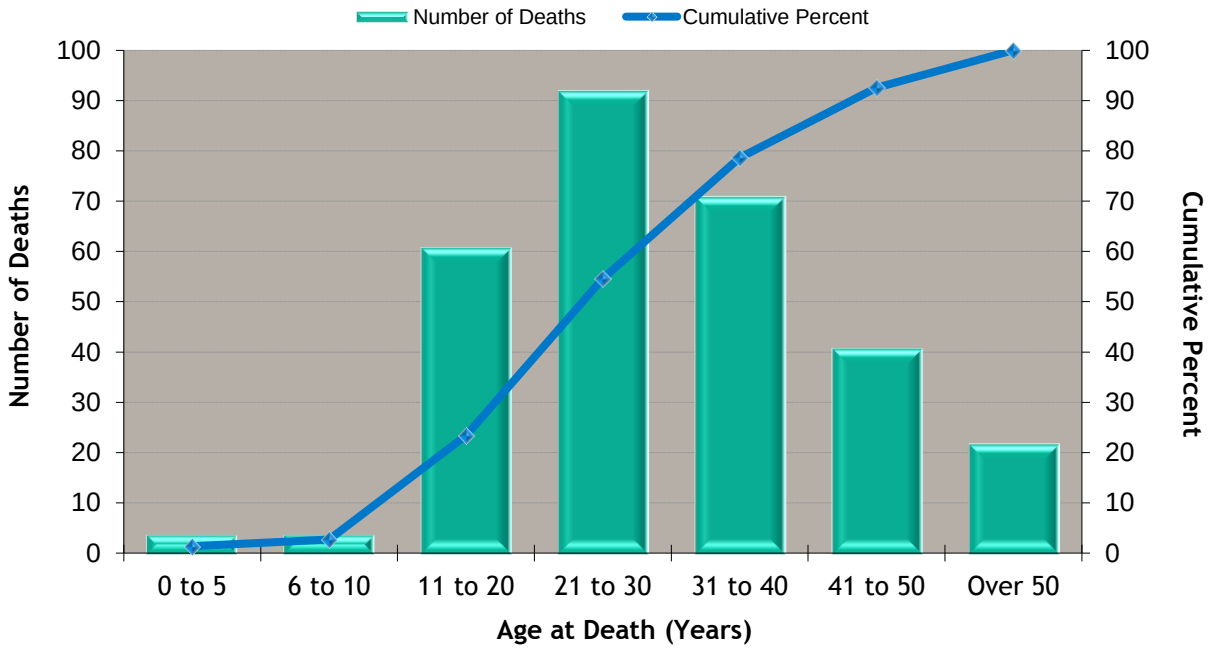
Table 3
Number of hospital days and home IV courses, 2011

	Total Number
Hospital Days	25,279
Hospitalizations	1,875
Clinic Visits	15,513
Home IV Courses	792
Home IV Days	12,088

Survival

There were 45 deaths recorded in the Registry for 2011. Since there are relatively few deaths per year, the sum of all deaths from 2007 to 2011 have been included in Figure 24. **The median age at death in 2011 was 34 years of age.** The most common cause of death was pulmonary-related.

Figure 24
Age at death, 2007 to 2011



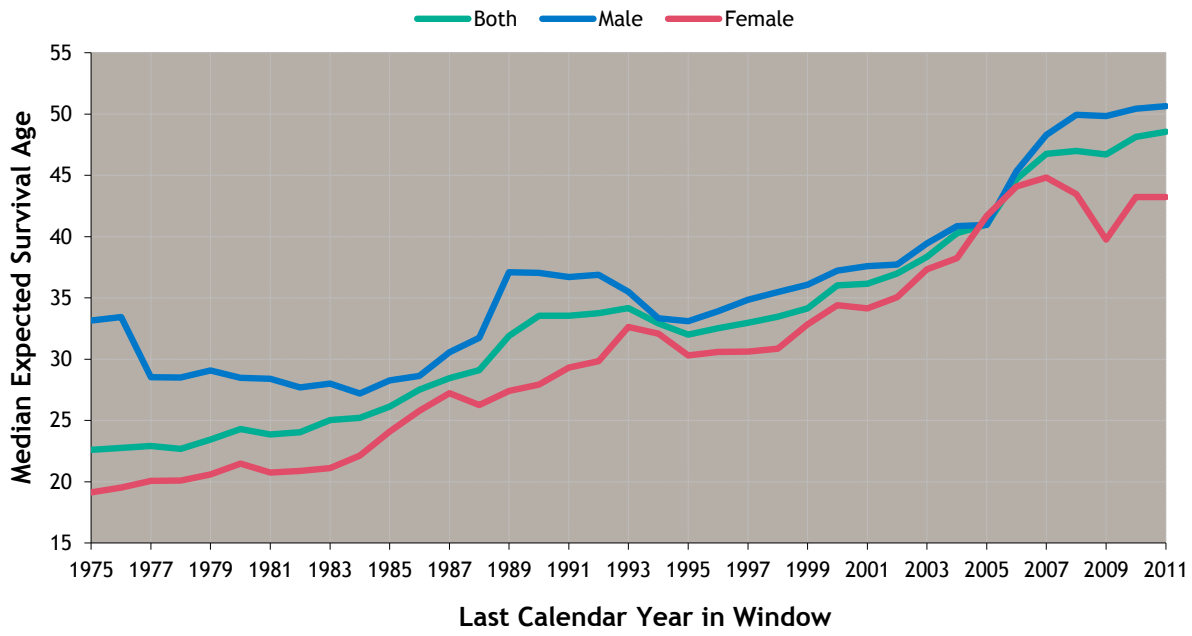
Survival

Median Age of Survival

The median age of survival for Canadians with cystic fibrosis is currently estimated to be **48.5 years of age** (Figure 25). Over the years, females have had higher mortality than males with cystic fibrosis which seems to persist in 2011. The cause of worse survival in females is not well understood but has been documented in the literature in many countries in addition to Canada.

It is not possible to know for certain the reason for the improvement seen in survival for Canadians with CF 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, Cystic Fibrosis Canada volunteers, partners, donors, researchers and CF clinic teams. Everyone can be very proud of this accomplishment.

Figure 25
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 (1/2) died before the median age of death and half (1/2) 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*.

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Jackie Ruderman, Program Director, Strategic Initiatives
Ian McIntosh, Program Director, Healthcare

Canadian CF Clinics

Victoria General Hospital	Hotel-Dieu Hospital, Kingston
Royal Jubilee Hospital, Victoria	Children's Hospital of Eastern Ontario, Ottawa
B.C. Children's Hospital, Vancouver	Ottawa General Hospital
St. Paul's Hospital, Vancouver	*Centre de santé et de services sociaux de Gatineau, Hull*
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Foothills Hospital, Calgary	Montreal Chest Institute
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Royal University Hospital, Saskatoon	*Hôtel-Dieu de Montréal*
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Hamilton Health Sciences Corporation	Saint John Regional Hospital
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®

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.

Since 1960, Cystic Fibrosis Canada has invested more than $140 million in innovative CF research and clinical care in Canada.

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 and it is more important than ever to support research and care programs in Canada.

For more information and to donate, please visit www.cysticfibrosis.ca.

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