



CYSTIC FIBROSIS

Cystic fibrosis (CF) is a rare disease affecting over 4,300 Canadians or roughly 1 in 3,600 live births. Cystic fibrosis is a progressive, degenerative multi-system disease that affects mainly the lungs and 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.

In addition to the physical effects of the disease, mental health concerns are emerging; anxiety and depression are common among this population. Individuals with cystic fibrosis may reach the point where they require a lung transplant; most fatalities of people with cystic fibrosis are due to lung disease. There is no cure.

CYSTIC FIBROSIS CANADA

Since being founded by parents in 1960, Cystic Fibrosis Canada has grown into a leading organization with a central role engaging people living with cystic fibrosis, parents and caregivers, volunteers, researchers and healthcare professionals, industry, government and donors. We work together going further to change lives; advocating for access to therapy, supporting delivery of care, funding research, and providing information and support. We will not stop until all people with cystic fibrosis can and do experience everything life has to offer — and enjoy everything life has to offer. For more information, visit www.cysticfibrosis.ca.

Published: February 2022

Scan the QR code below to access the online publication of the report, or please visit us at www.cysticfibrosis.ca.

Suggested citation (print): Cystic Fibrosis Canada. (2022). *The Canadian Cystic Fibrosis Registry 2020 Annual Data Report*. Toronto, Canada: Cystic Fibrosis Canada.

Suggested citation (online): Cystic Fibrosis Canada. (2022). *The Canadian Cystic Fibrosis Registry 2020 Annual Data Report*. Retrieved from <https://www.cysticfibrosis.ca/Registry/2020AnnualDataReport.pdf>.

Cover page: CF individual from Quebec



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OUR VISION IS A WORLD WITHOUT CYSTIC FIBROSIS

OUR MISSION IS TO END CYSTIC FIBROSIS

TABLE OF CONTENTS

Foreward	2
2020 Highlights	3
Introduction	4
Demographics	6
Diagnosis	14
Genotype	16
Ethnicity	18
Respiratory	19
Nutrition	23
Microbiology	30
Physiotherapy	34
Medications	35
Healthcare Encounters	37
Cystic Fibrosis-Related Diabetes (CFRD)	38
Mental Health	39
Transplants	40
Survival	41
COVID-19	46
Glossary of Terms	47
References	48



FOREWORD

We are pleased to share the 2020 Annual Data Report for the Canadian Cystic Fibrosis Registry (CF Registry). While the data in this report reflect the first year of the COVID-19 pandemic, we are relieved to announce that this doesn't appear to have had significant immediate effects on health outcomes, including the estimated median age of survival for a Canadian born with cystic fibrosis (CF). We are grateful to the committed healthcare teams, patients and caregivers, researchers and advocates whose dedication ensures these steady health outcomes, particularly through a pandemic.

The CF Registry has made a remarkable impact on our understanding and treatment of the disease, and its importance continues. We are grateful to our founders for having had the foresight to begin the national CF Registry back in the 1970's. The data help us to track trends in the Canadian CF community and understand the progress and impact that has been made to advance health outcomes. It has been instrumental for Canadian cystic fibrosis research and care and has also been a tremendous tool for our advocacy efforts.

As a result of the CF Registry, we have been able to provide decision makers with key and timely information to assist them in understanding the possible impact of funding drugs for this community. When the Canadian Agency for Drugs and Technology (CADTH) issued its draft recommendation on the disease modifying drug Trikafta, we were able to swiftly report on the number of Canadians with CF who could be eligible but would be excluded from accessing the drug.

In addition to the support in advocacy, the CF Registry enabled us to advance our knowledge on COVID-19 and cystic fibrosis. When the pandemic was announced in March of 2020, there were a lot of unknowns about how COVID-19 would affect a person with cystic fibrosis. The CF Registry team quickly communicated to the CF clinics on how to capture items related to COVID-19 in existing CF Registry fields and partnered with our global peers to research the new virus. We have been participating in global research related to COVID-19 since March 2020 and continue to study how the disease impacts our population today, using data from the CF Registry.

We continue to prioritize and leverage the CF Registry to support our programs, including Cystic Fibrosis Canada's Accelerating Clinical Trials network, where CF Registry data are used to help facilitate clinical trials and MyCFLifePortal, where patients at participating clinics can view their health data that is entered into the CF Registry.

We deeply value the information provided by the Canadian Cystic Fibrosis Registry and know how fortunate we are to have it. On behalf of Cystic Fibrosis Canada, we would like to thank the clinic staff and the patients who contribute to the CCFR and our donors, without your commitment and contributions, this incredible resource would not be possible.

Sincerely,



Kelly Grover
President and CEO
Cystic Fibrosis Canada



Dr. Anne Stephenson
Medical Director, Registry, Cystic Fibrosis
Canada
CF Physician, Unity Health Toronto, St.
Michael's site, Toronto

2020 HIGHLIGHTS FROM THE CANADIAN CF REGISTRY



4,332 CANADIANS WITH
CYSTIC FIBROSIS

DEMOGRAPHICS

87 NEW CF
DIAGNOSES
64 THROUGH
NEWBORN SCREENING



62%
ARE ADULTS

36% TRAVELLED MORE
THAN **100 KM** TO
RECEIVE CF CARE



CYSTIC FIBROSIS TREATMENT & CARE

 **18,000+** CLINIC VISITS

 **17,100+** HOSPITAL DAYS

 **13,600+** HOME IV DAYS

 **21** LUNG TRANSPLANTS



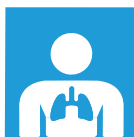
32% HAVE *PSEUDOMONAS*
AERUGINOSA INFECTIONS



34% OF ADULTS HAVE
CF-RELATED DIABETES

HEALTH OUTCOMES

SURVIVAL



ESTIMATED MEDIAN POST-LUNG
TRANSPLANT SURVIVAL IS **10.7 YEARS**



ESTIMATED MEDIAN AGE OF
SURVIVAL IS **55.4 YEARS OF AGE**

64% OF ADULTS
AND
75% OF CHILDREN

HAVE AN
ADEQUATE WEIGHT



MEDIAN
LUNG FUNCTION



64.7%
FOR
ADULTS



93.4%
FOR
CHILDREN

INTRODUCTION

The Canadian Cystic Fibrosis Registry (CF Registry) is a collection of national cystic fibrosis patient data used to support and improve our knowledge and understanding of CF. This extensive resource has been involved in many important studies resulting in achievements in improving health outcomes for those living with cystic fibrosis.

Participating individuals who attend any of the accredited 42 CF clinics across Canada are represented in the CF Registry. Data are submitted by the CF clinics on behalf of patients. Given that most people living with CF attend one of these clinics, and nearly all consent to contributing their data, we are confident that the CF Registry includes data on virtually all Canadians diagnosed with cystic fibrosis — giving a comprehensive picture of the CF population in this country.

Cystic Fibrosis Canada publishes the Canadian CF Registry Annual Data Report on national summary statistics to further educate and promote awareness of CF. We would like to acknowledge the involvement and continued participation of those living with cystic fibrosis who consent to having their data submitted, and the exceptional effort and contribution from CF clinic team members who collect and enter the data.

■ HOW TO READ THIS REPORT

This Annual Data Report contains data from individuals diagnosed with CF who have consented to participate in the CF Registry and who were reported on by a Canadian CF clinic in 2020, including those who were born, diagnosed with CF or died in 2020. Data from individuals with a diagnosis of CF screen positive, inconclusive diagnosis (CFSPID) or CFTR-related disease are excluded from this report.

All the data analyses presented in this report have been recalculated in order to include data that might have been updated or missed in previous years. These recalculations ensure that data can be compared accurately between different years within this report. It also means that discrepancies might occur when comparing historical reports with the current one.

Individuals who are under 18 years of age are categorized as children and those 18 years of age or older are categorized as adults. For the purposes of this report, age is calculated as of December 31, 2020.

The year 2020 saw many disruptions to typical CF care and submission of data to the CF Registry, and some of these are reflected in the data presented in this report. The information presented below is aimed to provide some context to the information presented in the Canadian CF Registry 2020 Annual Data Report, and the reader is encouraged to interpret any temporal trends with caution, and in the context of these changes.

■ CHANGES TO THE CANADIAN CF REGISTRY AND CF CARE IN CANADA IN 2020

The COVID-19 pandemic has undoubtedly changed the face of CF care delivery at most, if not all, CF clinics across Canada, leading to restrictions on in-person interactions, temporary suspension of some procedures (for example, pulmonary function testing, sputum cultures), and re-deployment of CF clinic staff. Throughout the course of the pandemic, CF Canada has worked closely with CF clinics to complete data entry for 2020. However, these changes may be reflected in CF Registry data collection and reporting in 2020 due to disruptions in the CF care process and potentially incomplete data entry. Some key considerations when reviewing the 2020 Annual Data Report are described below.

The first confirmed case of COVID-19 in Canada was on January 25, 2020, and the World Health Organization (WHO) formally declared a COVID-19 pandemic on March 11, 2020. In the face of the COVID-19 pandemic, existing fields within the CF Registry were adapted to collect data on COVID-19 testing and infection, ensuring continued progress for our mission by capturing and identifying trends related to the health of people living with CF.

On April 10, 2020, new options for other therapies, “ECMO (extracorporeal membrane oxygenation)”, “mechanical ventilation”, and “in-hospital oxygen” were implemented to better capture any potential interventions in the CF Registry. Following feedback from clinics, a “location” field was also added to capture the location in which clinical measurements were taken in order to identify and differentiate between virtual and in-person visits. This option was implemented on December 9, 2020, with nearly 70% of clinical measurements in 2020 having a location entered.

While the COVID-19 pandemic certainly had a measurable impact on CF care, it is also important to note that starting in late January 2020, some eligible Canadians living with CF were approved to access the triple combination CFTR modulator therapy elxacaftor/tezacaftor/ivacaftor through the Special Access Program which provides compassionate access to those who meet the eligibility criteria.

DATA COVERAGE IN 2020

Data from 2020 showed that there were fewer overall healthcare encounters compared to previous years, with a concomitant decrease in the number of clinical measurements, hospitalizations, and home IV courses, as well as microbiology and mycobacteria samples. It follows that the overall percentage of individuals with at least one data measurement reported in 2020 also decreased from 2019, with 9% fewer individuals aged 6 years old and older having a recorded FEV₁ percent predicted.

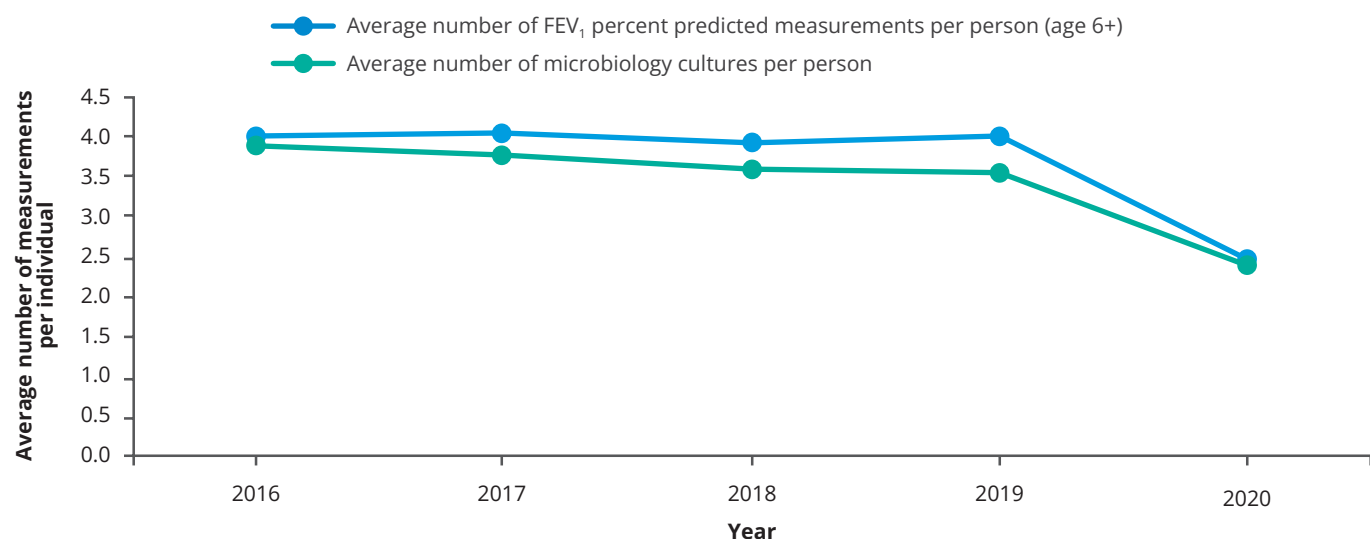
Percentage of individuals with at least one data measurement reported

	Report Year					Change from 2019-2020
	2016	2017	2018	2019	2020	
BMI or BMI percentile	98.4%	98.2%	98.3%	98.8%	91.6%	-7.1%
FEV₁ percent predicted (age 6+)	97.1%	96.4%	96.6%	97.6%	88.8%	-8.8%
Microbiology Culture (excluding COVID-19 testing)	93.7%	92.5%	92.3%	91.7%	85.5%	-6.2%
Mycobacterial Culture	27.6%	30.4%	33.5%	35.7%	30.4%	-5.3%

From 2016 to 2019, the average number of FEV₁ percent predicted measurements per individual was 4.0 measurements per year, which fell to an average of 2.5 measurements per individual in 2020 among individuals aged 6 years and older.

Similarly, the number of microbiology cultures reported per individual prior to 2020 was an average of 3.7 cultures per year, which dropped to an average of 2.4 cultures per individual in 2020. It should be noted that the number of microbiology cultures sampled can impact the likelihood of detection of microorganisms.

Number of FEV₁ percent predicted measurements and microbiology cultures entered per individual, 2016 to 2020.



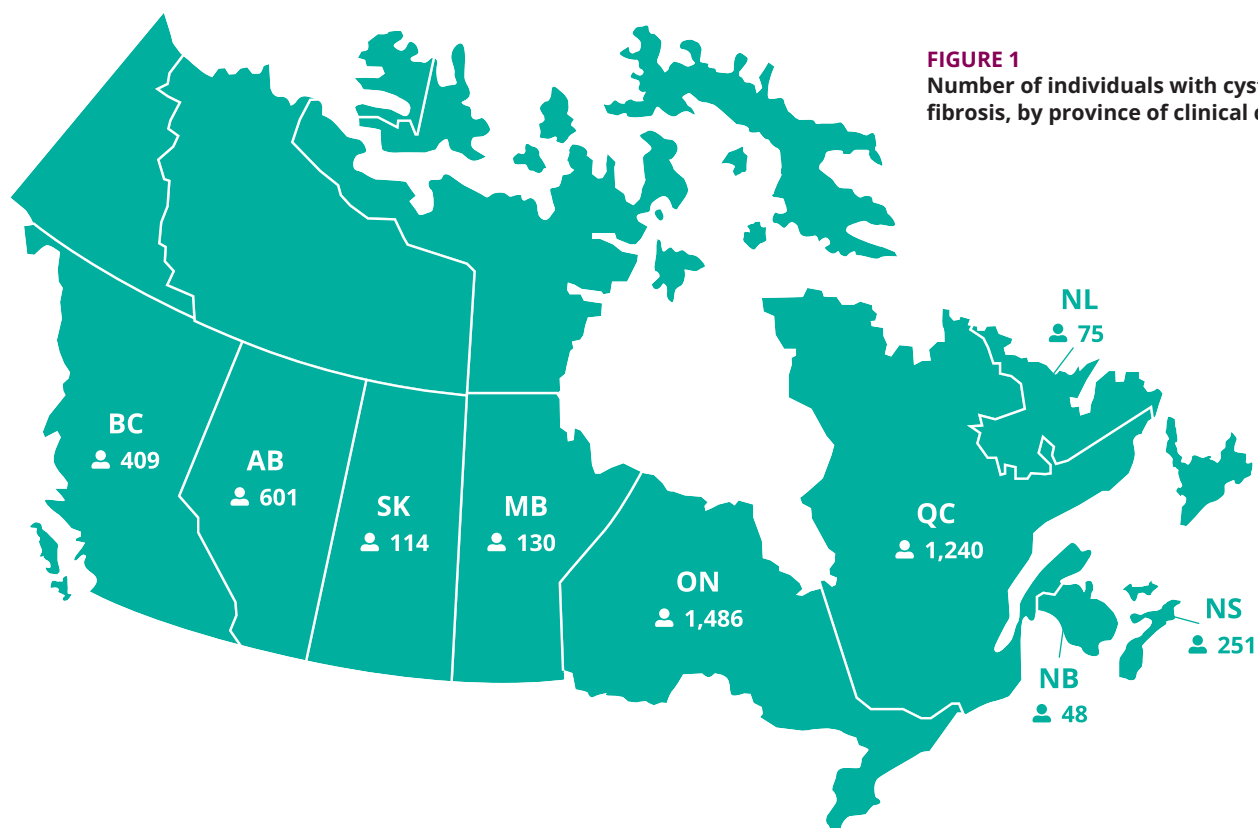
At the end of the report, we present a section on COVID-19, comparing the testing and characteristics of Canadians with CF who tested positive for SARS-CoV-2 (the virus that causes COVID-19) in 2020 against the general Canadian CF population. The CF Registry team continues to monitor the effects of COVID-19 and highly effective CFTR modulator therapies on the CF population.

DEMOGRAPHICS

CANADIANS WITH CYSTIC FIBROSIS

In 2020, there were a total of 4,332 individuals with cystic fibrosis who attended one of the 42 accredited CF clinics across Canada (Figure 1), with 87 of those being newly diagnosed with cystic fibrosis. Fewer people were reported on in 2020 compared to 2019, which may be due to the ongoing COVID-19 pandemic and potential disruptions to typical CF care. Overall, the total Canadian CF population has been steadily increasing and in the last two decades, has grown by 31% since 2001. (Figure 2).

Individuals are associated with the province in which they attended a CF clinic. Those who attended CF clinics in multiple provinces in 2020 will be counted in each of those provinces for provincial-level statistics, and therefore these figures should not be summed to obtain a national total. However, individuals are only counted once (i.e. unique individuals) in the national reported numbers.

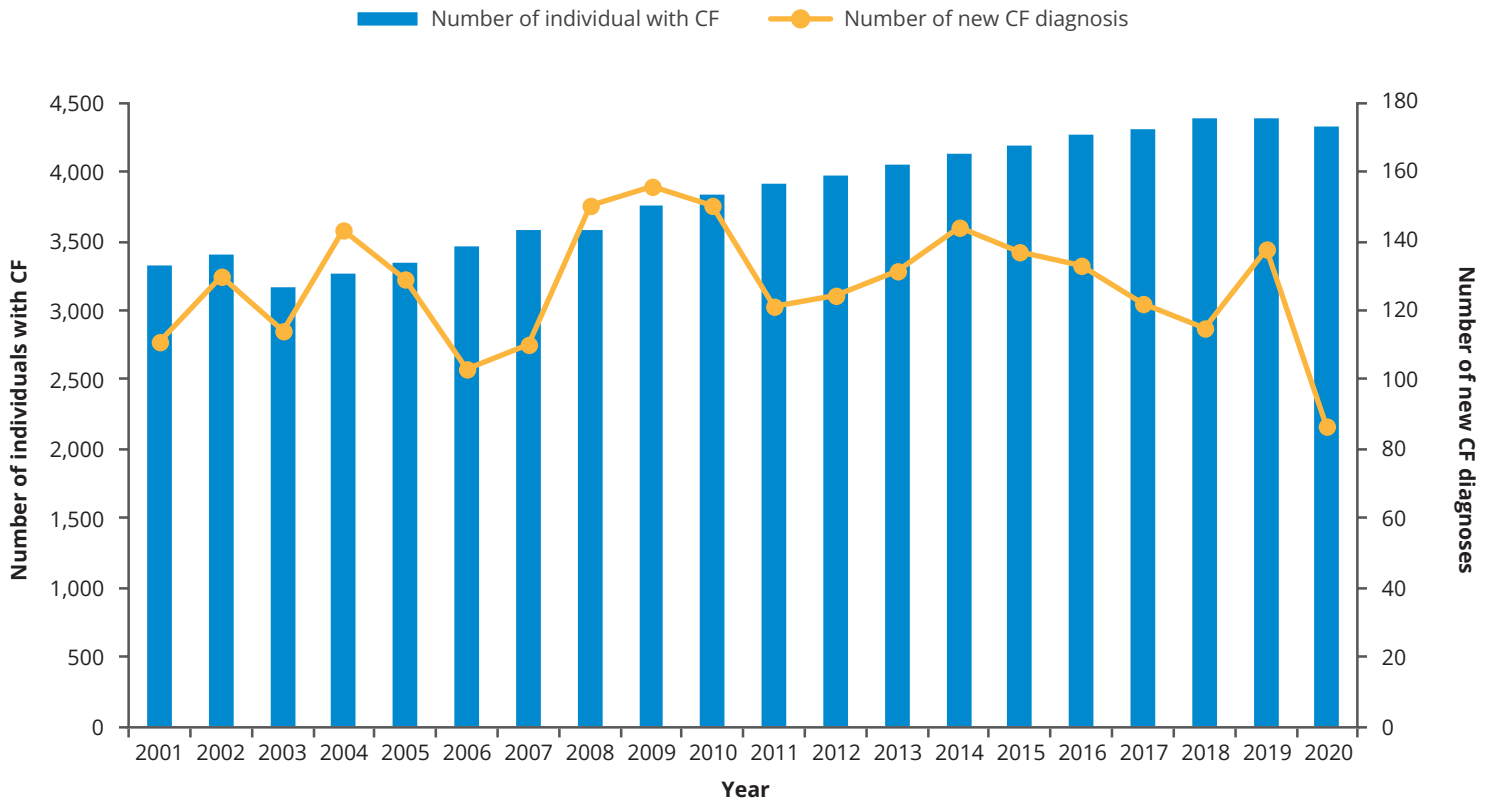


PROVINCE OF CLINICAL CARE*	NUMBER OF INDIVIDUALS WITH CF	FEMALE	MALE	ADULT	CHILDREN
BC	409	178	231	261	148
AB	601	295	306	350	251
SK	114	45	69	58	56
MB	130	57	73	70	60
ON	1,486	710	776	921	565
QC	1,240	587	653	800	440
NB	48	26	22	35	13
NS	251	118	133	154	97
NL	75	28	47	51	24

* Individuals with cystic fibrosis living in provinces or territories not listed here are included, if reported on by CF clinics in other jurisdictions.

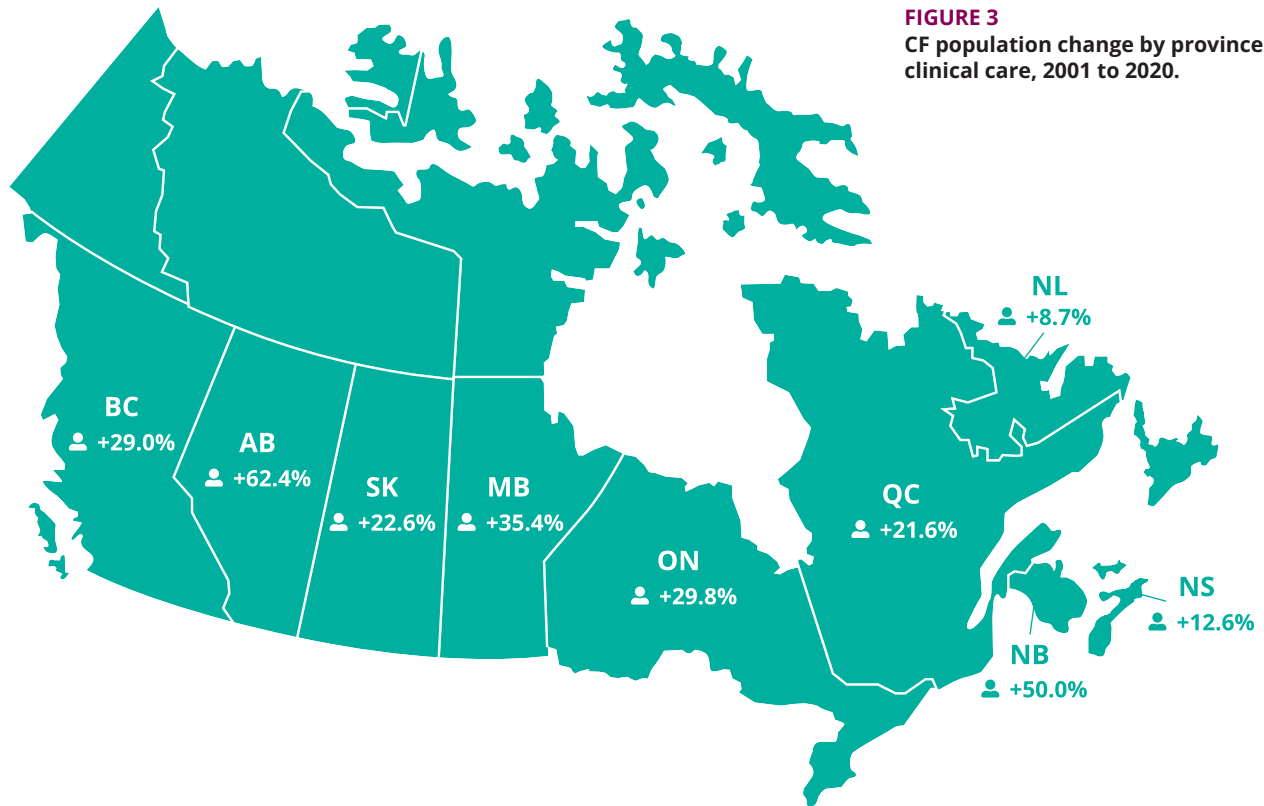
CANADIANS WITH CYSTIC FIBROSIS

FIGURE 2
Total number of individuals with cystic fibrosis and new CF diagnoses, 2001 to 2020.



PROVINCIAL POPULATION CHANGE

Figure 3 shows the number of individuals with CF reported on by clinics within each province in 2001 and 2020, along with the percent change. It should be noted that during this time period, provinces began including CF within their newborn screening (NSB) programs, beginning with Alberta in 2007.



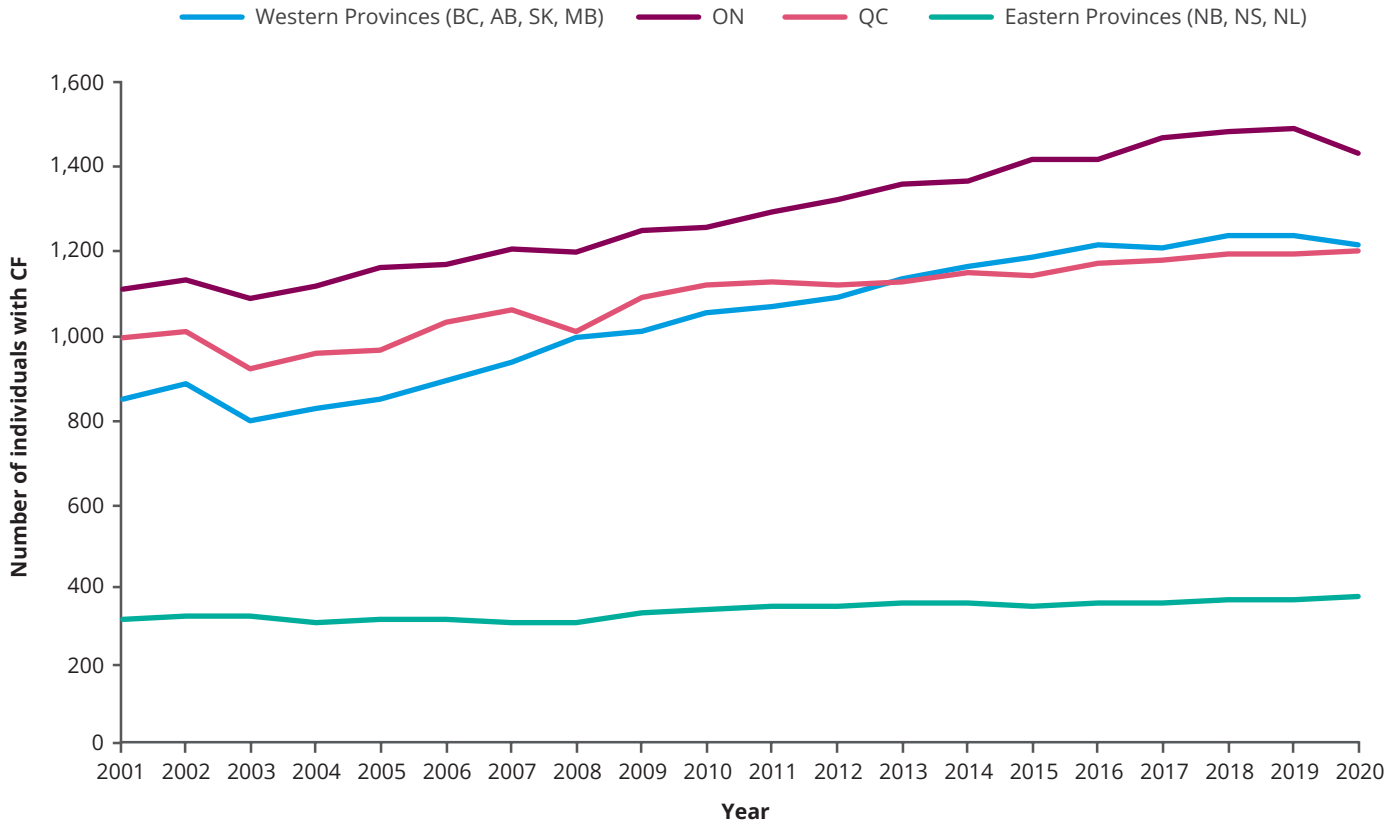
PROVINCE OF CLINICAL CARE*	2001	2020	PERCENT CHANGE
BC	317	409	29.0%
AB	370	601	62.4%
SK	93	114	22.6%
MB	96	130	35.4%
ON	1,145	1,486	29.8%
QC	1,020	1,240	21.6%
NB	32	48	50.0%
NS	223	251	12.6%
NL	69	75	8.7%

* Individuals with cystic fibrosis living in provinces or territories not listed here are included, if reported on by CF clinics in other jurisdictions.

PROVINCIAL POPULATION CHANGE

Figure 4 shows the growth of the Canadian CF population in four regions across the country, over the past 20 years.

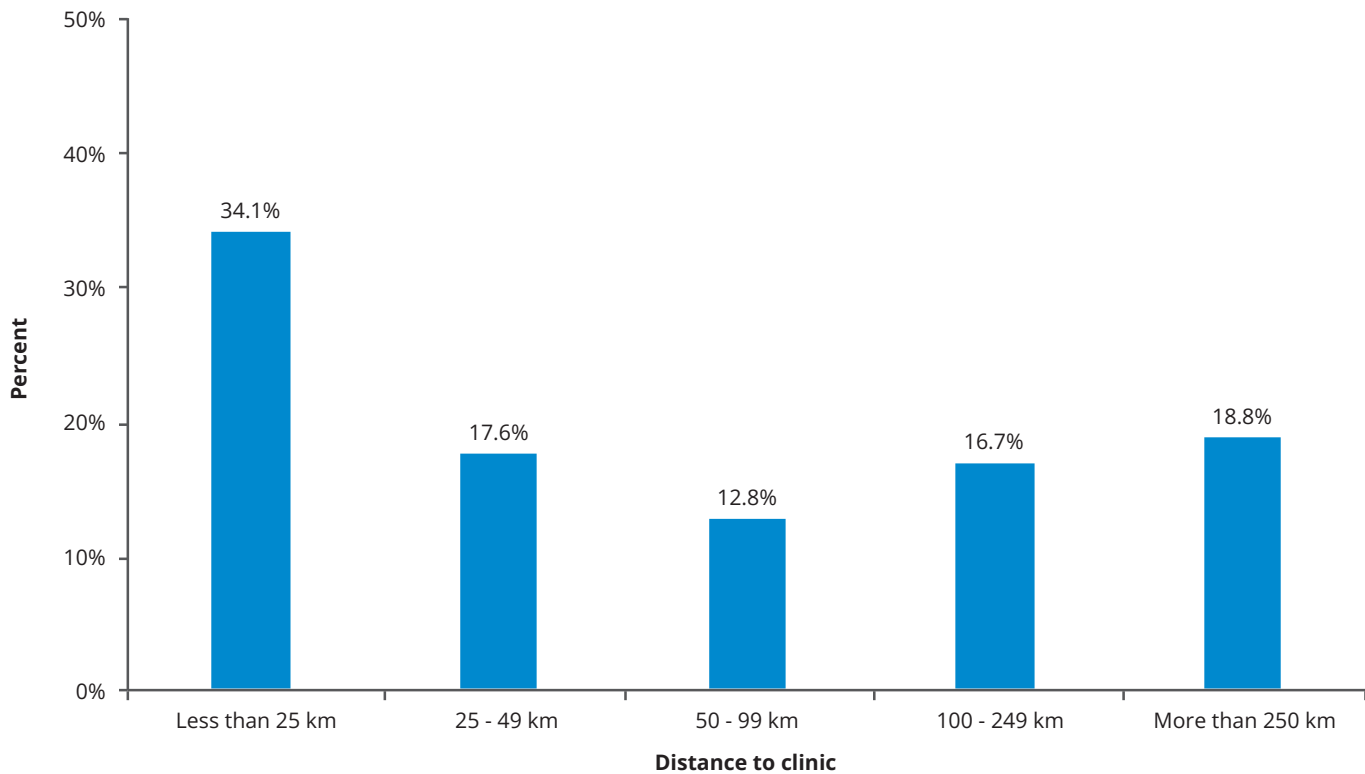
FIGURE 4
CF population growth by region of clinical care, 2001 to 2020.



DISTANCE TO CLINICS

The CF Registry began collecting the location of residence of those living with cystic fibrosis in 2015, through the first three digits of their postal code, or the forward sortation area (FSA). Distances to the reporting clinic were calculated in kilometers (km) using the fastest driving route. In 2020, there were 1,811 (41.8%) cystic fibrosis individuals with at least one valid location recorded (Figure 5). While 51.7% of those with a reported location attended a CF clinic within 50 km of where they live, 35.5% travelled more than 100 km and 18.8% travelled more than 250 km for their CF care.

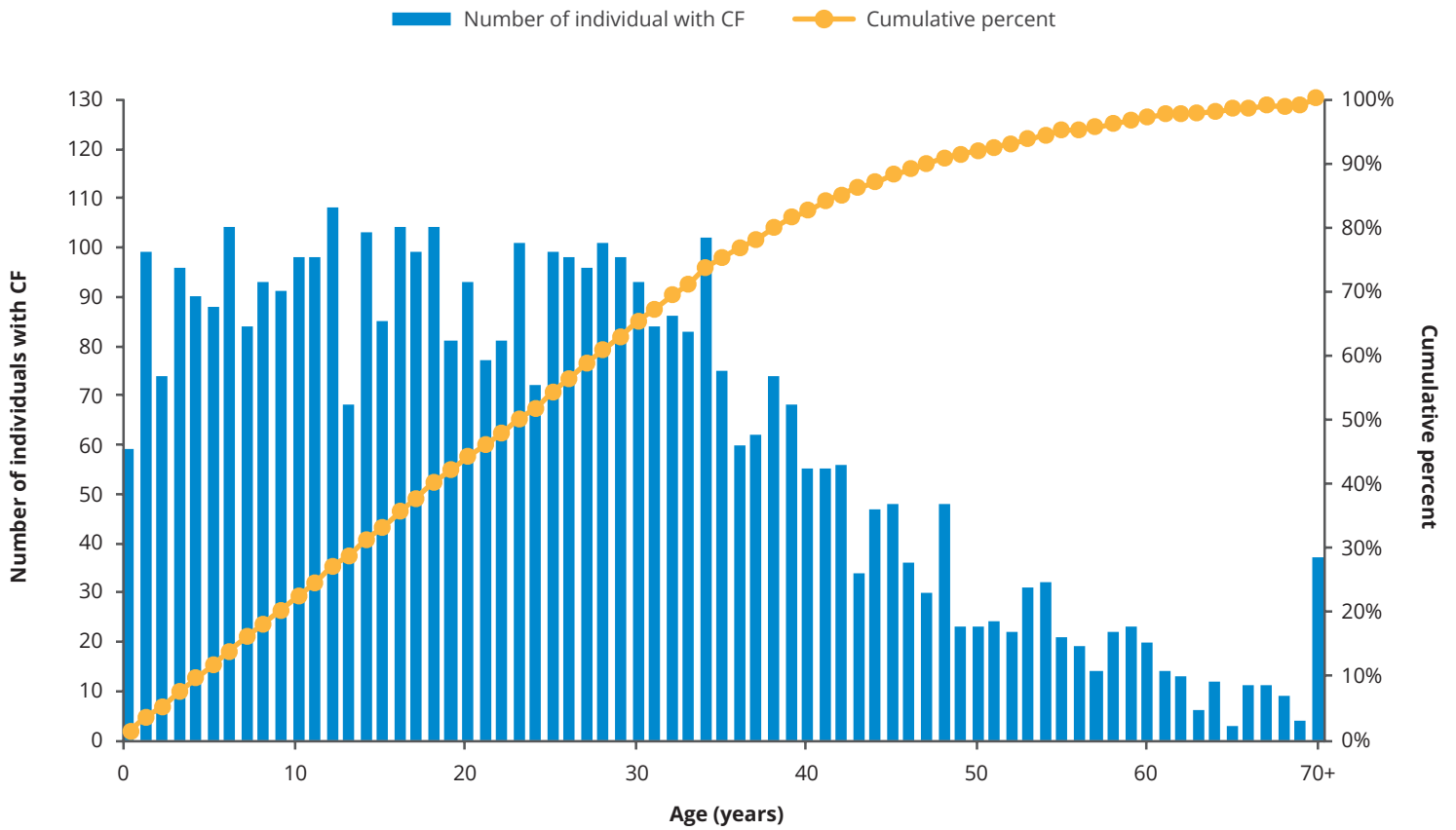
FIGURE 5
Distance travelled to clinic for individuals with cystic fibrosis (N = 1,811), 2020.



AGE DISTRIBUTION OF CANADIANS WITH CYSTIC FIBROSIS

Figure 6 shows the age distribution of the 4,332 Canadians living with cystic fibrosis in 2020.

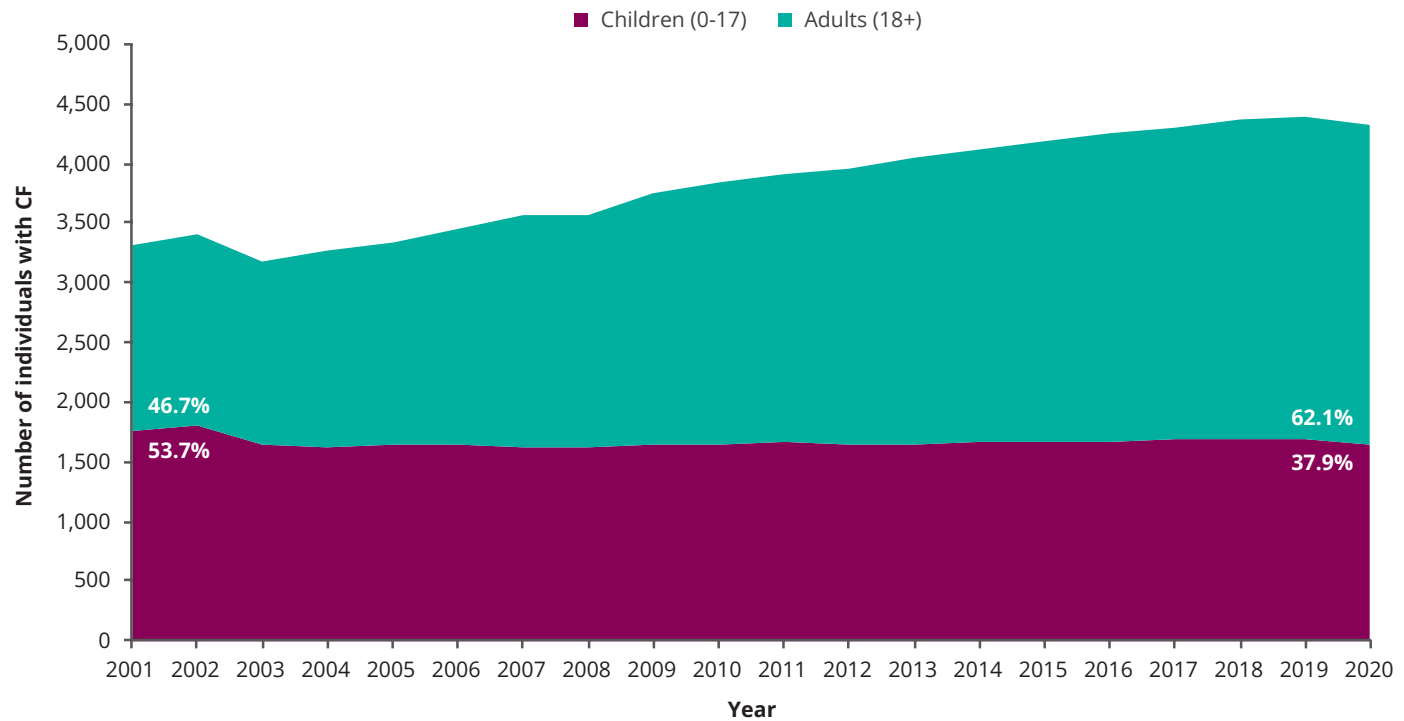
FIGURE 6
Age distribution of individuals with cystic fibrosis, as of December 31, 2020.



AGE DISTRIBUTION OF CANADIANS WITH CYSTIC FIBROSIS

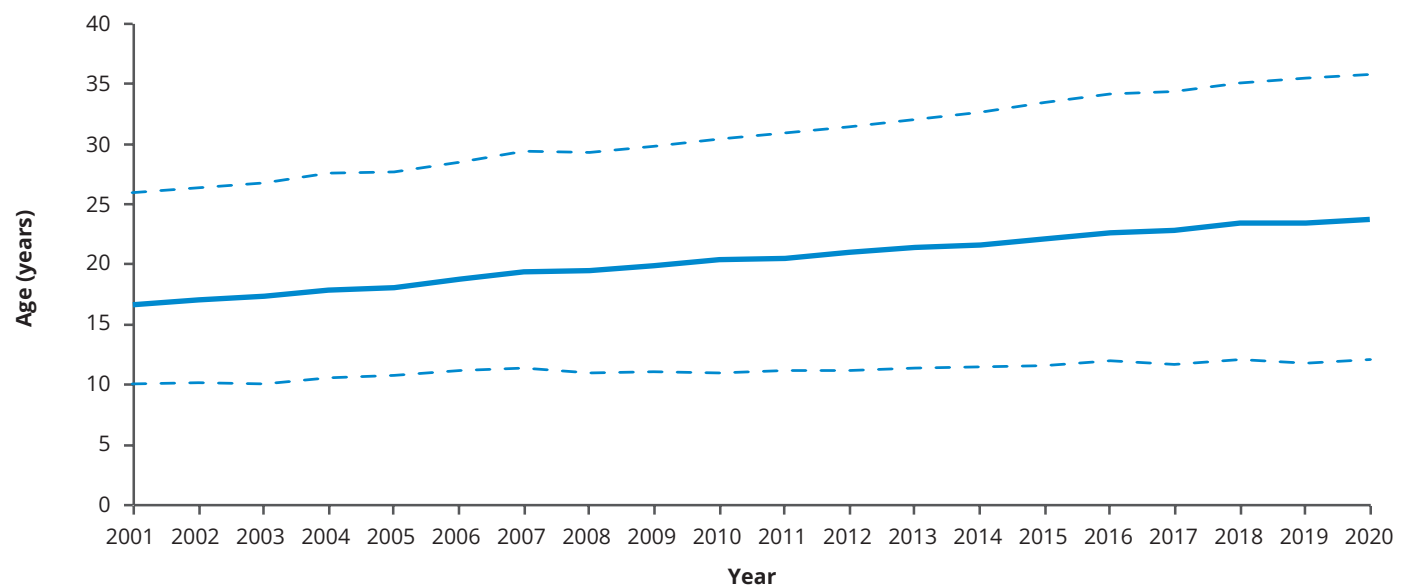
Improvements in treatment and care in the last few decades have led to an increase in the number of Canadian adults living with cystic fibrosis. Twenty years ago, less than half of all Canadians living with cystic fibrosis were adults (individuals aged 18 years and older) (Figure 7). In 2020, there were 2,691 adults living with CF, accounting for 62.1% of the Canadian CF population, and 803 (18.5%) adults aged 40 years and over.

FIGURE 7
Number of children and adult living with cystic fibrosis, 2001 to 2020.



It follows that the median age of individuals with cystic fibrosis has increased steadily over the past 20 years. From just under 17 years in 2001, to 23.8 years among those reported on in 2020 (shown in Figure 8 along with the 25th and 75th percentile of ages).

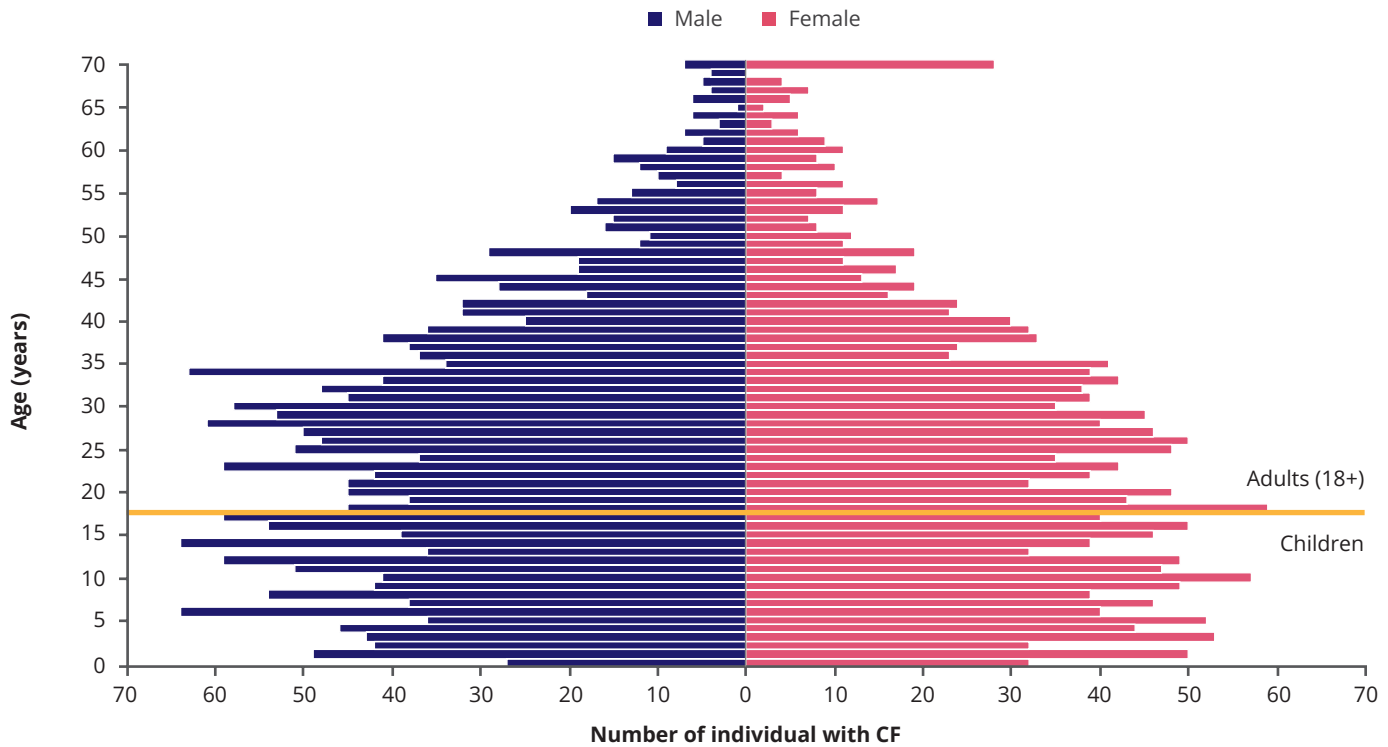
FIGURE 8
Median age of individuals with cystic fibrosis 2001 to 2020.



AGE AND SEX DISTRIBUTION OF CANADIANS WITH CYSTIC FIBROSIS

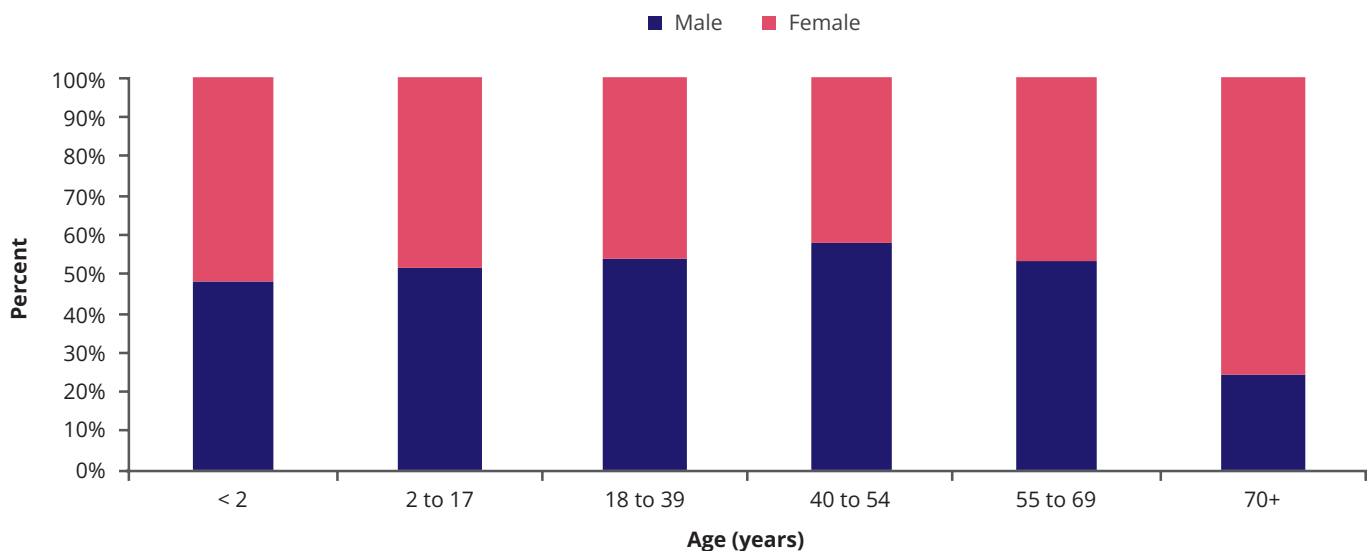
Figure 9 shows the age-sex distribution (referring to biological sex) for all individuals reported on in 2020. Of the 4,332 individuals reported on in 2020, 2,304 (53.2%) were male and 2,028 (46.8%) were female.

FIGURE 9
Population distribution of individuals (N = 4,332) with cystic fibrosis, by age and sex, as of December 31, 2020.



The sex distribution of those living with CF differed by age group. As seen in Figure 10, children under age 18 years were fairly evenly distributed between the sexes, with the proportion of males increasing into adulthood before reaching a peak of 58.2% male for those aged 40 to 54 years. After age 55, the proportion of females begins to increase to 75.7% female for those aged 70+.

FIGURE 10
Sex distribution of individuals with cystic fibrosis, by age group, as of December 31, 2020.



DIAGNOSIS

AGE AT DIAGNOSIS

There were 87 new diagnoses of CF in 2020. 68.5% of individuals with cystic fibrosis reported on in 2020 and with a recorded diagnosis date, were diagnosed before the age of one year, and nearly three quarters (74.0%) were diagnosed by the age of two years (Figure 11). Adults diagnoses, those diagnosed at 18 years and older, accounted for only 7.5% of all individuals diagnosed in 2020.

FIGURE 11
Age at diagnosis of cystic fibrosis individuals, as of December 31, 2020 (N = 4,257).

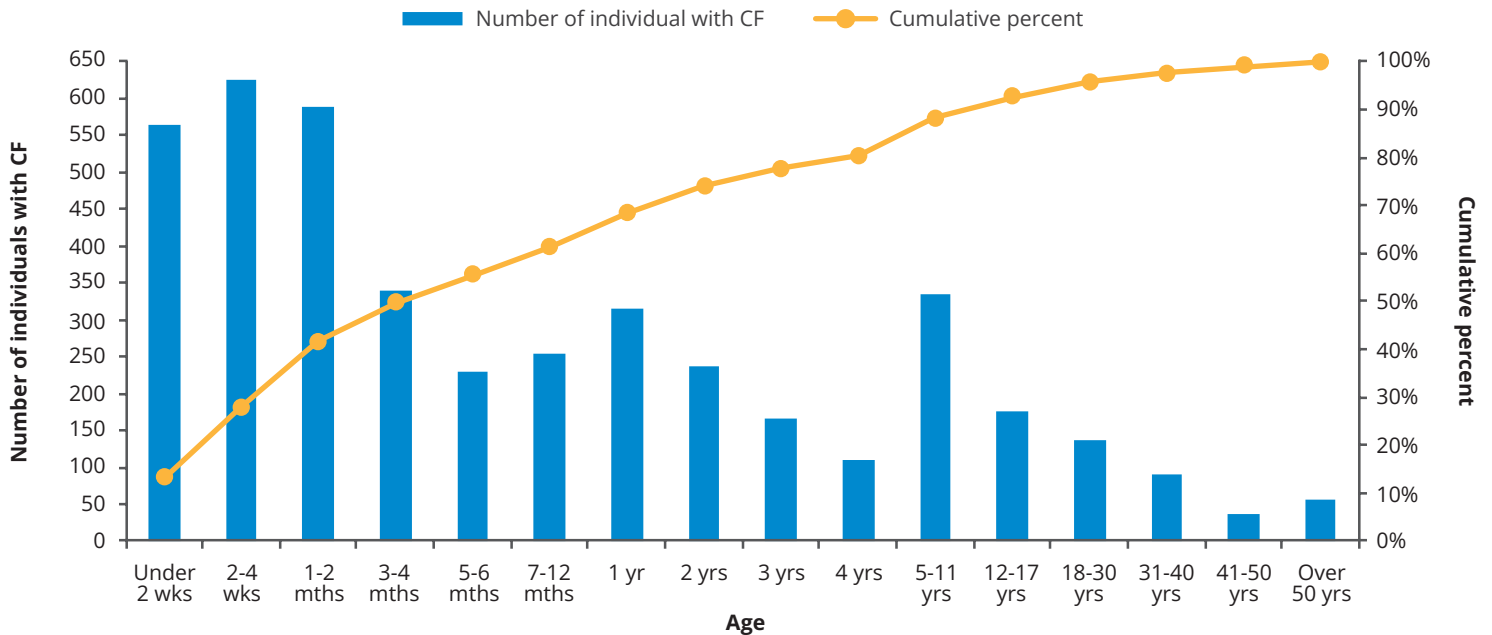
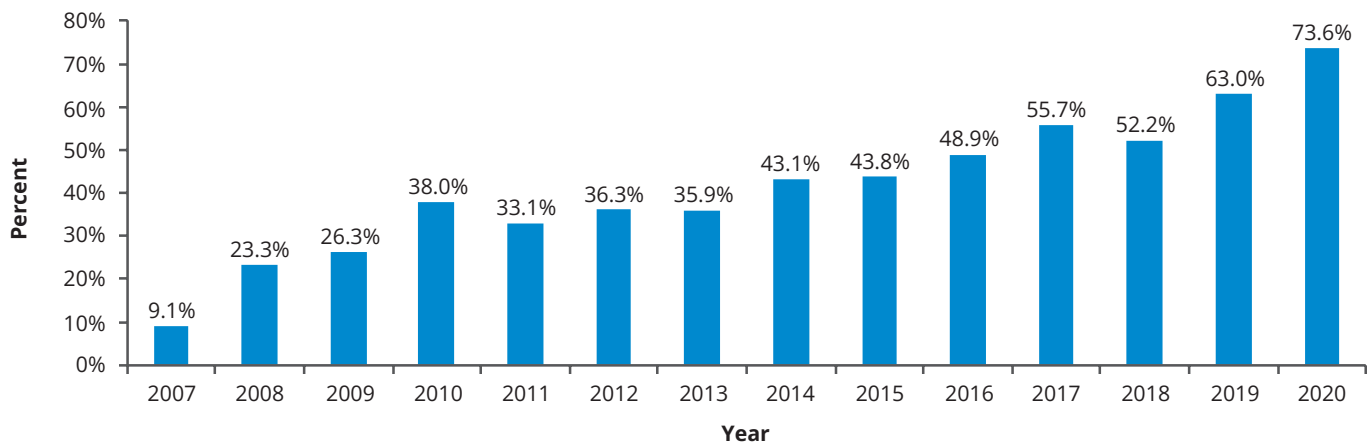


Figure 12 shows the percentage of newborns diagnosed through provincial newborn screening (NBS) programs since 2007, when NBS for CF started in Alberta. At that time, only 9% of new CF diagnoses were identified through NBS. In the spring of 2018, Quebec became the last jurisdiction to start screening newborns for cystic fibrosis. In 2020, nearly three quarters of new diagnoses (64; 73.6%) were made through NBS. Newborn screening is now in practice for all provinces across Canada and remains essential for early diagnosis and intervention.

FIGURE 12
Percentage of all new CF diagnoses made through the NBS program, 2007 to 2020.



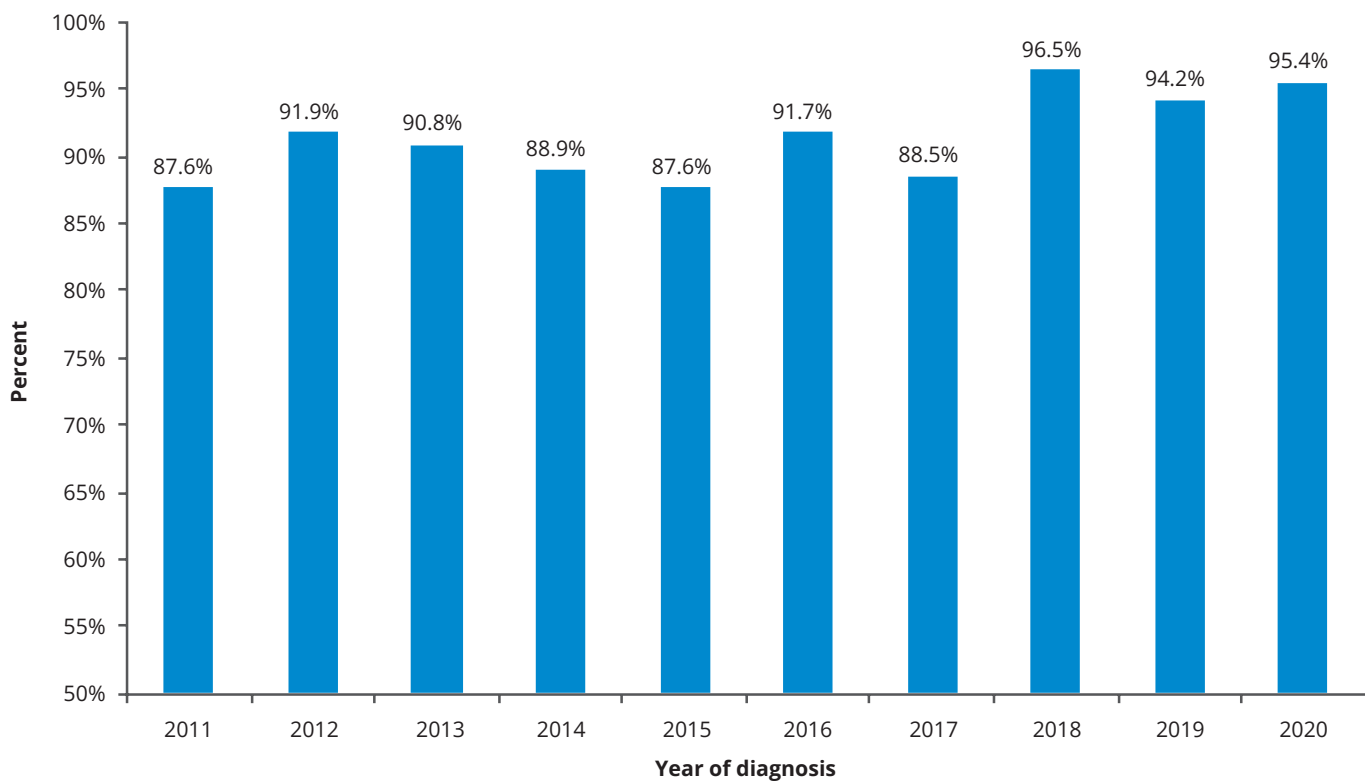
SWEAT CHLORIDE TESTING

Sweat chloride testing can help provide a CF diagnosis by measuring the concentration of salt in a person's sweat. Doctors will order a chloride sweat test for kids with positive newborn screen for cystic fibrosis, a family history of cystic fibrosis, or symptoms of the disorder. Sweat chloride testing is the most reliable way to diagnosis CF. It is also routinely used as part of CFTR modulator initiation and monitoring.

Individuals with cystic fibrosis typically have a sweat chloride value greater than 60 mmol/L whereas values between 40 and 59 mmol/L are indeterminate. Values lower than 40 mmol/L are considered in the normal range.

The CF Registry began capturing sweat chloride test results in 2011. Since 2011, the number of newly diagnosed individuals with at least one sweat chloride test has remained fairly stable (Figure 13). In 2020, 83 of the 87 (95.4%) newly diagnosed individuals had at least one sweat chloride test result recorded.

FIGURE 13
Percentage of newly diagnosed individuals with at least one sweat chloride test, 2011 to 2020.



GENOTYPE

CF is caused by mutations in one or more alleles in a single gene located on chromosome 7, called the cystic fibrosis transmembrane conductance regulator (CFTR) gene. The CFTR gene codes for the CFTR protein which functions as a chloride channel and is involved in many cellular functions. To date, more than 2,000 different mutations in the *CFTR* gene have been identified ¹.

By far, the most common CF mutation in Canada 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**. F508del is also the most common mutation worldwide, however, the distributions of mutations can vary widely depending on location, ethnic background and other factors. CF disease-causing mutations can be classified into five major categories depending on how the mutation impacts the production and function of the CFTR protein. There are some mutations where the impact on the CFTR protein is not clear or unknown therefore these cannot be classified. CFTR protein modulator medications target specific classes of mutations. Table 1 summarizes the classification of CFTR gene mutations.

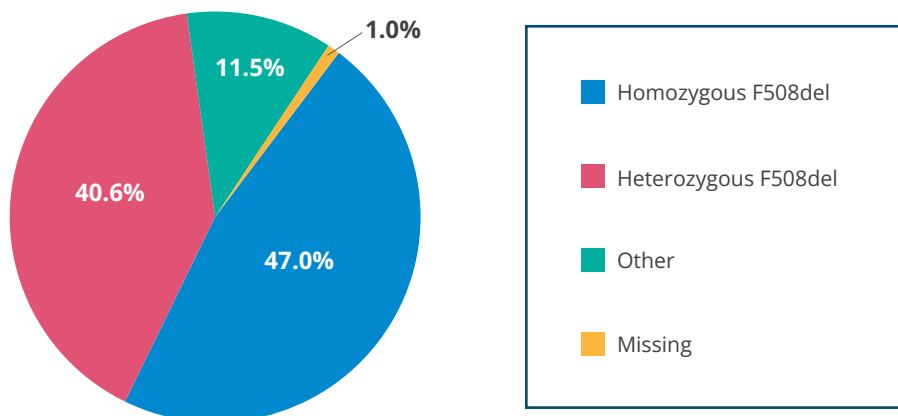
TABLE 1
Classification of CFTR gene mutations based on the impact on the CFTR protein.

CLASS	HOW CFTR PROTEIN IS AFFECTED	EXAMPLES
I	No functional CFTR protein is made.	G542X, W1282X, 621+1G->T
II	CFTR protein is abnormal and destroyed by the cell before it reaches the cell membrane.	F508del, G85E
III	CFTR protein reaches the cell membrane but the channel is blocked.	G551D
IV	CFTR protein reaches the cell membrane but the channel does not move chloride the way it should.	R117H, R334W
V	The CFTR protein is made and works properly but the quantity of protein made is insufficient.	3849+10kbc->T

Out of 5,446 people ever reported on in the CF Registry who are still alive, only 382 people have no mutations recorded. Out of those, the majority (91.6%) are adults, 8.4% are under 18 years of age, and only 0.3% are under 1 year of age.

Nearly all individuals with cystic fibrosis reported on in 2020 (4,290; 99%) had at least one CFTR gene mutation recorded. Almost half (2,034; 47%) have two copies of the F508del mutation (referred to as homozygous F508del) and 40.6% carry a single copy of the F508del mutation (referred to as heterozygous F508del). In total, almost 90% carry at least one copy of the F508del mutation (Figure 14). Individuals with more severe disease symptoms are generally diagnosed earlier; milder forms of cystic fibrosis may only be diagnosed in adulthood.

FIGURE 14
Genotype distribution of CF population (N = 4,332), 2020.



GENOTYPE

Figure 15 shows the genotype distribution of the CF population by the age of diagnosis. Those diagnosed as a child (under 18 years) were more likely to be homozygous F508del (50.5%) while those diagnosed as an adult (18 years and older) were more likely to be heterozygous F508del (63.9%).

FIGURE 15
Genotype distribution of individuals with cystic fibrosis, by diagnosis age group (N = 4,332), 2020.

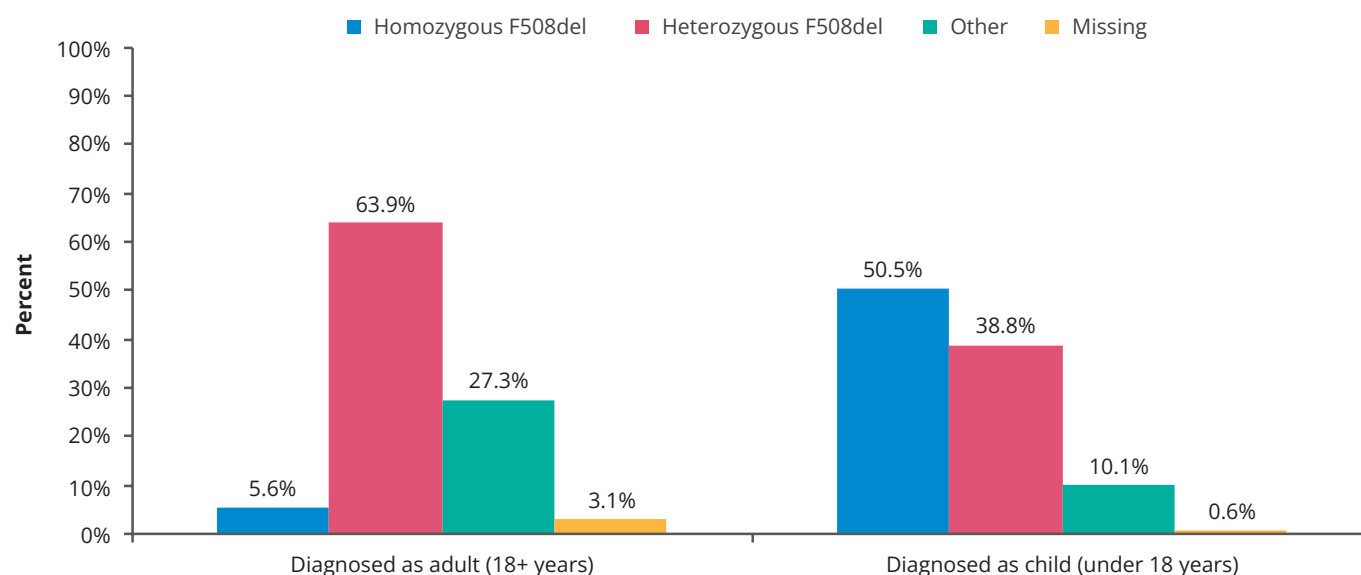


Table 2 lists the most common CFTR gene mutations for individuals with cystic fibrosis reported in 2020. After F508del, 621+1G->T is the next most frequent CFTR gene mutation identified in 6% of the population.

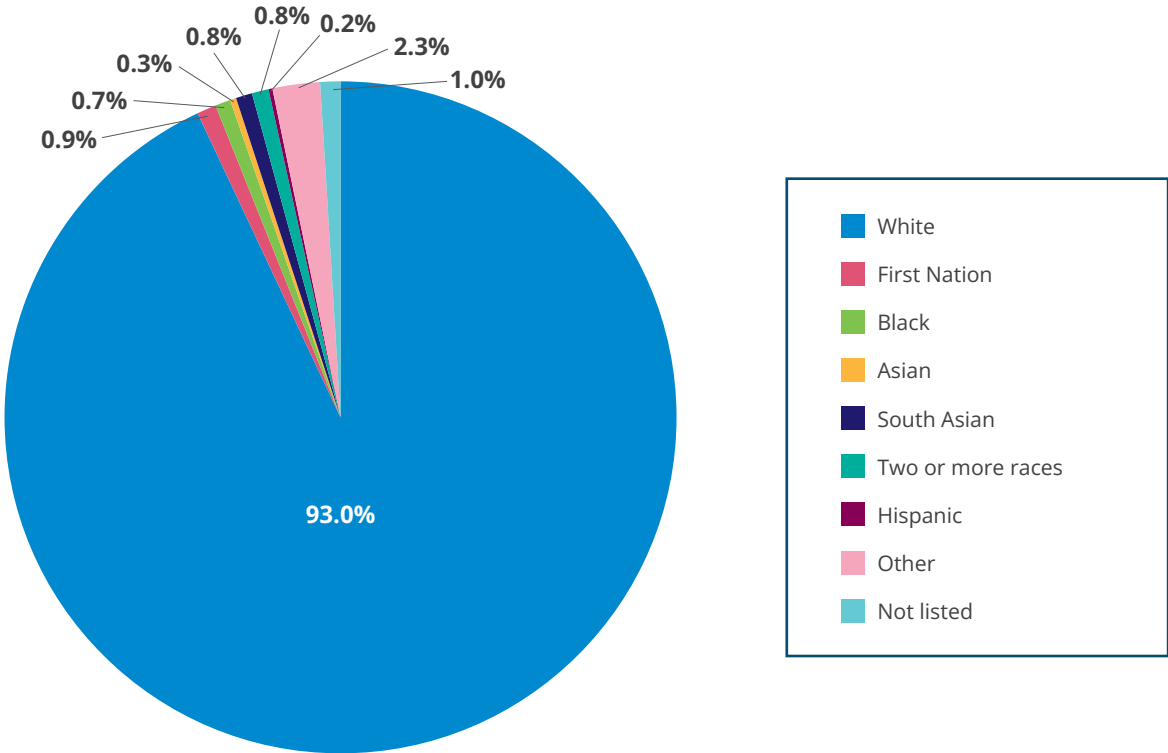
TABLE 2
Frequency of the top 10 most common CFTR gene mutations on one or both alleles of cystic fibrosis individuals with recorded mutations (N = 4,290), 2020.

MUTATION	NUMBER	PERCENTAGE
F508del	3,791	88.4%
621+1G->T	259	6.0%
G542X	146	3.4%
G551D	136	3.2%
711+1G->T	118	2.8%
A455E	112	2.6%
L206W	110	2.6%
N1303K	91	2.1%
M1101K	69	1.6%
G85E	68	1.6%

ETHNICITY

Cystic fibrosis can affect people of all ethnicities anywhere in the world. The majority (93%) of the Canadian CF population is White. Of those remaining who have an identified ethnicity (Figure 16), they are divided among five other ethnic groups (First Nations, Black, Asian, South Asian, and Hispanic). Ethnicity is captured through self-report.

FIGURE 16
Ethnicity distribution of CF population, 2020.



RESPIRATORY

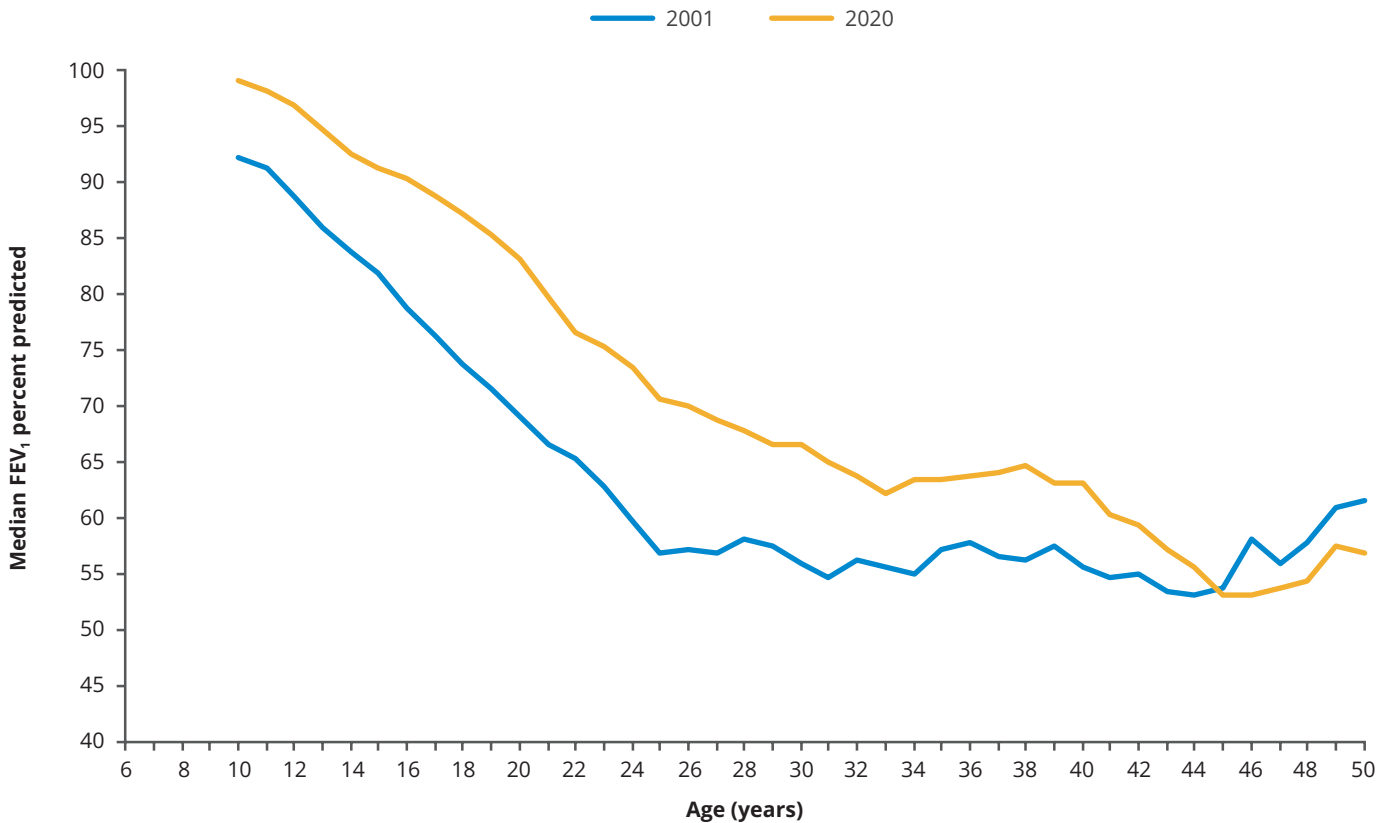
MEDIAN FEV₁ PERCENT PREDICTED

In cystic fibrosis, mucus in the lungs is linked to chronic infections, making it harder to breathe and potentially causing permanent damage to the airways. As such, lung function measurements are critical for evaluating lung health and are reliably measured starting at six years of age. 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 an individual is calculated by comparing the measured FEV₁ to the average FEV₁ of a healthy population of similar age, height, ethnicity, and sex. Global Lung Initiative (GLI) equations are used to calculate the FEV₁ percent predicted values². Though FEV₁ percent predicted is a commonly used measure of lung function, it may not be sensitive enough to detect mild changes in the airways or early lung disease.

In this report, the first complete and stable lung measurement of the year was used per individual with cystic fibrosis to summarize lung function. If none exist, the first complete measurement regardless of the lung status was used. Individuals who have received a lung transplant are excluded from all FEV₁ percent predicted data, as their new non-CF lungs may have lung health similar to a person without CF.

Figure 17 shows the median FEV₁ percent predicted from ages 6 to 50 years using a 5-year moving average window. While at an individual patient level, lung function tends to decline with age, the median FEV₁ percent predicted has increased since 2001 at a population level. The median FEV₁ at 24 years of age (the median age of an individual living with cystic fibrosis) was 71.3% predicted in 2020 compared to 55.7% predicted in 2001, marking an improvement of 15.7% over the last two decades.

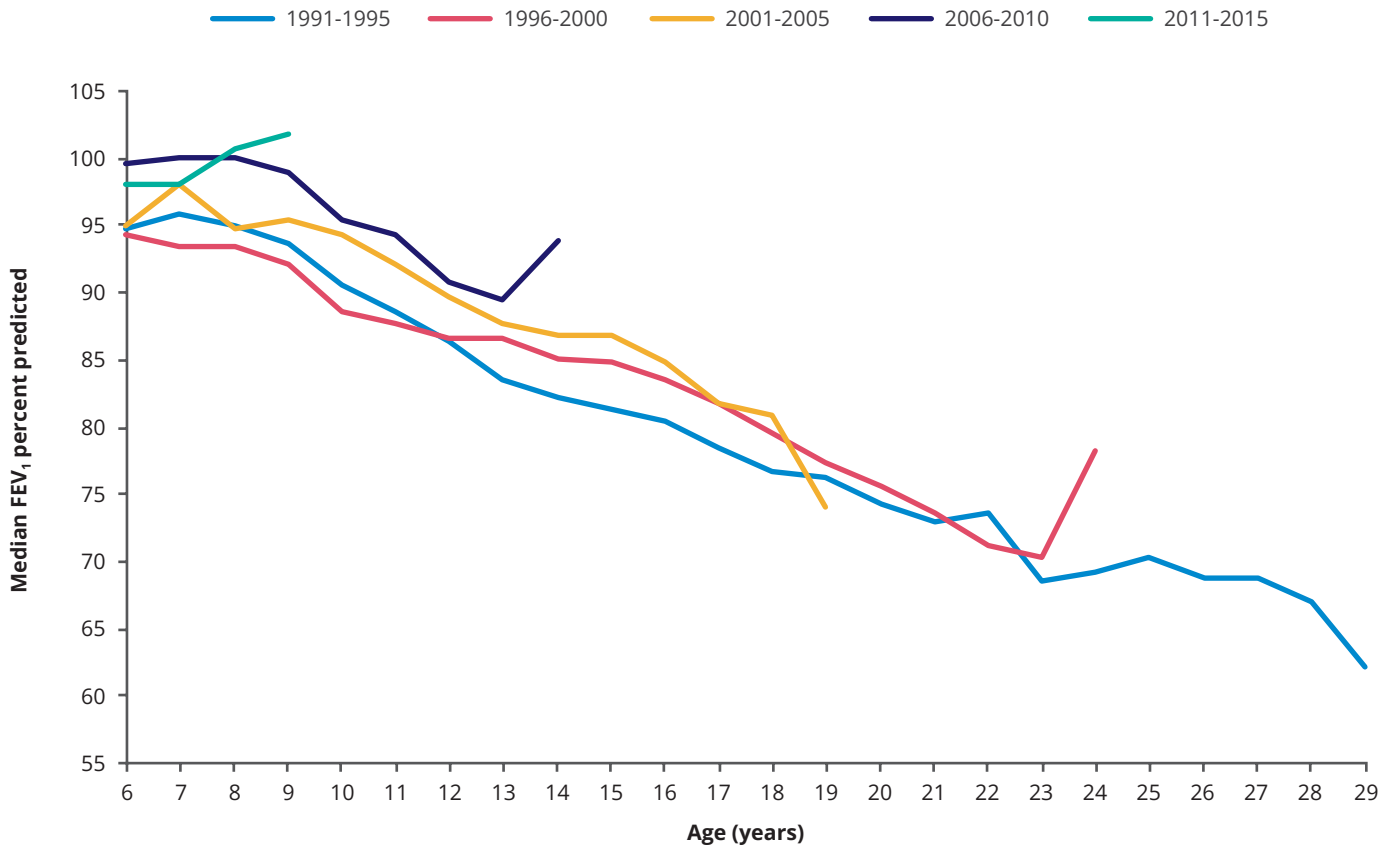
FIGURE 17
Median FEV₁ percent predicted vs. age of cystic fibrosis individuals (5-year moving window), 2001 and 2020.



MEDIAN FEV₁ PERCENT PREDICTED

Individuals born recently have a higher median FEV₁ percent predicted at age 6 years and have a slower rate of decline than those born earlier (Figure 18). The deviations in trends present in some of the birth cohort lines for the later ages are due to the small sample sizes resulting from the method used to group ages in the calculations.

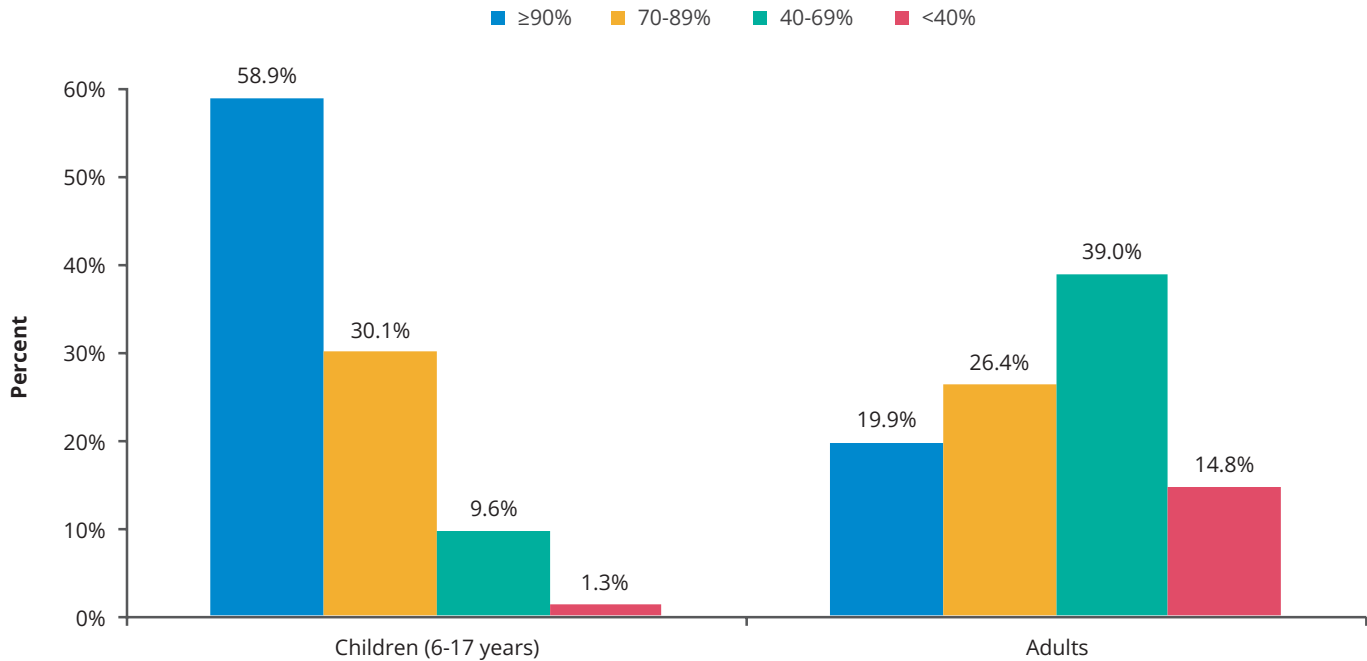
FIGURE 18
Median FEV₁ percent predicted of individuals with cystic fibrosis, by birth cohort, 2020.



RESPIRATORY STATUS

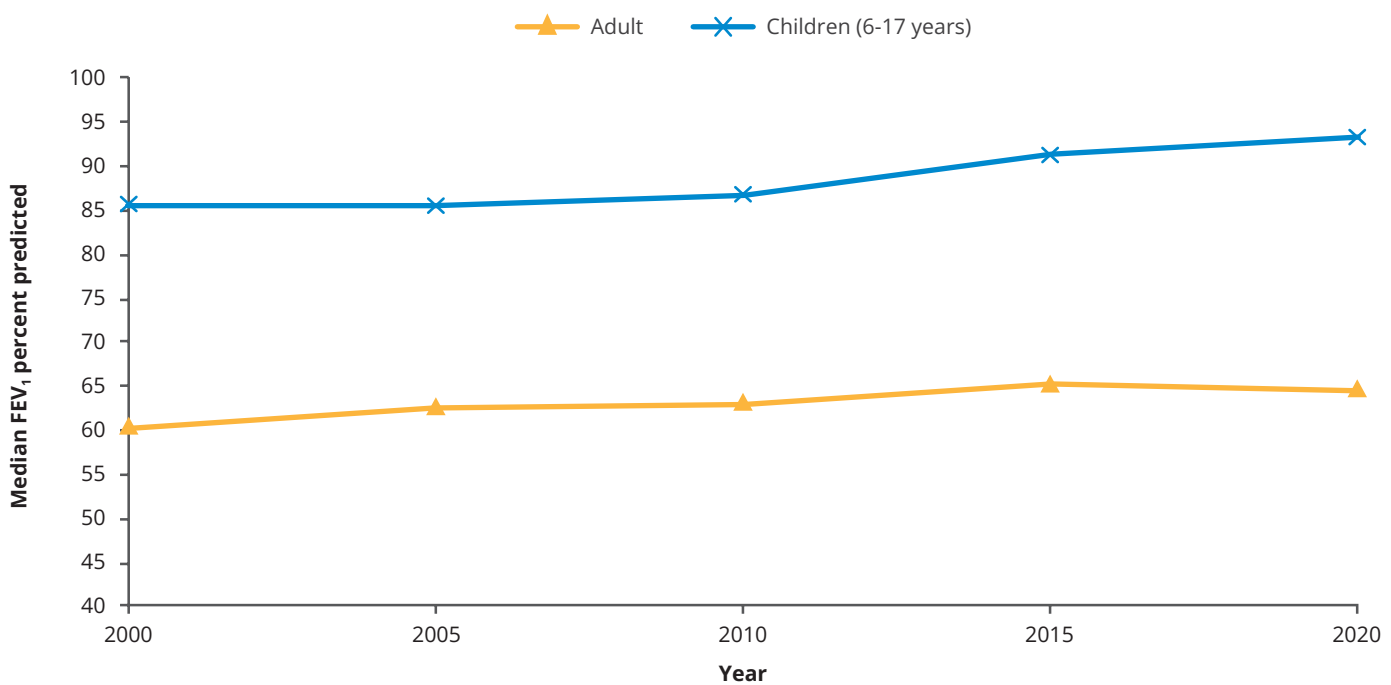
The majority (58.9%) of children, aged 6 to 17 years in 2020, have lung function $\geq 90\%$ predicted, while only 19.9% of adults have lung function in this range, as shown in Figure 19.

FIGURE 19
Respiratory status of children and adults with cystic fibrosis, 2020.



Over time, the median FEV₁ percent predicted has been steadily increasing for both age groups, and in 2020 these values were 64.7% for adults and 93.4% for children (6-17 years of age), as shown in Figure 20.

FIGURE 20
Median FEV₁ percent predicted values for children and adults with cystic fibrosis, 1995 to 2020.



RESPIRATORY STATUS BY SEX

Figure 21 and Figure 22 show that between males and females for both children and adults, the distribution of lung function severity is fairly similar for people with FEV₁ percent predicted above 70%. The vast majority of children have FEV₁ percent predicted at or above 90%, function whereas the majority of adults have FEV₁ percent predicted less than 70%.

FIGURE 21
Respiratory status of children (6 to 17 years) with cystic fibrosis, by sex, 2020.

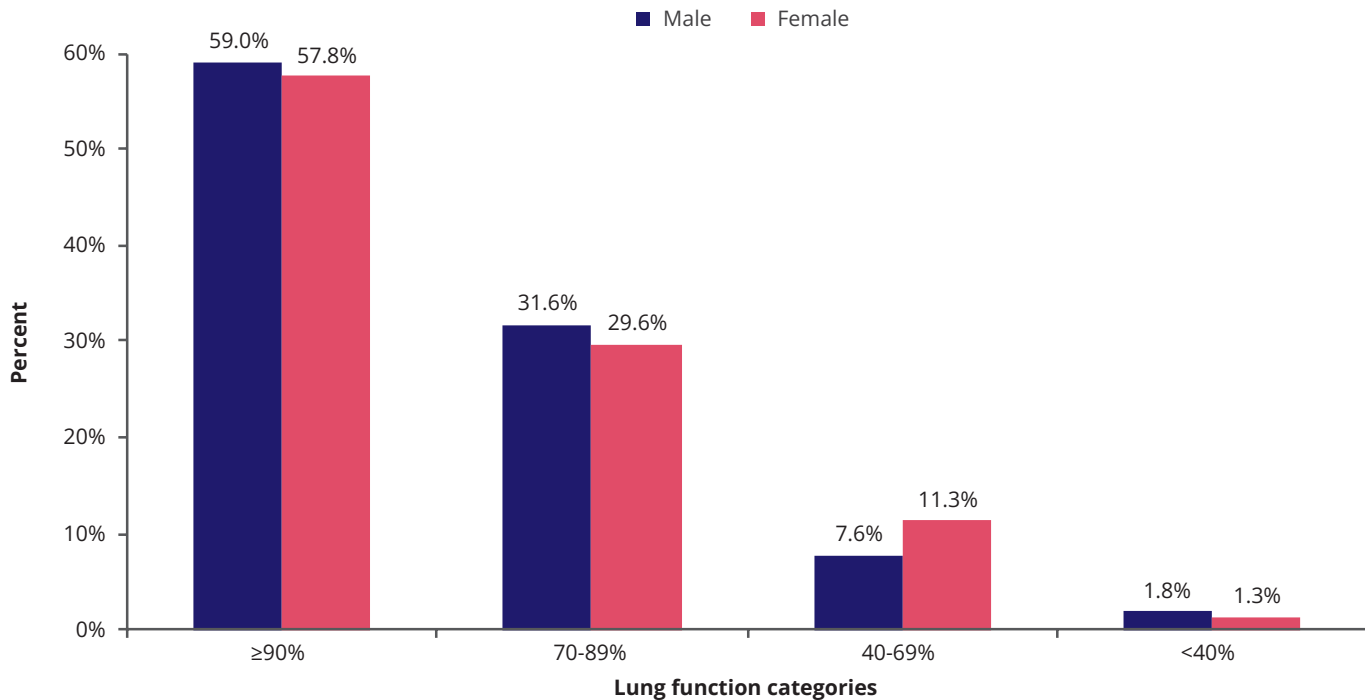
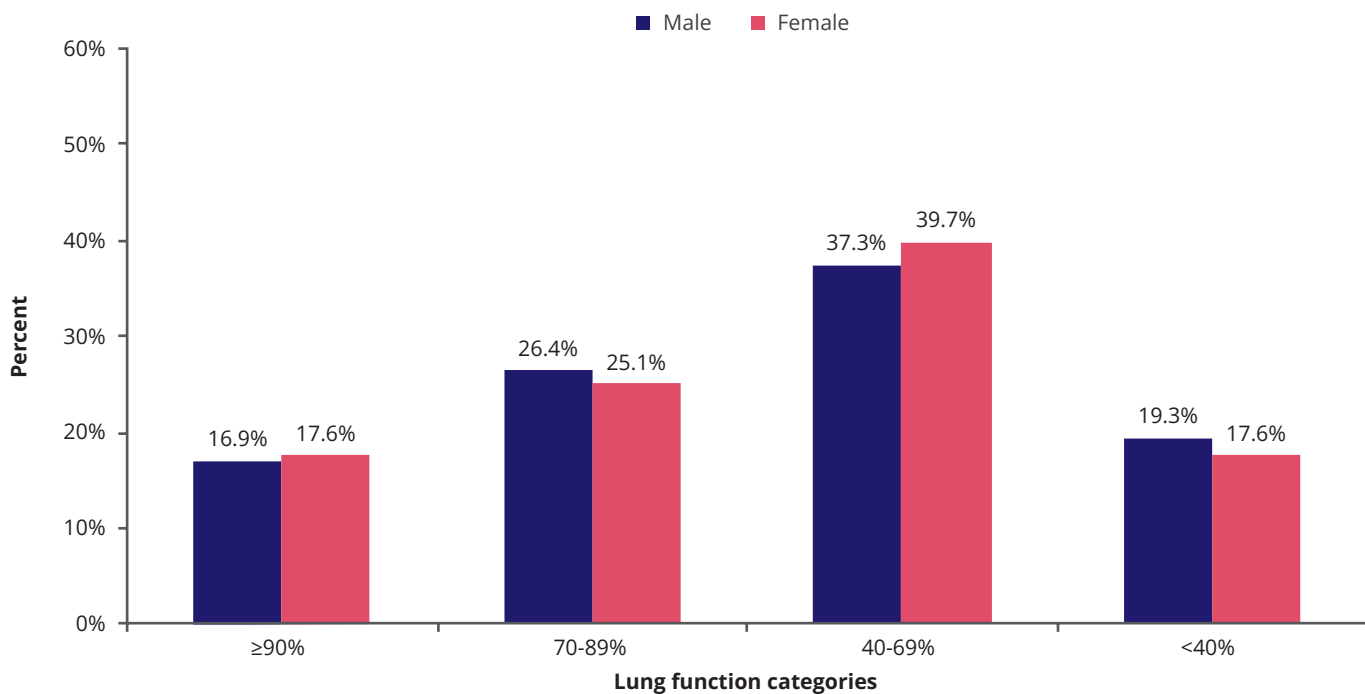


FIGURE 22
Respiratory status of adults (18 years of age and older) with cystic fibrosis, by sex, 2020.



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 2020, the majority (84.3%) of individuals with cystic fibrosis were taking supplemental pancreatic enzymes (and identified as pancreatic insufficient) compared to 15.7% who were not (identified as pancreatic sufficient), as shown in Figure 23. For individuals 40 years of age or older, 28.5% were pancreatic sufficient (Figure 24).

FIGURE 23
Pancreatic status of individuals with cystic fibrosis, 2020.

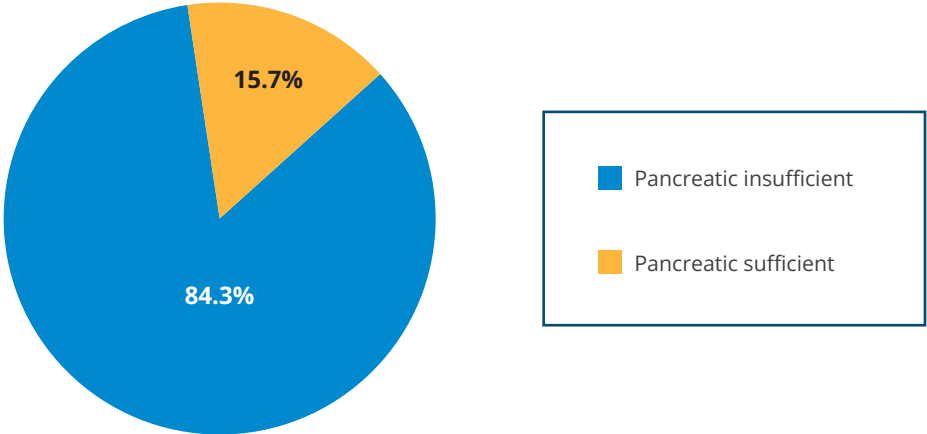
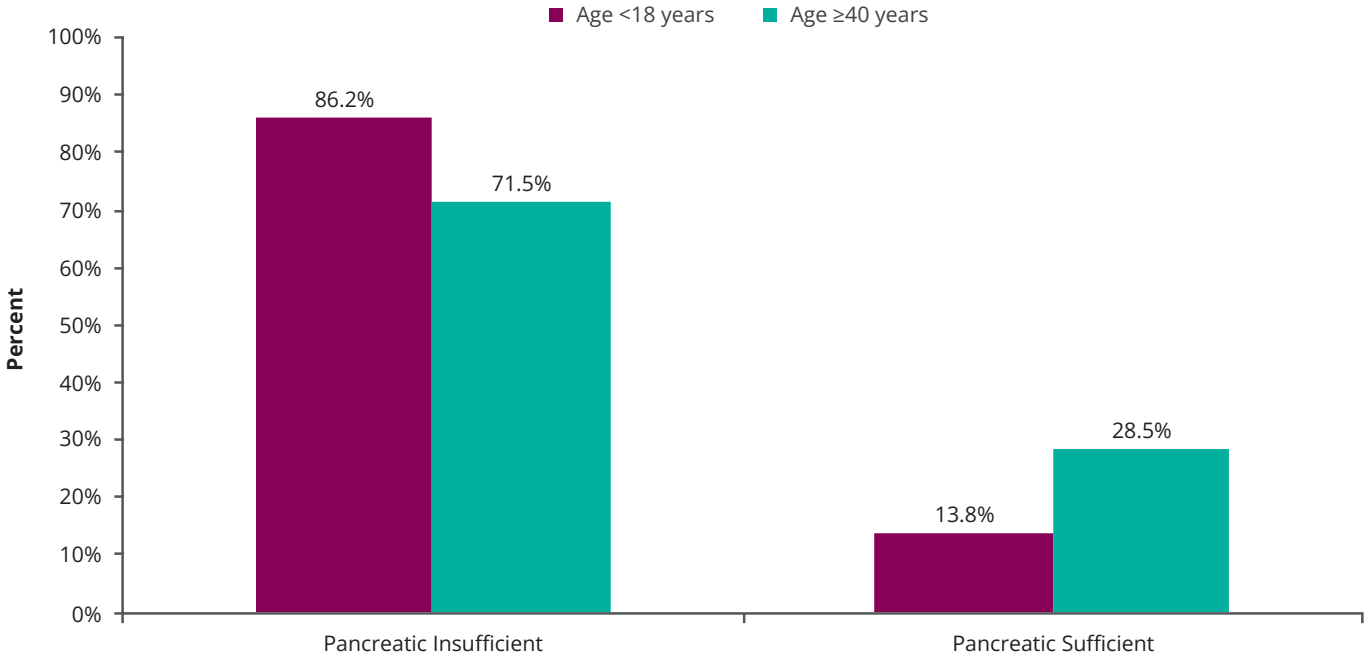


FIGURE 24
Pancreatic status of individuals with cystic fibrosis, by age group, 2020.



BODY MASS INDEX (BMI) PERCENTILE

Body mass index (BMI) is a measure of a person's nutritional status and is based on their weight (in kilograms) and height (in metres). Typically, BMI is only reported for adults because they have attained their maximal height. As children are rapidly growing, one must consider the child's age when assessing their nutritional status, thus using BMI percentiles are a more appropriate measure.

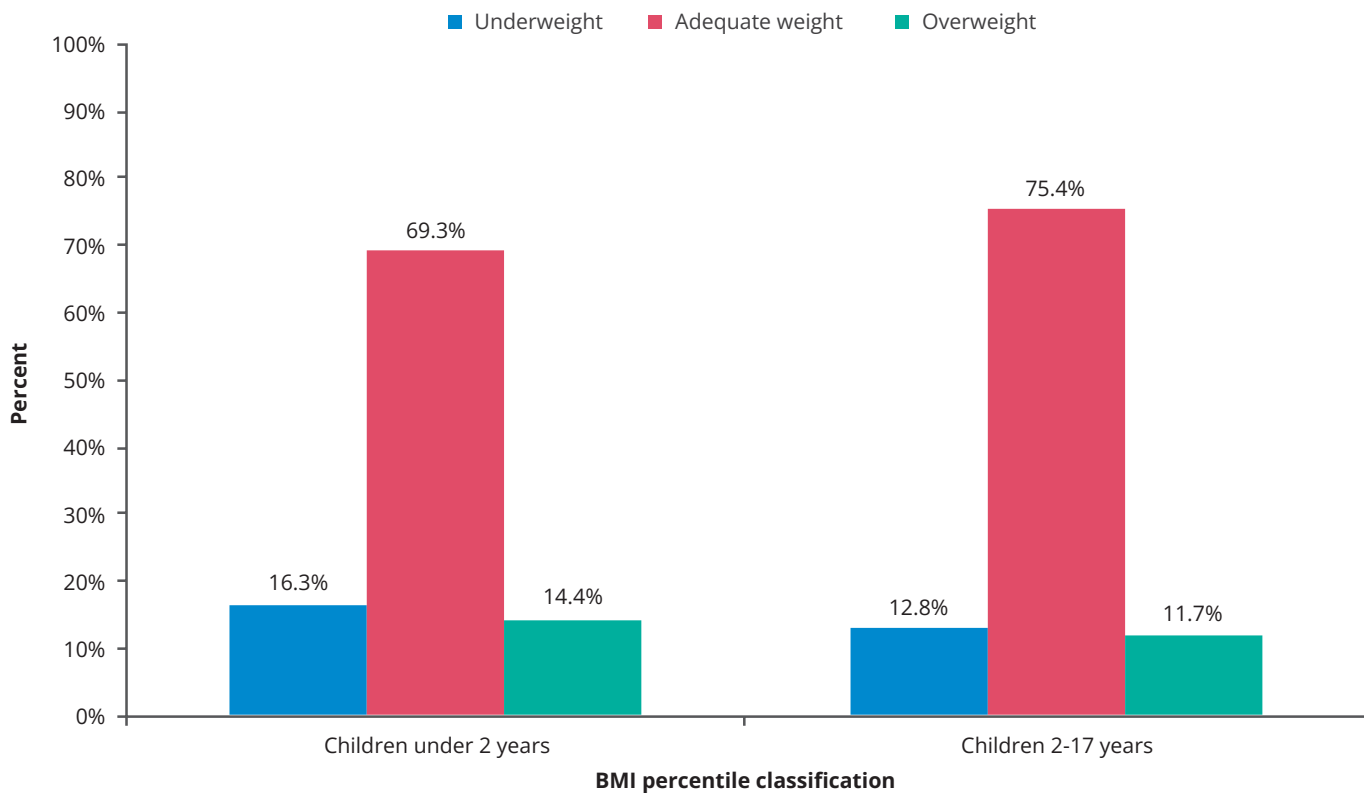
BMI percentiles are calculated following the World Health Organization (WHO) guidelines for children under 2 years of age, and the Centers for Disease Control and Prevention (CDC) guidelines for children ages 2 to 17 years³. BMI percentiles allow comparisons to be made between the individual's height and weight and other children who are the same age and sex. Table 3 details the BMI percentile classification categories following the respective WHO or CDC guidelines⁴.

TABLE 3
BMI percentile classification.

CLASSIFICATION	RANGE
Underweight	≤ 12 th percentile
Adequate weight	13 th percentile - 84 th percentile
Overweight	≥ 85 th percentile

The national median BMI percentile for children under 2 and children between 2 and 17 years of age are 42.1 and 45.1 respectively. Most children with cystic fibrosis (69.3% of children under 2 years and 75.4% of children 2-17 years) have an adequate weight (Figure 25). The 50th BMI percentile is the national goal for children with cystic fibrosis and in 2020, 50.6% of children under 2 years and 47.8% of children 2-17 years exceeded this goal.

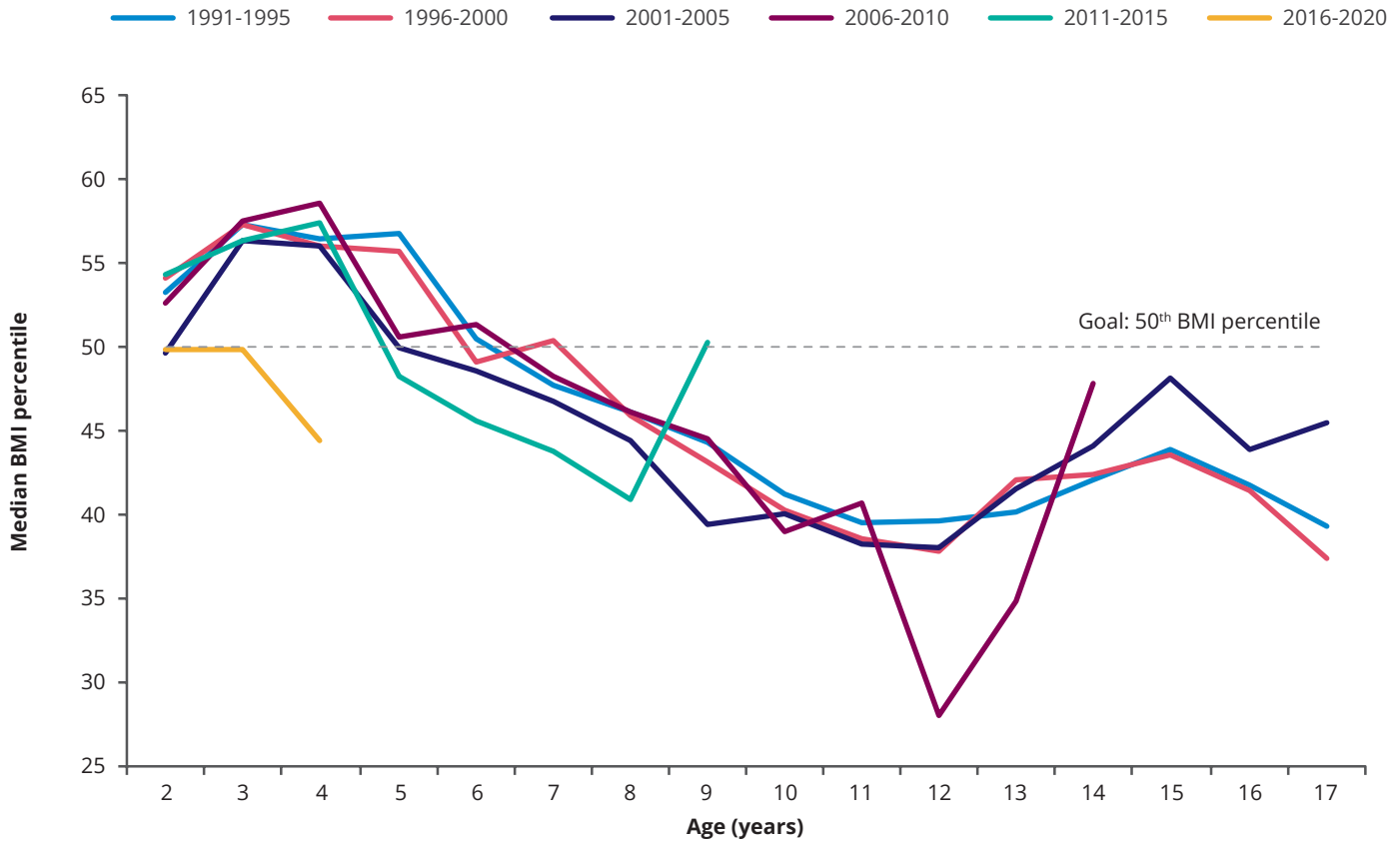
FIGURE 25
BMI percentile status for children with cystic fibrosis, 2020.



BODY MASS INDEX (BMI) PERCENTILE

Figure 26 below shows the median BMI percentile for children between 2 and 17 years of age by birth cohort. In more recent birth cohorts, the median BMI percentile at age 2 years increases for the most part. The nutritional status is relatively stable in the early ages (2 to 4 years) followed by a gradual decline in BMI percentiles over the ages until approximately age 10 years. Median BMI percentile seems to stabilize after 10 years of age. The deviations in trends present in some of the birth cohort lines for the later ages are due to the small sample sizes resulting from the method used to group ages in the calculations.

FIGURE 26
Median BMI percentile for children (2-17 years) with cystic fibrosis, by birth cohort, 2020.



BMI PERCENTILE BY SEX

Figure 27 and Figure 28 show the BMI percentile status for males and females in children under 2 years (N = 202) and children 2-17 years (N = 1,466).

FIGURE 27
BMI percentile status for children (under 2 years) with cystic fibrosis, by sex, 2020.

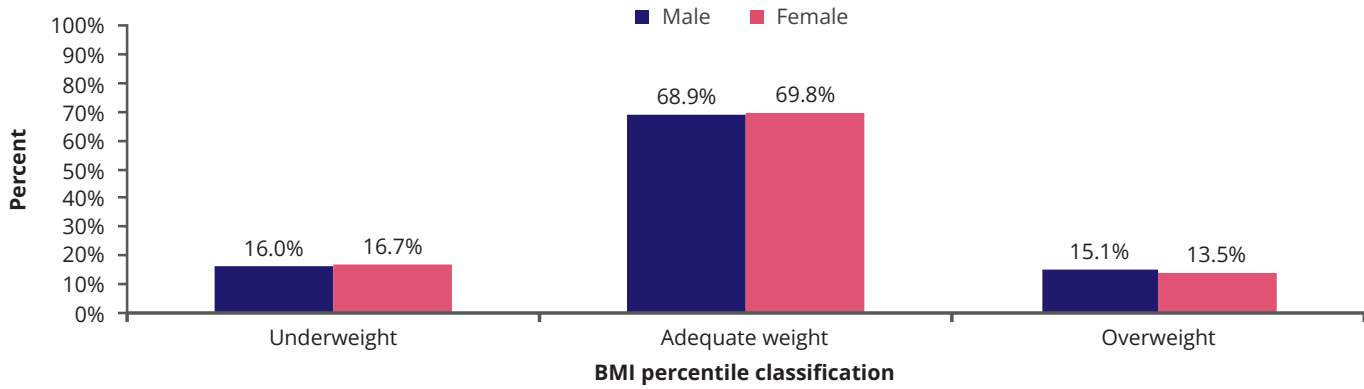
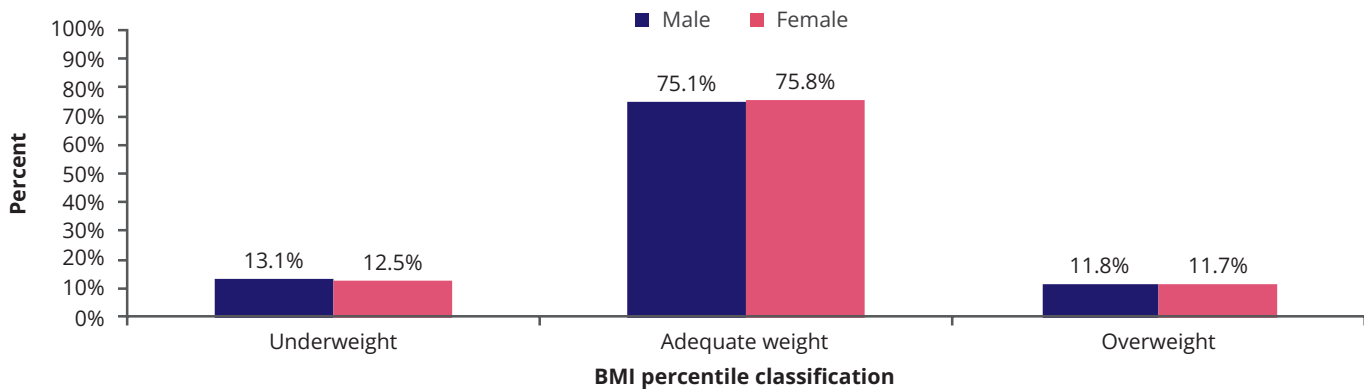
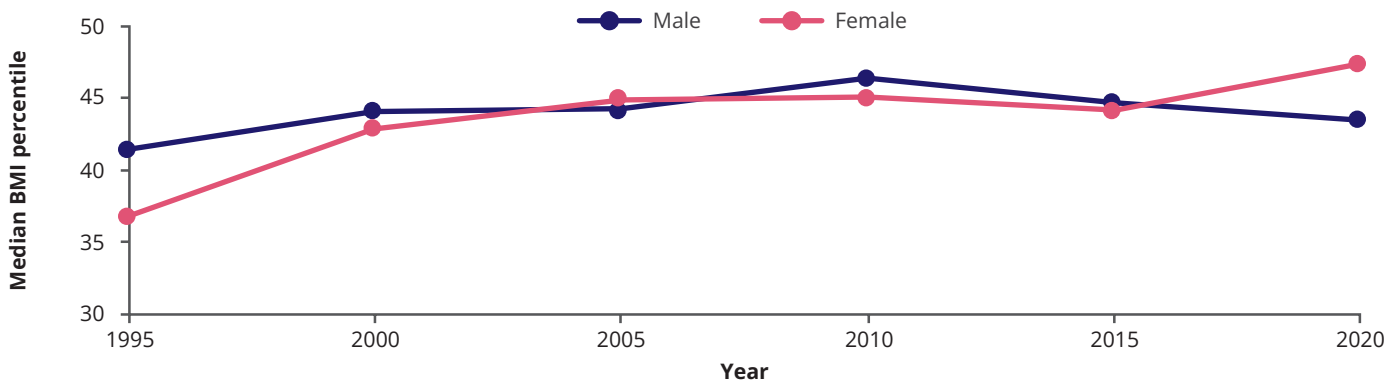


FIGURE 28
BMI percentile status for children (2-17 years) with cystic fibrosis, by sex, 2020.



For both males and females, the median BMI percentiles have been increasing over time. While males show a slightly higher median BMI percentile in earlier years, the gap between sexes diminishes over time. In 2020, females show a higher median BMI percentile than males (Figure 29).

FIGURE 29
Median BMI percentiles for children (2-17 years) with cystic fibrosis, by sex, 1995 to 2020.



BMI

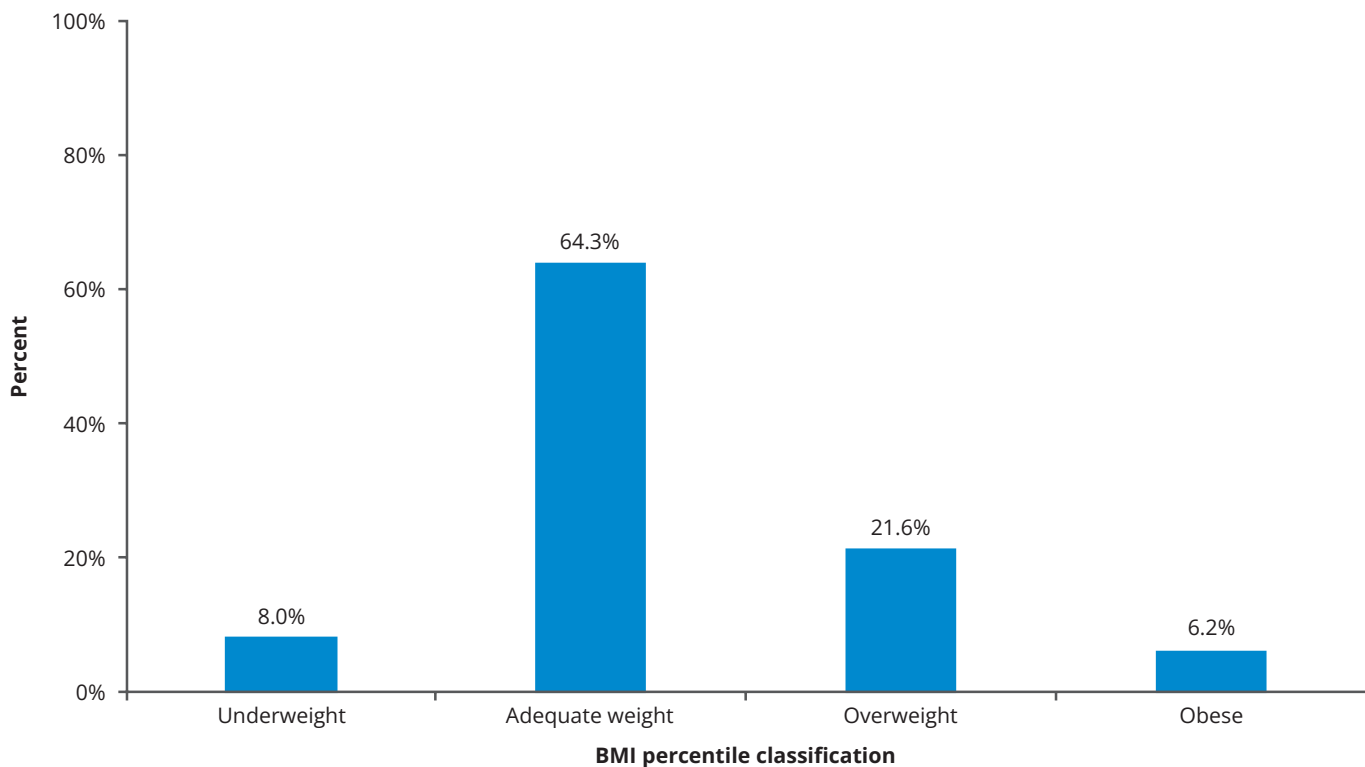
Table 4 below describes the BMI classifications and their BMI ranges according to the WHO guidelines⁵. These guidelines were updated in 2016 and as such, the proportions of BMI classifications will be different from those described in reports prior to 2016. In 2020, the national median BMI for adults (aged 18 and older) was 22.6 kg/m².

TABLE 4
BMI classification.

CLASSIFICATION	RANGE
Underweight	< 18.5 kg/m ²
Adequate weight	18.5 - 24.9 kg/m ²
Overweight	25 - 29.9 kg/m ²
Obese	≥ 30 kg/m ²

The majority (64.3%) of the adult cystic fibrosis population had an adequate weight, while 8% were considered underweight and 27.7% were considered overweight or obese (Figure 30).

FIGURE 30
BMI status for adults with cystic fibrosis, 2020.



BMI BY SEX

Figure 31 shows the breakdown of BMI categories for adult males and females. Individuals who are muscular may have a higher BMI due to increased weight from larger amounts of muscle mass.

In 2020, while more females (10.4%) were considered underweight compared to males (5.9%), the median BMI over the past 25 years has been steadily rising within the cystic fibrosis adult population for both sexes (Figure 32) and can be attributed to fewer individuals who are underweight and more adults classified as either overweight or obese (Figure 33 and Figure 34).

FIGURE 31
BMI status for adults with cystic fibrosis, by sex, 2020.

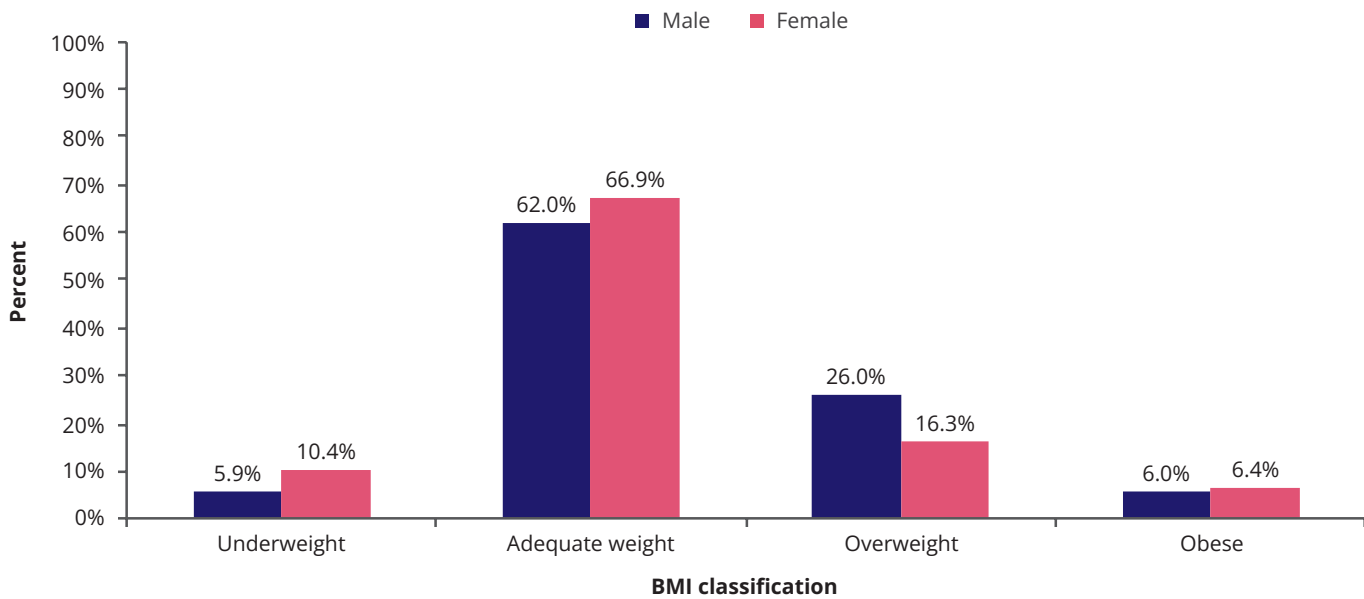
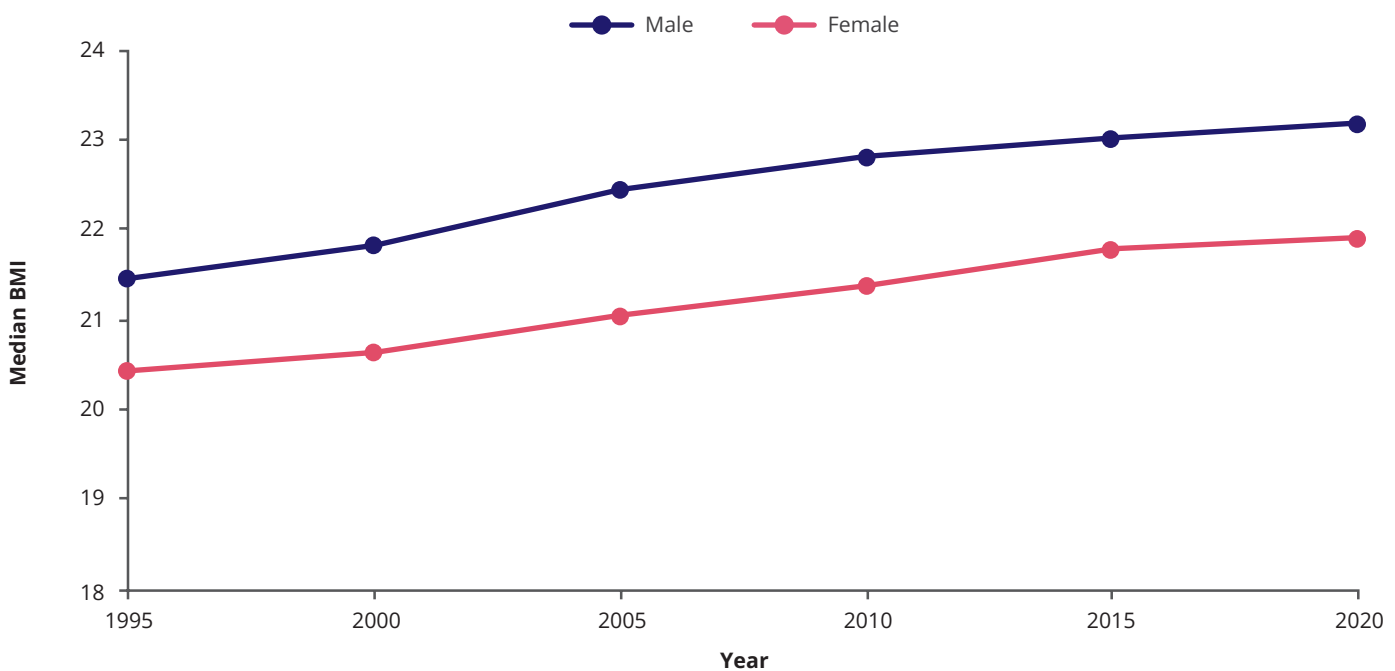


FIGURE 32
Median BMI values for adults with cystic fibrosis, by sex, 1995 to 2020.



BMI BY SEX

FIGURE 33
Percentage of male adults with cystic fibrosis, by BMI status, 1995 to 2020.

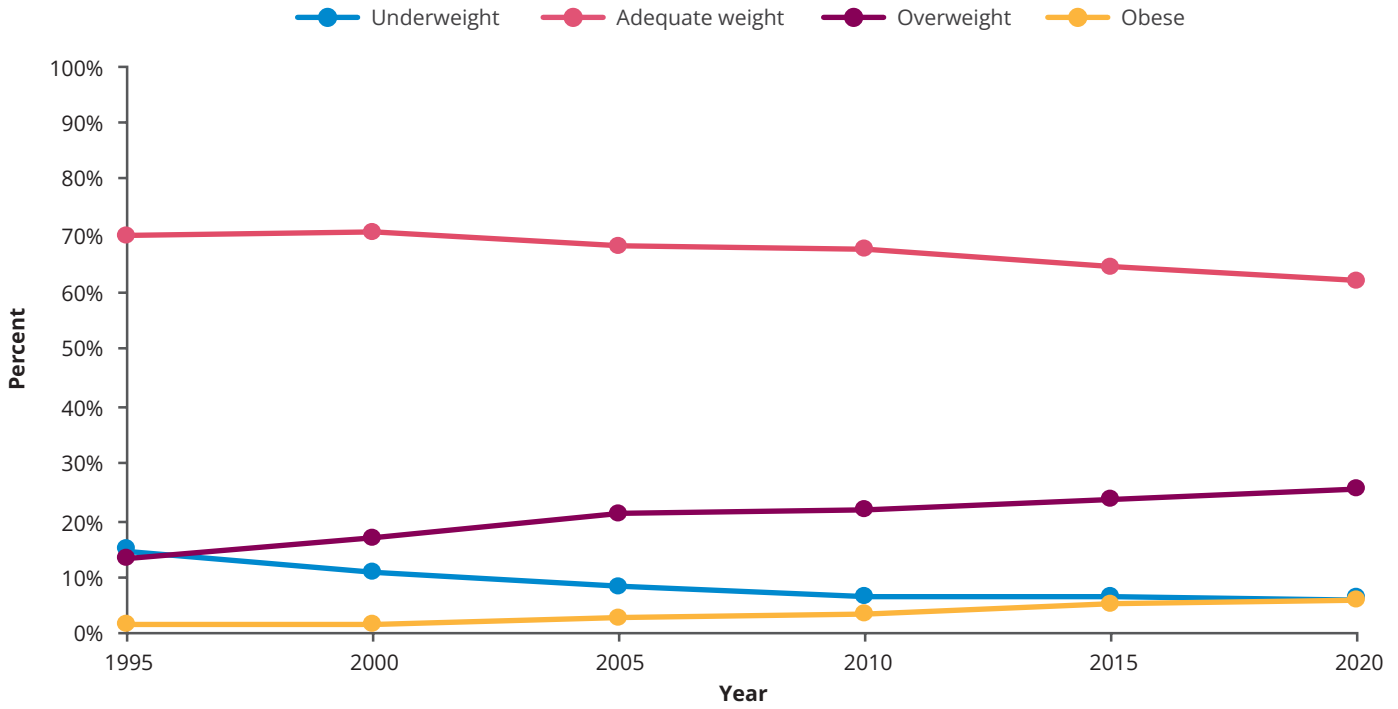
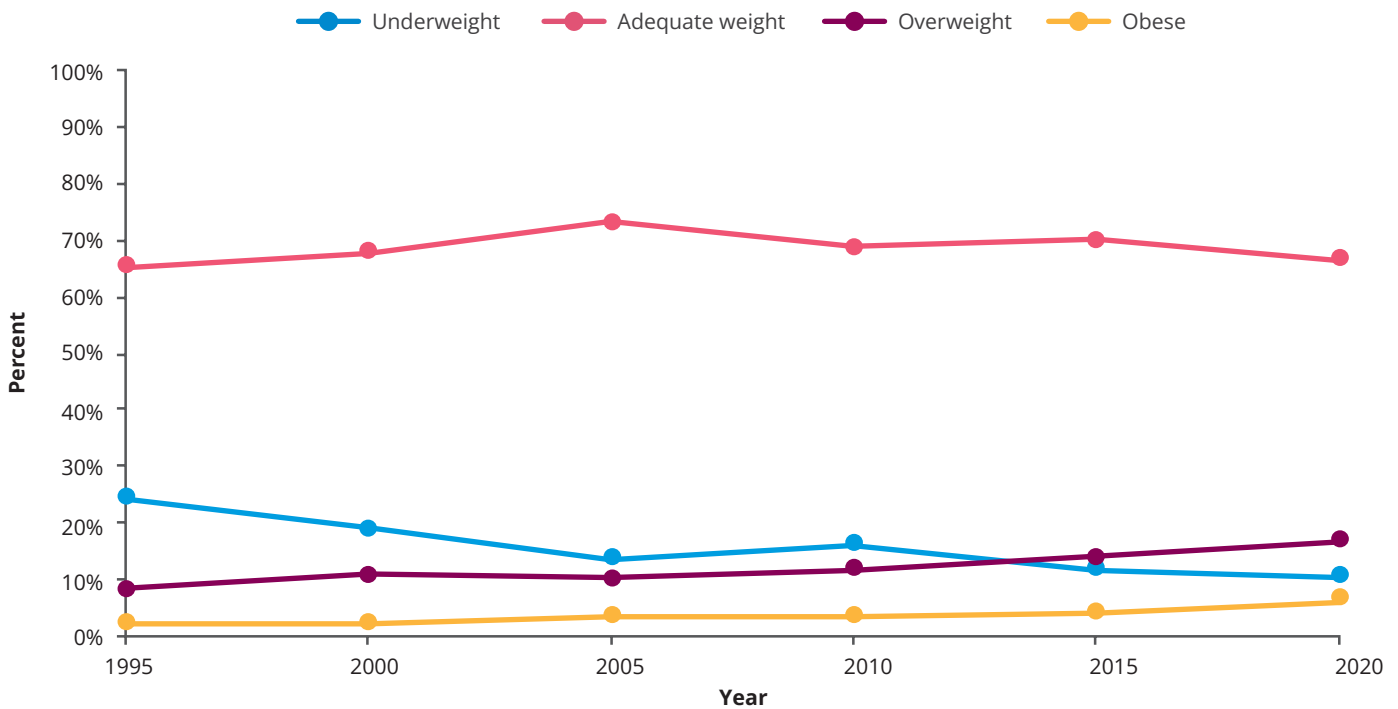


FIGURE 34
Percentage of female adults with cystic fibrosis, by BMI status, 1995 to 2020.



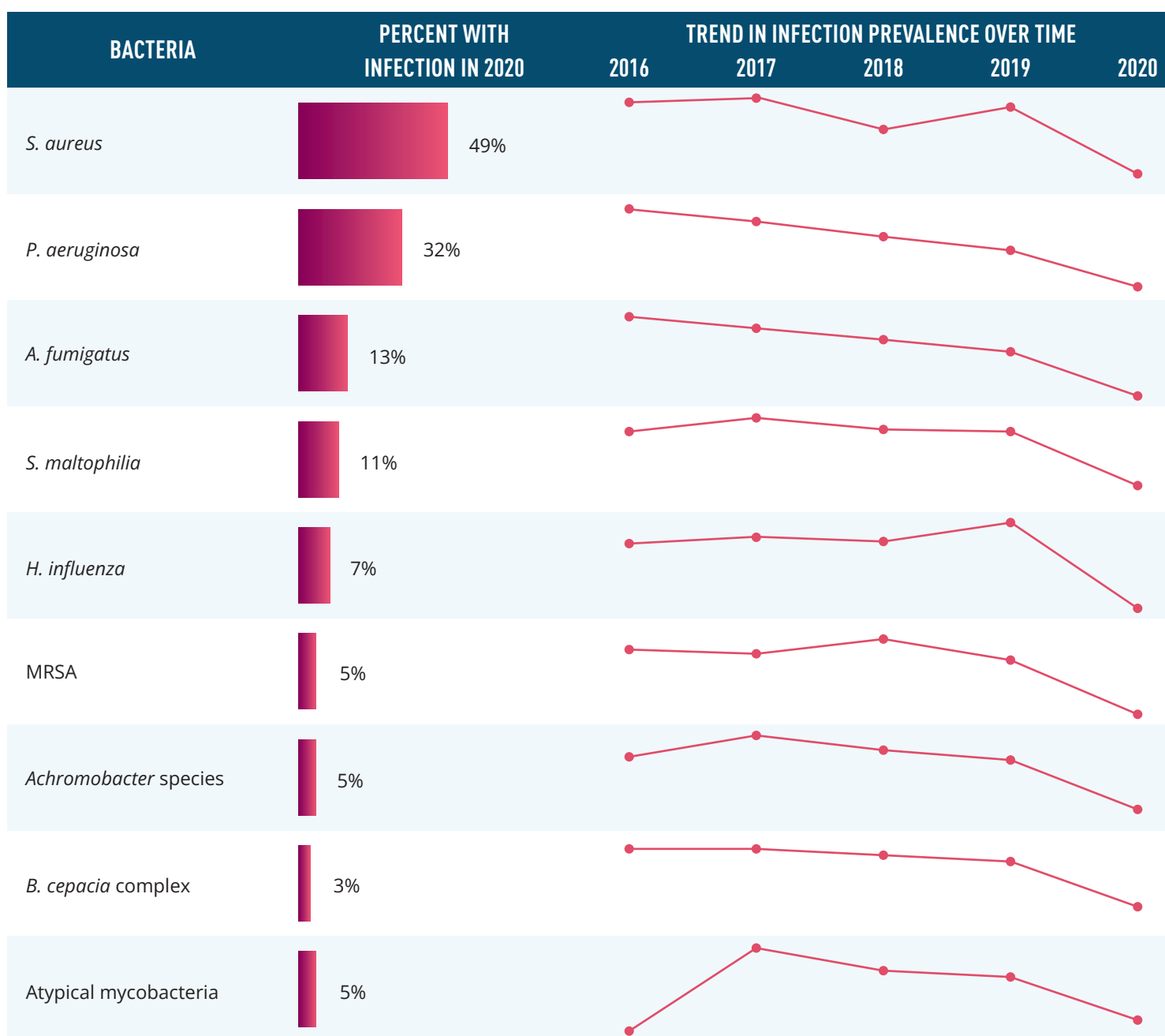
MICROBIOLOGY

BACTERIAL SPECIES AND RESPIRATORY INFECTIONS

Chronic and recurrent infection of the airways is one of the most severe consequences of cystic fibrosis. The most common pulmonary pathogens are *Staphylococcus aureus* (*S. aureus*) and *Pseudomonas aeruginosa* (*P. aeruginosa*), and found to be prevalent in 48.9% and 32.2% of all Canadians with cystic fibrosis, respectively (Table 5). Over the past 5 years, the infection prevalence of the three most common bacteria (*S. aureus*, *P. aeruginosa*, and *A. fumigatus*) has steadily decreased. However, we note that the prevalence of bacteria identified in cultures was lower in 2020 compared to 2019, likely due to the effects of the on-going pandemic.

The CF Registry aims to track relevant bacterial species for the CF population and several have been added in recent years including MRSA (2003), *Achromobacter* species (formally called *Alcaligenes* species) (2011), and atypical mycobacteria (2011).

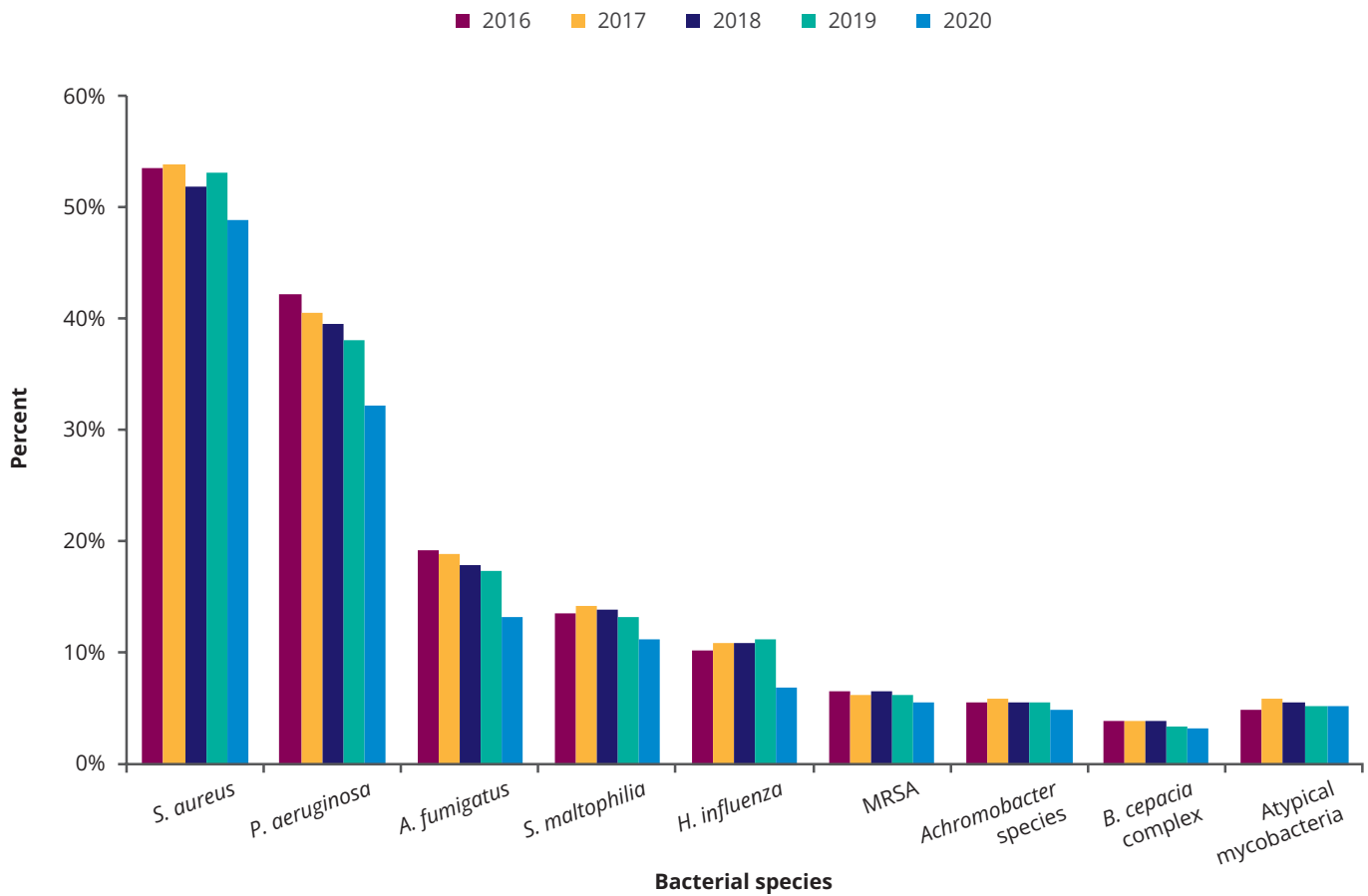
TABLE 5
Prevalence of bacterial species cultured from airways of individuals with cystic fibrosis (all ages), 2020.



BACTERIAL SPECIES AND RESPIRATORY INFECTIONS

While decreasing, Figure 35 shows that over the past several years, *S. aureus* and *P. aeruginosa* remain the two most prevalent pulmonary pathogens among individuals with cystic fibrosis. There has also been a slight increase in the less frequently found pathogens such as *Achromobacter* species (formerly *Alcaligenes* species) and atypical mycobacteria. This may be due, in part, to an increase in reporting of these organisms rather than a true increase in prevalence.

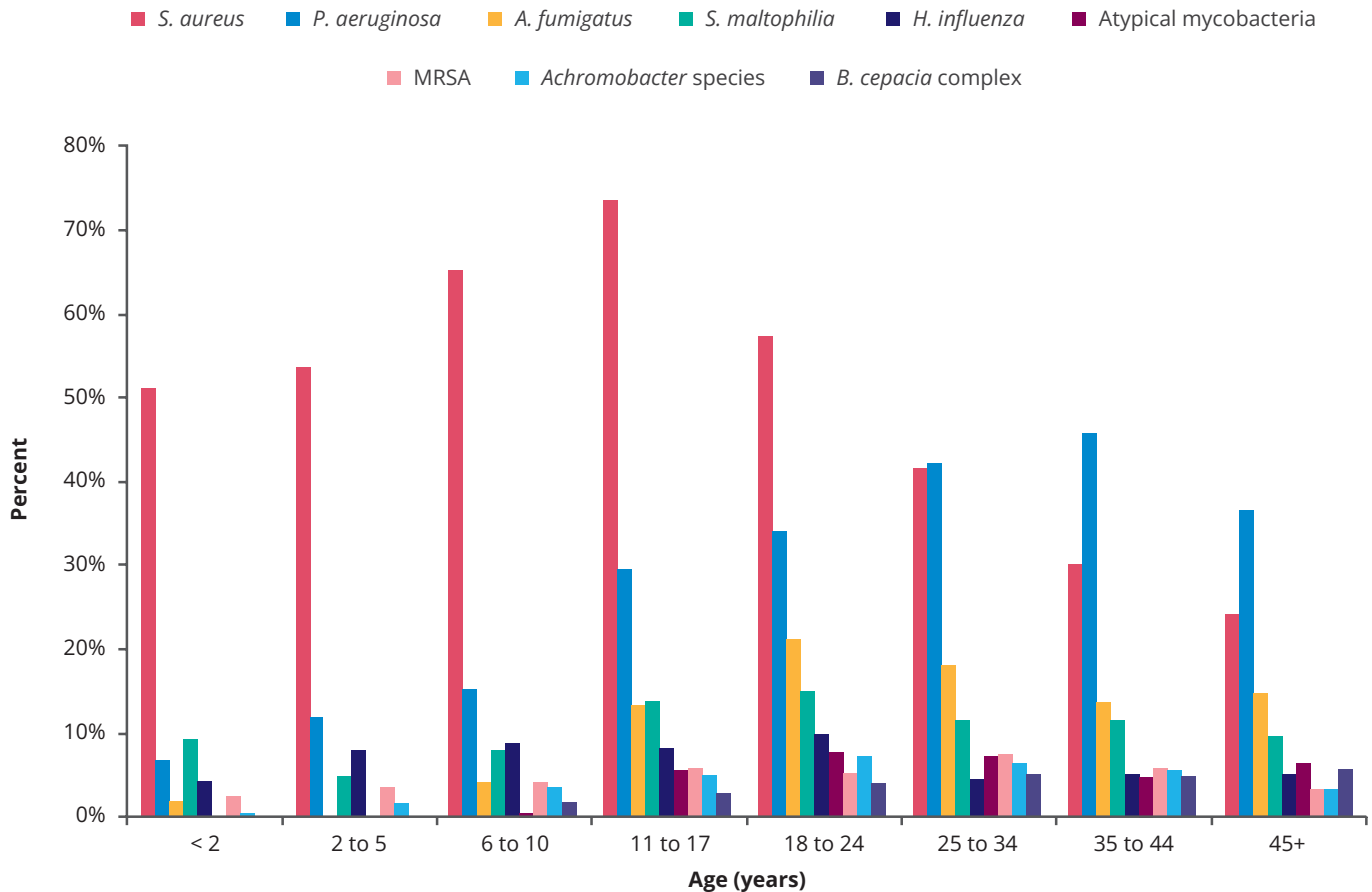
FIGURE 35
Prevalence of respiratory infections of individuals with cystic fibrosis, 2016 to 2020.



BACTERIAL SPECIES AND RESPIRATORY INFECTIONS

When examining the prevalence of pathogens by age (Figure 36), it appears that *S. aureus* is more common in children with cystic fibrosis and *P. aeruginosa* is more common in the adult CF population. *Burkholderia cepacia* complex (*B. cepacia* complex or BCC) is more commonly seen in older individuals with cystic fibrosis, but the prevalence is low for the entire CF population (4.3%). Furthermore, new acquisition of BCC is infrequent and typically, the *Burkholderia* species that is reported is an environmental strain rather than the epidemic *cenoecepacia* strain (for more details see Figure 37 and Figure 38).

FIGURE 36
Age-specific prevalence of respiratory infections in individuals with cystic fibrosis, 2020.



BURKHOLDERIA CEPACIA COMPLEX (BCC)

Out of all individuals with cystic fibrosis with bacterial species recorded in 2020, 140 (3.2%) unique individuals who grew at least one *Burkholderia cepacia* complex (BCC) species. The two most common types of BCC species are *B. cenocepacia* (50.0%) and *B. multivorans* (37.9%) (Figure 37). Of the unique individuals who had BCC in 2020, 122 (87.1%) were adults and 42 (30%) were over the age of 40 (Figure 38). Not all BCC bacteria have been speciated, as 13.9% of the BCC species in the CF Registry were classified as unknown. Although BCC has been reported on for decades, the ability to specify the type of BCC species was added to the CF Registry in 2011.

Note: The prevalence of *B. gladioli* was 7.5%, though it was not included in Figure 37, because it is not officially recognized as a BCC species.

FIGURE 37
Burkholderia cepacia complex species prevalence in individuals with cystic fibrosis (N = 140), 2020.

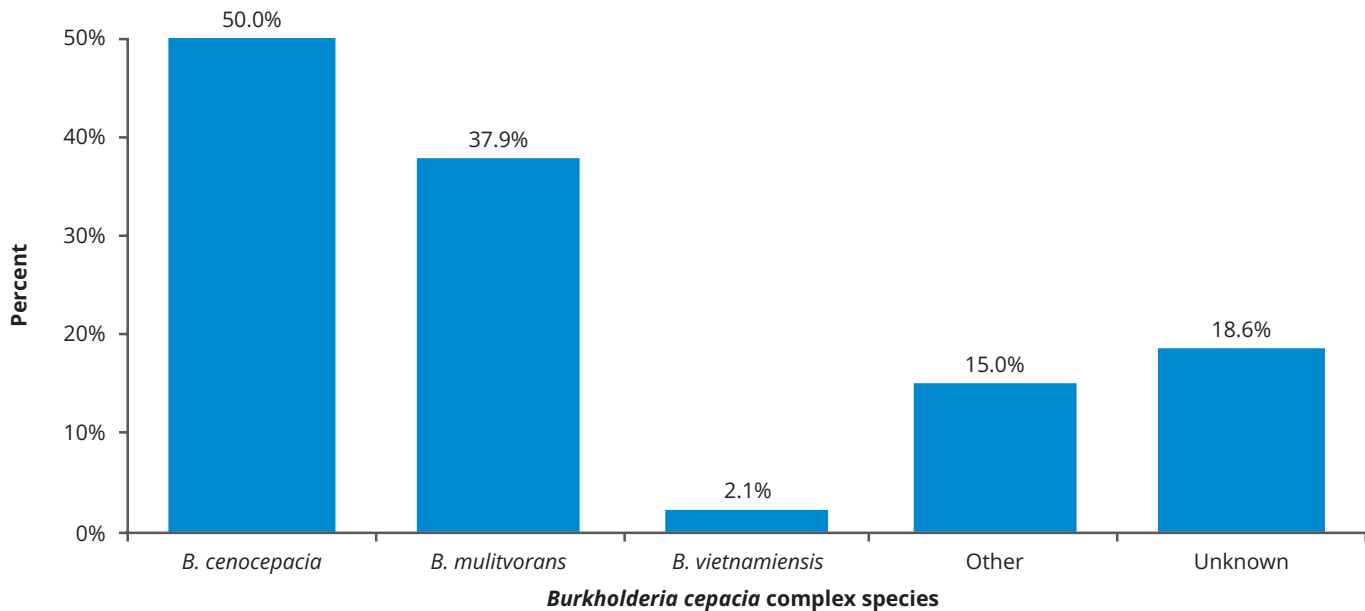
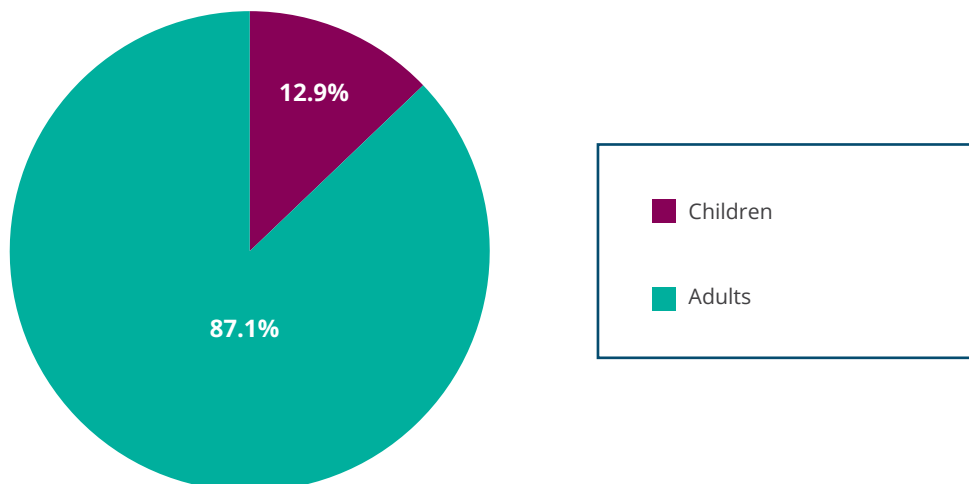


FIGURE 38
Burkholderia cepacia complex distribution of cystic fibrosis individuals, by age, 2020.

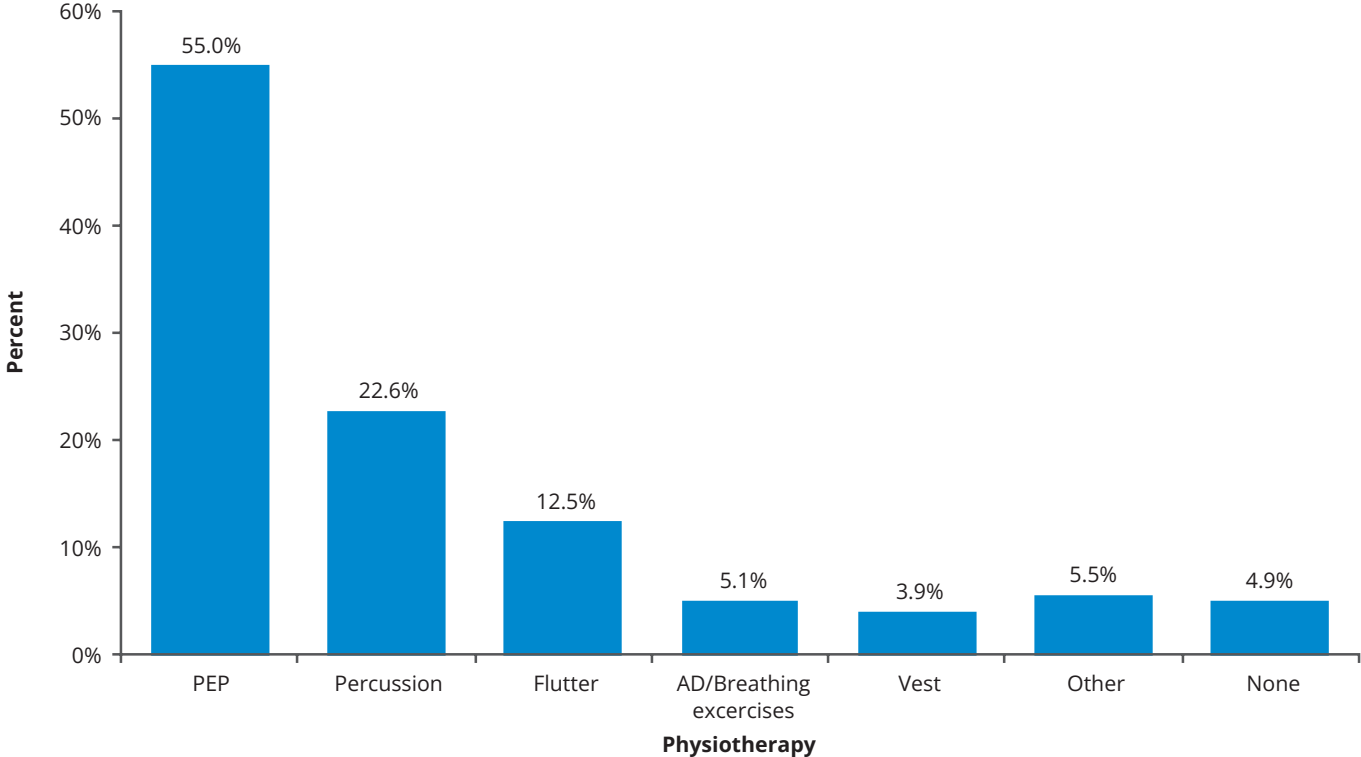


PHYSIOTHERAPY

Physiotherapy is done to help clear mucus from airways using a variety of methods. In 2020, information on physiotherapy was entered for 89.8% of people living with CF (including indication of no physiotherapy regimen). Figure 39 shows the distribution of physiotherapy recorded in the CF Registry. The most commonly used form of therapy are positive expiratory pressure (PEP) (55.0%) and percussion (22.6%), while 4.9% were reported as not doing any form of physiotherapy.

Note: Individuals who have ever received a lung transplant (7.1% of the 2020 reported CF population) were excluded from these calculations because, typically, chest physiotherapy is not part of routine post-transplant treatment.

FIGURE 39
Physiotherapy usage of cystic fibrosis individuals (N = 4,023), 2020.

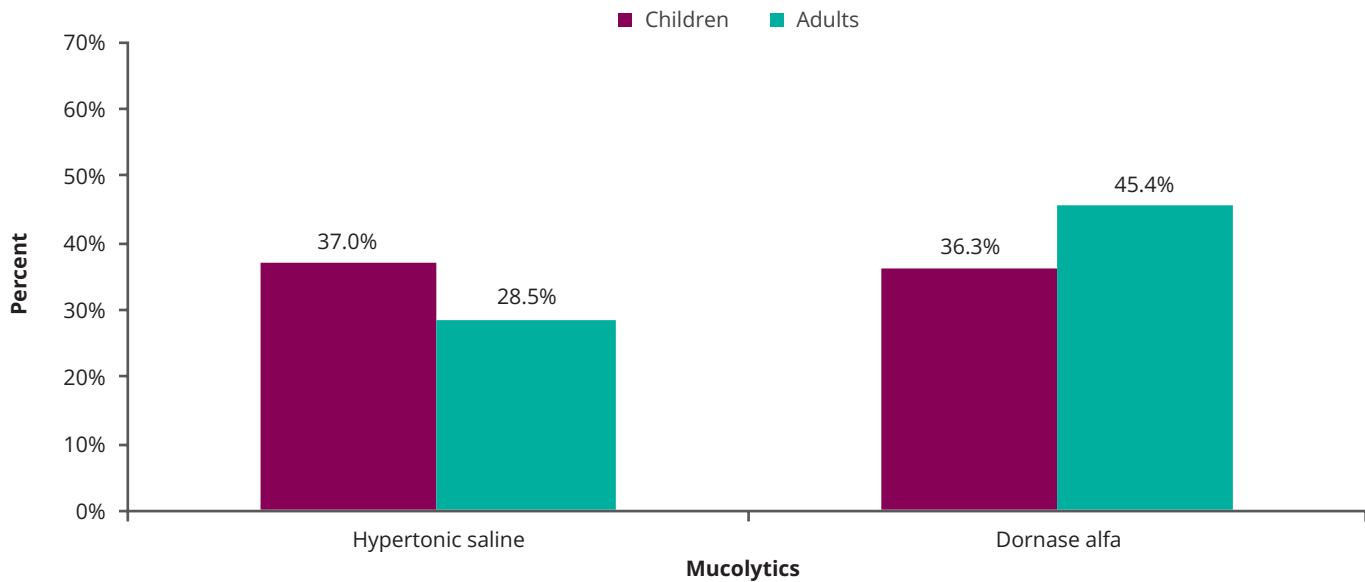


MEDICATIONS

In 2020, there were a total of 4,023 individuals (1,638 children and 2,385 adults) who never received a transplant. Individuals who ever received a transplant (any organ) were excluded from the following figures, because the medications listed are not typically part of routine post-transplant treatment.

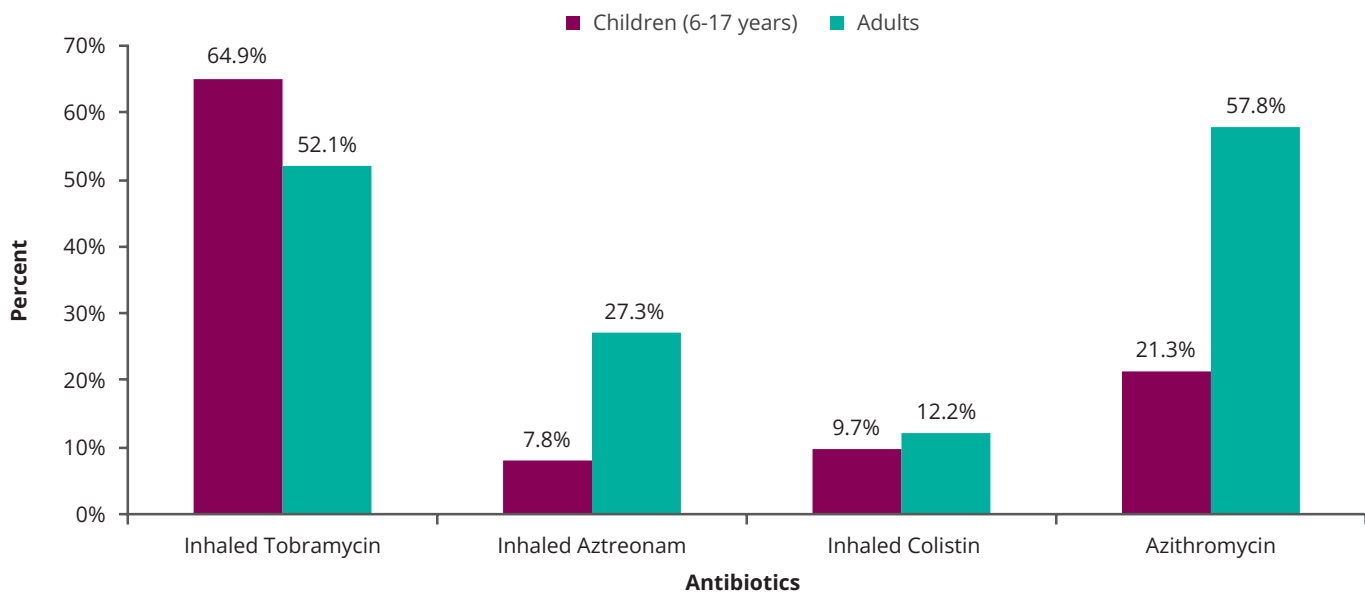
Figure 40 shows that of individuals who never received a transplant, 2,505 (57.8%) were prescribed mucolytic therapy during the calendar year (hypertonic saline and/or dornase alfa).

FIGURE 40
Percentage of cystic fibrosis individuals on mucolytics, by age group, 2020.



There were 1,273 individuals over the age of 6 years who have never received a transplant and were reported to have *Pseudomonas aeruginosa* in 2020, which include 268 children (6-17 years) (21.1%) and 1,005 adults (78.9%). Of those, there were 174 children (64.9%) and 524 adults (52.1%) who were prescribed inhaled tobramycin treatment, and 57 children (21.3%) and 581 adults (57.8%) who were prescribed macrolide therapy (azithromycin) (Figure 41).

FIGURE 41
Percentage of cystic fibrosis individuals on chronic antibiotics, by age group, 2020.



■ CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE (CFTR) MODULATORS

CFTR modulator therapies are designed to improve the production, intracellular processing, and function of the malfunctioning protein made by the CFTR gene. These drugs are an important advance in managing CF, however their efficacy depends on the specific mutations in an individual patient since different mutations result in different defects.

Single agent ivacaftor was approved by Health Canada on November 26, 2012 for patients with the G551D mutation. Ivacaftor approval for an additional 9 mutations was received in June 2014, followed by the approval for the R117H mutation in March 2015. Lumacaftor/ivacaftor was approved in January 2016 and ivacaftor/tezacaftor in January 2018. In June 2021, Health Canada has approved the triple combination therapy elexacaftor/ivacaftor/tezacaftor for sale in Canada.

In 2020, there were 908 unique individuals (274 children and 634 adults) on CFTR modulator therapies (Table 6).

TABLE 6
Number of cystic fibrosis individuals on CFTR modulators by age group, 2020.

CFTR MODULATOR	CHILDREN	ADULTS	TOTAL
ivacaftor	59	102	161
lumacaftor/ivacaftor	145	203	348
ivacaftor/tezacaftor	44	242	286
elexacaftor/ivacaftor/tezacaftor	28	169	197

* For more information about CFTR modulators, please visit <https://www.cysticfibrosis.ca/our-programs/advocacy/access-to-medicines>

HEALTHCARE ENCOUNTERS

A total of 4,289 (99%) individuals with cystic fibrosis visited a CF clinic (had a recorded clinic visit date and/or clinical measurement) at least once in 2020 with 3,355 (78.2%) having three or more clinic visits. These clinic visits can include telemedicine or virtual appointments, during which patients receive medical education, or health advice and information via telecommunication technologies. Of the people having three or more clinic visits, 1,477 were children and 1,878 were adults, making up 90.0% and 69.8% of all children and adults, respectively. In 2020, there were a total of 18,044 clinic visits (Table 7).

The CF Registry captures the date of admission and discharge from hospital. From these data, the total number of hospitalizations per patient are calculated. In 2020, there were 933 (21.5%) individuals with cystic fibrosis who altogether spent 17,111 days in hospital from a total of 1,525 recorded hospitalizations, which do not include visits to the out-patient CF clinics.

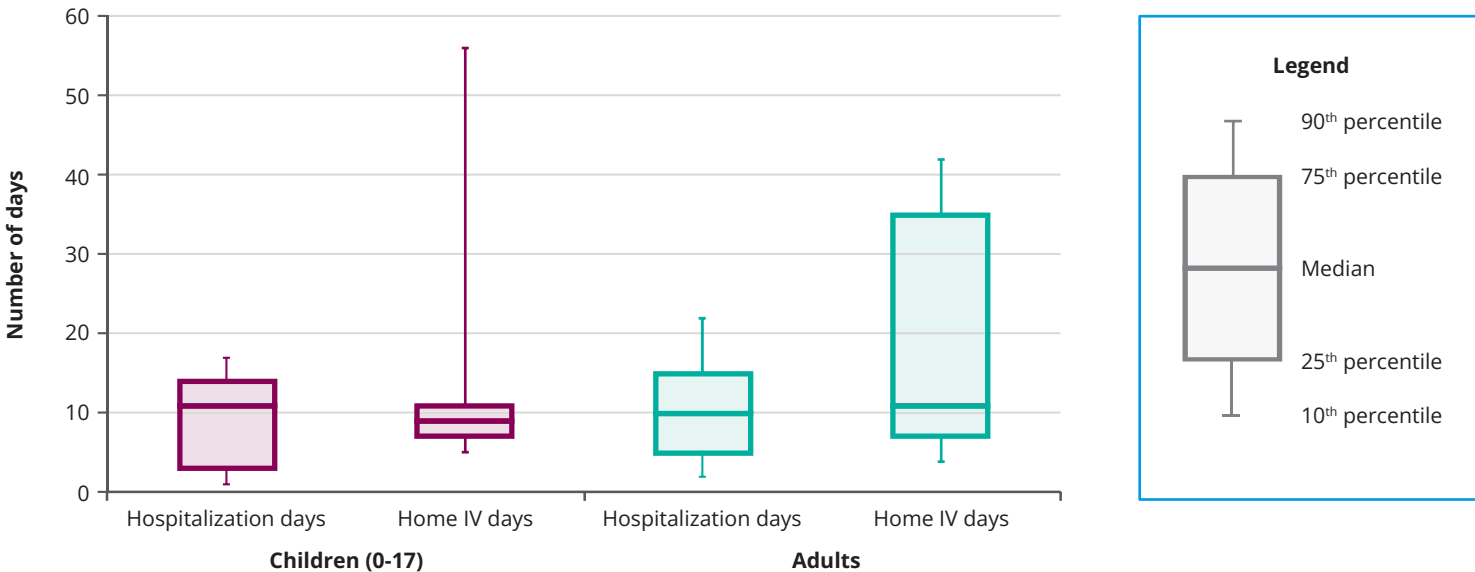
At home, individuals with cystic fibrosis had 13,606 days on IV antibiotics from a total of 749 courses. Note that home IV may be used as part of treatment prior to, or following hospitalization, and as such, may not represent unique episodes of care.

TABLE 7
Total number of healthcare encounters recorded for individuals with cystic fibrosis, 2018 to 2020.

	2018	2019	2020
Clinic Visits	18,916	18,960	18,044
Hospitalizations	2,138	1,952	1,525
Hospital Days	26,573	25,246	17,111
Home IV Courses	941	842	749
Home IV Days	17,730	15,530	13,606

Figure 42 shows the distribution of hospitalizations and home IV courses by age group. In 2020, the distribution of hospital days is relatively similar between age groups, where children were hospitalized for an average of 11 days and adults for 10 days. However, the number of days on home IV varied more among adults compared to children. Though the median number days spent on home IV was similar between children and adults (9 days vs.11 days), a quarter of all adults spent over 35 days on home IV in 2020.

FIGURE 42
Distribution of hospitalizations and home IV courses, by age group, 2020.



CYSTIC FIBROSIS-RELATED DIABETES (CFRD)

Cystic fibrosis-related diabetes (CFRD) is a unique type of diabetes common in individuals living with cystic fibrosis. CFRD is often associated with weightloss and lung function decline, but with early diagnosis and proper treatment, CFRD can be managed successfully. In 2020, CFRD was reported in 958 (22.1%) individuals with cystic fibrosis. While CFRD is not routinely screen in children younger than 10 years of age, 54 (3.3%) children and 904 (33.6%) adults (Figure 43) were recorded as having CFRD in 2020. Of those individuals with CFRD, 49.0% were female, 22.2% have received a transplant, and 53.2% were 35 years of age or older. While there are very few children reported as having CFRD, there is an increasing prevalence of CFRD in the adult population (Figure 44).

FIGURE 43
Percentage of children and adults reported to have CFRD, 2020.

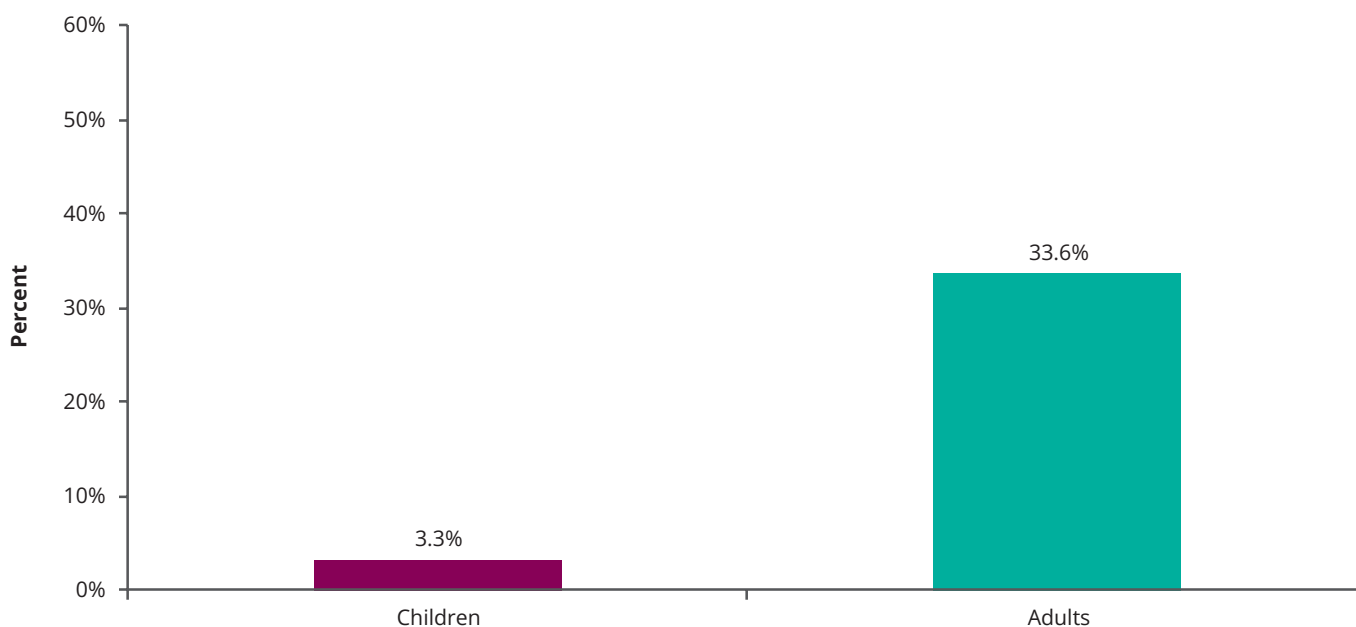
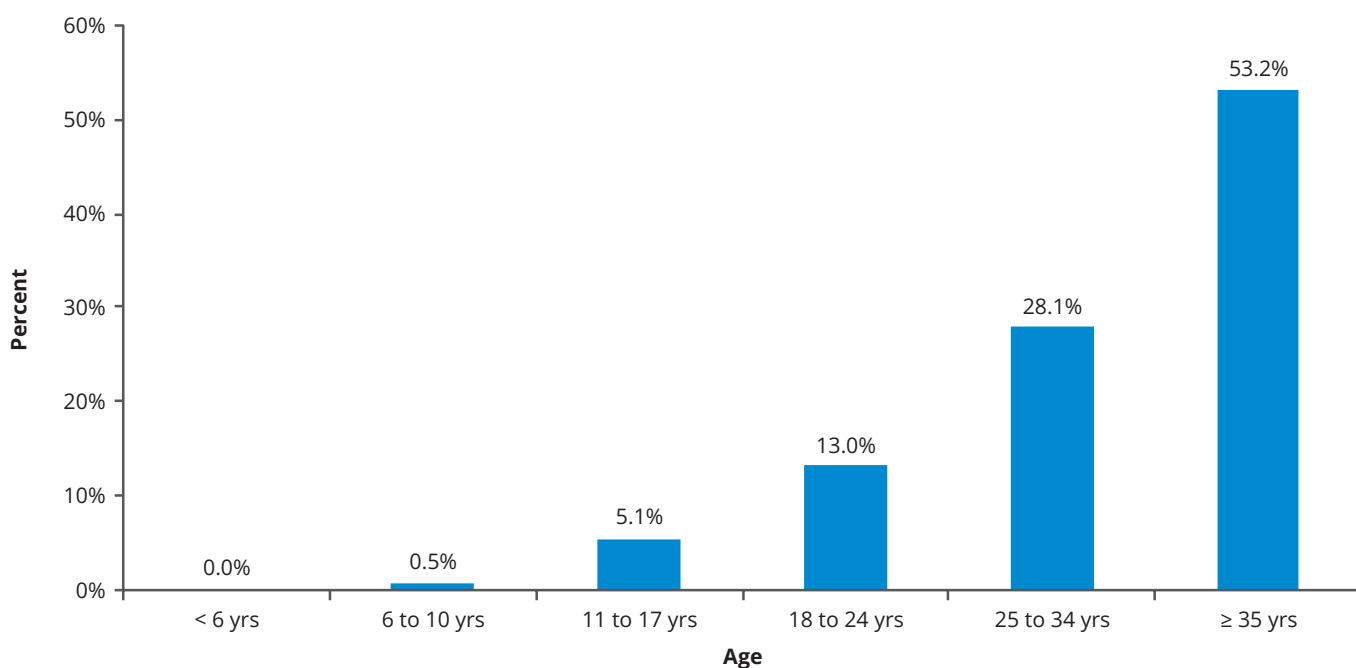


FIGURE 44
Percentage of individuals with CFRD by age, 2020.

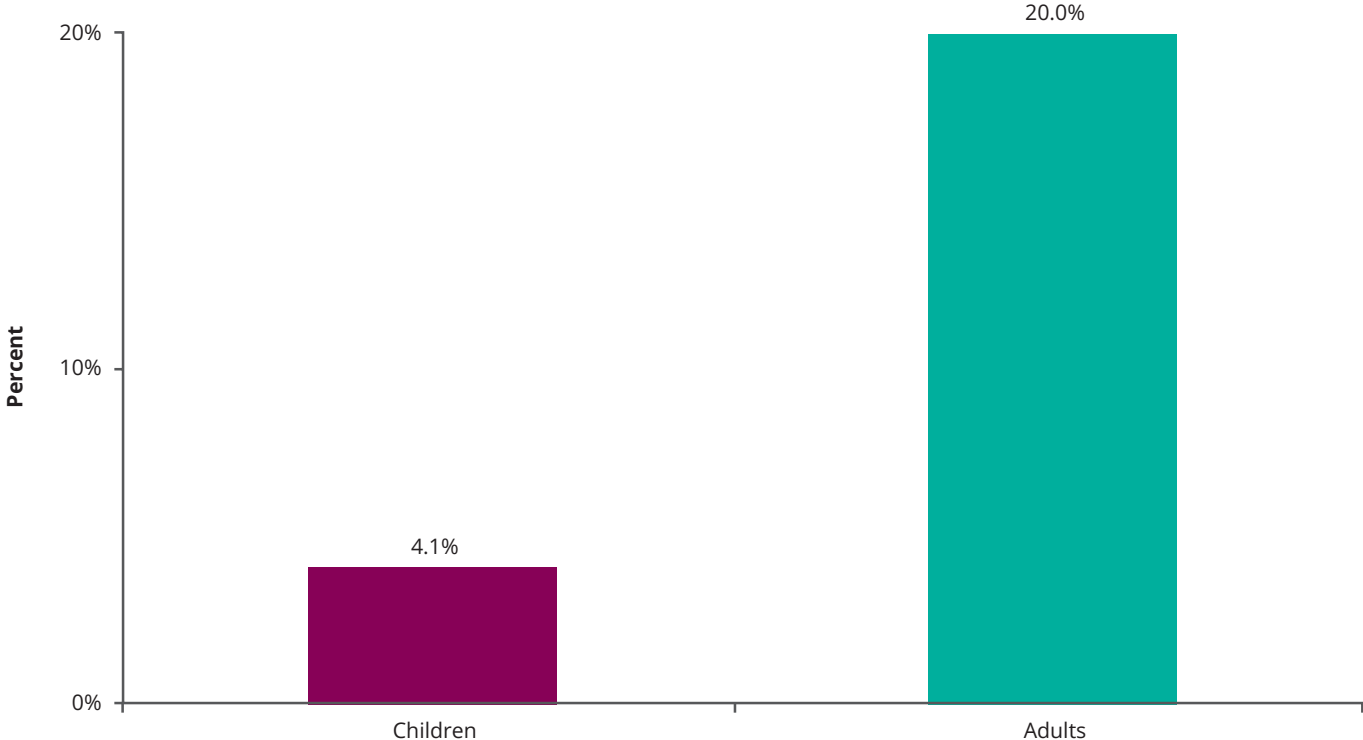


MENTAL HEALTH

In 2020, there were 605 individuals with cystic fibrosis (14% of all individuals) with reported clinically diagnosed depression or anxiety. 67 of these diagnoses were children and 538 were adults, representing 4.1% of all children and 20.0% of all adults living with cystic fibrosis (Figure 45).

These prevalence rates are in line with findings from The International Depression/Anxiety Epidemiology Study (TIDES) which showed elevated rates of depression and anxiety among individuals with cystic fibrosis and their parents/caregivers ^{6 7}.

FIGURE 45
Percentage of children and adults diagnosed with depression or anxiety, 2020.

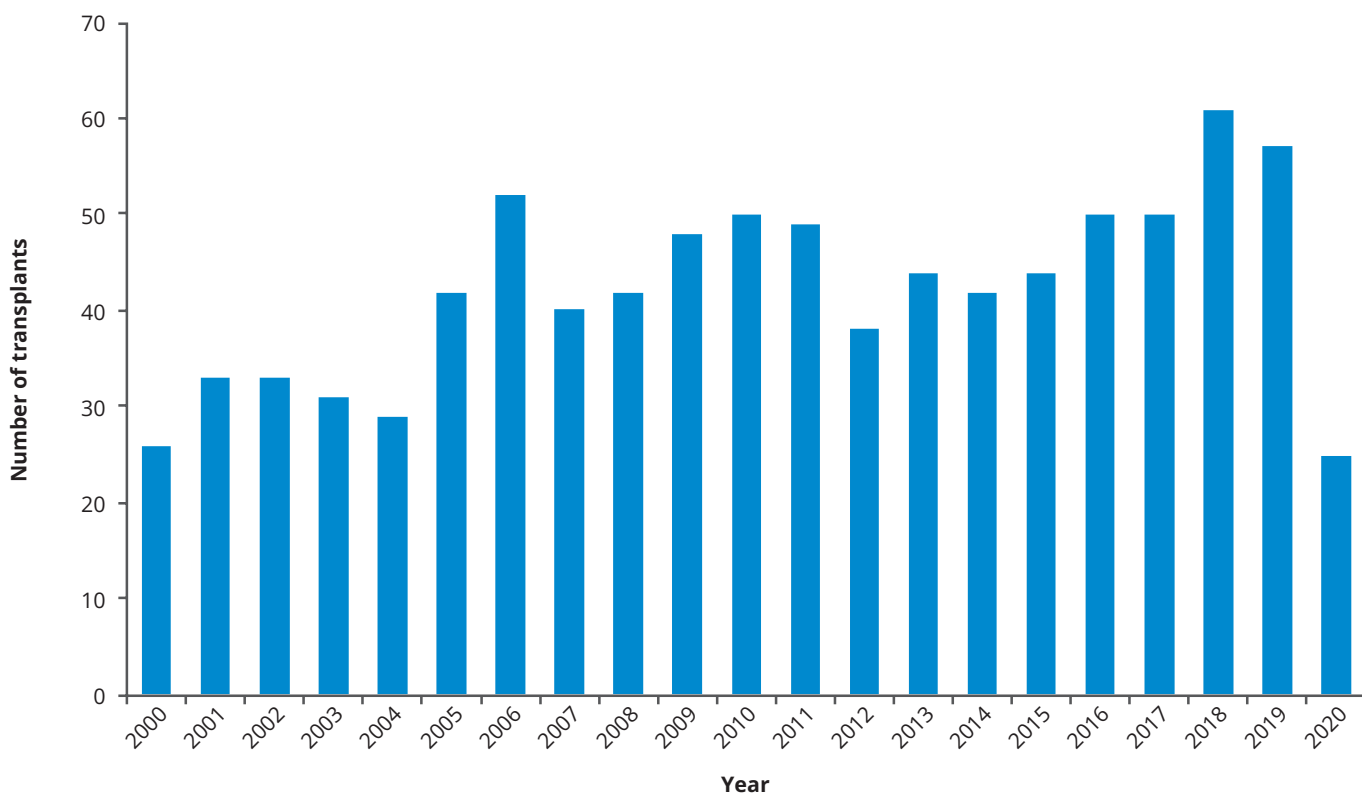


TRANSPLANTS

For some individuals with advanced disease, transplantation may be the next step to help regain health. Figure 46 shows the number of transplants carried out per year as reported in the CF Registry. In 2020, 25 individuals with cystic fibrosis received a transplant with a median age at the time of transplant of 32.9 years. The number of transplants dropped over 50% in 2020, compared to 57 transplants conducted in 2019. Although the numbers provided represent primarily lung transplants (21 lung transplants in 2020), individuals who received other combinations or organs (e.g. lung and liver, liver, heart and lung, heart) are also included in the total.

The first transplant recorded in the CF Registry was performed in 1988. As of December 31, 2020, there were 941 unique individuals with cystic fibrosis reported as having received one or more transplants (any organ). Of these patients, 63 (7%) have received at least two lung transplants, 526 (55.9%) were reported as being alive, and 288 (54.8%) of those living patients were male.

FIGURE 46
Number of transplants per year of cystic fibrosis individuals, 2000 to 2020.



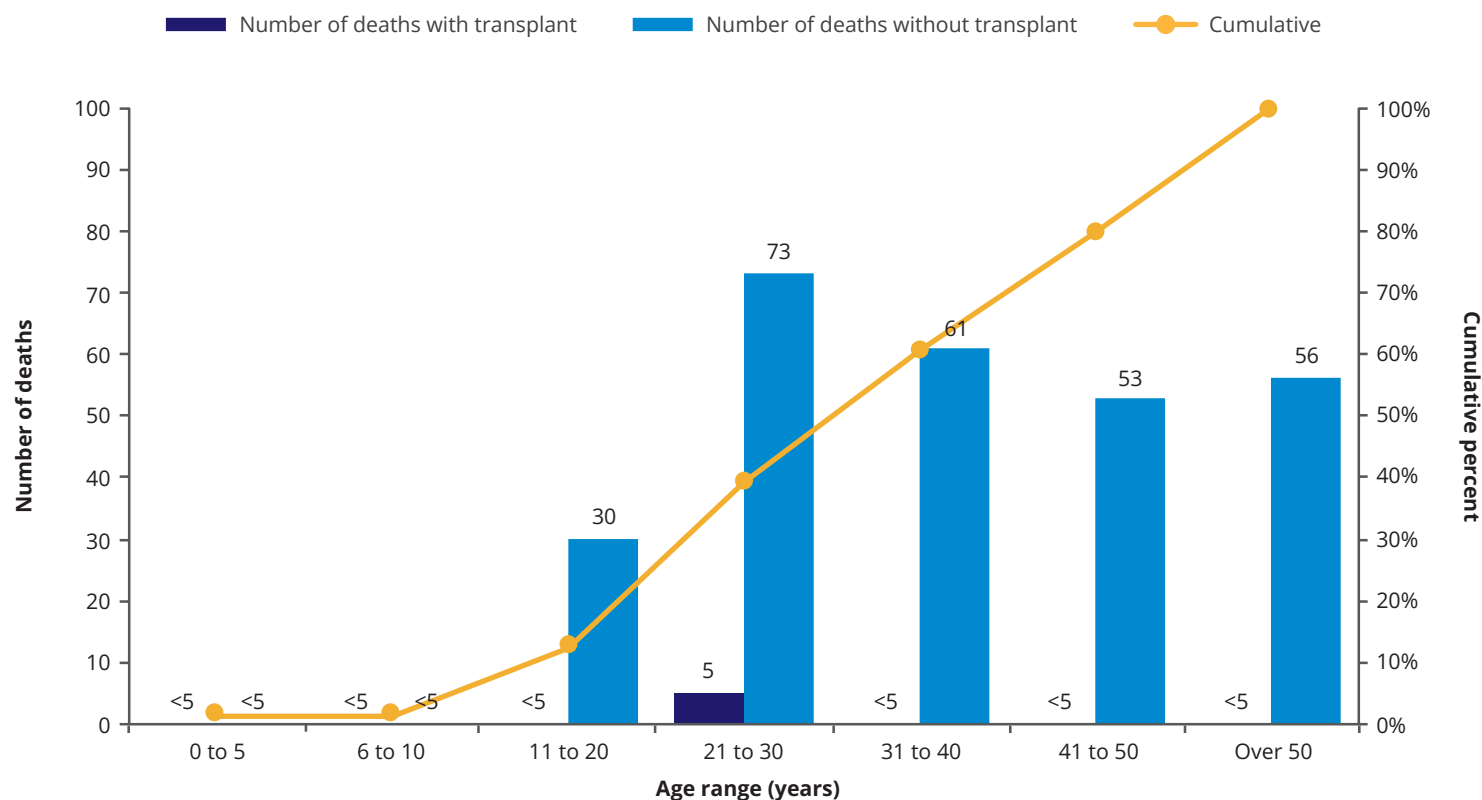
SURVIVAL

The survival and health outcomes in Canadians living with cystic fibrosis continues to improve over time. In 2020, there were 31 deaths recorded in the CF Registry, compared to 50 deaths in 2019. Figure 47 shows the cumulative number of deaths and the age at death from 2015 to 2020.

Risk factors such as pulmonary exacerbations and malnutrition are often associated with increased risk of death. In 2020, of the deaths with a recorded cause of death (31 deaths; 100%), 72.4% of deaths were reported as pulmonary/infection/cardiovascular complications.

20 (64.5%) individuals with cystic fibrosis who died in 2020 had never received a transplant (any organ).

FIGURE 47
Cumulative number of deaths and age at death, 2015 to 2020.



SURVIVAL

Over the past two decades, a gradual increase in the median age of death can be seen. The median age of death was 42.0 years in 2020, compared to 41.9 years in 2019, and 25.2 years in 2001 (Figure 48). The median age of death tells us that half of those who died were younger than 42.0 years of age and the other half who died were older. Large fluctuations in the median age of death can be seen each year because there are relatively few deaths in a given year. However, the annual death rate (calculated as the number of deaths divided by the total number of individuals reported in the year) has been steadily decreasing since 2000 (Figure 49). In 2020, this value was 0.7%.

FIGURE 48
Median age at death per year, 2001 to 2020.

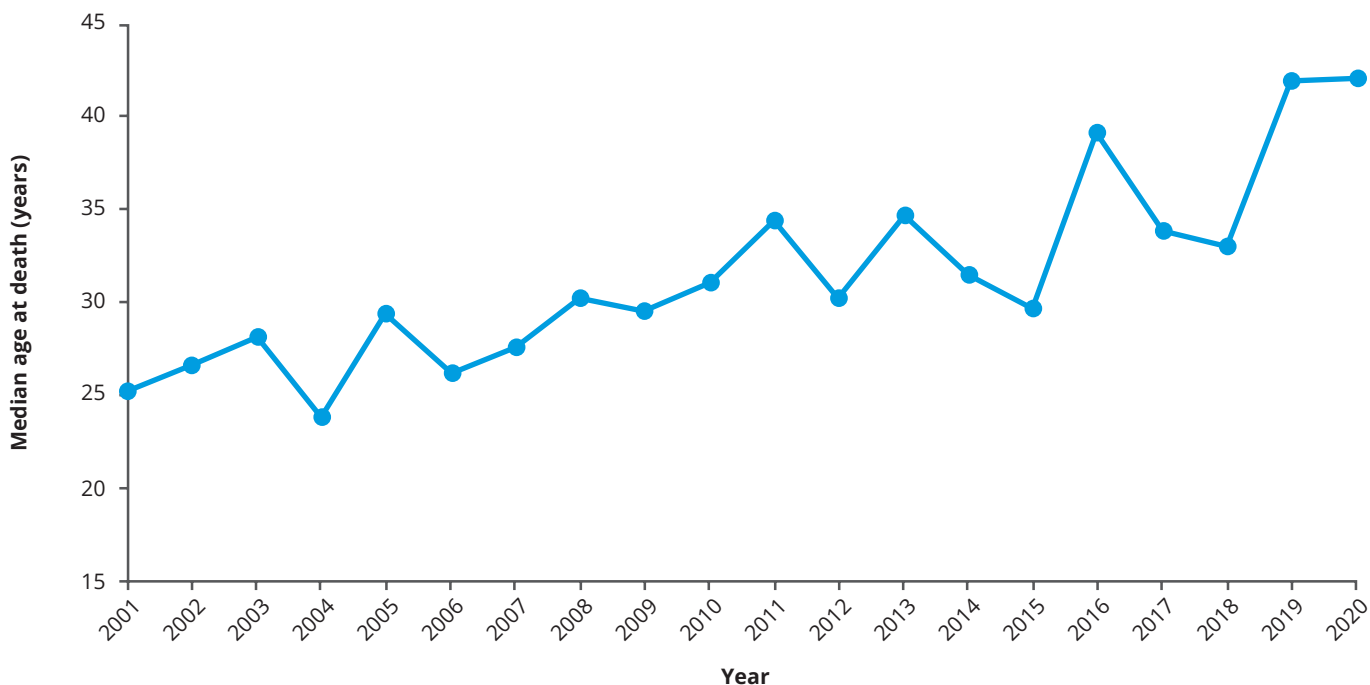
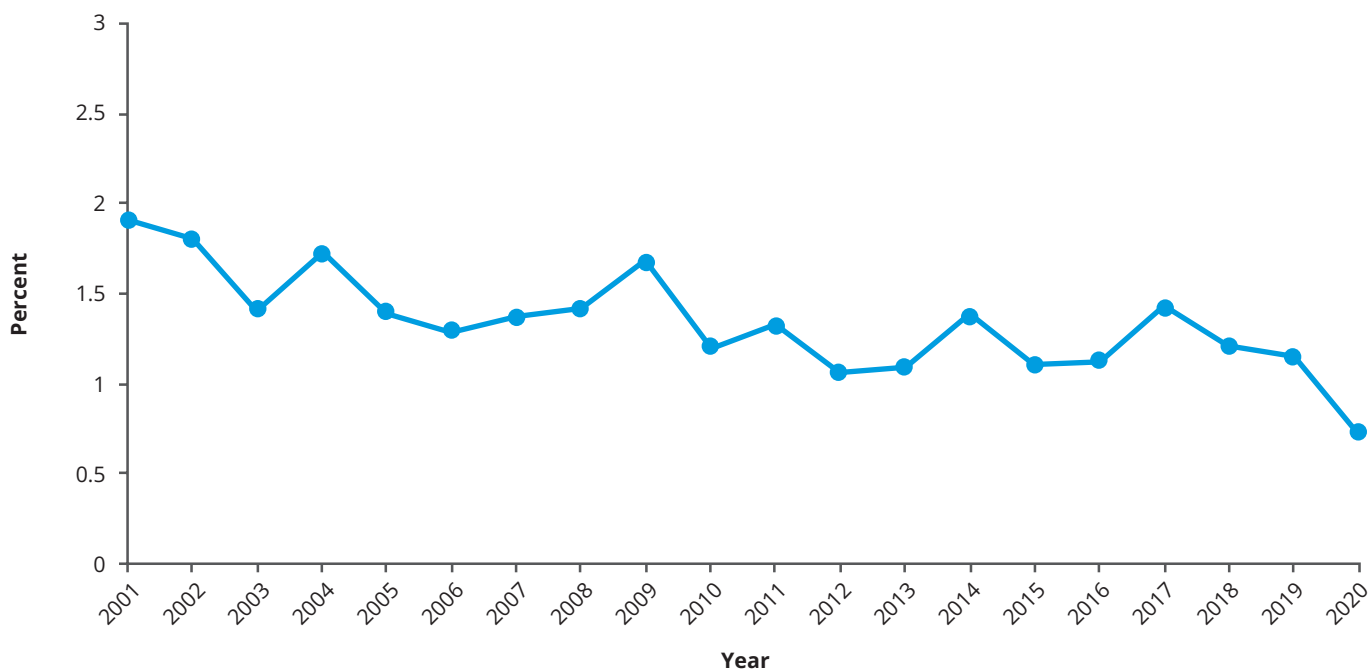


FIGURE 49
Death rate per year, 2001 to 2020.

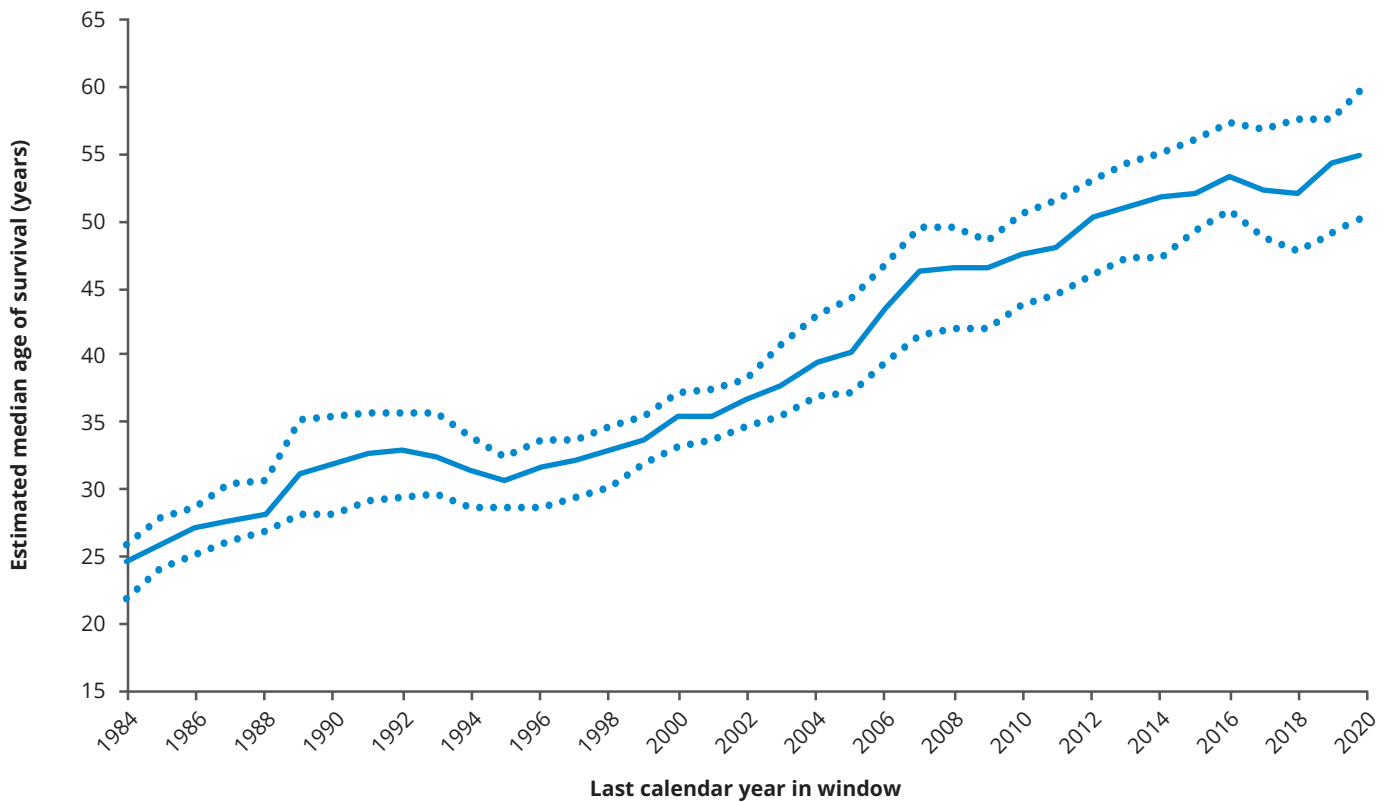


ESTIMATED MEDIAN AGE OF SURVIVAL

A 5-year rolling window, to stabilize the estimates over time, was used to calculate the median age of survival using a Cox proportional hazards model. The most recent 5-year window (2016 to 2020) included 5,101 people with cystic fibrosis and 245 deaths. Out of these people, the number of individuals with cystic fibrosis lost-to-follow-up (defined as individuals with cystic fibrosis we assume are alive but haven't been reported on in the past 2 years) was 273 (5.4%).

In 2020, the median age of survival was estimated to be 55.4 years of age (Figure 50). In 2012, the estimated median age of survival passed 50 years of age for the first time and it has remained steady since. The estimated median age of survival is the age beyond which we expect 50% of babies with cystic fibrosis born today to live, under the assumption that current age-specific mortality rates will remain stable. Transplanted individuals are included in the survival analysis because transplant is considered a form of therapy for end-stage CF. Excluding deaths post-transplant would overestimate the median age of survival ⁸.

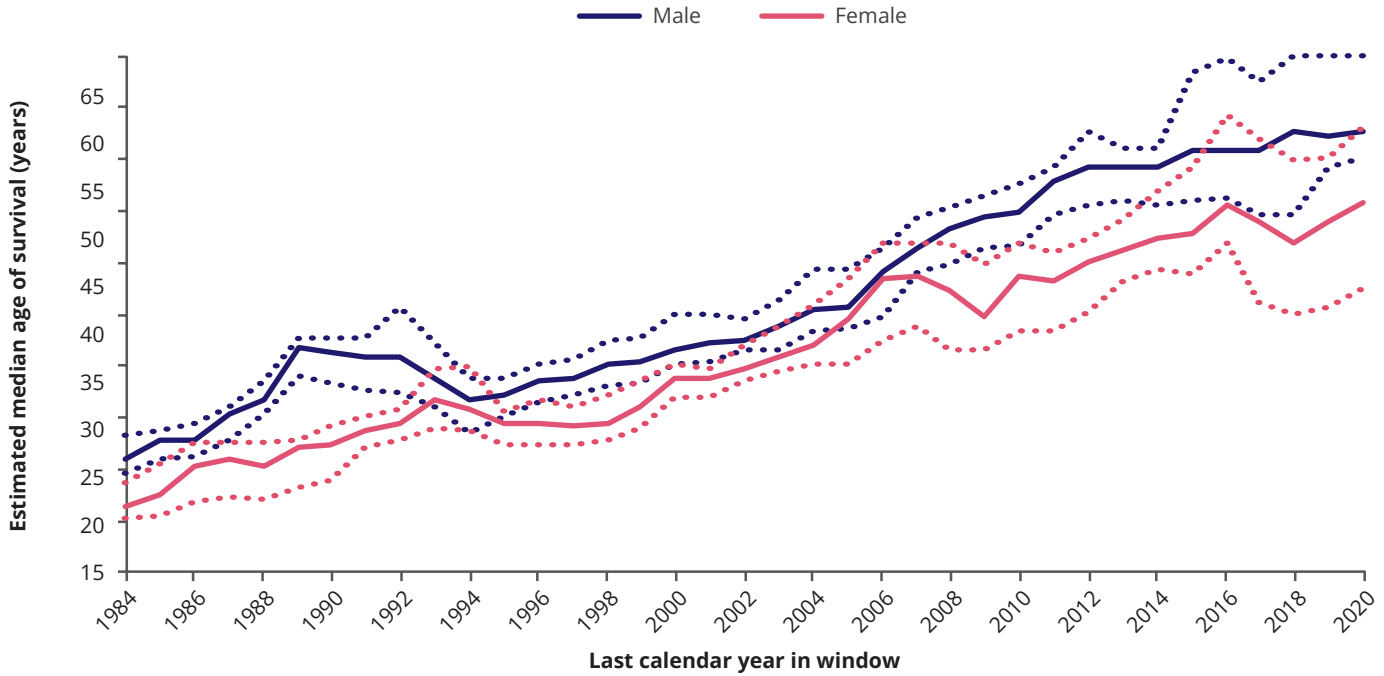
FIGURE 50
Estimated median age of survival for a moving 5-year window with 95% confidence intervals, 1984 to 2020.



ESTIMATED MEDIAN AGE OF SURVIVAL

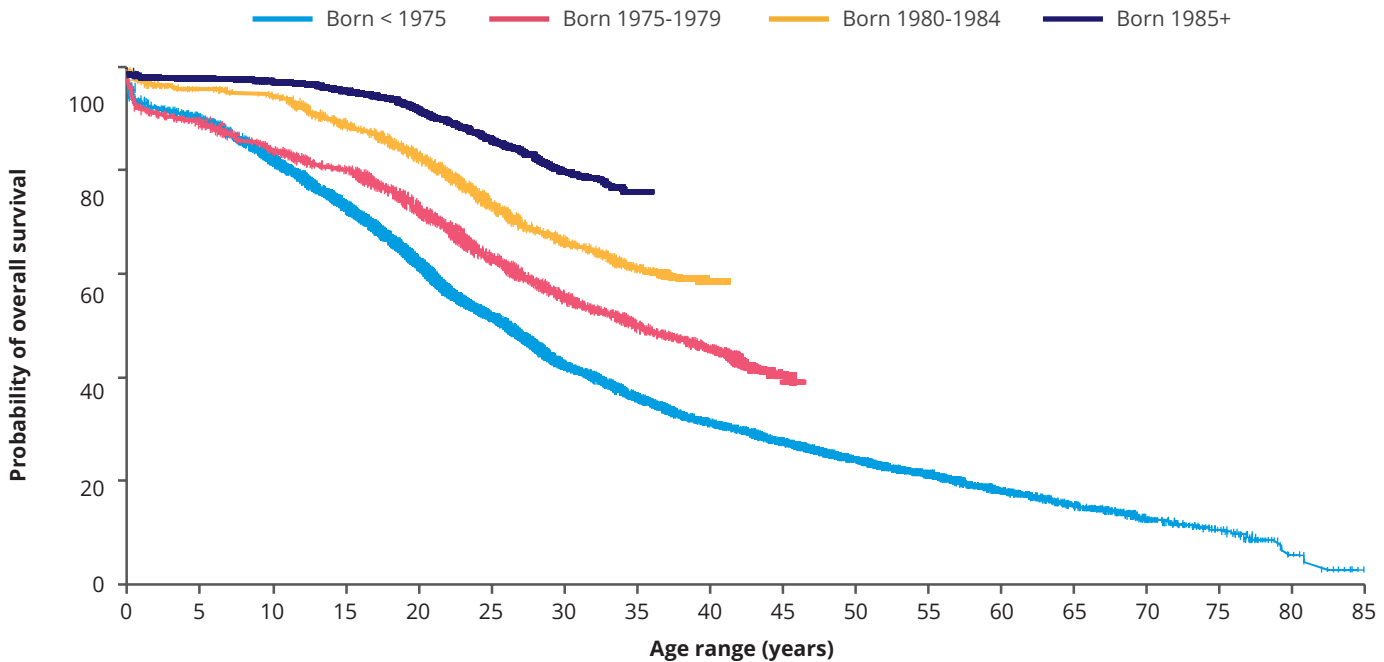
The median age of survival remains stable for both males and females with males continuing to have a higher median age of survival compared to females (Figure 51). While the cause of lower survival in females is not well understood, it has been documented in published CF literature^{9 10 11}.

FIGURE 51
Estimated median age of survival for a moving 5-year window with 95% confidence intervals, by sex, 1984 to 2020.



Survival by birth cohort is presented in Figure 52 and indicates that the expected median age of survival is higher when considering more recent cohorts. The probability of surviving beyond age 20 years is 91.8% for those born in 1985 or later compared to 62.2% for those born before 1975.

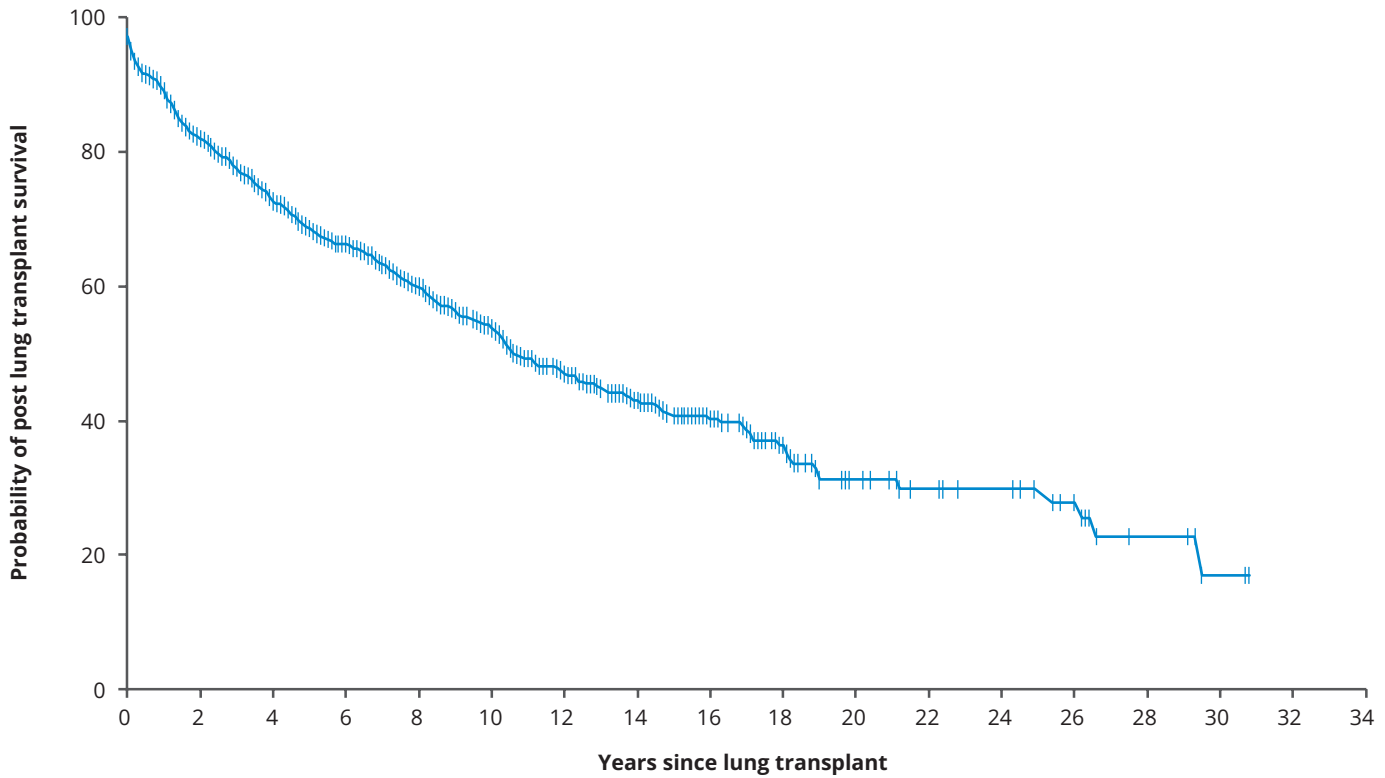
FIGURE 52
Overall survival of individuals with cystic fibrosis, by birth cohorts, 2020.



POST LUNG TRANSPLANT SURVIVAL

Between 1988 and 2020, there were 906 lung transplant recipients and 398 deaths post-lung transplant. Figure 53 shows the probability of survival post lung transplant which is 89.1% at one year, 77.6% at three years and 68.6% at five years. Overall, 50% of those patients transplanted today would be expected to live beyond 10.7 years following lung transplantation.

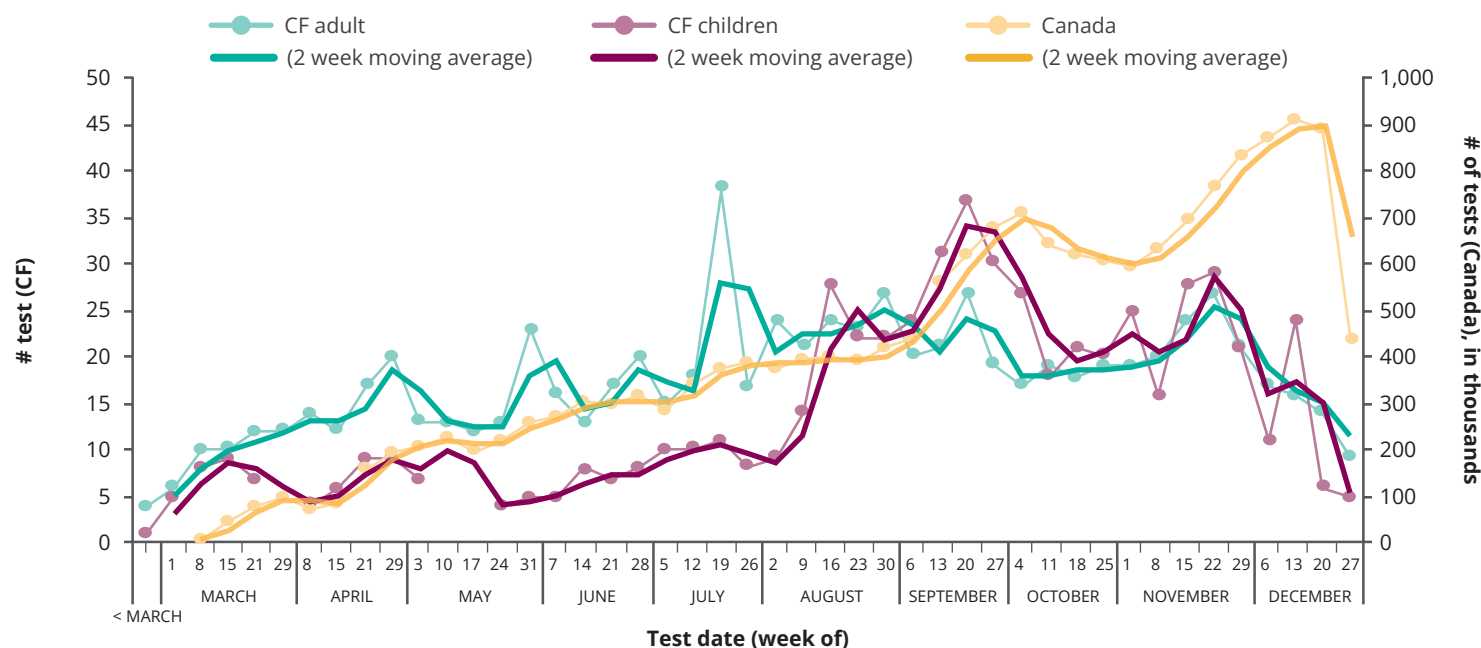
FIGURE 53
Post lung transplant survival, 2020.



COVID-19

Between January 1 and December 31, 2020, 1,422 tests for SARS-CoV-2 (the virus that causes COVID-19) were recorded in the CF Registry. These tests were recorded among 911 unique individuals (21.0% of the Canadian CF population), and by 35 of the 42 CF clinics across Canada. Figure 54 shows the weekly number of tests conducted for children and adults living with CF alongside testing among the general Canadian population. Through the first half of the year, adults were routinely tested more than children, however beginning in late summer (coinciding with the start of the new school year), tests among children increased.

FIGURE 54
Weekly SARS-CoV-2 testing among individuals with CF and general Canadian population.



In 2020, a total of 26 people living with CF (7 children and 19 adults) tested positive for SARS-CoV-2. As seen in Table 8, these individuals were younger and more often female compared to the Canadian CF population.

TABLE 8
Characteristics of Canadians with CF who tested positive for SARS-CoV-2 in 2020 compared with the general Canadian CF population.

	CANADIAN CF POPULATION (N = 4,332)	SARS-COV-2 (N = 26)
Median age (years) (range)	24 (0 to 83)	27 (2 to 66)
Female	2,028 (47%)	14 (54%)
Overweight	812 (19%)	5 (19%)
Post-transplant	309 (7%)	5 (19%)
Median best FEV ₁ percent predicted(%):		
All (IQR)	81% (60-98)	79% (54-99)
Adult (IQR)	70% (51-88)	68% (43-91)
Children (IQR)	98% (87-106)	104% (91-106)

The course of disease appeared to be milder than originally anticipated, with 9 (34.6%) requiring hospitalization. There were no deaths recorded among those who tested positive in 2020. Follow-up clinical information (including lung function post-infection) were available for 18 (69.2%) individuals, and the median change in lung function after COVID-19 infection was -5.6%.

The CF Registry will continue to report on the mid-to-long term clinical and health system impacts of COVID-19 infection.

GLOSSARY OF TERMS

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 80 years for males and 84 years for females based on data from the World Health Organization ¹². This means that, on average, a male baby born today will be expected to live 80 years and a female baby, on average, will be expected to live to 84 years of age. Life expectancy is not the same as median age of survival. 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.

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.

Median Age at Death

The median age at death is very different from 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 **age beyond which we expect 50% of babies with cystic fibrosis born today to live, under the assumption that recent age-specific mortality rates will hold for the rest of their lives** ¹³. This is NOT the age at which people with cystic fibrosis would be expected to die, (i.e. how long someone can expect to live, on average - see life expectancy above). 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 cystic fibrosis are living (for example, median age at death and annual death rate).

When we say that the median age of survival in 2020 is 55.4 years, we are saying that if a child with cystic fibrosis is born in Canada in 2020, they have a 50% chance of living beyond 55.4 years of age based on current mortality rates. In other words, half of the CF population would be expected to live to an age older than 55.4 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 in 2020.

It is important to note that these survival estimates apply to a population of individuals and do not necessarily apply to any one individual.

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ACKNOWLEDGMENTS

Thank you to the following groups and people who made outstanding contributions to the Canadian Cystic Fibrosis Registry and this 2020 Registry Annual Data Report.

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**FUNDING FOR THE DESIGN AND DISTRIBUTION OF THIS REPORT WAS GENEROUSLY
SUPPORTED BY AN UNRESTRICTED GRANT FROM**



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