

# 2019 ANNUAL DATA REPORT



## THE CANADIAN CYSTIC FIBROSIS REGISTRY

# 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. Currently, there is no cure.



## CYSTIC FIBROSIS CANADA

Cystic Fibrosis Canada has achieved firsts, breakthroughs and quantified improvements in lifespan for people living with cystic fibrosis. We have never worked alone. Our team is committed to working with, and for, the people living with cystic fibrosis.

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, government and donors. We work together going further to change lives through treatments, research, 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](http://www.cysticfibrosis.ca).

**OUR VISION IS A  
WORLD WITHOUT  
CYSTIC FIBROSIS**  
**OUR MISSION IS TO END CYSTIC FIBROSIS**

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This publication is also available online. Please visit us at [www.cysticfibrosis.ca](http://www.cysticfibrosis.ca)

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Cover page: CF individual from Ontario



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# THE CANADIAN CYSTIC FIBROSIS

## 2019 ANNUAL DATA REPORT

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# THE CANADIAN CYSTIC FIBROSIS REGISTRY

The *Canadian Cystic Fibrosis Registry* (CCFR) 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 CCFR. Data are submitted by the CF clinics on behalf of patients. Given that most CF patients attend one of these clinics, we are confident that the CCFR 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 THE 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.

Patients whose data were reported by any of the 42 accredited Canadian CF clinics in 2019 are included in this report. In light of the on-going global COVID-19 pandemic, CF Canada has worked closely with CF clinics to complete data entry for 2019. However, data in this report may not be complete for all clinics.

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, 2019.

I am pleased to share the 2019 Annual Data Report for the Canadian Cystic Fibrosis Registry. Since its inception in the 1970's, the Registry, continues to be an important resource that helps to deepen our understanding of cystic fibrosis. Data collected from the Registry is used by researchers, clinicians and policy makers and more recently has supported clinical trials through the Cystic Fibrosis Canada Accelerating Clinical Trials Network (CF CanACT), an integral step in the development of new cystic fibrosis treatments and therapies.

This year, through the Registry, we were able to quickly assess the impact COVID-19 could have on the cystic fibrosis community through a global collaborative study, where data was collected from the Registries of eight countries, including Canada. Released on April 30, 2020, the results, while early, suggested encouraging health outcomes for people living with cystic fibrosis who have contracted COVID-19. We were happy we could support this global review of the impact of the pandemic on our community.

The Registry also helps us to advance our advocacy efforts for improved access to life-changing cystic fibrosis medications. For instance, by using data from the Registry, important research released in August predicted the impact that access to Trikafta would have on the CF population in the future and the impact of delayed access to this life-changing drug. This research helps us to demonstrate to key government decision makers the urgency of our issues and the impact of action or inaction on our community.

Without the support of cystic fibrosis patients and the dedication and hard work of clinic staff, the Registry and the impact of its data would not be possible. I would also like to acknowledge our many donors, whose contributions support this important work. To all, on behalf of Cystic Fibrosis Canada, thank you.

Sincerely,



**Kelly Grover**

President and CEO

Cystic Fibrosis Canada

It is my pleasure to present the 2019 CF Registry Annual Data Report summarizing the Canadian CF population. I want to acknowledge the incredible effort that goes into maintaining the Registry both by individuals with CF and their families who allow their data to be captured, staff at clinics who enter the data, and the team at CF Canada who process and maintain the data. All this effort has really paid off over the years and the power of the registry has never been more evident, both at the population and individual level.

Many years ago, Registry data were pivotal in showing the importance of nutritional supplementation in CF and its impact on survival. More recently, Registry data was used to publish a study showing the dramatic life-saving benefits of the breakthrough cystic fibrosis modulator drug, Trikafta. Amidst the current global COVID-19 pandemic, the Canadian CF Registry also worked in collaboration with international registries to publish a manuscript in the Journal of Cystic Fibrosis, capturing the most detailed information to date on how COVID-19 impacts this high-risk population. The impact of Registry data has been felt not only within Canada, but also internationally with between-country comparisons shining a spotlight on disparities. At a patient level, Registry data is used by clinics to identify potentially eligible subjects for clinical trials of new therapies and to identify areas for improvement that can be the basis for quality improvement initiatives.

As we look towards the future, we hope the Registry will continue to support clinics and patients to meet the evolving need of the Canadian CF population.

Sincerely,



**Dr. Anne Stephenson**


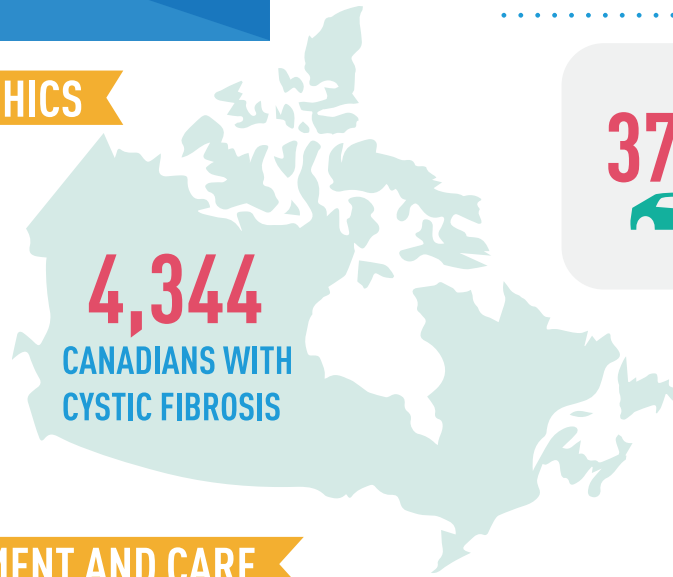
Medical Director, Registry, Cystic Fibrosis Canada and CF Physician, Unity Health Toronto, St. Michael's site, Toronto

# 2019 HIGHLIGHTS FROM THE CYSTIC FIBROSIS REGISTRY

## DEMOGRAPHICS

**116** NEW CF  
DIAGNOSES

**76** THROUGH  
NEWBORN  
SCREENING

**37%** TRAVELLED MORE  
THAN **100 KM** TO  
RECEIVE CF CARE



**62%**  
ARE ADULTS

## CF TREATMENT AND CARE



**18,900+**  
CLINIC VISITS



**25,200+**  
HOSPITAL DAYS



**15,500+**  
HOME IV DAYS



**46**  
LUNG TRANSPLANTS



**38%** HAVE *PSEUDOMONAS  
AERUGINOSA* INFECTIONS



**34%** OF ADULTS HAVE  
CF-RELATED DIABETES

## HEALTH OUTCOMES

**63%** of ADULTS  
AND  
**73%** of CHILDREN

HAVE AN  
ADEQUATE WEIGHT

MEDIAN LUNG FUNCTION:



**67.9%**  
FOR  
ADULTS



**93.4%**  
FOR  
CHILDREN

## SURVIVAL

ESTIMATED MEDIAN POST-LUNG  
TRANSPLANT SURVIVAL IS

**10.6 YEARS**



ESTIMATED MEDIAN AGE OF  
SURVIVAL IS

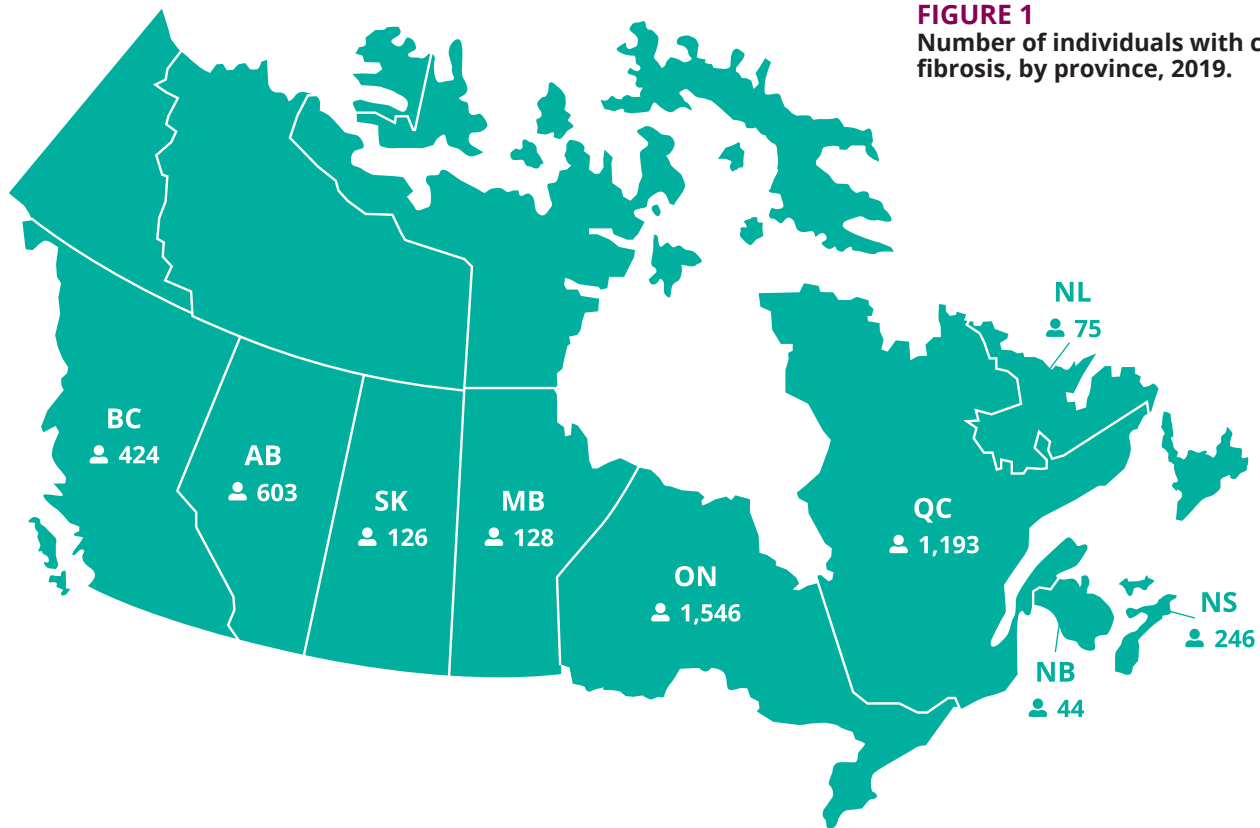
**54.3 YEARS OF AGE**



# DEMOGRAPHICS

## CANADIANS WITH CYSTIC FIBROSIS

In 2019, there were a total of 4,344 individuals with cystic fibrosis who attended one of the 42 accredited CF clinics across Canada (Figure 1), with 116 of those being newly diagnosed with cystic fibrosis. Overall, the total Canadian CF population has been steadily increasing and in the last two decades, has grown by 33.3% (Figure 2). Individuals are associated with the province in which they attended a CF clinic. Those who attended CF clinics in multiple provinces in 2019 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.



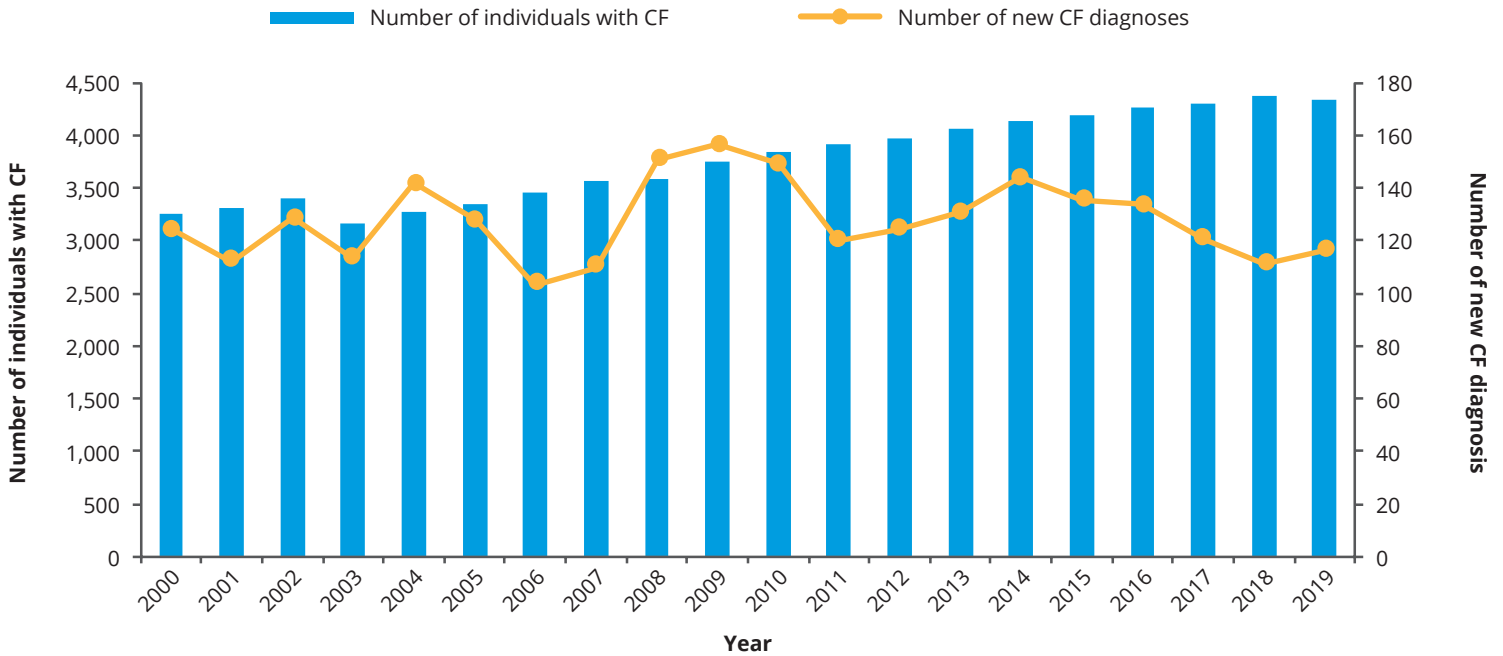
**FIGURE 1**  
Number of individuals with cystic fibrosis, by province, 2019.

PROVINCE *	NUMBER OF INDIVIDUALS WITH CF	FEMALE	MALE	ADULTS	CHILDREN
BC	424	177	247	269	155
AB	603	295	308	353	250
SK	126	50	76	64	62
MB	128	58	70	67	61
ON	1,546	743	803	954	592
QC	1,193	552	641	781	412
NB	44	24	20	32	12
NS	246	112	134	153	93
NL	75	30	45	52	23

\* 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

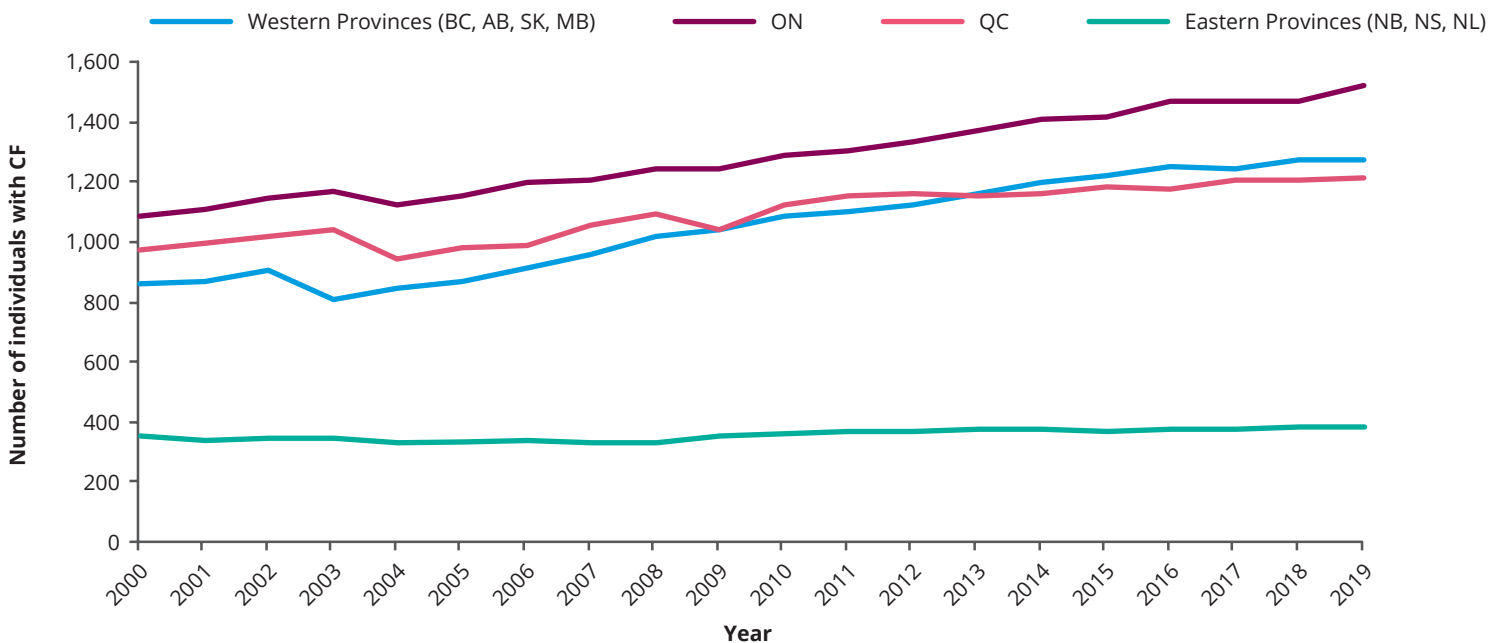
**FIGURE 2**  
Total number of individuals with cystic fibrosis and new CF diagnoses, 2000 to 2019.



## PROVINCIAL POPULATION CHANGE

Over the past two decades, the Canadian CF population has grown, with the largest increases seen in clinic visits in Ontario and the western provinces of British Columbia, Alberta, Saskatchewan, and Manitoba (Figure 3).

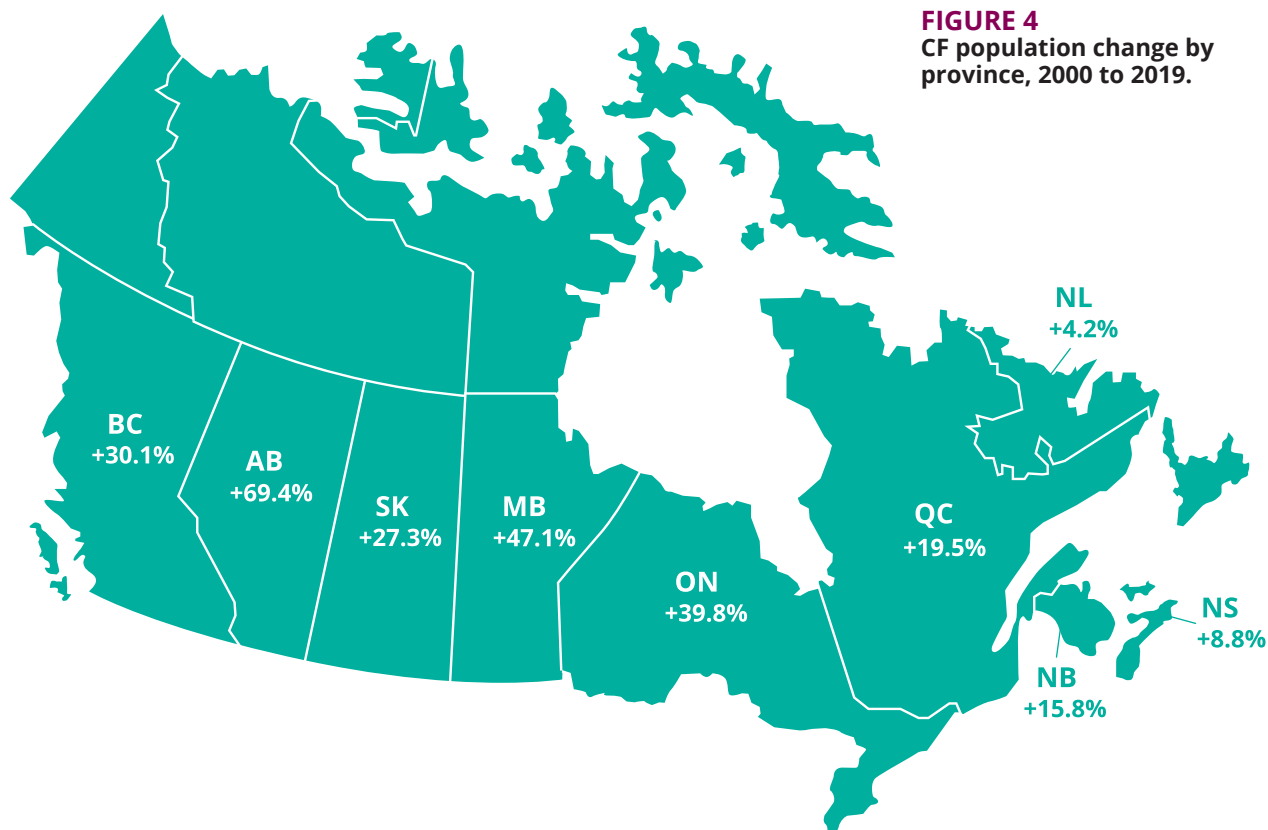
**FIGURE 3**  
CF population growth by region, 2000 to 2019.





# PROVINCIAL POPULATION CHANGE

Figure 4 shows the number of individuals with CF reported on by clinics within each province in 2000 and 2019, 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.



**FIGURE 4**  
CF population change by province, 2000 to 2019.

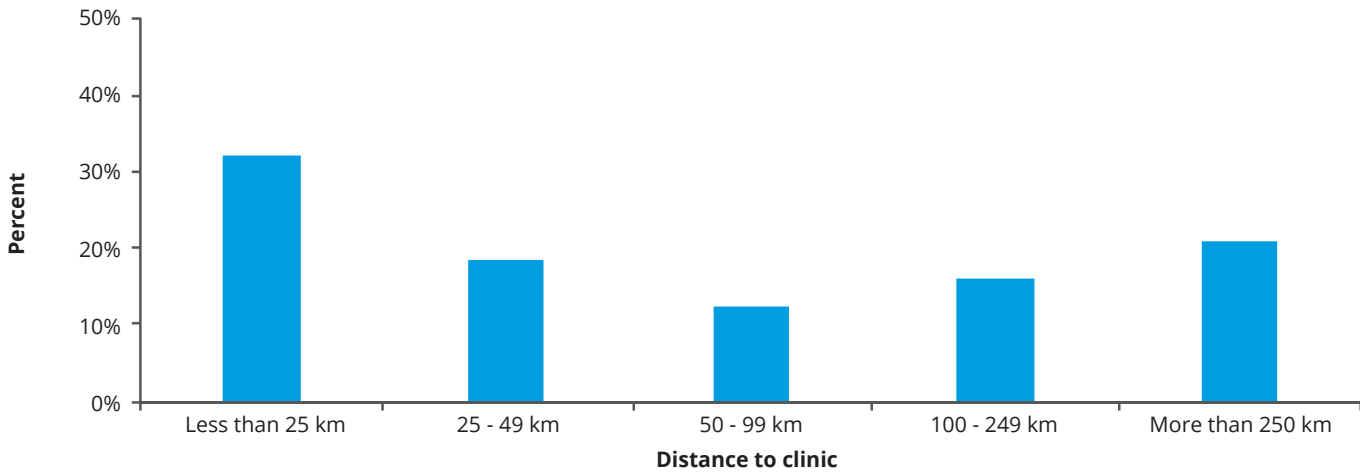
PROVINCE *	2000	2019	PERCENT CHANGE
BC	326	424	+30.1%
AB	356	603	+69.4%
SK	99	126	+27.3%
MB	87	128	+47.1%
ON	1,106	1,546	+39.8%
QC	998	1,193	+19.5%
NB	38	44	+15.8%
NS	226	246	+8.8%
NL	72	75	+4.2%

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

# DISTANCE TO CLINICS

The CCFR 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 2019, there were 1,456 (33.5%) cystic fibrosis individuals with at least one valid location recorded (Figure 5). While 51% of those with a reported location attend a CF clinic within 50 km of where they live, 37% travel more than 100 km and 21% travel more than 250 km for their CF care.

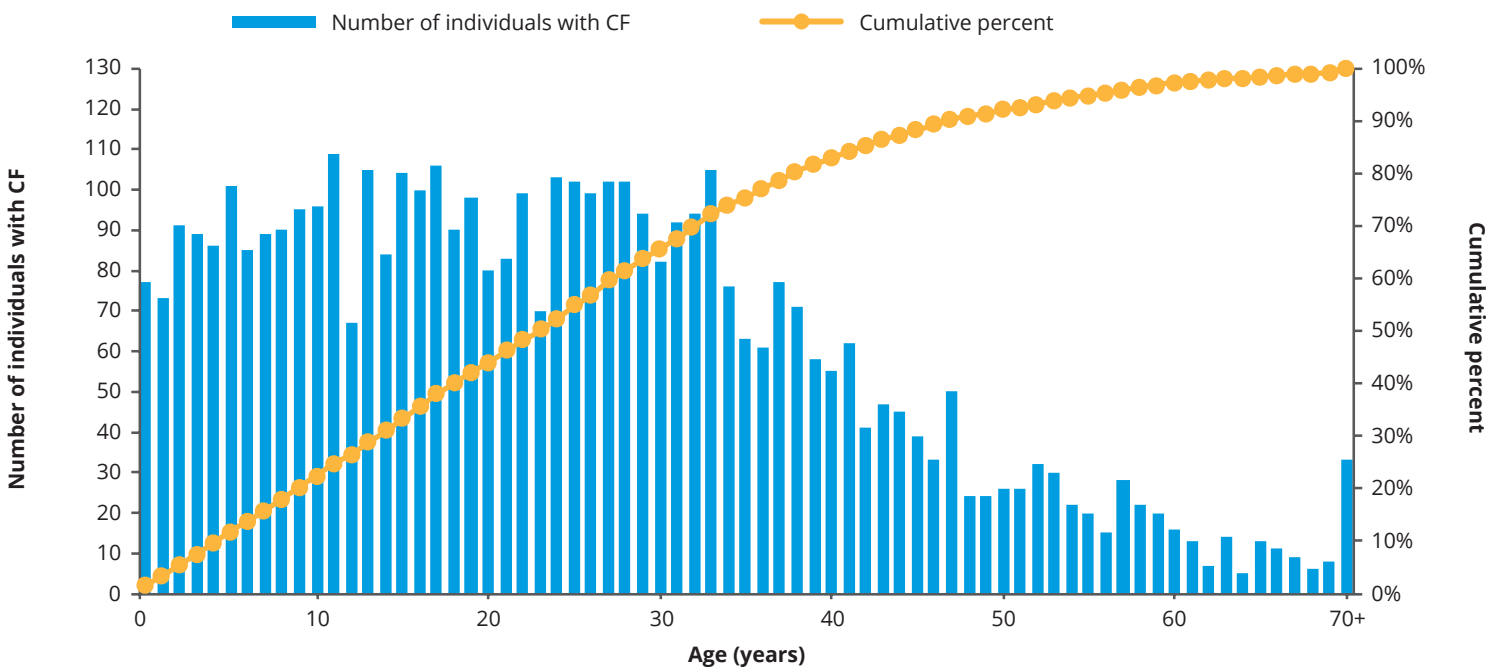
**FIGURE 5**  
Distance travelled to clinic for individuals with cystic fibrosis (N = 1,456), 2019.



# AGE DISTRIBUTION OF CANADIANS WITH CYSTIC FIBROSIS

Figure 6 shows the age distribution of the 4,344 Canadians living with cystic fibrosis in 2019.

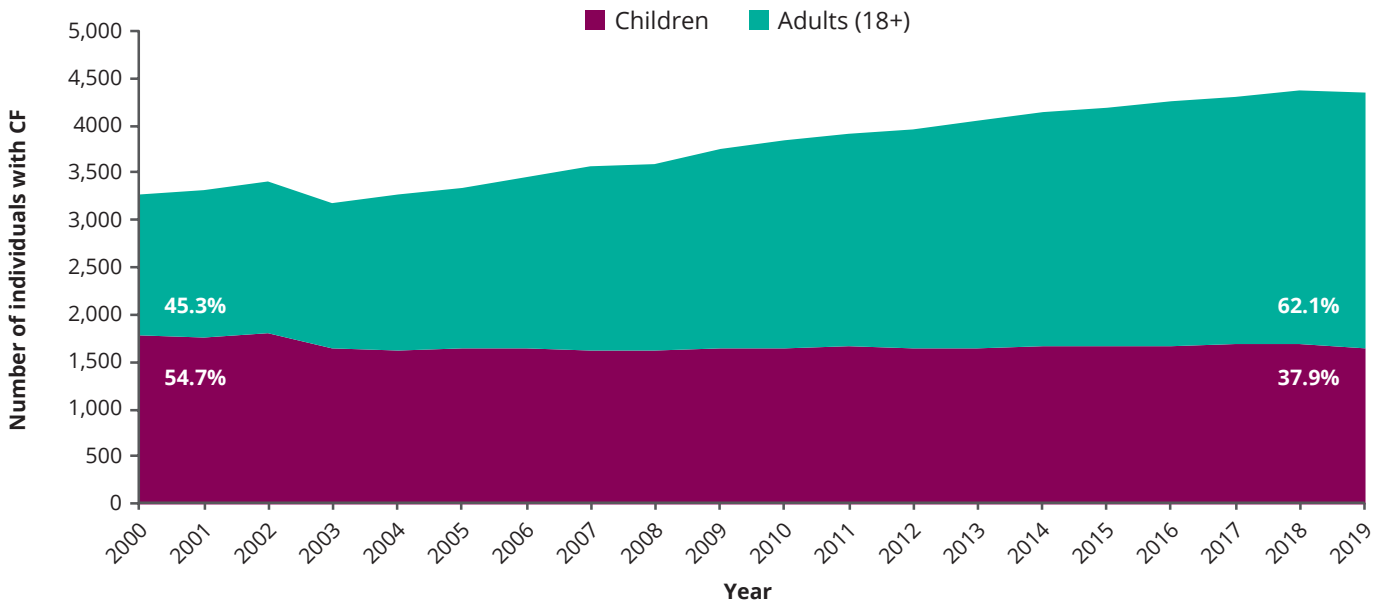
**FIGURE 6**  
Age distribution of individuals with cystic fibrosis, as of December 31, 2019.



# AGE DISTRIBUTION OF CANADIANS WITH CYSTIC FIBROSIS

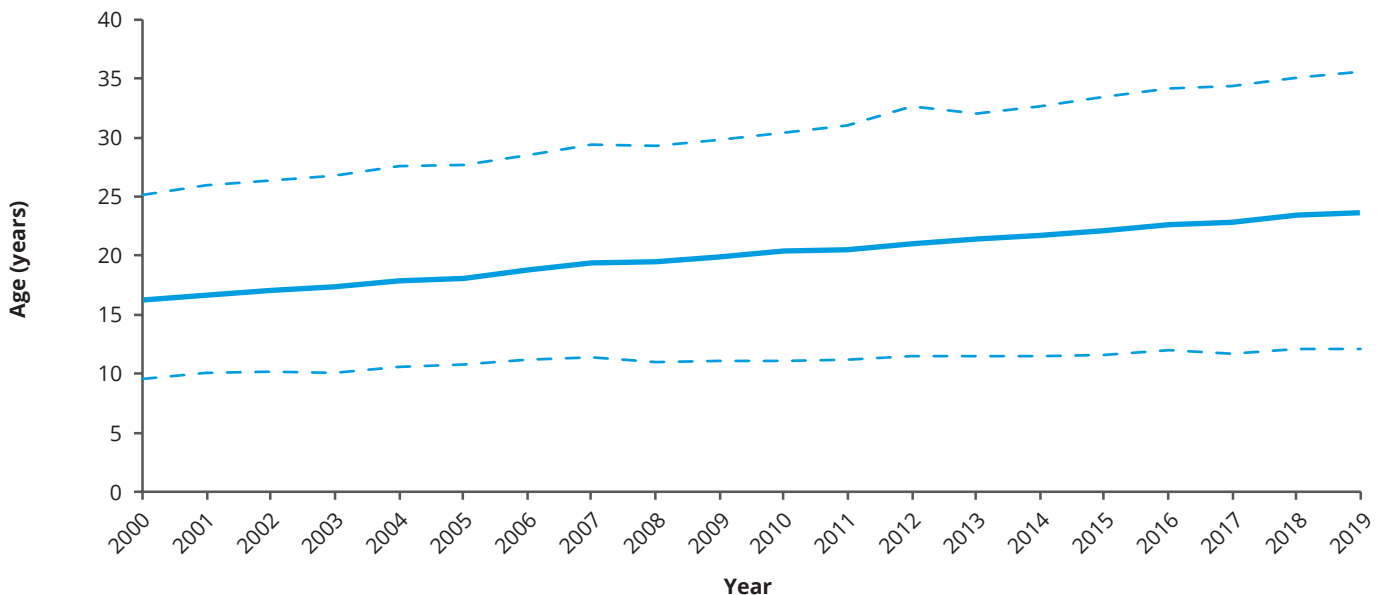
Improvements in treatment and care in the last few decades has led to an increase in the number of Canadian adults living with cystic fibrosis. Twenty years ago, less than half of all Canadians living with cystic fibrosis were adults (individuals aged 18 years and older) (Figure 7). In 2019, adults accounted for 62.1% of the Canadian CF population, with 18.3% of adults aged 40 years and over.

**FIGURE 7**  
Number of children and adults with cystic fibrosis, 2000 to 2019.



It follows that the median age of individuals with cystic fibrosis has increased steadily over the past 20 years. From just over 16 years in 2000, to 23.7 years among those reported on in 2019 (shown in Figure 8 along with the 25<sup>th</sup> and 75<sup>th</sup> percentile of ages).

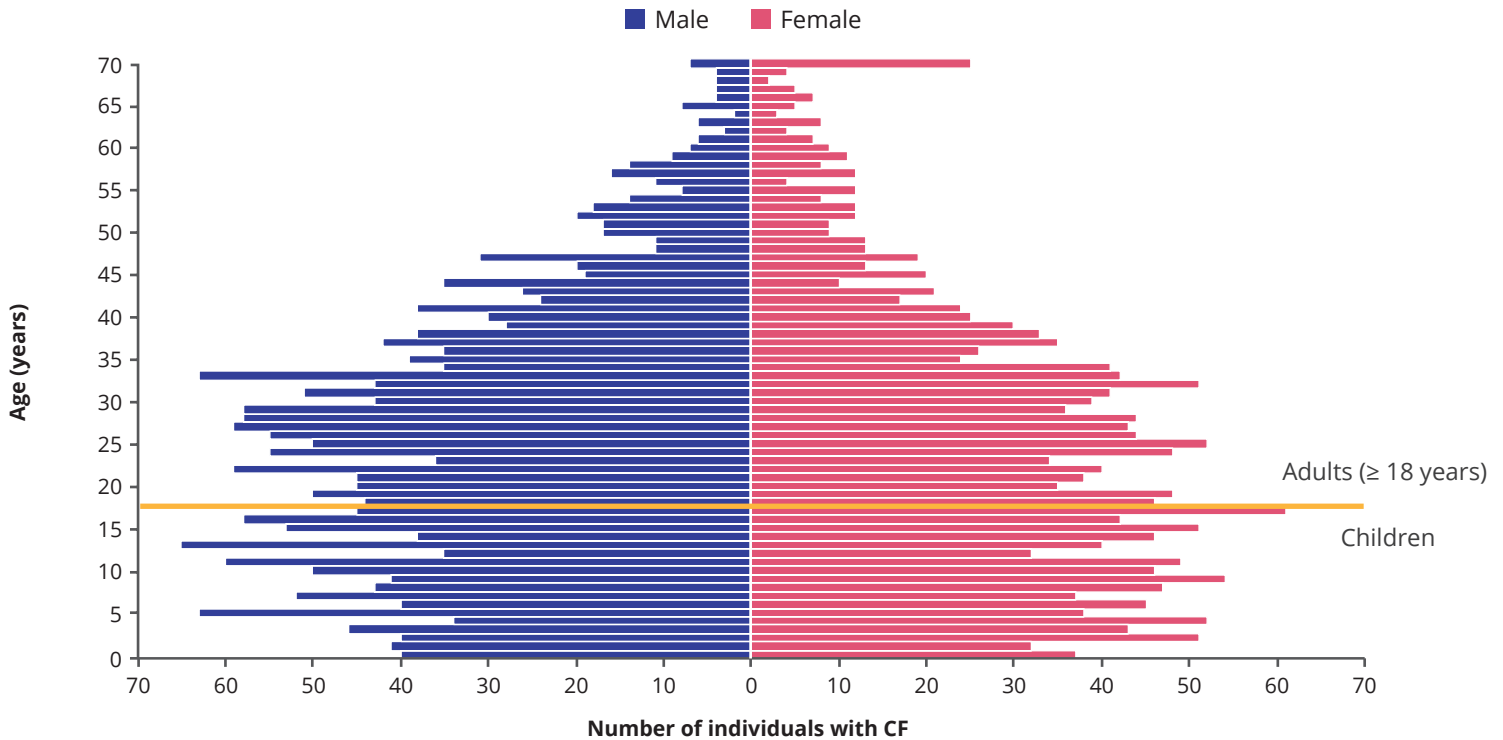
**FIGURE 8**  
Median age of individuals with cystic fibrosis, 2000 to 2019.



# AGE AND SEX DISTRIBUTION OF CANADIANS WITH CYSTIC FIBROSIS

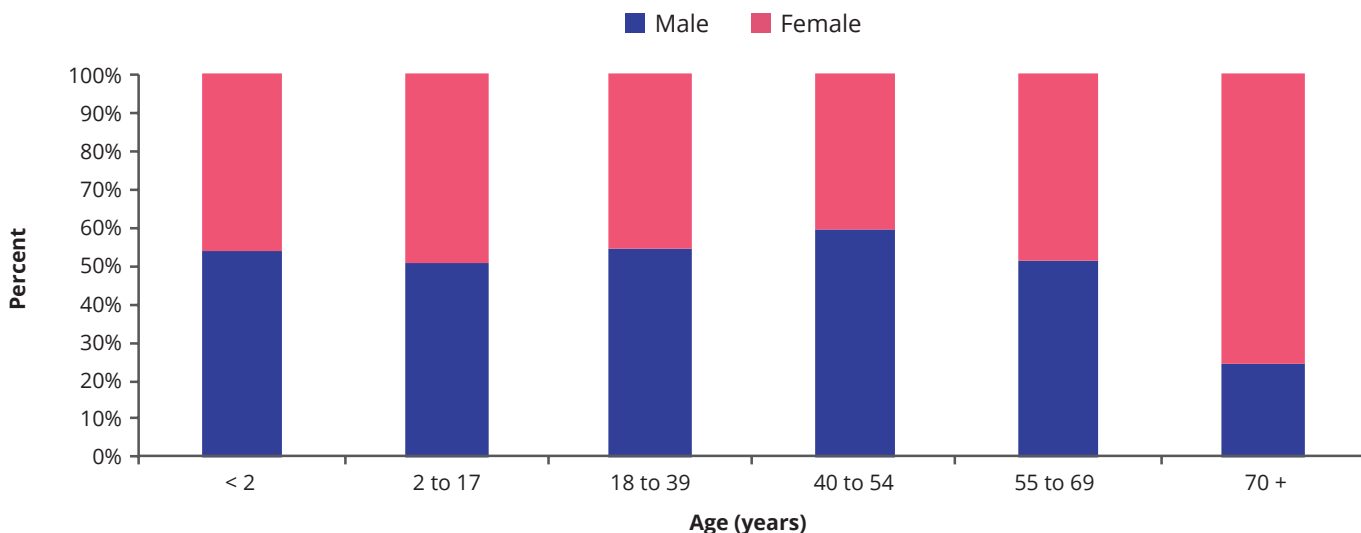
Figure 9 shows the age-sex distribution (referring to biological sex) for all individuals reported on in 2019. Overall, males accounted for 53.4% of individuals living with cystic fibrosis, however the proportion of males varied by age group.

**FIGURE 9**  
Population distribution of individuals (N = 4,344) with cystic fibrosis, by age and sex, as of December 31, 2019.



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 59.5% male for those aged 40 to 54 years. After age 55, the proportion of females begins to increase to 75.8% female for those aged 70+.

**FIGURE 10**  
Sex-distribution of individuals with cystic fibrosis, by age group, as of December 31, 2019.



# DIAGNOSIS

## AGE AT DIAGNOSIS

More than half (60.5%) of individuals with cystic fibrosis reported on in 2019 were diagnosed before the age of one year, and nearly three quarters (73.4%) were diagnosed by the age of two years (Figure 11). Adults diagnosed later in life (18 years and older) accounted for only 7.9% of all individuals diagnosed in 2019.

**FIGURE 11**  
Age at diagnosis of cystic fibrosis individuals, as of December 31, 2019 (N = 4,258).

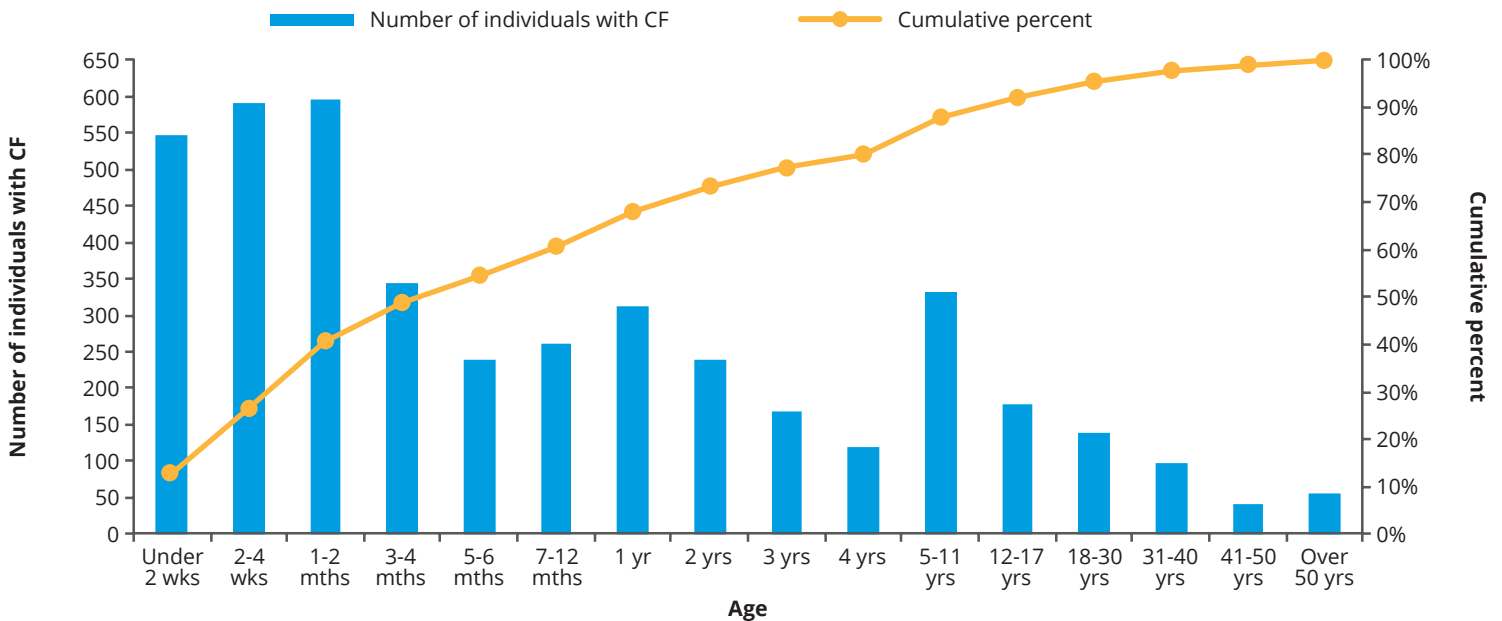
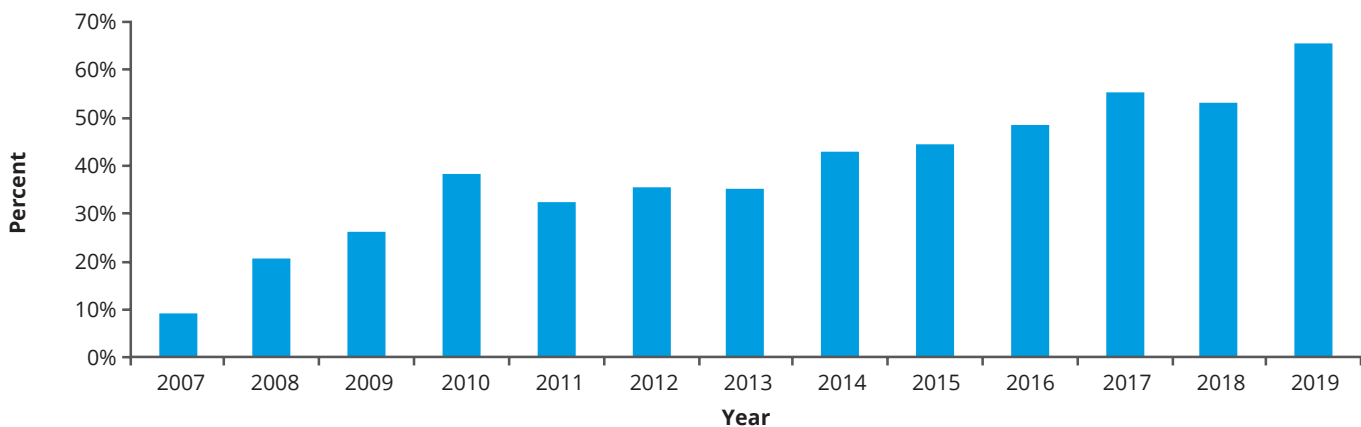


Figure 12 shows the percentage of newborns diagnosed through provincial newborn screening (NBS) programs since 2007, when NBS for CF started in Alberta. At that time, only 9% of new CF diagnoses were identified through NBS. In the spring of 2018, Quebec became the last jurisdiction to start screening newborns for cystic fibrosis. In 2019, nearly two-thirds of new diagnoses (76; 65.5%) were made through NBS, meaning more babies are receiving an early CF diagnosis than ever before. 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 2019.



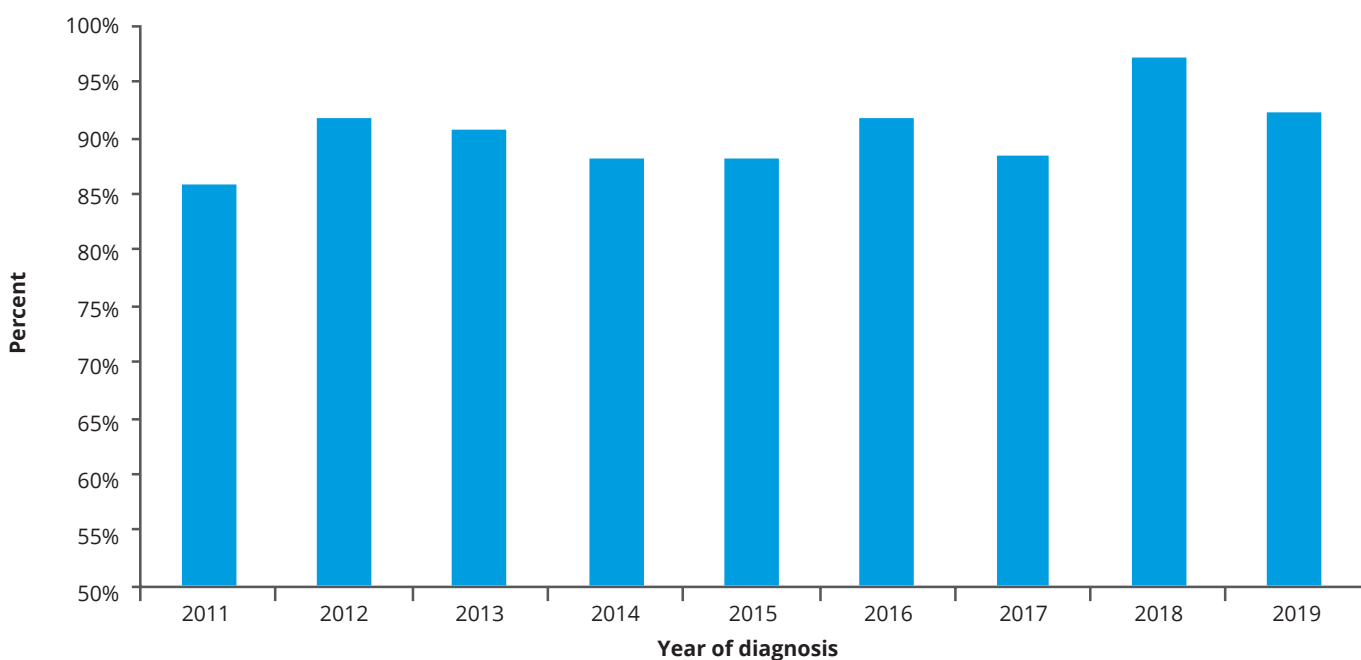
# SWEAT CHLORIDE TESTING

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.

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 CCFR 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 2019, 107 of the 116 (92.2%) 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 2019.



# 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 2,090 different mutations in the *CFTR* gene have been identified.

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.

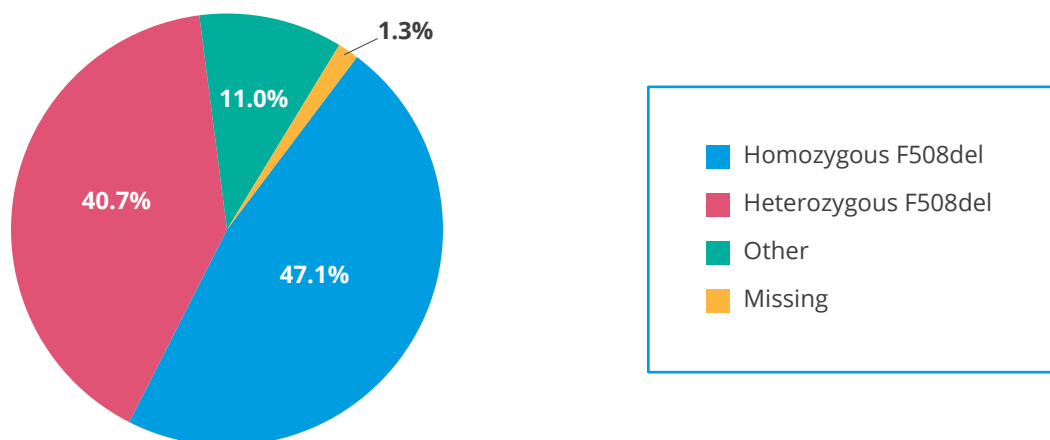
**TABLE 1**  
Classification of CFTR mutations based on the impact on the CFTR protein.

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 all the people ever reported on in the CCFR, 390 people have no mutations recorded. The majority (90.3%) are adults, 9.7% are under 18 years of age, and 0.5% are under 1 year of age.

Nearly all individuals with cystic fibrosis reported on in 2019 (4,289; 98.7%) had at least one CF mutation recorded. Almost half (2,044; 47.1%) have two copies of the F508del mutation (referred to as homozygous F508del) and 40.7% 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 = 4,344), 2019.



# 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 (51.0%) while those diagnosed as an adult (18 years and older) were more likely to be heterozygous F508del (64.2%).

**FIGURE 15**  
Genotype distribution of individuals with cystic fibrosis, by diagnosis age group (N = 4,344), 2019.

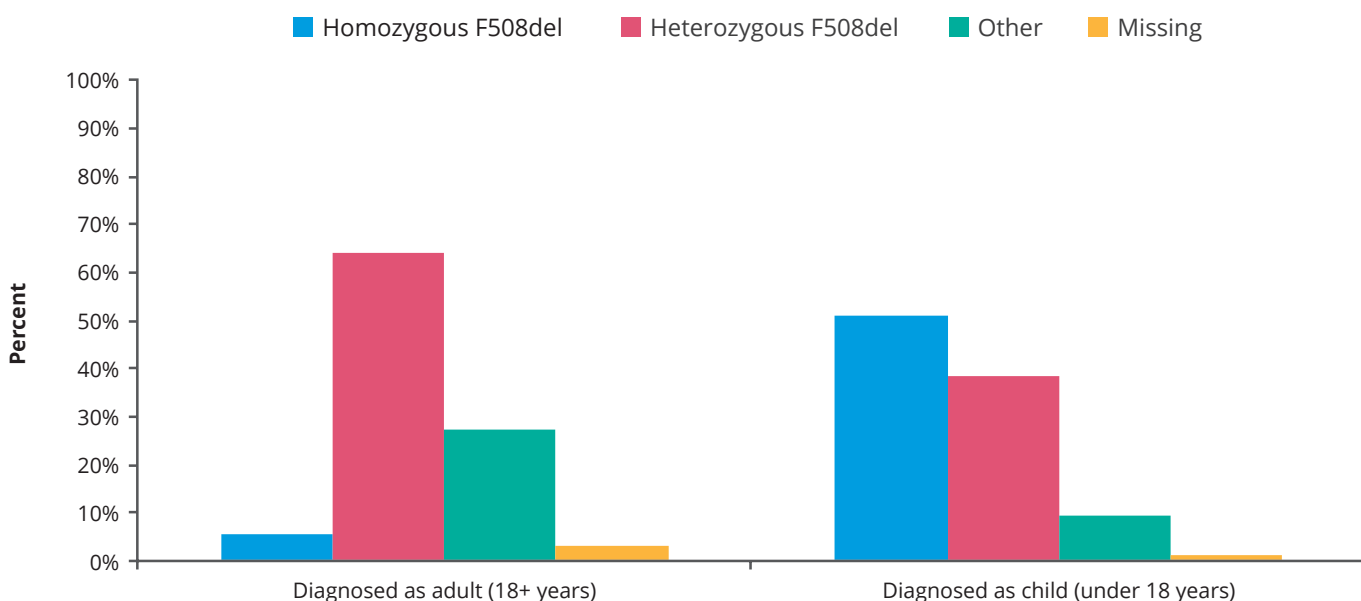


Table 2 lists the most common mutations for individuals with cystic fibrosis reported in 2019. After F508del, 621+1G->T is the next most frequent mutation identified in 5.8% of the population.

**TABLE 2**  
Frequency of the top 10 most common CF mutations on one or both alleles of cystic fibrosis individuals with recorded mutations (N = 4,289), 2019.

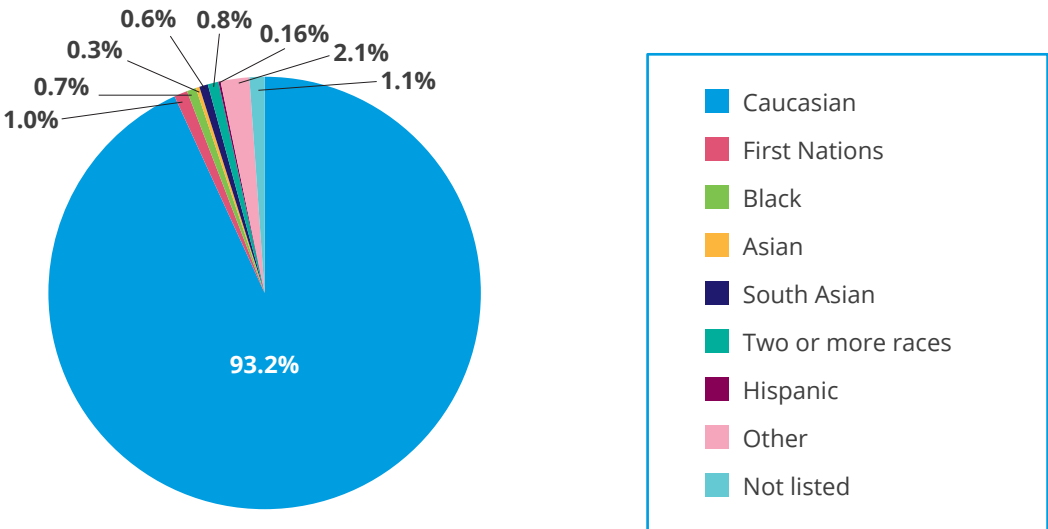
MUTATION	NUMBER	PERCENTAGE
F508del	3,812	88.9%
621+1G->T	250	5.8%
G542X	144	3.4%
G551D	140	3.3%
711+1G->T	116	2.7%
L206W	107	2.5%
A455E	106	2.5%
N1303K	89	2.1%
M1101K	70	1.6%
R117H	69	1.6%



# ETHNICITY

Cystic fibrosis can affect people of all ethnicities anywhere in the world. Caucasians account for the majority (93.2%) of the Canadian CF population. 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, 2019.



# RESPIRATORY

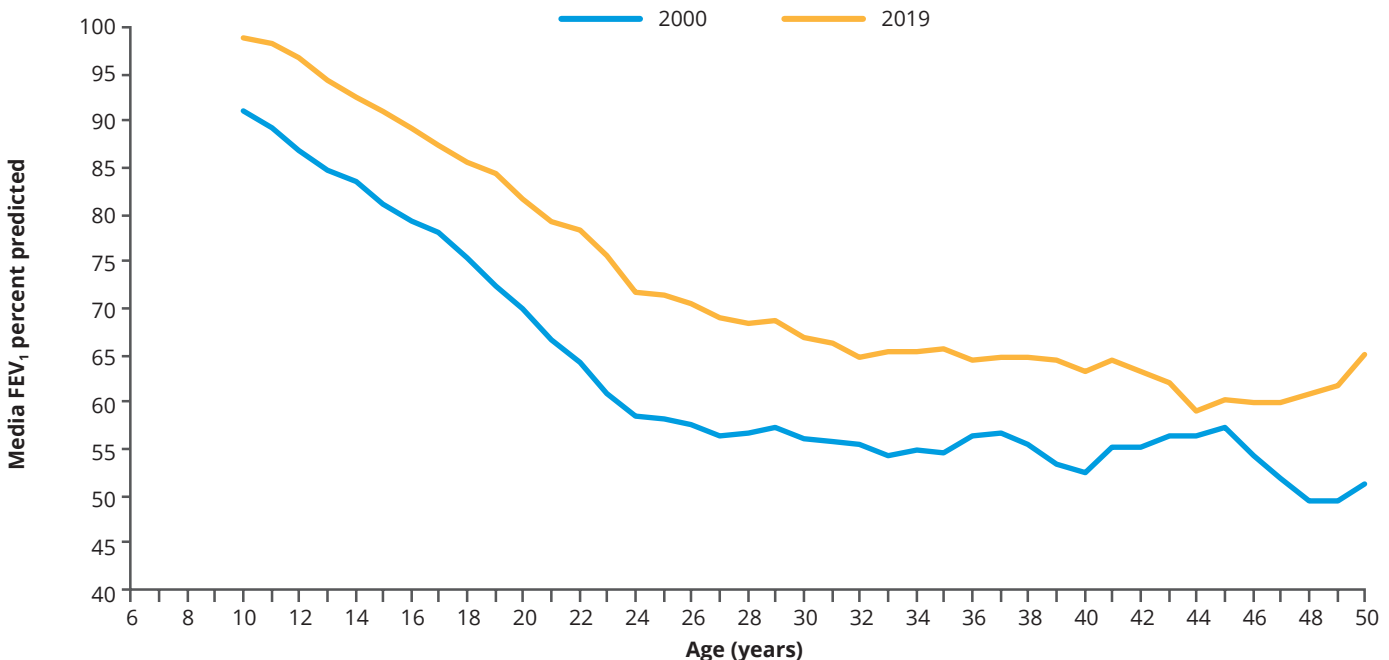
## MEDIAN FEV<sub>1</sub> PERCENT PREDICTED

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. FEV<sub>1</sub> (forced expiratory volume in one second) is the volume of air that a person can forcibly blow out in one second. FEV<sub>1</sub> percent predicted for an individual is calculated by comparing their FEV<sub>1</sub> to the average FEV<sub>1</sub> of a healthy population of similar age, height, ethnicity, and sex. Global Lung Initiative (GLI) equations<sup>1</sup> are used to calculate the percent predicted FEV<sub>1</sub> values.

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.

For people with cystic fibrosis, an FEV<sub>1</sub> % predicted of 90% or higher indicates normal lung function. Figure 17 shows the median FEV<sub>1</sub> 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, at a population level the median FEV<sub>1</sub> percent predicted has increased since 2000. The median FEV<sub>1</sub> at 24 years of age (the median age of an individual living with cystic fibrosis) was 64.0% predicted in 2019 compared to 53.6% predicted in 2000, marking an improvement of 10.4% over the last two decades.

**FIGURE 17**  
Median FEV<sub>1</sub> percent predicted vs. age of cystic fibrosis individuals (5-year moving average window), 2000 and 2019\*.

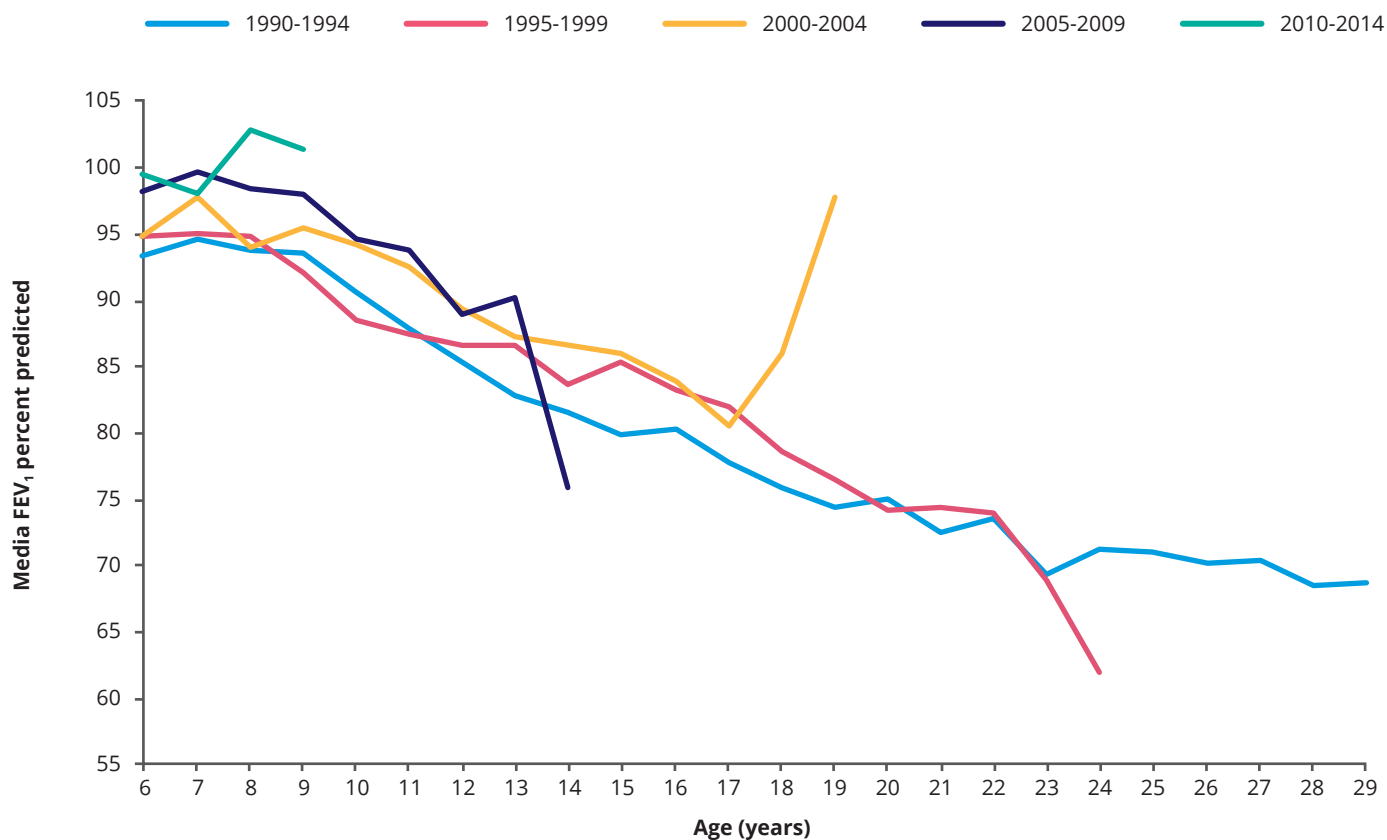


\* GLI reference equations used to calculate FEV<sub>1</sub> percent predicted values.

# MEDIAN FEV<sub>1</sub> PERCENT PREDICTED

Individuals born recently have a higher median FEV<sub>1</sub> percent predicted at age 6 years and have a slower rate of decline than those born earlier (Figure 18). The upticks seen 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 FEV<sub>1</sub> percent predicted of individuals with cystic fibrosis, by birth cohort, 2019\*.



\* GLI reference equations used to calculate FEV<sub>1</sub> percent predicted values.

# RESPIRATORY STATUS

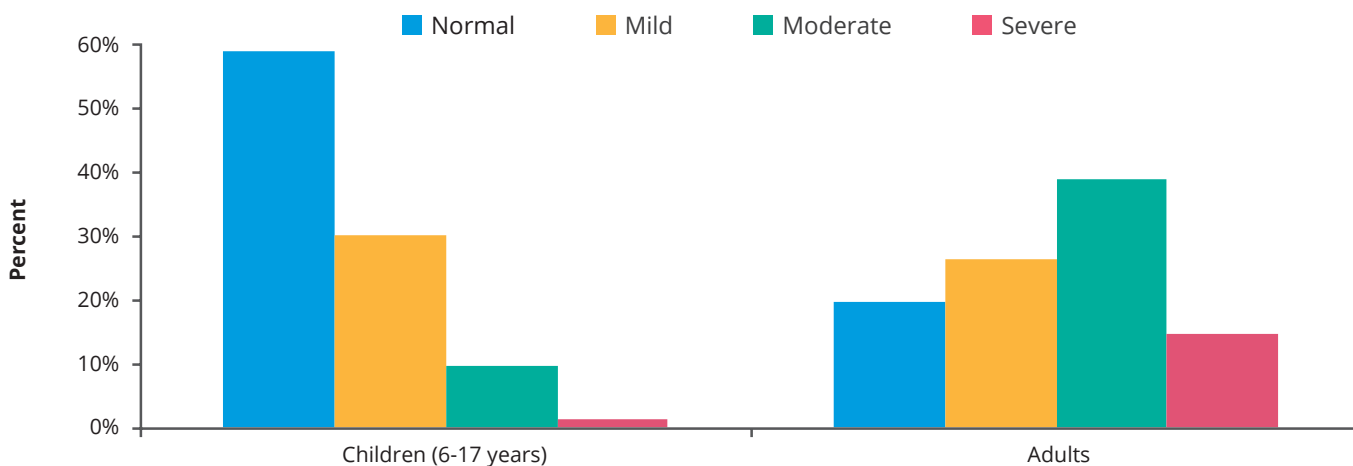
Table 3 defines severity of lung disease using FEV<sub>1</sub> percent predicted categories.

**TABLE 3**  
Lung function classification by FEV<sub>1</sub> percent predicted.

CLASSIFICATION	RANGE
Normal	≥ 90%
Mild	70 – 89%
Moderate	40 – 69%
Severe	< 40%

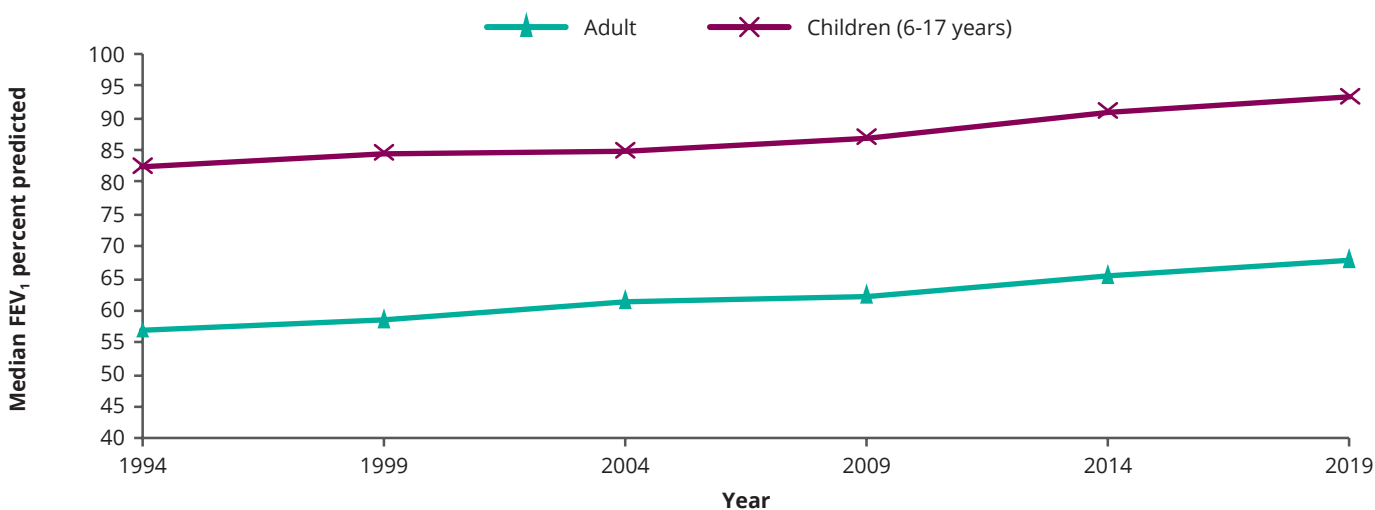
The majority (58.9%) of children, aged 6 to 17 years in 2019, have normal lung function (defined as ≥90% predicted) while only 19.9% of adults have normal lung function, as shown in Figure 19.

**FIGURE 19**  
Respiratory status of children and adults with cystic fibrosis, 2019.



Over time, the median FEV<sub>1</sub> percent predicted has been steadily increasing for both age groups, and in 2019 these values were 67.9% for adults and 93.4% for children (6-17 years of age), as shown in Figure 20.

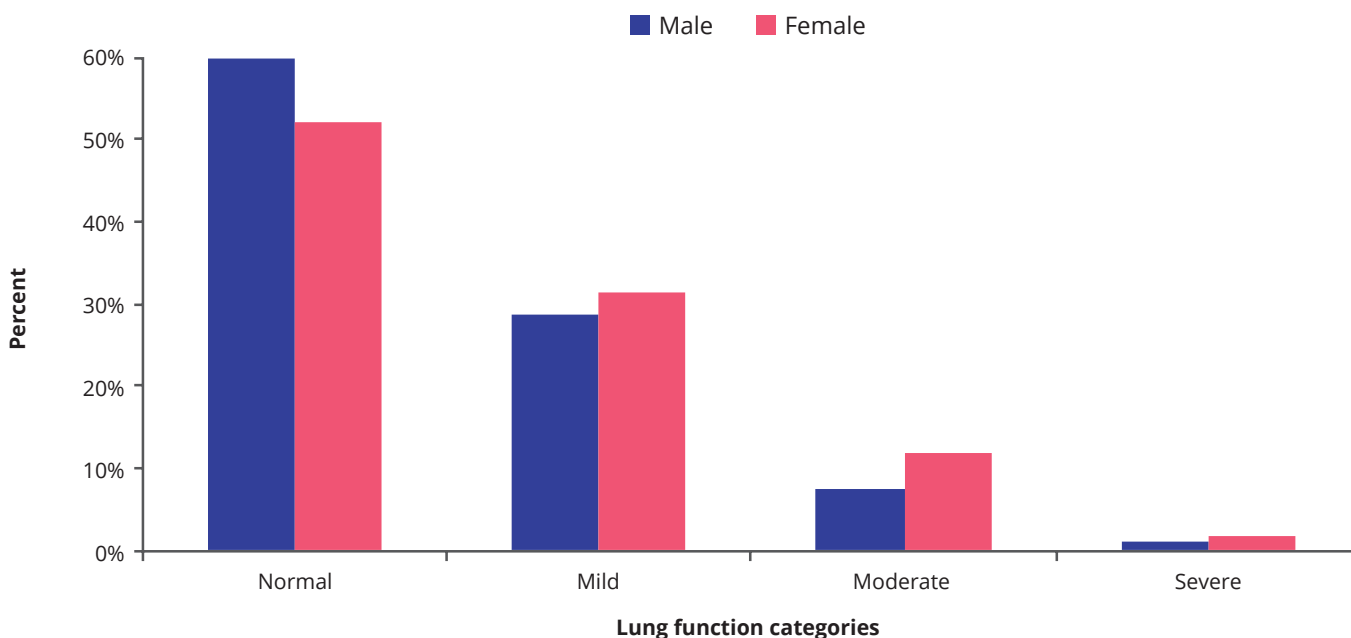
**FIGURE 20**  
Median FEV<sub>1</sub> percent predicted values for children and adults with cystic fibrosis, 1994 to 2019.



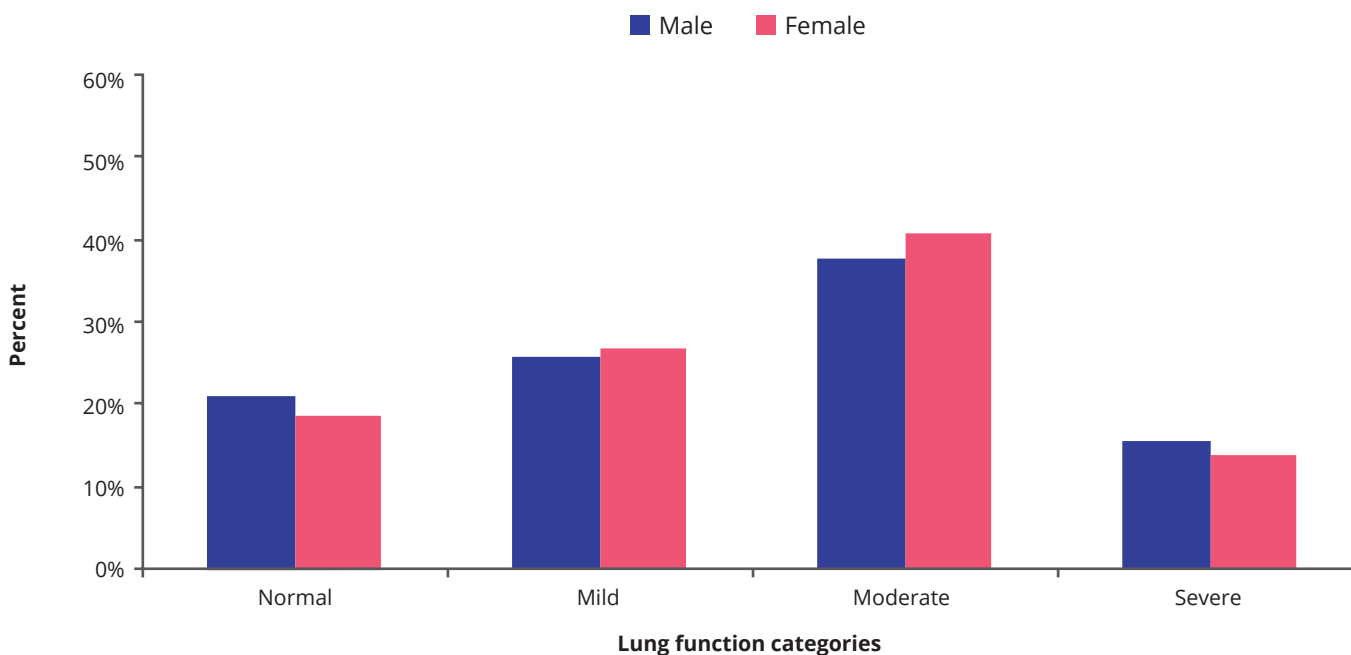
# RESPIRATORY STATUS BY SEX

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 across each category.

**FIGURE 21**  
Respiratory status of children (6 to 17 years) with cystic fibrosis, by sex, 2019.



**FIGURE 22**  
Respiratory status of adults (18 years of age and older) with cystic fibrosis, by sex, 2019.

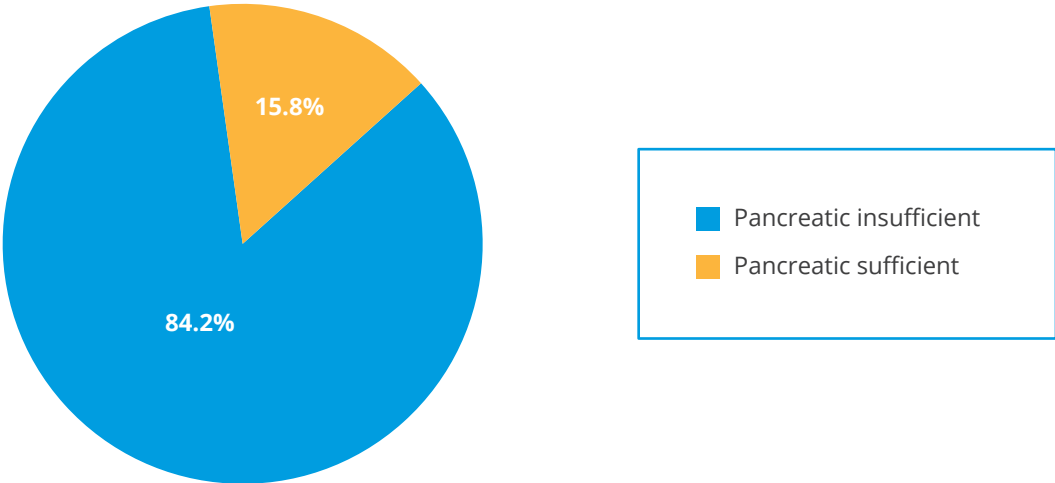


# NUTRITION

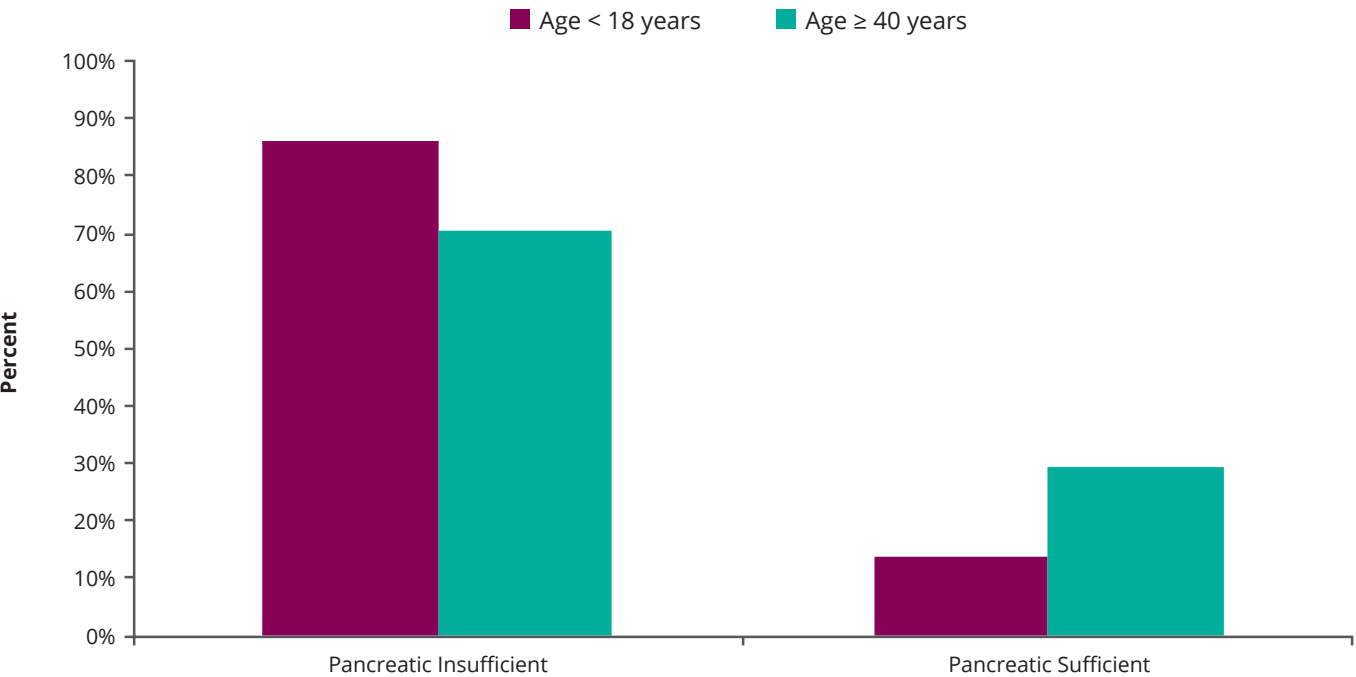
## PANCREATIC STATUS

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 2019, the majority (84.2%) of individuals with cystic fibrosis were taking supplemental pancreatic enzymes (and identified as pancreatic insufficient) compared to 15.8% who were not (identified as pancreatic sufficient), as shown in Figure 23. For individuals 40 years of age or older, only 29.5% were pancreatic sufficient (Figure 24).

**FIGURE 23**  
Pancreatic status of individuals with cystic fibrosis, 2019.



**FIGURE 24**  
Pancreatic status of individuals with cystic fibrosis, by age group, 2019.



# 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, this is only calculated for adults because they have attained their maximal height. Children are rapidly growing and therefore, one must consider the child's age when assessing their nutritional status, therefore using BMI percentiles are a more appropriate measure.

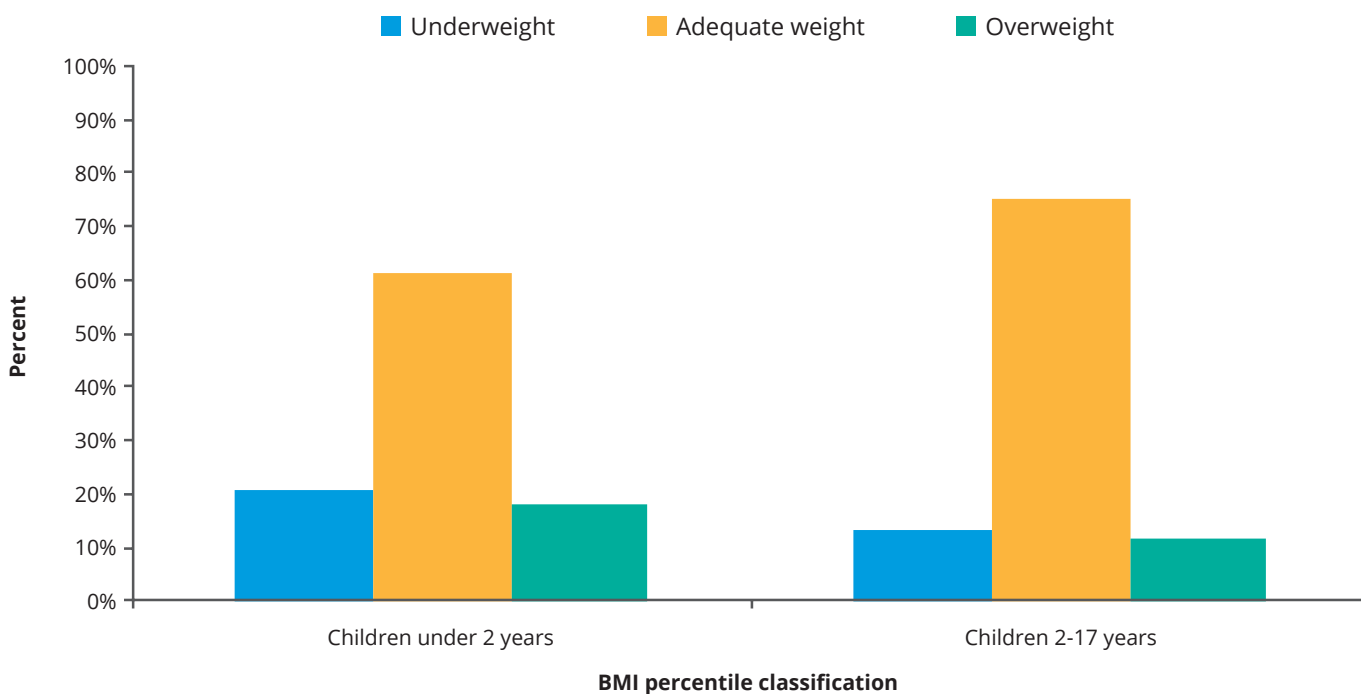
BMI percentiles<sup>2</sup> are calculated following the World Health Organization (WHO) guidelines for children under 2 years of age, and the Centers for Disease Control and Prevention (CDC) guidelines for children ages 2 to 17 years. BMI percentiles allow comparisons to be made between the individual's height and weight and other children who are the same age and sex. Table 4 details the BMI percentile classification categories following the respective WHO or CDC guidelines<sup>3</sup>.

**TABLE 4**  
BMI percentile classification.

CLASSIFICATION	RANGE
Underweight	≤ 12 <sup>th</sup> percentile
Adequate weight	13 <sup>th</sup> percentile - 84 <sup>th</sup> percentile
Overweight	≥ 85 <sup>th</sup> percentile

The national median BMI percentile for children under 2 and children between 2 and 17 years of age are both 46.0. Most children with cystic fibrosis (61.4% of children under 2 years and 75.2% of children 2-17 years) have an adequate weight (Figure 25). The 50<sup>th</sup> BMI percentile is the national goal for children with cystic fibrosis and in 2019, 47.4% of children under 2 years and 45.8% of children 2-17 years exceeded this goal.

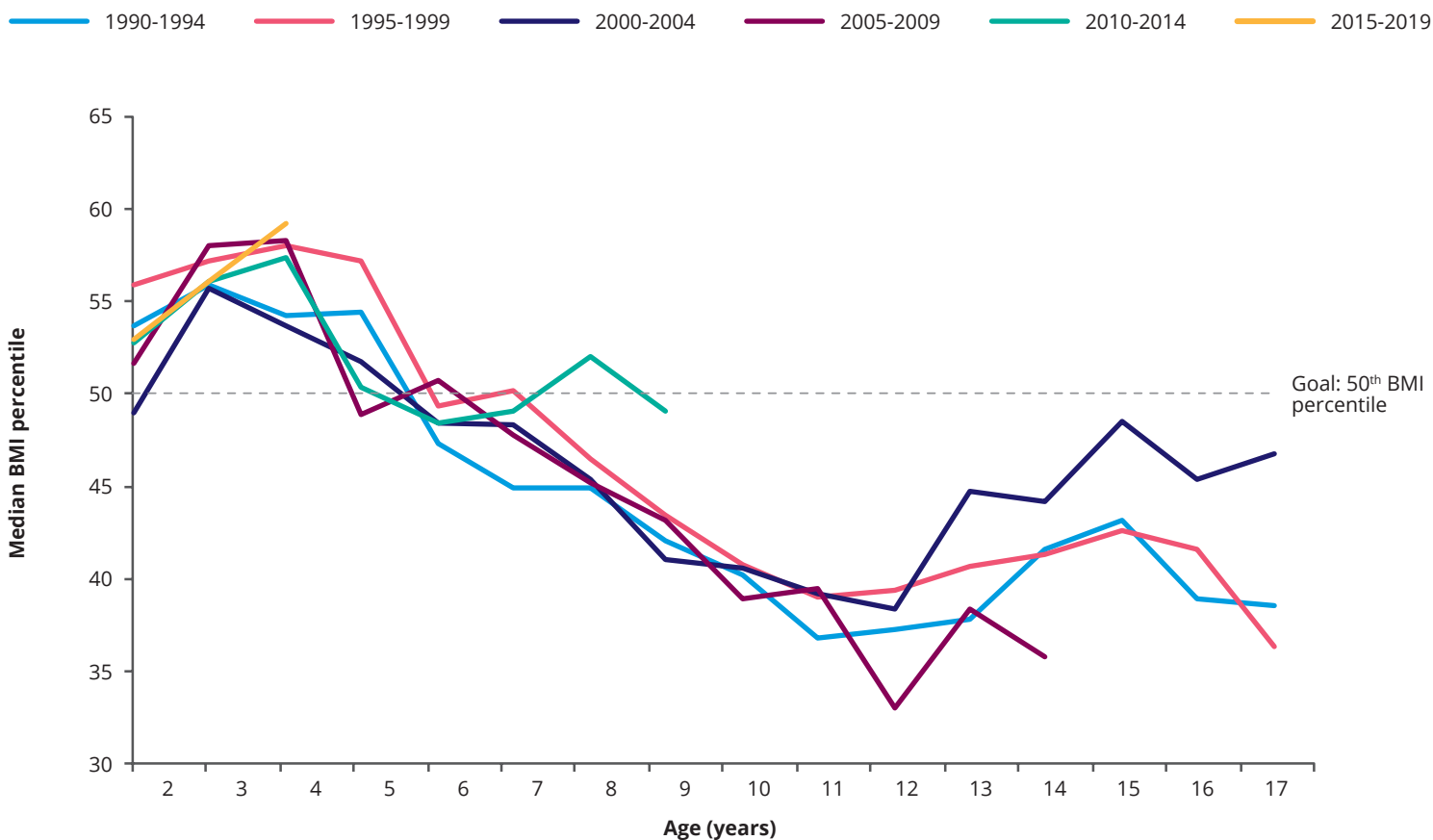
**FIGURE 25**  
BMI percentile status for children with cystic fibrosis, 2019.



# 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.

**FIGURE 26**  
Median BMI percentile for children (2-17 years) with cystic fibrosis, by birth cohort, 2019.

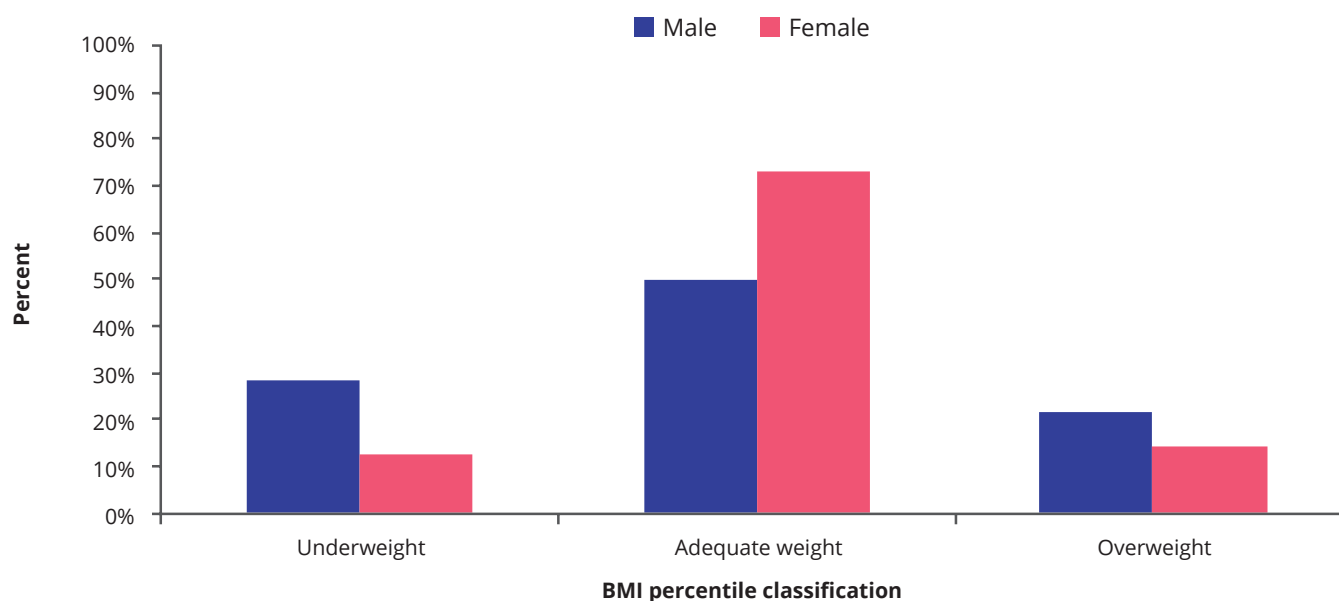




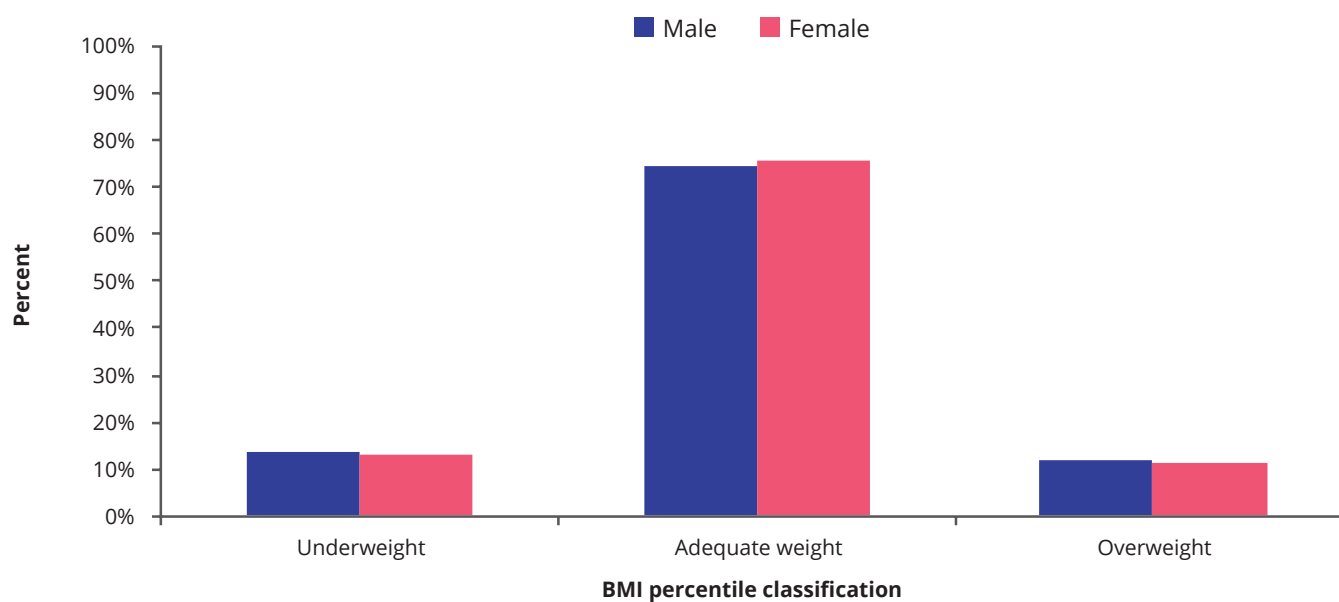
# BMI PERCENTILE BY SEX

Figure 27 and Figure 28 show the BMI percentile status for males and females in children under 2 years (N = 228) and children 2-17 years (N = 1,486).

**FIGURE 27**  
BMI percentile status for children (under 2 years) with cystic fibrosis, by sex, 2019.



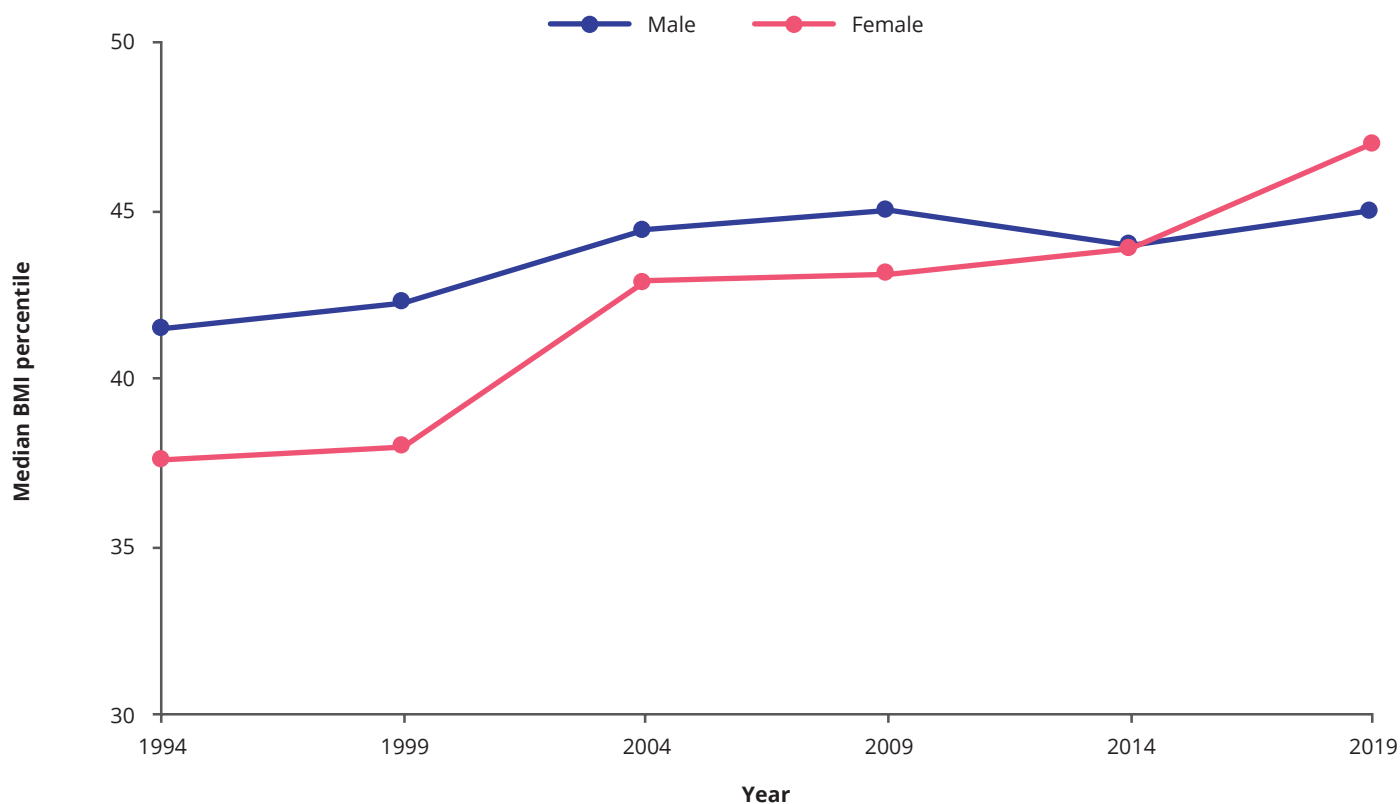
**FIGURE 28**  
BMI percentile status for children (2-17 years) with cystic fibrosis, by sex, 2019.



# BMI PERCENTILE BY SEX

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. By 2019, 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, 1994 to 2019.



# BMI

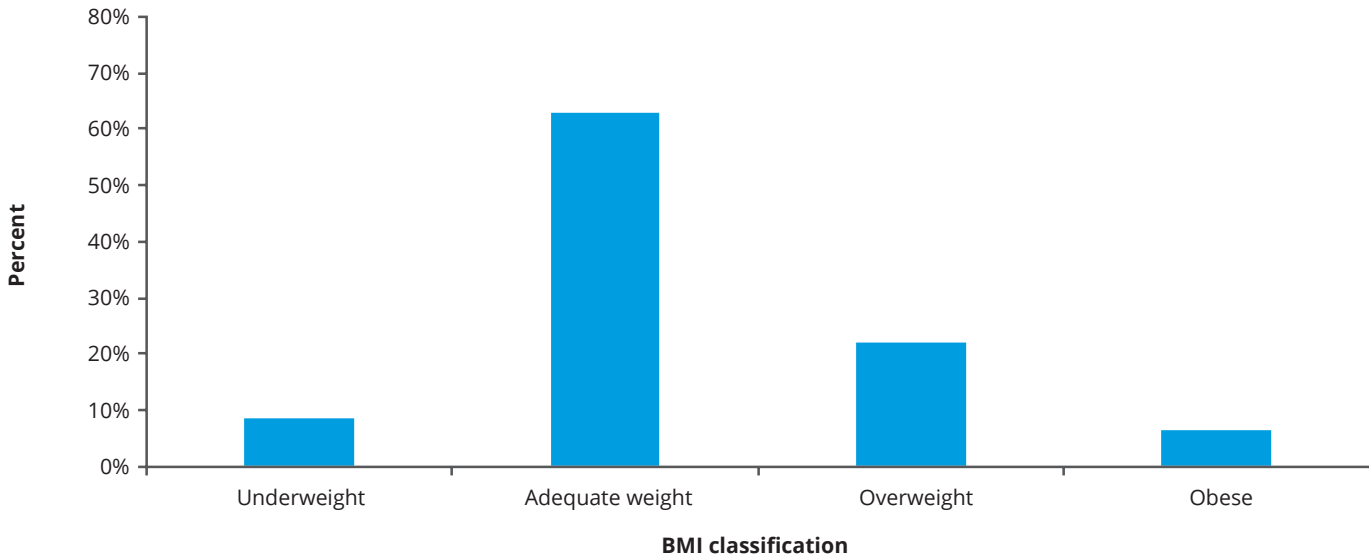
Table 5 below describes the BMI classifications and their BMI ranges according to the WHO guidelines<sup>4</sup>. These guidelines were updated in 2016 and as such, the proportions of BMI classifications will be different from those described in reports prior to 2016. Also note that “adequate weight” was referred to as “normal weight” in prior reports. In 2019, the national median BMI for adults (aged 18 and older) was 22.7 kg/m<sup>2</sup>.

**TABLE 5**  
**BMI classification**

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

The majority (62.9%) of the adult cystic fibrosis population had an adequate weight, while 8.8% were considered underweight and 6.4% were considered obese (Figure 30).

**FIGURE 30**  
**BMI status for adults with cystic fibrosis, 2019.**

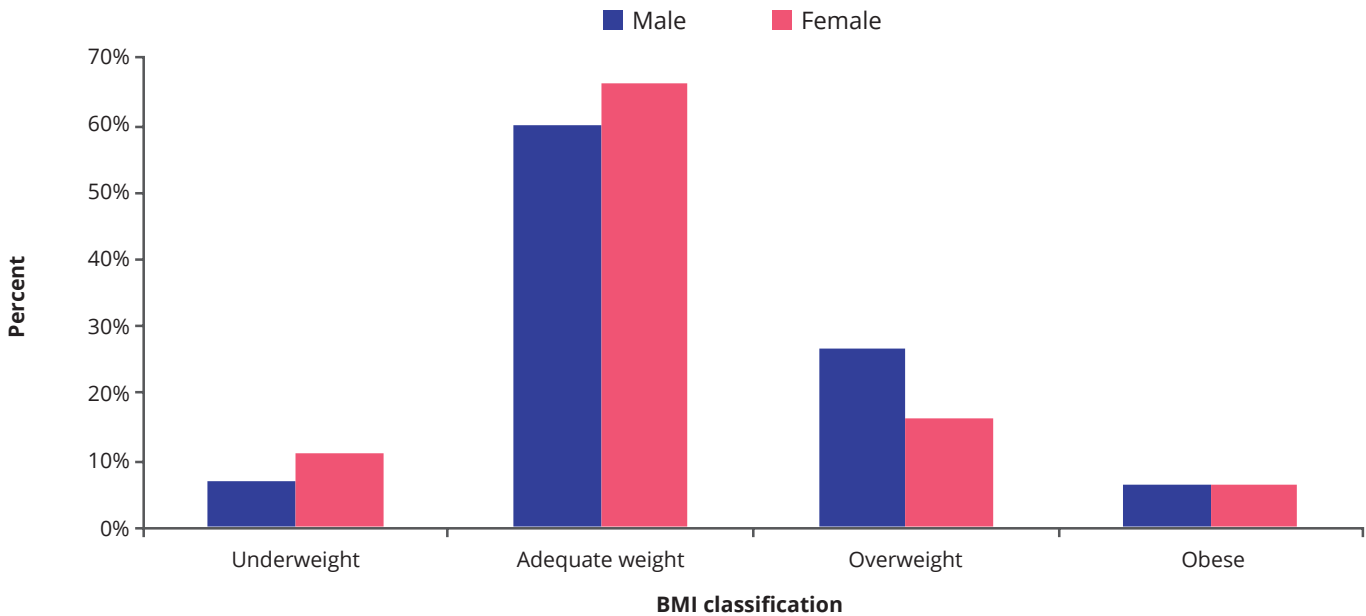


# BMI BY SEX

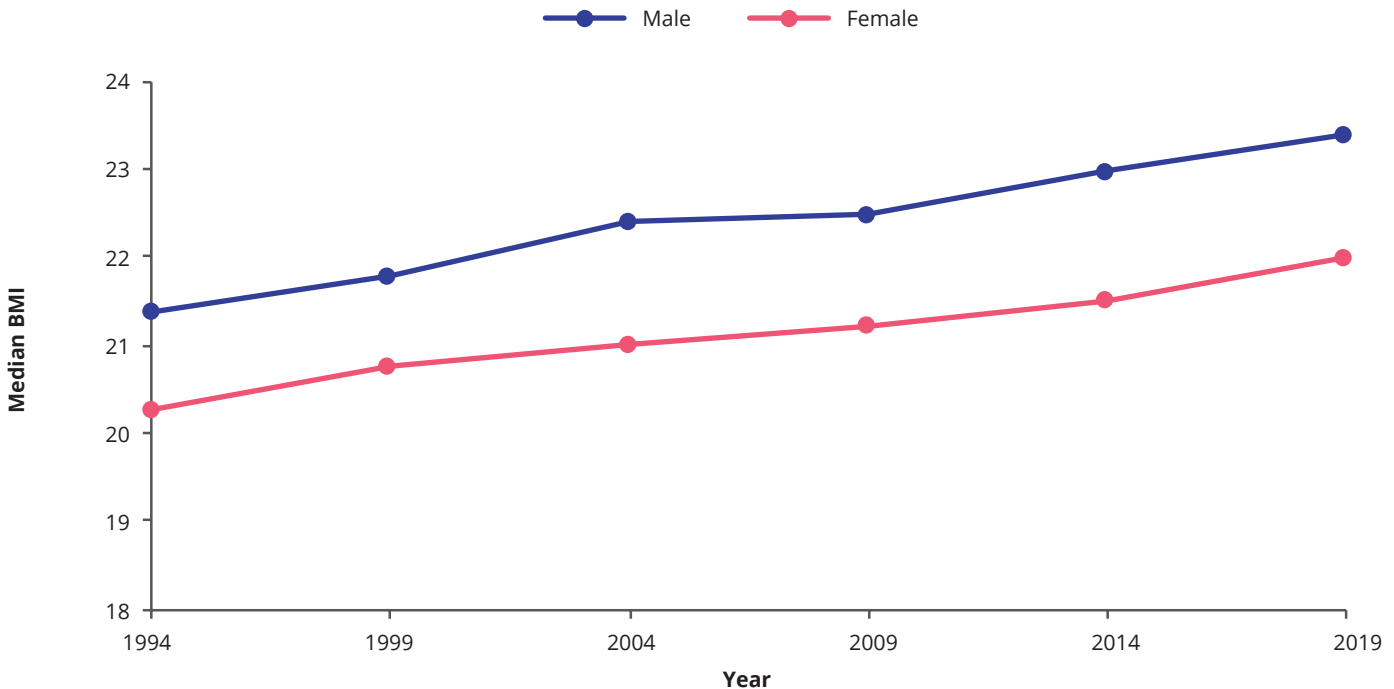
Figure 31 shows the breakdown of BMI categories for adult males and females. Individuals who are muscular may have a higher BMI due to increased weight from larger amounts of muscle mass.

In 2019, while more females (11.0%) were considered underweight compared to males (6.9%), 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, 2019.

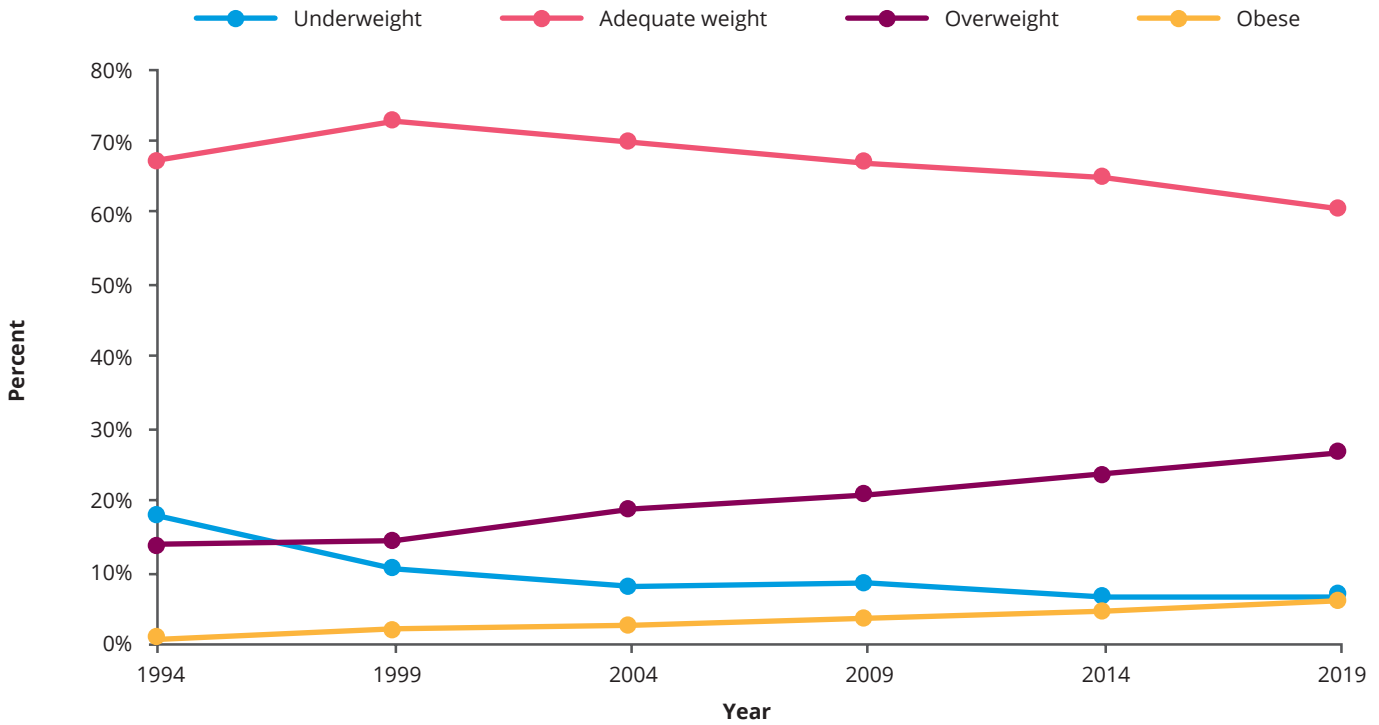


**FIGURE 32**  
Median BMI values for adults with cystic fibrosis, by sex, 1994 to 2019.

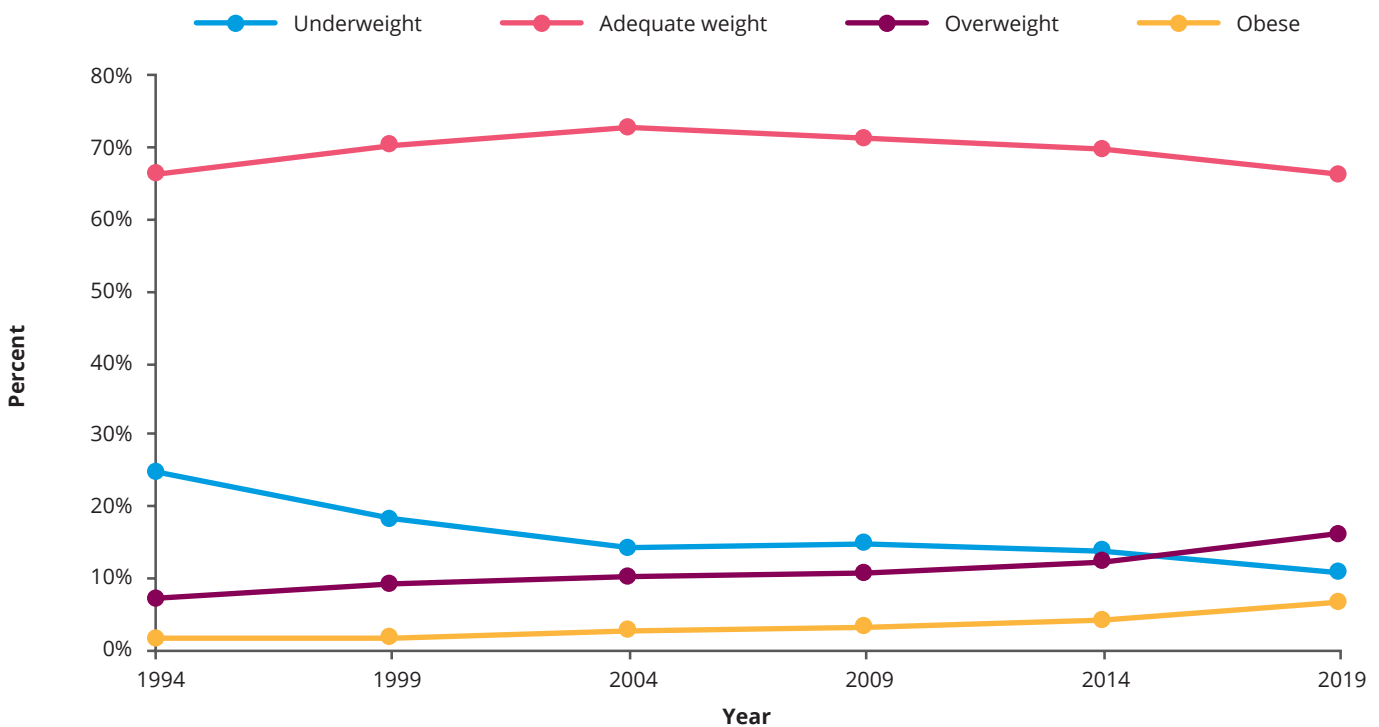


# BMI BY SEX

**FIGURE 33**  
Percentage of male adults with cystic fibrosis, by BMI status, 1994 to 2019.



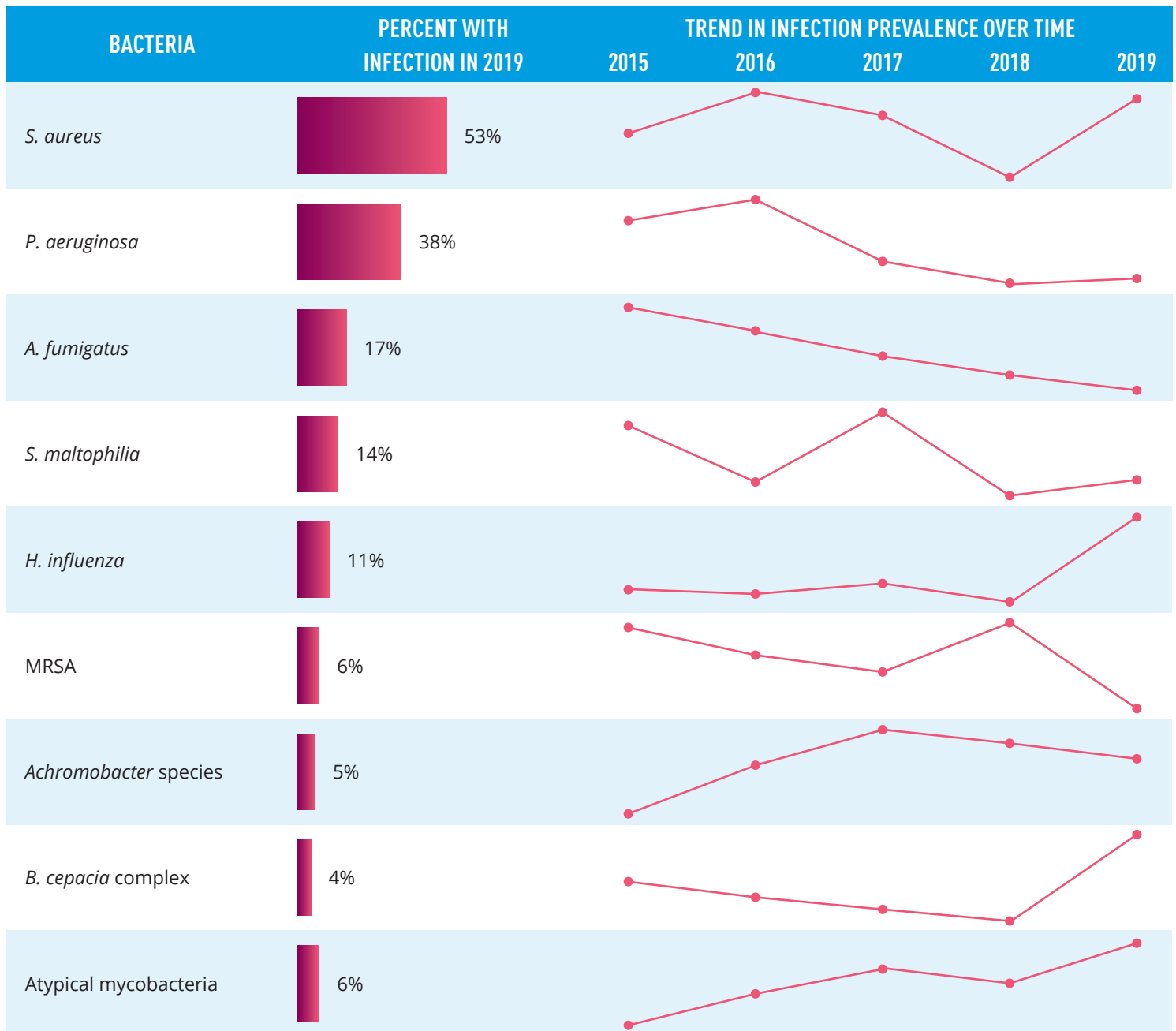
**FIGURE 34**  
Percentage of female adults with cystic fibrosis, by BMI status, 1994 to 2019.



## 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 52.9% and 38.3% of all Canadians with cystic fibrosis, respectively (Table 6). Over the past 5 years, the infection prevalence of the three most common bacteria (*S. aureus*, *P. aeruginosa*, and *A. fumigatus*) has steadily decreased. The CCFR 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).

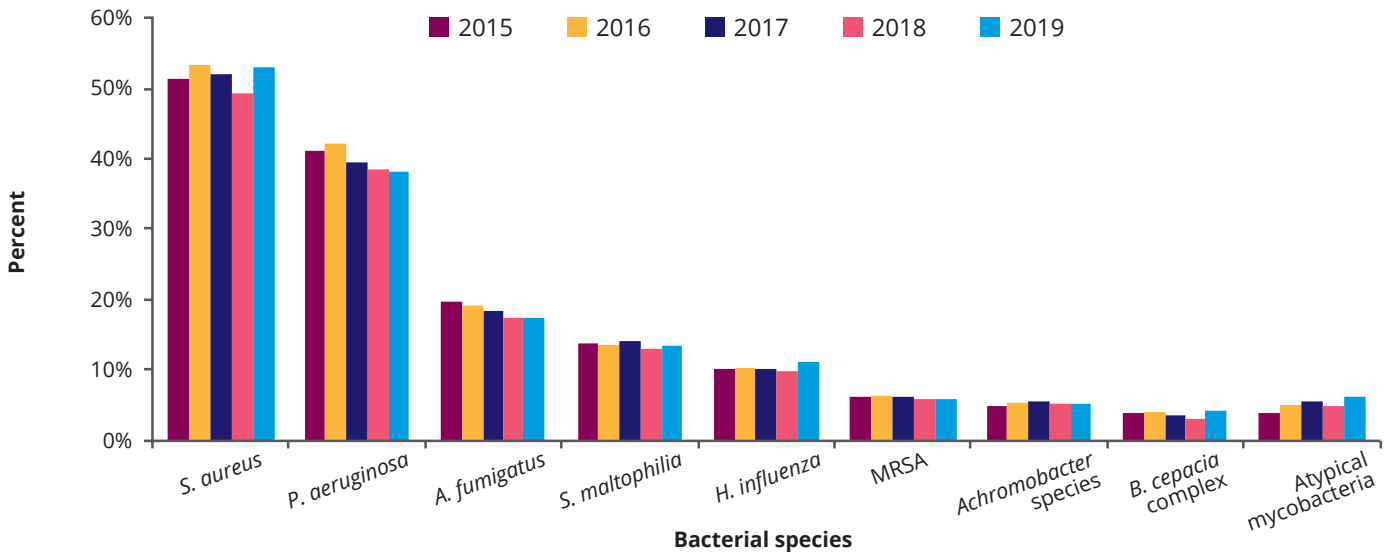
**TABLE 6**  
Prevalence of bacterial species cultured from airways of individuals with cystic fibrosis (all ages), 2019.



# BACTERIAL SPECIES AND RESPIRATORY INFECTIONS

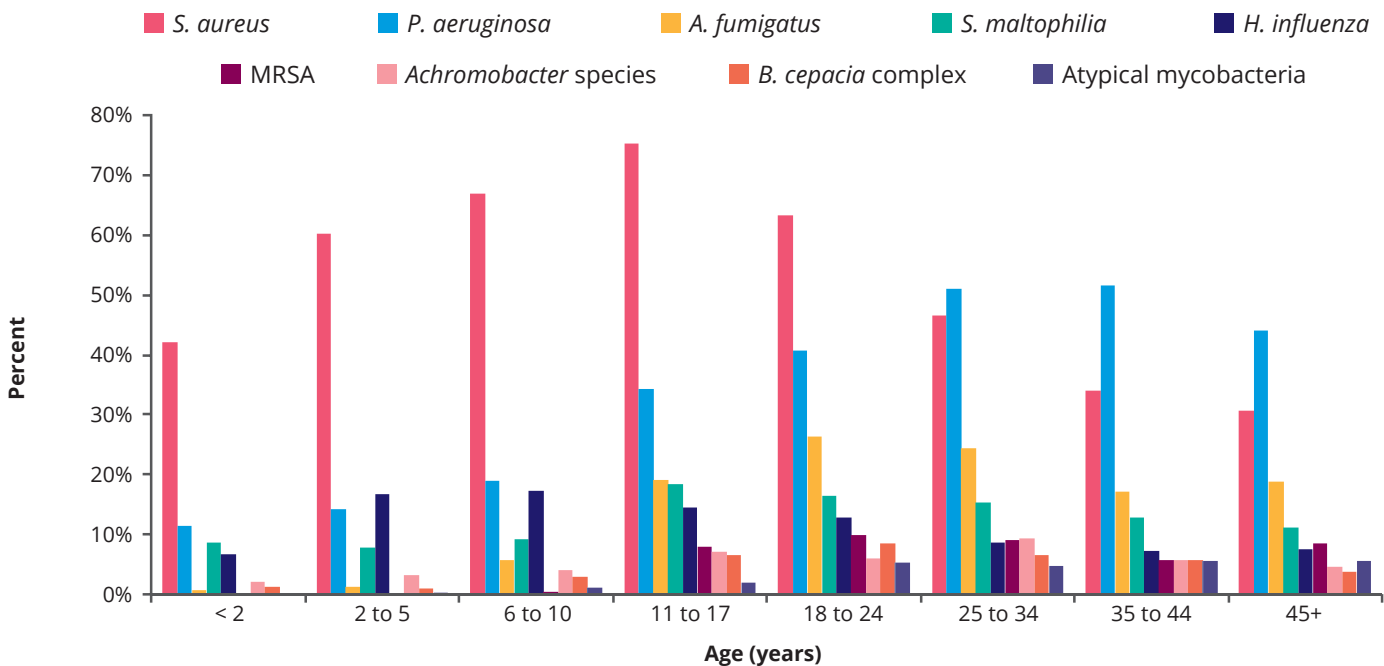
While decreasing, Figure 35 shows that over the past several years, *S. aureus* and *P. aeruginosa* remain the two most prevalent pulmonary pathogens among individuals with cystic fibrosis. There has also been a slight increase in the less frequently found pathogens such as *Achromobacter* species (formerly *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, 2015 to 2019.



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 (3.7%). 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, 2019.

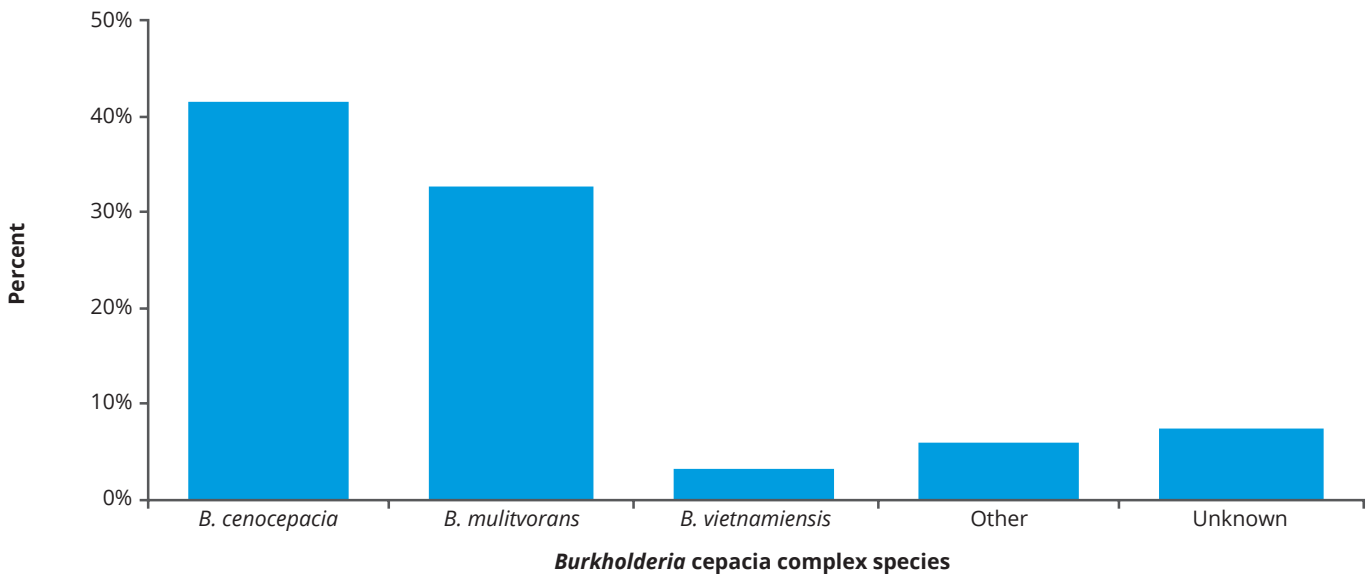


# BURKHOLDERIA CEPACIA COMPLEX (BCC)

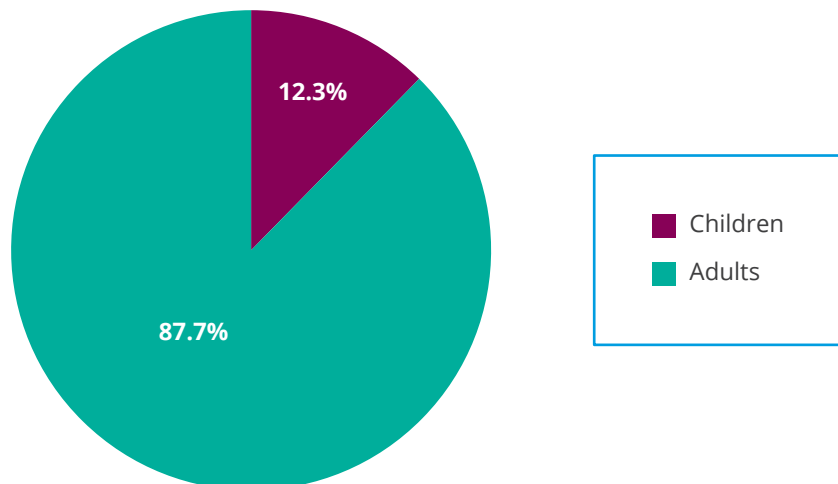
Out of all individuals with cystic fibrosis with bacterial species recorded in 2019, 162 (3.7%) unique individuals who grew at least one *Burkholderia cepacia* complex (BCC) species. The two most common types of BCC species are *B. cenocepacia* (41.5%) and *B. multivorans* (32.7%) (Figure 37). Of the unique individuals who had BCC in 2019, 142 (87.7%) were adults and 46 (28.4%) were over the age of 40 (Figure 38). Not all BCC bacteria have been speciated, as 7.4% of the BCC species in the CCFR 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 CCFR in 2011.

Note: The prevalence of *B. gladioli* was 9.2%, 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 = 162), 2019.



**FIGURE 38**  
*Burkholderia cepacia* complex distribution of cystic fibrosis individuals, by age, 2019.



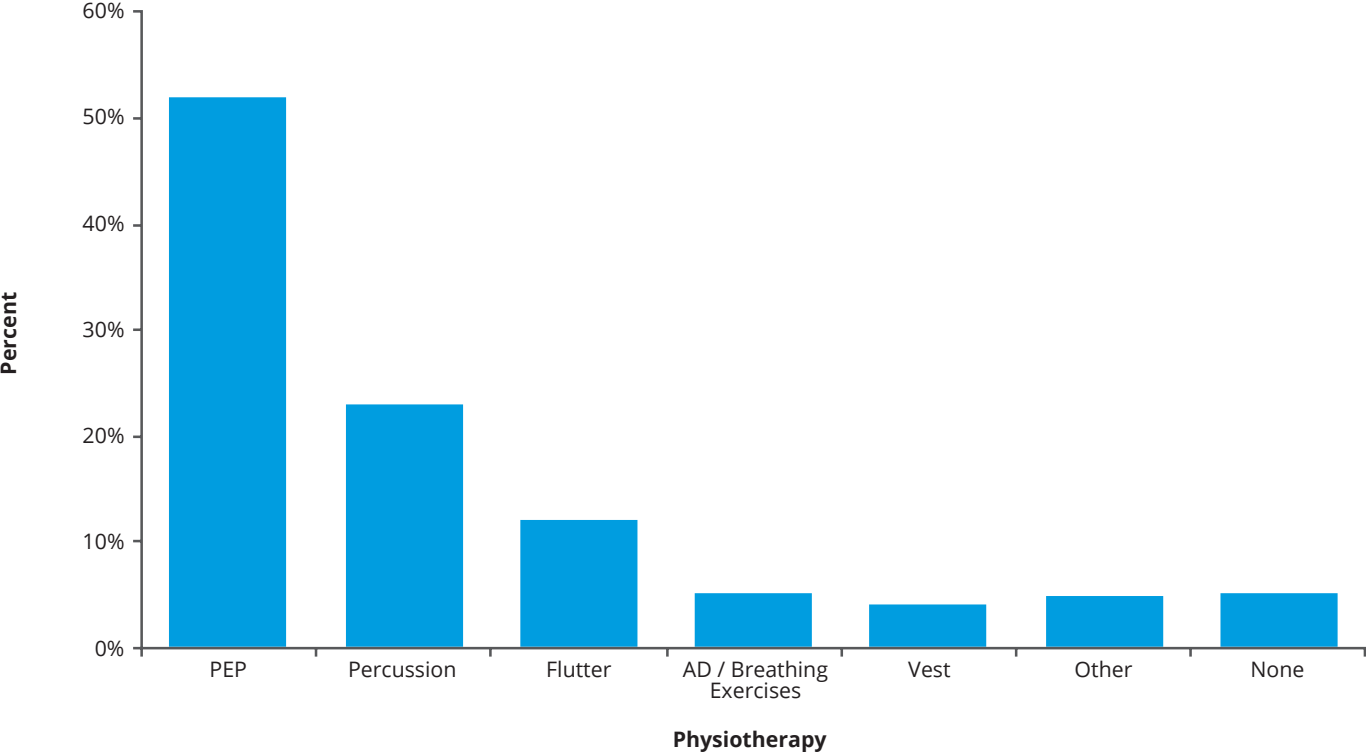


# PHYSIOTHERAPY

Physiotherapy is done to help clear mucus from airways using a variety of methods. Figure 39 shows the multiple forms of physiotherapy that are tracked in the CCFR. The most commonly used form of therapy are positive expiratory pressure (PEP) (51.9%) and percussion (22.9%), while 5.2% were reported as not doing any form of physiotherapy.

Note: Individuals who have ever received a lung transplant (7.8% of the 2019 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 = 4,006), 2019.

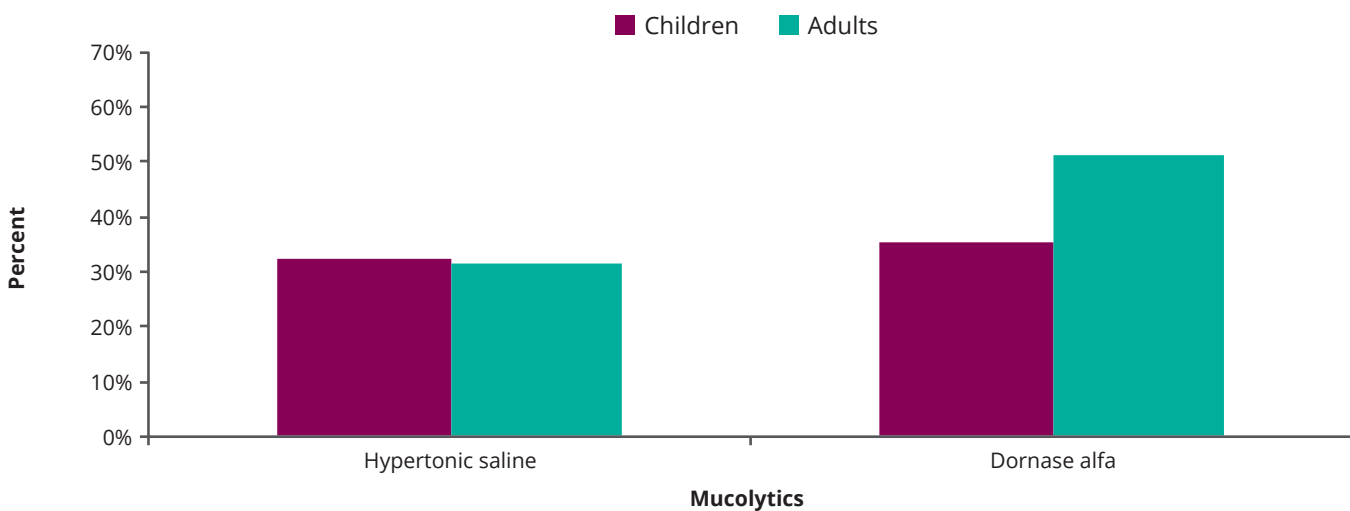


# MEDICATIONS

In 2019, there were a total of 4,006 individuals (1,640 children and 2,366 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.

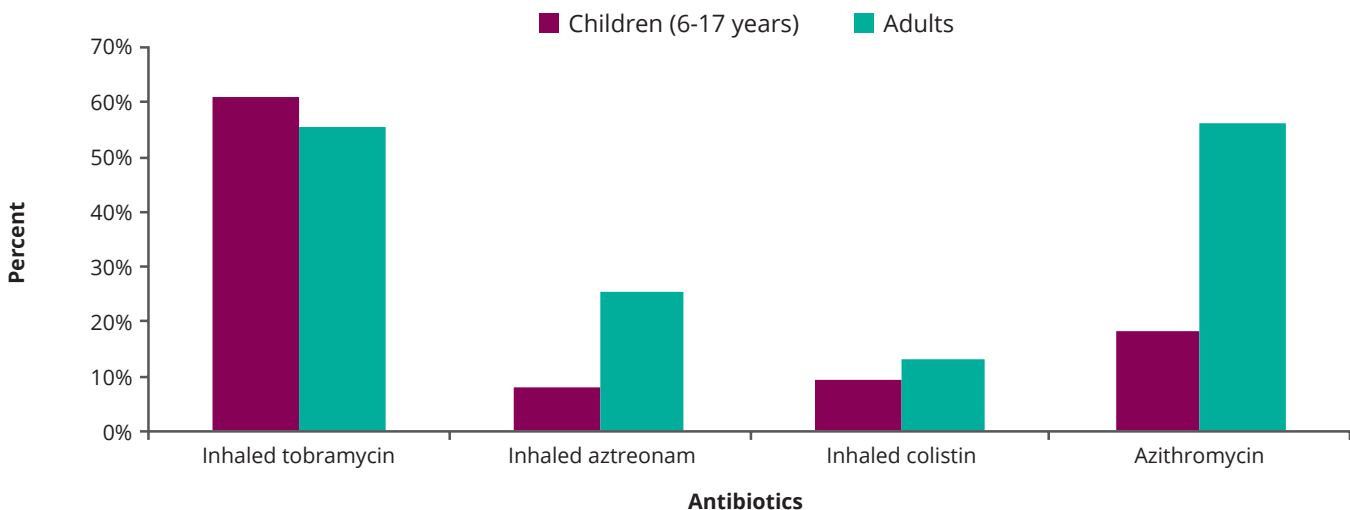
Figure 40 shows that of individuals who never received a transplant, 2,431 (60.7%) were prescribed mucolytic therapy during the calendar year (hypertonic saline and/or dornase alfa).

**FIGURE 40**  
Percentage of cystic fibrosis individuals on mucolytics, by age group, 2019.



There were 1,486 individuals over the age of 6 years who have never received a transplant and were reported to have *Pseudomonas aeruginosa* in 2019, which include 313 children (6-17 years) (21.1%) and 1,173 adults (78.9%). Of those, there were 191 children (61.0%) and 652 adults (55.6%) who were prescribed inhaled tobramycin treatment, and 57 children (18.2%) and 660 adults (56.3%) who were prescribed macrolide therapy (azithromycin) (Figure 41).

**FIGURE 41**  
Percentage of cystic fibrosis individuals on chronic antibiotics, by age group, 2019.



# CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE (CFTR) MODULATORS

Cystic fibrosis transmembrane conductance regulator (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 additional 9 mutations were 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. As of 2020, access to the triple combination therapy of elexacaftor/ivacaftor/tezacaftor remains limited to compassionate access programs.

In 2019, there were 658 unique individuals (216 children and 442 adults) on CFTR modulator therapies (Table 7).

**TABLE 7**  
**Number of cystic fibrosis individuals on CFTR modulators by age group, 2019.**

CFTR MODULATOR	DESCRIPTION	CHILDREN	ADULTS
ivacaftor	Indicated for the treatment of CF in patients 6 years of age and older who have the G551D mutation.	52	94
lumacaftor/ivacaftor	Indicated for the treatment of CF in patients 2 years of age and older who are homozygous for the F508del mutation in the CFTR gene.	132	236
ivacaftor/tezacaftor	Indicated for the treatment of CF in people who have a particular set of genetic mutations.	38	148
elexacaftor/ivacaftor/tezacaftor	Has not been submitted for approval in Canada.	N/A	N/A

# HEALTHCARE ENCOUNTERS

A total of 4,316 (99.4%) individuals with cystic fibrosis visited a CF clinic (had a recorded clinic visit date and/or clinical measurement) at least once in 2019 with 3,367 (77.5%) having three or more clinic visits. These clinic visits can include telemedicine appointments, during which patients receive medical education, or health advice and information via telecommunication technologies. Of the people having three or more clinic visits, 1,525 were children and 1,842 were adults, making up 92.6% and 68.3% of all children and adults, respectively. In 2019, there were a total of 18,960 clinic visits (Table 8).

The CCFR captures the date of admission and discharge from hospital. From these data, the total number of hospitalizations per patient are calculated. In 2019, there were 1,209 (27.8%) individuals with cystic fibrosis who altogether spent 25,246 days in hospital from a total of 1,952 recorded hospitalizations, which do not include visits to the out-patient CF clinics.

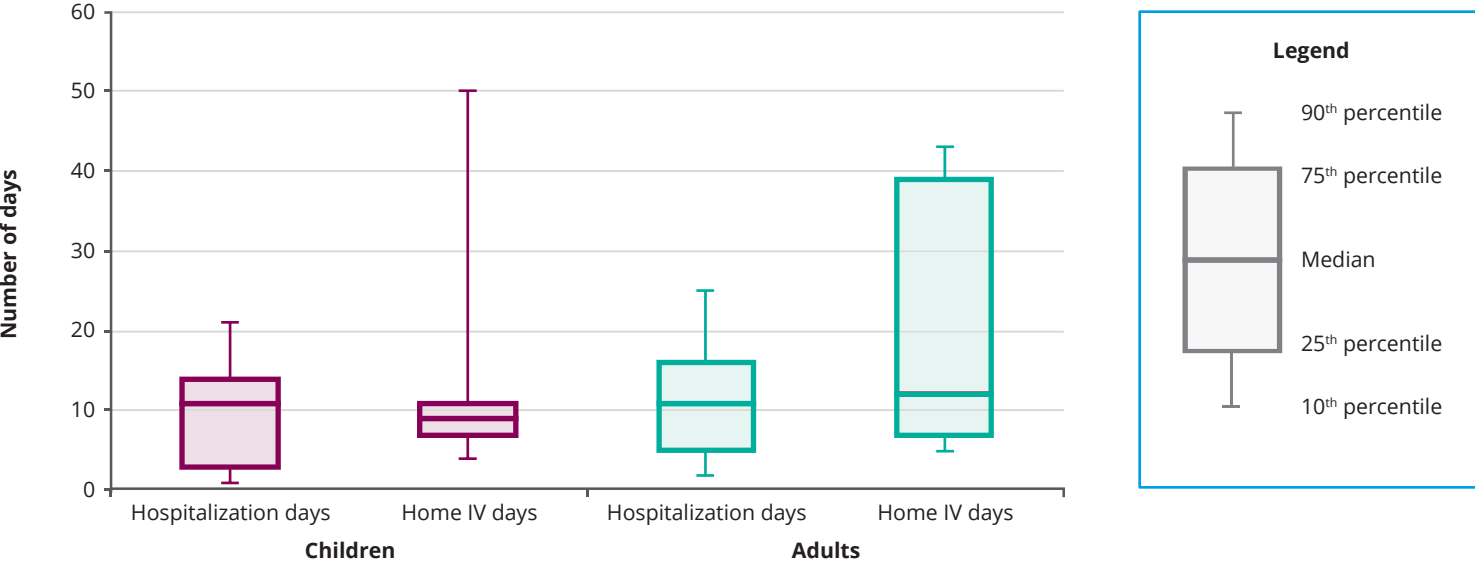
At home, individuals with cystic fibrosis had 15,530 days on IV antibiotics from a total of 842 courses.

**TABLE 8**  
**Total number of healthcare encounters recorded for individuals with cystic fibrosis, 2019.**

	TOTAL NUMBER
Clinic Visits	18,960
Hospitalizations	1,952
Hospital Days	25,246
Home IV Courses	842
Home IV Days	15,530

Figure 42 shows the distribution of hospitalizations and home IV courses by age group. In 2019, the distribution of hospital days is relatively similar between age groups, where children were hospitalized for an average of 11 days and adults for 14 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. 12 days), a quarter of all adults spent over 40 days on home IV in 2019.

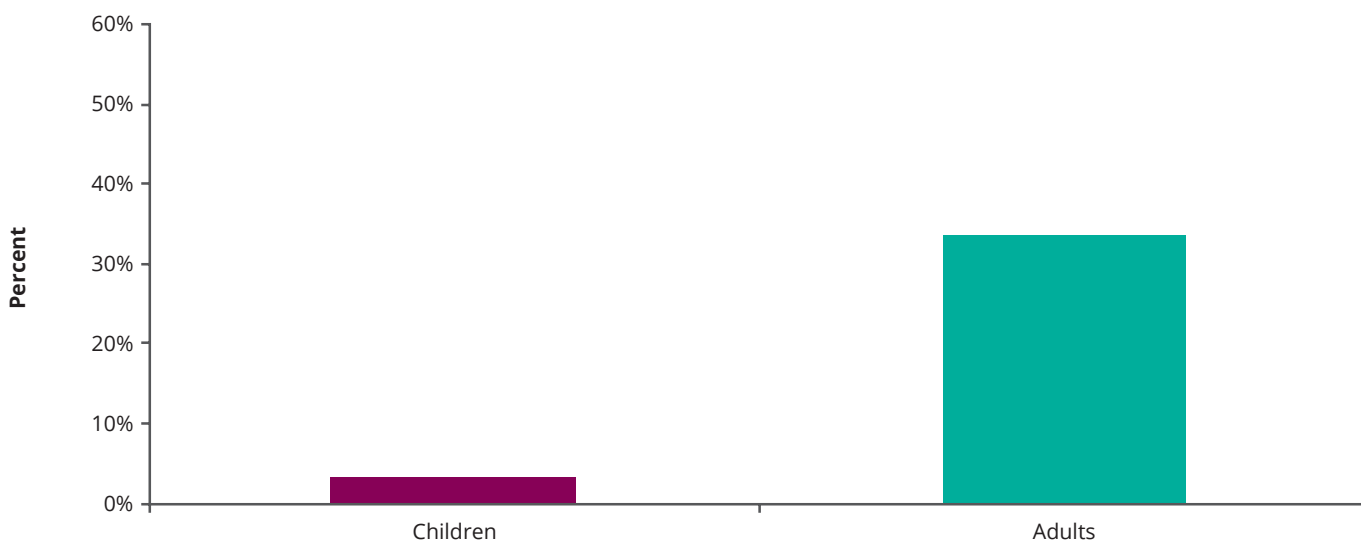
**FIGURE 42**  
**Distribution of hospitalizations and home IV courses, by age group, 2019.**



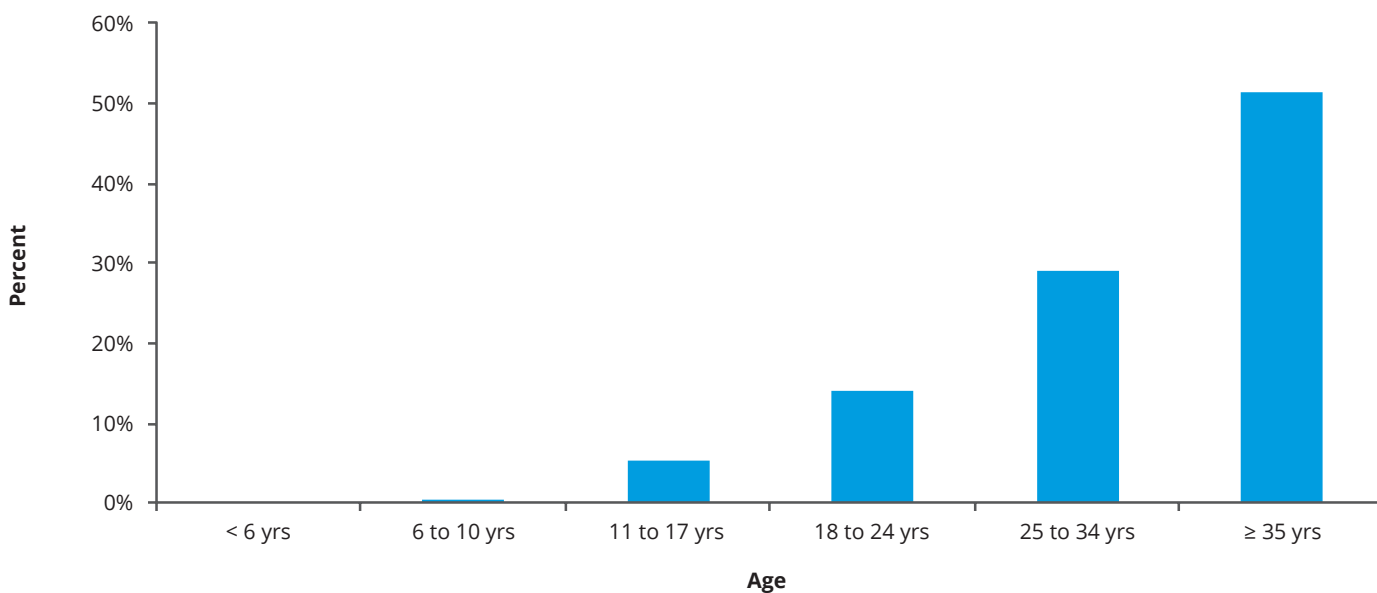
# CYSTIC FIBROSIS-RELATED DIABETES (CFRD)

Cystic fibrosis-related diabetes (CFRD) is a unique type of diabetes common in individuals living with cystic fibrosis. CFRD is often associated with weight-loss and lung function decline, but with early diagnosis and proper treatment, CFRD can be managed successfully. In 2019, CFRD was reported in 957 (22.0%) individuals with cystic fibrosis, affecting 54 (3.3%) children and 903 (33.5%) adults (Figure 43). Of those individuals with CFRD, 49.8% were female, 23.7% have received a transplant, and 51.2% 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, 2019.



**FIGURE 44**  
Percentage of cystic fibrosis individuals with CFRD by age, 2019.

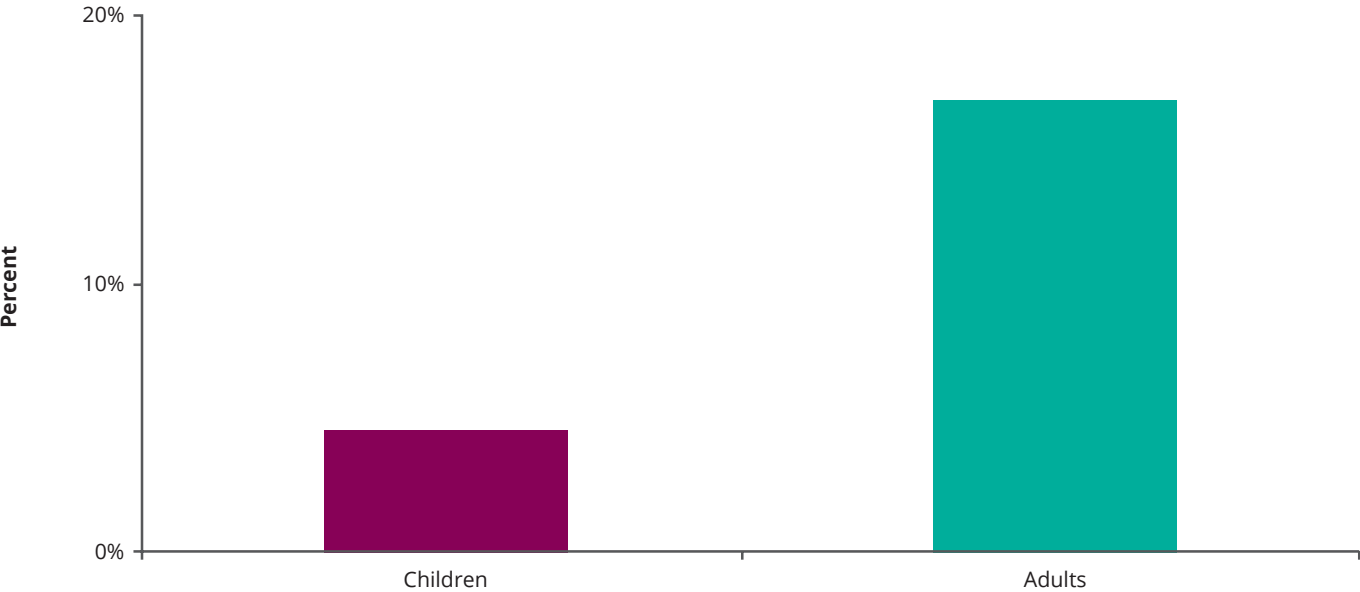


# MENTAL HEALTH

In 2019, there were 528 individuals with cystic fibrosis (12.2% of all individuals) with reported clinically diagnosed depression or anxiety. 75 of these diagnoses were children and 453 were adults, representing 4.6% of all children and 16.8% 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)<sup>5,6</sup> which showed elevated rates of depression and anxiety among individuals with cystic fibrosis and their parents/caregivers.

**FIGURE 45**  
Percentage of children and adults diagnosed with depression or anxiety, 2019.

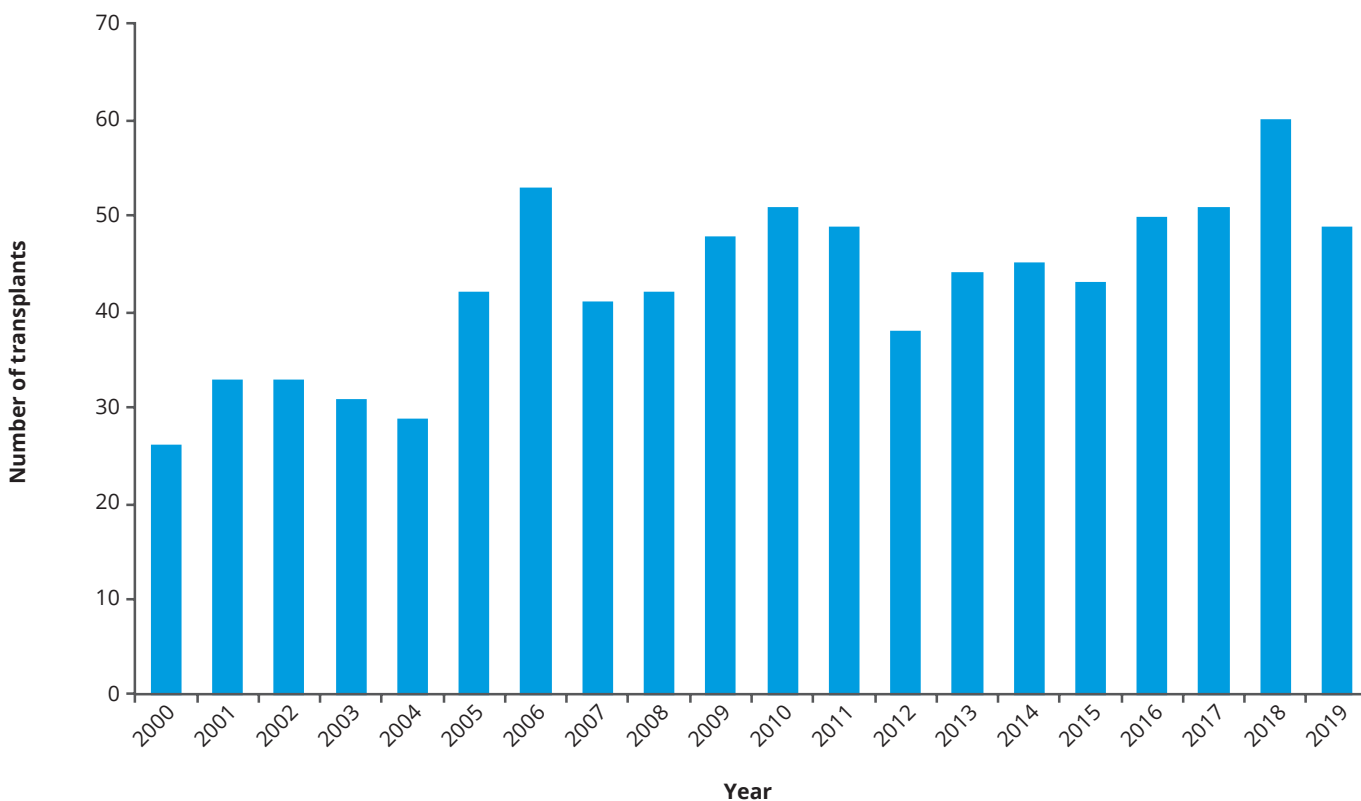


# TRANSPLANTS

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 CCFR. In 2019, 49 individuals with cystic fibrosis received a transplant with a median age at the time of transplant of 33.2 years. Although the numbers provided represent primarily lung transplants (46 lung transplants in 2019), 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 CCFR was performed in 1988. As of December 31, 2019, there were 916 unique individuals with cystic fibrosis reported as having received one or more transplants (any organ) with a median age at the time of transplant of 28.7 years. Of these patients, 60 (6.6%) have received at least two lung transplants, 517 (56.4%) were reported as being alive, and 284 (54.9%) of those living patients were male.

**FIGURE 46**  
Number of transplants per year of cystic fibrosis individuals, 2000 to 2019.



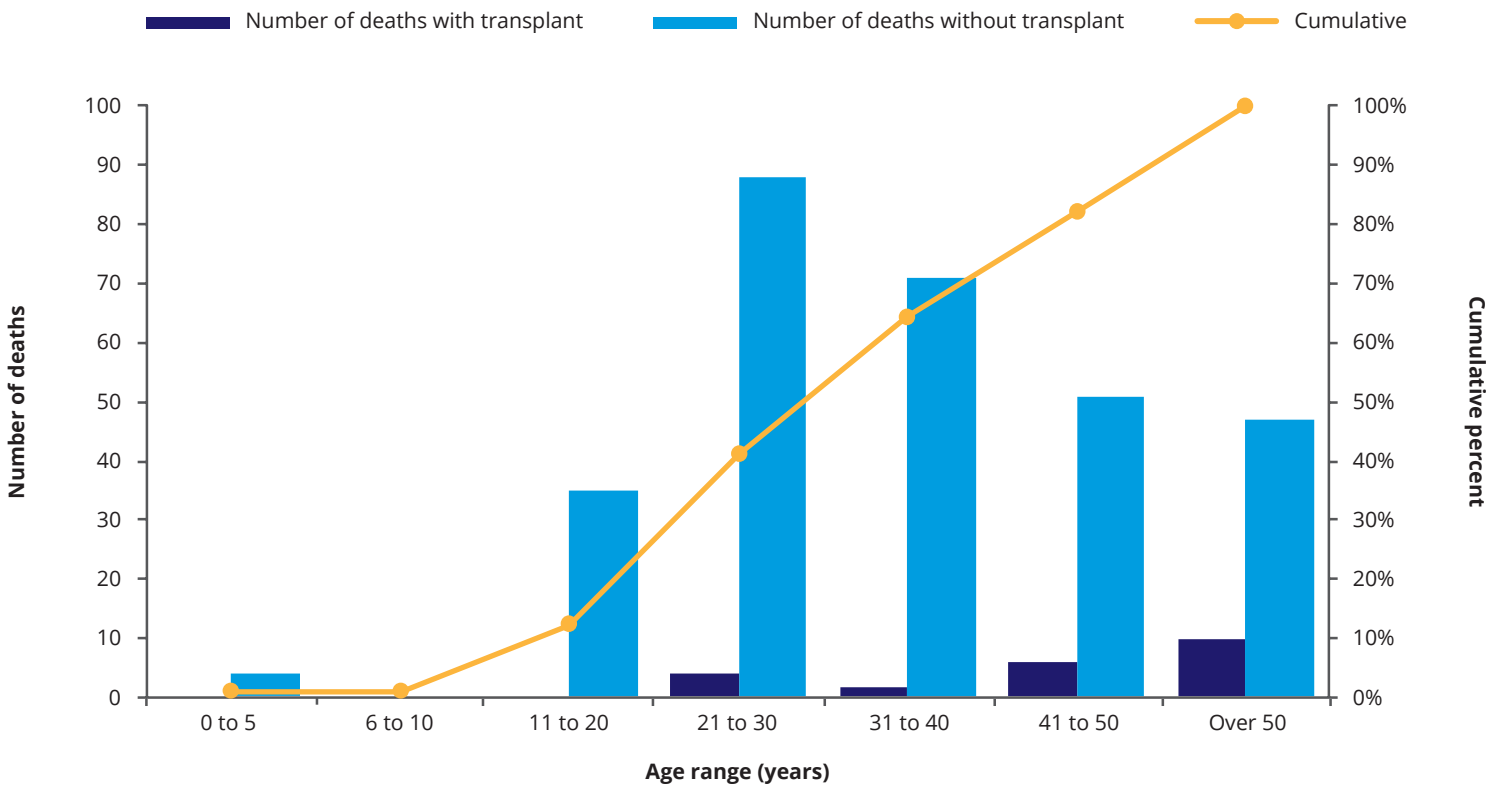
# SURVIVAL

The survival and health outcomes in Canadians living with cystic fibrosis continues to improve over time. In 2019, there were 49 deaths recorded in the CCFR, compared to 53 deaths in 2018. Figure 47 shows the cumulative number of deaths and the age at death from 2015 to 2019.

Risk factors such as pulmonary exacerbations and malnutrition are often associated with increased risk of death. In 2019, of the deaths with a recorded cause of death (41 deaths; 83.7%), 80.5% of deaths were reported as pulmonary/infection/cardiovascular complications.

29 (59.2%) individuals with cystic fibrosis who died in 2019 had never received a transplant (any organ).

**FIGURE 47**  
Cumulative number of deaths and age at death, 2015 to 2019.

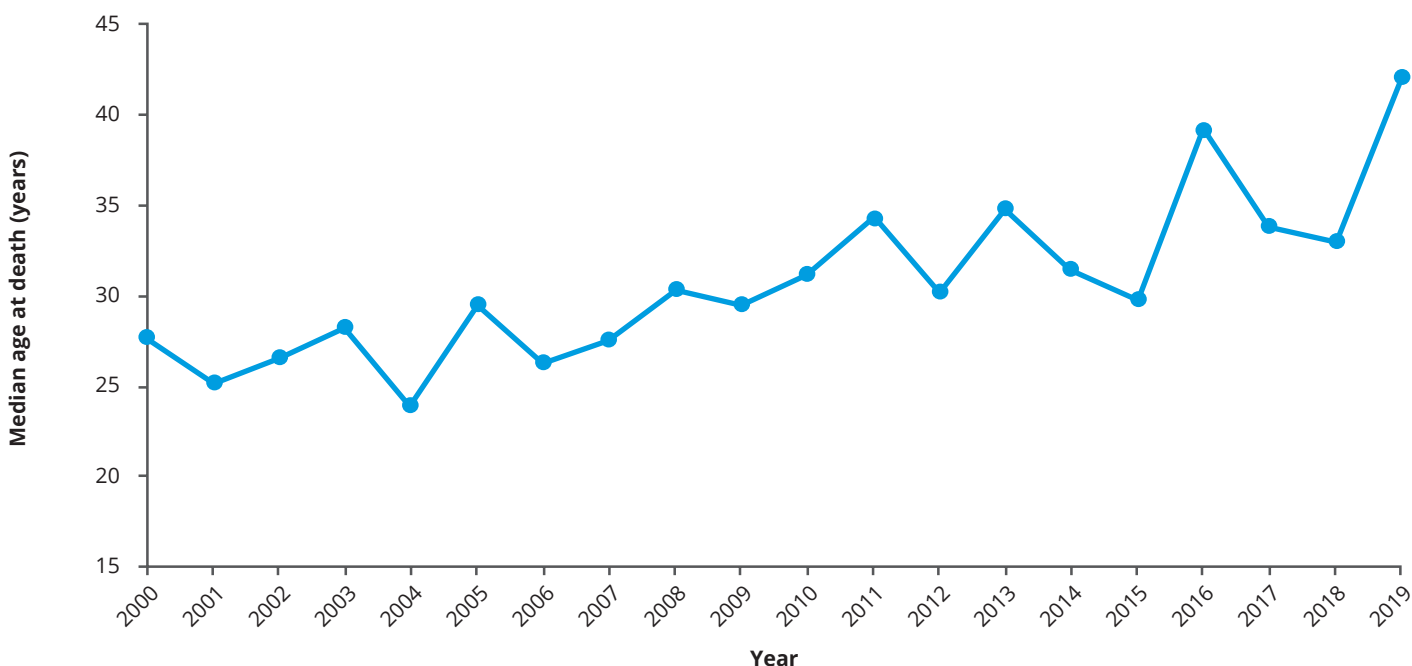




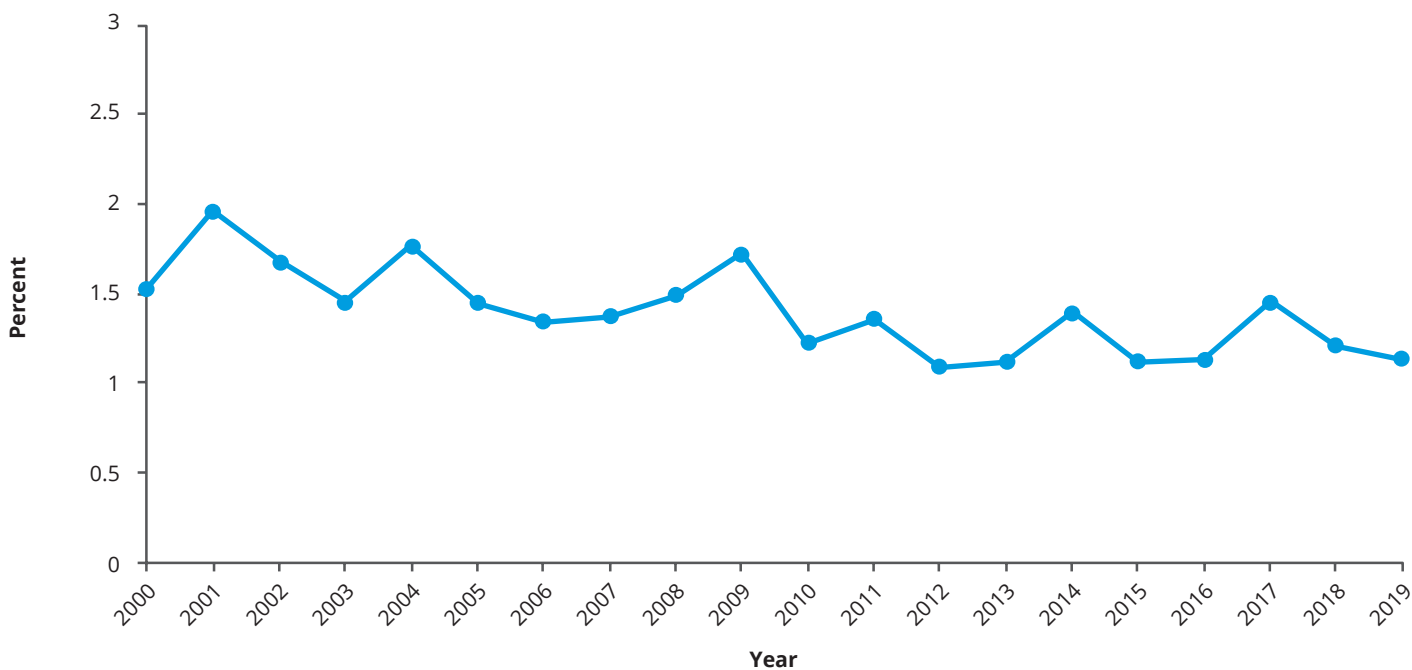
# SURVIVAL

Over the past two decades, a gradual increase in the median age of death can be seen. The median age of death is 42.1 years in 2019, compared to 33.0 years in 2018 and 27.7 years in 2000 (Figure 48). The median age of death tells us that half of those who died were younger than 42.1 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 2019, this value was 1.1%.

**FIGURE 48**  
Median age at death per year, 2000 to 2019.



**FIGURE 49**  
Death rate per year, 2000 to 2019.

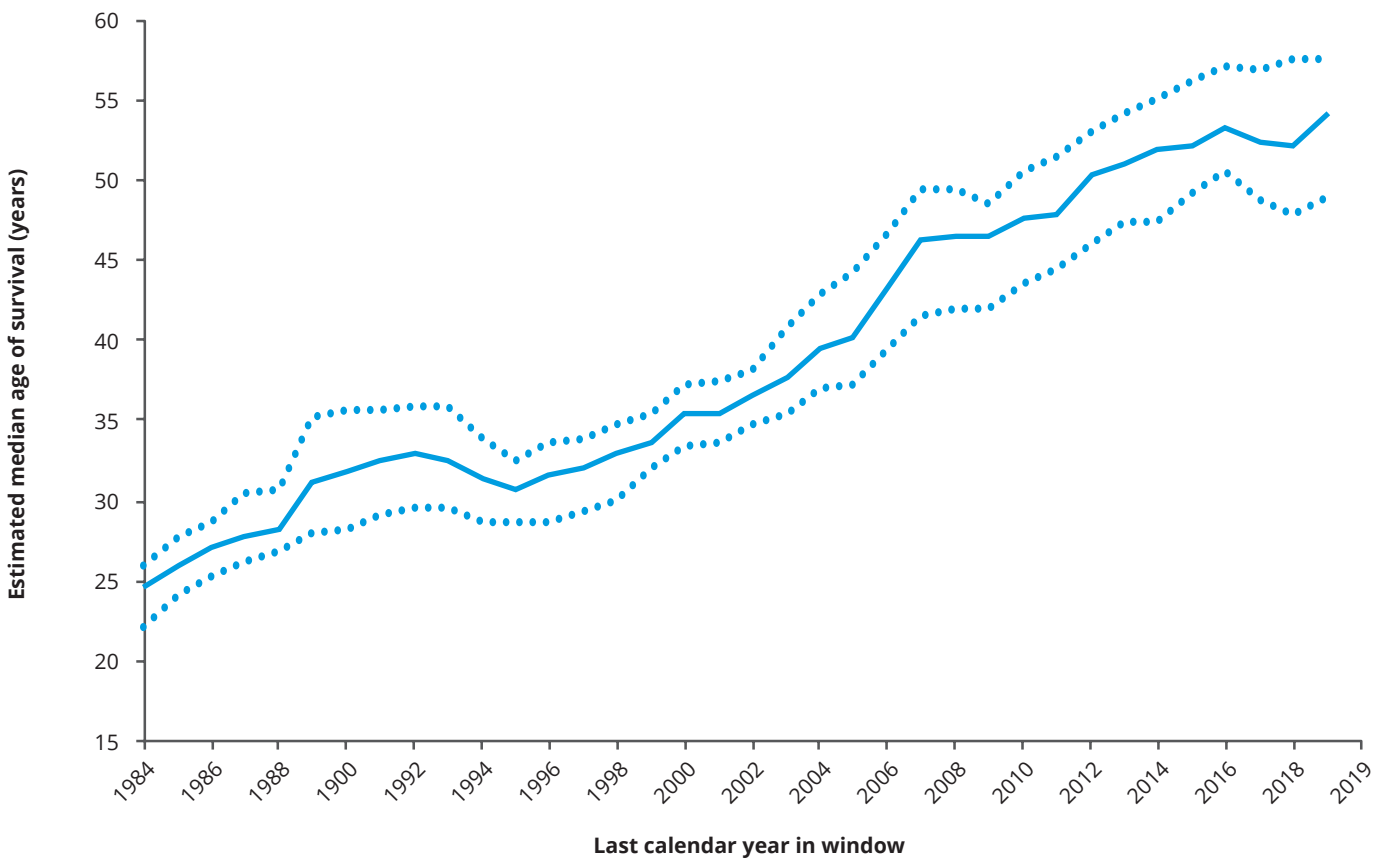


# ESTIMATED MEDIAN AGE OF SURVIVAL

A 5-year rolling window, to stabilize the estimates over time, was used to calculate the median age of survival using the Cox proportional hazards model. The most recent 5-year window (2015 - 2019) included 5,066 people with cystic fibrosis and 260 deaths. The number of individuals with cystic fibrosis lost-to-follow-up (defined as individuals with cystic fibrosis who are alive but haven't been reported on in the past 2 years) was 174 (3.4%).

In 2019, the median age of survival is currently estimated to be 54.3 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<sup>7</sup>.

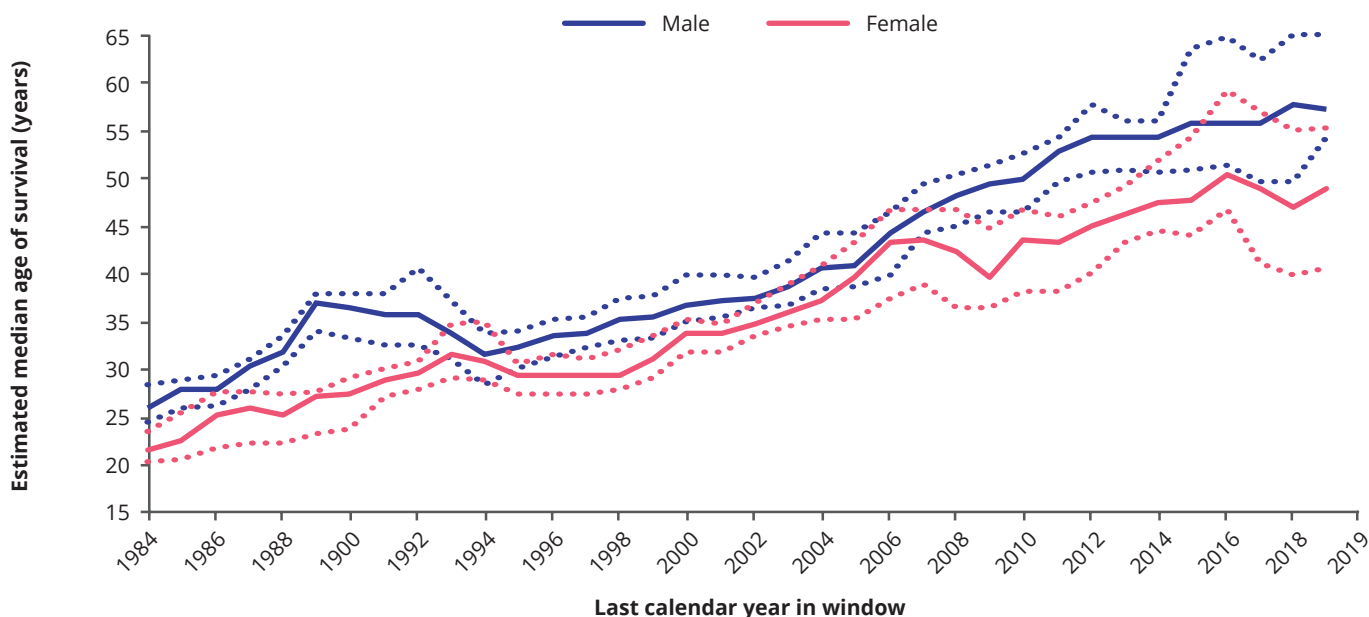
**FIGURE 50**  
Estimated median age of survival for a moving 5-year window with 95% confidence intervals, 1984 to 2019.



# ESTIMATED MEDIAN AGE OF SURVIVAL

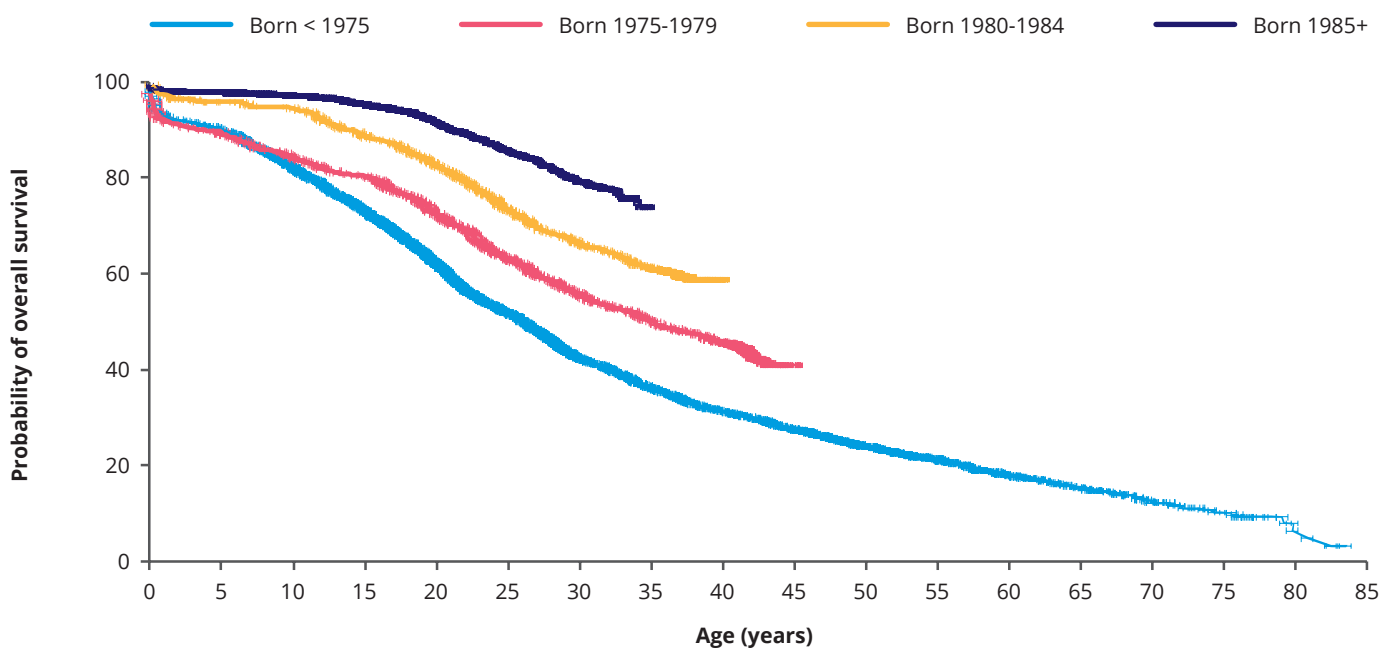
The median age of survival remains stable for both males and females with males continuing to have a higher median age of survival compared to females (Figure 51). While the cause of lower survival in females is not well understood, it has been documented in published CF literature<sup>8,9,10</sup>.

**FIGURE 51**  
Estimated median age of survival for a moving 5-year window with 95% confidence intervals, by sex, 1984 to 2019.



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 91.6% for those born in 1985 or later compared to 62.1% for those born before 1975.

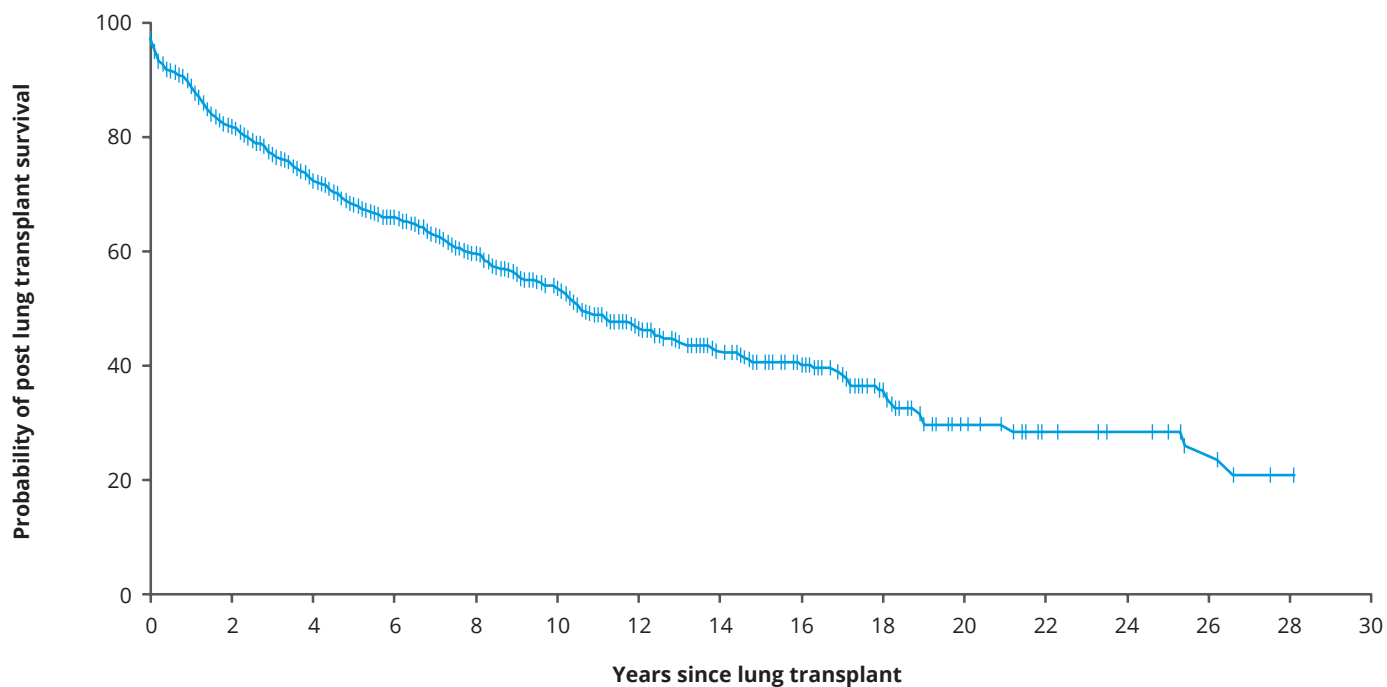
**FIGURE 52**  
Overall survival of individuals with cystic fibrosis, by birth cohorts, 2019.



# POST LUNG TRANSPLANT SURVIVAL

Between 1988 and 2019, there were 884 lung transplant recipient and 385 deaths post lung transplant. Figure 53 shows the probability of survival post lung transplant which is 88.9% at one year, 77.0% at three years and 68.2% at five years. Overall, 50% of those patients transplanted today would be expected to live beyond 10.6 years following lung transplantation.

**FIGURE 53**  
Post lung transplant survival, 2019.



# 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<sup>11</sup>. 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**<sup>12</sup>. 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 2019 is 54.3 years, we are saying that if a child with cystic fibrosis is born in Canada in 2019, they have a 50% chance of living beyond 54.3 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 54.3 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 2019.

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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Foothills Hospital, Calgary	Centre de santé et de services sociaux de Gatineau, Hull
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Grand River Hospital, Kitchener	QEII Health Sciences Centre, Halifax
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