

2022 CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY HIGHLIGHTS



OVERVIEW

The positive clinical trends seen in 2020 and 2021 were sustained in 2022 with continued improvement in lung function and median survival. The number of pulmonary exacerbations and lung transplants remained low, while the number of pregnancies among women with CF remained much higher than historical trends. All of this occurred in the context of decreased use of chronic therapies, likely as a result of improved lung health for the many benefitting from elexacaftor/tezacaftor/ivacaftor (ETI). Although life was returning to “normal” in many regards, the number of clinical encounters remained lower than pre-pandemic levels. More visits were conducted in clinic in 2022 as compared 2020 and 2021. The many positive trends in 2022 are cause for optimism, but many challenges remain. The unanticipated effects of ETI and the pandemic such as excessive weight gain and obesity as well an increase in mental health concerns underscore the continued importance of holistic CF care delivery with a multi-disciplinary approach.

REGISTRY BY THE NUMBERS



32,621

People in the Registry



97,127

Clinic Visits



15,212

Telehealth Visits



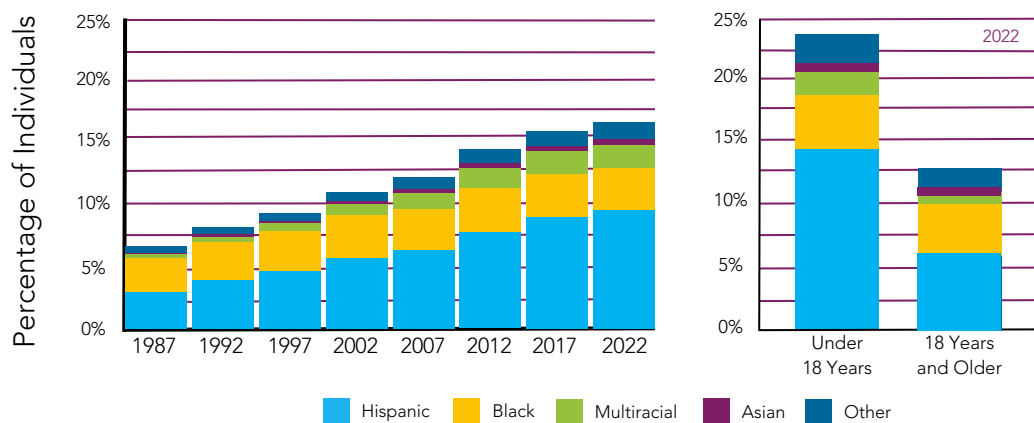
9,632

Hospitalizations

RACE AND ETHNICITY

The CF population as reported to the Registry is growing more diverse every year. In 2022, over 15 percent of people with CF were identified as either Hispanic, Black, multiracial, Asian or as other than White. This trend towards increased diversity will likely continue, as it is being driven by younger individuals with CF.

DIVERSITY IN THE CF POPULATION

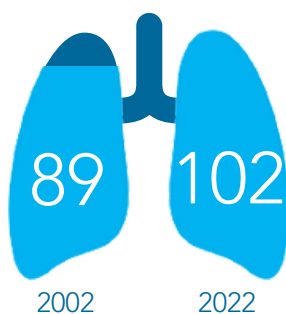


LUNG FUNCTION

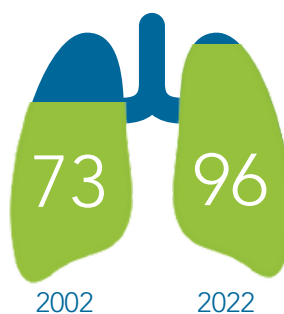
Lung function is a primary indicator of health for people with CF. FEV₁, a measure of lung function, is the Forced Exhaled Volume of air in the first second of an exhaled breath. It is shown as a percent predicted based on the FEV₁ of healthy, non-smoking people of the same age, height, and gender.

Median FEV₁ Percent Predicted

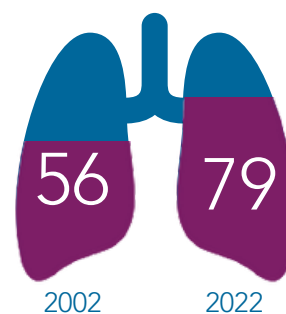
For 10 year olds



For 18 year olds



For 30 year olds



INSURANCE

CF care is expensive and includes many out-of-pocket costs for outpatient medical care, hospitalizations, and medications. Almost all people with CF have health insurance and many are enrolled in government-funded insurance programs.



58%

of all people with CF on a private insurance plan.



51%

of adults with CF 18-25 years old are on their parents' insurance plan.



13%

of adults with CF 18-64 years old are on Medicare.









39%

of all people with CF are on Medicaid.



54%

of children with CF under 10 years old are on Medicaid.

Bacteria	2018 Percent With Infection	2022 Percent With Infection
 <i>Pseudomonas aeruginosa</i>	44%	26%
 <i>Stenotrophomonas maltophilia</i>	12%	5%
 <i>Methicillin-resistant Staphylococcus aureus</i>	25%	16%
 <i>Achromobacter xylosoxidans</i>	6%	2%
 <i>Burkholderia cepacia complex</i>	3%	1%
 <i>Nontuberculous mycobacteria</i>	14%	10%

MICROBIOLOGY

People with CF are vulnerable to airway infections. To help prevent the spread of germs, infection prevention and control guidelines were created for clinics hospitals, homes, schools, and workplaces. Compared to 2018, the collection of respiratory cultures decreased in 2022, likely due to a continued decrease of in-person clinic attendance and inability to produce sputum for some. This may influence detection rates.

MENTAL HEALTH

PERCENT OF PEOPLE WITH CF AGE 12 YEARS AND OLDER WITH REPORTED DEPRESSION OR ANXIETY



2018 **23%** **19%**
Depression Anxiety

2022 **25%** ↑ **26%** ↑
Depression Anxiety

Addressing the mental health of all people with CF is critical to maintaining overall health and quality of life. Over the course of the pandemic there have been increased rates of anxiety and depression among people with CF similar to overall national trends.

MILESTONES

EMPLOYMENT



57%

of adults with CF have full-time or part-time jobs.

EDUCATION

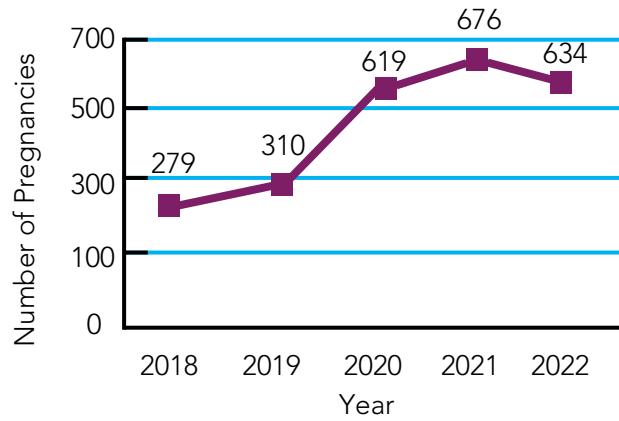


41%

of adults with CF have a college degree.

PREGNANCIES

The number of pregnancies remained relatively stable in 2022 as compared to 2020 and 2021. The total number of pregnancies reported among women with CF remained relatively constant between 2000 - 2019, with a marked increase starting in 2020.



NUTRITION

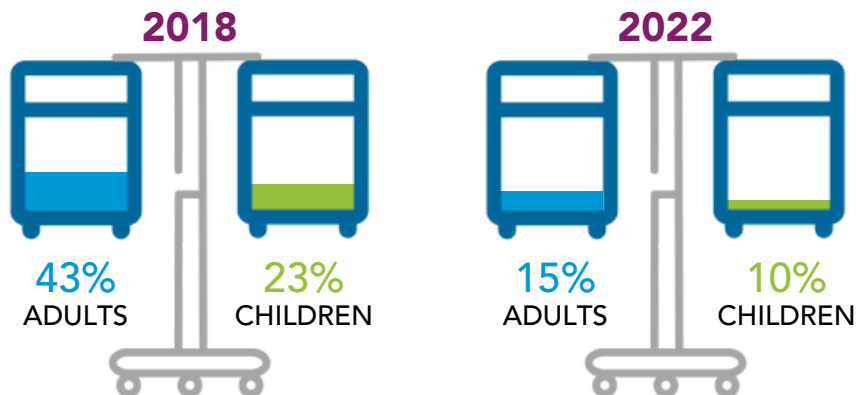


Proper nutrition is associated with better lung function. CFTR modulator use is associated with weight gain. This has resulted in a healthy weight for most people, but 13% are now considered obese as defined by a BMI greater than 30.

EXACERBATIONS

PEOPLE WITH CF WITH ONE OR MORE PULMONARY EXACERBATION

Pulmonary exacerbations, when lung symptoms worsen, are common in people with CF. This graphic shows those treated with intravenous (IV) antibiotics. Two likely contributors to this decrease include the additional attention to infection prevention control induced by the pandemic and the wide adoption of elexacaftor/tezacaftor/ivacaftor.



ADULTS MEETING BMI GOALS

65%

Over age 20, the BMI (Body Mass Index) goal is 23 for men and 22 for women.

MEDIAN BMI PERCENTILE FOR 2-19 YEAR OLDS

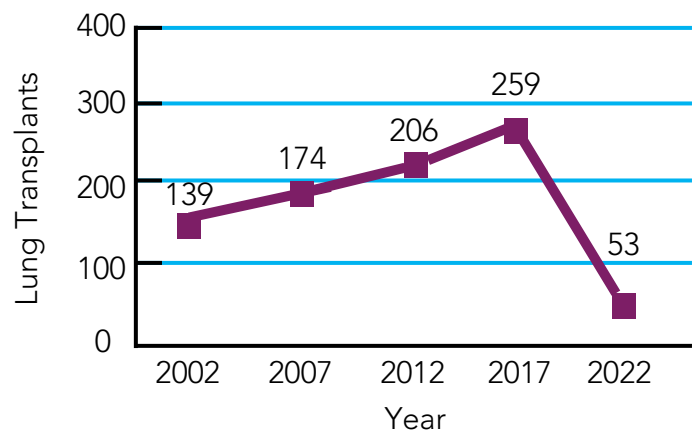
61

The BMI percentile goal is 50 or greater for children and adolescents.

TRANSPLANTATION

Lung transplantation remains an option for some individuals with severe lung disease. The number of lung transplant procedures for people with CF fluctuates. In 2022, 53 individuals with CF were reported to have undergone a lung transplant, including 14 re-operations for individuals with a previous transplant. The number of transplants in 2022 was a dramatic decrease from 2019 and prior years which corresponds to improvements in lung health.

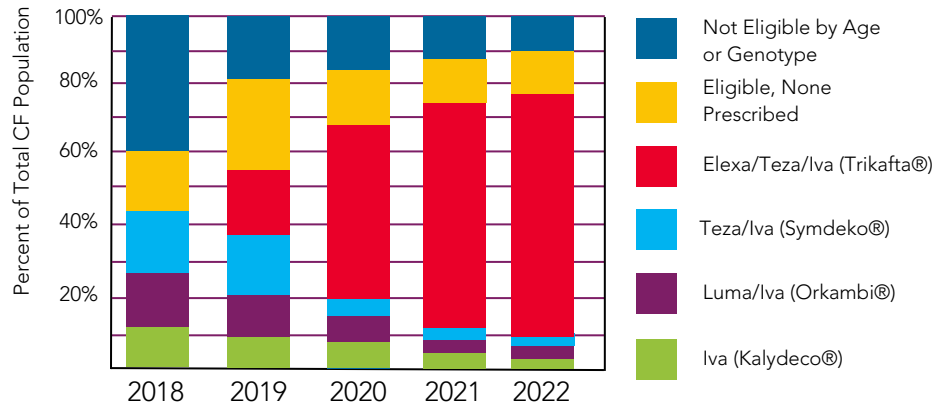
LUNG TRANSPLANTS BY YEAR



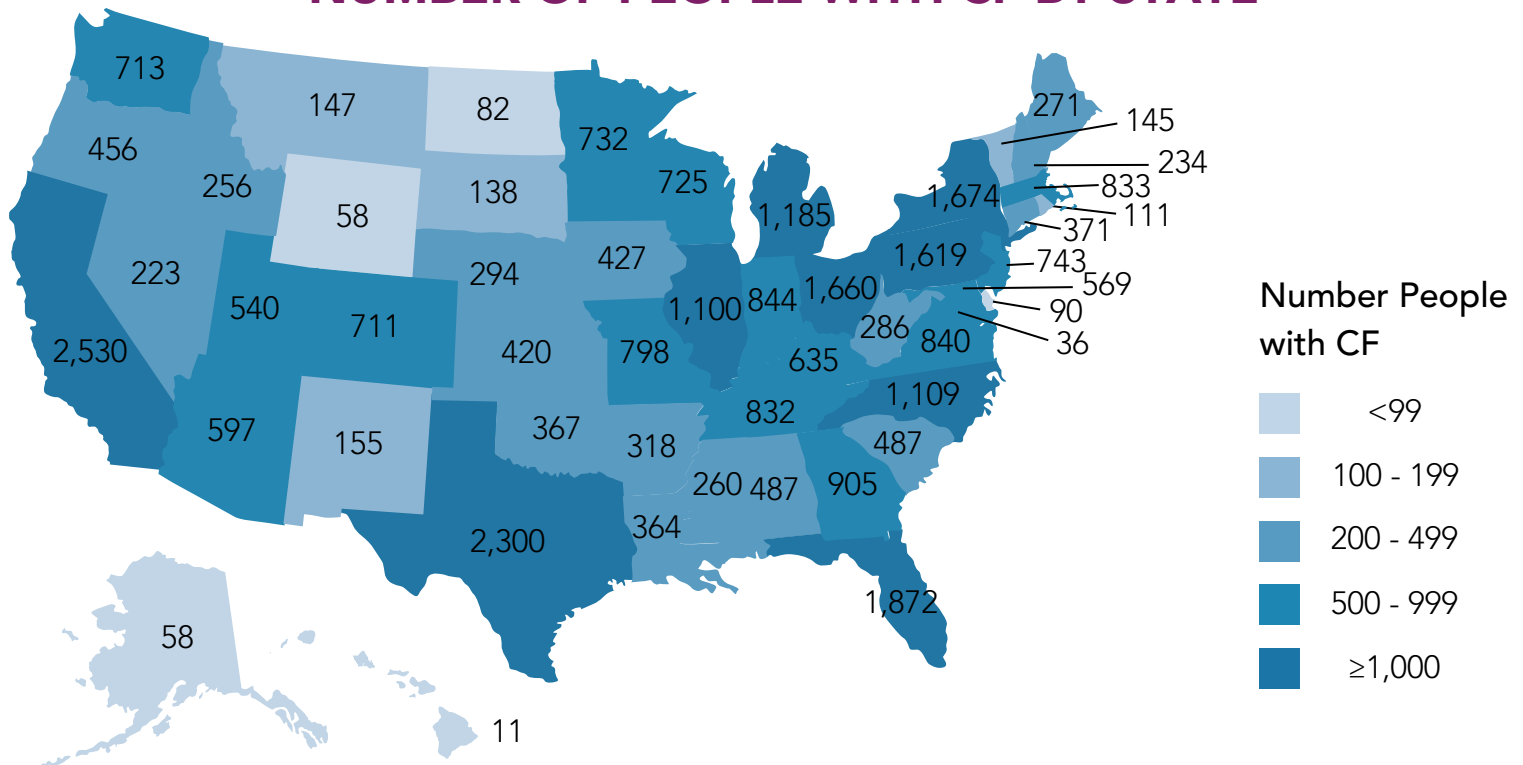
CFTR MODULATORS

The number of people with CF who are using CFTR modulator therapies continued to increase. The expanded approval of lumacaftor/ivacaftor for children with cystic fibrosis ages 1-2 years in September 2022 resulted in approximately 250 becoming eligible. More than 24,000 people with CF were taking a CFTR modulator by the end of 2022.

CFTR MODULATOR BY YEAR



NUMBER OF PEOPLE WITH CF BY STATE

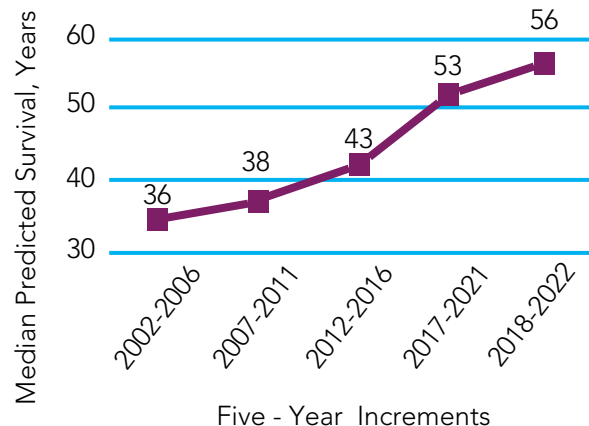


SURVIVAL

56
YEARS

2018 - 2022

Among people with CF born between 2018 and 2022, half are predicted to live to 56 years of age or more. This does not reflect individual variability in survival among people with CF.



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SOURCE DATA
Cystic fibrosis patients under care at CF Foundation-accredited care centers in the United States, who consented to have their data entered.

SUGGESTED CITATION
2022 Cystic Fibrosis Foundation Patient Registry Highlights
Bethesda, Maryland
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