

# 2021 CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY HIGHLIGHTS



## OVERVIEW



The year 2021 was the second year of the COVID-19 pandemic and the third year elexacaftor/tezacaftor/ivacaftor was available for people with CF. Telehealth accounted for 20% of CF care team visits. The 2021 Registry data reflect a combination of factors that impacted the lives of people with CF during the year. The favorable trends in lung function, pulmonary exacerbations, infection rates, and survival that were observed in 2020 data continued in 2021. These are strong indicators that broad uptake of elexacaftor/tezacaftor/ivacaftor is contributing to better health outcomes.

## PATIENT REGISTRY BY THE NUMBERS



32,100

People in the Registry



92,566

Clinic Visits



23,149

Telehealth Visits



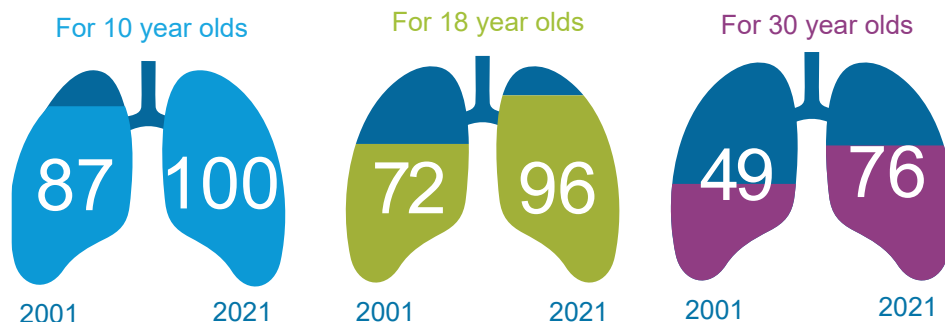
8,967

Hospitalizations

## LUNG FUNCTION

Lung function is a primary indicator of health for people with CF. FEV<sub>1</sub>, 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 FEV<sub>1</sub> of healthy, non-smoking people of the same age, height, and gender.

Median FEV<sub>1</sub> Percent Predicted

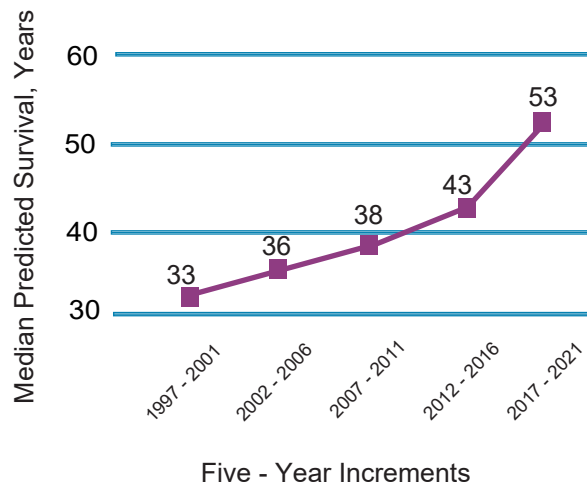


## SURVIVAL

53  
YEARS

2017 - 2021

Among people with CF born between 2017 and 2021, half are predicted to live to 53 years of age or more. This does not reflect individual variability in survival among people with CF.



## 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.



52%

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.



42%

of all people with CF are on Medicaid.



52%

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

## MICROBIOLOGY

Bacteria	2019 Percent With Infection	2021 Percent With Infection
<i>Pseudomonas aeruginosa</i>	43%	28%
<i>Stenotrophomonas maltophilia</i>	12%	6%
Methicillin-resistant <i>Staphylococcus aureus</i>	25%	18%
<i>Achromobacter xylosoxidans</i>	6%	3%
<i>Burkholderia cepacia</i> complex	3%	1%
Nontuberculous mycobacteria	14%	10%

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 2019, the collection of respiratory cultures decreased in 2021, likely due to decreased 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



2019 **24%** Depression **21%** Anxiety

2021 **25%** ↑ Depression **25%** ↑ 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



55%

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

EDUCATION

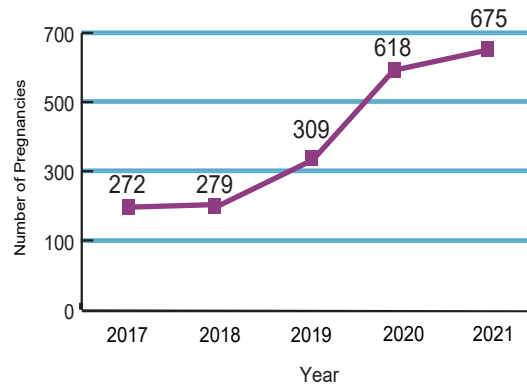


40%

of adults with CF have a college degree.

## PREGNANCIES

The number of pregnancies continued to increase in 2021. Registry data show that 675 people with CF were pregnant in 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. With CFTR modulators, adults with CF can gain weight more easily. This has resulted in a healthy weight for most people, but 12% are now considered obese as defined by a BMI greater than 30.

### 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

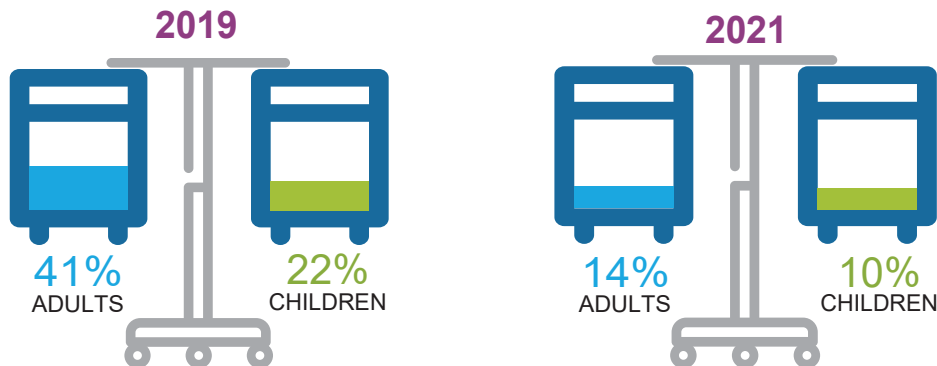
**62**

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

## EXACERBATIONS

### PEOPLE WITH CF WITH ONE OR MORE PULMONARY EXACERBATION

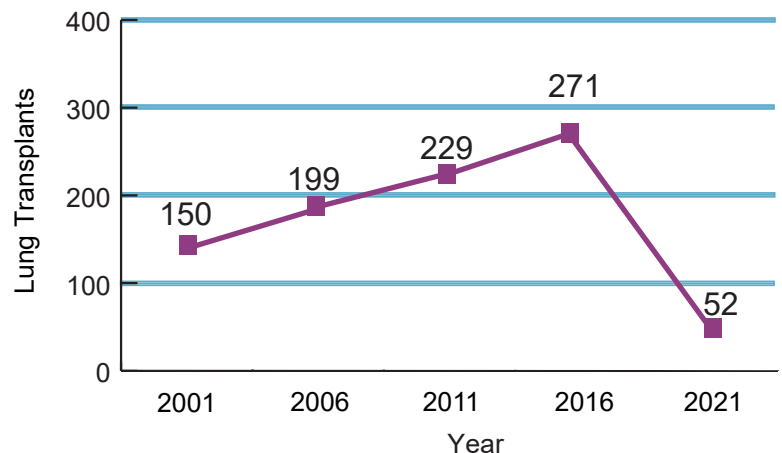
Pulmonary exacerbations, when lung symptoms worsen, are common in people with CF. This graphic shows those treated with intravenous (IV) antibiotics. In 2021, a substantial decrease in number of reported exacerbations was observed, particularly in adolescents and adults. The pandemic triggered attention to infection prevention and the use of elexacaftor/tezacaftor/ivacaftor may have contributed to this decrease.



## 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 2021, 52 individuals with CF were reported to have received a lung transplant, a dramatic decrease from previous years corresponding to improvements in health status.

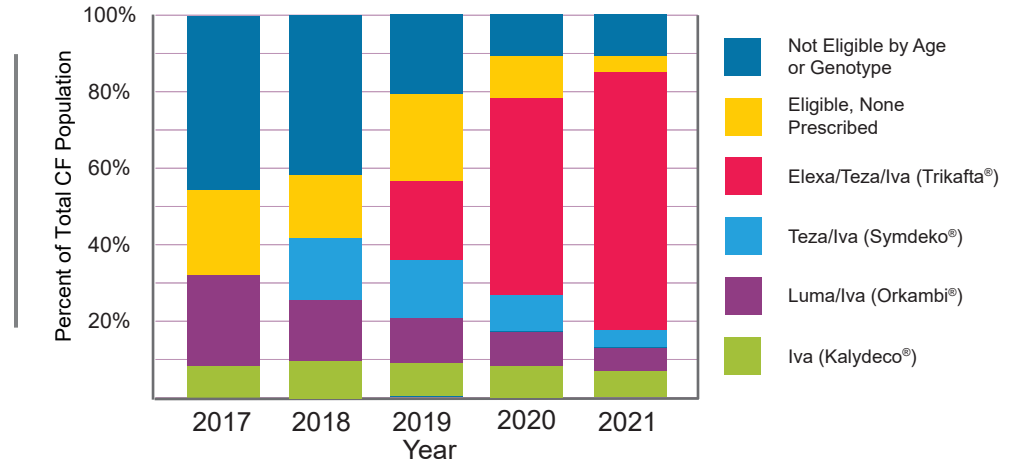
### LUNG TRANSPLANTS BY YEAR



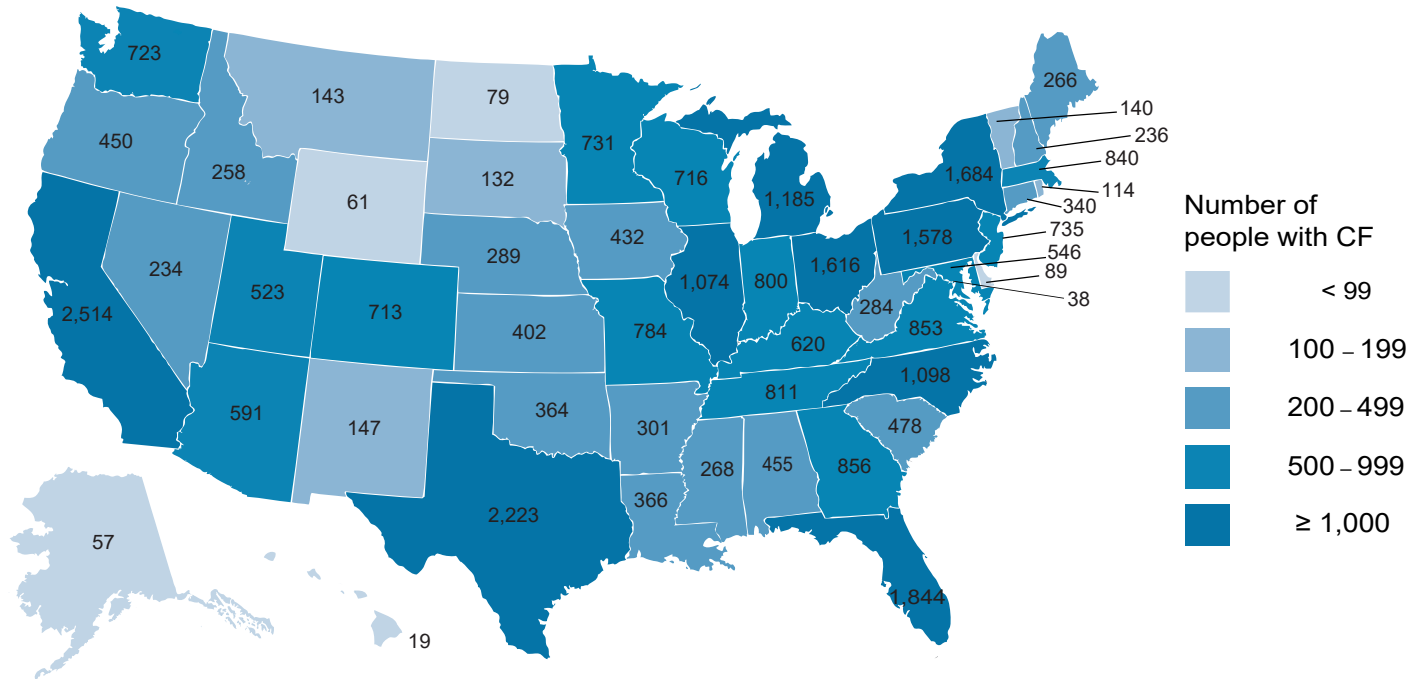
# CFTR MODULATORS

The number of people with CF who are using CFTR modulator therapies continued to increase. The expanded approval of elexacaftor/tezacaftor/ivacaftor for children with cystic fibrosis ages 6 - 11 in June 2021 resulted in approximately 1,500 becoming eligible. Ivacaftor was approved for children as young as 4 months in September 2021. More than 23,000 people with CF were taking a CFTR modulator by the end of 2021.

CFTR MODULATOR BY YEAR



## NUMBER OF PEOPLE WITH CF BY STATE



CFSmartReports.com uses CF Foundation Patient Registry data to support care teams in quality improvement and research.



98%

of all CF care programs accessed CFSmartReports



22

New study protocols identifying potentially eligible clinical trial participants



50,150

Total number of patient summary reports downloaded



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**SOURCE OF 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**  
2021 Cystic Fibrosis Foundation Patient Registry Highlights  
Bethesda, Maryland  
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