Uses of the Cystic Fibrosis Foundation Patient Registry

- Track Our Progress
- Support Clinical Trials
- Assess Therapies After Approval
- Improve Quality of Care
- Compare Treatment Options
- Inform Clinic Visits

Patient Registry by the Numbers

- 282 CF care programs
- 29,887 People in the Registry
- 129,542 Clinic visits
- 22,535 Hospitalizations

Number of People with CF in the Registry by State

- <99
- 100 – 199
- 200 – 499
- 500 – 999
- ≥1,000

2017 CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY HIGHLIGHTS
EXACERBATIONS

PEOPLE WITH CF WITH ONE OR MORE PULMONARY EXACERBATION

Pulmonary exacerbations, when lung symptoms worsen, are common in people with CF. Shown to the right are those treated with intravenous (IV) antibiotics.

43% ADULTS
24% CHILDREN

OF PEOPLE WITH CF TAKE THREE OR MORE INHALED MEDICATIONS

TREATMENT

TREATMENT COMPLEXITY

Therapies to manage CF symptoms include pills, airway clearance, exercise, inhaled medications, insulin, and nutritional supplementation. These therapies help people lead healthier lives, and can require complicated treatment plans that take two to three hours per day.

36% Inhaled Antibiotics
36% Hypertonic Saline
36% Dornase Alfa

LUNG FUNCTION

Median FEV1, Percent Predicted

For 10 year olds

1997 88
2017 97

For 18 year olds

1997 69
2017 85

For 30 year olds

1997 50
2017 61

LUNG FUNCTION

LIFE STATISTICS

Employment

51% of adults with CF have full-time or part-time jobs.

Marital Status

43% of adults with CF report being married or living together.

Education

31% of adults with CF have a college degree.

LIFE STATISTICS

CYSTIC FIBROSIS-RELATED DIABETES

Cystic Fibrosis-Related Diabetes (CFRD) is associated with weight loss, lung function decline, and increased mortality. Early diagnosis and treatment may minimize the impact of CFRD.

43% of people with CF age 10 and older were given an OGTT in 2017.

The CF Foundation and American Diabetes Association recommends screening all individuals annually, starting at age 10, with an oral glucose tolerance test (OGTT).
**NUTRITION**

**MEDIAN BMI PERCENTILE FOR 2-19 YEAR OLDS**

- 49 (2007)
- 57 (2017)

Children and teens with CF need adequate nutrition, which is a challenge due to the impact of CF on their digestive systems. The Body Mass Index (BMI) percentile goal is 50 or greater.

**ADULTS MEETING BMI GOALS**

- 41% (2007)
- 52% (2017)

Proper nutrition is associated with better lung function. Over age 20, the BMI goal is 23 for men and 22 for women.

**MICROBIOLOGY**

**PEOPLE WITH CF WHO CULTURED POSITIVE IN 2017**

- **Pseudomonas aeruginosa**
  - 46%
- **Methicillin-resistant Staphylococcus aureus**
  - 26%
- **Burkholderia cepacia complex**
  - 3%
- **Nontuberculous mycobacteria**
  - 13%

**MENTAL HEALTH SCREENING**

**DEPRESSION OR ANXIETY SCREENING FOR PEOPLE WITH CF AGED 12 AND OLDER**

- 58% (2016)
- 71% (2017)

Addressing the mental health of all individuals with CF is critical to maintaining their overall health and quality of life.

**CFTR MODULATORS**

**PEOPLE WITH CF ELIGIBLE FOR CFTR MODULATORS**

- 4% (2013)
- 38% (2015)
- 53% (2017)

Our Goal

**LUMACAFTOR/IVACAFTOR**

- 10,669 people with CF are eligible to take lumacaftor/ivacaftor

**IVACAFTOR**

- 4,293 people with CF are eligible to take ivacaftor

CFTR modulators are a new group of CF drugs that improve the function of the CFTR protein. The first CFTR modulator was approved in 2012 for people aged 6 and older with a G551D mutation. Since then, more people have become eligible to take modulators because the FDA approved a second drug and expanded the use of both drugs to younger age groups and more mutations.
The overall number of CF transplants reported to the Registry continues to increase.

Organ Transplants

Lung transplantation remains an option for some individuals with severe lung disease. The number of lung transplant procedures for people with CF fluctuates yearly, with an overall upward trend.

Reported to the Registry

<table>
<thead>
<tr>
<th>Year</th>
<th>Liver</th>
<th>Lung</th>
<th>Kidney</th>
</tr>
</thead>
<tbody>
<tr>
<td>2007</td>
<td>194</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2017</td>
<td>269</td>
<td></td>
<td>11 (Liver)</td>
</tr>
</tbody>
</table>

CF care can be 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.

Insurance

- 58% of adults with CF 18-25 years old are on their parents’ insurance plan.
- 52% of children with CF under 10 years old are on Medicaid.
- 17% of adults with CF 18-64 years old are on Medicare.
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Survival

People with CF continue to enjoy longer and healthier lives. Life expectancy estimates do not yet account for the potential benefits of newly available CFTR modulators and other recent clinical care improvements.

- 44 years (2013-2017) Among people with CF born between 2013 and 2017, half are predicted to live to 44 years old or more. This does not reflect individual variability in survival seen among people with CF.

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

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