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
1998 2018
For 10 year olds
1998 2018
For 18 year olds
1998 2018
For 30 year olds

PATIENT REGISTRY BY THE NUMBERS

<table>
<thead>
<tr>
<th>CF care programs</th>
<th>People in the Registry</th>
<th>Clinic visits</th>
<th>Hospitalizations</th>
</tr>
</thead>
<tbody>
<tr>
<td>285</td>
<td>30,775</td>
<td>130,836</td>
<td>22,377</td>
</tr>
</tbody>
</table>

SURVIVAL
44 YEARS
2014 - 2018
Among people with CF born between 2014 and 2018, half are predicted to live to 44 years old or more. This does not reflect individual variability in survival seen among people with CF.

Median Predicted Survival

NUMBER OF PEOPLE WITH CF BY STATE

CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY HIGHLIGHTS
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.

54% of adults with CF 18-25 years old are on their parents’ insurance plan.

16% of adults with CF 18-64 years old are on Medicare.

51% of children with CF under 10 years old are on Medicaid.

EXACERBATIONS

PEOPLE WITH CF WITH ONE OR MORE PULMONARY EXACERBATION

Pulmonary exacerbations, when lung symptoms worsen, are common in people with CF. The graphic on the right shows those treated with intravenous (IV) antibiotics.

43% of adults
23% of children

MICROBIOLOGY

<table>
<thead>
<tr>
<th>Bacteria</th>
<th>Percent With Infection</th>
<th>Median Age in Years at First Infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>44%</td>
<td>5</td>
</tr>
<tr>
<td>Methicillin-resistant Staphylococcus aureus</td>
<td>25%</td>
<td>11</td>
</tr>
<tr>
<td>Burkholderia cepacia complex</td>
<td>3%</td>
<td>20</td>
</tr>
<tr>
<td>Nontuberculous mycobacteria</td>
<td>14%</td>
<td>22</td>
</tr>
</tbody>
</table>

People with CF are more vulnerable to lung infections due to the sticky mucus in their lungs. To help prevent the spread of germs, infection prevention and control guidelines were created for clinics, hospitals, homes, schools, and workplaces.

CFTR MODULATORS

In people with CF, mutations in the CFTR (cystic fibrosis transmembrane conductance regulator) gene can result in a defective protein being produced. CFTR modulators are a group of drugs that improve the function of the CFTR protein.

<table>
<thead>
<tr>
<th>IVACAFTOR</th>
<th>LUMACAFTOR/IVACAFTOR</th>
<th>TEZACAFTOR/IVACAFTOR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age 1 year or older with an approved CFTR mutation</td>
<td>Age 2 years or older with an approved CFTR mutation</td>
<td>Age 12 years or older with an approved CFTR mutation</td>
</tr>
<tr>
<td>NUMBER ELIGIBLE</td>
<td>PERCENT PRESCRIBED</td>
<td>NUMBER ELIGIBLE</td>
</tr>
<tr>
<td>4,773</td>
<td>65%</td>
<td>12,167</td>
</tr>
</tbody>
</table>
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, but may require treatment plans that take hours each day.

COMPLICATIONS

<table>
<thead>
<tr>
<th>Complication</th>
<th>Age &lt; 18</th>
<th>Age ≥ 18</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asthma</td>
<td>29%</td>
<td>34%</td>
</tr>
<tr>
<td>Acid Reflux</td>
<td>34%</td>
<td>40%</td>
</tr>
<tr>
<td>CF Related-Diabetes</td>
<td>5%</td>
<td>31%</td>
</tr>
<tr>
<td>Sinus Disease</td>
<td>22%</td>
<td>54%</td>
</tr>
</tbody>
</table>

Some CF complications are more common in adults than children. Management of complications secondary to CF is important for maintaining an individual’s health and quality of life. 94% of people with CF were reported as having complications.

TRANSPLANTATION

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. In 2018, there were 1,715 post-lung transplant individuals in the Registry, including 253 individuals who were reported to have received a lung transplant this year.

NUTRITION

Proper nutrition is associated with better lung function.

ADULTS MEETING BMI GOALS

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

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

EMPLOYMENT

52%
of adults with CF have full-time or part-time jobs.

EDUCATION

39%
of adults with CF have a college degree.

MENTAL HEALTH

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

23% Depression
19% Anxiety

Addressing the mental health of all people with CF is critical to maintaining their overall health and quality of life. Since 2015, the CF Foundation has awarded mental health coordinator grants to care programs to conduct screening and treatment for depression and anxiety.

CARE TEAM CONSULTATIONS

PERCENT OF PEOPLE WITH CF SEEN BY EACH SPECIALIST

- Dietitian: 93%
- Pharmacist: 40%
- Physical Therapist: 29%
- Respiratory Therapist: 86%
- Social Worker: 90%

CF is a complex disease that requires partnership among multidisciplinary teams of health care professionals, people with CF, and their families to provide expert care.

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

- 99% of all CF care programs accessed CFSmartReports
- 18 Study protocols identifying potentially eligible clinical trial participants
- 68,522 Total number of patient summary reports downloaded