Highlights of the

2014 PATIENT REGISTRY

Data

CYSTIC FIBROSIS FOUNDATION

ADDING TOMORROWS

Preston W. Campbell, III, M.D.

Since the Cystic Fibrosis Foundation's Patient Registry was created 50 years ago, it has become an indispensable tool in our efforts to ensure all people with CF receive the highest quality, specialized care and the most effective treatments. The Registry captures a broad range of data on the health of those living with CF, providing critical information to help advance many important initiatives — from improvements in the quality of CF care and the development of care guidelines to the design of clinical trials to test new CF therapies.

This report highlights the steady progress we are making and also reflects the dedicated teamwork of many, including CF care centers, researchers, the Patient Registry team, and above all, those living with CF and their families. I thank each of you for helping make these gains possible and look forward to working with you to achieve our ultimate goal of a cure for all people with CF.

Preston W. Campbell, III, M.D.
President and CEO
Cystic Fibrosis Foundation

The Mission of the Cystic Fibrosis Foundation

The Mission of the Cystic Fibrosis Foundation is to cure cystic fibrosis and to provide all people with the disease the opportunity to lead full, productive lives by funding research and drug development, promoting individualized treatment, and ensuring access to high quality, specialized care.

CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

To the CF Community and Friends,

Bruce C. Marshall, M.D.

We are pleased to present the Cystic Fibrosis Foundation's *Highlights of the 2014 Patient Registry Data*.

Thanks to the steady progress in CF care and treatment, the face of the disease has changed dramatically over the last few decades. We achieved an important milestone in 2014, with more than half of those with CF in the United States now 18 years and older. This milestone is a tribute to people with CF and their families who follow through with the rigorous treatment regimen and to the commitment and leadership of care teams across the country that serve them. Such progress also presents a challenge to the CF community, as we strive to serve the needs of the growing number of adults living with the disease.

This positive trend and others deserve celebration, but the data also show that much work remains so all people with CF can lead healthy and fulfilling lives. Many individuals with CF still require hospitalization for treatment of exacerbations and, as the CF population ages, many now face other health problems like CF-related diabetes and depression, which add to the daily demands of living with this disease. Some people with CF pursue lung transplantation as an option to extend the length and quality of life. We are committed to addressing all of the challenges that those living with CF and their families face.

We hope you find this year's report rich and interesting and that you participate in the discussions about what the information means for you and for the CF community. This is a truly exciting time in CF, with advances in health care and new treatments that have the potential to transform the lives of many. Together, we will track these and other important developments in the Registry.

We are deeply grateful to all who have contributed to this report, especially people with CF and their families who so generously agree to share their information. Thank you for your continued commitment to our mission to cure CF.

Sincerely,

Bruce C. Marshall, M.D.
Senior Vice President
Clinical Affairs

Highlights of CF Foundation Patient Registry Data

From the analysis of the CF Foundation Patient Registry data in 2014

More than
28,000
people with CF
were seen at a
CF Foundation-accredited
care center.

49%
of people with CF
under the age of 10
were on Medicaid.

About
36%
of adults with CF are
college graduates, representing an
increase of almost
10
percentage points in the past
decade.

Over
50%
of those followed in the Registry
were age 18 years or older.

The
MRSA
infection rate
has remained stable
at about
25%
since 2010.

Among people with CF
over the age of 40,
1 in **6**
have received a
lung transplant.

Reporting of
fecal elastase values,
a measurement of
pancreatic sufficiency,
has increased from
40%
in 2011 to
60%
in 2014.

Pseudomonas
infections have steadily declined
over the last 20 years
from about
60%
in 1994 to
48%
in 2014.

28%
of adults with CF had depression
or anxiety reported.

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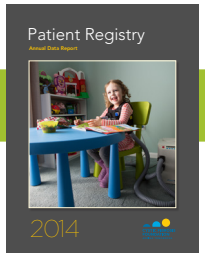
For more information

Please access the Patient Registry Annual Data Report at
https://www.cff.org/2014-Annual-Data-Report/
or search "patient registry report" on www.cff.org



About the CF Foundation Care Model

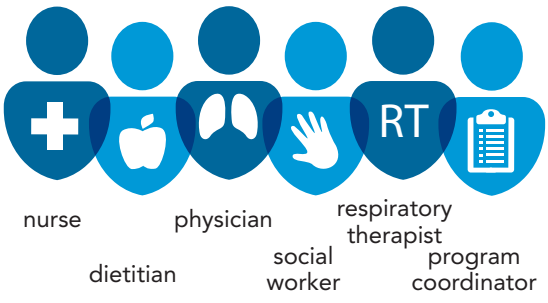
Care Center Network

The CF Foundation accredits a nationwide network of more than 120 care centers. Care centers are comprised of adult, pediatric, and affiliate centers. As more individuals with cystic fibrosis (CF) reach adulthood, the number of adult programs continues to expand. Multidisciplinary health care professional teams at the care centers work together to provide expert care tailored to meet the unique needs of individuals living with CF.

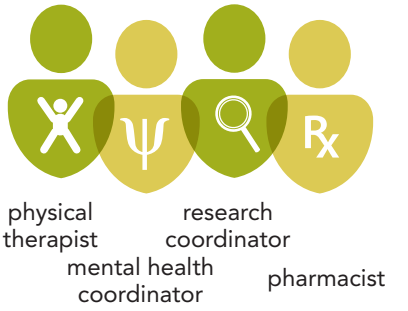
The CF Foundation's care center network is recognized as a national model for chronic disease care and for driving improvements in care. The CF Foundation is supported in this mission by a Center Committee that fosters exemplary care of all individuals with CF through the promotion of standards of care, the accreditation of care centers, the education of providers, and the advancement of research in all aspects of CF.

Each center undergoes an assessment by the CF Foundation's Care Center Committee to receive accreditation and funding. Accredited centers are reevaluated annually to ensure that people with CF receive effective and consistent levels of care and state-of-the-art treatments.

The Multidisciplinary Team



Required Team Members



Recommended Team Members

CF Foundation Clinical Care Practice Guidelines

The CF Foundation provides accredited care centers with clinical care practice guidelines, which are updated regularly based on current research, care, and treatments. The Foundation brings together committees of subject-matter experts, including physicians, nurses, respiratory therapists and dietitians, along with individuals with CF and their families, to develop care recommendations on each topic.

Quality Improvement Initiative

The CF Foundation works closely with care centers to ensure all people with CF receive the highest quality of care. The Quality Improvement Initiative provides training and resources that enable CF health care professionals and individuals with CF and their families to work together to improve care delivery and implement best practices. People with CF and their families are vital partners in this effort as they provide important insights on the experience of receiving care at their center and how they sustain daily care at home.

About the Cystic Fibrosis Foundation Patient Registry

Each year, information on the health status of children and adults with CF who receive care at CF Foundation-accredited care centers is entered into the Registry. This information provides critical data to help care teams and researchers identify new health trends, recognize the most effective treatments, design CF clinical trials and develop clinical care practice guidelines.

Uses of the Cystic Fibrosis Foundation Patient Registry

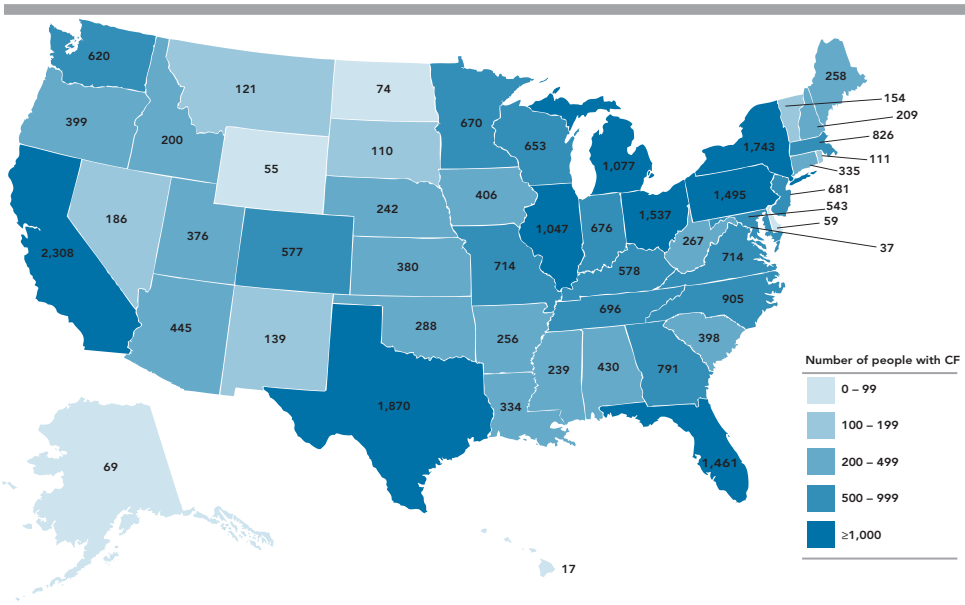
DISEASE SURVEILLANCE	FRAMEWORK FOR CLINICAL TRIALS	POST-MARKETING SURVEILLANCE STUDIES	QUALITY IMPROVEMENT	COMPARATIVE EFFECTIVENESS RESEARCH	INTERNATIONAL COMPARISONS
					
Track progress in curing CF and the impact of treatments	Test promising new therapies	Ensure safety and effectiveness of approved products	Provide all people with CF with high quality care	Promote evidence-based clinical decision making	Compare impact of different health systems

Overview of the Health of People with CF and the Standards of Care in the CF Foundation Care Center Network

Demographics

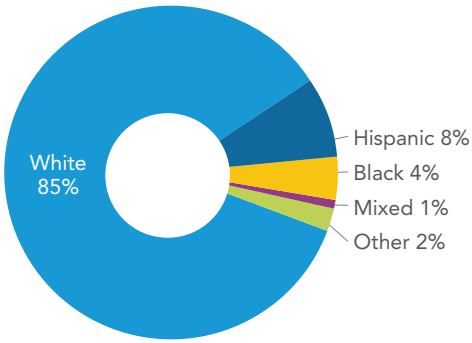
The records of 28,676 people with CF were included in the Registry in 2014. Individuals with CF are dispersed throughout the fifty United States, District of Columbia, and Puerto Rico. Males and females are relatively evenly distributed amid the CF population.

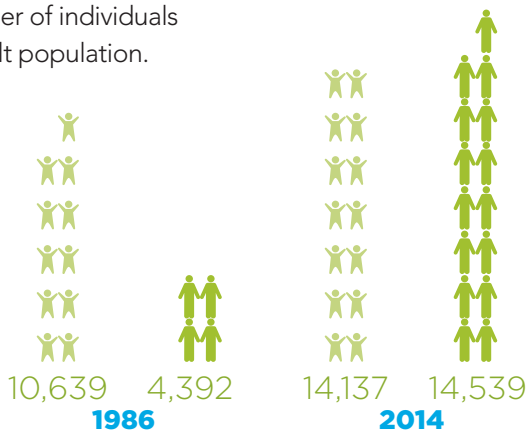
Number of People with CF Included in the Registry in Each State



The vast majority of individuals with CF are Caucasian. Over time, there has been minimal change in the racial distribution of individuals with CF; however, there has been a continual increase in the number of Hispanics in the registry, likely reflecting the growth of the Hispanic population in the general US population. This increase has led to efforts to develop more bilingual materials and capabilities within the centers and the CF Foundation. Since 1986, there has been a modest increase in the number of individuals younger than 18 in the registry along with more than tripling of the adult population.

Distribution of Race/Ethnicity among People with CF





Changes in adult versus pediatric population in CF Patient Registry

Insurance and Assistance Programs

CF care is multi-faceted and involves substantial costs for outpatient medical care, hospitalizations, and medications, thereby making health insurance for individuals with CF a necessity. Information on insurance is collected annually in the registry. The data suggest that across all age groups, almost all individuals report having medical insurance. Half of individuals with CF received at least part of their health insurance through federal or state-funded programs. A majority of people with CF age 18 to 25 received health insurance through their parents' plan in 2014.

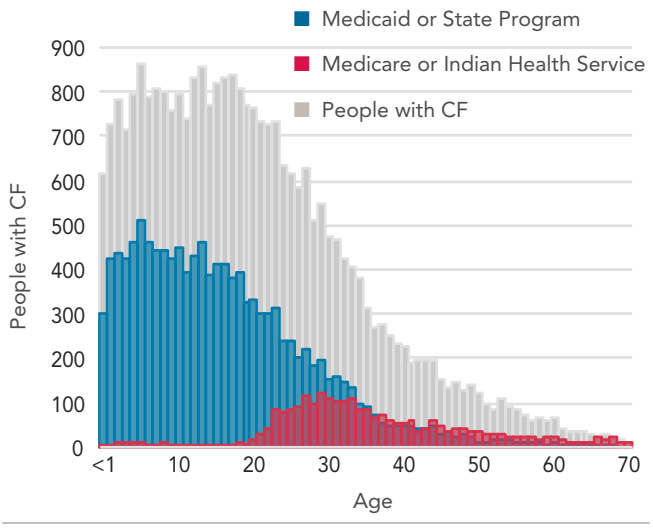
Insurance Coverage in 2014 for People with CF

Type of Insurance	Percent
Health insurance policy (e.g. private insurance)	60
Medicare/Indian Health Services	10
Medicaid/state programs	44
TriCare or other military health plan	3
Other	4
No health insurance	1

People with CF can be on more than one insurance type during the year or two or more types at one time.

57%
of people with CF age 18–25 are on their parents' insurance plan

Individuals on Government-funded Insurance Programs by Age, 2014



49%
of people with CF under the age of 10 are on Medicaid

17%
of people with CF age 18–64 are on Medicare

In 2014,
27%
of people with CF participated in a patient assistance program through CFF or another source

Accessing and affording treatment remains a major challenge for individuals with CF. To support individuals and families, the CF Foundation connects individuals to resources for assistance with co-pays, co-insurance and deductibles, offers reimbursement counseling to maximize insurance benefits, provides referrals to alternative funding sources for CF drug therapies and supports people with CF to navigate the social security application process.

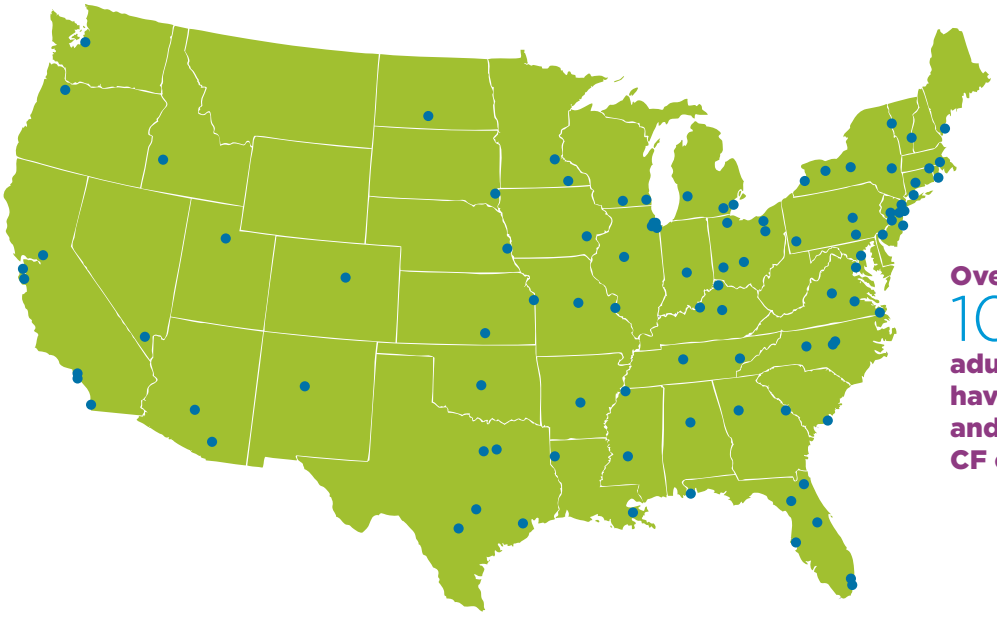
Adults with CF

In 2014, for the first time, there were more individuals in the Registry who were age 18 and older than individuals younger than age 18. This steady progression from CF as primarily a pediatric disease to one with a substantial adult population has necessitated an expansion of the care model. There are currently over 100 adult care programs within the CFF care center network. To ensure continuity of care, the Foundation works with care teams to help develop processes to support the hundreds of individuals transferring each year from pediatric to adult programs.

For the first time, adults 18 and over outnumber children in the Registry

1986
29.2%

2014
50.7%



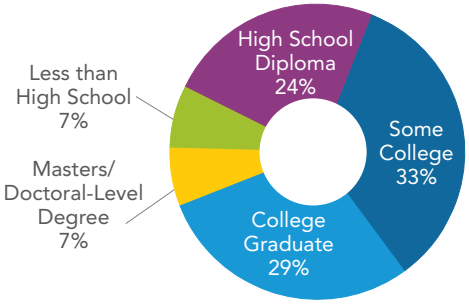
Over
100
adult care programs have been developed and accredited within the CF care center network

As a growing number of individuals with CF reach adulthood, more are pursuing higher education, employment, and starting families of their own. Since the late 1990s, the number of adults with CF with college degrees has more than doubled.

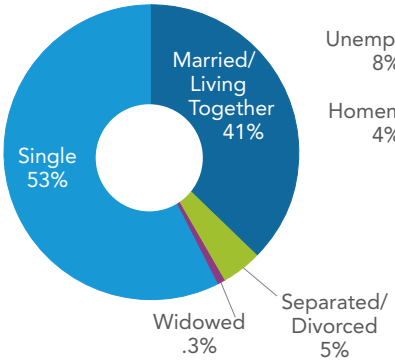
About
36%
of adults with CF are college graduates

Characteristics of Adults 18 Years and Older with CF in 2014

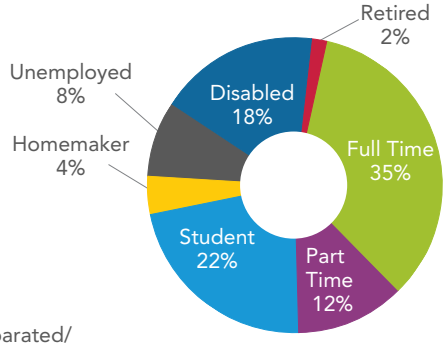
Education



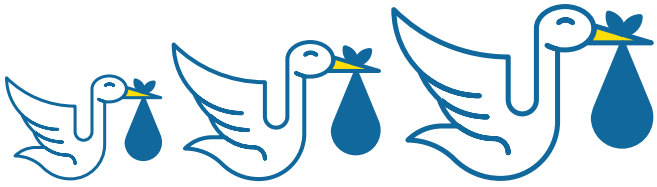
Marital Status



Employment



Number of Reported Pregnancies



139	185	232
1994	**2004**	**2014**

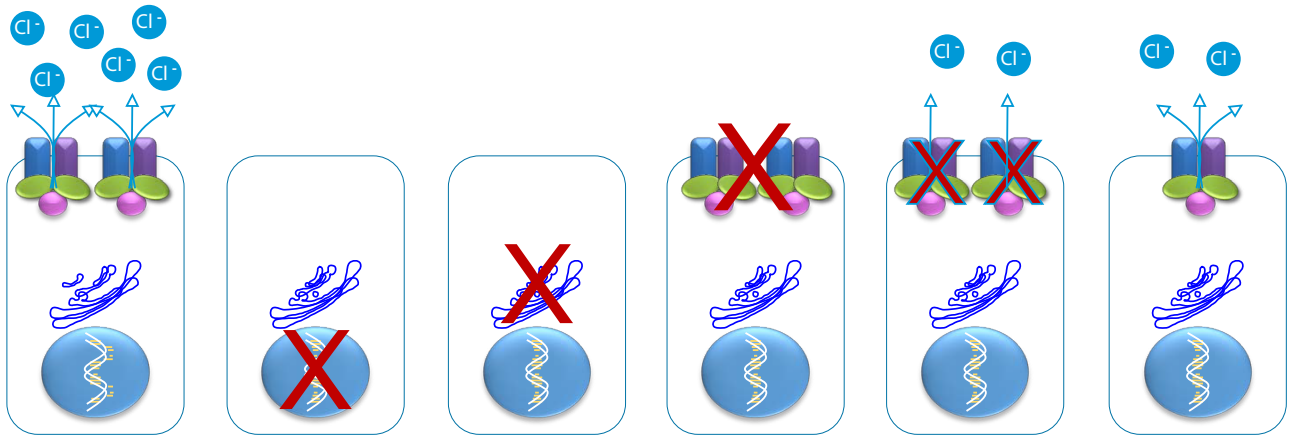
CF Genetics

In people with CF, mutations in both copies of the cystic fibrosis transmembrane conductance regulator (CFTR) gene disrupt the normal production of the CFTR protein. Different mutations cause CFTR to malfunction in different ways. In some people with CF, little to no CFTR protein is produced. In others, the defective protein is produced but cannot move to the surface of the cell where it is needed to regulate the transfer of chloride and other ions in and out of cells. In others, CFTR protein is produced and moves to the surface of the cell, but the gate that controls the movement of ions does not open properly.

There are more than 1,800 known CFTR mutations, many of which have been categorized by researchers into different groups. People with two mutations in classes I, II, and III typically exhibit more severe pulmonary disease and pancreatic insufficiency as compared to people with at least one mutation in classes IV or V.

CFTR Mutation Classes

The chart below shows just one of the ways that researchers classify CFTR mutations. Individuals with CF can consult with a genetic counselor or other member of their care team to learn more about their specific mutations. To find more information on CFTR and mutation classes, visit www.cftr2.org.



	Normal	Class I	Class II	Class III	Class IV	Class V
DESCRIPTION	CFTR is created, reaches cell surface and functions properly, allowing transfer of chloride and water.	No functional CFTR created.	CFTR protein is created, but misfolded, keeping it from reaching the cell surface.	CFTR protein is created and reaches cell surface, but the gate does not function properly.	The opening in the CFTR protein ion channel is faulty.	CFTR is created in insufficient quantities.
EXAMPLES		G542X W1282X R553X	F508del N1303K I507del	G551D S549N V520F	R117H D1152H R347P	3849+10kbC->T 2789+5G->A A455E

Adapted from: http://www.umd.be/CFTR/W_CFTR/gene.html

The prevalence of mutations varies; the majority of individuals with CF have at least one copy of the F508del mutation and almost half of the population has two copies. However, there are hundreds of mutations that are found in less than 0.5 percent of individuals with CF. Research is ongoing to understand how mutations affect the CFTR protein and to consider best approaches for treatment.

Prevalence of the 25 Most Common CFTR Mutations in 2014

MUTATION	NUMBER OF PEOPLE	PERCENT OF PEOPLE WITH ONE OR MORE COPIES OF THE MUTATION
F508del	24,157	86
G542X	1,289	5
G551D	1,219	4
R117H	784	3
N1303K	676	2
W1282X	631	2
R553X	503	2
1717-1G->A	445	2
621+1G->T	429	2
3849+10kbC->T	405	1
2789+5G->A	360	1
3120+1G->A	273	1
I507del	227	1
D1152H	205	1
R1162X	202	1
3659delC	195	1
1898+1G->A	184	1
G85E	179	1
R347P	164	1
R560T	164	1
2184insA	152	1
A455E	146	1
R334W	138	1
Q493X	130	1
E60X	117	less than 1

Homozygotes (two copies) - 46%
Heterozygotes (one copy) - 40%

The number and percent of individuals with a given mutation include those with one or two copies of the mutation.

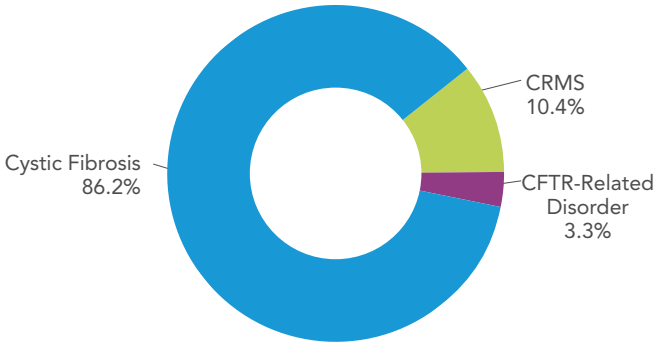
Diagnosis of CF

Nationwide newborn screening for CF has been in place since 2010. As a result, more people with CF are now diagnosed in infancy, often before symptoms of the disease appear. Almost two-thirds of new diagnoses were detected by newborn screening (NBS) in 2014. Early diagnosis allows for earlier treatment, which may lead to better lung function and nutritional outcomes later in life.

In 2014,
64%
of new diagnoses were detected by newborn screening

A challenge of newborn screening is the potential for misdiagnosing infants. In these cases, infants may receive unneeded CF treatments. To address this, the term CFTR-Related Metabolic Syndrome (CRMS) was developed to categorize a positive newborn screening test with diagnostic results that are not conclusive for CF. It is still unknown whether infants with CRMS will progress to develop symptoms of CF. Early indicators are that infants with CRMS typically have normal growth indices but about 10% culture positive for *Pseudomonas aeruginosa*. CFTR-related disorder is typically diagnosed in older people with CF who experience problems such as male infertility or pancreatitis, which cannot be explained by other causes.

CF, CRMS and CFTR-Related Disorder Diagnoses in 2014

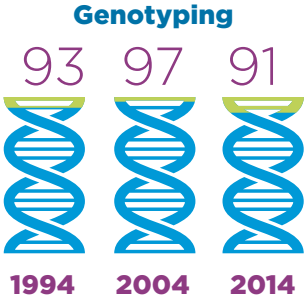


We estimate that the true prevalence of CRMS is greater than is indicated by the registry since it is not required to enter data on these infants.

Percent of People with CF with Diagnostic Test Results Reported by Year of Diagnosis

Sweat Test

Genotyping



It is extremely important for people newly diagnosed with CF to visit a CF Foundation-accredited care center to receive a complete diagnostic evaluation. A complete diagnostic evaluation includes a sweat test, genetic test, and clinical evaluation. It is encouraging that a majority of individuals diagnosed with CF receive genetic testing (genotyping) to determine their specific CFTR mutations. However, the current trend of decreased use of the sweat test as a diagnostic tool is concerning because even among individuals with two disease-causing mutations of the CFTR gene, a sweat test can provide information about severity of disease.

CF Clinical Care Practice Guidelines

CF clinical care practice guidelines are developed by expert multidisciplinary committees, based on published evidence and clinical experience. Guidelines are intended to inform care centers about CF care and treatment best practices and to be adapted by care center teams to the needs, preferences and values of the individual with CF and his or her family. Foundation-accredited care centers continue to increase the percentage of individuals who see dietitians, respiratory or physical therapists, and social workers each year.

71%
of people with CF were seen by all recommended specialists in 2014

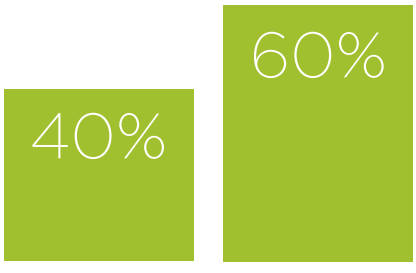
The majority of people with CF followed in the Registry receive care as recommended by the guidelines with almost all eligible individuals with CF receiving the influenza vaccine. Many people with CF receive testing for liver enzymes and fat-soluble vitamins. Of concern, there is less oral glucose tolerance testing, especially among adults. This may be due to external social and economic factors as teens and adults may be busy with school or jobs.

Care, Screening and Prevention Guidelines for People with CF

PERCENT OF PEOPLE WITH CF WHO HAD	2014			ELIGIBILITY CRITERIA FOR RECOMMENDATION
	UNDER 18 YEARS	18 YEARS AND OLDER	ALL	
An influenza vaccine (flu shot)	96	93	95	If 6 months of age or older
Fat-soluble vitamins measured	90	80	86	All
An oral glucose tolerance test (OGTT)	50	29	39	If 10 years of age or older and doesn't have Cystic Fibrosis-Related Diabetes
A blood test to measure liver enzymes	84	76	82	All
Seen by all recommended specialists	80	61	71	All

Given the importance of early life interventions, it is encouraging to observe that, on average, infants are seen frequently and according to recommendations at their CF care centers. Pancreatic insufficiency is best measured by the fecal elastase test and there has been considerable improvement in its use in recent years.

Reporting of Fecal Elastase Values for Children under 2 with CF



Median number of clinic visits during the first year of life for infants diagnosed by newborn screening

Growth and Nutrition

Nutritional outcomes are a key indicator of health in people with CF. Research suggests that maintaining a healthy weight is important to lung function. Children and teens with CF need adequate nutrition to grow and thrive, and it is important for adults to maintain proper nutrition to stay healthy.

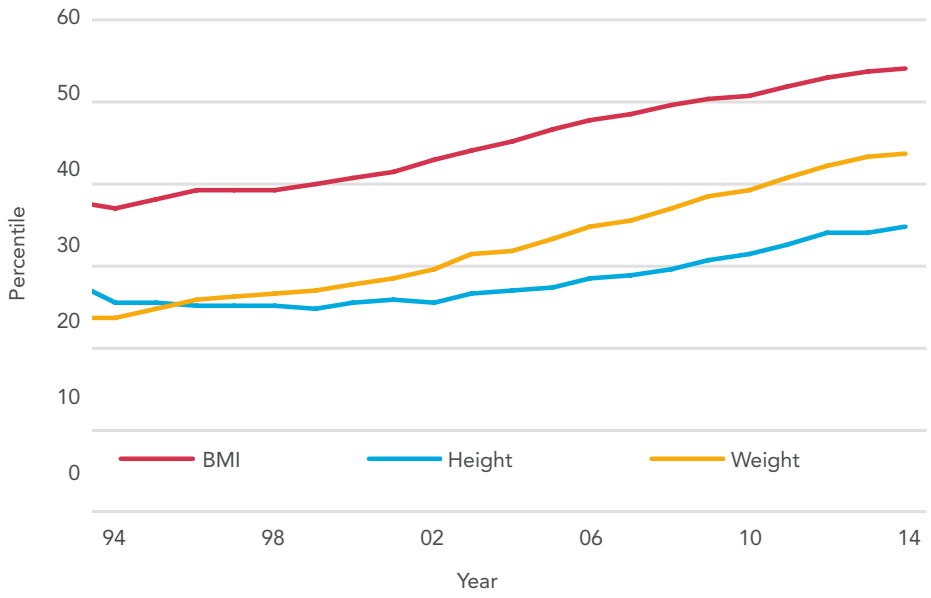
BMI Percentile

BMI percentile matches a child's BMI to other children in the United States of the same age and gender. A BMI of the 50th percentile means half of the children of the same age and gender are larger and half are smaller.

WHO Nutritional Outcomes for Individuals Under 24 Months

	MEDIAN
Weight-for-Length Percentile	63
Weight Percentile	45
Length Percentile	33

Median Nutritional Outcome Percentiles for Children and Adolescents Age 2 to 19 years, 1994-2014



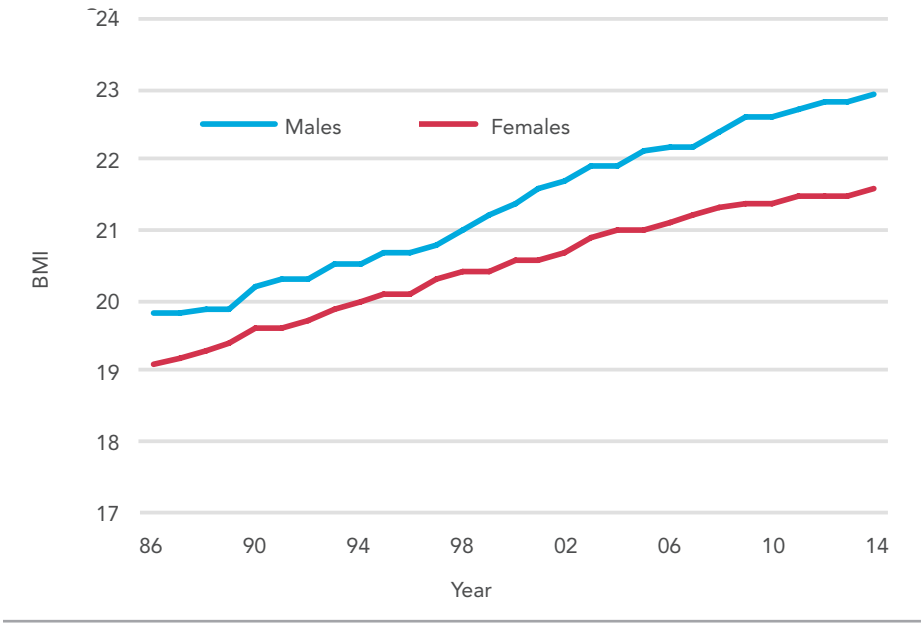
Growth and Nutrition

Nutritional outcomes have improved noticeably for both children and adults over the past decade. More than half of infants and children have a weight for length or **body mass index (BMI) percentile** that is greater than the recommendation. While these height and weight combined measures are promising, looking at height independently reveals that infants and children with CF are shorter on average than the general US population. Almost half of adults meet recommendations for body mass index and there have been improvements over time. However, **BMI** measurements for women with CF tend to be lower than for men with CF.

BMI

Body mass index (BMI) is based on a person's weight and height. It is used to help monitor nutritional health. To calculate, go to http://www.cdc.gov/healthyweight/assessing/bmi/index.html

Median BMI Value for People with CF 20 Years and Older, 1986-2014



People with CF Meeting BMI Goals

AGE 2–19

1994	2014
37%	55%

AGE 20+

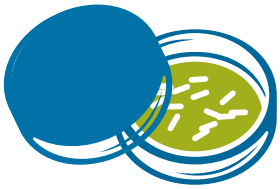
1994	2014
23%	48%

Microbiology

Pulmonary infections are a serious and chronic problem for many living with CF. People with CF are at greater risk of getting lung infections because thick and sticky mucus accumulates in their lungs and allows germs to thrive and multiply. To help reduce the spread of germs, infection prevention and control guidelines for CF were created. These CF Foundation guidelines provide recommendations for people with CF, their families, and CF health care professionals to help reduce the spread of germs in the clinic and hospital setting and at home, school, or work.

There are a number of microorganisms that are found in the lungs of people with CF. The microorganisms differ with regard to the method of exposure, how common they are and how often individuals have strains of the microorganisms that are resistant to antibiotics.

The prevalence of non-tuberculous microbacterial (NTM) infections is increasing in the general population. This is of concern because people with CF are at a higher risk of developing NTM infections which require long periods of treatment with multiple antibiotics. The majority of eligible individuals (i.e. individuals able to produce sputum) were tested for NTM in 2014.



69%

of people with CF who produced a sputum culture were tested for mycobacterial species

Microorganisms that are Dangerous to the Lungs of People with CF

BACTERIA	% WITH INFECTION	CURRENT OBSERVED TREND	MEDIAN AGE AT FIRST INFECTION	DISTINCTIVE FEATURES IN CF
Pseudomonas aeruginosa	48	Decline in rate of infection	6	• Leading cause of airway infection • Associated with a decline in lung function • 10% of strains are multi-drug resistant
Burkholderia cepacia complex	3	Slight decline in rate of infection	20	• Small proportion of people with CF infected • Can lead to rapid decline in pulmonary function • Most often multi-drug resistant
Methicillin-Resistant *Staphylococcus aureus*	26	Negligible increase in rate of infection	12	• Widespread among people with and without CF • Multi-drug resistant
Stenotrophomonas maltophilia	13	Unchanged rate of infection in the last 3 years	10	• Found in water, soil, plants, animals and hospital environments • Often multi-drug resistant
Alcaligenes species	6	Slight decrease in rate of infection	14	• Inhabits natural environment, including soil and water • Often multi-drug resistant
Non-tuberculous mycobacteria	12	Significant increase in rate of infection in recent years	22	• Of concern due to increase in reported rate of infection via inter-person spread • Treatment of infection is difficult • Typically multi-drug resistant

Lung Health

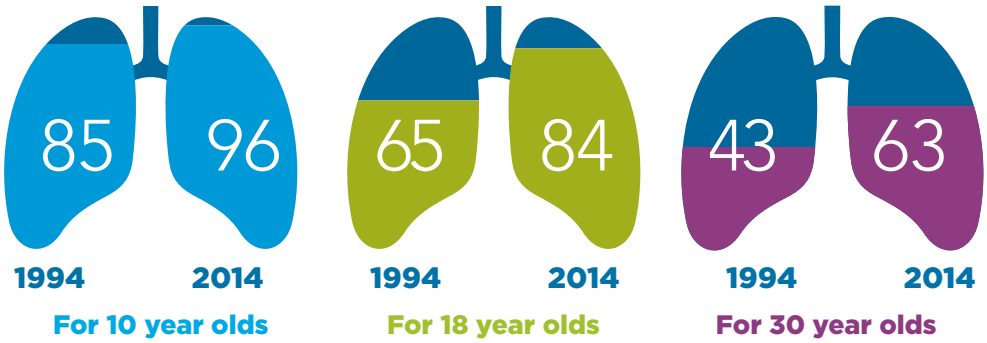
Pulmonary function is an important clinical health indicator of individuals with CF. Research shows that people with CF of all ages, including infants, have some lung damage — even when **FEV_1 percent predicted** is within the normal range.

Over the past 20 years, there have been considerable improvements in lung function among children, adolescents, and adults with CF.

FEV_1 Percent Predicted

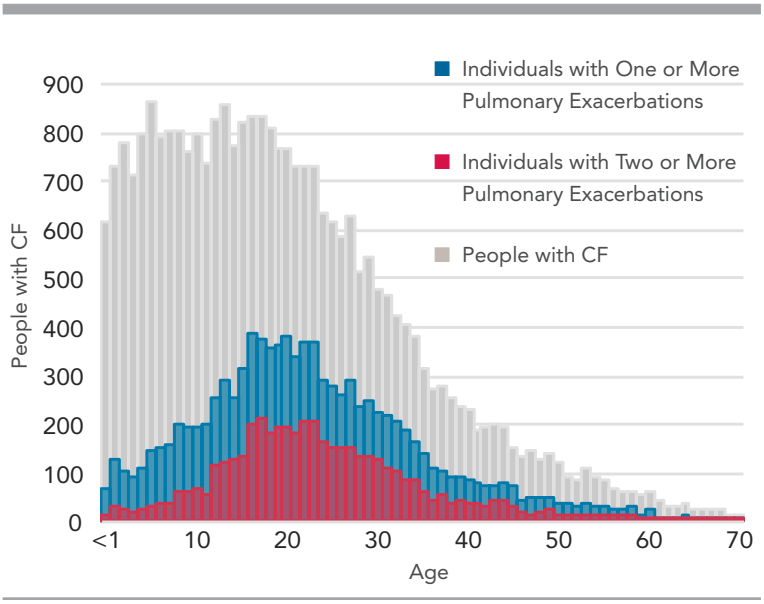
FEV_1 is a measure of lung function. It is the forced exhaled volume of air in the first second of an exhaled breath. It is shown as a percent predicted, based on the FEV_1 of healthy, non-smoking people of the same age, height and gender.

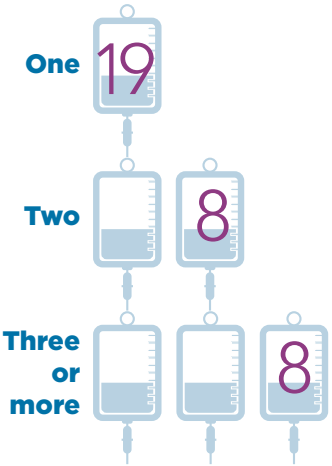
Median FEV_1 Percent Predicted in 1994 and 2014



Despite these improvements in pulmonary function, exacerbations (periods of increased symptoms requiring antibiotics) are still very common events for people with CF and their frequency has not changed over the past decade. About one third of individuals had at least one pulmonary exacerbation in 2014 that required treatment with IV antibiotics. Adults are more likely to experience exacerbations than children. Research efforts are underway to determine best practices for treating pulmonary exacerbations.

Pulmonary Exacerbations by Age, 2014

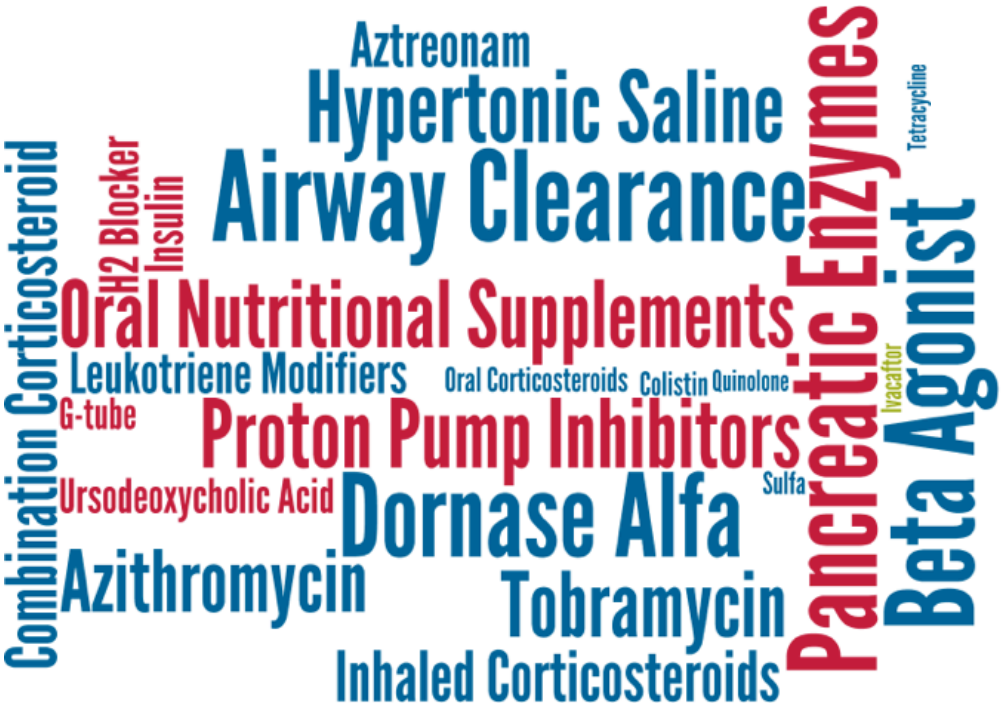




Percentage of people with CF treated for pulmonary exacerbations in 2014

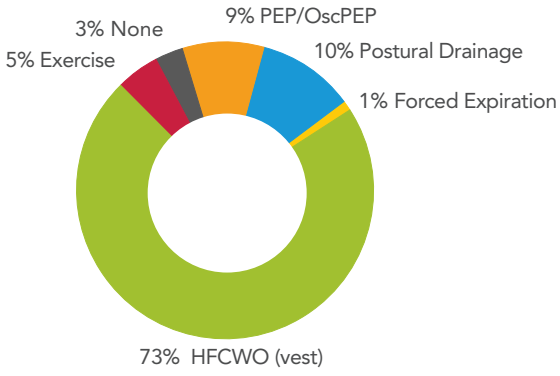
Treatment Used For Individuals With CF

Individuals with CF use a wide range of treatments to manage the symptoms and complications of their disease. These treatments include: airway clearance, drugs to thin mucus in the lungs, oral and inhaled antibiotics, pancreatic enzymes, insulin, drugs to reduce stomach acidity, bronchodilators, corticosteroids, vitamins, CFTR-modulators, oral and IV nutritional supplementation. The treatments reported in the Registry for at least 5 percent of individuals are included in the word cloud below. These treatments have provided opportunities for individuals with CF and lead to healthier lives. However, they necessitate complicated treatment plans that require intensive time commitments (often estimated as 2 – 3 hours / day). The CF Foundation's Partnerships for Sustaining Daily Care Program is currently investigating strategies to support individuals and their families.



CF Treatments
This word cloud represents the frequencies of the most common treatments prescribed for CF. Treatments for gastrointestinal symptoms are in red; treatments for respiratory symptoms are in blue; CFTR modulators are in green.

Primary Airway Clearance Techniques



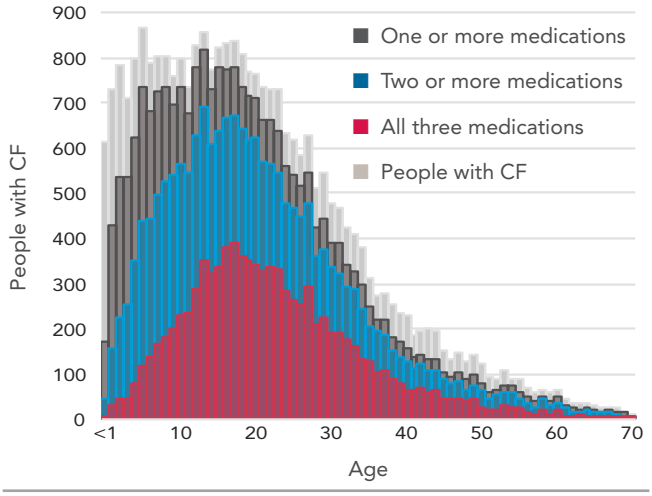
46%
of adults with CF use exercise as a primary or secondary airway clearance technique

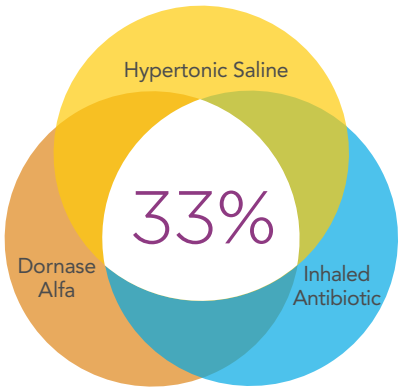
In 1994, dornase alfa was the first drug to be approved that specifically treated symptoms of CF. Subsequently, additional inhaled and oral antibiotics were determined to be effective in treating lung infections in individuals with CF. These medications are used extensively among eligible individuals.

Percentage (%) of People Prescribed CF Medications

RECOMMENDED CHRONIC MEDICATIONS FOR LUNG HEALTH	% OF THOSE ELIGIBLE	2014 CRITERIA FOR PRESCRIBING MEDICATION
Dornase alfa (Pulmozyme®)	86	• At least 6 years old
Hypertonic saline	66	• At least 6 years old
Tobramycin for inhalation solution (such as TOBI®)	70	• At least 6 years old • *P. aeruginosa* in cultures
Aztreonam for inhalation solution (such as Cayston®)	43	• At least 6 years old • *P. aeruginosa* in cultures
Azithromycin (such as Zithromax®)	68	• At least 6 years old • *P. aeruginosa* in cultures • Weight over 25 kg (55 lbs) • FEV_1 over 30% predicted
Ivacaftor (Kalydeco®)	89	• At least 6 years old • A G551D gene mutation

Inhaled Medication Use by Age, 2014



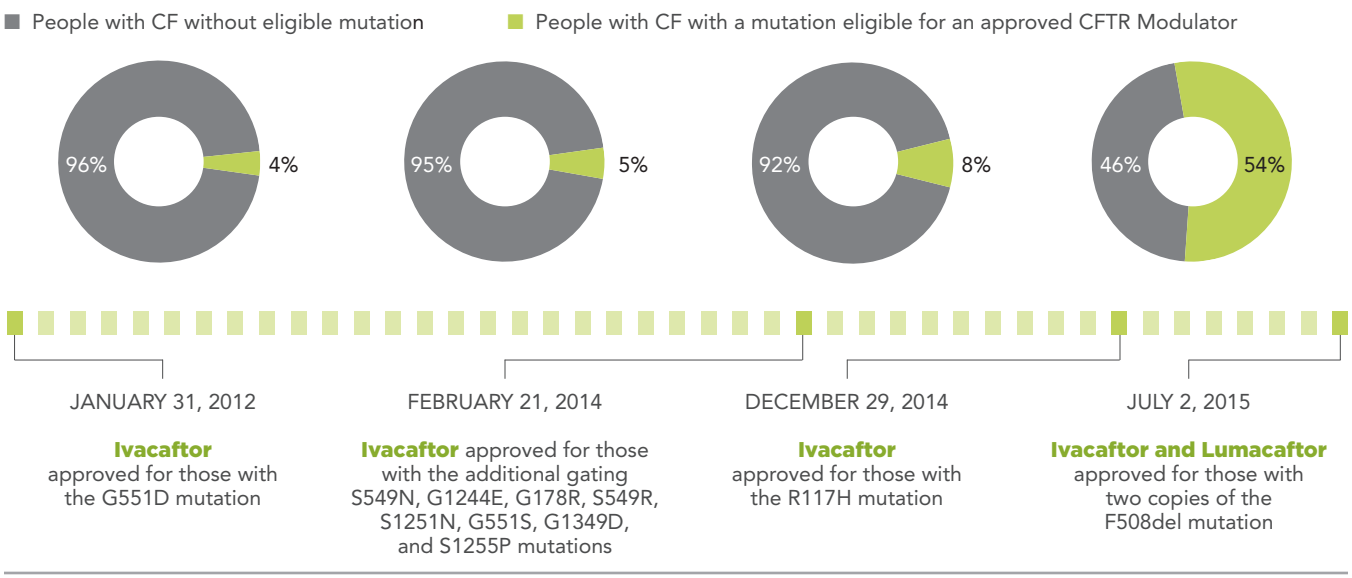


of people with CF take all three inhaled medications

Treatment Used For Individuals With CF continued

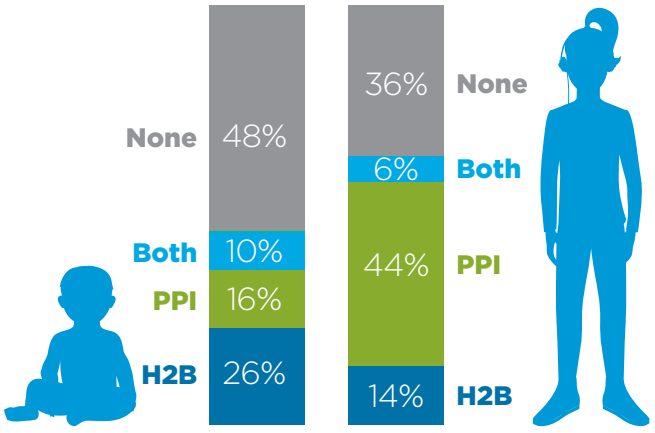
In 2012, the U.S. Food and Drug Administration approved the first of a new group of CF drugs, known as CFTR modulators, that address the basic genetic defect in CF. Different CFTR modulators, alone or in combination, are designed to treat the problems in the CFTR protein caused by specific mutations in the CF gene. The proportion of individuals with CF who can be treated with CFTR modulators continues to increase, with the aim of making these drugs available quickly to even greater numbers of people with CF.

Timeline of Approved CFTR Modulator Therapies



83%
of people with CF in 2014 were using pancreatic enzyme replacement therapy (PERT)

Acid Blocker Use

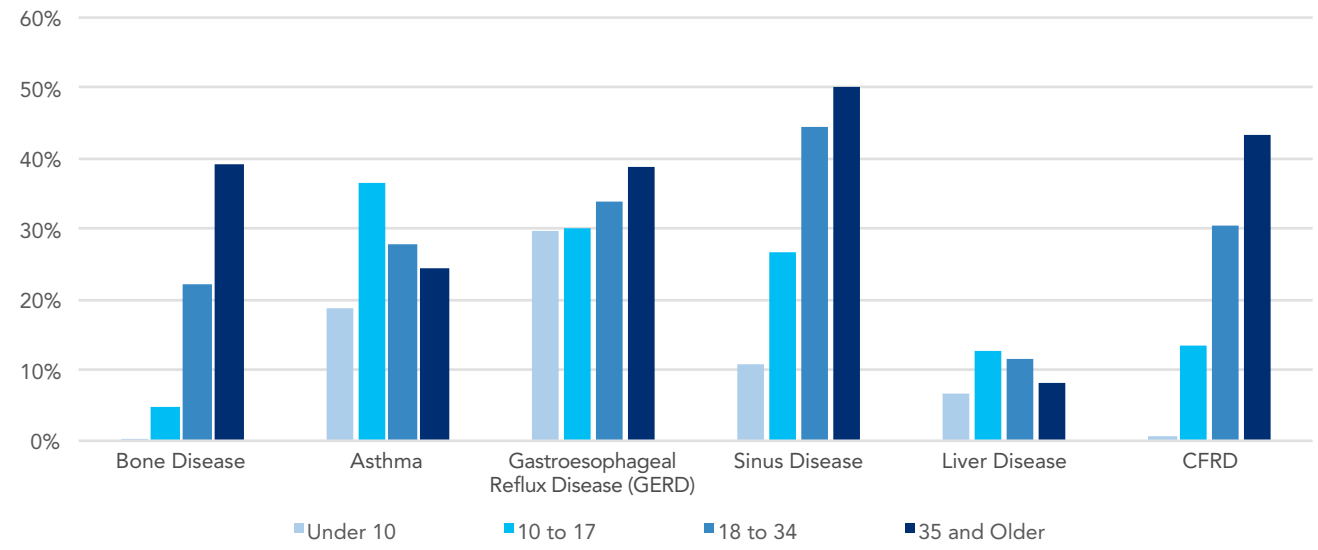


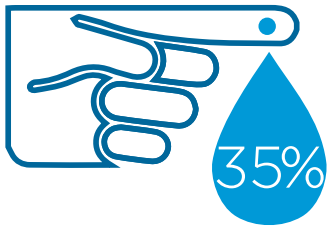
H2B = H2 Blockers
PPI = Proton Pump Inhibitor

Complications of CF

CF is often associated with complications other than lung disease and impaired nutritional status. These complications can be a direct result of the disease or a result of treatments for CF. Detecting complications early and managing them properly is crucial for the health and well-being of those with CF. The prevalence of complications increases with age, most notably **CF-related diabetes (CFRD)**, bone and sinus disease. These complications are more often reported among individuals with more severe lung disease.

Prevalence of Common Complications by Age in 2014





of people with CF age 18 years and older have CFRD

Cystic Fibrosis-Related Diabetes

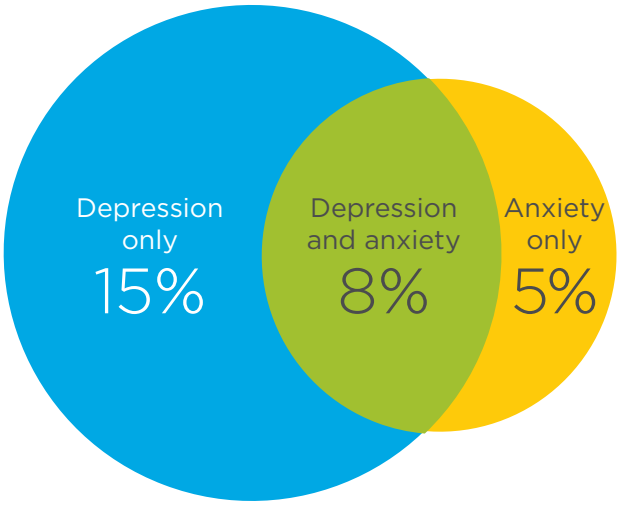
Cystic Fibrosis-Related Diabetes (CFRD) is a unique type of diabetes that is common in people with CF. CFRD can be treated with medication and monitoring blood sugar levels, eating a high-calorie diet and staying active.

Complications of CF continued

Mental Health

Mental health plays a central role in the long-term well-being, health, and quality of life of individuals with CF and their families. Therefore, it is critical to address the mental health needs of our community. Substantial proportions of individuals living with CF report anxiety, depression or both. The CF Foundation awarded grants for mental health coordinators for **depression and anxiety screening** and treatment to address this growing need.

Reported Prevalence of Depression and Anxiety in Adults with CF



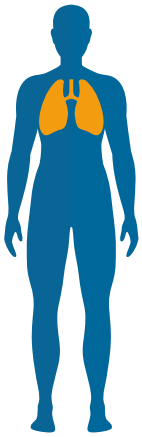
Depression and Anxiety Screening

Current recommendations are that individuals age 12 and older with CF and at least one of their caregivers receive yearly depression and anxiety screenings during routine clinic visits.

Transplantation

Lung transplantation remains an option for some people with CF who have severe lung disease. The annual number of lung transplant procedures for CF fluctuates yearly with an overall upwards trend. However, lung transplantation has its own risks and requires lifelong, post-transplant care.

As lung transplants become more common and recipients are living longer, a considerable proportion of adults with CF over 40 are post-transplant.

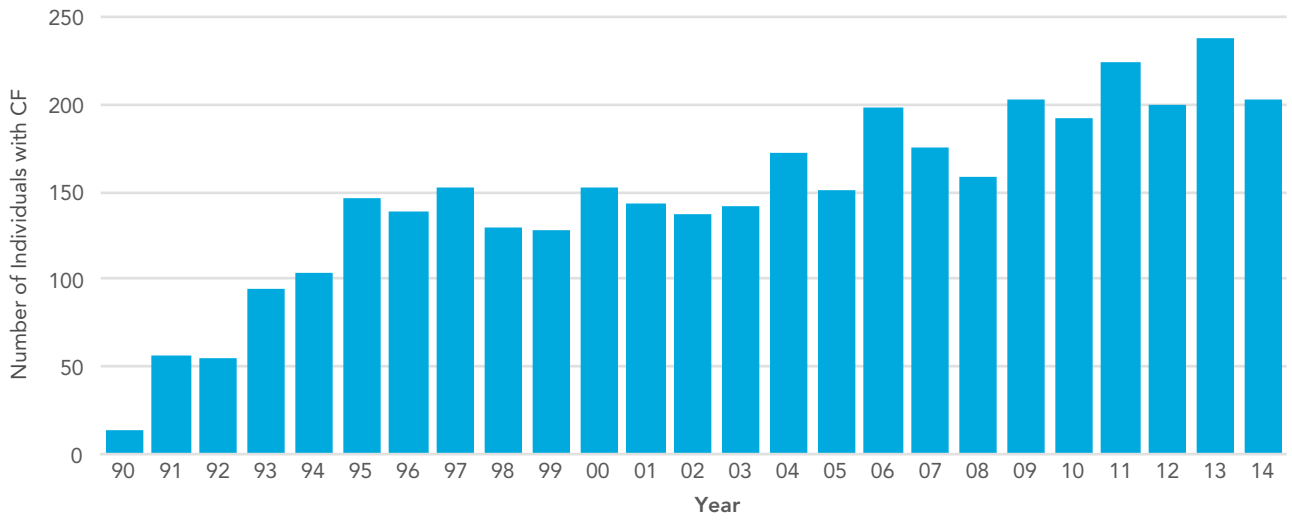


In 2014,

202

people with CF received a lung transplant

Number of Individuals with CF Receiving a Lung Transplant, 1990–2014



1 in 6 people with CF over 40 have received a lung transplant

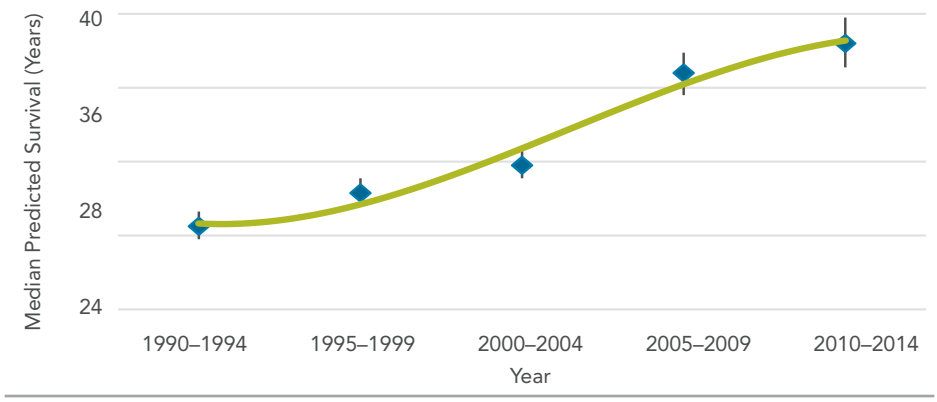
Survival

People with CF continue to enjoy longer and healthier lives thanks to advances in research and medical care. The **median predicted survival age** continues to increase. For all people with CF currently in the Registry, the median age for survival was 39.3 years, meaning 50 percent are expected to live to 39.3 years of age or beyond.

Median Predicted Survival Age

The median predicted survival age is the age to which half of the current Registry population would be expected to survive, given their ages in 2014 and assuming that mortality rates do not change. The median predicted survival age does not equal life expectancy.

Median Predicted Survival Age, 1990–2014 (in 5 year bands)



39.3
years
Median predicted survival age in 2014

The CF Foundation is committed to promoting individualized treatment and ensuring access to high quality, specialized care for people with CF. The findings from 2014 registry data provide insight into the current health of the CF population and provide clinicians with a road map for improving care. As new treatments and therapies emerge, the registry will remain a rich resource for researchers, clinicians, people with CF and families as they partner to improve care, treatment, and research.

Appendix of Resources

About the CF Foundation Care Model

Care Center Network
https://www.cff.org/For-Caregivers/CF-Care-Centers/

CF Foundation Clinical Care Practice Guidelines
CF Care Guidelines
https://www.cff.org/Care-Guidelines

Quality Improvement Initiative
Quality Improvement
https://www.cff.org/PDF-Archive/Partnering-for-Improvement/

Overview of the Health of People with CF and the Standards of Care in the CF Foundation Care Center Network

Demographics
Patient Registry Annual Data Report to Center Directors
https://www.cff.org/Patient-Registry-Reports
Resources in Spanish
https://www.cff.org/En-Espanol/

Insurance and Assistance Programs
https://www.cff.org/Living-with-CF/Navigating-Insurance/
https://www.cff.org/Compass/

Adults with CF
https://www.cff.org/Adult-Care-Guidelines
https://www.cff.org/Program-for-Adult-Care-Excellence

Diagnosis of CF
Testing for Cystic Fibrosis
https://www.cff.org/What-is-CF/Testing/
https://www.cff.org/What-is-CF/Diagnosed-with-Cystic-Fibrosis/

CF Genetics
About CF Genetics
https://www.cff.org/What-is-CF/CF-Genetics/
CF Mutation Analysis Program
https://www.cff.org/About-Us/CF-Mutation-Analysis-Program/
Types of CF Mutations
https://www.cff.org/What-is-CF/Genetics/Know-Your-CF-Mutations/

Microbiology
Additional Clinical Initiatives
https://www.cff.org/Pipeline
Staying Healthy with CF
https://www.cff.org/GermSmart

CF Care Guidelines – Infection Prevention and Control
https://www.cff.org/Infection-Prevention-and-Control-Care-Guidelines/
Information about Nontuberculosis Mycobacteria (NTM)
https://www.cff.org/NTM

Growth and Nutrition
Nutrition and Cystic Fibrosis
https://www.cff.org/Living-with-CF/Treatments-and-Therapies/Nutrition/
WHO Growth Standards and Growth Charts
www.cdc.gov/growthcharts/who_charts.htm

Treatments Used for Individuals with CF
Drug Development Pipeline
https://www.cff.org/Pipeline
About Clinical Trials
https://www.cff.org/Our-Research/Clinical-Trials/
Information about CF and Exercise
https://www.cff.org/Living-with-CF/Treatments-and-Therapies/Fitness/
Therapies for Cystic Fibrosis
https://www.cff.org/Living-with-CF/Treatments-and-Therapies/
Chronic Medications: Guidelines for Lung Health Webcast
https://www.cff.org/Basics-of-Lung-Care-Webcast

Lung Health
CF Care Guidelines – Respiratory
https://www.cff.org/Respiratory-Care-Guidelines
Chronic Medications: Guidelines for Lung Health Webcast
https://www.cff.org/Basics-of-Lung-Care-Webcast

Complications
CF and Depression and Anxiety
https://www.cff.org/Living-with-CF/Emotional-Wellness/
CFRD
https://www.cff.org/Living-with-CF/Cystic-Fibrosis-Related-Diabetes/
Nutrition and Bone Health
https://www.cff.org/PDF-Archive/Bone-Health-and-Cystic-Fibrosis/
Sinus Disease
https://www.cff.org/PDF-Archive/Partnering-for-Care-Slides--Sinus-Disease-in-CF/

Transplantation
Information on Lung Transplantation
https://www.cff.org/Living-with-CF/Lung-Transplantation/

Survival
About Cystic Fibrosis
https://www.cff.org/What-is-CF/About-Cystic-Fibrosis/

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 into the registry.

SUGGESTED CITATION: Cystic Fibrosis Foundation Patient Registry
2014 Annual Data Report
Bethesda, Maryland

COVER PHOTO: Cade Martin Photography

If you have any questions about CF care, please talk with your CF care center or contact the Foundation at **1.800.FIGHT.CF** *or* **info@cff.org**.

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