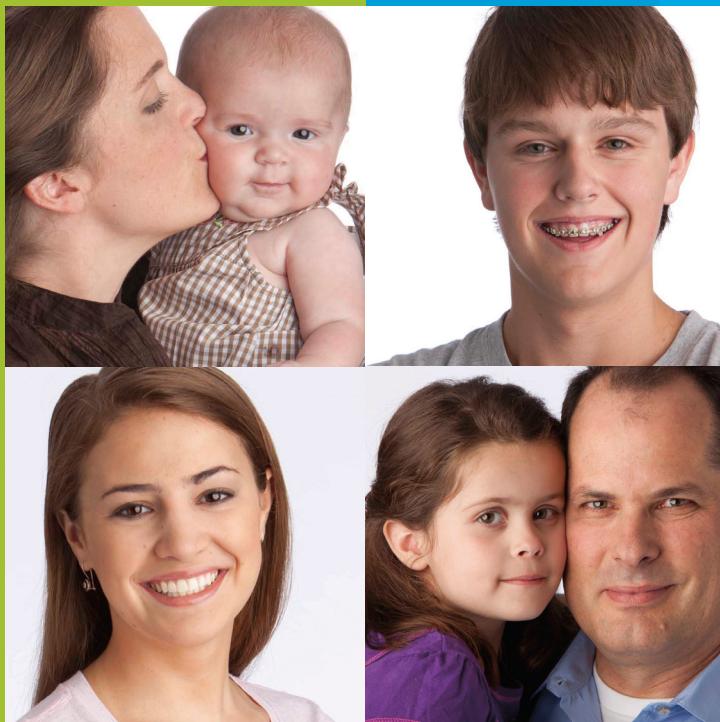


# Patient Registry

Annual Data Report

# 2011



THE CF FOUNDATION WISHES TO THANK  
the people with CF and their families who  
contributed their photos to this report.

PHOTOGRAPHY BY

- Cade Martin Photography
- Dakota Fine
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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 in 2011.

SUGGESTED CITATION

Cystic Fibrosis Foundation Patient Registry  
2011 Annual Data Report  
Bethesda, Maryland  
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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](mailto:info@cff.org).



*To the CF Community and Friends,*

Each year, the Cystic Fibrosis Foundation collects information on the health status of the more than 27,000 people with CF who receive care at Foundation-accredited centers across the country.

This information is shared with the wider community through the CF Foundation's *Patient Registry Annual Data Report*, highlighting progress, trends and areas for improvement in CF care.

Based on the most recent data in the Patient Registry, the outlook for people with CF continues to improve:

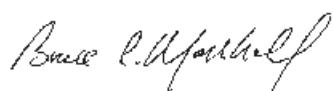
- Adults and children with CF are experiencing better overall lung function.
- Nutrition and growth in people with CF has improved, with most children who have CF at or near the median body mass index (BMI) percentile for their age.
- Thanks to nationwide newborn screening, greater numbers of infants are now diagnosed with CF in the first year of life.
- Close to half of all people with CF are now ages 18 and older.

These steady gains reflect the strong partnerships in the CF team, made up of people with CF, their families and care center staff. We hope the information in this report helps to strengthen the CF team by serving as a tool for shared learning and a springboard for open communication and continued improvement.

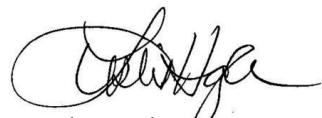
People with CF and their families are experts in living with the day-to-day challenges of the disease. Your input and active partnership with your care center are key to continued advances in CF care and improved health for those living with the disease. We encourage you to use this report to start a conversation with your CF care center to improve CF care.

We are deeply grateful to the people with CF and their families who generously agree to share their data. We look forward to working together to add more tomorrows for everyone with CF.

Sincerely,



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



Leslie Hazle, M.S., R.N.  
Director of Patient Resources



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## ABOUT CYSTIC FIBROSIS AND THE CF FOUNDATION

### WHAT IS CYSTIC FIBROSIS?

CF is a life-threatening genetic disease that causes mucus to build up and clog some of the body's organs. This leads to multiple problems, especially in the lungs and digestive system.

People with CF often get lung infections and have inflammation or swelling in the lungs. Over time, this leads to damage and lower lung function. CF also can make it hard for the body to absorb nutrients in food. This makes it difficult for a person with CF to grow normally and maintain a healthy body weight.

About one in 3,500 babies is born with CF each year in the United States. CF affects all racial and ethnic groups, but is more common among white people (Caucasians). An estimated 30,000 people in the United States have the disease.

### WHAT IS THE CYSTIC FIBROSIS FOUNDATION?

The CF Foundation was founded in 1955 by parents of children with CF. The mission of the Foundation is to assure the development of the means to cure and control CF and to improve the quality of life for those with the disease.

The Foundation accredits a network of more than 110 CF care centers across the United States to meet the health care needs of people with CF. The Foundation provides care centers with:



- Grants to give some funding for care and research.
- Training in quality improvement.
- CF care guidelines based on research published in medical journals.
- Updates on CF care and research at the annual North American CF Conference.
- Education and advocacy resources for people with CF and their families.
- Patient Registry data to track the health of people with CF.

The Foundation also supports the work of researchers to discover and develop new therapies to improve the length and quality of life for those with the disease. Developing new CF drugs takes time and is expensive. To help offset the costs and speed up the process, the Foundation supports drug discovery as well as later-stage clinical trials.

A top research priority of the Foundation is to find drugs that target the underlying cause of CF. The Foundation also supports the development of therapies that treat the symptoms of the disease. To speed up the development of all potential new therapies, CF Foundation-accredited care centers do clinical research.

Learn more about CF, the Foundation, its research and its drug development pipeline at [www.cff.org](http://www.cff.org).

## WHAT IS THE CYSTIC FIBROSIS FOUNDATION'S PATIENT REGISTRY?

The Cystic Fibrosis Foundation's Patient Registry tracks the health and treatments of people with cystic fibrosis (CF) across the United States. Information is collected every year on more than 27,000 people who receive care at CF Foundation-accredited care centers (adult, affiliate and pediatric programs) and agree to participate in the Registry. The data includes state of residence, height, weight, gender, CF mutations, pulmonary function test (PFT) results, medication use and problems (complications) related to CF.

The Patient Registry gives health care professionals and researchers the information they need to:

- Improve the delivery of care.
- Study the effects of treatments on people with CF.
- Develop care guidelines.
- Design clinical trials to test new therapies.

The Registry helps people with CF and their families, as well as health care professionals, compare the overall health of those receiving care at one CF center with all other centers nationwide. It is vital for everyone with CF to agree to have their data entered into the Registry. The participation of all people with CF provides data that are more complete and helps create a better picture of the current state of CF care. This data can also be used to find areas where more work can be done to improve the health of those with the disease.

The CF Foundation developed the following seven goals focused on improving the health of people with CF. This report highlights these goals and the work being done to meet them.

The goals are:

**Goal 1:** People with CF and their families will be full members of the care team.

Communication will be open so everyone can be involved in care decisions.

Care will be respectful of the person with CF's needs, preferences and values.

**Goal 2:** Children, adolescents and adults with CF will have normal growth and nutrition.

**Goal 3:** People with CF will receive appropriate therapies for maintaining lung function.

Pulmonary exacerbations will be detected early and treated aggressively to return people with CF to their previous levels of lung function.

**Goal 4:** People with CF, their families and CF care center staff will be well-informed and active partners in reducing the spread of germs, particularly *Pseudomonas aeruginosa* (*Pseudomonas*) and *Burkholderia cepacia* (*B. cepacia*) complex.

**Goal 5:** People with CF will be screened and managed aggressively for complications of the disease, particularly CF-related diabetes (CFRD).

**Goal 6:** People with CF and their families will be supported by their CF care center when facing decisions about transplantation and end-of-life care.

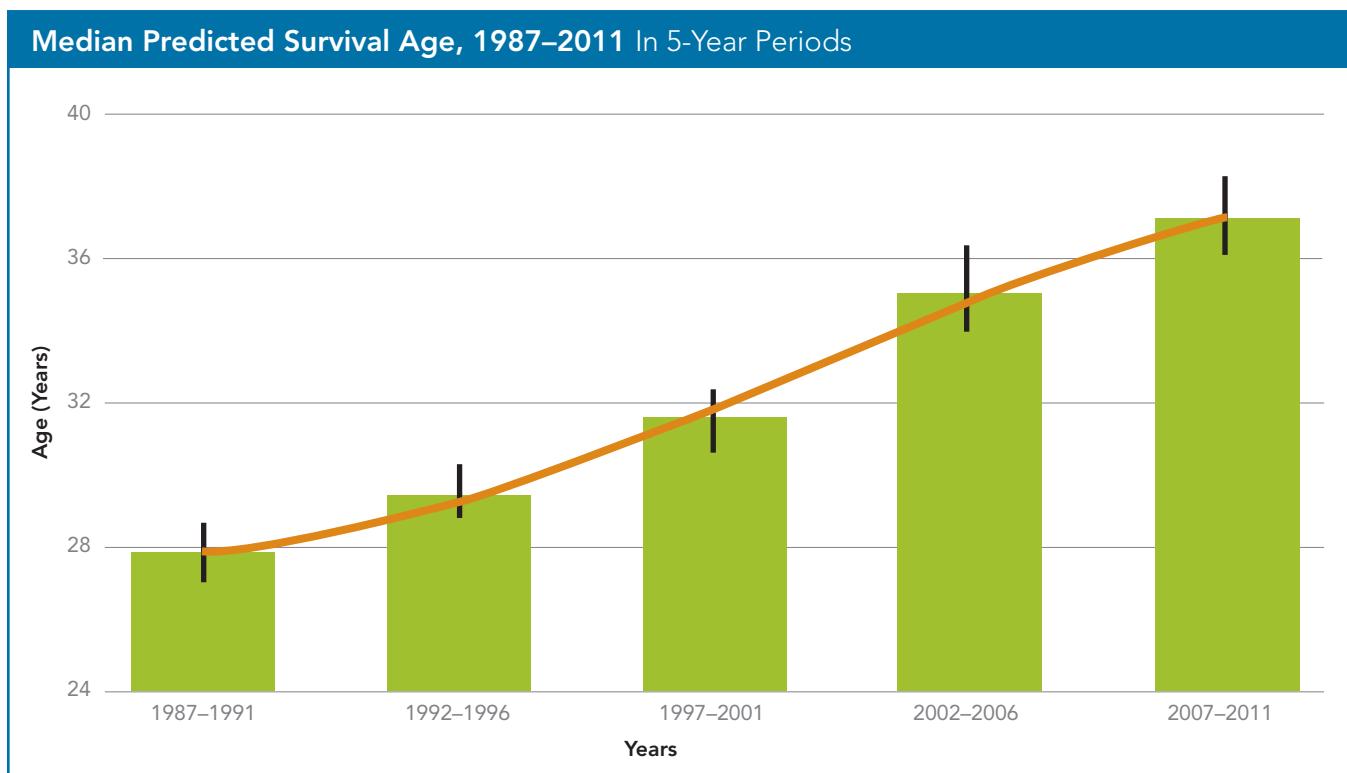
**Goal 7:** People with CF and their families will have access to appropriate therapies, treatments and support regardless of race, age, education or ability to pay.

## OVERVIEW OF THE HEALTH OF PEOPLE WITH CF

The Patient Registry is a tool used to measure the improvements in the health and survival of people with CF in the United States. This section is an overview of the data from diagnosis through adulthood.

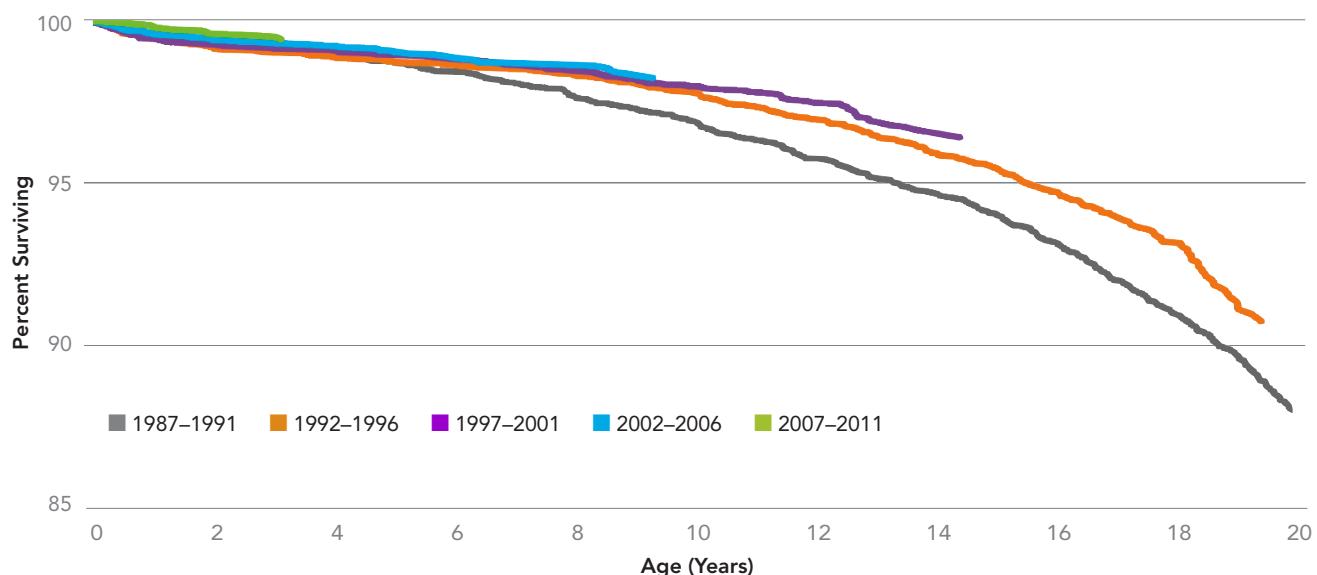
The median predicted age of survival for people with CF has risen steadily over the last 25 years. The chart below shows the rise in median predicted survival. In 2011, the age was 36.8 years. This is less than what it was in 2010. However, year-to-year fluctuations are normal when measuring health outcomes for any disease or medical condition. What is important is the steady increase in survival over time.

Median predicted age of survival is calculated using a standard method called life table analysis. This is best known for its use in the life insurance industry. Using this calculation, half of the people in the patient registry are expected to live beyond the median predicted survival age, and half are expected to live less than the median predicted survival age.



The graph on the next page shows another way to look at how survival is improving. Of people with CF born between 1987 and 1991 (gray line), 94.7 percent were alive at age 14. For children with CF born between 1997 and 2001 (purple line), 96.6 percent were alive at age 14. The younger the group of children with CF, the better they are doing. There are many reasons for this increase in survival. A key reason is people with CF, their families and CF care center working together to improve CF care.

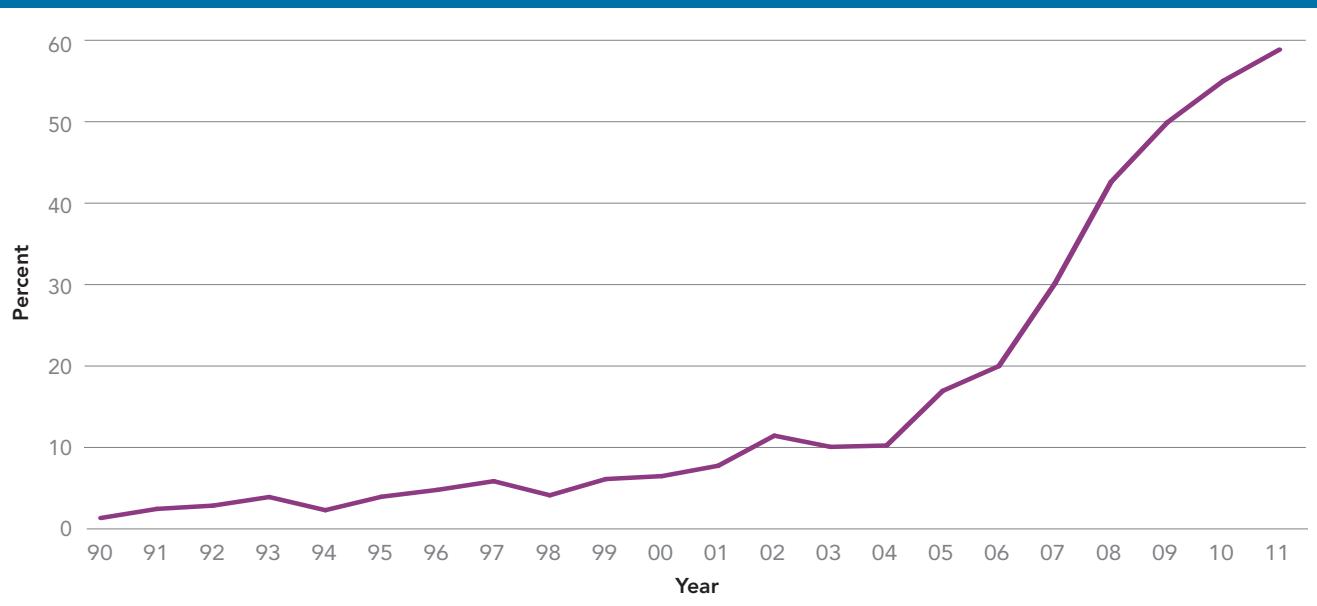
## Survival by Birth Cohort



It is thought that early diagnosis may play an important role in improving survival. Since 2010, all 50 states and the District of Columbia screen all newborns for CF. Research shows that people with CF who are diagnosed because of newborn screening have better weight and healthier lungs later in life than those diagnosed because of symptoms.

The earlier CF is diagnosed the sooner treatment can begin. As shown in the graph below, almost 60 percent of those diagnosed with CF in 2011 were diagnosed because of CF newborn screening.

## Percent Diagnosed Because of Newborn Screening



To help CF doctors and nurses care for babies with CF diagnosed because of newborn screening, the Foundation worked with experts in CF and infant care to develop care guidelines. The CF infant care guidelines outline CF care to keep babies with CF as healthy as possible. To read the Foundation's care guidelines, visit [www.cff.org/treatments/CFCareGuidelines/AgeSpecificCare](http://www.cff.org/treatments/CFCareGuidelines/AgeSpecificCare).

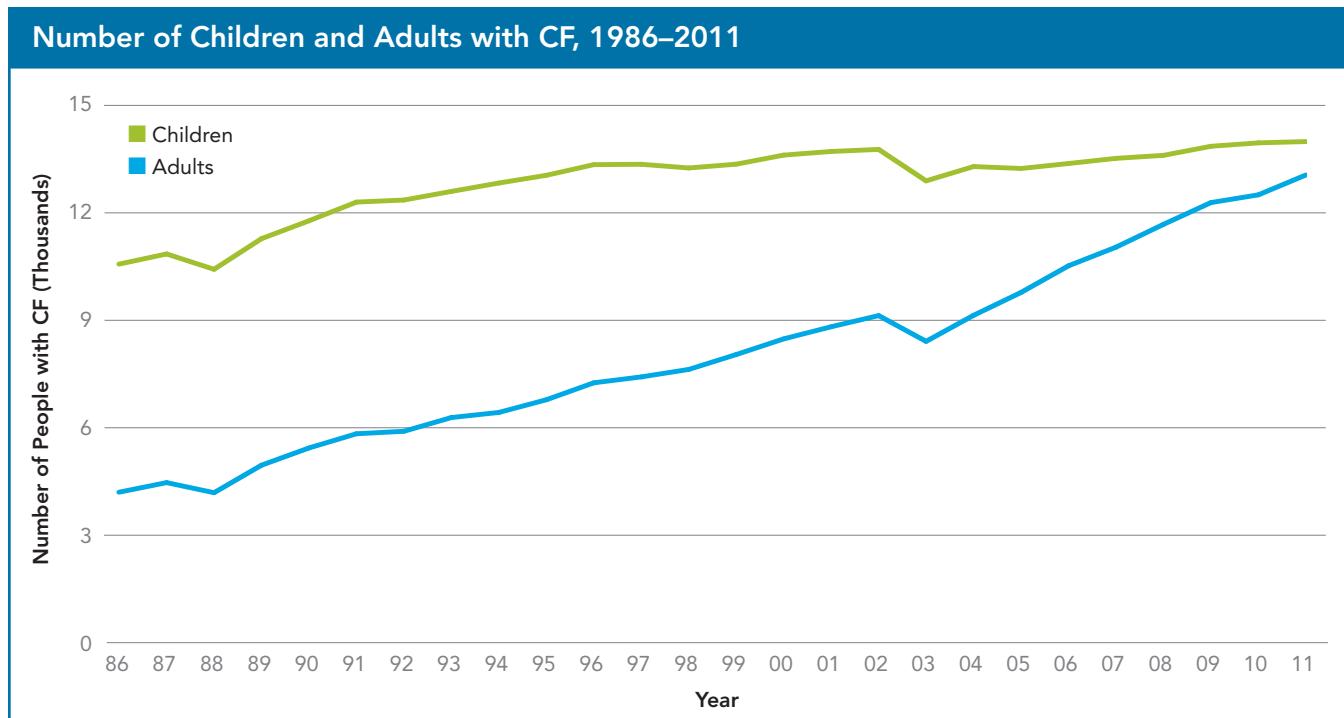
Research also shows that infants with CF still may have poor growth even with early diagnosis and treatment. The Foundation is supporting research to find out why. For example, the Baby Observational and Nutrition Study (BONUS) is looking at what might interfere with good growth in a baby with CF. To learn more visit [www.cff.org/research/ClinicalResearch/Find](http://www.cff.org/research/ClinicalResearch/Find), click on "Advanced Search" and type in keyword "infant" to find this and other studies for infants with CF.

Although many people with CF are diagnosed because of newborn screening, other children and adults are still diagnosed each year because of symptoms. You can learn more about the testing for CF at [www.cff.org/AboutCF/Testing](http://www.cff.org/AboutCF/Testing).

### ADULTS WITH CF

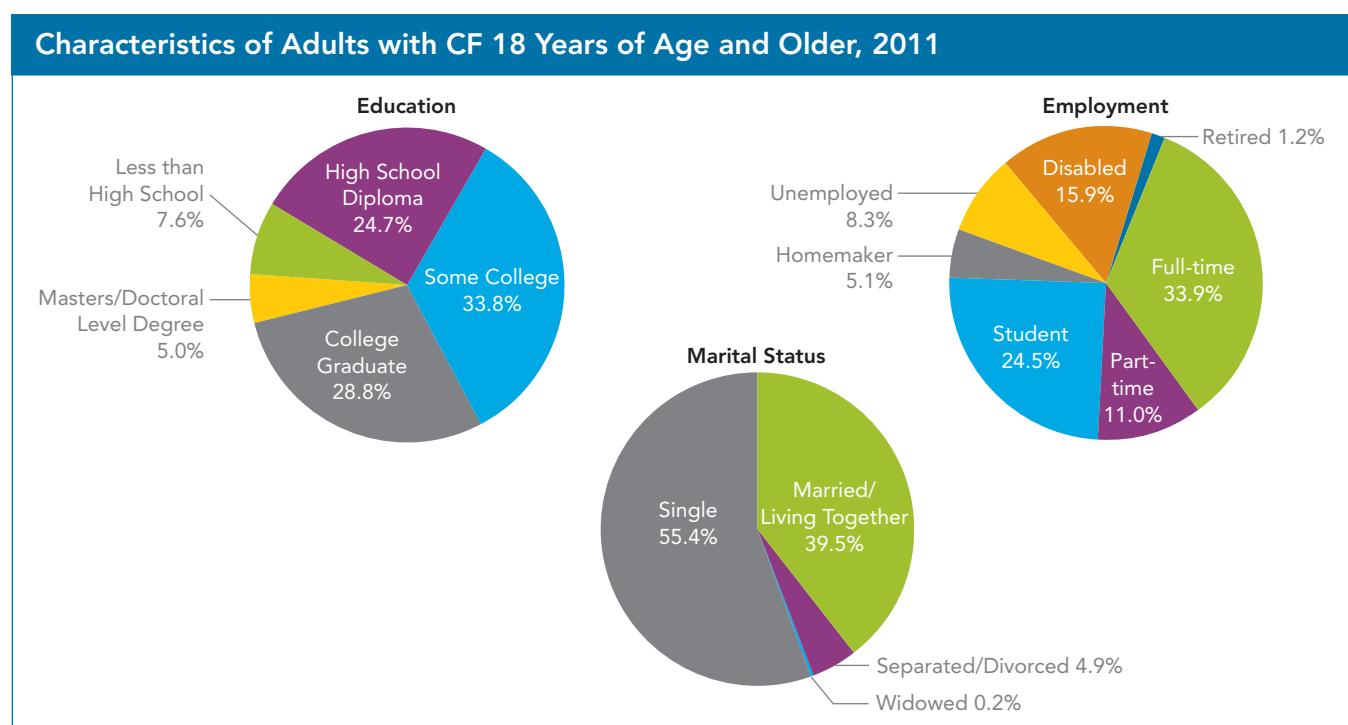
The graph below shows how fast the number of adults with CF is growing in relation to the number of children with CF in the Patient Registry. The data show that today more people with CF are living well into adulthood. In 2001, the oldest person with CF was 74 years old. In 2011, the oldest was 81 years.

In 1991, over 32 percent of people in the Patient Registry were ages 18 years or older. In 2011, more than 48 percent of people with CF in the Patient Registry were adults, and that number continues to grow. The CF Foundation supports adult programs in CF care centers. One way it does this is by helping to support more doctors as they learn to care for adults with CF. To learn more about CF in adults, visit the "For Adults with CF" pages at [www.cff.org/Adults](http://www.cff.org/Adults).



It is important to help teens with CF as they move toward taking charge of their own health. The care center can help teach children, teens and adults how to manage CF. To learn more about adult CF care and the transition from pediatric to adult care, watch the “Partnering for Care” webcast series at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/PartneringCare](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/PartneringCare). You can also read the guidelines on adult CF care at [www.cff.org/treatments/CFCareGuidelines/AgeSpecificCare](http://www.cff.org/treatments/CFCareGuidelines/AgeSpecificCare).

The charts below show that many adults with CF are leading active lives.



## CF AND FERTILITY

Many adults with CF wish to have children. In the 1980s, it was thought to be too risky for a woman with CF to get pregnant and impossible for a man with CF to biologically father a child. Now, thanks to better nutrition and lung function, many women with CF are able to have a healthy pregnancy. Advances in fertility medicine have also given men with CF the option to father children.

In 2011, the Patient Registry reported that 211 women with CF were pregnant. To learn more about male and female fertility issues in CF, watch the webcasts at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/AgeSpecific](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/AgeSpecific). Talk with your CF care center about what people with CF should think about before starting a family.

## CF GENETICS

People with CF should know which mutations they have. According to the Patient Registry, 91.8 percent of people with CF have had at least one CF mutation identified. If you do not know if you or your child has had your CF mutations identified, talk to your CF care center.

The table below lists the most common CF mutations. Almost 87 percent of people with CF have at least one copy of the F508del (Delta F508) mutation, and about 47 percent have 2 copies.

While the table below shows the most common mutations, remember, there are more than 1,800 known mutations of the CF gene. The Foundation is supporting research to find out which of these mutations cause CF. Learn more about CF genetics, the research and specific CF mutations, visit [www.cff.org/AboutCF/Testing/Genetics](http://www.cff.org/AboutCF/Testing/Genetics).

The Foundation is working with companies to develop drugs that target the underlying cause of CF. Much of this research is looking at specific mutations, or genotypes. In January 2012, Kalydeco™ was approved for people with CF ages 6 years and older who have at least one copy of the G551D mutation. Research showed that people with G551D who took the drug had major improvements in lung function and weight, compared with those who did not take the drug. For the first time, a drug also lowered the sweat chloride level of people with CF. You can read more about Kalydeco at [www.cff.org/treatments/Therapies/Kalydeco](http://www.cff.org/treatments/Therapies/Kalydeco).

The Foundation continues to support research to speed up the development of drugs that help people with the F508del (Delta F508) and other CF mutations. Find out more about this research, called “CFTR Modulation,” on the CF Foundation’s drug development pipeline at [www.cff.org/research/DrugDevelopmentPipeline](http://www.cff.org/research/DrugDevelopmentPipeline).

You can also sign up to get email alerts when clinical trials are posted on the Web and when results of a trial are available at [www.cff.org/research/ClinicalResearch/Find/ClinicalTrialAlerts](http://www.cff.org/research/ClinicalResearch/Find/ClinicalTrialAlerts).



**Most Common CF Mutations, 2011**

Mutation	Percentage of People with One or Two of the Mutation
F508del	86.8
G542X	4.6
G551D	4.4
R117H	2.7
N1303K	2.5
W1282X	2.4
R553X	1.8
621+1G->T	1.7
1717-1G->A	1.6
3849+10kbC->T	1.5
2789+5G->A	1.3
3120+1G->A	1.0

## MEETING THE GOALS OF CF CARE

The Foundation works to improve CF care based on the seven goals it has developed (see page 5). The data in the Registry shows how the work being done in CF care centers helps people with CF and their families reach these goals.

### Goal 1: FULL MEMBERS OF THE CF CARE TEAM

People with CF and their families will be full members of the care team. Communication will be open so everyone can be involved in care decisions. Care will be respectful of the person with CF's needs, preferences and values.

The CF Foundation's Patient Registry creates reports about:

- The health of each person with CF.
- All people seen at a CF Foundation-accredited care center.
- The health of everyone with CF throughout the United States.

These reports help the full CF team find new ways to improve care. The sample *Patient Summary Report* on page 12 is a snapshot of a person's lung health, weight and other information. People with CF and families should ask their CF care center for a copy of their *Patient Summary Report* at clinic visits.

When looking at your or your child's *Patient Summary Report*, think about asking your CF care center the following questions:

- What does the data mean?
- How can we work together to have the best possible health for me or my child with CF?
- What can we do as a team to improve care at the CF center?

Learn more about the different ways you can be a full member of your CF care team.

Watch the webcasts series on "Partnering for Care" and "Quality Improvement" at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts). Read about how others have worked with their care centers to improve care at [www.cff.org/LivingWithCF/QualityImprovement](http://www.cff.org/LivingWithCF/QualityImprovement).

### CF Care Guidelines

Care or practice guidelines are used in health care for many different diseases and conditions. These guidelines recommend treatments for health care professionals to use when caring for patients with specific diseases or conditions, based on published medical research.

To develop CF care guidelines, the Foundation gathers CF health experts, adults with CF and parents of children with CF to review the medical literature as well as data from the Patient Registry. These guidelines are about diagnosis, nutrition, lung health, infection control and age-specific care. Learn more about CF care guidelines at [www.cff.org/treatments/CFCareGuidelines](http://www.cff.org/treatments/CFCareGuidelines).

To help keep people with CF as healthy as possible, the care guidelines list a certain number of clinic visits, throat or sputum cultures and lab tests that they should have every year. The table on page 13 lists these and reports the percentage of people with CF in the Patient Registry who met these care guidelines in 2011, compared with 2010.

## Sample Patient Summary Report

VISIT DATE: _____							
Last Hospitalization:	Date of Birth: /1963						
Last Home IV Therapy:	Genotype: F508del / F508del						
Last Clinical Visit: /2012	Centers Visited: Adult Program						
Current diagnosis: Cystic Fibrosis							
Diagnosis Date: /1964	Symptoms: Acute or persistent respiratory abnormalities						
Sweat Test Date: /1992	Sweat Test Value 107.0 (mmol/L):	Sweat Test QNS: Quantity sufficient					
Last Care Episode Date:							
Number of Segments in Care Episode	Total:	Home IV:	Hospital Stay:				
Reason(s) for Care Episode:							
Culture Results							
Last Culture:							
Positive Tests:	Pseudomonas aeruginosa	Multi-drug Resistant Pseudomonas aeruginosa+	Burkholderia species	Stenotrophomonas maltophilia	MSSA	MRSA	H. influenzae
	/2012 /2011 /2011 /2011 /2011	/2012 /2011 /2011 /2011 /2011			/2012 /2011 /2011 /2011 /2011		
Last Mycobacterial Culture:	/2011						
Mycobacterial Species Detected:							
<p>PFTs - % Predicted</p> <p>■ FEV1 Percent Predicted    ● FVC Percent Predicted    ▲ FEF25-75 Percent Predicted</p>							
Last FEV1 Date: /2012	Last FEV1 Value: 1.95						
Last FVC Date: /2012	Last FVC Value: 3						
Last FEF25-75 Date: /2012	Last FEF25-75 Value: 0.87						
Nutritional Trend							
<p>Percentile</p> <p>■ Weight-for-Length Percentile    ● Height Percentile    ▲ Weight Percentile    ◆ BMI Percentile</p> <p>BMI</p>							
Complications Active at Last Visit:		Complications Previously Noted:					
Routine Evaluations							
Last PFT:	/2012	Last Dietary Visit:	/2011				
Last CXR:		Last LFT:	/2012				
Last SW Visit:	none	Last Glucose Screening:	/2012				
Last RT/PT Visit:	/2010	Last Creatinine:	/2012				

## Care, Screening and Prevention Guidelines for People with CF

Guidelines to be met every year	People with CF Who Met the Guidelines (%)	
	2010	2011
4 or more clinic visits	62.7	63.4
4 or more respiratory cultures (sputum or throat)	44.3	45.9
2 or more pulmonary function tests (PFTs) if 6 years of age or older and physically able	82.0	82.5
An influenza vaccine (flu shot) if 6 months of age or older	88.9	91.0
Fat-soluble vitamin levels measured (Vitamins A, E & D)	82.7	83.3
An oral glucose tolerance test (OGTT) if 10 years of age or older	24.3	31.5
Test to measure liver enzymes in the blood	71.4	77.6

The newest care guidelines focus on the diagnosis and treatment of low vitamin D levels in people with CF. Vitamin D helps keep bones and teeth strong. The *Guide to Bone Health and Disease in Cystic Fibrosis* says that people with CF at the age of 18 years should have a DXA (also spelled DEXA) bone density scan to measure the thickness of the bone. If the bone is thick enough, then a DXA scan should be done every 5 years. But if the bone is thin or less dense (called osteoporosis), a DXA scan should be done every 2 to 4 years.

Ask your CF care center what you can do to keep your or your child's bones healthy and strong. Learn more about bone health and vitamin D in CF at [www.cff.org/treatments/CFCareGuidelines/Nutrition](http://www.cff.org/treatments/CFCareGuidelines/Nutrition).

### Goal 2: NORMAL GROWTH AND NUTRITION

**Children, teens and adults with CF will have normal growth and nutrition.**

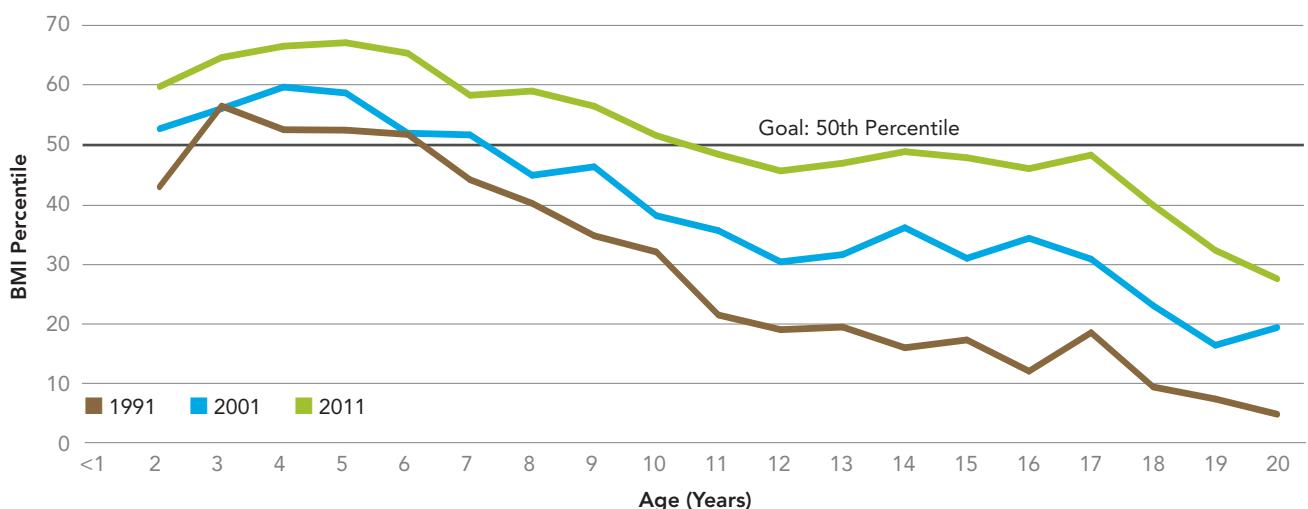
The nutrition of people with CF is better today than it was 10 years ago. But there is more work to be done so their nutrition can continue to improve. The care guidelines set goals for body mass index (BMI) for children and adults with CF. BMI is based on a person's weight and height. BMI is calculated by dividing body weight in kilograms by the person's height in meters squared (weight kg/height m<sup>2</sup>=BMI).

For children and teens with CF, BMI is stated as a percentile compared with children in the United States of the same age and gender who do not have CF. The goal is for children with CF to grow and develop at the same rate as children without CF. The average BMI percentile for children in the United States who do not have CF is around the 50th percentile for their age. This is the goal for children with CF too.

To calculate a child's BMI percentile or an adult's BMI, visit [www.cdc.gov/healthyweight/assessing/bmi](http://www.cdc.gov/healthyweight/assessing/bmi).

The next graph shows how much the BMI percentile of children with CF has improved since 1991. It also shows that more work can be done to improve the BMI percentile of children with CF older than 10 years. This graph starts at age 2 because the growth of younger children is measured as weight for length on a growth chart. These charts are available at [www.cdc.gov/growthcharts/clinical\\_charts.htm](http://www.cdc.gov/growthcharts/clinical_charts.htm).

## Median BMI Percentiles by Age 1991, 2001 and 2011

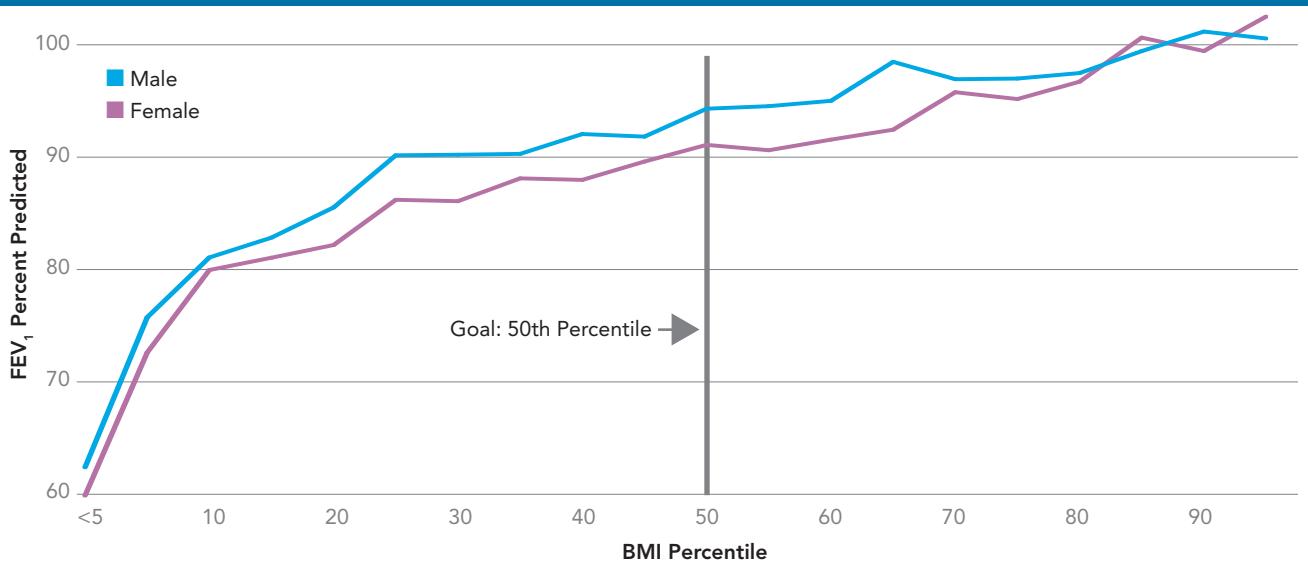


Normal growth and nutrition are important because the Patient Registry data show a strong association between a higher BMI percentile and better lung function in children with CF. Pulmonary function tests (PFTs) measure how well the lungs work. Lung damage is caused by inflammation and infections that injure the lungs and airways and may lower PFTs. With every lung infection, a little more damage may happen.

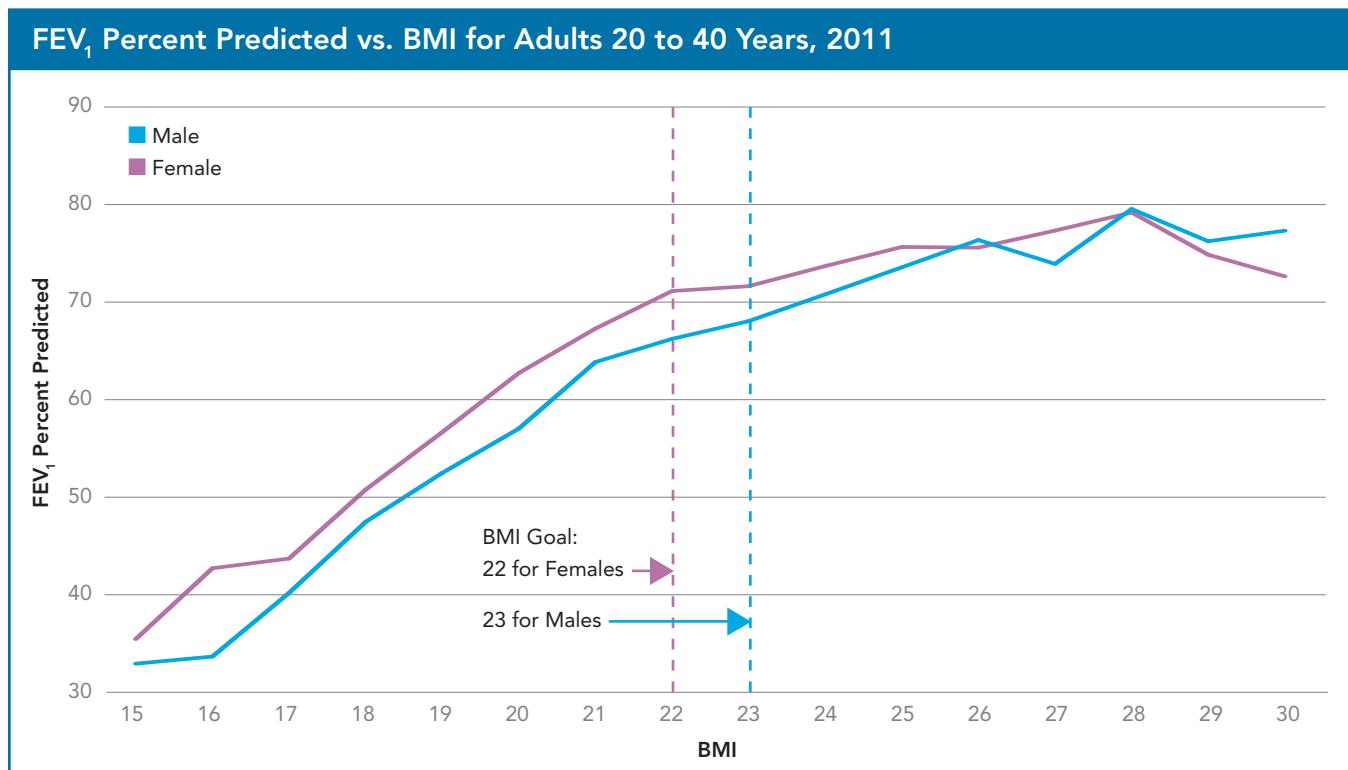
FEV<sub>1</sub> is one measure of pulmonary function. It measures the forced exhaled volume in the first second. It is shown as a percent predicted, based on the FEV<sub>1</sub> of healthy, non-smoking people of the same age, height and gender. The following graphs show that good nutrition and lung health seem to go hand in hand.

The graph below shows the association between higher BMI percentile and better lung function in children ages 6 through 19 years. The bar in the middle is the BMI 50th percentile goal for children with CF.

## FEV<sub>1</sub> Percent Predicted vs. BMI Percentile for Children 6 to 19 Years



The graph below shows the association between higher BMI and better lung function for adults with CF. The BMI goals are 23 for males and 22 for females.



To help you improve or maintain your or your child's growth and nutrition, the Foundation suggests that you work closely with your CF care center to reach these goals. Learn more about CF and nutrition at [www.cff.org/LivingWithCF/StayingHealthy](http://www.cff.org/LivingWithCF/StayingHealthy) or ask your CF care center. You can also watch webcasts about CF nutrition at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Nutrition](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Nutrition).

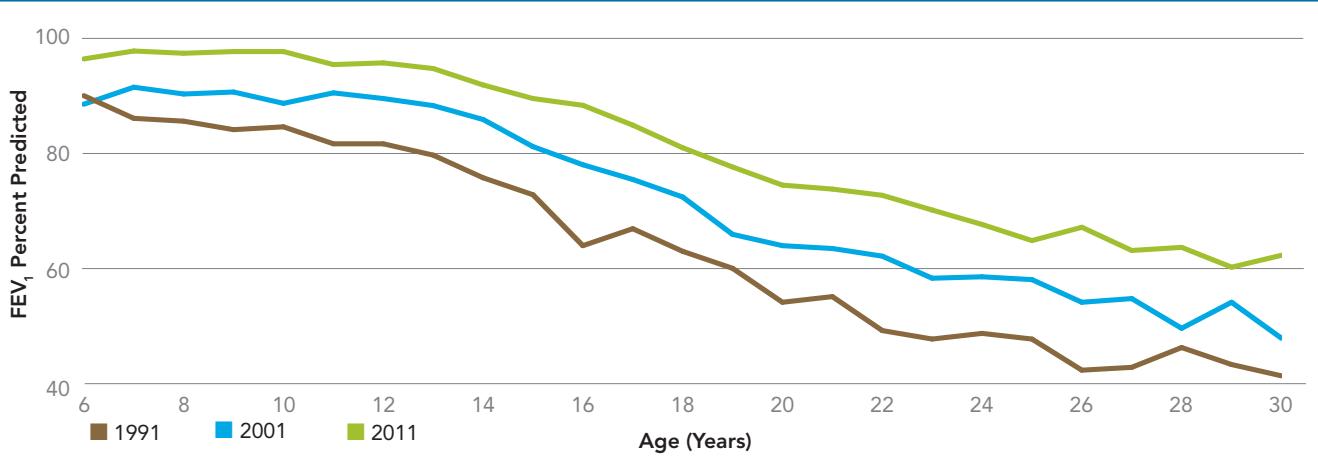
#### Goal 3: MAINTAIN LUNG HEALTH

All people with CF will receive appropriate therapies for maintaining lung function and reducing acute episodes of infection. Pulmonary exacerbations will be detected early and treated aggressively to return lung function to previous levels.

People with CF often have pulmonary exacerbations — when respiratory symptoms get worse and more treatments are needed. This can be caused by infection and inflammation. If you or your child are coughing more, have more sputum, a loss of appetite or energy or more shortness of breath, it may be a pulmonary exacerbation. It is important to call your CF care center to start treatment for an exacerbation early.

The graph at the top of the next page shows that the lung health of people with CF has improved since 1991. FEV<sub>1</sub> is usually in a normal range (90 to 100 percent predicted) when first measured around 6 years of age.

## Median FEV<sub>1</sub> Percent Predicted vs. Age, 1991, 2001 and 2011

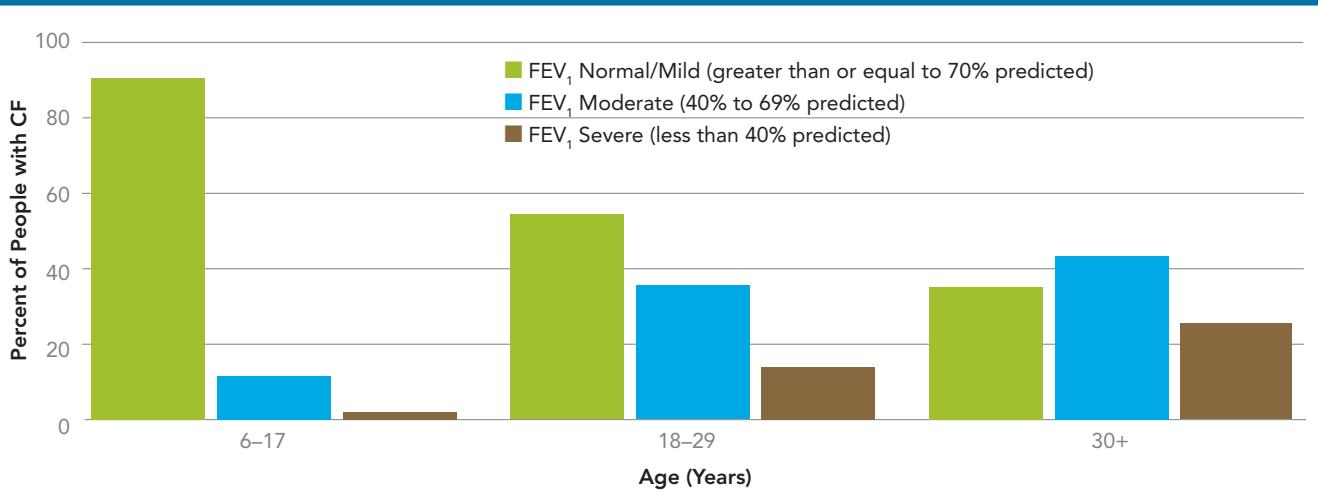


Research shows that people of all ages with CF have some lung damage — even when their lung function is normal. Over time, FEV<sub>1</sub> decreases because of damage from exacerbations. In CF, the lower a person's FEV<sub>1</sub> percent predicted, the more severe the lung disease. Here is a guide to FEV<sub>1</sub> percent predicted and what it may mean for a person with CF:

- FEV<sub>1</sub> greater than or equal to 90 percent predicted is looked at as normal. However, even with normal lung function, some lung disease may be present.
- 70 to 89 percent predicted shows mild lung disease.
- 40 to 69 percent predicted shows moderate lung disease.
- Less than 40 percent predicted is a sign of severe lung disease.

The graph below shows the percentage of people with CF by age group with lung function from normal to severe, based on FEV<sub>1</sub> percent predicted. This is better than in 2001, when 77.2 percent of children with CF had an FEV<sub>1</sub> greater than or equal to 70 percent predicted. In 2011, more than 87 percent of children ages 6 to 17 years had a lung function that shows mild disease or normal lung function.

## Lung Function by Age Group, 2011



## Lung Health Research

Research is ongoing to find new drugs to slow or stop lung function from getting worse, improve FEV<sub>1</sub> and keep the lungs of people with CF as healthy as possible. Learn more about potential new medicines for the lungs on the CF Foundation's drug development pipeline at [www.cff.org/research/DrugDevelopmentPipeline](http://www.cff.org/research/DrugDevelopmentPipeline).

All drugs must be proven safe and effective and approved by the U.S. Food and Drug Administration (FDA) before they can be prescribed. They must be tested in people during clinical trials to make sure they are safe and work. People with CF play a critical role by volunteering to take part in CF clinical trials and research to help find new therapies and learn more about the disease. The Foundation encourages you to learn, ask about and consider joining a CF clinical trial.

You can learn more about how to help find new drugs and therapies to treat CF, and hear personal stories from other people with CF, family members and research teams about participating in CF research, at [www.cff.org/research/ClinicalResearch](http://www.cff.org/research/ClinicalResearch). To watch webcasts about CF research and clinical trials and how you can help find a cure for CF, visit [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Research](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Research).

You can also find CF clinical trials by using the clinical trial search tool at [www.cff.org/research/ClinicalResearch/Find](http://www.cff.org/research/ClinicalResearch/Find).

Sign up to receive clinical trial email alerts when a trial is posted on the Foundation's website and when results of a trial are available at [www.cff.org/research/ClinicalResearch/Find/ClinicalTrialAlerts](http://www.cff.org/research/ClinicalResearch/Find/ClinicalTrialAlerts).

## Therapies for Lung Health

The following table shows the percentage of people with CF who are taking the most prescribed medications for lung health and the criteria for prescribing them. To find out if you or your child might be helped by a daily medication, talk with your CF care center. The *Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Function* lists daily medications that help people with CF take care of their lungs. Learn more about these guidelines at [www.cff.org/treatments/CFCareGuidelines/Respiratory](http://www.cff.org/treatments/CFCareGuidelines/Respiratory).

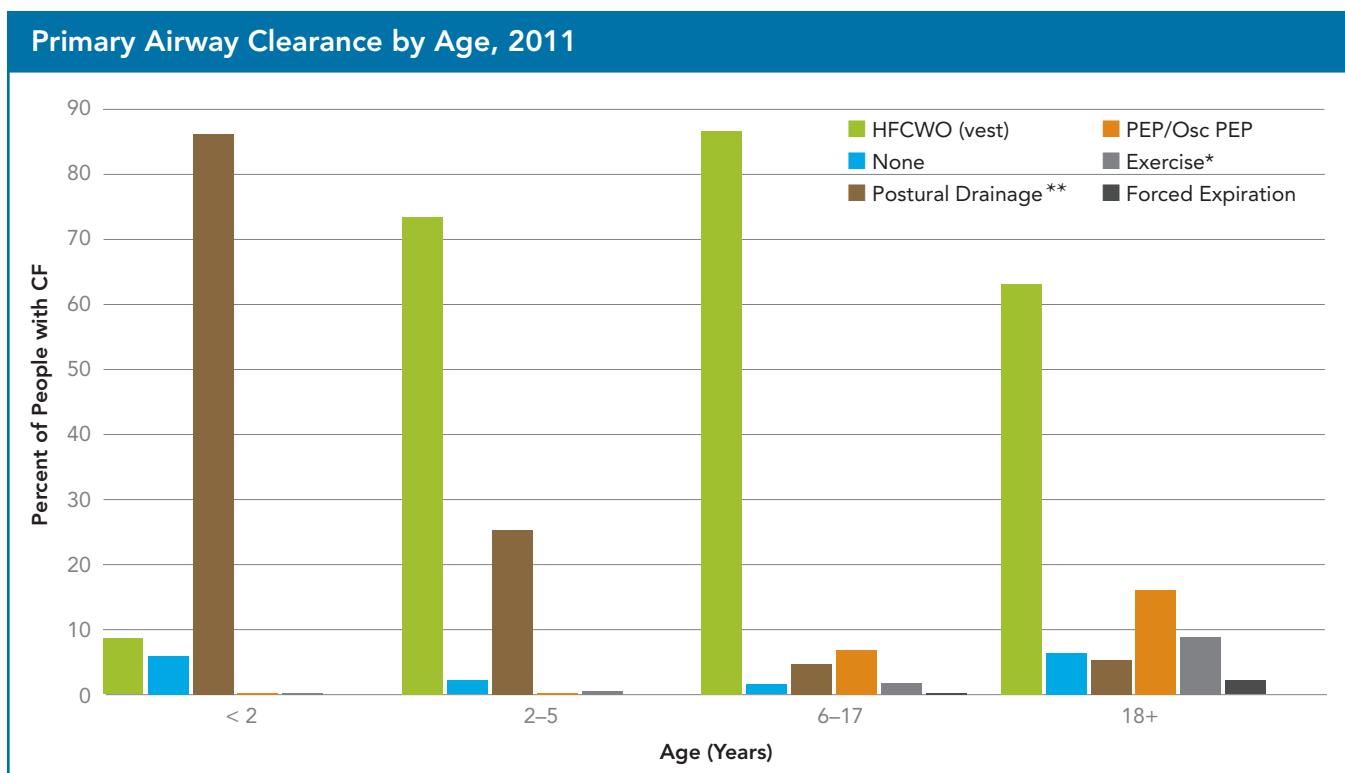
### Percentage (%) of People with CF Who Met the Criteria and Were Prescribed the Drug, 2011

Chronic Medication*	People with CF Prescribed the Drug (%)	Criteria for Prescribing Drug
Dornase alfa (Pulmozyme®)	82.1	<ul style="list-style-type: none"><li>• At least 6 years old</li></ul>
Hypertonic saline	55.2	<ul style="list-style-type: none"><li>• At least 6 years old</li></ul>
Tobramycin solution for inhalation (TOBI®)	65.9	<ul style="list-style-type: none"><li>• At least 6 years old</li><li>• <i>P. aeruginosa</i> in cultures</li></ul>
Aztreonam for inhalation solution (Cayston®)	35.8	<ul style="list-style-type: none"><li>• At least 6 years old</li><li>• <i>P. aeruginosa</i> in cultures</li></ul>
Azithromycin (chronic oral macrolide antibiotic)	70.6	<ul style="list-style-type: none"><li>• At least 6 years old</li><li>• <i>P. aeruginosa</i> in cultures</li><li>• Weight over 25 kg (55 lbs)</li><li>• FEV<sub>1</sub> over 30% predicted</li></ul>

Learn more information about medications and treatments for people with CF at [www.cff.org/treatments/Therapies](http://www.cff.org/treatments/Therapies).

Medication alone can't keep CF lungs healthy. Getting the thick mucus out of the lungs is key. Airway clearance techniques (ACTs) help move mucus out of the lungs. The body's normal and basic ACT is a cough. It's a reflex that clears mucus with high-speed airflow. However, in CF, the mucus cannot be cleared by coughing alone. The guidelines for ACT state that people with CF should do airway clearance every day to keep lungs healthy, even when they are feeling well. When sick, airway clearance should be done more often.

People with CF often use one or more types of ACT. The chart below shows the type of ACT used most often by people with CF.



\*\* Postural drainage is also called postural drainage & percussion (PD&P) or chest physiotherapy (CPT).

\*The Cystic Fibrosis Pulmonary Guidelines: Treatment of Pulmonary Exacerbations states that while exercise is important, it does not replace ACT ([www.cff.org/treatments/CFCareGuidelines/Respiratory](http://www.cff.org/treatments/CFCareGuidelines/Respiratory)).

Learn more about the different ways to clear mucus from the lungs, at [www.cff.org/treatments/Therapies/Respiratory/AirwayClearance](http://www.cff.org/treatments/Therapies/Respiratory/AirwayClearance). You can also watch the CF Education webcasts "Airway Clearance Techniques" under Respiratory and "Help Your Respiratory or Physical Therapist Help You Thrive" under Partnering for Care at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts).

## Keeping Lungs Healthy

Each time a person with CF has a pulmonary exacerbation or an infection, lung damage may occur.

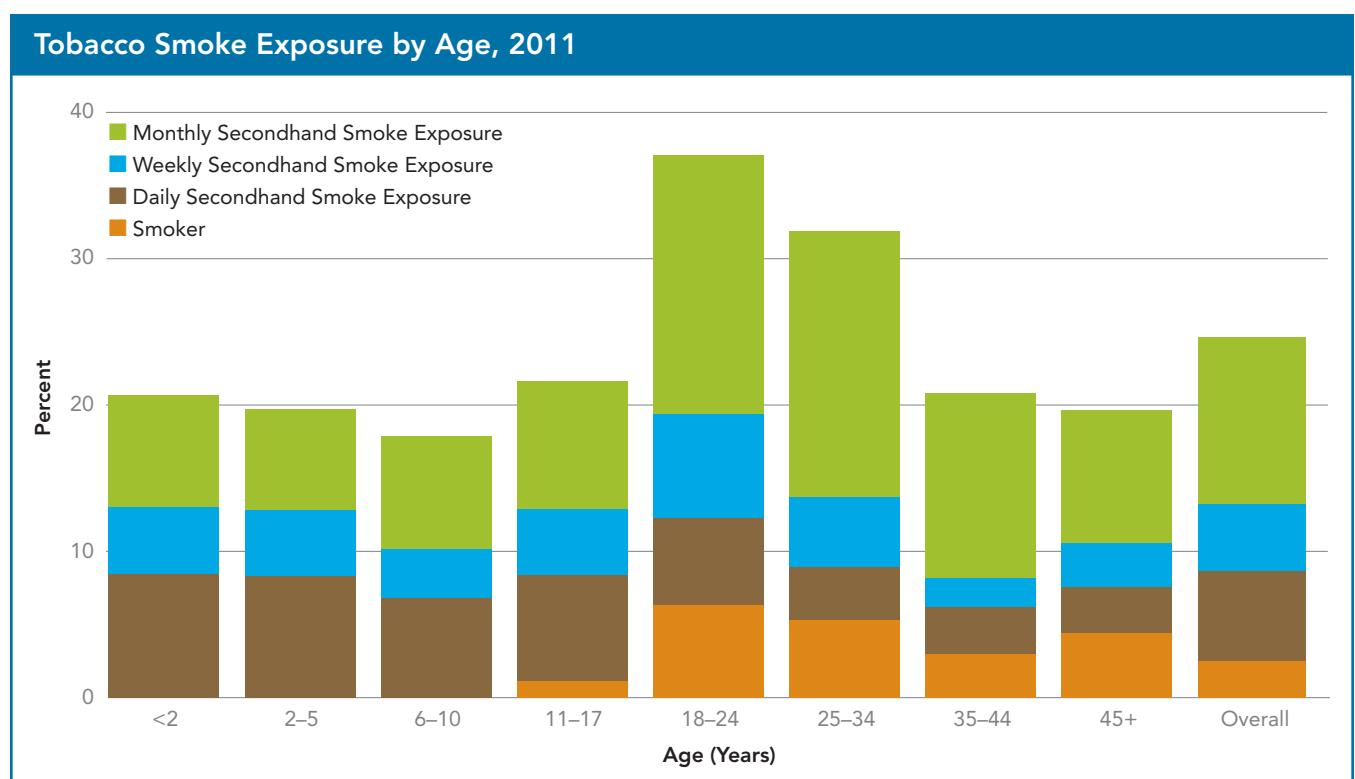
Here are some things you or your child and others can do to keep healthy and reduce the risk of getting sick:

- Do airway clearance every day to keep lungs as clear as possible.
- Take medications as prescribed by your CF care center.
- Clean and disinfect nebulizers after every treatment.
- Get a flu shot every fall for everyone ages 6 months and older with and without CF.
- Avoid germs; see page 20 for ways to reduce respiratory germs.
- Exercise regularly to keep muscles strong.
- Avoid tobacco smoke.

If you have CF and smoke, you are adding to your lung disease and damage. Ask your doctor or CF care center staff to help you quit. People with CF should not smoke and should avoid secondhand smoke to keep their lungs as healthy as possible.

Secondhand smoke — tobacco smoke exhaled by a smoker or produced by burning tobacco — is harmful for people with CF. Research has shown that children without CF who breathe in secondhand smoke have more respiratory infections. For adult non-smokers, secondhand smoke can cause lung cancer and heart disease. The U.S. Surgeon General has concluded that breathing even a little secondhand smoke can be harmful to your health. You can learn more at [www.cdc.gov/tobacco](http://www.cdc.gov/tobacco).

The graph below shows, by age group, the percentage of people with CF who smoke or are exposed to secondhand smoke.





If you smoke and live with someone with CF, think about quitting. In the meantime, only smoke outside and away from others. Keeping your home and car smoke-free can also help you quit smoking.

The only way to protect yourself or your child from secondhand smoke is to be in a 100 percent smoke-free environment. To make a smoke-free environment, you should:

- Not let anyone smoke in your home or car.
- Ask people around you not to smoke, even when outside.
- Stay away from people when they are smoking and teach children to do the same.
- Ask your employer for a smoke-free work and eating space.
- Make sure that your day care center or school is smoke-free.
- Choose restaurants and other businesses that are smoke-free.
- Let owners of businesses that are not smoke-free know that secondhand smoke is harmful to you or your family's health and that it will stop you from going to their businesses.

#### Goal 4: REDUCE RESPIRATORY GERMS

People with CF, their families and CF care center staff will be well-informed and active partners in reducing the spread of germs, particularly *Pseudomonas aeruginosa* (*Pseudomonas*) and *Burkholderia cepacia* (*B. cepacia*) complex.

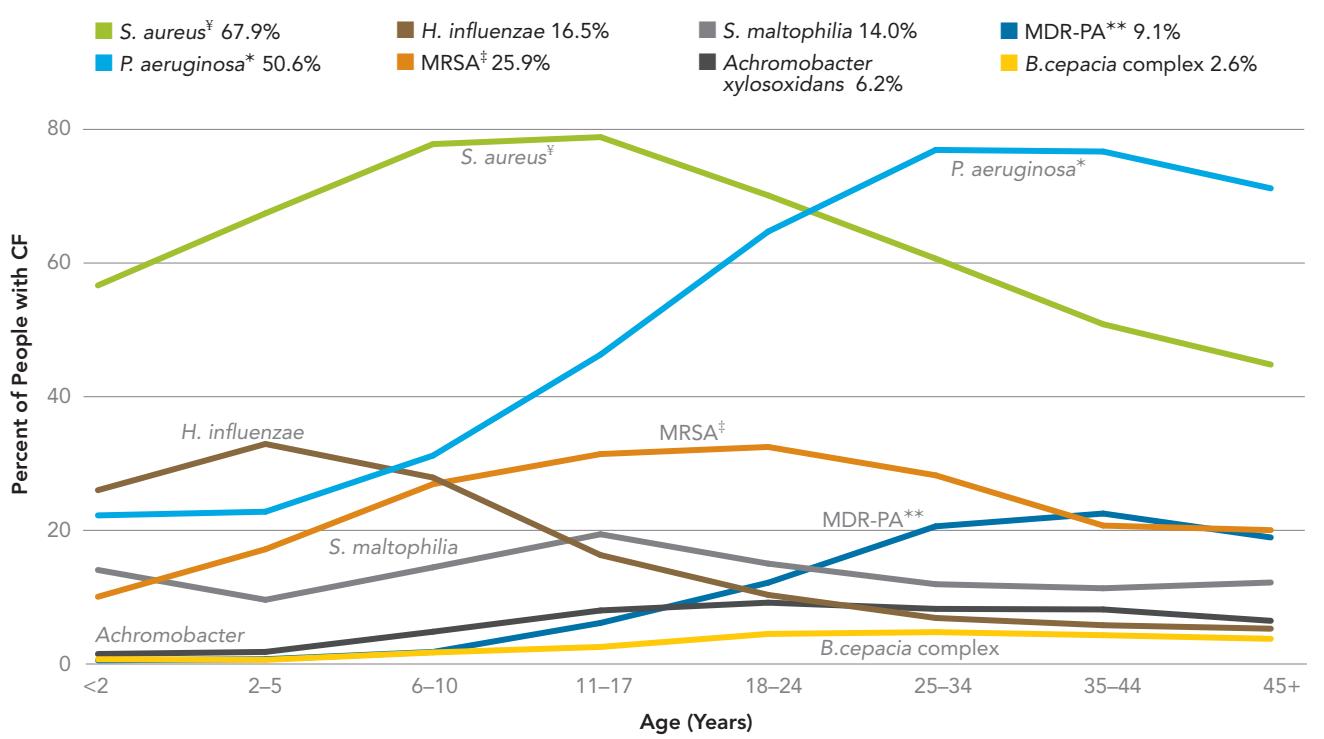
The best way for anyone to reduce germs is to:

- Clean hands often with soap and water or 60 percent alcohol-based hand gels.
- Use a tissue when coughing or sneezing, then clean your hands.
- Limit how much you touch your eyes, nose or mouth.
- Stay away from people who are sick.
- If you are ill, stay away from others.
- Get all immunizations, including a yearly influenza vaccine (flu shot).

Repeated lung infections and exacerbations can cause damage and worsen CF lung disease. When the lungs are damaged, infections happen more often. This is why avoiding germs is important for people with CF.

The graph on the next page shows some of the germs that are found in the lungs of people with CF. Learn more about how to reduce germs by talking to your CF care center.

## Respiratory Germs by Age, 2011



\*P. aeruginosa includes people with MDR-PA.

\*\*MDR-PA is multi-drug resistant Pseudomonas aeruginosa (P. aeruginosa).

<sup>¥</sup>S. aureus includes people with MRSA.

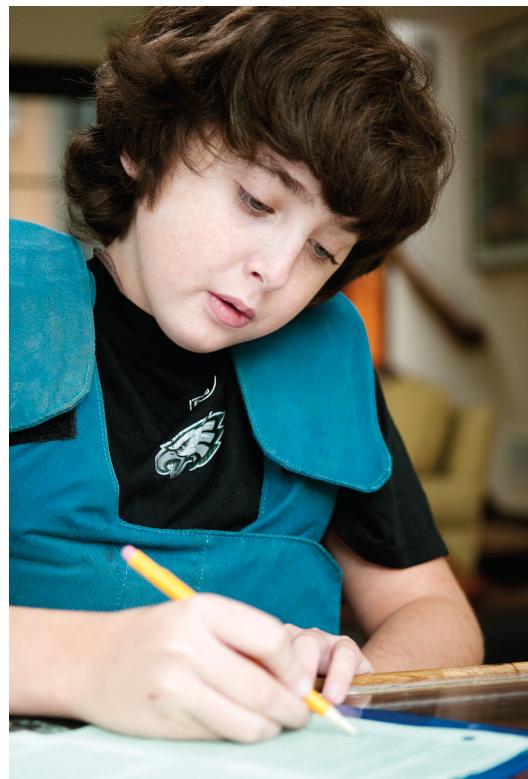
<sup>‡</sup>MRSA is methicillin-resistant Staphylococcus aureus (S. aureus).

The Foundation supports research to learn more about the germs found in the lungs of people with CF. For example, studies are looking at the best way to treat methicillin-resistant *Staphylococcus aureus* (MRSA) in people with CF. Learn more at [www.cff.org/research/ClinicalResearch/Find](http://www.cff.org/research/ClinicalResearch/Find); click on “Advanced Search” and type in the keyword “MRSA.”

The Foundation also supports the *Burkholderia cepacia* Research Laboratory and Repository at the University of Michigan, Ann Arbor. Learn more about *B. cepacia* complex and how it spreads at [www.cff.org/LivingWithCF/StayingHealthy/Germs/Bcepacia](http://www.cff.org/LivingWithCF/StayingHealthy/Germs/Bcepacia).

Watch a webcast and learn about germs, how they affect people with CF and how to avoid them at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Germs](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Germs).

Even if you are careful and do all you can to prevent a lung infection, it's hard to avoid them completely. If you or your child starts to feel ill — for example, are coughing more, have a loss of energy or appetite — call your CF care center so treatment can be started as soon as possible.



## Goal 5: COMPLICATIONS RELATED TO CF

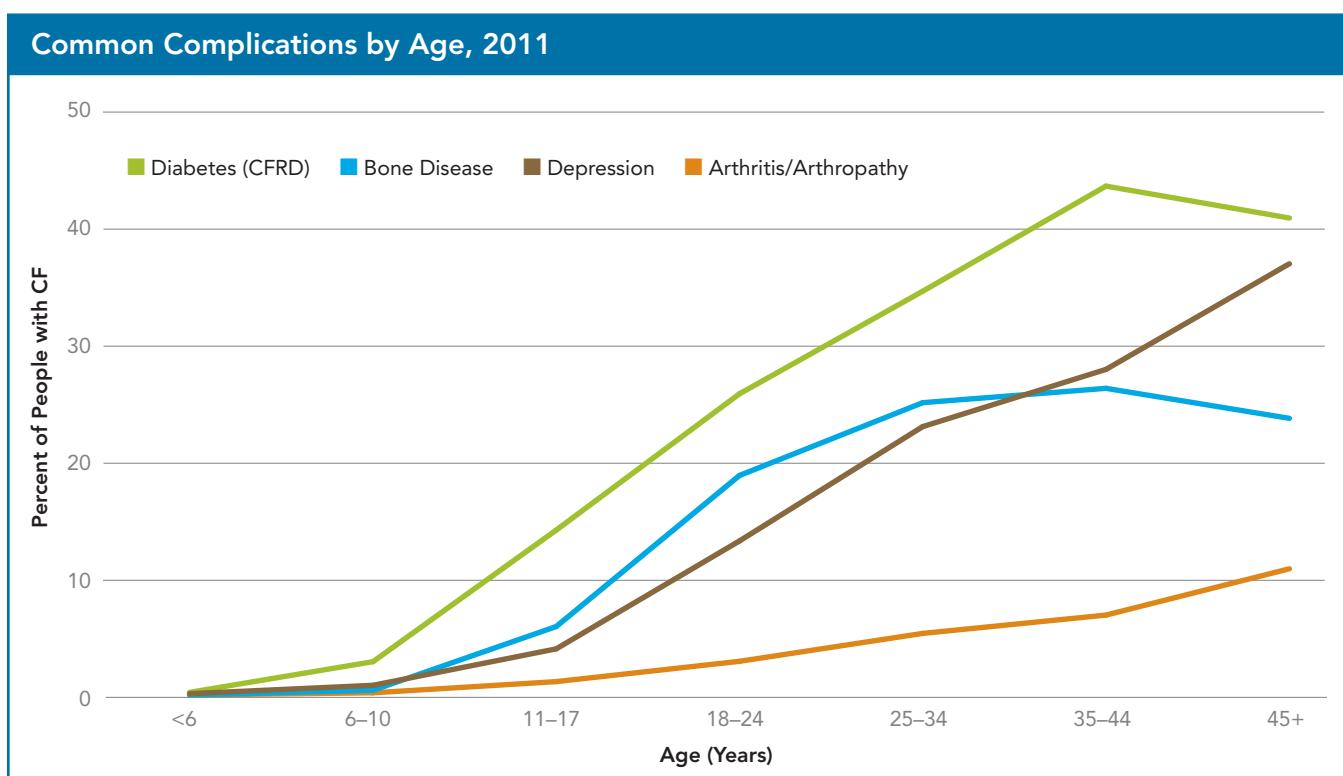
People with CF will be screened and managed aggressively for complications of the disease, particularly CF-related diabetes.

Complications are problems related to CF such as cystic fibrosis-related diabetes (CFRD).

CFRD is one of the most common CF complications. It is different from diabetes in people without CF. In 2011, 19 percent of people of all ages in the Patient Registry had CFRD, and 32.6 percent of people ages 18 years and older had CFRD.

People with CF ages 10 years and older should be tested every year for CFRD. The test is called an oral glucose tolerance test (OGTT). In 2011, 31.5 percent of people with CF who did not have CFRD and were 10 years of age or older had an OGTT.

Research shows that early diagnosis and treatment of CFRD leads to better nutrition and health. To hear how an adult with CFRD manages daily life, and to learn more about CFRD and how to manage it, watch the webcast series “Cystic Fibrosis-Related Diabetes” at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Nutrition](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Nutrition). You can also read the 2010 *Clinical Care Guidelines for Cystic Fibrosis-Related Diabetes* at [www.cff.org/treatments/CFCareGuidelines/Nutrition](http://www.cff.org/treatments/CFCareGuidelines/Nutrition).



Other common complications of CF are bone disease and joint problems, such as arthritis or arthropathy. Data from the Patient Registry show that 11.0 percent of people with CF reported bone disease (e.g., fracture, osteopenia or osteoporosis) and 2.7 percent had joint problems.

Preventing bone disease begins in childhood when bones are growing. A diet with enough calories, vitamin D, calcium and other vitamins and minerals, plus a healthy weight and exercise, can help. To learn more about keeping your or your child's bones and joints healthy, talk with your CF care center.

Depression is another problem people with CF often face. Registry data show that 22.1 percent of adults with CF have signs of depression. This is also common in people living with other chronic diseases. If you or your child are feeling hopeless, helpless or like you might be depressed, talk with a health care professional. It is important to be diagnosed and treated early for depression. People often respond well to treatment for depression, by both working with a therapist and taking medicine. Learn more about depression, the symptoms and treatments at [www.Medlineplus.gov](http://www.Medlineplus.gov), using the search word “depression.”

The Registry data also show other complications of CF not included on the graph on page 22. For example, 29.2 percent of people with CF reported having sinus disease in 2011. Symptoms include, but are not limited to, headaches, dental pain and feeling facial congestion or fullness. If you or your child are having these symptoms, talk with your CF care center. Learn more about CF and sinus disease by watching a webcast at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Respiratory](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Respiratory).

Other problems tracked by the Patient Registry include gastroesophageal reflux (GERD), distal intestinal obstructive syndrome (DIOS), liver disease and asthma (see the table below). Learn more about these and other problems of CF and watch a webcast at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Nutrition](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Nutrition).

The Foundation continues to work to find ways to prevent or treat all of these complications.

### Other Problems Related to CF, 2011

Complication	Percentage of people with CF (%) who report this
Gastroesophageal reflux (GERD)	28.9
Asthma	23.9
Liver disease	9.0
Distal intestinal obstructive syndrome (DIOS)	4.7

### Goal 6: TRANSPLANTATION AND END-OF-LIFE CARE

People with CF and their families will be supported by their CF care center when facing decisions about transplantation and end-of-life care.

People with CF who have severe lung disease often think about having a lung transplant. The supply of donor lungs for transplant is limited, plus organ transplantation carries its own set of risks and long-term medical care. It is important to know who might benefit from a lung transplant and when to start the conversation about transplantation. The graph on the next page shows the number of people with CF who have had a lung transplant each year.

To learn more about lung transplantation — the risks, benefits and evaluation process — and hear from an adult with CF who had a lung transplant, watch the CF webcast series at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Respiratory](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Respiratory).

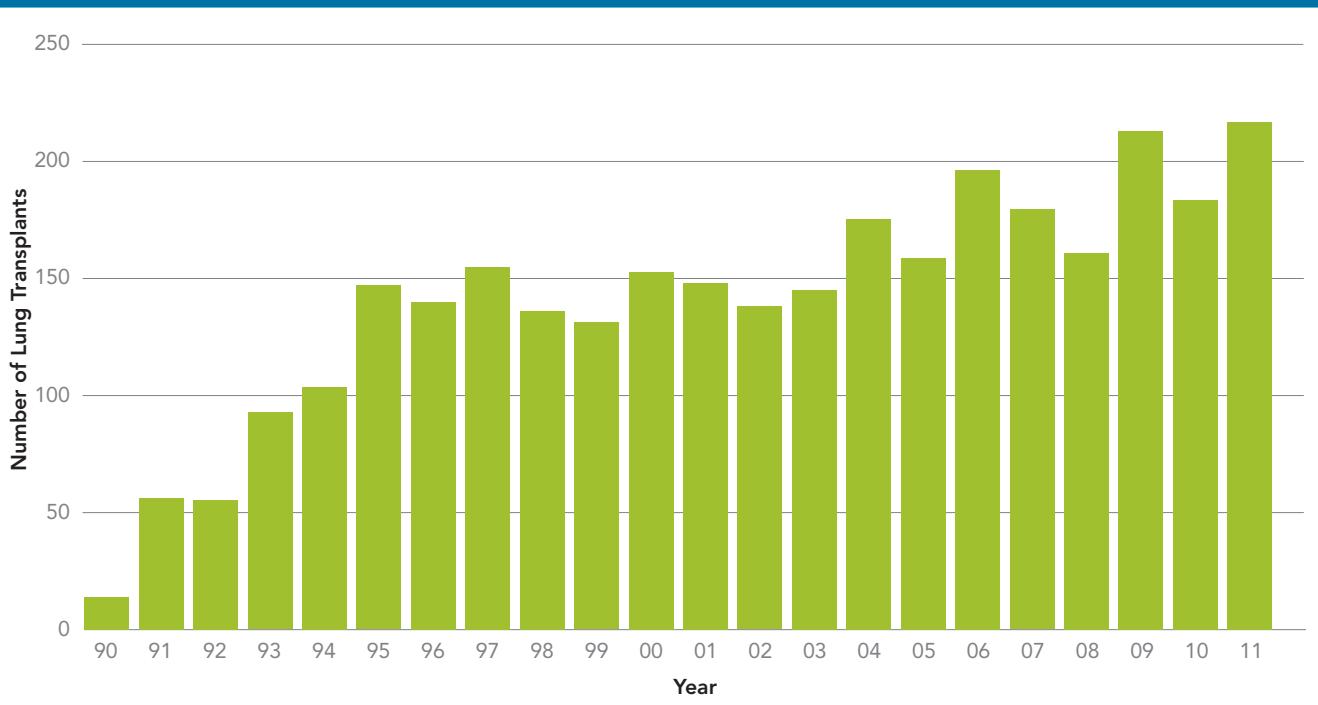
Learn more about living with a transplant at <http://transplantliving.org>.

You can learn more about research related to lung transplant at [www.clinicaltrials.gov/ct2/search](http://www.clinicaltrials.gov/ct2/search), using the search term “lung transplant.”

You can find data about transplantation at <http://optn.transplant.hrsa.gov/data>.

Before a person can get a transplant, someone else has to be willing to donate an organ. To learn about donating organs, visit [www.OrganDonor.gov](http://www.OrganDonor.gov).

## Number of People with CF Who Had a Lung Transplant by Year, 1990–2011



### Goal 7: ACCESS TO CARE

People with CF and their families will have access to appropriate therapies, treatments and support regardless of race, age, education or ability to pay.

Research suggests that people with CF who live in households with lower incomes are more likely to have lower lung function and lower body mass index (BMI) or BMI percentile. This pattern of health in lower-income households is also common in other chronic diseases, such as diabetes.

There are a number of programs available to help people with CF afford the care and medications they need. The Cystic Fibrosis Patient Assistance Foundation (CFPAF), a nonprofit subsidiary of the CF Foundation, helps people with CF living in the U.S. get their prescribed FDA-approved drugs and devices, regardless of health insurance coverage or financial resources. Learn more and visit the CFPAF's website at [www.cfpaf.org](http://www.cfpaf.org) or call toll free 1-888-315-4154.

To find more assistance for people with CF, visit the Patient Assistance Resources page on the Foundation's website at [www.cff.org/LivingWithCF/AssistanceResources](http://www.cff.org/LivingWithCF/AssistanceResources).

The Foundation supports changes to health care that help people with CF. The health care reform law, passed in 2010, allows children to stay on their parents' health insurance until the age of 26. In 2011, 49.8 percent of adults with CF ages 18 through 25 years in the Patient Registry were covered by their parents' health insurance.

The table on the next page shows the percentage of people with CF and the types of health insurance.

## Percentage of People with CF by Type of Health Insurance Coverage, 2011\*

Type of Insurance	17 Years and Younger	18 Years and Older
Health Insurance Policy (e.g. Private Insurance)	55.8	65.0
Medicare/Indian Health Services	0.6	16.6
Medicaid/State Programs	52.6	33.6
TriCare or Other Military Health Plan	2.7	2.1
Other	2.2	2.7
No Health Insurance**	0.4	1.8

\* Data are not mutually exclusive, except the “no health insurance” category, as people with CF may have more than one type of insurance.

\*\*The “no health insurance” category shows the percentage of people with CF who said they did not have any health insurance in 2011.

Data from the Patient Registry show that drugs such as Pulmozyme®, TOBI® and Cayston® are available to people with CF, regardless of income level. To learn more about medication assistance and how others manage CF, watch the webcast series “CF Healthcare Coverage and Advocacy” and “Building Life Skills to Manage CF” at [www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/PartneringCare](http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/PartneringCare).

A local CF care center is the best source for up-to-date information on health care coverage programs in your area and state. Call your CF care center to find help in your area.

To see more information about people with CF in the Patient Registry, including race, gender and age range, see the “Summary of the Data” on pages 26–27.



## SUMMARY TABLES

Summary of the Data: People in the CF Foundation's Patient Registry			
Characteristic	2001	2010	2011
People with CF (number)	22,703	26,532	27,111
Newly diagnosed (number)##	1,077	1,138	883
Age at diagnosis (median)	6 months	5 months	5 months
Age range (years)	0–74.8	0–82.7	0–81.7
Total number of deaths (number)	419	416	444
Predicted median survival (years)	32.6	38.1	36.8
Predicted median survival (years)—95% confidence interval	30.6–35.1	35.4–40.2	34.7–40.3
Adults 18 years and older (%)	39.4	47.3	48.3
<b>Race/Ethnicity (%)</b>			
Caucasian	95.4	94.3	94.3
African American	3.9	4.3	4.3
Hispanic	5.5	7.0	7.3
Males (%)	52.8	51.7	51.8
<b>Home Therapy (%)</b>			
Home IV antibiotics	20.7	19.1	19.1
Received any oxygen	6.0	10.8	10.9
<b>Nutrition</b>			
Pancreatic enzyme supplements use (%)	92.5	86.4	87.4
BMI percentile ages 2 to 19 years (median)*	41.0	50.4	51.3
BMI ages 20 to 40 years (median)*	21.2	22.1	22.1
Supplemental feeding (%)			
Tube	8.5	11.2	11.1
By mouth only	30.8	40.9	41.2
<b>Respiratory</b>			
FEV <sub>1</sub> % predicted (mean)	72.5	77.1	77.1
People with CF with respiratory cultures positive for (%)			
<i>Pseudomonas aeruginosa</i> ( <i>P. aeruginosa</i> ) ¥	58.7	51.2	50.6
Multiple drug resistant <i>P. aeruginosa</i> (MDR-PA)	4.2	9.8	9.1
<i>Burkholderia cepacia</i> complex ( <i>B. cepacia</i> )	3.1	2.5	2.6
<i>Staphylococcus aureus</i> ( <i>S. aureus</i> )**	52.9	67.0	67.9
Methicillin-resistant <i>S. aureus</i> (MRSA)	7.3	25.8	25.9
Therapies*** (%)			
Dornase alfa (i.e., Pulmozyme®)	60.1	81.2	82.1
Hypertonic Saline‡	-	51.5	55.2
Tobramycin solution for inhalation (i.e., TOBI®)	66.7	70.3	65.9
Aztreonam for inhalation solution (i.e., Cayston®)‡	-	22.4	35.8
Azithromycin (chronic oral macrolide antibiotic)‡	-	70.1	70.6
High-dose ibuprofen (e.g., 25–30 mg/kg)	7.5	3.5	3.2

## Summary of the Data: People in the CF Foundation's Patient Registry

Characteristic	2001	2010	2011
People with problems (complications) related to CF (%)			
Gastroesophageal reflux (GERD) ‡	-	26.9	28.9
Asthma‡	-	22.8	23.9
CF-related diabetes (CFRD)	8.8	17.7	19.0
Depression	6.1	11.7	11.8
Liver disease	2.3	8.0	9.0
Distal intestinal obstructive syndrome (DIOS)	3.0	4.3	4.7
Transplants (number)			
Lung (any type)	150	186	220
Liver	19	11	7
Kidney‡	-	4	7
Clinical research participation (number)	1,813	5,872	5,387
Pregnancies (number)	184	224	211
Live birth rates (per 100 women ages 14–45 years)	1.6	1.9	1.6

‡Due to data adjustments, some people diagnosed with CF in 2011 will be entered into the Registry in 2012.  
Next year the number of people who were diagnosed in 2011 is expected to be higher.

\*The Centers for Disease Control and Prevention provide body mass index (BMI) calculators. The national goal for children with CF ages 2 to 19 years is 50th BMI percentile. For adults with CF, the national goal for weight is a BMI of 23 for males and 22 for females. For more information, see [www.cdc.gov/healthyweight/assessing/bmi](http://www.cdc.gov/healthyweight/assessing/bmi).

¥Includes people with CF with MDR-PA.

\*\*Includes people with CF with MRSA.

\*\*\*This is the percentage of people who were eligible for a therapy and had it prescribed at least once in 2011.

‡Not available in 2001.

## Number of People with CF by State\*

State	Number	Percent	State	Number	Percent
Alabama	473	1.74	Nebraska	235	0.87
Alaska	60	0.22	Nevada	182	0.67
Arizona	389	1.43	New Hampshire	199	0.73
Arkansas	255	0.94	New Jersey	653	2.41
California	2,261	8.34	New Mexico	126	0.46
Colorado	550	2.03	New York	1,575	5.81
Connecticut	301	1.11	North Carolina	841	3.10
Delaware	69	0.25	North Dakota	73	0.27
District of Columbia	41	0.15	Ohio	1,498	5.53
Florida	1,290	4.76	Oklahoma	212	0.78
Georgia	776	2.86	Oregon	352	1.30
Hawaii	7	0.03	Pennsylvania	1,438	5.30
Idaho	167	0.62	Puerto Rico	2	0.01
Illinois	1,043	3.85	Rhode Island	96	0.35
Indiana	648	2.39	South Carolina	362	1.34
Iowa	361	1.33	South Dakota	102	0.38
Kansas	349	1.29	Tennessee	607	2.24
Kentucky	518	1.91	Texas	1,667	6.15
Louisiana	326	1.20	Utah	368	1.36
Maine	239	0.88	Vermont	145	0.53
Maryland	495	1.83	Virginia	671	2.48
Massachusetts	835	3.08	Washington	606	2.24
Michigan	1,040	3.84	West Virginia	241	0.89
Minnesota	605	2.23	Wisconsin	629	2.32
Mississippi	239	0.88	Wyoming	48	0.18
Missouri	691	2.55	Unknown	14	0.05
Montana	113	0.42	Foreign	28	0.10

\*These are only people with CF who agree to have their data in the Patient Registry.





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