Patient Registry

Annual Data Report 2010

Cystic Fibrosis Foundation

| Adding *tomorrows* every day.

Cystic Fibrosis Foundation | Adding *tomorrows* every day.

To the CF Community and Friends,

The Cystic Fibrosis Foundation works hard so that all people with cystic fibrosis have access to specialized CF care through its network of more than 110 CF care centers.

A key component of the effort to improve the quality of care is the data collected on the health of more than 26,000 people with CF receiving care at Foundation-accredited CF care centers nationwide.

This data is reported and shared with the wider community through the Patient Registry Annual Data Report, which each year highlights progress and areas for improvement in CF care.

We are proud to report that, based on the most recent data in the Patient Registry, the median predicted age of survival is now more than 38 years.

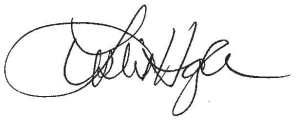
The data in this report, collected in 2010, show promise in many other areas, including:

- Better overall lung function for both adults and children.
- Increased body mass index (BMI) for adults and higher BMI percentile for children.
- Growing numbers of adults with CF ages 18 and older.
- Greater numbers of infants diagnosed at birth with CF, thanks to newborn screening efforts across the country.

These gains reflect the strong partnerships among people with CF, their families and health care professionals. We remain deeply grateful to the people with CF and their families who generously consent to share their data.

We hope that this report serves as a springboard to open discussions throughout the CF community. These conversations will help you improve the quality of care at your CF care center.

Sincerely,



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WHAT IS THE CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY?

The Cystic Fibrosis Foundation's Patient Registry tracks the treatments and health of people with cystic fibrosis (CF) across the United States. Information is collected every year on the more than 26,000 people who receive care at CF Foundation-accredited care centers and agree to participate in the Registry. The data include state of residence, height, weight, gender, genotype, pulmonary function test (PFT) results, medication use, length of hospital stays, home IV use and problems related to CF.

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

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

The Registry helps people with CF and their families, as well as health care professionals, compare the overall health of patients in one care center with all other centers nationwide. For this reason, it is vital for everyone with CF to agree to have their data entered into the Registry. Patient participation provides more complete data and helps create a better picture of the current state of CF care. This data can also be used to identify areas where more work can be done to improve the health of those living with the disease.

To improve the health of people with CF, the Foundation developed the following seven goals of CF care, described in more detail in this report:

1. People with CF and their families will be full members of the care team.
2. Children, adolescents and adults with CF will have normal growth and nutrition.
3. 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 them to previous levels of lung function.
4. People with CF, their families and CF health care professionals will be well-informed and active partners in reducing respiratory infections, particularly *Pseudomonas aeruginosa* (*Pseudomonas*) and *Burkholderia cepacia* (*B. cepacia*) complex.
5. People with CF will be screened and managed aggressively for complications of the disease, particularly CF-related diabetes (CFRD).
6. People with CF and their families will be supported by their CF health care professionals when facing decisions about transplantation and end-of-life care.
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.

The information presented in this report describes the work being done to meet these goals.

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 organs in the body. This leads to multiple problems, especially in the lungs and digestive system.

People with CF often develop lung infections and have inflammation in the lungs. Over time, this leads to damage and decreased 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 keep a healthy body weight.

About one in 3,500 children in the United States is born with CF each year. 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 a group of parents who had children with CF. Their mission was clear: to save their children. The Foundation built on this mission and works to assure the development of the means to cure and control CF and to improve the quality of life for those with the disease.

To further this mission, the Foundation set up a network of more than 110 accredited care centers across the United States to care for the needs of people with CF. The Foundation provides care centers with:

- Grants
- Quality improvement training
- CF care guidelines based on research from medical journals
- Updates on CF care and research at the North American CF Conference
- Educational resources for people with CF and their families
- Patient Registry data to track the health of people with CF and show care improvements.

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

A top research priority of the Foundation is to find treatments that target the underlying cause of CF. The Foundation also continues to support the development of therapies that treat the different symptoms of the disease. To speed up the development of all potential new therapies, CF Foundation-accredited care centers do clinical research. People with CF join in the testing or trial of potential CF drugs.

To learn more about CF, the Foundation and its research and drug development pipeline, visit **www.cff.org**.

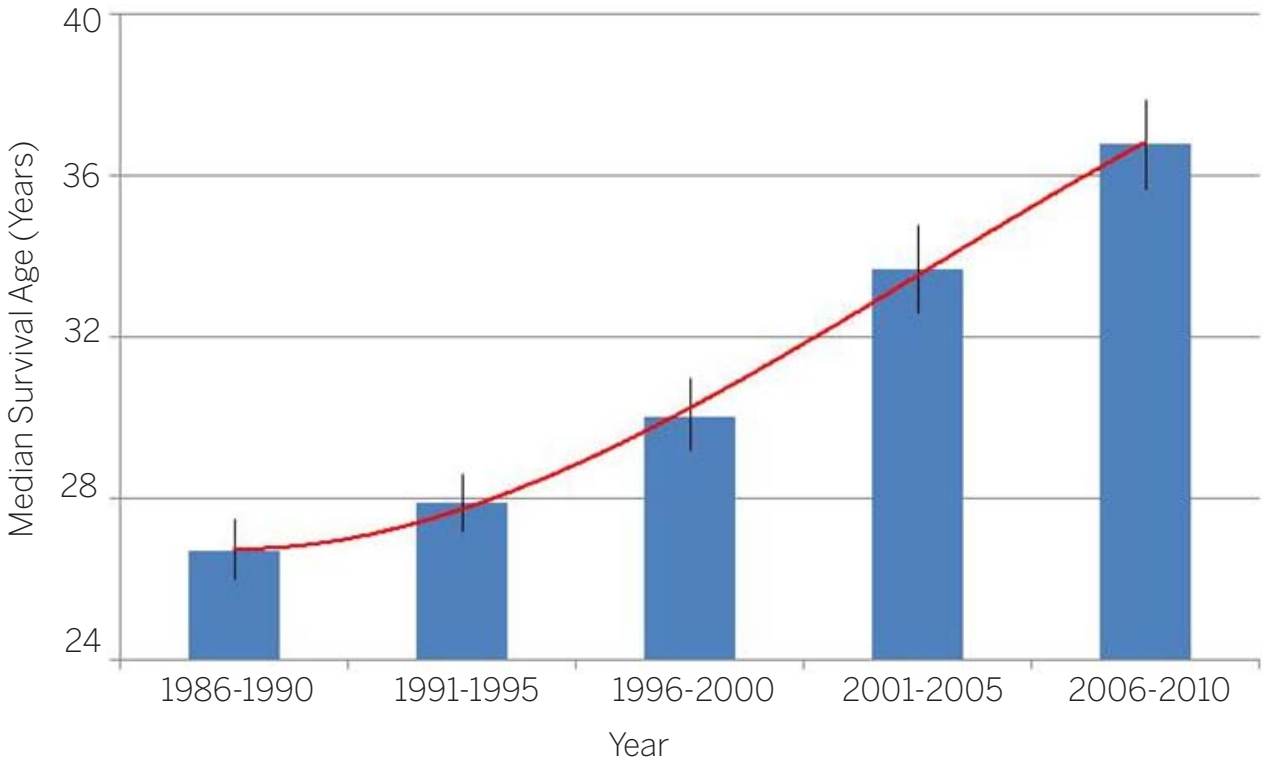
OVERALL HEALTH OF PEOPLE WITH CF IS IMPROVING

The median predicted age of survival for people with CF has risen dramatically over the past two decades. In 1986, the median predicted age of survival was 27. In 2010 it was 38.3.

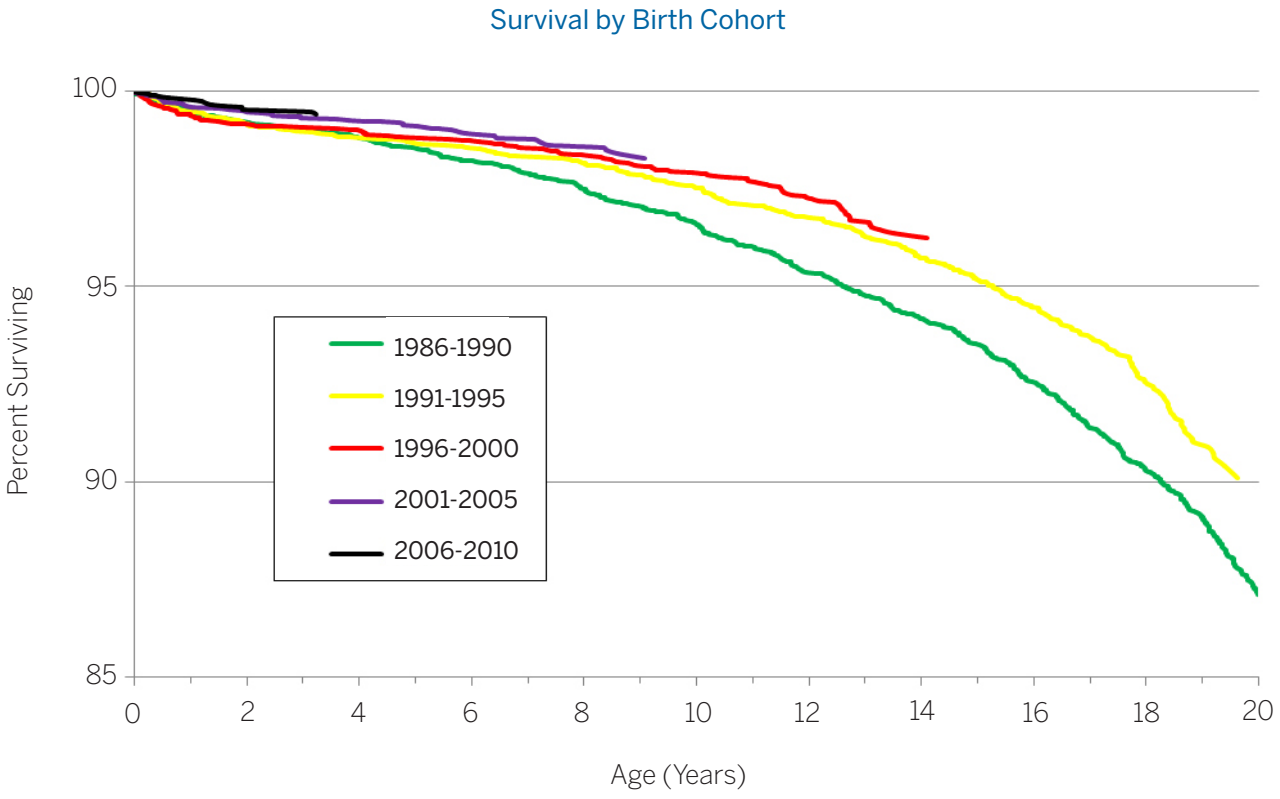
Median predicted age of survival is calculated using a standard method called life table analysis. This is best known for its use in the life insurance industry. Using this calculation, half of the people in the patient registry are expected to live beyond the median predicted survival age (38.3 years in 2010), and half are expected to live less than the median predicted survival age.

As the chart below shows, in 5-year periods, the survival age for people with CF continues to increase.

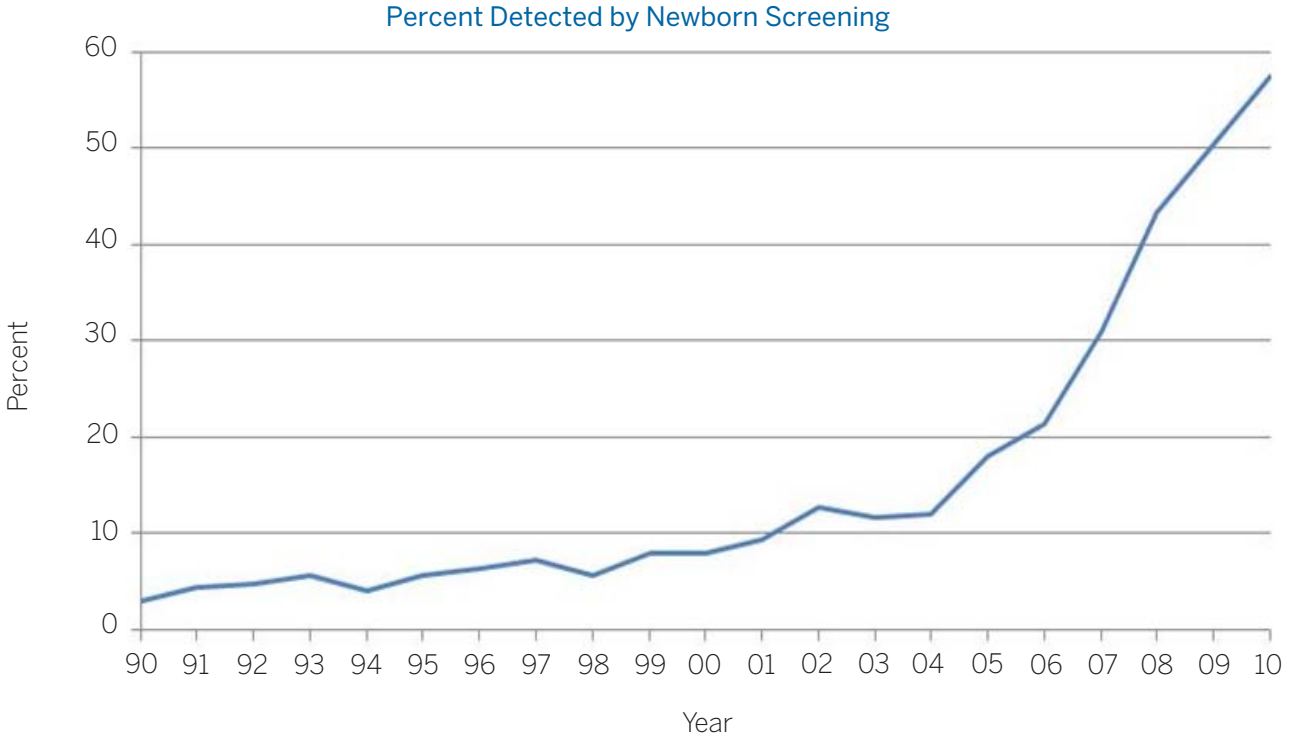
Median Predicted Survival Age, 1986-2010
Over 5 Year Bands



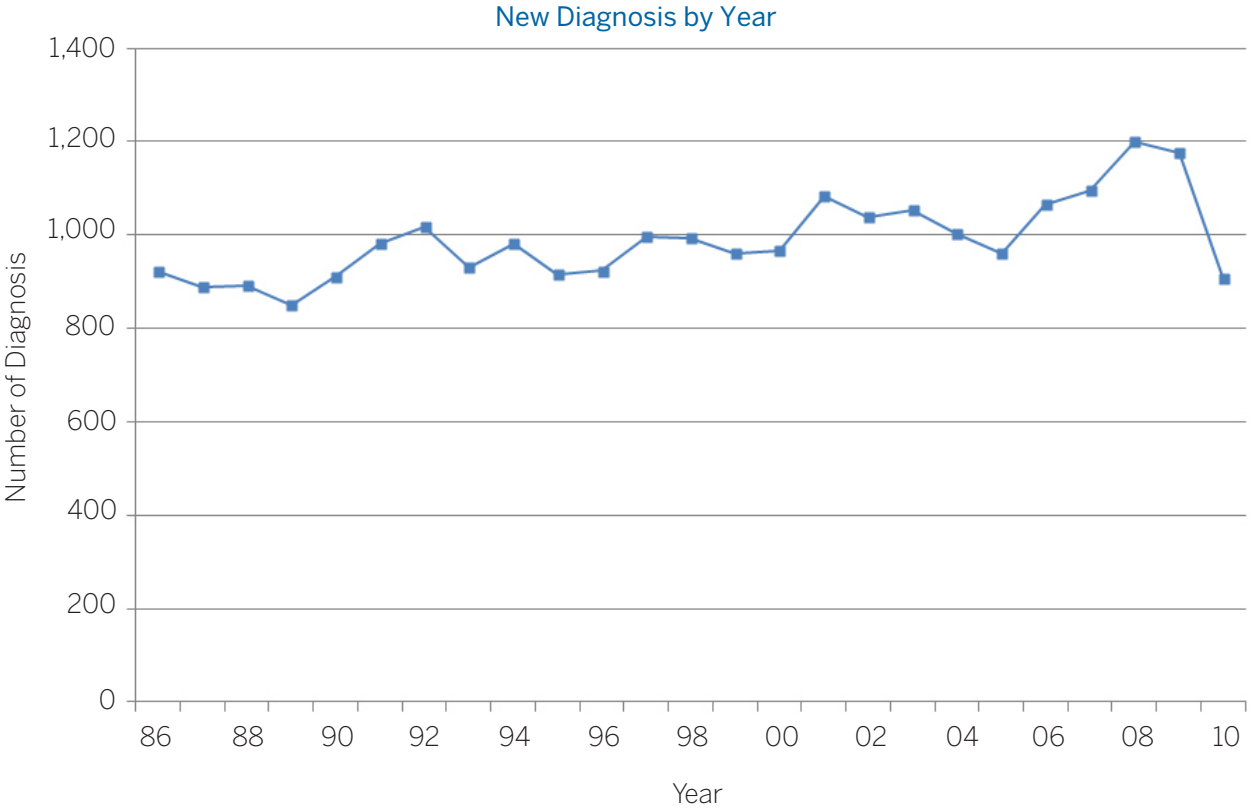
The graph on the next page shows another way to look at how survival is improving. Of people with CF born between 1986 and 1990 (green line), 93.5 percent were alive at age 15. For children with CF born between 1991 and 1995 (yellow line), 95.1 percent were alive at age 15.



There are many reasons for this increase in survival. Stronger partnerships among people with CF, their families and CF care center staff are key. Earlier diagnosis because of nationwide newborn screening also plays a critical role. The earlier CF is diagnosed, the sooner treatment can begin. As shown in the graph below, more than half of those diagnosed with CF in 2010 were diagnosed through CF newborn screening.



However, there are still many children and adults being diagnosed with cystic fibrosis. The graph below shows that more than 800 people are diagnosed with CF each year.



Since 2004, the CF Foundation has worked to have all 50 states and the District of Columbia include CF in their standard newborn screening tests. This goal was met in January 2010. 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 at an older age because of symptoms such as lung infections or not growing well.

To help CF doctors and nurses care for babies with CF who may not show symptoms, the CF Foundation worked with experts in CF and infant care to develop the Infant Care Guidelines. These guidelines outline CF care that helps to stop or slow the progress of the disease.

To read the Foundation's care guidelines, visit **www.cff.org/treatments/CFCareGuidelines/AgeSpecificCare**.

Research also shows that infants with CF, even those who show no symptoms, may already have lung damage. The CF Foundation supports research to find drugs suitable for infants with CF that can help keep their lungs healthier.

For example, a Phase 2 study of inhaled saline in infants (ISIS) is looking at inhaled hypertonic (7%) saline for use in children under 6 years of age. This therapy, now prescribed for others with CF, may help to stop or slow lung damage in a younger children. To learn more about this study, visit **www.cff.org/research/ClinicalResearch/Find** and search for CF clinical trials by age.

The CF Foundation is working with companies to develop drugs that target the underlying cause of CF. Much of this research is on specific mutations, or genotypes. Recent results from clinical trials of one potential therapy in people with at least one copy of the G551D mutation of CF showed that those people who took the drug had major improvement in lung function and weight gain, compared with those who did not take the drug.

To learn more about this and other CF clinical trials, visit **www.cff.org/research/ClinicalResearch**.

It is helpful for you to know which mutations you or your child has. According to the Patient Registry, 91.7 percent of people with CF have had their CF mutations identified. More than 88 percent of people with CF have at least one copy of the Delta F508 mutation.

However, there are more than 1,800 known mutations of the CF gene. The CF Foundation is supporting research to find out which mutations cause CF. Below is a list of the most common CF mutations. If you do not know if you or your child has had your CF mutations identified, talk to your CF care center.

Most Common CF Mutations or Genotype Reported in the CF Foundation's Patent Registry

Mutation	Percentage (%) of People with CF with at Least One Mutation
Delta F508	88.5
G542X	4.6
G551D	4.4
R117H	2.7
N1303K	2.5
W1282X	2.4
R553X	1.9

To learn more about CF genetics, see "About Cystic Fibrosis" at **www.cff.org**.

MEETING THE GOALS OF CF CARE

The CF Foundation is committed to improving CF care based on the seven goals of CF care. The data in the Registry show how the work being done in CF care centers helps the CF community reach these goals.

Full Members of the CF Care Team

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.

The CF Foundation's Patient Registry creates reports about the health of each person with CF, each CF center as a whole and the combined health of people with CF throughout the United States. These reports help the CF care team find ways to improve the care and health of people with CF.

The sample "Patient Summary Report" on page 8 is a snapshot of a person's lung health, weight and other information. Ask your CF care center for a copy of your or your child's "Patient Summary Report" at your next clinic visit.

SAMPLE PATIENT SUMMARY REPORT

VISIT DATE:	_____				
Last Hospitalization:	None	**Date of Birth:**	/1963		
Last Home IV Therapy:	None	**Genotype:**	F508del / F508del		
Last Clinical Visit:	/2011	**Centers Visited:**			
Current diagnosis:	Cystic Fibrosis				
Diagnosis Date:	/1964	**Symptoms:**	Acute or persistent respiratory abnormalities		
Sweat Test Date:	/1992	**Sweat Test Value (mmol/L):**	107.0	**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

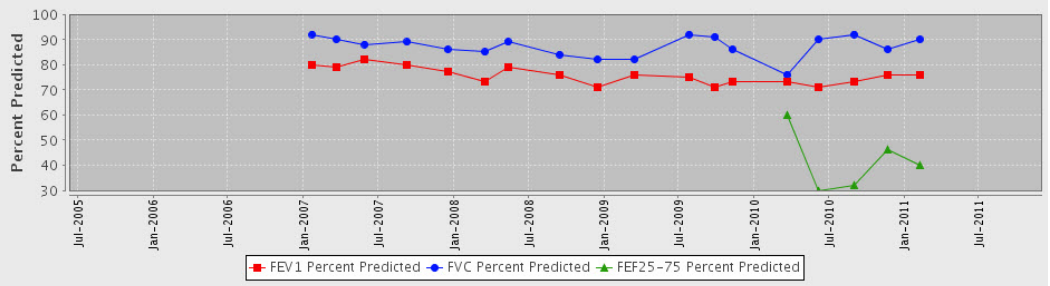
Last Culture: /2011

Positive Tests:	Pseudomonas aeruginosa	Multi-drug Resistant Pseudomonas aeruginosa+	Burkholderia species	Stenotrophomonas maltophilia	MSSA	MRSA	H. influenzae
	/2011 /2010 /2010 /2010 /2010	/2011 /2010 /2010 /2010 /2010			/2011 /2010 /2010 /2010	/2010	

Last Mycobacterial Culture: /2010

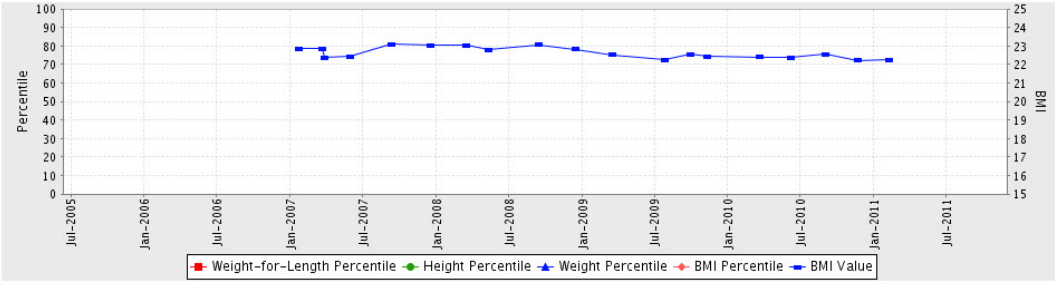
Mycobacterial Species Detected:

PFTs - % Predicted



Last FEV1 Date:	/2011	**Last FEV1 Value:**	
Last FVC Date:	/2011	**Last FVC Value:**	
Last FEF25-75 Date:	/2011	**Last FEF25-75 Value:**	

Nutritional Trend



Complications Active at Last Visit:		**Complications Previously Noted:**	

Routine Evaluations

Last PFT:	/2011	**Last Dietary Visit:**	/2010
Last CXR:	2008	**Last LFT:**	/2011
Last SW Visit:	none	**Last Glucose Screening:**	/2011
Last RT/PT Visit:	/2010	**Last Creatinine:**	/2011

Comments

The Information contained within Patient Registry reports reflect data entered by personnel at the care center. As such, CFF assumes no responsibility for, or in any way warrants the accuracy of the data submissions. Users must verify Registry data with the individual's medical record before making care decisions.

After reviewing your or your child's Patient Summary Report, think about asking the following questions:

- What does the data mean?
- How can I help improve my or my child's health?
- What is my center doing to improve care?
- What can I do to help my center improve?

To learn more, visit **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/** and watch the archived webcasts "Partnering for Improvement," "Partnering for Care," "One Team's Story: Raising the Bar for CF Care" and "Quality CF Care Is More Than the Numbers." To read success stories of how others have worked with their care centers to improve care, visit **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 provide information for health care professionals about the best treatment for specific diseases or conditions, based on published medical articles and research.

To develop CF care guidelines, the CF Foundation gathers CF health experts, adults with CF and parents of children with CF to review the medical literature and research on CF care, along with data from the Patient Registry. These guidelines give information about diagnosis, nutrition, lung health, infection control and age-specific care. To learn more, visit **www.cff.org/treatments/CFCareGuidelines**.

Published guidelines in 2010 include:

- "Clinical Care Guidelines for Cystic Fibrosis-Related Diabetes"
- "Pulmonary Complications: Hemoptysis and Pneumothorax"

The clinical care guidelines state that people with CF have certain lab tests, throat or sputum cultures and exams each year. The table below lists these and also reports how people with CF in the Patient Registry met these guidelines in 2010, compared with the data from 2009.

Care, Screening and Prevention Guidelines for People with CF	People with CF Who Met the Guidelines (%)	
	2009	**2010**
4 or more clinic visits	57.97	63.19
4 or more respiratory cultures	42.65	44.77
2 or more pulmonary function tests (PFTs) if 6 years of age or older and physically able	81.49	81.50
An influenza (flu) vaccine if 6 months of age or older	71.11	73.00
Fat-soluble vitamin levels measured	78.54	80.59
An oral glucose tolerance test (OGTT) if 10 years of age or older	13.02	19.33
Test to measure liver enzymes in the blood	78.18	72.26

NORMAL GROWTH AND NUTRITION

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

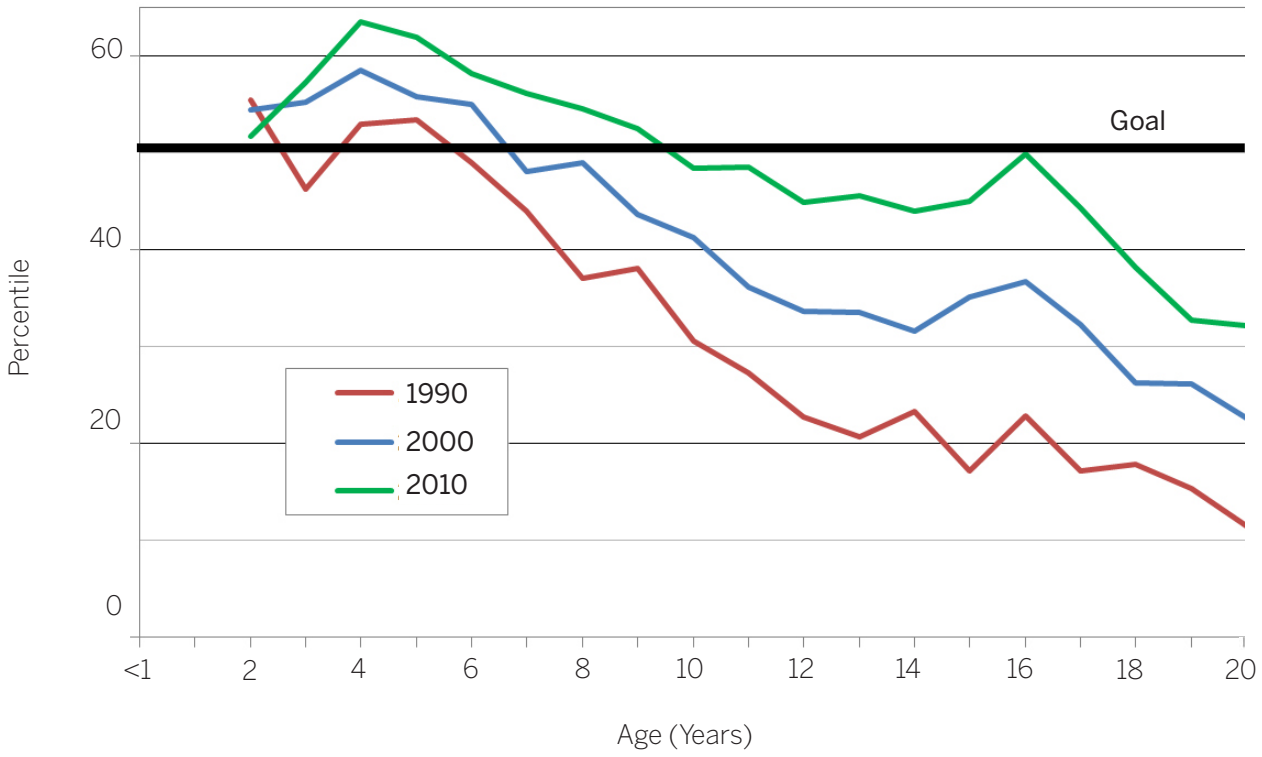
The nutrition of people with CF is better, but more work needs to be done. The CF care guidelines set goals for weight (measured as body mass index, or BMI) for children and adults with CF. BMI is based on a person's weight and height. It is used to screen for health problems. BMI is calculated by dividing body weight in kilograms by the person's height in meters squared (weight kg/height m2=BMI).

To calculate your or your child's BMI percentile, visit **www.cdc.gov/healthyweight/assessing/bmi**.

For children and teens with CF, BMI is stated as a percentile compared to 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 and to have a BMI around the 50th percentile for their age. (The 50th percentile is the average BMI percentile for children in the United States who do not have CF.)

The next graph shows how much the BMI percentile of children with CF has improved since 1990. It also shows that more work can be done to improve the BMI percentile of children with CF older than 10 years of age. The graph on page 11 starts at age 2 because the growth of younger children is measured as weight to length on a growth chart. To find infant growth charts, visit **www.cdc.gov/growthcharts/clinical_charts.htm**.

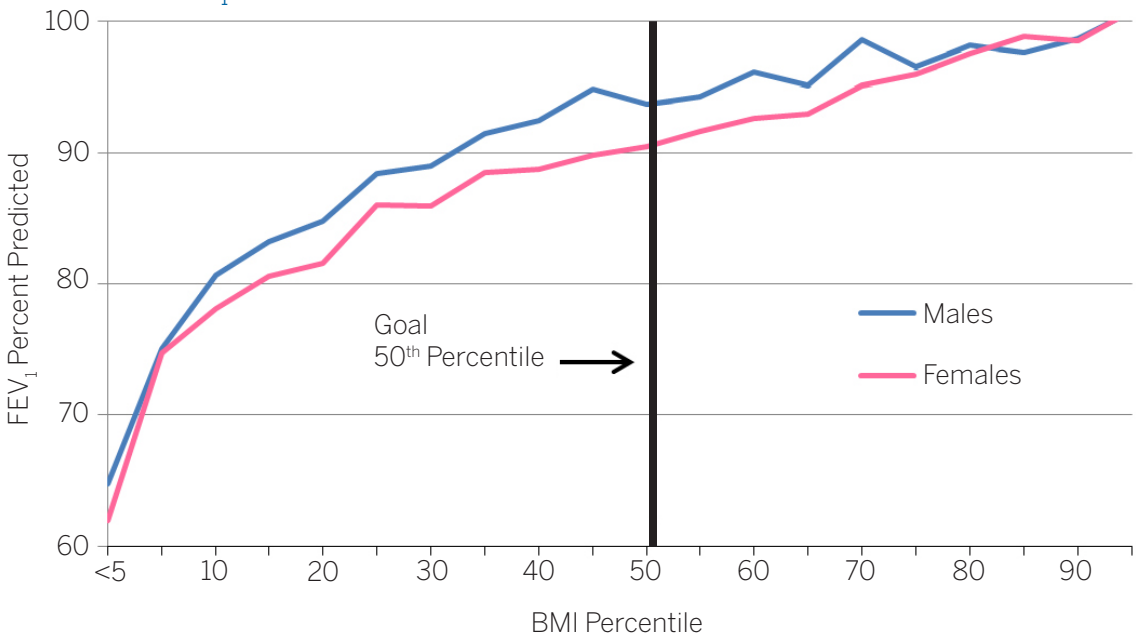
Median CDC BMI Percentile vs. Age



The Patient Registry shows a strong association between a higher BMI percentile and better lung function in children with CF. Lung function, or FEV_1, is one test that shows how well the lungs work. Lung damage is caused by inflammation or swelling from infections that injure the lungs and airways and do not heal. This type of injury often leads to scar tissue or fibrosis. With every lung infection, a little more damage can happen.

FEV_1 (forced expired volume in 1 second) is shown as the percent predicted, based on the FEV_1 of healthy, non-smoking people of the same age, height and gender. The following graph shows that good nutrition and lung health seem to go hand in hand. The bar in the middle is the BMI percentile goal for children with CF.

FEV_1 Percent Predicted vs. BMI Percentile in Patients 6 to 19 Years

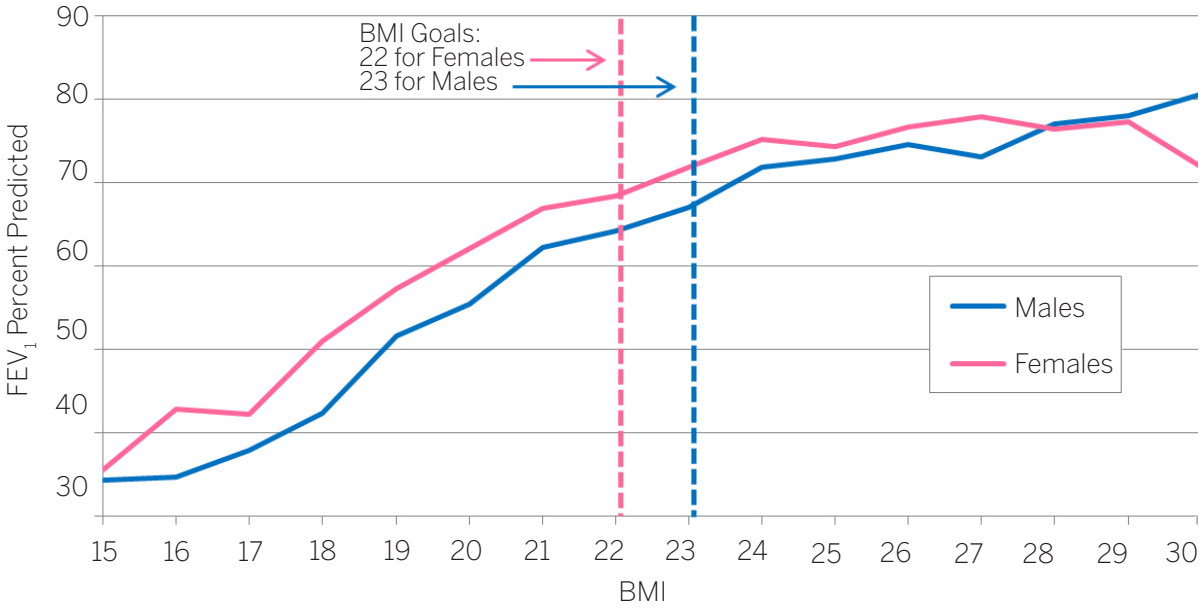


The association between a higher BMI and better lung function is also seen in adults with CF. The following graph shows the BMI goals for men and women with CF — 23 and 22, respectively.

To improve or maintain your or your child's weight, the Foundation suggests that you work closely with your CF care center staff. To learn more about CF and nutrition, ask your CF care center or visit **www.cff.org/LivingWithCF/StayingHealthy**.

In 2010, the U.S. Food and Drug Administration (FDA) approved three pancreatic enzyme supplements that help people with CF absorb food and have better nutrition and weight. While enzymes have been in use for decades, the FDA approval means all pancreatic enzymes that people with CF take have been reviewed and meet the same standards. The CF Foundation urged the FDA to do this review to make sure that all pancreatic enzymes are safe and effective so people with CF can have better nutrition and growth.

FEV$_1$ Percent Predicted vs. BMI in Patients 20 to 40 Years



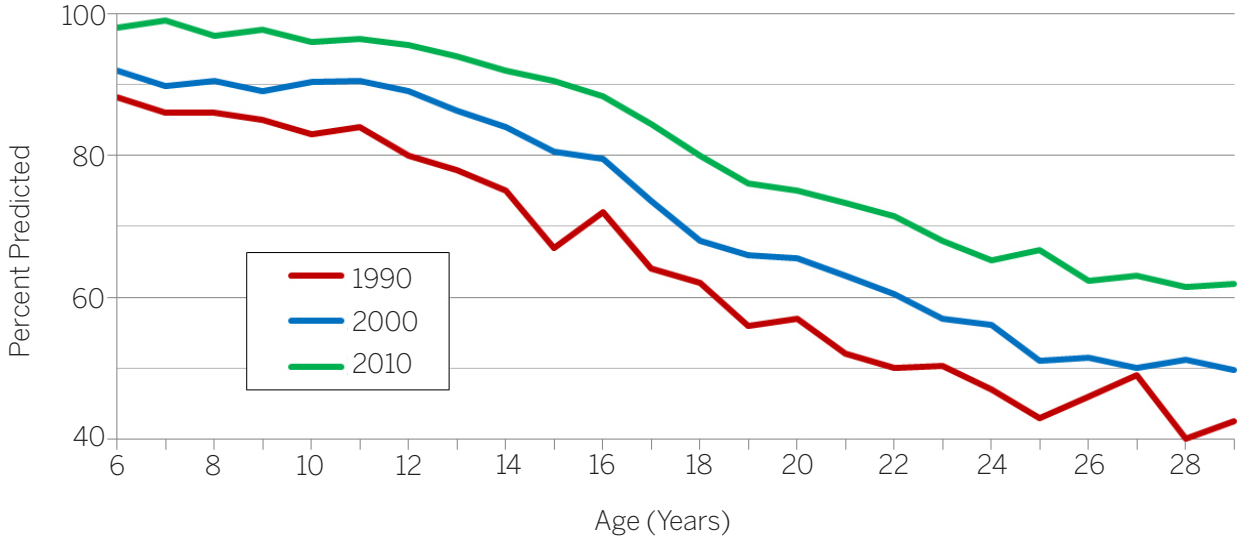
LUNG HEALTH

Goal 3: 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 aggresively to return lung function to previous levels.

People with CF often have pulmonary exacerbations — when the lungs need treatment to get better. This can be caused by an infection or inflammation. If you or your child are coughing more, have more sputum, a drop in lung function or a loss of appetite, it may be a pulmonary exacerbation.

The graph on the next page shows that the lung health of people with CF has improved since 1990. FEV$_1$ (lung function) is usually between 90 to100 percent predicted when first measured around 6 years of age.

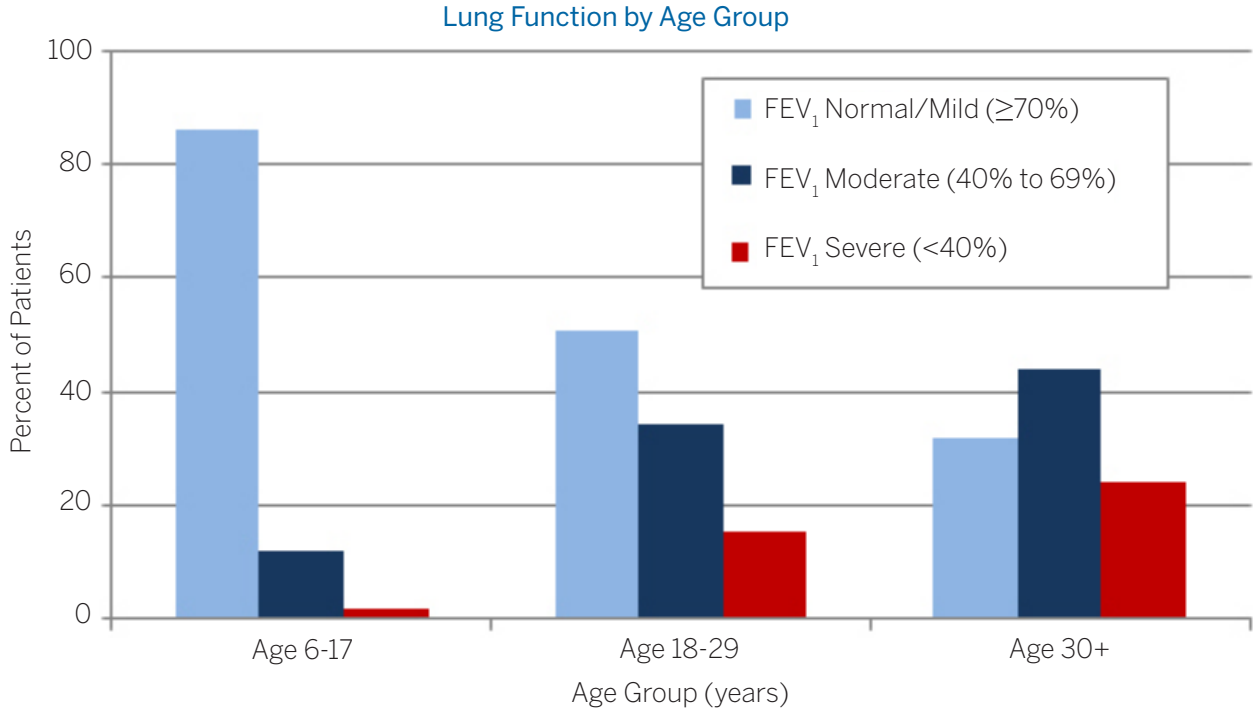
Median FEV$_1$ Percent Predicted vs. Age, 1990, 2000 and 2010



Research shows that people with CF, babies too, have some lung damage. Even when their lung function is normal there may be damage. However, over time, FEV_1 decreases because of damage from exacerbations. In CF, the lower a person's FEV_1, the more severe the lung disease and damage. Here is a guide to FEV_1 percent predicted and what it may mean for a person with CF:

- FEV_1 greater than or equal to 90 percent is normal. However, even with normal lung function, some lung disease and damage may be present.
- 70 to 89 percent shows mild lung disease and damage.
- 40 to 69 percent shows moderate lung disease and damage.
- Less than 40 percent is a sign of severe lung disease and damage.

The graph below shows the percentage of people with CF by age group with lung function from normal to severe, based on FEV_1 percent predicted. In 2000, 76.4 percent of children with CF had an FEV_1 of greater than or equal to 70 percent. Today, more than 85 percent of children ages 6 to 17 years have a lung function that shows mild disease or better lung health.



Researchers are working to find new drugs to slow or stop lung function decline, to improve FEV_1 and keep the lungs of people with CF as healthy as possible. One example of this work is the 2010 FDA approval of Cayston® (aztreonam for inhalation solution), developed by Gilead Sciences, Inc., with CF Foundation support. To learn more about Cayston, other therapies and CF research, visit **www.cff.org**.

The "Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Function" lists medications that help people with CF take care of their lungs. To learn more about these guidelines, visit **www.cff.org/treatments/CFCareGuidelines/Respiratory**. The following table shows the criteria for each medication, along with the percentage of people with CF who are taking each one. To learn whether you or your child might be helped from one of these medications, talk to your CF doctor.

Therapy Use in the CF Population

Percentage (%) of people who fit criteria for the medication and have had it prescribed:

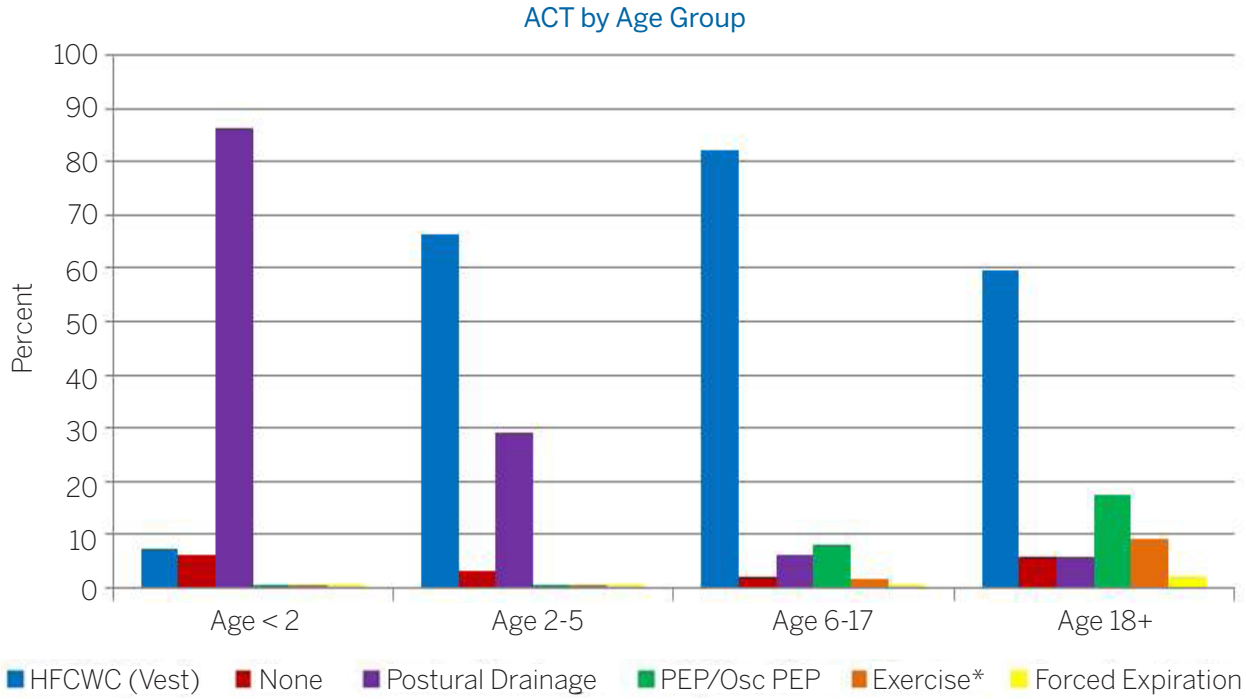
Chronic Medication*	Patients Prescribed the Medication (%)	Criteria for Prescribing Medication
Dornase alfa (Pulmozyme®)	78.4	• At least 6 years old
Hypertonic saline	49.9	• At least 6 years old
Tobramycin solution for inhalation (TOBI®)	70.2	• *P. aeruginosa* in cultures • At least 6 years old
Chronic oral macrolide antibiotic (not for treatment of an exacerbation)	70.2	• *P. aeruginosa* in cultures • At least 6 years old • Weight over 25 kg (55 lbs) • FEV_1 over 30% predicted
High-dose ibuprofen (e.g., 25-30 mg/kg)	3.5	• 6-12 years old • FEV_1 over 60% predicted

**Cayston® is not included in this report because it was not available for prescription in all 12 months of 2010. The percentage of use for this inhaled antibiotic will be reported in next year's CF Foundation Patient Registry Annual Data Report.*

Medication alone can't keep CF lungs healthy. Getting the thick mucus out of the lungs is key. Airway clearance techniques (ACT) 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. People with CF often use one or more types of ACT.

For more information, visit **www.cff.org/treatments/CFCareGuidelines/Respiratory**. The guidelines state that people with CF should do airway clearance every day to keep lungs healthy, even when they are feeling well.

The chart on the next page shows the percentage of people with CF by age and which ACT they use.



** The "Cystic Fibrosis Pulmonary Guidelines: Treatment of Pulmonary Exacerbations" recommends that airway clearance therapy be done more often when sick and does not consider exercise as a form of airway clearance (www.cff.org/treatments/CFCareGuidelines/Respiratory).*

To learn more, visit **www.cff.org/treatments/Therapies/Respiratory/AirwayClearance**, and watch the CF Education webcasts "Airway Clearance Techniques" and "Help Your Respiratory or Physical Therapist Help You Thrive" at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**.

Keeping Lungs Healthy

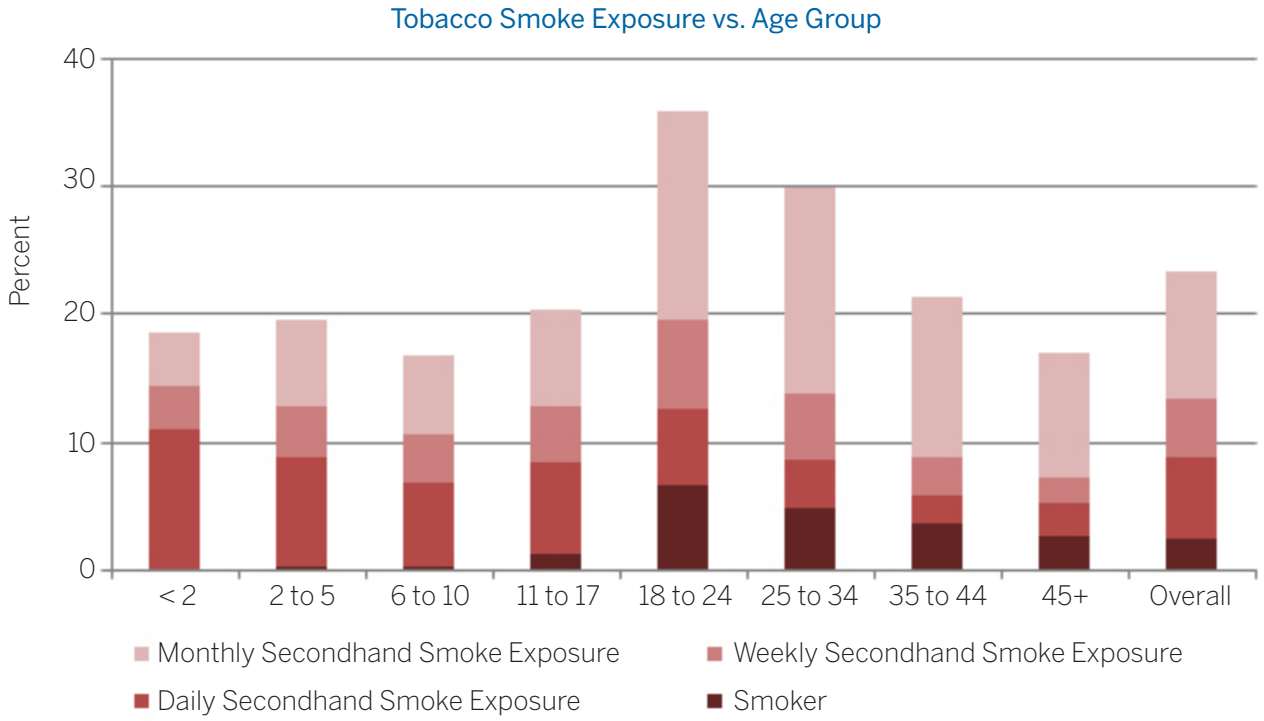
Each time a person with CF has an infection or pulmonary exacerbation, more lung damage may occur.

Here are some things you 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 doctor.
- Get a flu shot every fall for you or your child and for everyone age 6 months and older who lives in your house.
- Exercise regularly to strengthen your muscles.
- Avoid germs by:
  - Cleaning your hands often with soap and water or alcohol-based hand gel.
  - Using a tissue when coughing or sneezing, then cleaning your hands.
  - Cleaning and disinfecting nebulizers after every treatment.
- Avoid tobacco and marijuana smoke.

Secondhand smoke — tobacco smoke exhaled by a smoker or produced by burning tobacco — is harmful for people with CF. Research has shown that children 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.

The following graph shows, by age group, the percentage of people with CF who breathe in secondhand tobacco smoke.



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

- Not let anyone smoke in your home or car.
- Ask people not to smoke around you or your child, even when outside.
- Stay away from people when they are smoking and teach your child to do the same.
- Make sure that your child's day care center or school is smoke-free.
- Ask your employer for a smoke-free work and eating space.
- Choose restaurants and other businesses that are smoke-free and thank them for being 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.

If you smoke, 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.

If you have CF and smoke, you are adding to your lung disease and damage. Ask your doctor to help you quit. Recent research has shown that marijuana smoke can also worsen lung function and damage the airways. In any case, people with CF should avoid secondhand smoke and not smoke to keep their lungs as healthy as possible.

To learn more about the effects of smoking and tobacco use, visit the Centers for Disease Control and Prevention website at **www.cdc.gov/tobacco**. To learn the facts about the long-term effects of marijuana use, visit **www.teens.drugabuse.gov**.

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.

To learn more, visit **www.cff.org/treatments/Therapies**. It is important that you and your CF care center work together to create a plan to stay healthy. You can also view webcasts about CF lung health and disease at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**.

RESPIRATORY INFECTIONS

Goal 4: People with CF, their families and CF health care professionals will be well-informed and active partners in reducing respiratory infections, particularly from *Pseudomonas aeruginosa* (*Pseudomonas*) and *Burkholderia cepacia* (*B. cepacia*) complex.

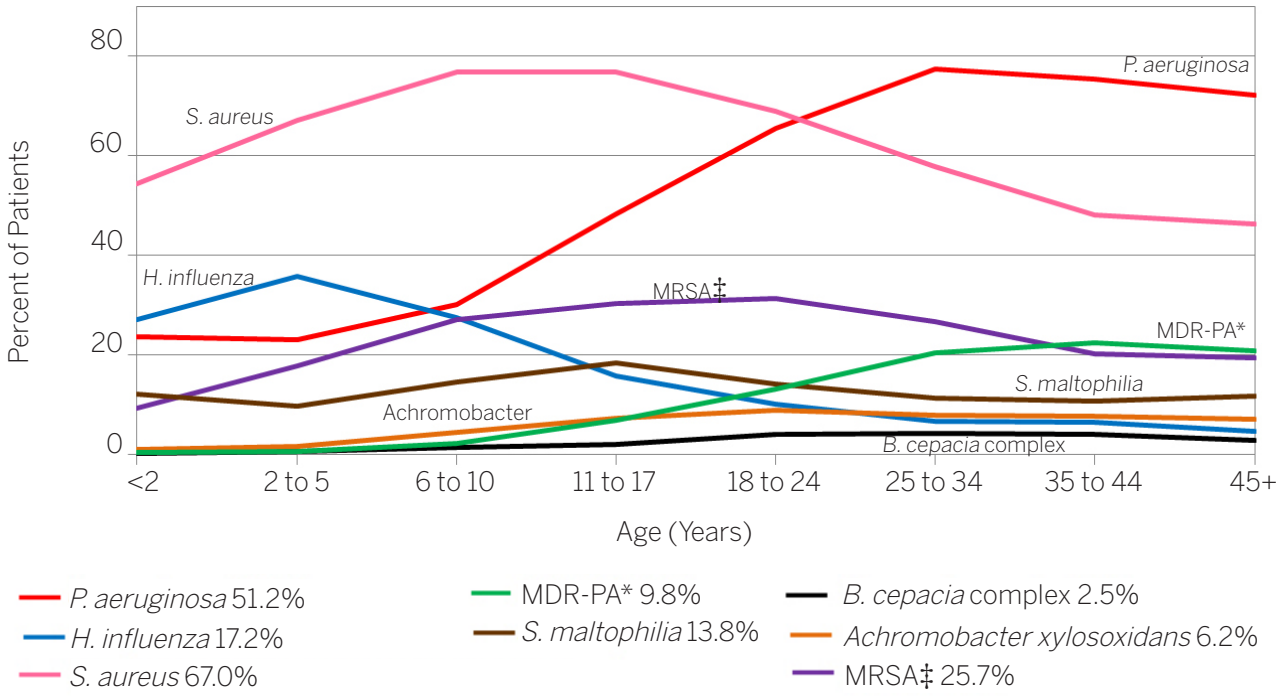
The best way for anyone to avoid germs is to:

- Clean hands often with soap and water or alcohol-based hand gels.
- Use a tissue when coughing or sneezing, then clean your hands.
- Avoid touching your eyes, nose or mouth.
- If you are ill, stay away from others.

Repeated lung infection and inflammation worsen lung disease and causes damage. This damage causes lung function (FEV_1) to get worse. When the lungs are damaged, infections happen more often. This is why avoiding the spread of germs is so important to people with CF and their families.

The graph on the next page shows some of the germs that are found in the lungs of people with CF. To learn more about how to avoid respiratory infections, talk to your CF care center staff.

Respiratory Germs by Age, 2010



P. aeruginosa 51.2%
H. influenza 17.2%
S. aureus 67.0%
MDR-PA* 9.8%
S. maltophilia 13.8%
B. cepacia complex 2.5%
Achromobacter xylosoxidans 6.2%
MRSA‡ 25.7%

*MDR-PA is multi-drug resistant *Pseudomonas aeruginosa (P. aeruginosa)*
‡MRSA is methicillin-resistant *Staphylococcus aureus (S. aureus)*

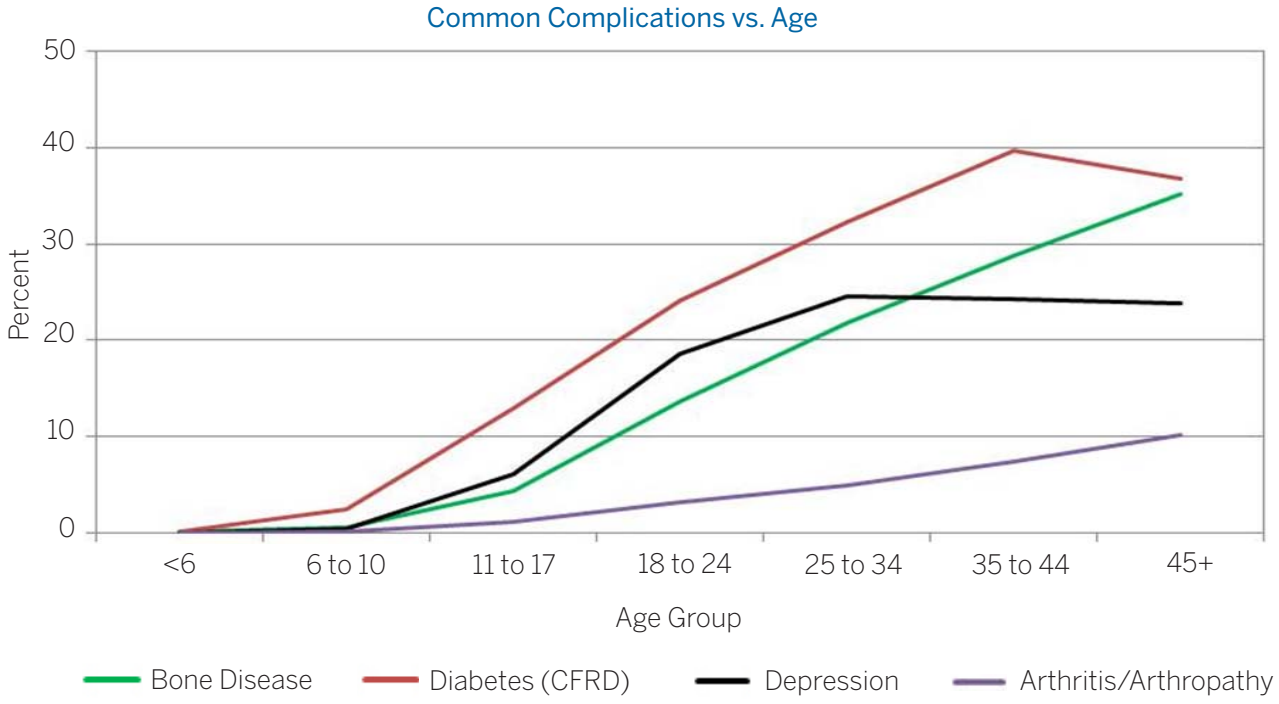
PROBLEMS OF CF

Goal 5: People with CF will be screened and managed aggressively for complications of the disease, particularly CFRD.

Complications are problems that happen related to CF. Cystic fibrosis-related diabetes (CFRD) is one of the most common CF complications. CFRD is different from diabetes in people without CF.

People with CF age 10 years and older should be tested every year for CFRD. In 2010, more than 17.5 percent of people of all ages in the Patient Registry have CFRD, and 30.5 percent of people age 18 years and older have CFRD.

The Patient Registry data show that early diagnosis and treatment of CFRD leads to better nutrition, weight and better health. To learn more about CFRD and how to manage it, watch the webcast "Cystic Fibrosis-Related Diabetes" at **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**.



The Registry data show other complications of CF. It is estimated that about 12 percent of people without CF in the United States have at least one sinus infection (acute sinusitis) per year. Almost 27 percent of people with CF reported having symptoms of sinus problems in 2010.

Chronic sinusitis is when symptoms last for eight or more weeks. Symptoms include, but are not limited to, headaches, dental pain and feeling facial congestion or fullness. If you or your child are having symptoms of chronic sinusitis, talk with your CF doctor. To learn more about CF and sinus disease, watch an archived webcast at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**.

Another common complication of CF is bone disease and joint problems such as arthritis or arthropathy (joint problems). Data from the Patient Registry show that about 10.9 percent of people with CF reported bone disease (e.g., fracture, osteopenia or osteoporosis) and 2.7 percent had joint problems.

Preventing bone disease and joint problems begins in childhood when bones are growing. Good nutrition, 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 dietitian and physical therapist.

Depression is another problem of CF. Registry data show that 22 percent of adults with CF have signs of depression. This is also common in people with other chronic diseases. People often respond well to treatment for depression so it is important to be diagnosed and treated early.

Other problems tracked by the Patient Registry include gastroesophageal reflux (GERD), distal intestinal obstructive syndrome (DIOS) and asthma. To learn more about these and other problems of CF, watch an archived webcast at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**. The CF Foundation is working to find ways to prevent or treat all of these complications of CF.

Complication	Percentage of People with CF (%)
Gastroesophageal reflux (GERD)	26.7
Distal Intestinal Obstructive Syndrome (DIOS)	4.2
Asthma	22.2

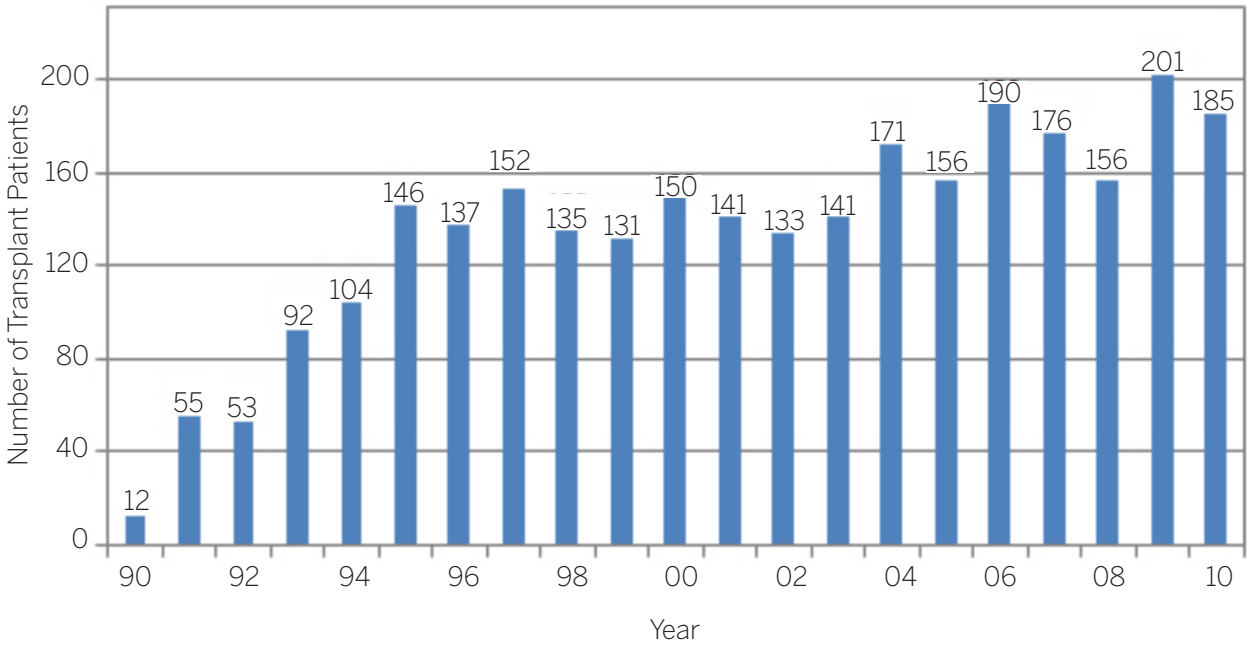
TRANSPLANTATION AND END-OF-LIFE CARE

Goal 6: People with CF and their families will be supported by their CF health care professionals 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 good donor lungs for transplant is limited and transplant is risky. It is important to know who can benefit from a lung transplant and when the time has come for a transplant.

Research based on data from the Patient Registry is used to help find out which person might benefit most from transplantation. To learn more about lung transplants and organ donation, watch the CF Education webcasts on lung transplantation, when considering, the evaluation process and a patient perspective at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**. You can find more information at **www.OrganDonor.gov** or **http://transplantliving.org**.

Number of CF Patients Who Had a Lung Transplant Each Year, 1990-2010



ACCESS TO 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.

Research suggests that people with CF who live in households with lower incomes are more likely to have lower lung function and lower BMI or BMI percentile. This pattern of poor 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 United States get their prescribed FDA-approved drugs and devices, regardless of health insurance coverage or financial resources.

To learn more, visit the CF Patient Assistance Foundation's website at **www.cfpaf.org** or call toll free 1-888-315-4154.

In addition, the Cystic Fibrosis Services Pharmacy, a full-service, mail-order pharmacy and wholly owned subsidiary of the CF Foundation, offers CF-specific patient assistance programs to help people with CF get their medications. To learn more, visit **www.cfservicespharmacy.com**.

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 webcasts "CF Healthcare Coverage" and "Building Life Skills to Manage CF" at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**.

If you have trouble paying for health care, contact your local CF care center to find out what kind of help is available in your area. Your CF care center is the best source for up-to-date information on health care coverage programs in your state. To learn more about patient assistance programs, visit **www.cff.org/LivingWithCF**.

To see information about the self-reported races and ages of the people in the Patient Registry, see the "Summary of the 2010 Data" on page 25.

Insurance Coverage 2010*

	17 and Younger	18 and Older
Number of Patients	13,822	12,476
Health Insurance Policy (e.g. Private Insurance)	56.7	63.8
Medicare/Indian Health Services	1.2	15.5
Medicaid/State Programs	51.2	34.7
TriCare or Other Military Health Plan	2.6	1.9
Other	2.1	2.7
No Health Insurance	0.7	2.3

*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 2010.

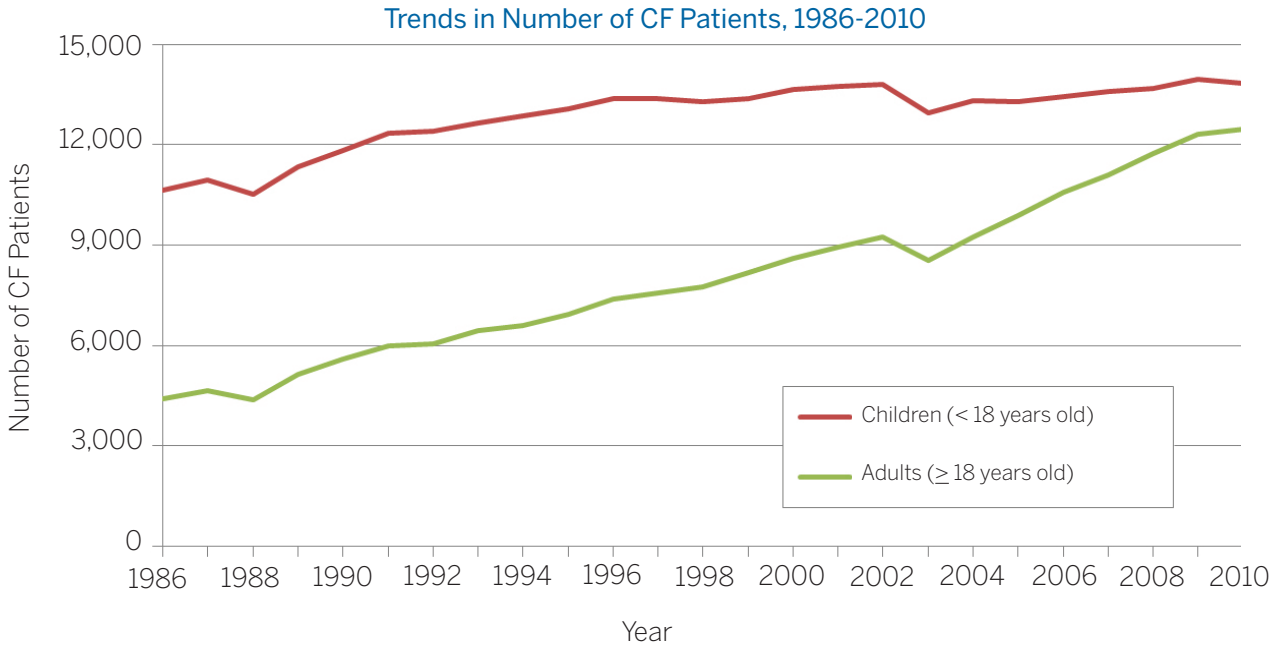
The Foundation supports changes to health care that help people with CF. Health care reform that passed in 2010 includes laws allowing children to stay on their parents' health insurance until the age of 26. Among people with CF in the patient registry, 43.1 percent of adults age 18 to 25 were covered by their parents' health insurance.

OTHER SIGNS OF IMPROVING HEALTH AND LIFE

The CF Foundation's Patient Registry also looks at other measures beyond the seven goals for CF care to study how the health and quality of life for people with CF is improving. The following information looks at adults with CF, the importance of CF clinical research and other data about people with CF in the Patient Registry.

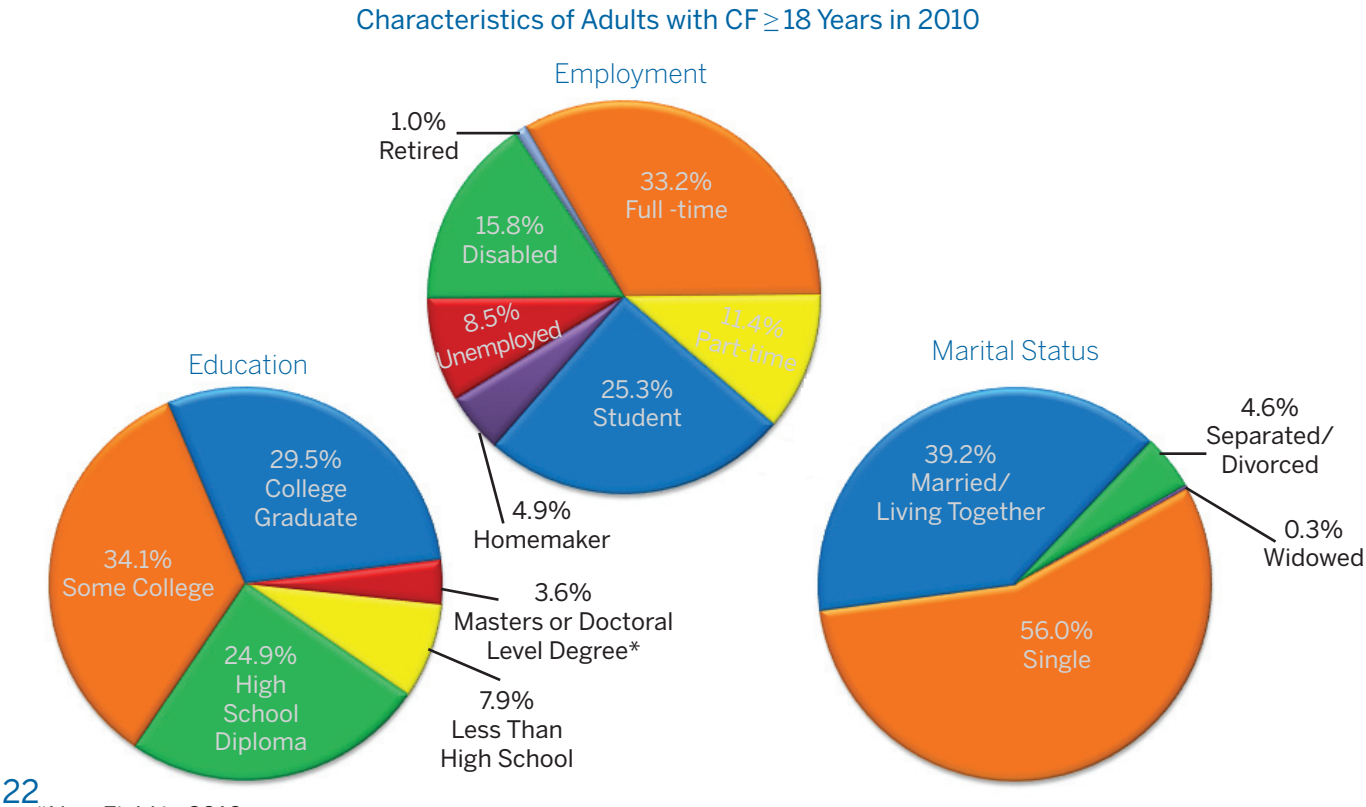
Adults with CF

This graph shows how fast the number of adults with CF is growing in relation to the number of children with CF in the Patient Registry.



In 1990, about 30 percent of people in the Patient Registry were age 18 or older. In 2010, more than 47 percent of people with CF in the Patient Registry were adults, and that number continues to grow. The CF Foundation supports adult programs in the CF care centers and gives grants to help more doctors train in the care of adults with CF. To learn more about adult care, read the CF Foundation guidelines at **www.cff.org/treatments/CFCareGuidelines**.

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



*New Field in 2010

It is important to help teens as they move from relying on their parents or another adult to taking charge of their health to managing their own health. Your care center can help teach children and teens how to manage CF.

To learn more about CF adult care and the transition from pediatric to adult clinics, watch the "Partnering for Care" webcast series at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**.

CF and Pregnancy

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 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 2010, the Patient Registry reported that 225 women with CF were pregnant. Talk with your doctor to learn what you should think about before starting a family. To learn more about CF male and female fertility issues, watch the webcasts at **www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts**.

Clinical Trials

All drugs must be proven safe and effective and approved by the FDA before a doctor can prescribe them. To do this, they must be tested in people with CF during clinical trials. People with CF play a critical role by volunteering to take part in clinical research.

In April 2011, thanks to the CF Foundation's efforts, a new law went into effect that would allow more people with CF to learn, ask and join clinical trials. Those with CF who receive Supplemental Security Income (SSI) can now accept compensation for participating in clinical trials without losing eligibility for SSI benefits. To learn more about the law and how it affects clinical trial participation, visit **www.cff.org/aboutCFFoundation/NewsEvents/4-4-Improving-Access-to-Clinical-Trials-Act.cfm**.

The CF Foundation encourages you to consider joining a clinical trial. To learn how you can help find a cure for CF, ask your care center about clinical trials. You can also search for clinical trials at **www.cff.org/research/ClinicalResearch/Find**.

You can learn more about the CF Foundation's Drug Development Pipeline at **www.cff.org/research/DrugDevelopmentPipeline**.

WHO ARE THE PATIENTS IN THE CF FOUNDATION'S PATIENT REGISTRY — A SUMMARY OF THE 2010 DATA.

	2000	2010
People with CF (number)	22,240	26,272
Newly diagnosed in 2010 (number)	966	882
Age at diagnosis (median)	6 months	5 months
Age range (years)	0-74	0-82
Total number of deaths in 2010	421	407
Predicted median survival	31 years	38.3 years
Predicted median survival (years) – 95% confidence interval	29.2-32.6	35.5-41.1
Patients 18 years and older (%)	38.7	47.5
Race/Ethnicity (%)		
White (Caucasian)	95.3	94.3
Black or African American	3.9	4.3
Hispanic (any race)	5.3	6.9
Male (%)	52.9	51.7
Home Therapy		
Home IV antibiotics	20.7	18.0
Received any oxygen	6.5	11.5
Nutrition		
Pancreatic enzyme use (%)	92.6	86.2
Median BMI percentile (2-20 years)* (%)	40.9	48.7
Median BMI (>20 years)*	21	21.9
Supplemental Feeding		
Tube	8.7	10.6
Oral only	27.8	38.7
Respiratory		
FEV_1 percent predicted (mean)	72	76.8
Respiratory cultures positive for (%)		
Pseudomonas aeruginosa (P. aeruginosa)	58.8	51.2
Multiple drug resistant *P. aeruginosa*‡	-	19
B. cepacia complex	3.2	2.5
*Staphylococcus aureus*** (*S. aureus*)	49.8	67
MRSA	6.1	25.7
Therapies***		
Dornase alfa (i.e., Pulmozyme®)	56.6	78.4
Hypertonic saline‡	-	49.9
Tobramycin solution for inhalation (i.e., TOBI®)	58.7	70.2
Chronic oral macrolide antibiotic (i.e., not to treat an exacerbation)‡	-	70.2
High-dose ibuprofen (e.g., 25-30 mg/kg)	4.2	3.5
Problems related to CF (complications %)		
Liver disease	6.6	4.9
Nasal polyps requiring surgery	2.6	4
Depression	3.1	12
Transplants (number)		
Lung (any type)	165	185
Liver	19	14
Kidney‡	-	5

	2000	2010
Clinical research participation	1,813	5,965
Pregnancies of females with CF	180	225
Live birth rate	1.9	1.9

‡Not available in 2000.

*The Centers for Disease Control and Prevention provide Body Mass Index (BMI) calculators. The national goal for children with CF ages 2-20 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**.

**Includes people with CF with MRSA.

***This is the percentage of patients who are eligible for a therapy and had it prescribed at least once in 2010.

NUMBER OF PATIENTS BY STATE IN THE CF PATIENT REGISTRY

State	Number	Percent
Alabama	378	1.44
Alaska	58	0.22
Arizona	343	1.30
Arkansas	249	0.95
California	2,195	8.35
Colorado	547	2.08
Connecticut	305	1.16
Delaware	62	0.24
District of Columbia	22	0.08
Florida	1,264	4.81
Georgia	759	2.89
Hawaii	5	0.02
Idaho	165	0.63
Illinois	1,027	3.91
Indiana	613	2.33
Iowa	396	1.51
Kansas	344	1.31
Kentucky	527	2.00
Louisiana	310	1.18
Maine	229	0.87
Maryland	486	1.85
Massachusetts	825	3.14
Michigan	1,021	3.88
Minnesota	567	2.16
Mississippi	232	0.88
Missouri	691	2.63

State	Number	Percent
Montana	110	0.42
Nebraska	226	0.86
Nevada	173	0.66
New Hampshire	195	0.74
New Jersey	651	2.48
New Mexico	123	0.47
New York	1,566	5.95
North Carolina	844	3.21
North Dakota	70	0.27
Ohio	1,471	5.59
Oklahoma	218	0.83
Oregon	341	1.30
Pennsylvania	1,389	5.28
Puerto Rico	1	0.00
Rhode Island	96	0.37
South Carolina	348	1.32
South Dakota	101	0.38
Tennessee	562	2.14
Texas	1,536	5.84
Utah	361	1.37
Vermont	138	0.52
Virginia	621	2.36
Washington	607	2.31
West Virginia	229	0.87
Wisconsin	610	2.32
Wyoming	48	0.18
Foreign	9	0.03

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 2010

SUGGESTED CITATION:
Cystic Fibrosis Foundation Patient Registry
2010 Annual Data Report
Bethesda, Maryland

Cystic Fibrosis Foundation

Adding *tomorrows* every day.

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