

# Patient Registry

Annual Data Report 2008





| Adding *tomorrows* every day.

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Patients and families,

The Cystic Fibrosis Foundation's mission is to develop the means to cure and control cystic fibrosis (CF) and to improve the quality of life for those with the disease. We are encouraged by the improvements in CF care shown in this report. With your help, we believe we can make CF care even better.

A strong partnership among people with CF, families and healthcare providers working as a team is critical to achieve the best possible health outcomes. Our intention in reporting these data, as well as the health outcomes data from each CF Foundation-accredited care center, available on our Web site ([www.cff.org](http://www.cff.org)), is to educate and help build strong relationships through open communication and information sharing.

Locally, CF care centers have invited people with CF and their families to serve as members of quality improvement teams, act as center advisors, form peer-support groups and share information via newsletters.

We encourage you to use this report and the health outcomes data on [www.cff.org](http://www.cff.org) to start a conversation with your care center staff about improving CF care. Across the country, CF care centers are focusing on quality improvement to advance care, but they need your help. They need your input, your opinions and your involvement. After all, you are the expert in receiving care from a CF center and living with the day-to-day challenges of CF.

We hope you will learn more about what you can do to improve CF care for you or your child and join us in our quest to add *tomorrows* every day to the lives of those with CF.

Sincerely,

A handwritten signature in black ink that reads "Bruce C. Marshall".

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

A handwritten signature in black ink that reads "Leslie Hazle".

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

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## WHAT IS THE CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY?

The Cystic Fibrosis Foundation's Patient Registry tracks the health of people with cystic fibrosis (CF) across the U.S. This information includes data on the more than 25,500 people who receive care at CF Foundation-accredited care centers. It is collected and added to the Patient Registry every year. The collected data includes state of residence, height, weight, gender, genotype, pulmonary function test (PFT) results, pancreatic enzyme use, length of hospitalizations, home IV use and complications related to CF.

It is important that people with CF agree to have their data entered into the Patient Registry. The information helps CF caregivers and researchers identify new trends in the health of people with CF, develop care guidelines, design clinical trials to test new therapies and improve the delivery of care. The following pages contain information from the 2008 Patient Registry, including the health of people with CF, guidelines for care and the areas where more work can be done.

The CF Foundation's seven goals to accelerate the rate of improvement in CF care are:

- 1) People with CF and their families are full members of the care team;
- 2) Children and adolescents will have normal growth and nutrition. Adults' nutrition will be kept as near normal as possible;
- 3) People with CF will receive the right therapies to keep lung function steady and reduce infections. Infections will be diagnosed early and treated;
- 4) To decrease the spread of germs between people with CF;
- 5) People with CF will be screened for complications and treated as needed;
- 6) To provide care regardless of race, age, education or insurance coverage; and
- 7) To support all transplantation and end-of-life care decisions.

The information presented in this report is based on these seven goals.

## ABOUT CYSTIC FIBROSIS

### 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 problems in the lungs and digestive system. Many people with CF have lung infections and inflammation (swelling). These slowly damage the lungs and reduce their ability to provide oxygen to the body. Digestive problems prevent people with CF from absorbing food, making it difficult 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. However, it is more common among Caucasians. An estimated 30,000 people in the United States have the disease.

### WHAT IS THE CYSTIC FIBROSIS FOUNDATION?

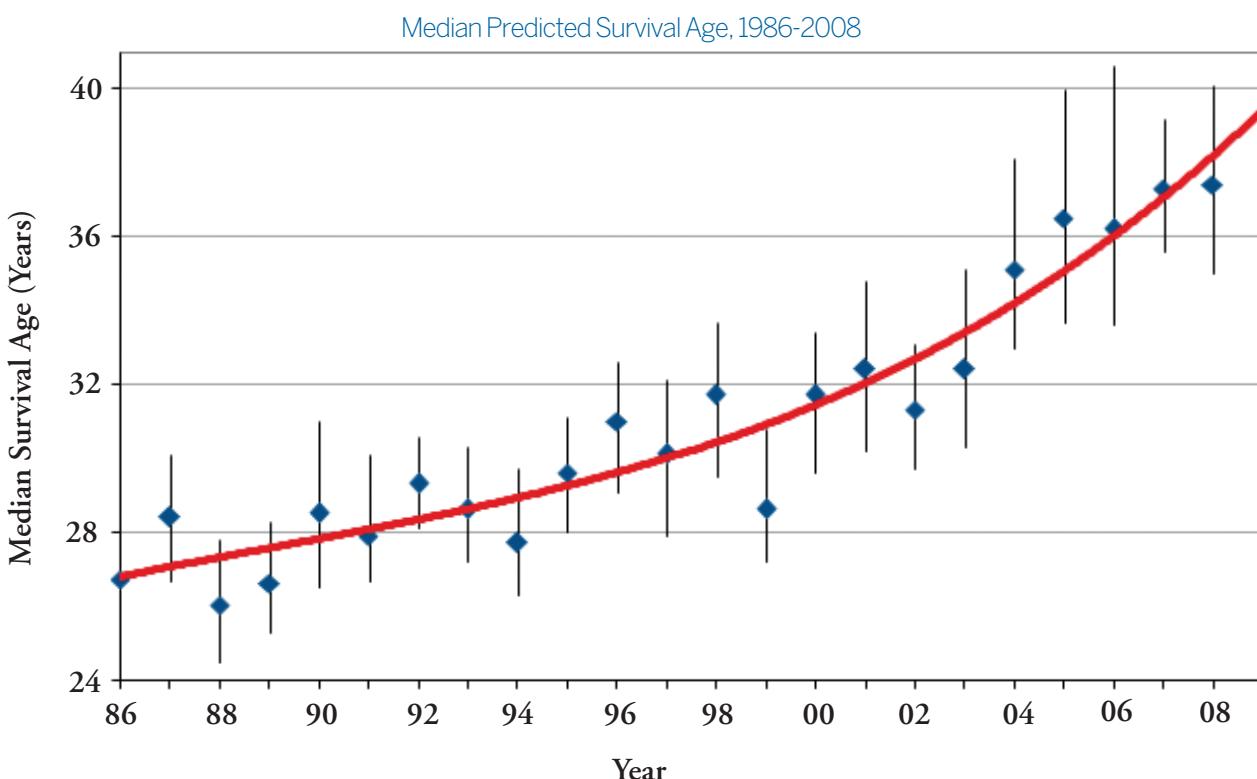
The CF Foundation was started in 1955 by a group of parents who had children with CF. They had a clear mission — to develop the means to cure and control CF and to improve the quality of life for those with the disease.

To further this mission, the Foundation established a network of about 110 accredited care centers across the United States to care for people with CF. The Foundation provides care centers with grants, training in quality improvement, the latest CF care guidelines based on the medical literature and Patient Registry data to support the care of people with CF. The Foundation also awards grants to researchers working to learn more about CF and to discover and develop new therapies that improve the length and quality of life for those with the disease. In addition, the Foundation supports the development of drugs that treat the symptoms of CF as well as the underlying genetic cause of the disease. The goal is to have therapies that treat many different symptoms as well as the basic defect. A group of 77 Foundation-accredited care centers form a clinical trials network that help enable the development of new drugs. To learn more about CF, the Foundation and its drug development pipeline, visit [www.cff.org](http://www.cff.org).

## SURVIVAL IS IMPROVING

Because of the hard work and strong partnership among people with CF, their families and CF Foundation-accredited care centers, the median predicted survival age for people with CF is steadily improving. When the CF Foundation was established in 1955, few children with CF lived long enough to attend elementary school. Today, the median predicted survival age is 37.4 years. The graph below shows how much this number has increased since 1986.

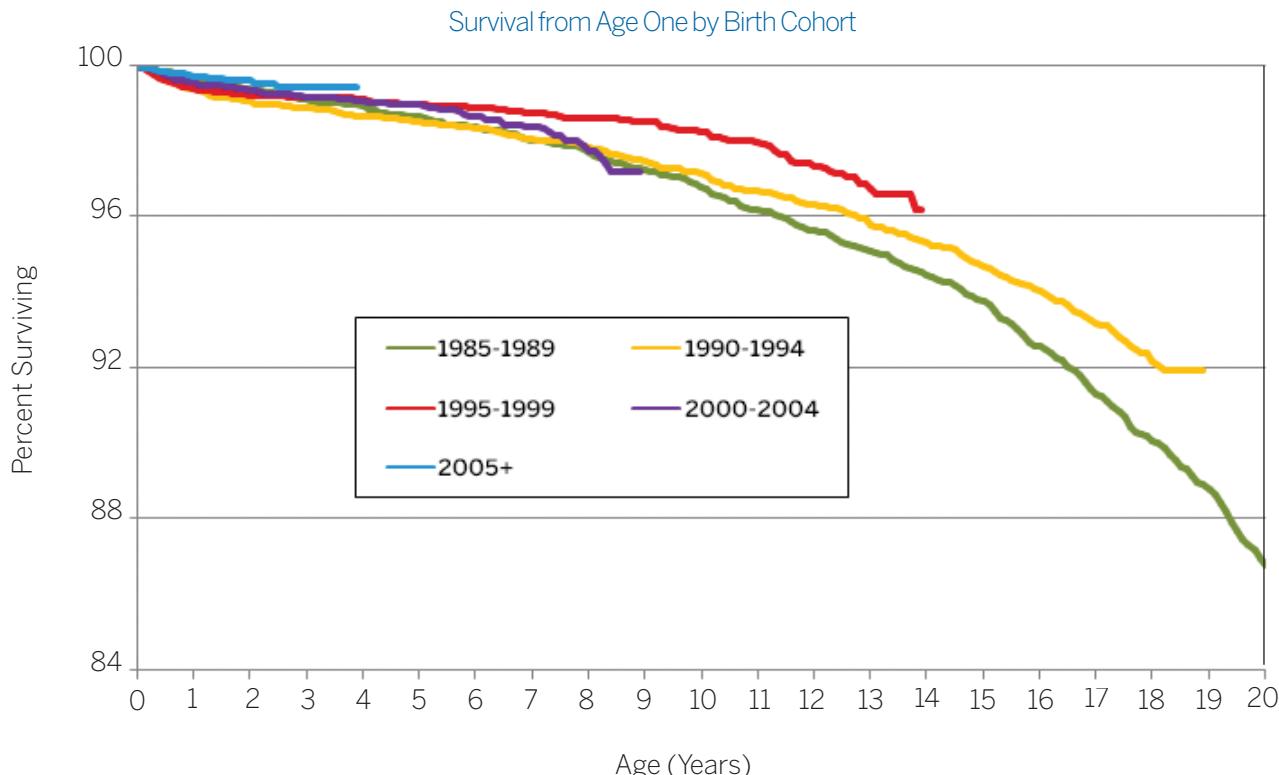
Median predicted survival age represents the age at which half of the people with CF currently in the Patient Registry are expected to survive. This number is calculated every year and is based on the deaths that occurred during that year. Continued improvement depends, in part, on gathering and using data in the Patient Registry from people with CF across the United States. The Foundation and its care centers continue to build partnerships with people with CF and their families to improve care and keep the median predicted survival age increasing.



Because of the Foundation's and the CF community's efforts, all 50 states and the District of Columbia are screening newborns for CF. The earlier CF is diagnosed, the sooner treatment can begin, slowing the course of the disease and preventing complications. Earlier diagnosis and treatment is one way to improve the quality and length of life for those with CF.

Research suggests that early treatment may help maintain lung health in infants. CF researchers are working to find a drug that maintains lung function and could be started in infants with CF right after diagnosis. The Phase 2 Infant Study of Inhaled Saline (ISIS) study is examining the effects of using inhaled hypertonic (7%) saline in children under 5 years of age. This may help prevent lung damage from the build up of mucus in the lungs. Visit [www.cff.org/research/ClinicalResearch/Find](http://www.cff.org/research/ClinicalResearch/Find) for more information.

The graph at the top of the next page is another way to show that survival continues to improve. Of people with CF born between 1985 and 1989 (green line), 88 percent were alive at age 19. For children born between 1990 and 1994 (yellow line), 92 percent were alive at age 19.



**Goal 1:** People with CF and their families are full members of the care team. Communication will be open so everyone can be involved in decisions about care. Care will be respectful of the individual's needs, preferences and values.

Through the CF Foundation's Patient Registry Annual Data Report, care centers record data about their patients and the combined health of people with CF throughout the United States. These reports help care centers talk with their patients and families about some of the ways patients can stay healthy with CF. One report is the "Patient Summary Report" on the next page. It shows trends in lung function, nutrition and other important information about an individual with CF. Ask your CF care center for a copy of your or your child's "Patient Summary Report" at your next CF clinic visit.

The Patient Registry also shows health trends of people with CF in the United States. Information on the CF Foundation's Web site ([www.cff.org/LivingWithCF/CareCenterNetwork/CareCenterData](http://www.cff.org/LivingWithCF/CareCenterNetwork/CareCenterData)) shows the average lung function and body mass index (BMI) for people with CF who are cared for at an individual center. The purpose of this data is to encourage people with CF and their families to get involved with their center to improve CF care. The data are updated every year. We encourage you to start a conversation with your care center about your center's data.

The following questions are good questions to ask.

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

Partner with your care center and be a full and active member of your CF care team.

To learn more about these data and how to work with your care center, watch the archived Web casts "One Team's Story: Raising the Bar for CF Care" and "Quality CF Care Is More Than the Numbers" at [www.cff.org/LivingWithCF/Webcasts](http://www.cff.org/LivingWithCF/Webcasts). You can also read the success stories of how others work with their care centers to improve care at [www.cff.org/LivingWithCF/QualityImprovement](http://www.cff.org/LivingWithCF/QualityImprovement).

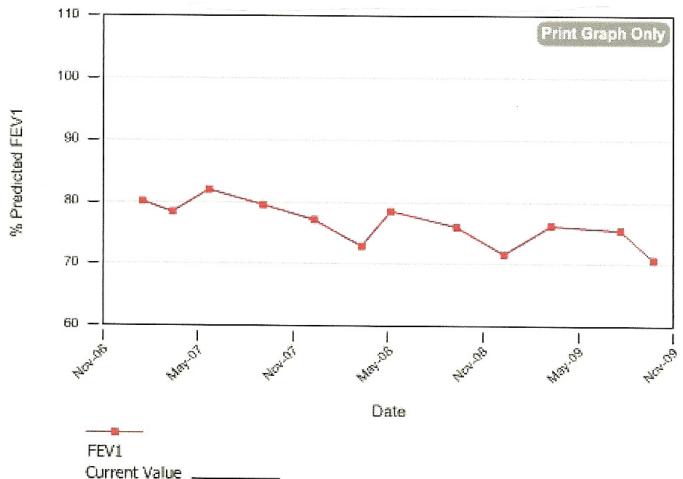
## Patient Report Example:

VISIT DATE: \_\_\_\_\_ Name: \_\_\_\_\_  
 Last Hospitalization: \_\_\_\_\_ /2006 - /2006 Date of Birth: \_\_\_\_\_ /1963  
 Last HomeIV Therapy: \_\_\_\_\_ /2006 - /2006 Genotype: Date Unknown ^F508 /  
 Last Clinical Visit: \_\_\_\_\_ /2009 ^F508

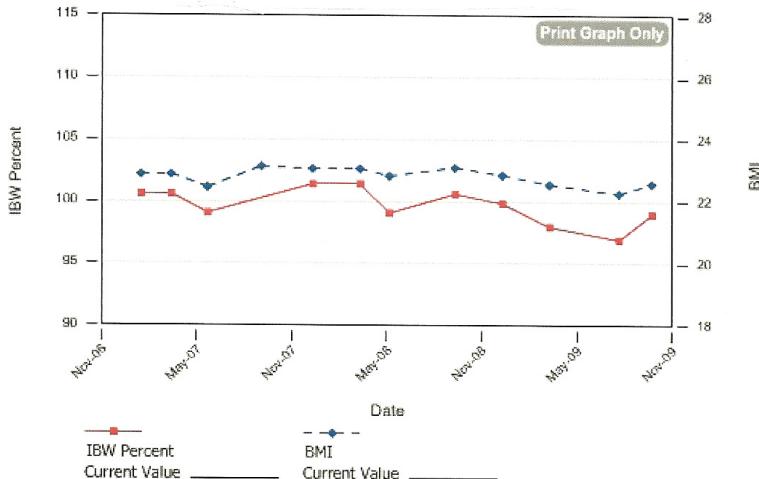
### Culture Results

Last Culture:	Pseudomonas aeruginosa, Staphylococcus aureus, :	/2009		
Last Positive:	B. cepacia None	MRSA /2006	PA /2009	MDR-PA+ /2009

### PFT Trend



### Nutritional Trend



### Complications

#### ACTIVE @ LAST VISIT

- CFRD
- Peptic ulcer disease
- Sinus Disease (symptomatic)

#### COMPLICATIONS PREVIOUSLY NOTED

- CFRD
- Dist Int Obst Synd (DIOS)
- Peptic ulcer disease
- Sinus Disease (symptomatic)

### Routine Evaluations

Last PFT: \_\_\_\_\_ /2009  
 Last CXR: 2008  
 Last SW Visit: \_\_\_\_\_ /2004

Last Dietary Visit: \_\_\_\_\_ /2008  
 Last LFT: \_\_\_\_\_ /2009  
 Glucose Screening: \_\_\_\_\_ /2004  
 Creatinine: \_\_\_\_\_ /2009

<sup>+</sup>Prior to 2003, multi-drug resistant *Pseudomonas Aeruginosa* (MDR-PA) status cannot be determined based on registry data.

## GUIDELINES FOR CF CARE

The CF Foundation gathers CF health experts to review the medical literature, research on CF care and data from the Patient Registry to develop guidelines for the care of everyone with CF. These experts recommend that each year people with CF should have at least:

- Four CF clinic visits;
- Four respiratory cultures, either by throat swab or sputum;
- Two pulmonary function tests (PFTs) if physically able;
- An influenza (flu) vaccine if  $\geq$  6 months of age;
- Vitamin A, D and E levels in the blood measured (sometimes called fat-soluble vitamin levels);
- An oral glucose tolerance test (OGTT) to screen for CF-related diabetes in people with CF age 10 years and older; and
- A test to measure liver enzymes in the blood (sometimes called a liver function test).

Below is an overview of how people with CF in the Patient Registry met these guidelines in 2008.

Guidelines for CF Care	People with CF who met the guidelines (%)
4 or more clinic visits	63.9
3 or more respiratory cultures, by throat swab or sputum	41.0
2 or more pulmonary function tests (PFTs) if physically able	83.3
Influenza (flu) Vaccine if $\geq$ 6 months of age	67.5
Fat Soluble Vitamin Levels Measured	82.7
Oral Glucose Tolerance Test (OGTT) if age 10 or older	12.8
Liver Function Test	77.8

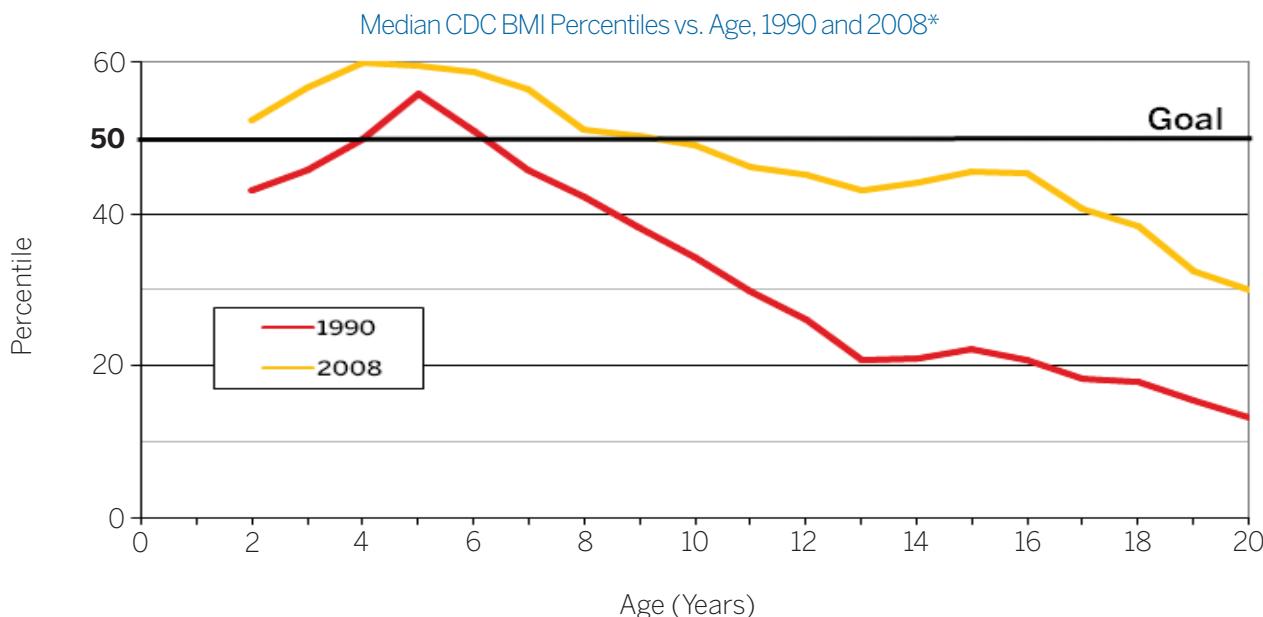
## MAINTAINING NORMAL NUTRITION

Goal 2: Children and adolescents will have normal growth and nutrition. Adults' nutrition will be kept as near normal as possible.

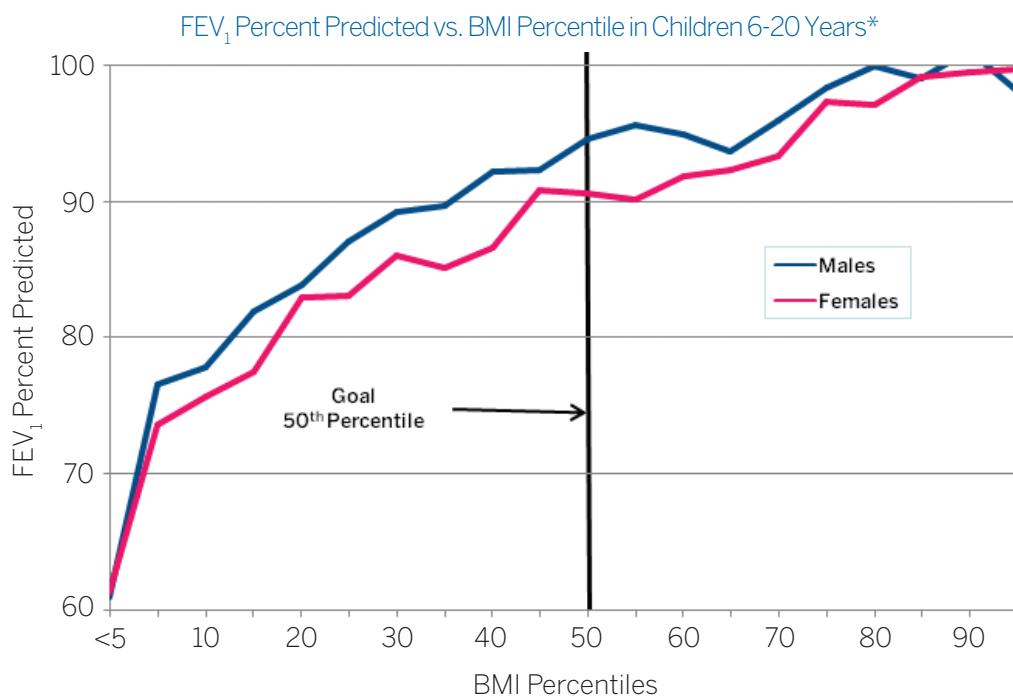
The nutrition of people with CF is getting better, but more work remains. The CF Foundation worked closely with experts in CF nutrition to review research, medical literature and data from the Patient Registry, to develop nutrition care guidelines and to set national body mass index (BMI) goals for people with CF. BMI is based on a person's weight and height and is used to screen people for health problems. For children and teens, BMI is stated as a percentile compared to children without CF of the same age and gender.

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). You can calculate your BMI or your child's BMI percentile on the Centers for Disease Control and Prevention's Web site ([www.cdc.gov/healthyweight/assessing/bmi](http://www.cdc.gov/healthyweight/assessing/bmi)).

For children with CF, the goal is to have a BMI around the 50th percentile for their age, which is the average BMI percentile for children in the United States who do not have CF. Ideally, children with CF should grow and develop the same as children without CF. The graph on the top of the next page shows how much the BMI percentile of children with CF has improved since 1990.



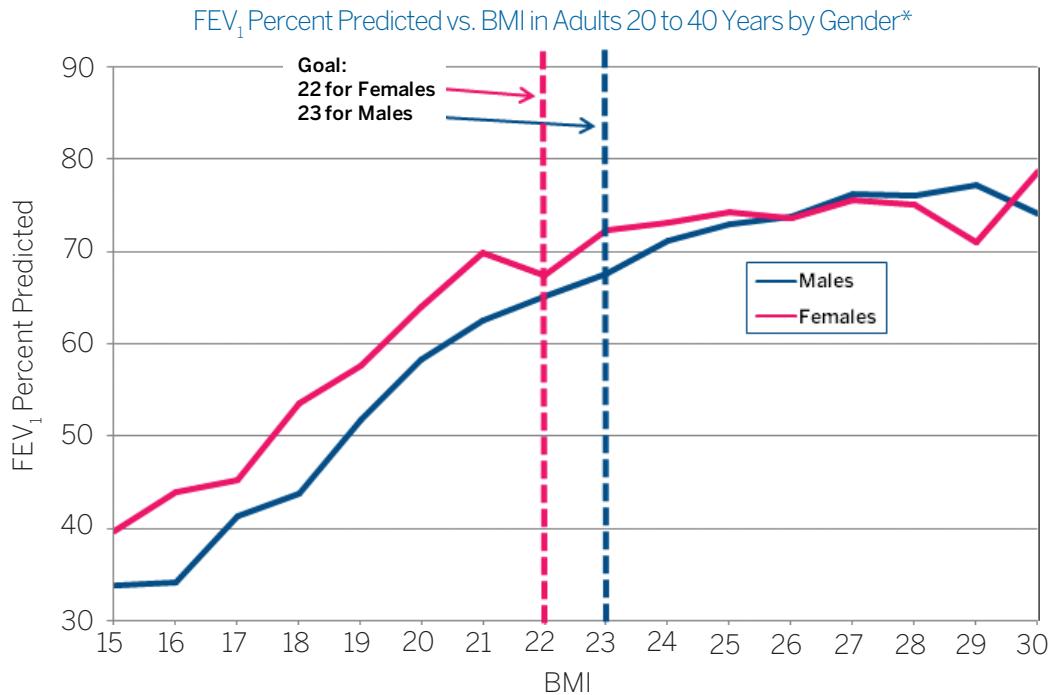
The Patient Registry shows a strong connection between a higher BMI percentile and better lung function in children. Lung function is measured by FEV<sub>1</sub>, or Forced Expiratory Volume over one second. This is shown as percent predicted based on healthy, non-smoking people of the same gender and age. The graph below shows that good nutrition and lung health seem to go hand in hand. The highlighted bar is the BMI percentile goal for children with CF.



The connection between a higher BMI and better lung function is also seen in adults with CF. Highlighted on the graph at the top of the next page are the national BMI goals for men and women with CF. For men, BMI should be 23 and for women, BMI should be 22.

To improve and/or maintain your or your child's weight, the Foundation suggests that you work closely with your CF care center. To learn more about CF and nutrition, ask your CF care center or visit [www.cff.org](http://www.cff.org).

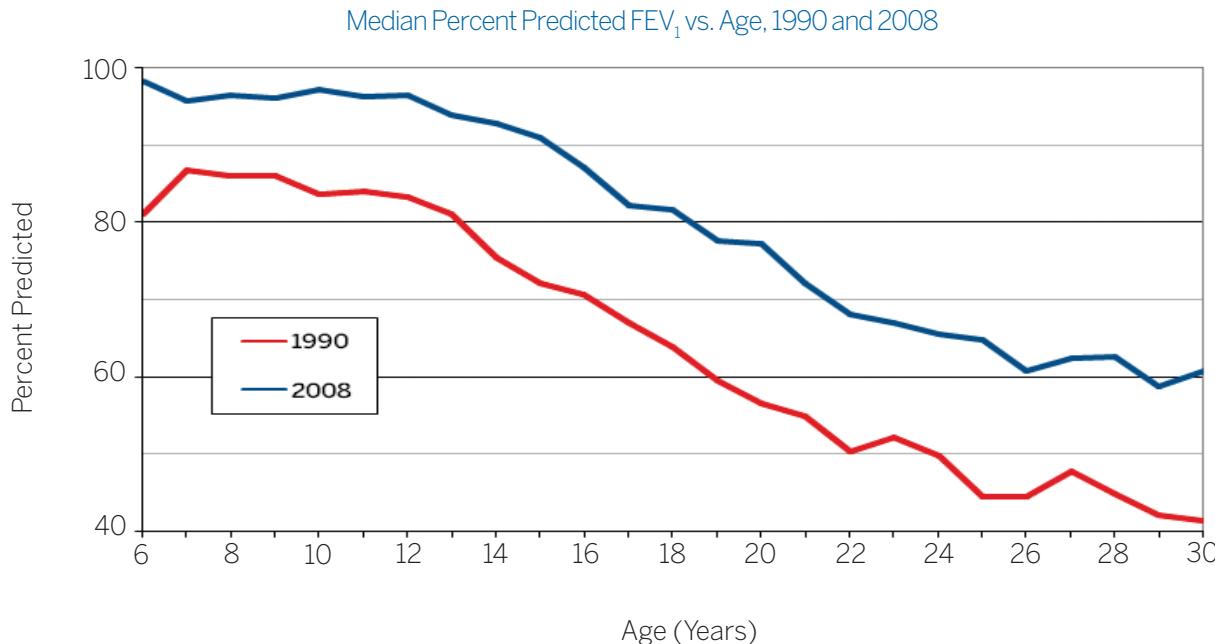
\* Centers for Disease Control and Prevention — [www.cdc.gov/healthyweight/assessing/bmi](http://www.cdc.gov/healthyweight/assessing/bmi)



## LUNG FUNCTION

Goal 3: People with CF will receive the right therapies to keep lung function steady and reduce infections. Infection will be diagnosed early and treated.

The graph below shows that there have been major improvements in the lung health of people with CF since 1990. FEV<sub>1</sub> (lung function) is usually near normal or just under 100 percent predicted when first measured around 6 years of age.

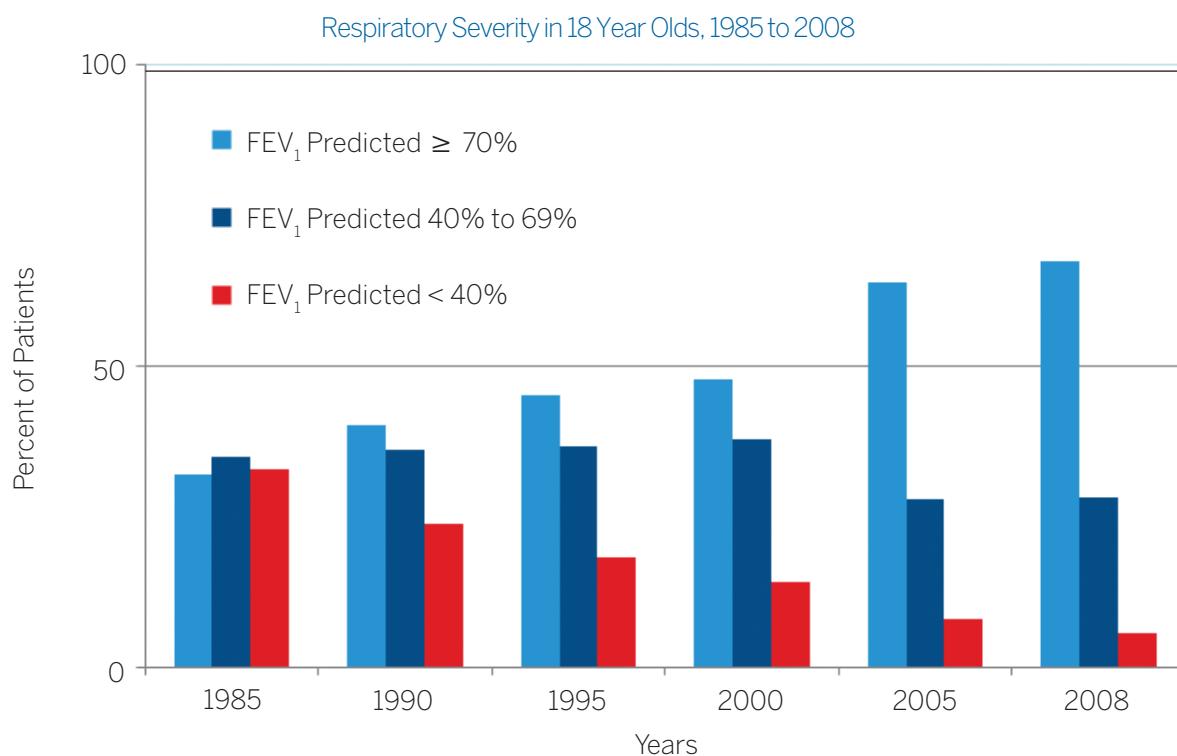


\* Centers for Disease Control and Prevention — [www.cdc.gov/healthyweight/assessing/bmi](http://www.cdc.gov/healthyweight/assessing/bmi)

Severity of lung disease is based on a person's lung function or FEV<sub>1</sub> percent predicted. In CF, the lower a person's FEV<sub>1</sub>, the more severe the lung disease. An FEV<sub>1</sub> greater than or equal to 90 percent is normal. An FEV<sub>1</sub> between 70 and 89 percent means mild lung disease. An FEV<sub>1</sub> between 40 and 69 percent indicates moderate lung disease. If the FEV<sub>1</sub> is less than 40 percent, severe lung disease is present.

Researchers are working to find new therapies to slow or stop lung function decline, improve FEV<sub>1</sub> percent predicted and keep the lungs of people with CF as healthy as possible. To learn more about CF research, visit [www.cff.org](http://www.cff.org).

The graph below shows an increase in the number of 18 year olds with CF in the Patient Registry and their lung function. The rising number of people with normal lung function or mild disease and the decreasing number with severe disease shows that the lungs of people with CF are much healthier now than 20 years ago.



In 2007, the "Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Function" was published in the November edition of the *American Journal of Respiratory and Critical Care Medicine*. These guidelines were developed by experts in CF lung disease who reviewed research data and recommended medications that help people with CF maintain lung health. The table on the next page shows the criteria for each medication to help maintain lung function, along with the percentage of people with CF who are taking each one. These criteria are set by the U.S. Food and Drug Administration (FDA) for the medicine.

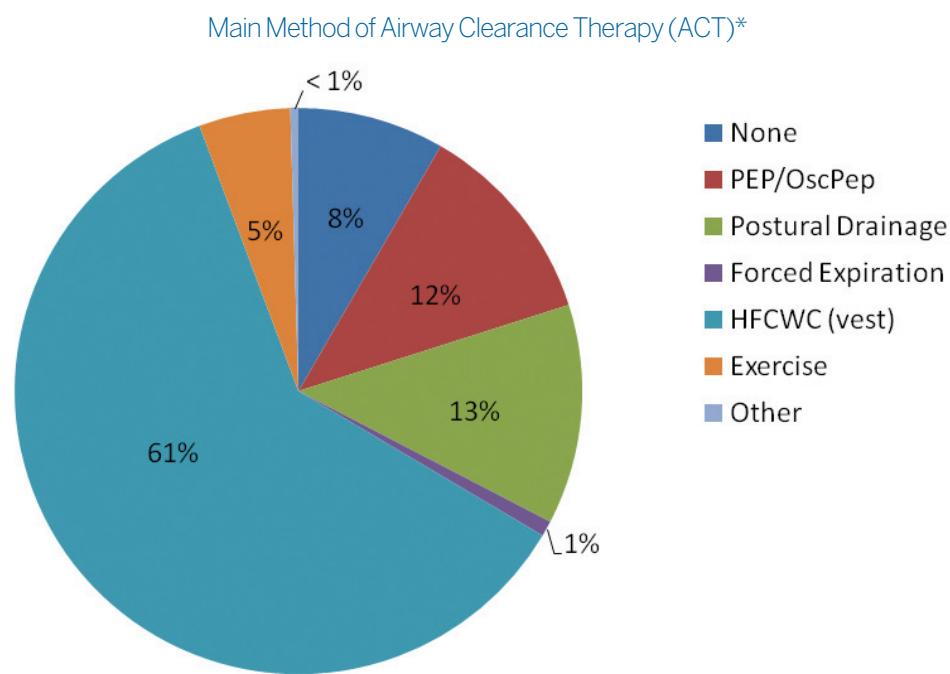
To find out if you or your child might do well on one of these medications, talk to your CF doctor. New therapies can only be available after they are tested by people with CF in clinical trials and approved by the FDA. Consider joining the search for new CF treatments by visiting [www.cff.org/research/ClinicalResearch/Find](http://www.cff.org/research/ClinicalResearch/Find). People with CF play a critical role in clinical trials and making new therapies available.

Talk to your care center to learn about clinical trials. The Foundation supports research to find new therapies and ways that CF centers can partner with people with CF and their families to get and use the right medicines regularly.

Chronic CF medications	Percentage (%) of people who fit the criteria and are on the medication	Criteria for medication
Inhaled tobramycin (TOBI)	67.4	<ul style="list-style-type: none"> <li>≥ 6 years of age</li> <li>Positive for <i>P. aeruginosa</i></li> </ul>
Dornase alfa (Pulmozyme)	76.1	<ul style="list-style-type: none"> <li>≥ 6 years of age</li> </ul>
Macrolides (azithromycin or Zithromax®)	65.6	<ul style="list-style-type: none"> <li>≥ 6 years of age</li> <li>Positive for <i>P. aeruginosa</i></li> <li>Weight ≥ 55 pounds (25kg)</li> </ul>
Ibuprofen	3.8	<ul style="list-style-type: none"> <li>6-12 years of age</li> <li>FEV<sub>1</sub> &gt; 60% predicted</li> </ul>
Hypertonic saline	40.3	≥ 6 years of age

Taking medicine is only one way to help keep CF lungs healthy. Getting the thick mucus out of the lungs is also important. Airway clearance techniques (ACT) help move mucus out of the lungs. The body's normal and most basic ACT is coughing. It is a reflex, which clears mucus with high-speed air-flow. Sometimes mucus cannot be cleared with coughing alone. There are many different airway clearance therapies that people with CF can use to help keep their lungs clear of extra mucus. The method of airway clearance you use varies by person and care center.

In 2007, the CF Foundation gathered experts in CF lung health to look at the latest research and set airway clearance guidelines for CF care. Experts noted that people with CF should do airway clearance to keep their lungs healthy, even when they are not sick. The method used can vary, but it is important that it be done daily. The chart below shows the percentage of people with CF and the methods of airway clearance used to clear the mucus from their lungs. You can learn more about airway clearance at [www.cff.org/treatments/Treatments/Respiratory/AirwayClearance](http://www.cff.org/treatments/Treatments/Respiratory/AirwayClearance).

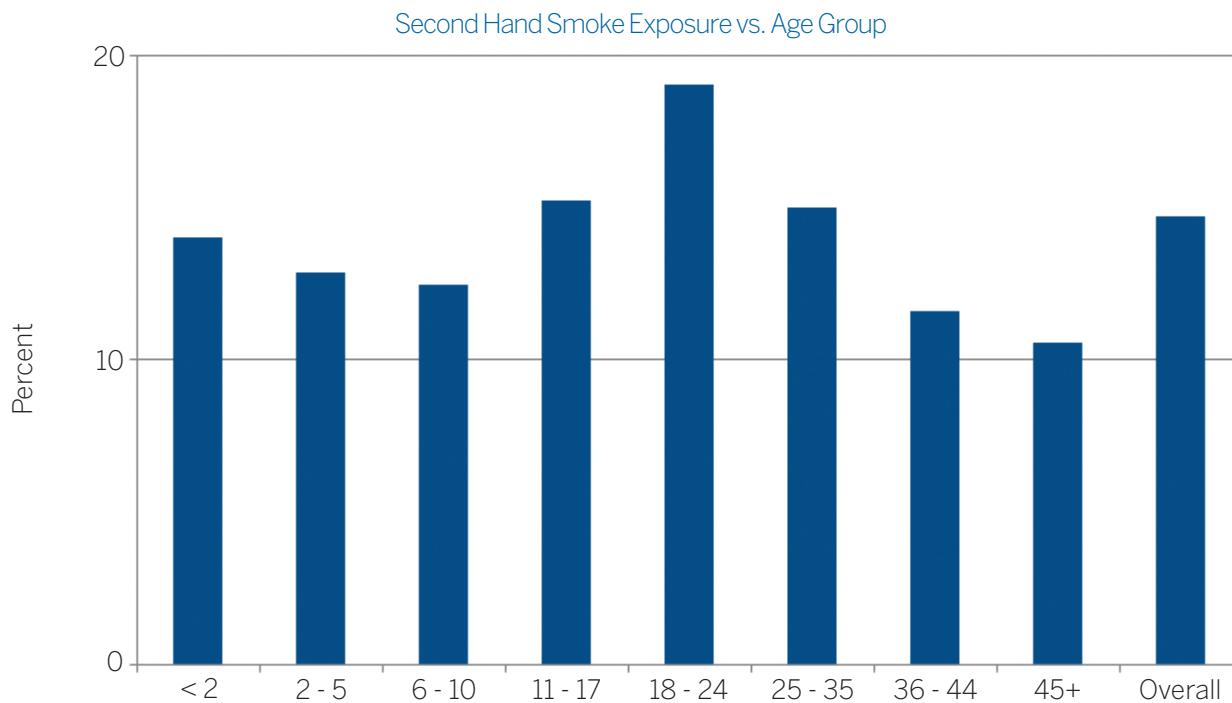


\* American Journal of Respiratory and Critical Care Medicine, September 2009. The "Cystic Fibrosis Pulmonary Guidelines: Treatment of Pulmonary Exacerbations" recommends that airway clearance therapy be done more often during an exacerbation or respiratory infection.

Each time a person with CF has a respiratory infection or exacerbation, there may be lung damage. However, there are things you can do to prevent or lessen the chance of an infection or exacerbation.

- Do airway clearance to keep your lungs as clear as possible of mucus;
- Take the medicines as your CF doctor prescribes;
- Get a flu shot every fall for you or your child and everyone living in the 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, and cleaning and disinfecting nebulizers; and
- Avoid secondhand smoke exposure.

The graph below shows, by age group, the percentage of people with CF who breathe in secondhand smoke. Secondhand smoke comes from burning tobacco in cigarettes, cigars, pipes and the smoke breathed out by people who smoke. It has been shown that children who breathe in secondhand smoke have more respiratory infections. In adults who do not smoke, breathing in secondhand smoke can cause lung cancer and heart disease. The Surgeon General has concluded that breathing even a little secondhand smoke can be harmful to health.



The only way to fully protect yourself and 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 allow anyone to smoke in your home or car;
- Ask people not to smoke around you or your child;
- Teach your child to stay away from people when they are smoking;
- Make sure that your child's day care center or school is smoke-free; and
- Choose restaurants and other businesses that are smoke-free. Thank businesses for being smoke-free. Let owners of businesses that are not smoke-free know that secondhand smoke is harmful to your family's health.

If you are a smoker, the single best way to protect you and your family from secondhand smoke is to quit smoking. In the meantime, only smoke outside to protect your family. Keeping your home and car smoke-free can also help you quit smoking. In addition, your doctor can help you quit smoking. For more information, please visit the “Smoking and Tobacco Use” section of the Centers for Disease Control and Prevention (CDC) Web site ([www.cdc.gov/tobacco](http://www.cdc.gov/tobacco)).

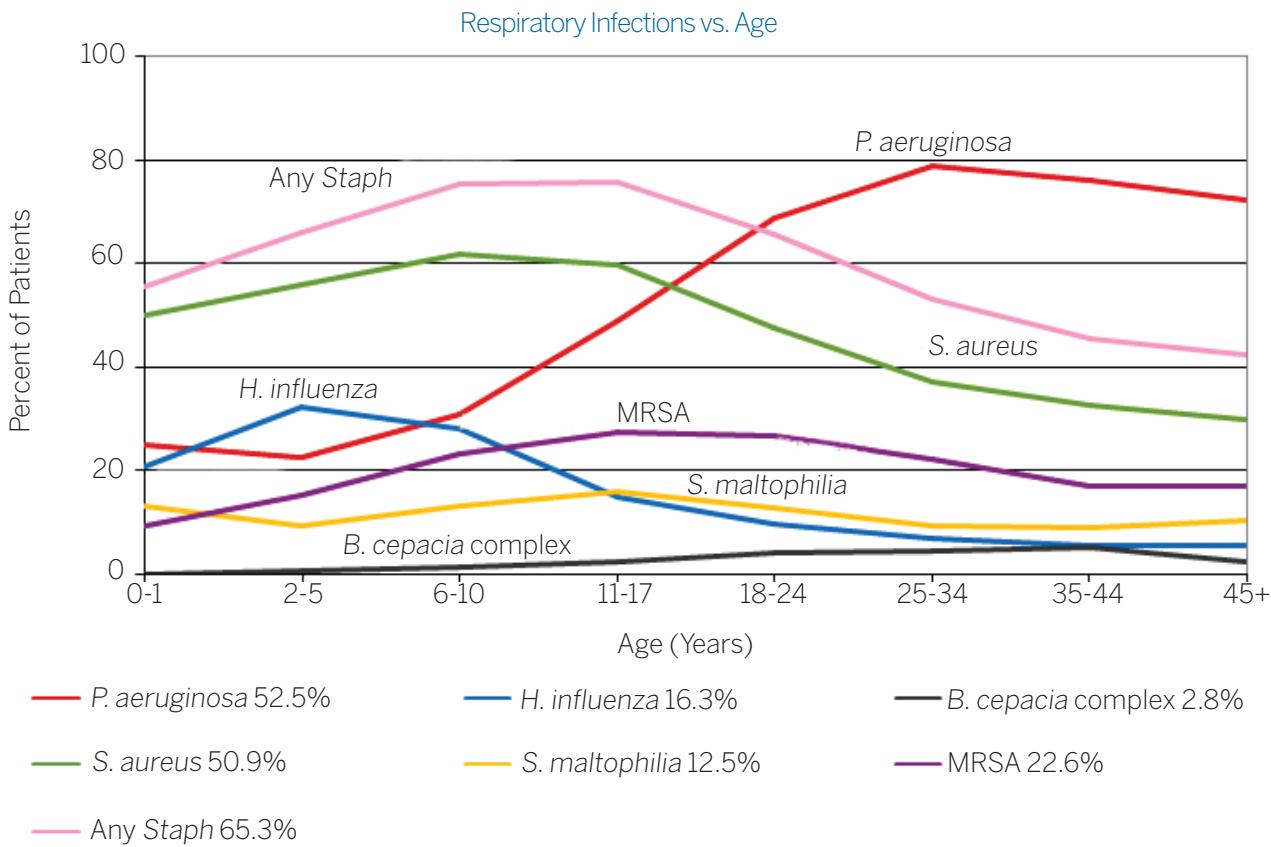
Even if you are careful and do all you can to prevent a lung infection, it is hard to avoid infections completely. If you or your child start to feel ill (e.g., are coughing more, experiencing loss of energy or appetite), call your CF care center so treatment can be started as soon as possible. To learn more about lung care and therapies for people with CF, visit [www.cff.org/LivingWithCF](http://www.cff.org/LivingWithCF). You can also watch archived Web casts about CF lung health and disease. It is important that you and your CF care center work together to create a plan to maintain your or your child’s health.

**Goal 4:** People with CF and their care centers will work together to decrease the chances of getting or spreading respiratory germs, particularly *Pseudomonas aeruginosa* (*P. aeruginosa*) and *Burkholderia cepacia* (*B. cepacia*) complex.

The best way to avoid germs is to:

- Clean your 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. Germs spread this way; and
- If you are ill, stay away from others. This helps stop the spread of germs.

Repeated lung infections are a concern for people with CF. It is the cycle of infection and inflammation that damages the lungs. This damage causes lung function (FEV<sub>1</sub>) to get worse. When the lungs are damaged, infections happen more often. The next graph shows some of the germs that are found in the lungs of people with CF. Talk to your CF care center to learn more about how to avoid respiratory infections. Information about CF germs and Web casts about “Germs, Infection Control and People With CF” are available at [www.cff.org/LivingWithCF/StayingHealthy](http://www.cff.org/LivingWithCF/StayingHealthy). You can also watch “Put Your Hands Together” at [www.cdc.gov/CDCTV/HandsTogether](http://www.cdc.gov/CDCTV/HandsTogether).



## COMPLICATIONS OF CF

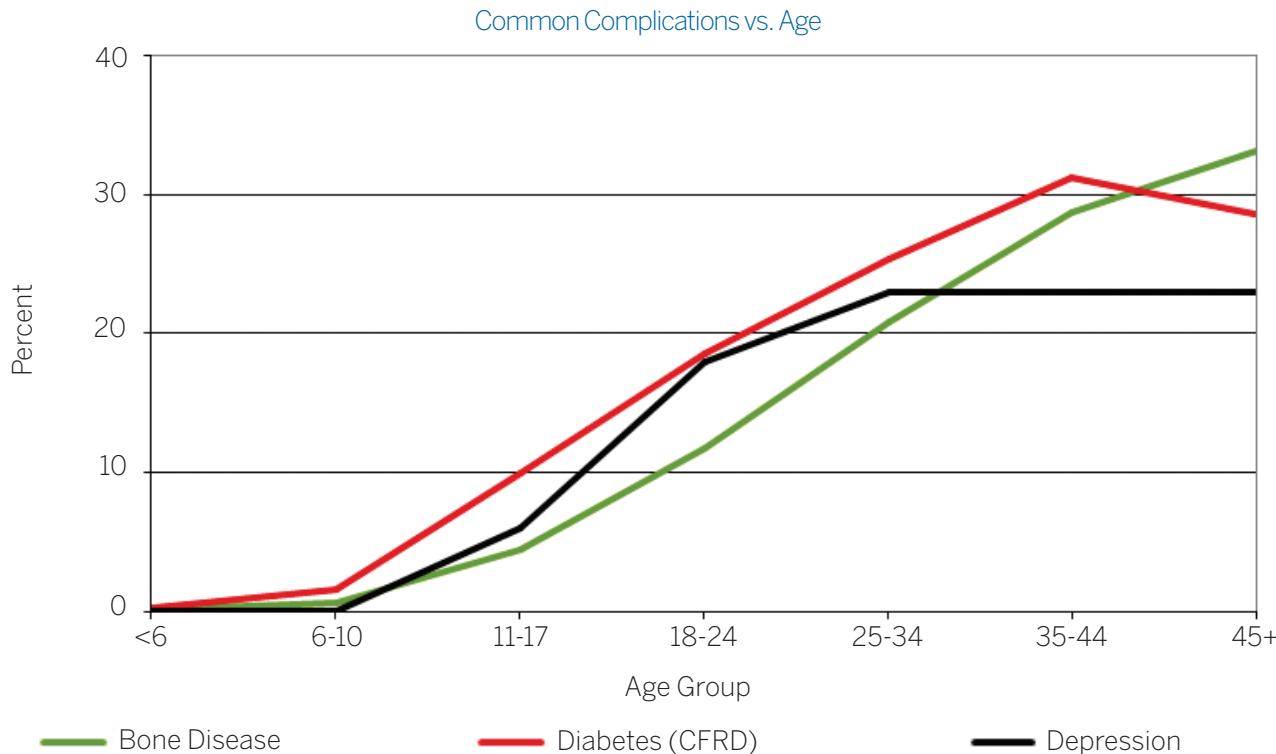
**Goal 5:** People with CF will be screened and treated early for complications of CF, especially CF-related diabetes (CFRD).

Cystic fibrosis-related diabetes (CFRD) is different from diabetes in people without CF because cystic fibrosis damages the pancreas. Anyone with CF, 10 years of age and older, should be tested every year for CFRD. Patient Registry data shows that early diagnosis and treatment of CFRD results in better nutrition and weight gain and, thus, better health. The CF Foundation continues to fund CFRD research. To learn about Foundation-supported research, visit [www.cff.org/research](http://www.cff.org/research).

The Patient Registry shows trends in other complications of CF as well. The importance of good nutrition and healthy bones is often in the news. Data from the Patient Registry show that about 11 percent of people with CF had bone disease in 2008. Preventing or lessening bone disease begins in childhood when bones are growing. Good nutrition, a healthy weight and exercise can help. Ask your CF dietitian or physical therapist what can be done to keep your or your child's bones healthy.

Another finding from the Patient Registry shows that almost 20 percent of adults with CF have symptoms of depression. This is a common complication that is often found in people with other chronic diseases as well. People with CF, their families and caregivers need to be aware of this complication so that diagnosis and treatment can be started early. People often respond well to treatment for depression.

Gastroesophageal reflux (GERD), asthma and sinus disease are some of the other complications people with CF may experience. The CF Foundation continues to promote prevention, early diagnosis and treatment of common CF complications.



## ACCESS TO CARE

Goal 6: Everyone with CF will have access to appropriate therapies, treatments and support regardless of race, age, education or insurance coverage.

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. The CF Foundation is working to find out why this happens in CF and how to change it.

In addition, CF Services Pharmacy, a full-service, mail-order pharmacy and wholly owned subsidiary of the CF Foundation, works hard to make proven CF therapies available to everyone. It runs CF-specific patient assistance programs to help people with CF get CF medications. Visit the CF Services Web site to learn more ([www.cfservicespharmacy.com](http://www.cfservicespharmacy.com)).

There are a number of programs available to help people with CF afford the medical care and medications they need. Many people don't realize that they qualify for these assistance programs. The Cystic Fibrosis Patient Assistance Foundation (a new subsidiary of the CF Foundation) helps people with CF get the medicines they need. Its mission is to ensure that everyone with CF living in the United States, regardless of health insurance coverage or financial resources can get their prescribed FDA-approved drugs for CF lung disease. You can visit the CF Patient Assistance Foundation's Web site ([www.cfpaf.org](http://www.cfpaf.org)) or call toll-free (1-888-315-4154) to learn more.

Data from the Patient Registry shows that therapies like Pulmozyme® and TOBI® are available to people with CF, no matter what their income level. You can learn more about assistance for these medications and hear how others manage CF by watching the archived Web casts "CF Healthcare Coverage" and "Building Life Skills to Manage CF," available [www.cff.org/LivingWithCF/Webcasts](http://www.cff.org/LivingWithCF/Webcasts).

To learn more about race and age of the people in the Patient Registry, "A Summary of the 2008 Data" can be found on page 18.

### Insurance Coverage 2008

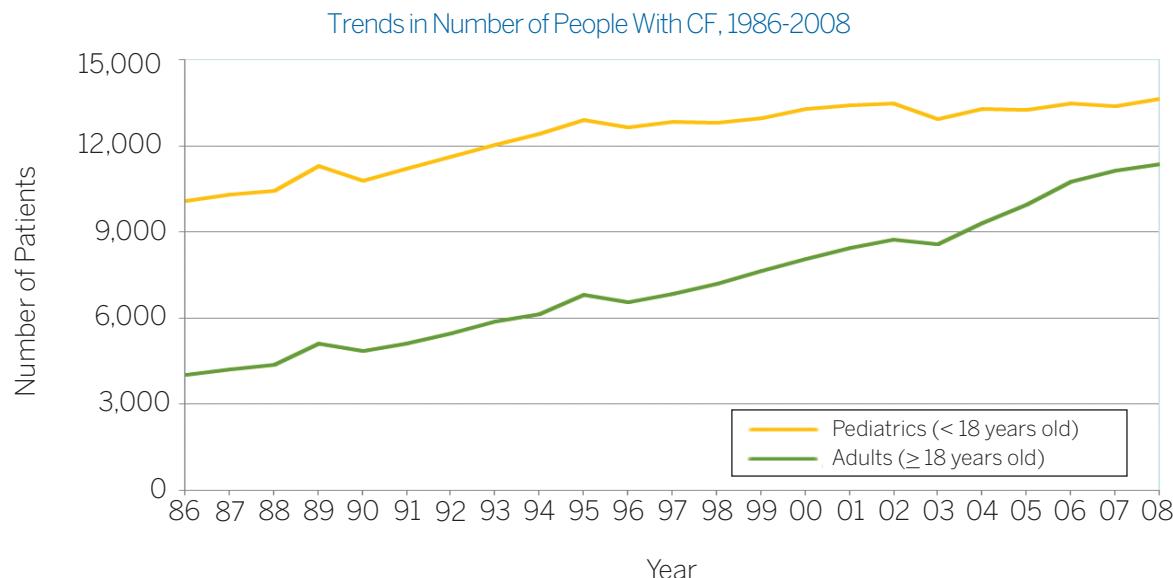
Type of Insurance*	Children <18 Years (%)	Adults ≥ 18 Years (%)
No Insurance	3.1	4.8
Private/HMO	58.6	65.1
Medicaid/State	48.1	33.1
CHAMPUS	2.0	1.5
Federal	1.0	12.6
Other	1.4	2.8

\*Data are not mutually exclusive, as people with CF may have more than one type of insurance.

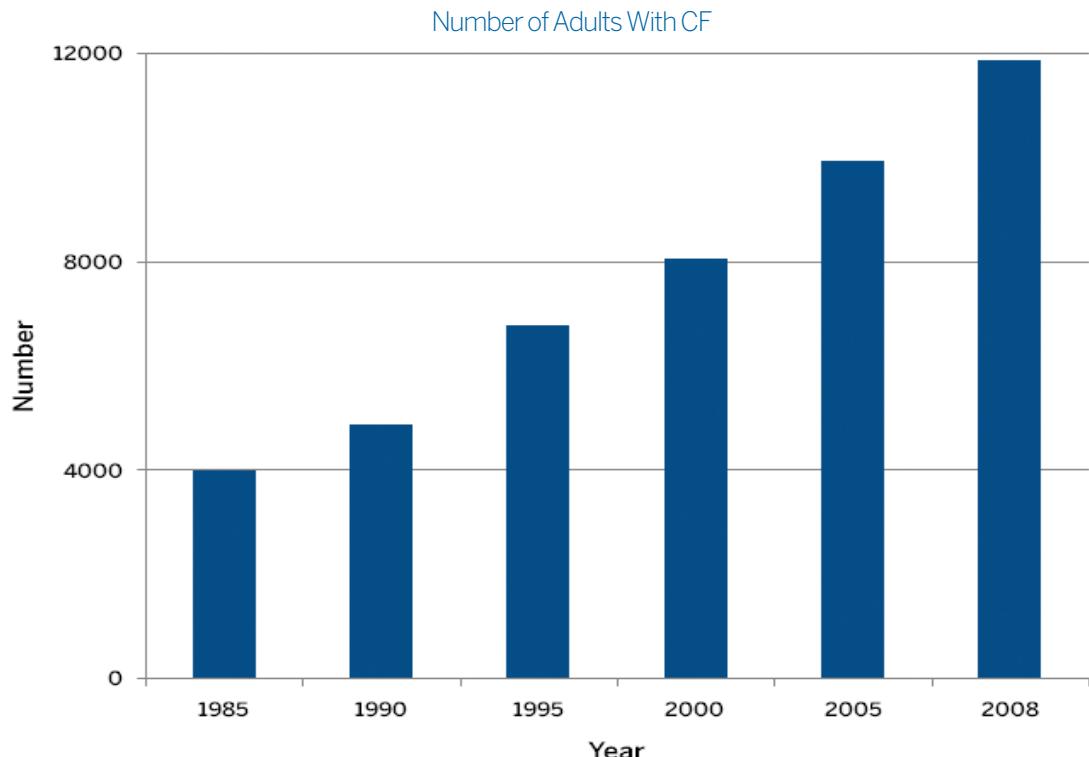
The Foundation has information about patient assistance programs on its Web site at [www.cff.org/LivingWithCF/AssistancePrograms](http://www.cff.org/LivingWithCF/AssistancePrograms). If you or a loved one with cystic fibrosis has trouble paying for your healthcare, the Foundation recommends that you 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 healthcare coverage programs in your state.

## ADULTS WITH CF

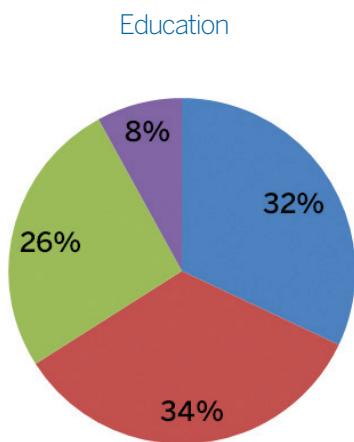
This graph shows how fast the number of adults with CF is growing in relation to the number of children with CF in the Patient Registry. The data indicate that children with CF are healthier and living well into adulthood.



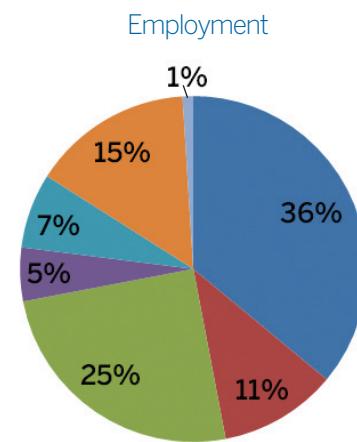
In 1990, about 30 percent of people in the Patient Registry were age 18 or older. In 2008, more than 46 percent of people with CF in the Patient Registry were adults, and that number continues to grow.



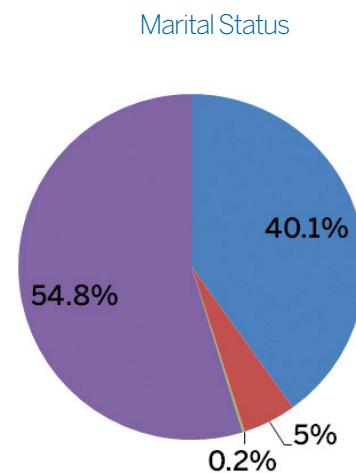
Because of the growing number of adults with CF, the CF Foundation has developed guidelines for the care of adults with CF. The CF Foundation also mandated the development of adult care programs, or clinics for adults with CF, and is providing grants to help more doctors train in the care of adults with CF. Also, it is important to help teens transition from depending on their parents or another adult to taking charge of managing their own health. Your care center can help teach children and teens how to manage CF. The charts below show that many adults with CF are leading active lives.



- College Graduate
- Some College
- High School Diploma
- Less Than High School



- Full-time
- Part-time
- Student
- Homemaker
- Unemployed
- Disabled
- Retired



- Married/Living Together
- Separated/Divorced
- Widowed
- Single

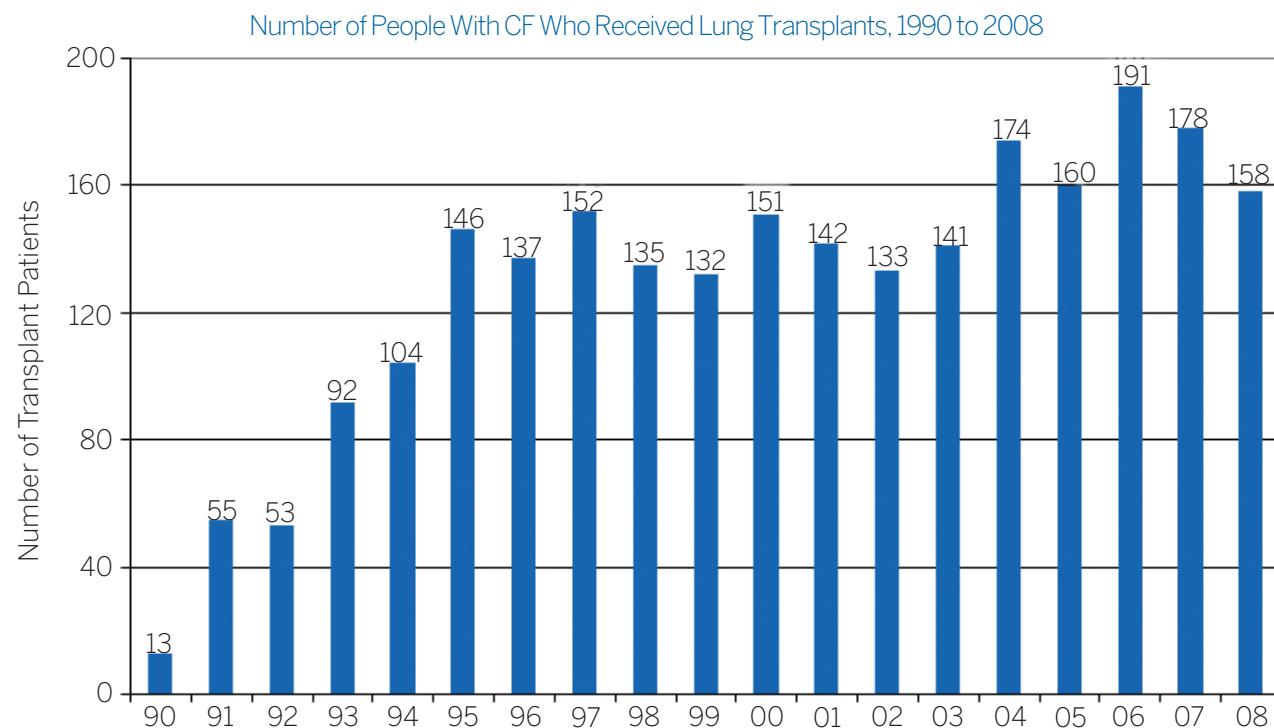
## CF AND PREGNANCY

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. Now, thanks to improvements in nutrition and lung function, many women with CF are able to have a healthy pregnancy and baby. In 2008, the Patient Registry reported that 240 women with CF were pregnant. Also, with advances in fertility medicine, more men with CF are able to father children than ever before. Ask your care center to learn what you should consider before starting a family. You can learn more about CF male and female fertility issues by watching the Web casts at [www.cff.org/LivingWithCF/Webcasts](http://www.cff.org/LivingWithCF/Webcasts).

## TRANSPLANTATION AND END-OF-LIFE CARE

**Goal 7:** Everyone with CF 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. However, a lung transplant is risky and the supply of good donor lungs for transplant is limited. It is important to understand who can benefit from a lung transplant and when the best times for a transplant are. Research with data from the Patient Registry has been used to help identify who is most likely to benefit. The ultimate goal of CF research and care is that no one with CF will need a lung transplant. To learn more about lung transplants and organ donation, visit [www.cff.org/treatments/LungTransplantation](http://www.cff.org/treatments/LungTransplantation).



## WHO ARE THE PATIENTS IN THE CF FOUNDATION'S PATIENT REGISTRY — A SUMMARY OF THE 2008 DATA.

CF patients (number)	25,651	FEV <sub>1</sub> % predicted (mean)	76.1
Newly diagnosed patients in 2008 (number)	1,006	Increased respiratory symptoms or a pulmonary exacerbation (%)	30.4
Patients diagnosed by NBS (%)	42.8	Therapies**	
Age at diagnosis (median)	6 months	TOBI® (tobramycin)	67.4
Age range	0 to 82 years	Pulmozyme® (rhDNase)	76.1
Total number of deaths	420	Ibuprofen	3.8
Predicted median survival	37.4 years	Macrolides (azithromycin)	65.6
Patients 18 years and older (%)	46.3	Hypertonic saline	40.3
Race/Ethnicity (%)		Oxygen	2.5
Caucasian	94.0	IV antibiotics	21.4
Hispanic (black or white)	7.2	Respiratory cultures positive for (%)	
African American	4.3	<i>P. aeruginosa</i>	52.5
Males (%)	51.7	<i>B. cepacia</i> complex	2.8
Genotyped (%)	89.5	<i>S. aureus</i> ***	50.9
Total pregnancies among women aged 13 to 45 (number)	240	MRSA	22.6
Live births (per 100 women age 13 to 45)	1.9	<i>S. maltophilia</i>	12.5
Clinical trial participation (number)	4,515	Complications (%)	
Median BMI percentile for patients 2-20 years* (%)	47.7	Diabetes (CFRD)/glucose intolerance	21.5
Median BMI for patients ≥ 21 years* (%)	21.4	Bone disease	11.1
Pancreatic enzyme supplements (%)	89.8	Liver disease	11.0
		Nasal polyps requiring surgery	3.6
		Distal intestinal obstruction syndrome	3.8
		Transplants (numbers)	
		Lung:	
		Bilateral	157
		Liver:	8

\*The Centers for Disease Control and Prevention have calculators for Body Mass Index (BMI). The national goal for children with CF ages 2-20 years is 50<sup>th</sup> 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).

\*\*This is the percentage of patients who are eligible for a therapy and had it prescribed at least once in 2008.

\*\*\*Does not include MRSA

## NUMBER OF PATIENTS BY STATE IN THE CF PATIENT REGISTRY

<u>State</u>	<u>Number</u>	<u>Percent</u>	<u>State</u>	<u>Number</u>	<u>Percent</u>
Alabama	425	1.66	Nebraska	225	0.88
Alaska	62	0.24	Nevada	169	0.66
Arizona	409	1.59	New Hampshire	176	0.69
Arkansas	214	0.83	New Jersey	657	2.56
California	1,996	7.78	New Mexico	121	0.47
Colorado	528	2.06	New York	1,617	6.30
Connecticut	306	1.19	North Carolina	824	3.21
Delaware	58	0.23	North Dakota	70	0.27
District of Columbia	22	0.09	Ohio	1,438	5.61
Florida	1,275	4.97	Oklahoma	243	0.95
Georgia	781	3.04	Oregon	299	1.17
Hawaii	2	0.01	Pennsylvania	1,404	5.47
Idaho	164	0.64	Puerto Rico	0	0.0
Illinois	963	3.75	Rhode Island	88	0.34
Indiana	581	2.27	South Carolina	350	1.36
Iowa	362	1.41	South Dakota	99	0.39
Kansas	324	1.26	Tennessee	553	2.16
Kentucky	482	1.88	Texas	1,401	5.46
Louisiana	304	1.19	Utah	328	1.28
Maine	217	0.85	Vermont	134	0.52
Maryland	515	2.01	Virgin Islands	0	0.0
Massachusetts	758	2.96	Virginia	679	2.65
Michigan	973	3.79	Washington	558	2.18
Minnesota	563	2.19	West Virginia	217	0.85
Mississippi	210	0.82	Wisconsin	638	2.49
Missouri	661	2.58	Wyoming	44	0.17
Montana	110	0.43	Foreign	54	0.21





SOURCE OF DATA:

Cystic fibrosis patients currently  
under care at CF Foundation-accredited  
care centers in the United States, 2008

SUGGESTED CITATION:

Cystic Fibrosis Foundation Patient Registry  
2008 Annual Data Report  
Bethesda, Maryland  
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| Adding *tomorrows* every day.

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