



Cystic Fibrosis Foundation

Patient Registry

Annual Data Report

2006

Suggested citation:

Cystic Fibrosis Foundation,
Patient Registry 2006 Annual Report,
Bethesda, Maryland.

© 2008 Cystic Fibrosis Foundation

Cystic Fibrosis Foundation

6931 Arlington Road
Bethesda, Maryland 20814

toll free (800) FIGHT CF

local (301) 951-4422

internet www.cff.org

e-mail info@cff.org

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. The *CF Foundation Patient Registry Annual Data Report* (Patient Registry) shows that CF care is improving, and we believe that with your help we can make it even better.


The CF Foundation recognizes that people with CF and their families are full members of the care team. A strong partnership between patients and healthcare providers working as a team is critical to achieve the best possible health for people with CF. Our intention with this report and the care center data from CF Foundation-accredited care centers on our Web site (www.cff.org) is to educate and foster strong partnerships between people with CF, their families and care center staff through open communication.

We encourage people with CF and their families to get involved with their care center. We encourage you to use this report and the center data to start a conversation with your care center staff. Across the country, CF care centers are focusing on quality improvement to improve 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. To learn more, read how other people with CF and their families worked with their care center to improve care on the CF Foundation's Web site (www.cff.org/LivingWithCF/QualityImprovement/).

The CF Foundation, in collaboration with the Institute for Family Centered Care, is supporting activities at CF care centers to promote patient- and family-centered care. 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 also continue to support and expand educational forums on patient- and family-centered care, including presentations at national meetings and Web cast programs.

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,



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



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

What Is the Cystic Fibrosis Foundation Patient Registry?

More than 40 years ago, the Cystic Fibrosis Foundation started a Patient Registry to track the health of people with CF across the United States. Today, information about more than 24,000 people who receive care at CF Foundation-accredited care centers is collected and added to the Patient Registry every year. The type of information collected 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.

Information in the Patient Registry helps caregivers and researchers see new trends, design clinical trials to test new therapies and improve the delivery of care for people with CF. To get the best information, it is important for people with CF to participate in the Patient Registry. The following pages contain information from the Patient Registry relating to the CF Foundation's seven worthy goals to improve CF care. These goals are:

- 1) People with CF and their families are full members of the care team;
- 2) People with CF will achieve normal growth and maintain normal nutrition;
- 3) People with CF will receive respiratory therapies that keep lung function steady and to diagnose infections early;
- 4) To decrease the spread of germs between people with CF;
- 5) To prevent complications and/or to diagnose and treat them early;
- 6) To provide care regardless of race, age, education or insurance coverage; and
- 7) To support all transplantation and end-of-life care decisions.

If You Are New to Cystic Fibrosis

What Is Cystic Fibrosis?

CF is a genetic disease caused by an altered gene. It results in the faulty transport of salt in organs such as the lungs and the pancreas. This leads to thick, sticky mucus that blocks the ducts in these organs, disrupting their normal functions. Many people with CF have a cycle of lung infection or exacerbation and inflammation (swelling). This cycle slowly damages the lungs and their ability to provide oxygen to the body. When the pancreas is affected, it causes problems with digestion and makes it difficult to grow normally and keep a healthy body weight.

Approximately one in 3,500 children in the United States each year is born with CF. It is found in 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 created in 1955 by a dedicated 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.

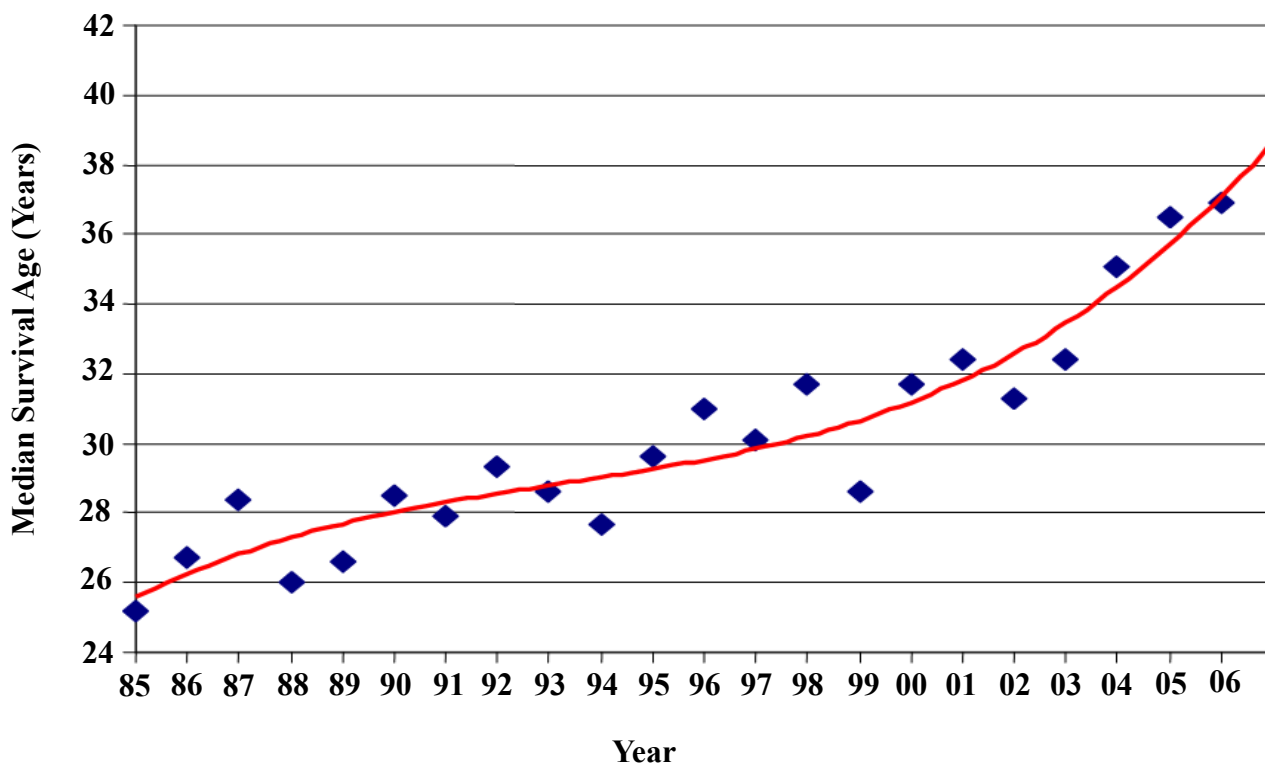
To continue this mission, the CF Foundation has a network of more than 115 accredited care centers across the United States for people with CF. The CF Foundation provides grants, training in quality improvement, and the latest CF care guidelines based on the medical literature to support the care centers as they care for people with CF. The CF Foundation also provides grants to researchers working to discover and develop new drugs and new therapies to improve the length and quality of life for those with the disease. To learn more about CF and the CF Foundation, visit www.cff.org.

Survival Is Improving

Because of the hard work and strong partnership between people with CF, their families and CF Foundation-accredited care centers, the predicted survival for people with CF has steadily improved. When the CF Foundation was created in 1955, few children with CF lived to go to school. Today, the predicted survival extends into the late 30s. The graph below shows how much the median predicted survival has improved since 1985.

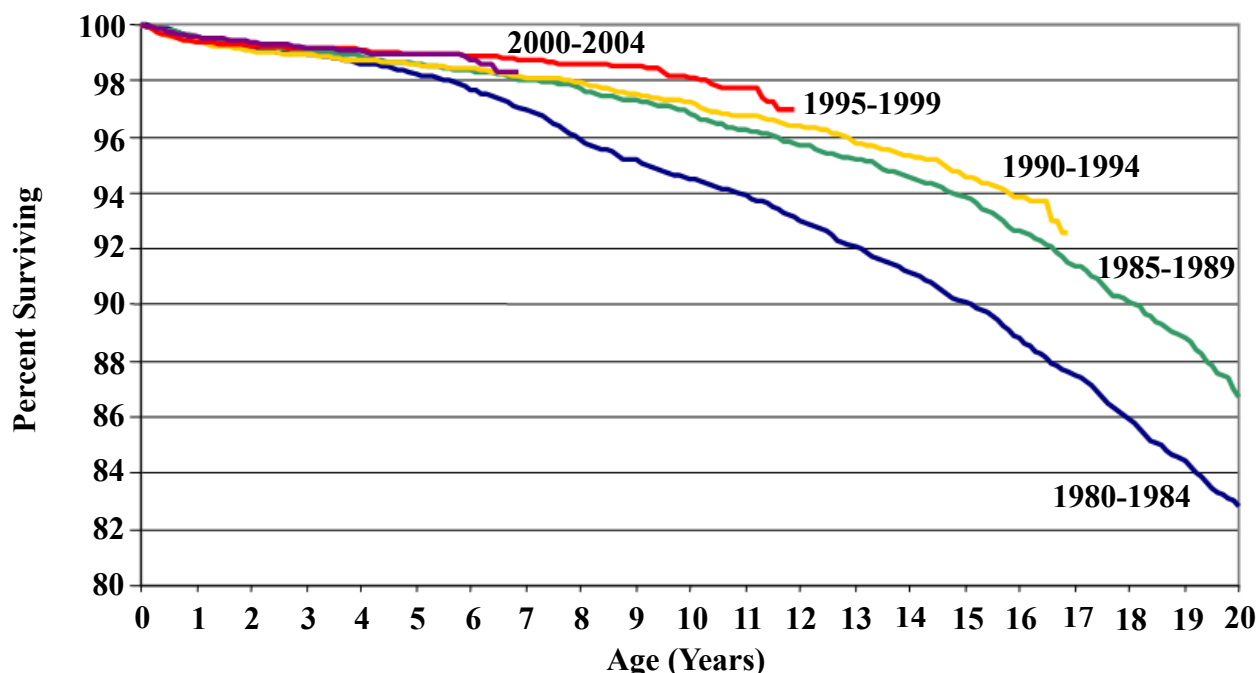
Median age of survival represents the age to which half of the current people with CF in the Patient Registry would be expected to survive. This number is calculated every year and is based on the deaths that occurred in that year. In 2006, the predicted survival reached 36.9 years. This continued improvement in survival depends, in part, on gathering and using data from people with CF across the United States through the Patient Registry. The CF Foundation-accredited care centers and the CF Foundation continue to partner with people with CF and their families to keep the line moving up.

Median Predicted Survival Age, 1985-2006



The graph at the top of the next page shows that survival continues to improve from 1 year of age and grouped by year of birth. Of people with CF born between 1980 and 1984 (dark blue line), 90.2 percent were alive at age 15. For children born between 1990 and 1994 (yellow line), 95.2 percent were alive at age 15. The CF Foundation is working with all 50 states in an effort to have all newborn babies screened for CF. The earlier CF is diagnosed, the sooner treatment can be started to slow the disease and prevent complications. Earlier diagnosis and treatment helps improve quality and length of life for those with CF.

Survival from Age One by Birth Cohort



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 patients' needs, preferences and values.

Through the Patient Registry, care centers get combined data about their patients. These reports help care centers communicate with their patients and families about some of the important aspects of the disease. They are available from your CF Foundation-accredited care center. The next page is an example of a "Patient Summary Report." It shows trends in lung function, nutrition and other important information about the patient. Ask your CF care center for a copy of your or your child's "Patient Summary Report" at your next CF clinic visit.

In December 2006, the CF Foundation made key health data for each of the accredited CF care centers available on its Web site (www.cff.org). The data are part of the effort to open communications and encourage people with CF and their families to get involved and improve CF care. This data is updated at the end of every year. We encourage you to start a conversation with your care center about your center's data. The following questions are good 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 become a full and active member of your CF care team.

To learn more about this 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" on the CF Foundation's Web site. You can also read the success stories of how others work with their care centers to improve care in the Quality Improvement section of the CF Foundation's Web site.

Cystic Fibrosis Foundation Patient Registry: Annual Data Report 2006

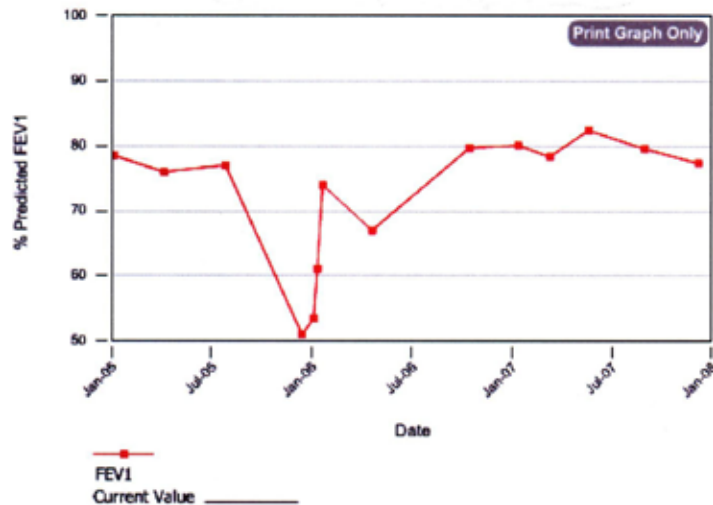
Patient Report Example:

VISIT DATE: _____ Name: _____
 Last Hospitalization: 1/18/ - 1/23/ Date of Birth: _____
 Last Home/IV Therapy: 1/23/ - 2/3/ Genotype: Date Unknown ^F508 /
 Last Clinical Visit: 12/18/ ^F508

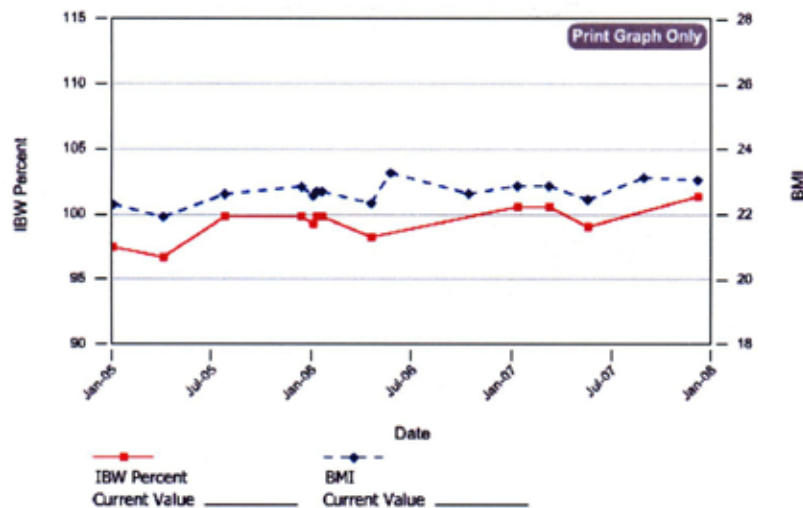
Culture Results

Last Culture: *Pseudomonas aeruginosa*, *Staphylococcus aureus*, : 9/11/
 Last Positive: **B. cepacia** **MRSA** **PA** **MDR-PA+**
 None 10/26/ 9/11/ 9/11/

PFT Trend



Nutritional Trend



Complications

ACTIVE @ LAST VISIT

- CFRD
- Dist Int Obst Synd (DIOS)
- 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: 12/18/
 Last CXR: 2006
 Last SW Visit: 4/6/

Last Dietary Visit: 6/6/
 Last LFT: 9/11/
 Glucose Screening: 10/19/
 Creatinine: 9/11/

Guidelines for CF Care

The CF Foundation gathers experts to review the medical literature and research on CF care to develop guidelines for the care of all people with CF. Below are the percentages of patients in the Patient Registry who met some of these recommendations in 2006.

Guidelines for CF Care	Children Who Meet Guidelines (%)	Adults Who Meet Guidelines (%)
Clinic Visits — 4 or More Per Year	66.8	52.1
Pulmonary Function Tests — (PFT) 2 or More Per Year	92.0	83.7
Respiratory Cultures — At Least 1 Per Year	97.3	90.3
Glucose — Every Year if ≥ 14 Years	77.7	70.1
Liver Enzymes — Every Year	79.0	74.4
Influenza (flu) Vaccine — Every Year ≥ 6 Months	67.5	58.1

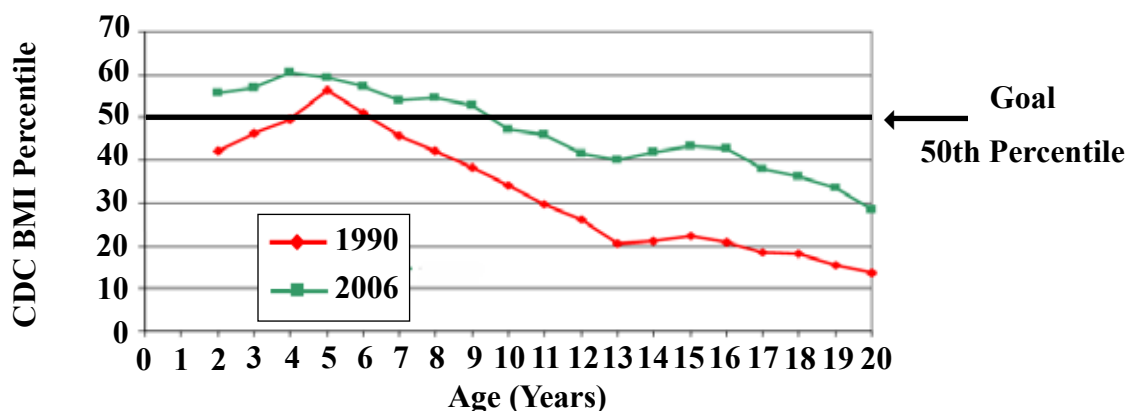
Maintaining Normal Nutrition

Goal 2: Children and teens will have normal growth and good nutrition. Adults' nutrition will be maintained as near to "normal" as possible.

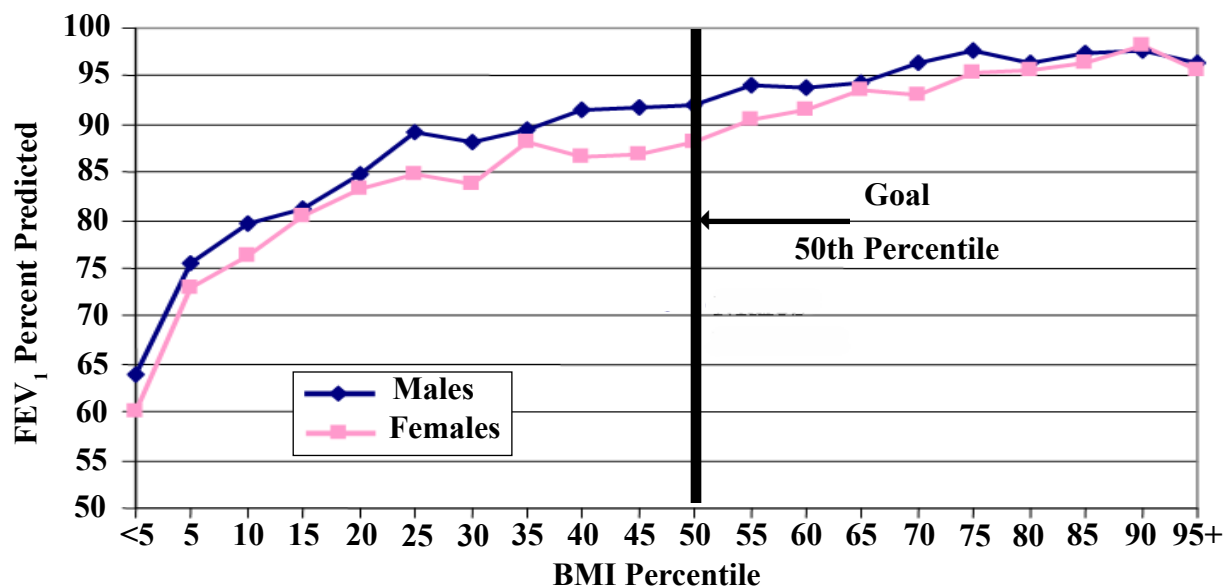
Steady progress toward this goal has been made, but more work remains. In 2005, the CF Foundation worked closely with experts in CF and nutrition to review the medical literature and data from the Patient Registry to set national goals for children and adults with CF as measured by body mass index (BMI). BMI is based on a person's weight and height. It is used to screen for people who may have health problems if that number is too high or too low. For children and teens, BMI is stated as a percentile compared to healthy children 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 m²=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/nccdphp/dnpa/bmi/index.htm).

The goal is for children to be at or above the 50th BMI percentile for their age. The graph on the top of the next page shows how much the BMI percentile of children with CF has improved since 1990. Children with CF who have good nutrition will most likely grow normally.

Median CDC BMI Percentiles vs. Age, 1990 and 2006*

The Patient Registry shows a strong association between a higher BMI percentile and better lung function in children. Lung function is measured by FEV₁, or Forced Expiratory Volume over one second, which 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 seems to go hand in hand. The highlighted bar is the goal for children with CF. The 50th percentile is the average BMI percentile for children in the United States who do not have CF. We want children with CF to grow and develop like children without CF.

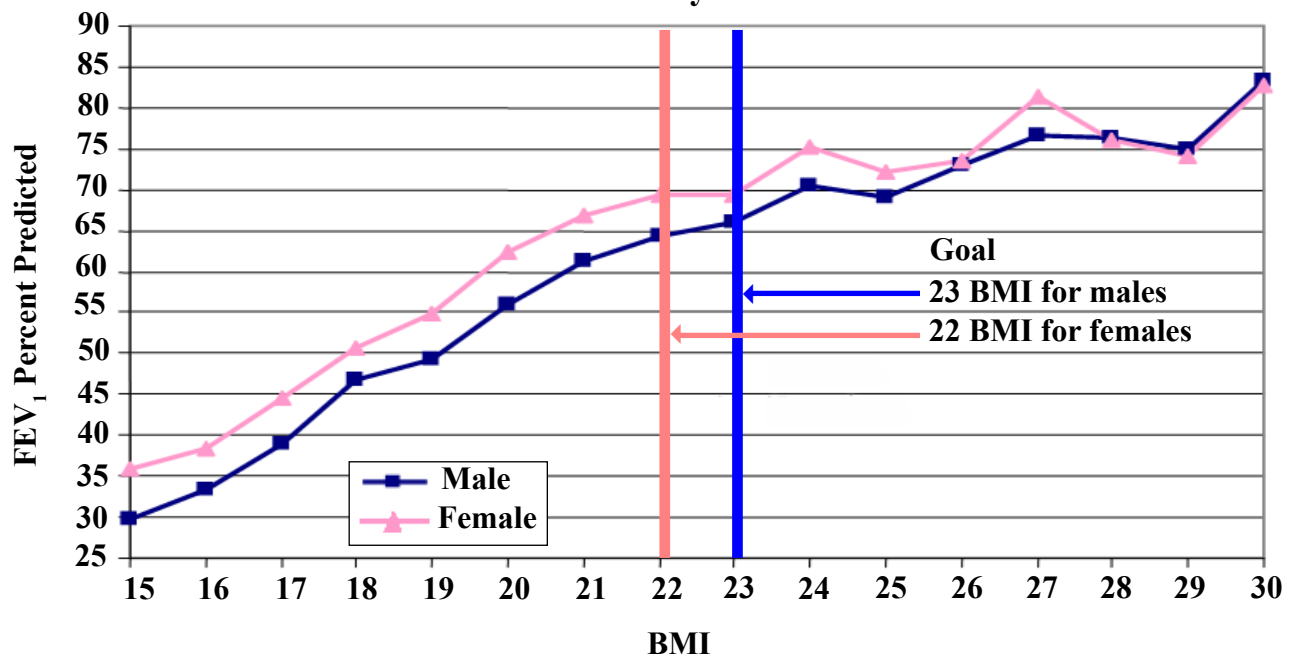
FEV₁ Percent Predicted vs. BMI Percentile in Children 2-20 Years*

Work close with your CF care center to improve your or your child's nutrition and help keep lungs healthy. To learn more about CF and nutrition, ask your CF care center or visit the CF Foundation's Web site (www.cff.org) to watch the archived Web cast about nutrition.

The association 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. Men should partner with their care center and work to maintain or reach for a BMI of 23. Women should also work with their care center to maintain or reach for a BMI of 22.

* Centers for Disease Control and Prevention — www.cdc.gov/nccdphp/dnpa/bmi/index.htm

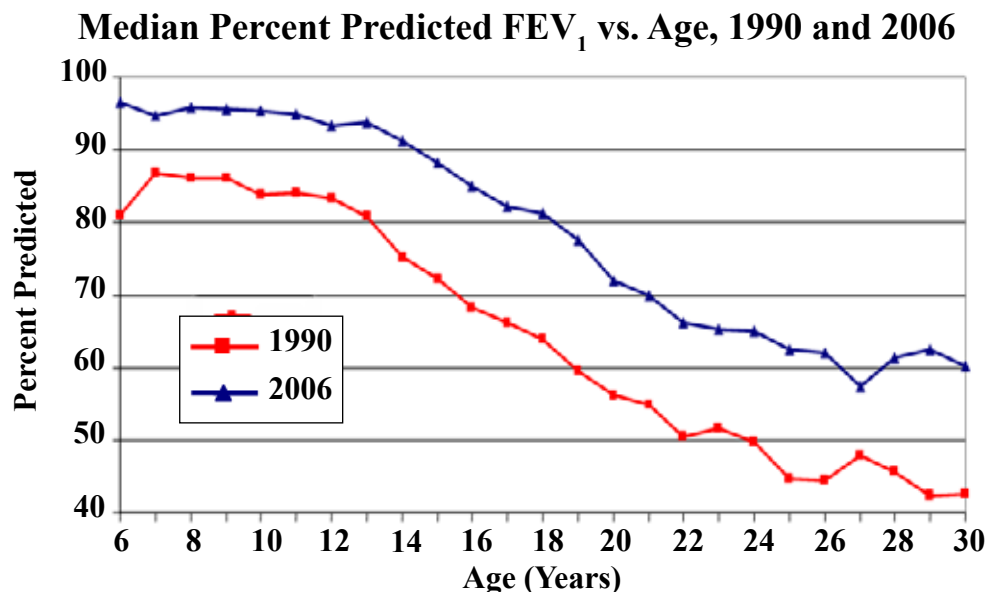
FEV₁ Percent Predicted vs. BMI in CF Patients 20 to 40 Years by Gender*



Lung Function

Goal 3: Everyone with CF will receive the right therapies to keep lung function steady and to decrease the number of pulmonary exacerbations or respiratory infections. Exacerbations will be diagnosed early and treated appropriately.

The graph below shows that since 1990 there have been major improvements in the lung health of people with CF. FEV₁ (lung function) is usually near normal or just under 100 percent predicted when first measured around 6 years of age.

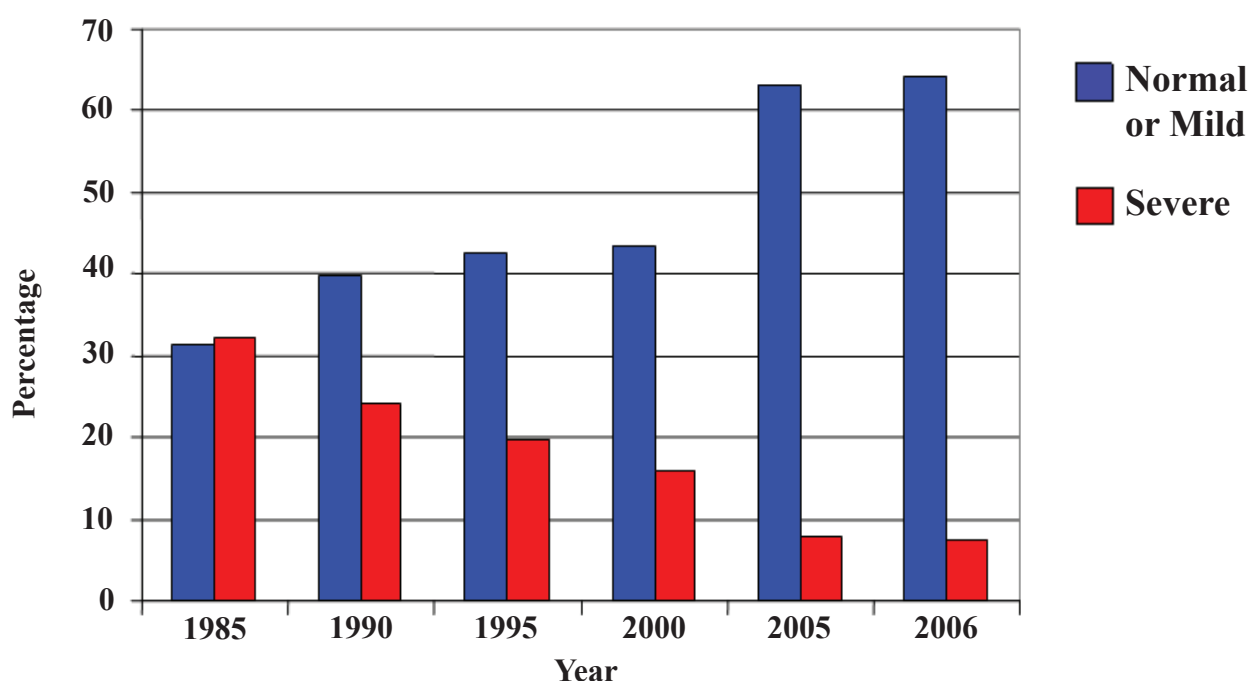


* Centers for Disease Control and Prevention — www.cdc.gov/nccdphp/dnpa/bmi/index.htm

Severity of lung disease is based on a person's FEV₁ percent predicted. In CF, the lower a person's FEV₁ the more severe the lung disease. An FEV₁ greater than or equal to 90 percent is normal. If the FEV₁ is between 70 and 89 percent, the person has mild lung disease. An FEV₁ between 40 and 69 percent means the person has moderate lung disease. If the FEV₁ is less than 40 percent, this means severe lung disease is present.

The graph below shows how the percentage of 18 year olds with CF in the Patient Registry with normal lung function or mild lung disease has increased as compared to those with severe lung disease. The rising number of people with normal lung function or mild disease and the dropping number with severe disease show that the lungs of people with CF are much healthier now than 20 years ago.

Lung Function of 18-Year-Olds With CF



In 2006, the CF Foundation brought experts in CF lung disease together to review data and recommend medications to maintain lung health in people with CF. These therapies are given to patients who meet certain criteria (e.g., age, severity of disease). The “Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Health” are in the November 15, 2007 edition of the *American Journal of Respiratory and Critical Care Medicine*.

The table on the next page shows the percentage of people with CF who meet the criteria and have the medication prescribed. It shows that hypertonic saline has been prescribed to 23.2 percent of people with CF 6 years of age or older.

In 2005, CF Foundation-supported research on hypertonic saline that showed it improved lung health and decreased the number of pulmonary exacerbations or respiratory infections in people with CF. Talk to your CF doctor to find out if you or your child might do well on one of these medications. Data from the Patient Registry will continue to help find and report the use of therapies that help people with CF.

Cystic Fibrosis Foundation Patient Registry: Annual Data Report 2006

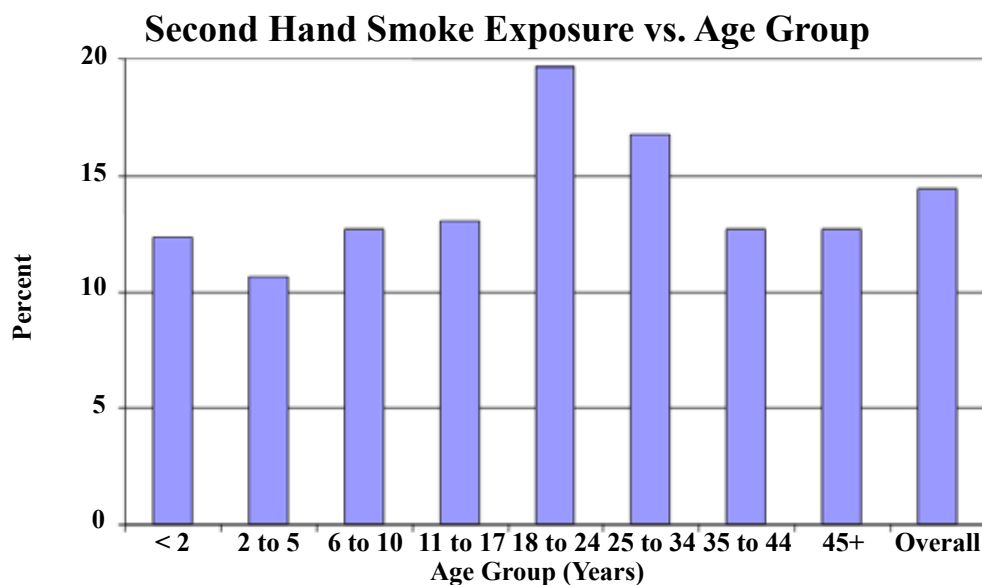
Along with these therapies, better nutrition, airway clearance and exercise also helps keep lungs healthy. All of these work to improve FEV₁ and lung health in people with CF.

Medication	Percentage (%) of people who fit the criteria and are on the medication	Criteria for medication
tobramycin (TOBI®)	61.8	<ul style="list-style-type: none"> • ≥ 6 years of age • Positive for <i>P. aeruginosa</i> • Moderate to severe lung disease
rhDNase (Pulmozyme®)	67.4	<ul style="list-style-type: none"> • ≥ 6 years of age
azithromycin (Zithromax®)	57.3	<ul style="list-style-type: none"> • ≥ 6 years of age • Positive for <i>P. aeruginosa</i> • Moderate to severe lung disease • Weight ≥ 55 pounds (25kg)
ibuprofen	4.5	<ul style="list-style-type: none"> • 6-12 years of age • Moderate to severe lung disease
hypertonic saline	23.2	<ul style="list-style-type: none"> • ≥ 6 years of age

To learn more about what you or your child can do to keep the lungs healthy, watch the Web cast on CF lung disease on the CF Foundation's Web site (www.cff.org).

Even if you are careful and do all you can to prevent a lung infection, it is hard to avoid them totally. If you or your child start to feel ill (e.g., increased cough, fever, loss of energy or appetite), call your care center so treatment can be started as soon as possible.

Secondhand smoke comes from burning tobacco in cigarettes, cigars, pipes and the smoke breathed out of the lungs of people who smoke. It is known 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. For someone with CF, it can cause even more problems. The graph below shows, by age group, the percentage of people with CF who breathe in secondhand smoke.



The only way to fully protect yourself and your child from secondhand smoke is through 100 percent smoke-free environments.

You can:

- Make your home and car smoke-free.
- Ask people not to smoke around you or your child.
- Make sure that your child's day care center or school is smoke-free.
- 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.
- Teach your child to stay away from secondhand smoke.

If you are a smoker, the single best way to protect your family from secondhand smoke is to quit smoking. In the meantime, you can protect your family by making your home and vehicles smoke-free and only smoking outside. A smoke-free home rule can also help you quit smoking. Your care center can also help you quit smoking and keep your loved ones with CF healthy.

For more information, please refer to "How to Protect Yourself and Your Loved Ones from Secondhand Smoke" available at www.cdc.gov/tobacco.

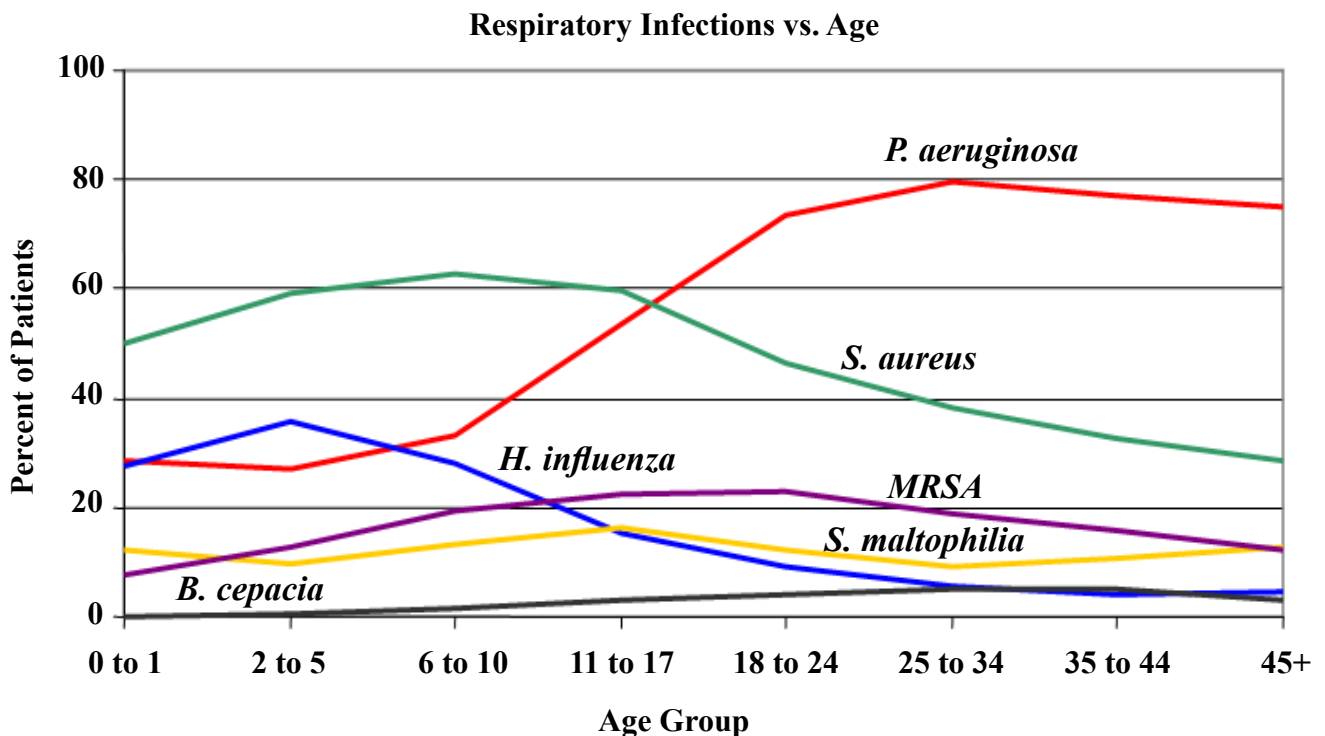
Each time a person has a respiratory infection or exacerbation, there may be lung damage. However, there are things you can do to prevent or lessen the chance of a lung infection, such as:

- Getting a flu shot every fall for you/your child and everyone living in the house;
- Regular exercise to strengthen your muscles;
- Doing airway clearance to keep the lungs as clear as possible of mucus;
- Taking the medicines as prescribed by your CF care center;
- Avoiding germs by using good hand hygiene and properly cleaning and disinfecting equipment; and
- Avoiding secondhand smoke exposure.

To learn more about lung care and therapies for people with CF, visit the CF Foundation's Web site (www.cff.org). You can watch an archived Web cast and read about ways to stay healthy. It is important that you and your CF care center partner and have a plan to maintain your or your child's health.

Goal 4: People with CF and their care centers will work together to eliminate the chances of patients getting respiratory pathogens or germs, particularly *Pseudomonas aeruginosa* (*P. aeruginosa*) and *Burkholderia cepacia* (*B. cepacia*) complex, in the hospital, clinic and home.

Repeated respiratory infections or exacerbations are a concern for people with CF. It is the cycle of infection and inflammation (swelling) that may damage the lungs. This damage causes lung function (FEV₁) to decline. When the lungs are damaged, exacerbations happen more often. The next graph shows the germs that are found in the lungs of people with CF. Talk to your CF care center to learn more about how to prevent respiratory infections. Information about what CF germs are, how they are spread and how to avoid germs is available on the CF Foundation's Web site (www.cff.org).



Overall Percentage in 2006:

— <i>P. aeruginosa</i> 55.0%	— <i>H. influenza</i> 16.9%	— <i>B. cepacia</i> complex 2.9%
— <i>S. aureus</i> 51.5%	— <i>S. maltophilia</i> 12.6%	— MRSA 18.9%

Complications of CF

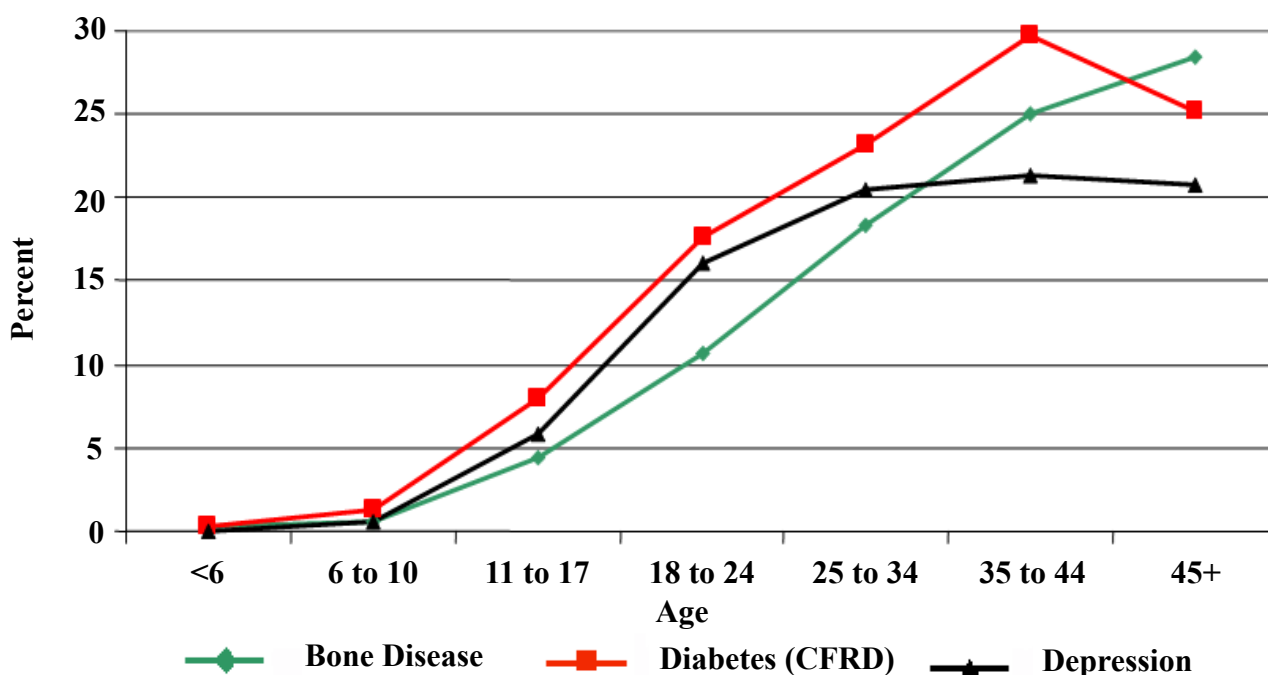
Goal 5: People with CF will be closely monitored for complications of CF, especially CF-related diabetes (CFRD). Prevention and early treatment of any CF complication is the goal. In the early 1990s, data in the Patient Registry showed an increase in the number of teens and adults with diabetes. CFRD was found to be different from diabetes in people without CF because of how cystic fibrosis damages the pancreas. The CF Foundation brought together experts in CF and diabetes and developed guidelines for the care of CFRD. Anyone with CF, 14 years of age or older, should be tested every year for CFRD. Data suggest that diagnosing and treating CFRD earlier results in better outcomes. The CF Foundation continues to fund CFRD research. To learn about CF Foundation-supported research, visit www.cff.org.

Cystic Fibrosis Foundation Patient Registry: Annual Data Report 2006

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 8.5 percent of people with CF had bone disease in 2006. Preventing or lessening bone disease begins in childhood when bones are growing. Ask your CF dietitian what can be done to keep your or your child's bones healthy.

Another finding from the Patient Registry is that almost 19 percent of adults with CF have symptoms of depression. This is a common complication of many chronic diseases. 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. The CF Foundation continues to promote prevention, early diagnosis and treatment of complications that people with CF may experience.

Common Complications vs. Age



Access to Care

Goal 6: Everyone with CF will be able to receive 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 poorer lung function and worse BMI or BMI percentile. This pattern of poor health in lower-income households is common with other chronic diseases. The CF Foundation is working to find out why this happens in CF and how to change it. CF Services Pharmacy (www.cfservicespharmacy.com), a mail-order pharmacy and wholly owned subsidiary of the CF Foundation, works hard to keep proven CF therapies available to everyone. For example, data from the Patient Registry show that people with CF, no matter their income, have therapies like Pulmozyme® and TOBI® available to them. You can learn more about your legal rights and hear how others manage CF by watching the archived Web casts “Patient Advocacy: Issues and Answers” and “Building Life Skills to Manage CF,” available on the CF Foundation’s Web site (www.cff.org).

Learn more about race and age of the people in the Patient Registry on page 16, “A Summary of the 2006 Data.”

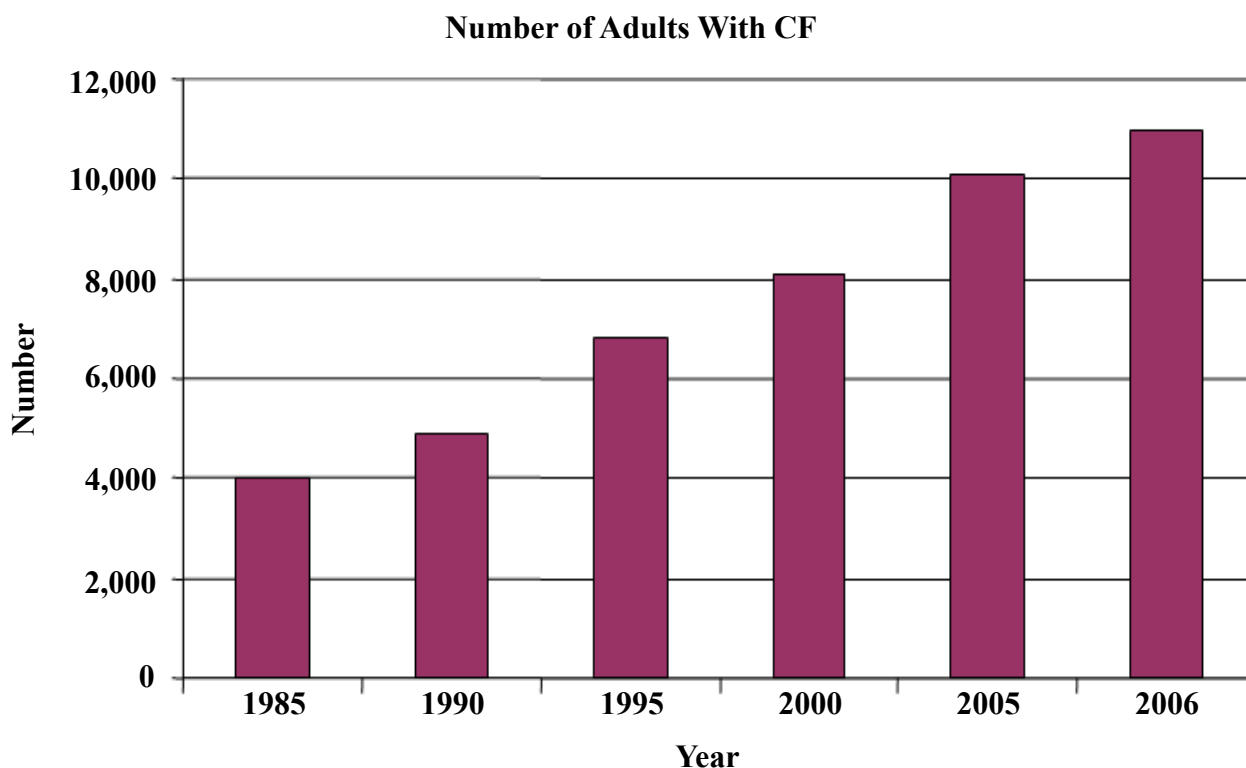
Cystic Fibrosis Foundation Patient Registry: Annual Data Report 2006

Insurance*	Children (%)	Adults (%)
No Insurance	2.4	5.4
Private/HMO	61.1	64.8
Medicaid/State	45.9	30.8
CHAMPUS	1.6	1.2
Federal	1.3	11.8

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

Adults With CF

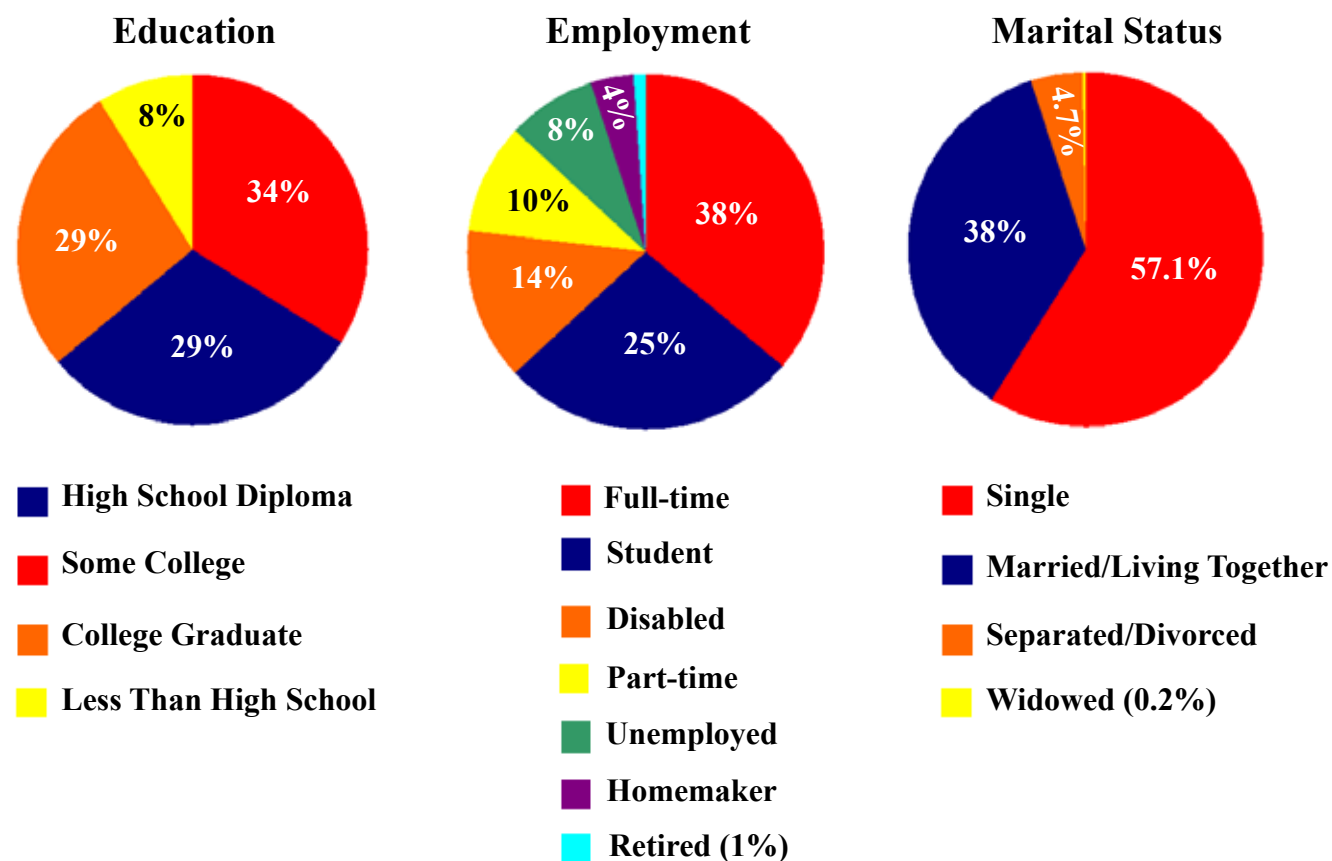
In 1990, about 30 percent of people in the Patient Registry were age 18 or older. In 2006, almost 45 percent of people with CF in the Patient Registry were adults, and that number continues to grow.



Because of the growing number of adults with CF, the CF Foundation brought together experts in CF and adult medicine and 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. It is important to help teens transition from depending on their parents or another adult to taking charge or managing their own health. Your care center can help teach teens how to manage CF.

Cystic Fibrosis Foundation Patient Registry: Annual Data Report 2006

Data from the Patient Registry show some facts about adults with CF. As you can see, they lead busy and productive lives while dealing with CF. It is important for adults with CF to partner with their care centers and find the best way to deal with the healthcare demands of the disease, while pursuing their life goals.



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 2006, the Patient Registry reported that 209 women with CF were pregnant. Ask your care center and visit the CF Foundation's Web site (www.cff.org) to learn what to consider before starting a family.

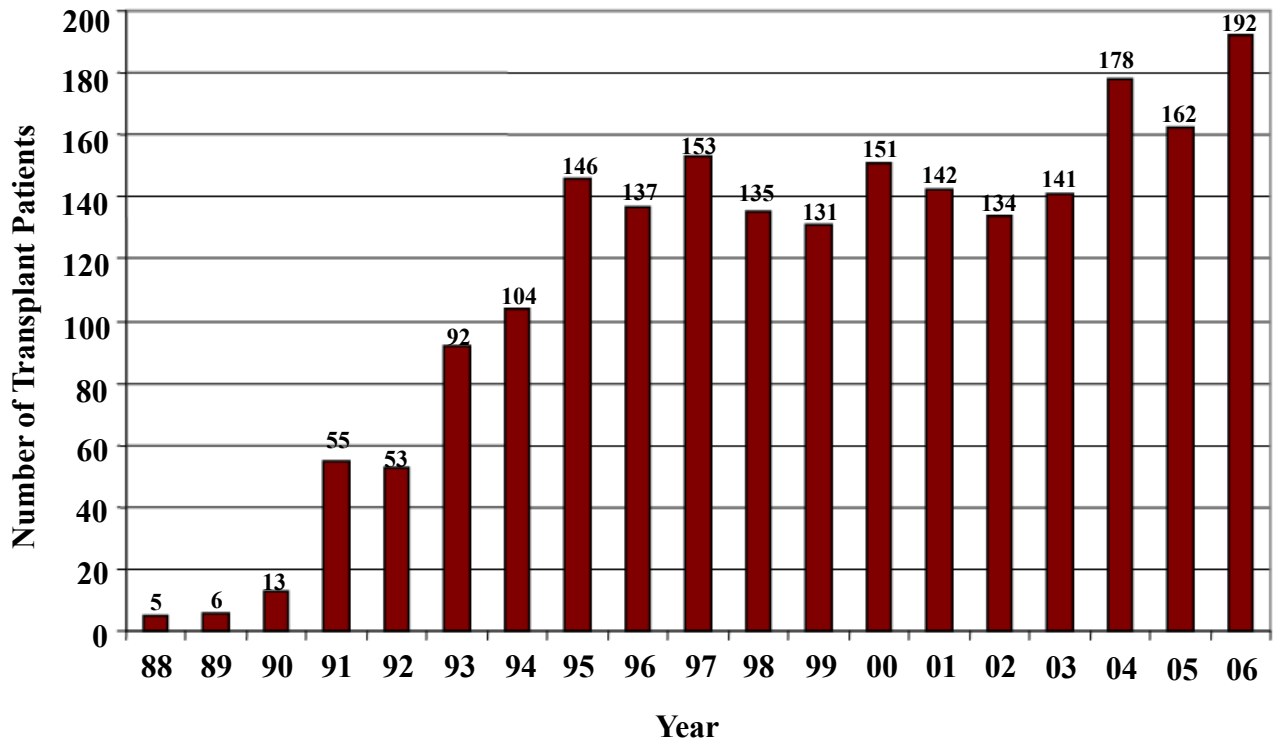
Also, with advances in fertility medicine, more men with CF are able to father children than ever before.

Transplantation and End-of-Life Care

Goal 7: Everyone with CF will be supported by their CF care center when making decisions about transplantation and end-of-life care.

People with CF who have severe lung disease often think about having a lung transplant. Unfortunately, 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 this procedure and when is the best time to pursue it. Research with data from the Patient Registry has been used to help identify who is most likely to benefit from a transplant. The national formula used to decide who is listed first to get donor lungs changed in 2005. Data from the CF Foundation Patient Registry shows more people with CF received lung transplants in 2006 than previous years. Of course, the ultimate goal of research and care is that no one with CF will need a lung transplant. To learn more about lung transplants and organ donation, visit the CF Foundation's Web site at www.cff.org.

Number of People With CF Who Received Lung Transplants, 1988 to 2006



Who Are the Patients in the CF Foundation's Patient Registry — A Summary of the 2006 Data.

CF patients (number)	24,487	Median BMI percentile for patients 2-20 years* (%)	47.1
Newly diagnosed patients in 2006 (number)	874	Median BMI for patients ≥ 21 years* (%)	21.2
Patients diagnosed by NBS (%)	21.6	Respiratory cultures positive for (%)	
Age at diagnosis (median)	6 months	<i>P. aeruginosa</i>	55.0
Age range	0 to 78 years	<i>B. cepacia</i> complex	2.9
Total number of deaths	362	<i>S. aureus</i>	51.5
Predicted median survival	36.9 years	<i>S. maltophilia</i>	12.6
Patients 18 years and older (%)	44.6	MRSA	18.9
Race/Ethnicity (%)		Complications (%)	
Caucasian	94.6	Diabetes (CFRD)/glucose intolerance	19.5
Hispanic (black or white)	6.8	Bone disease (patients ≥ 18 years)	16.7
African American	3.9	Liver disease	10.7
Males (%)	52.0	Nasal polyps requiring surgery	3.7
Genotyped (%)	85.6	Transplants (numbers)	
Home therapy (%)		Lung:	
IV antibiotics	20.9	Bilateral	187
Oxygen	5.2	Lobar-cadaveric	4
Supplemental feeding – tube	10.2	Lobar-living related donor	2
oral only	38.0	Liver:	11
Taking pancreatic enzyme supplements (%)	90.7	Therapies**	
Clinical trial participation (number)	3,037	TOBI® (tobramycin)	61.8
FEV ₁ % predicted (mean)	75.4	Pulmozyme® (rhDNase)	67.8
		Ibuprofen	4.5
		Zithromax® (azithromycin)	57.3
		Hypertonic Saline	23.2
		Total pregnancies among women aged 13 to 45 (number)	209
		Live births (per 100 women age 13 to 45)	1.6

*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 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/nccdp/hnpa/bmi/index.htm.

**This is the percentage of patients who are eligible for a therapy and had it prescribed at least once in 2006.

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	373	1.6	Nebraska	208	0.9
Alaska	53	0.2	Nevada	132	0.6
Arizona	343	1.4	New Hampshire	170	0.7
Arkansas	219	0.9	New Jersey	586	2.5
California	1,915	8.1	New Mexico	105	0.4
Colorado	465	2.0	New York	1,561	6.6
Connecticut	300	1.3	North Carolina	747	3.1
Delaware	45	0.2	North Dakota	65	0.3
District of Columbia	18	0.1	Ohio	1,363	5.7
Florida	1,092	4.6	Oklahoma	236	1.0
Georgia	700	2.9	Oregon	278	1.2
Hawaii	14	0.1	Pennsylvania	1,291	5.4
Idaho	140	0.6	Puerto Rico	14	0.1
Illinois	901	3.8	Rhode Island	102	0.4
Indiana	539	2.3	South Carolina	321	1.3
Iowa	299	1.3	South Dakota	92	0.4
Kansas	263	1.1	Tennessee	481	2.0
Kentucky	443	1.9	Texas	1,370	5.8
Louisiana	254	1.1	Utah	278	1.2
Maine	192	0.8	Vermont	118	0.5
Maryland	458	1.9	Virgin Islands	1	0.0
Massachusetts	775	3.3	Virginia	684	2.9
Michigan	856	3.6	Washington	535	2.3
Minnesota	515	2.2	West Virginia	177	0.7
Mississippi	215	0.9	Wisconsin	597	2.5
Missouri	613	2.6	Wyoming	44	0.2
Montana	101	0.4	Foreign	31	0.1

