Patient Registry

Annual Data Report 2009

Cystic Fibrosis Foundation

| Adding *tomorrows* every day.

Cystic Fibrosis Foundation | Adding *tomorrows* every day.

To the CF Community and Friends,

We are proud to report that, based on the most recent data in the CF Foundation's Patient Registry, the outlook for people with cystic fibrosis has continued to improve.

Each year, the Cystic Fibrosis Foundation collects information on the health status of the more than 26,000 people with CF receiving care at Foundation-accredited care centers nationwide.

The CF Foundation's Patient Registry Annual Data Report shares this information with the wider community, highlighting progress, trends and areas for improvement in the treatment of cystic fibrosis.

Information collected in 2009 and contained in this report shows promise in many areas, including:

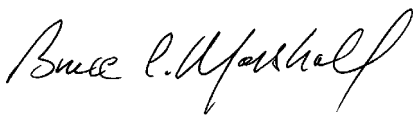
- Better overall lung function for both adults and children
- Increased body mass index (BMI) for adults and BMI percentile for children
- Growing numbers of adults with CF 18 years of age and over
- Greater numbers of infants diagnosed at birth with CF, thanks to newborn screening nationwide

These gains are the result of the strong partnerships among those with CF, families and health care professionals. We applaud the contributions of the many people with CF and families who have become actively involved by serving as members of advisory boards and quality improvement teams or in other critical roles at their care centers.

These strong partnerships will remain crucial to achieving the best possible health outcomes for people with CF. We encourage you and your CF care center to use this report—and other resources available on www.cff.org—as a tool for discussion about ways to improve the quality of care at your care center.

We hope you will learn more from this report about what you can do to join us and add tomorrows every day to the lives of those with CF.

Sincerely,



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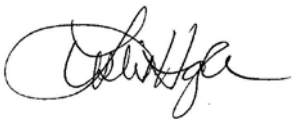

Leslie Hazle, M.S., R.N.
Director of Patient Resources
Cystic Fibrosis Foundation

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WHAT IS THE CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY?

The Cystic Fibrosis Foundation's Patient Registry tracks the 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 includes state of residence, height, weight, gender, genotype, pulmonary function test (PFT) results, pancreatic enzyme 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 how treatments help people with CF;
- develop care guidelines;
- design clinical trials to test new therapies; and
- improve the delivery of care.

It is vital for everyone with CF to agree to have their data entered into the Registry. This data helps to create a more accurate picture of the current state of CF care and to point out 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 seven worthy goals of CF care:

1) People with CF, their families and CF health care professionals make up the CF 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 what work is being done to meet these goals. We hope it will provide you and all members of the CF care team with ideas on ways to improve care even further.

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.

Many people with CF have lung infections and inflammation, which over time lead to lung damage and decreased lung function. CF also makes it hard for the body to absorb food, making 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 and 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. Their mission was clear: to save their children. The Foundation expanded upon their desire 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 has a network of more than 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 and CF care guidelines based on research from medical journals and the Patient Registry.

The CF Foundation also provides support to researchers working to learn more about CF and to discover and develop new therapies 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 development. While the Foundation is focused on finding a therapy that can treat the basic cause of CF, it also continues to fund the development of therapies that treat the many different symptoms as well.

To speed up the development of these new therapies, the CF Foundation-accredited care centers participate in clinical research. Researchers and people with CF at the centers help in the testing or trials of drugs being developed.

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

OVERALL SURVIVAL FOR CYSTIC FIBROSIS

The median predicted age of survival for people with cystic fibrosis has risen dramatically over the past two decades.

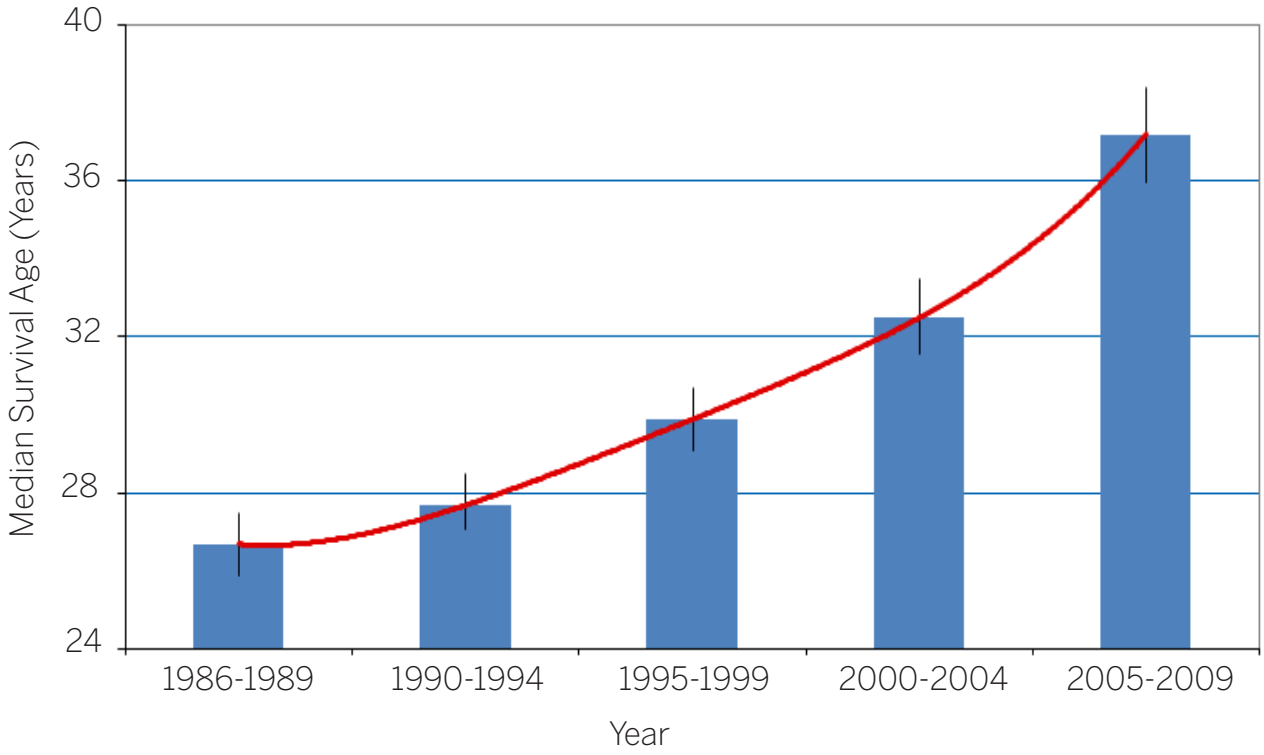
In 1985, the median age of survival was age 27. In 2009, it was 35.9.

While the survival age is less than what it was in 2008 (37.4 years), year-to-year fluctuations are normal when measuring health outcomes for a disease or medical condition. What is important is to see a trend of improvement over time.

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

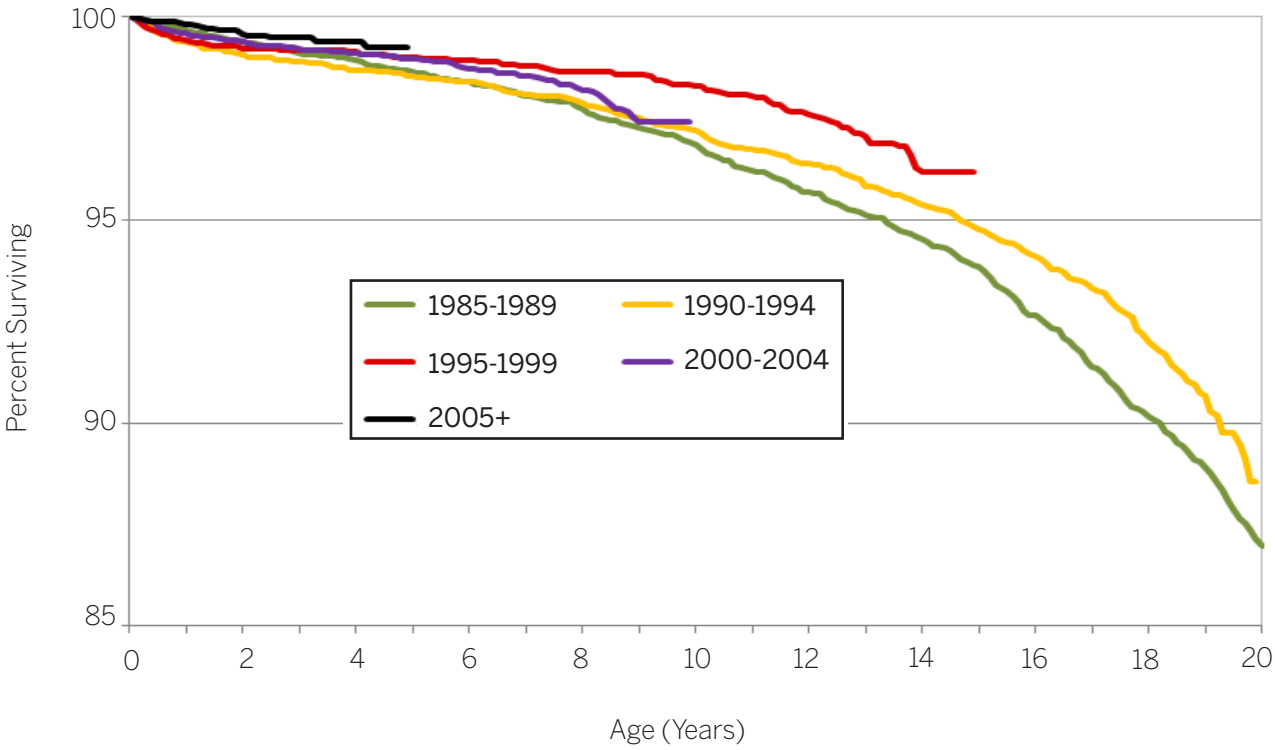
As the chart below shows, survival age for CF has consistently climbed to new levels.

Median Predicted Survival Age, 1986-2009
Over 5 Year Bands



Another way to look at how survival is improving is demonstrated in the following graph, which shows that fewer children with CF are dying in childhood. Of people with CF born between 1985 and 1989 (green line), 93.9 percent were alive at age 15. For children born between 1995 and 1999 (red line), 96.2 percent were alive at age 15.

Survival by Birth Cohort



There are many reasons for this increase in survival. The stronger partnerships among people with CF, their families and care centers are central. Another factor is earlier diagnosis, due to all states screening newborns for the disease. The earlier CF is diagnosed, the sooner treatment can begin. This helps to slow the disease and lessen or prevent problems. The infant care guidelines outline CF care that works to stop or slow the progression of CF. You can read these on the Foundation's website at www.cff.org/treatments/CFCareGuidelines/AgeSpecificCare.

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

The Phase 2 Infant Study of Inhaled Saline (ISIS) is looking at inhaled hypertonic (7%) saline in children under 5 years of age. This therapy may help to stop or slow damage from the buildup of mucus in the lungs. For more information on this trial, visit www.cff.org/research/ClinicalResearch/Find and search for clinical trials by age.

Research based on specific CF mutations or genotypes also continues. It is helpful for you to know your or your child's CF mutations. According to the Patient Registry, 90.7 percent of people with CF have had their CF mutations identified. More than 87% of people with CF have at least one ΔF508 (delta F508) mutation. However, there are more than 1,800 known mutations of the CF gene. The CF Foundation currently supports research to find out which ones cause CF. Below is a list of the most common CF mutations. If you do not know if you or your child have had your genotype identified, talk to your CF care center.

Most Common CF Mutations or Genotype

ΔF508 (delta F508)	G542X	G551D	R117H
W1282X	N1303K	R553X	

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.

All members of the CF care team will:

- be informed of how care decisions are made and carried out;
- tailor care to fit the needs of the person with CF; and
- treat everyone with respect.

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 CF care teams identify new ways to improve the health of people with CF at their center. The "Patient Summary Report" on the next page shows 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 CF clinic visit.

The care center data shows the average lung function and body mass index (BMI) for people with CF who are cared for at a CF center. You can learn more at www.cff.org/LivingWithCF/CareCenterNetwork/CareCenterData.

We encourage you to talk with your CF care center about the data. Consider 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?

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 "Partnering for Care" series, "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, under "Managing Life." You can also read the success stories of how others work with their care centers to improve care at www.cff.org/LivingWithCF/QualityImprovement.

CF CARE GUIDELINES

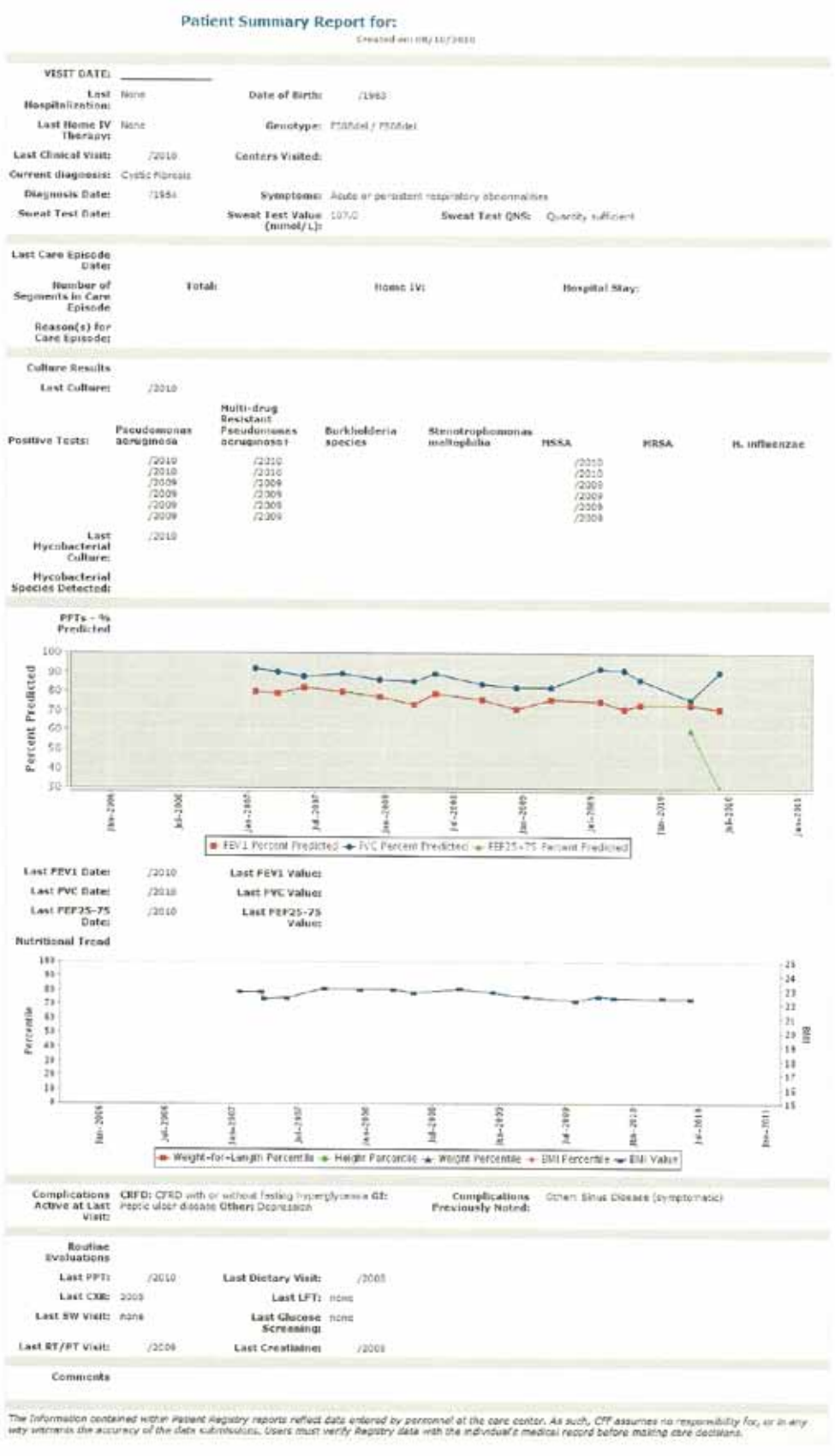
Care or practice guidelines are used in health care for many different diseases and conditions. These guidelines provide the healthcare professional with guidelines for how to best treat a disease or condition, based on published reports of clinical research.

To develop CF care guidelines, the CF Foundation gathers CF health experts to review the medical literature and research on CF care, along with data from the Patient Registry. These guidelines (www.cff.org/treatments/CFCareGuidelines) have information about diagnosis, nutrition, lung health and disease, infection control and age-specific care. Guidelines recently published include:

- Management of Infants with Cystic Fibrosis;
- Management of Infants with Cystic Fibrosis Transmembrane Conductance Regulator (CFTR)-Related Metabolic Syndrome (CRMS);
- Pulmonary Exacerbations; and
- Pulmonary Complications.

The care guidelines state that each year, people with CF should have certain lab tests, throat or sputum cultures and exams. The table on page 8 lists these and reports how people with CF in the Patient Registry met these guidelines in 2009, as compared to 2008.

SAMPLE PATIENT SUMMARY REPORT



Patient Summary Report for:

VISIT DATE: _____

Last Hospitalization: None — Date of Birth: /1963

Last Home IV Therapy: None — Genotype: F508del / F508del

Last Clinical Visit: /2010 — Centers Visited:

Current diagnosis: Cystic Fibrosis

Diagnosis Date: /1964 — Symptoms: Acute or persistent respiratory abnormalities

Sweat Test Date: — 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: /2010

Positive Tests:	Pseudomonas aeruginosa	Multi-drug Resistant Pseudomonas aeruginosa†	Burkholderia species	Stenotrophomonas maltophilia	MSSA	MRSA	H. influenzae
	/2010	/2010			/2010		
	/2010	/2010			/2010		
	/2009	/2009			/2009		
	/2009	/2009			/2009		
	/2009	/2009			/2009		
	/2009	/2009			/2009		

Last Mycobacterial Culture: /2010

Mycobacterial Species Detected:

PFTs - % Predicted

Last FEV1 Date: /2010 — Last FEV1 Value:

Last FVC Date: /2010 — Last FVC Value:

Last FEF25-75 Date: /2010 — Last FEF25-75 Value:

Nutritional Trend

Complications Active at Last Visit: CFRD: CFRD with or without fasting hyperglycemia GE: Peptic ulcer disease Others: Depression — Complications Previously Noted: Chronic Sinus Disease (symptomatic)

Routine Evaluations

Last PFT: /2010 — Last Dietary Visit: /2008

Last CXR: 2009 — Last LFT: none

Last SW Visit: none — Last Glucose Screening: none

Last RT/PT Visit: /2009 — Last Creatinine: /2009

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.

Guidelines for Yearly CF Care	People With CF Who Met the Guidelines (%)	
	2008	2009
4 clinic visits	63.9	65.0
4 respiratory cultures, by either throat swab or sputum	41.0	43.1
2 pulmonary function tests (PFTs) if physically able	83.3	84.0
An influenza (flu) vaccine if ≥ 6 months of age	67.5	69.5
Vitamin A, D and E levels in the blood measured (sometimes called fat-soluble vitamin levels)	82.7	83.1
An oral glucose tolerance test (OGTT) to screen for CF-related diabetes in people with CF age 10 and older	12.8	14.6
A test to measure liver enzymes in the blood (sometimes called a liver function test)	77.8	78.5

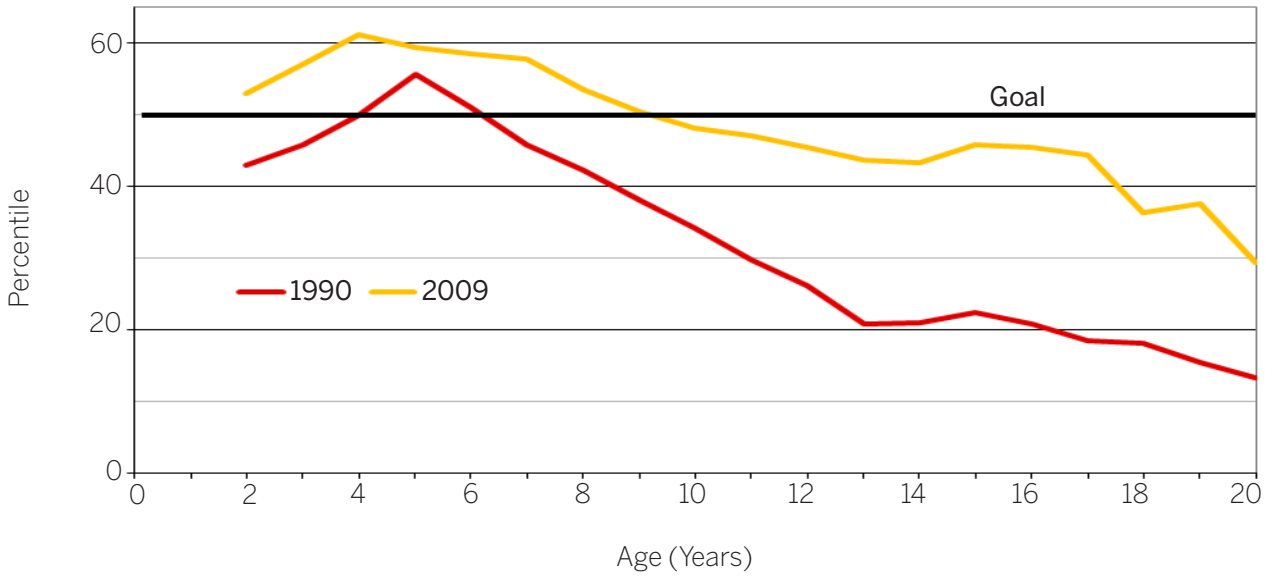
MAINTAINING NORMAL NUTRITION

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

The nutrition of people with CF is getting better, but more work remains to be done. The CF Care Guidelines set the goals for weight (measured as BMI) for children and adults with CF. BMI is based on a person's weight and height and is used to screen people for health problems. BMI is calculated by dividing body weight in kilograms by the person's height in meters squared (weight kg/height m^2=BMI). You can calculate your BMI or your child's BMI percentile on the Centers for Disease Control and Prevention's website (www.cdc.gov/healthyweight/assessing/bmi).

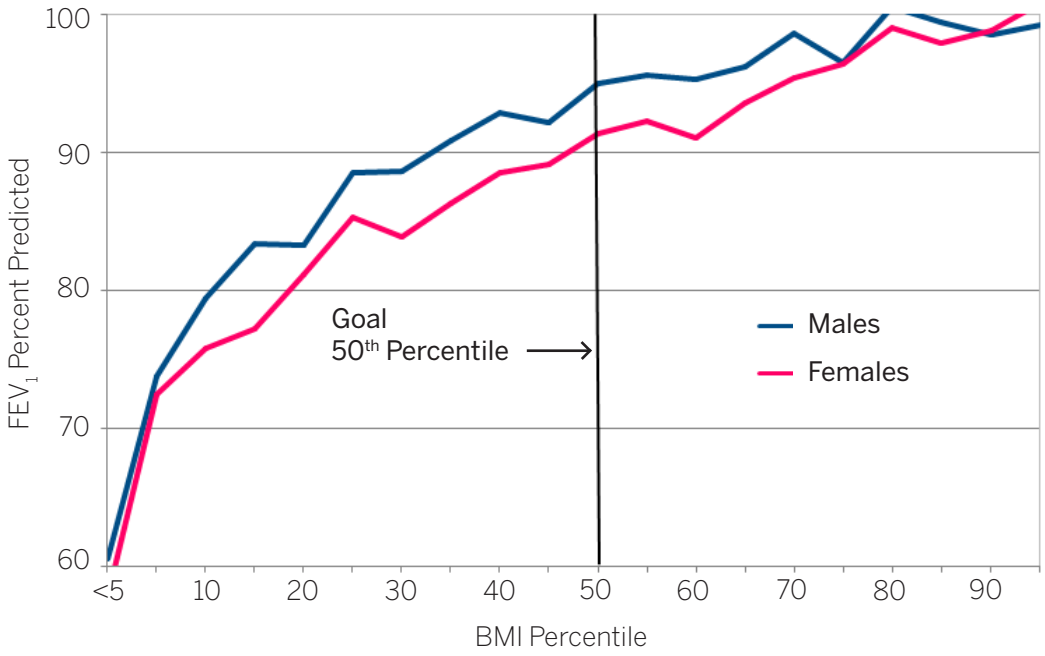
For children and teens, BMI is stated as a percentile matched up to children without CF of the same age and gender. The goal is for children with CF to grow and develop the same as children without CF and to have a BMI around the 50th percentile for their age. (The 50th percentile is the average BMI percentile for children in the United States who do not have CF.) The following graph shows how much the BMI percentile of children with CF has improved since 1990.

Median CDC BMI Percentiles vs. Age, 1990 and 2009



The Patient Registry shows a strong link between a higher BMI percentile and better lung function in children. One important measure of lung function in CF is FEV_1, or Forced Expiratory Volume over one second. This 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 highlighted bar is the BMI percentile goal for children with CF.

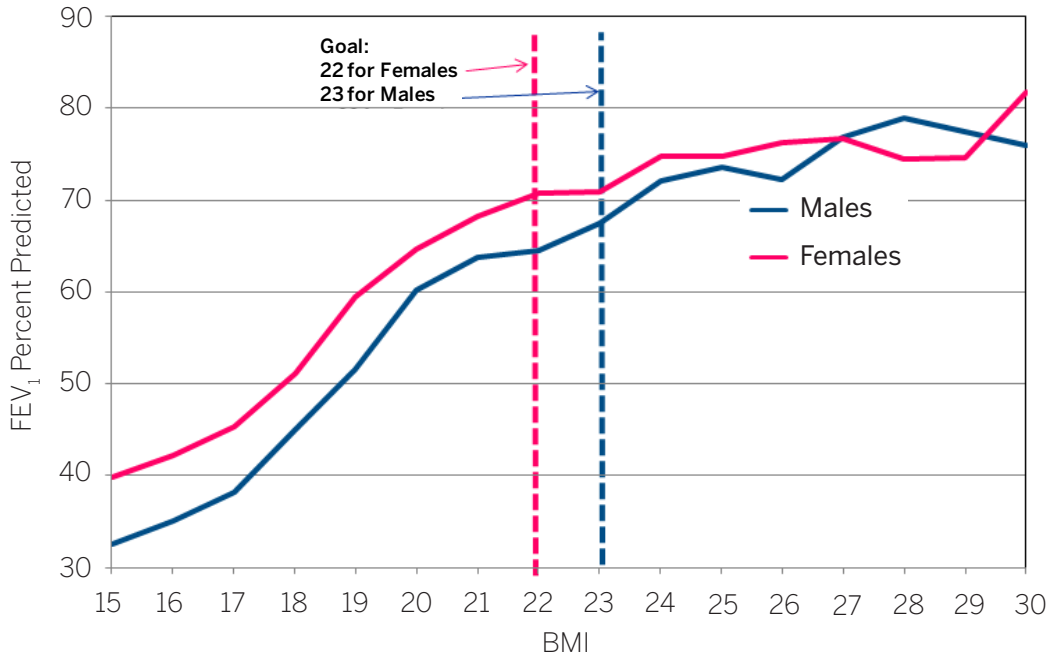
FEV_1 Percent Predicted vs. BMI Percentile in Patients 6 to 20 Years



The connection between a higher BMI and better lung function is also seen in adults with CF. Highlighted on the following graph 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.

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



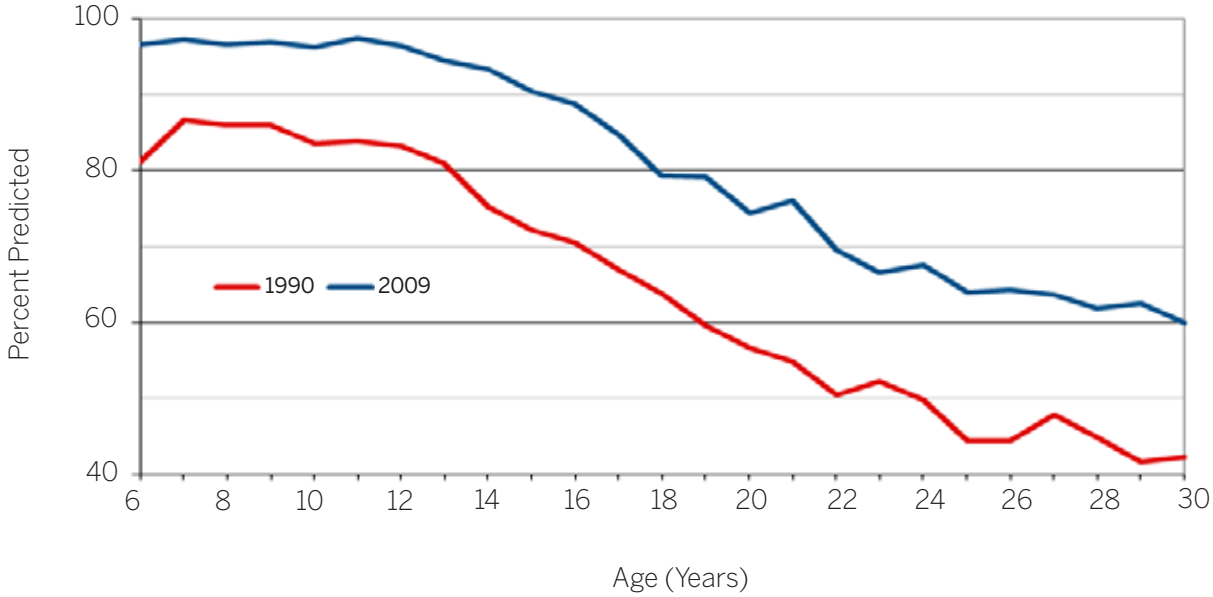
LUNG FUNCTION

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 them to previous levels of lung function.

People with CF often have pulmonary exacerbations — a period when lung disease gets worse. This can be an infection, an increase in cough and sputum, a drop in lung function and/or weight loss.

The graph below shows that the lung health of people with CF has improved since 1990. FEV$_1$ (lung function) is usually near normal or just under 100 percent predicted when first measured around 6 years of age.

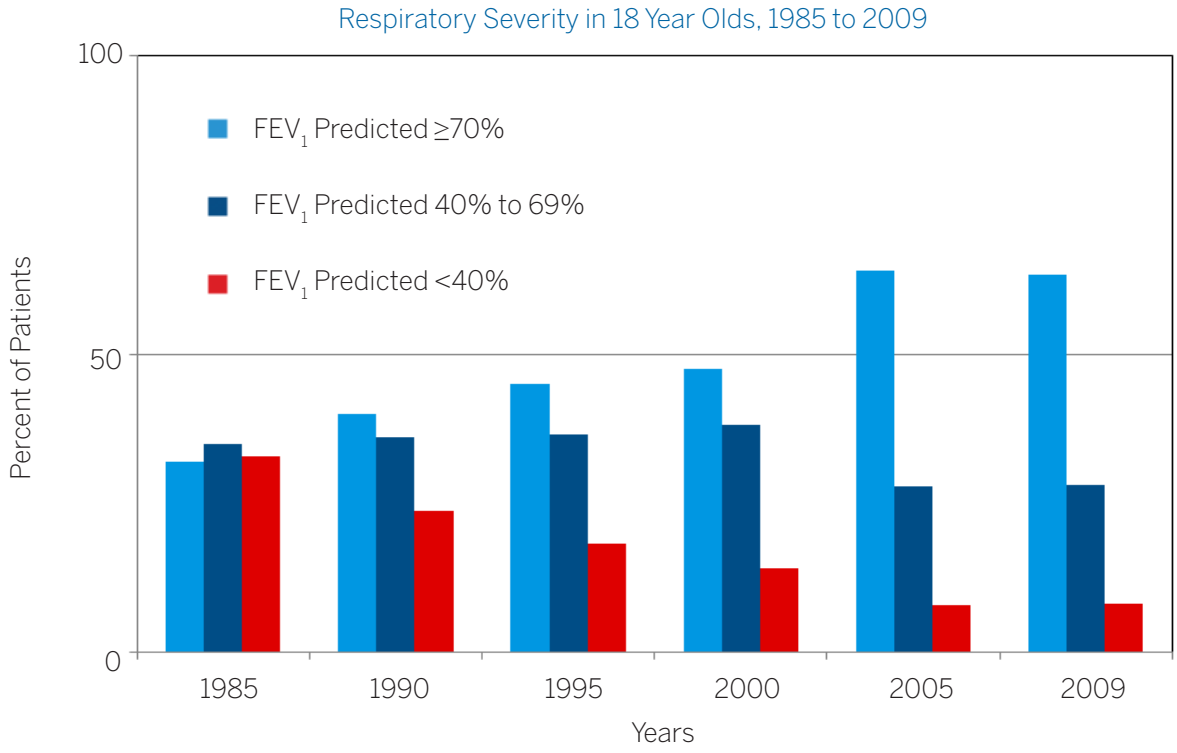
Median FEV$_1$ Percent Predicted vs. Age, 1990 and 2009



Research shows that a person with CF, even a baby, has lung damage even when their lung function or FEV_1 percent predicted is normal. In CF, the lower a person's FEV_1, the more severe the lung disease. An FEV_1 greater than or equal to 90 percent is normal. An FEV_1 from 70 to 89 percent indicates mild lung disease. An FEV_1 from 40 to 69 percent means moderate lung disease. An FEV_1 of less than 40 percent suggests severe lung disease.

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. To learn more about CF research, visit www.cff.org.

The graph below shows the growing number of 18-year-olds with CF in the Patient Registry with normal lung function or mild disease and the falling number of those with severe disease. This means that the lungs of people with CF are much healthier now than 20 years ago.



The "Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Function" lists medications that help people with CF maintain lung health. The following table shows the criteria for each medication, along with the percentage of people with CF who are taking each one. Who should take these drugs is set by the U.S. Food and Drug Administration (FDA) for each medicine. To find out if you or your child might do well on one of these medications, talk to your CF doctor.

In February 2010, the FDA approved the use of Cayston® (aztreonam for inhalation solution) as an inhaled antibiotic for people with CF who have *Pseudomonas aeruginosa* (*Pseudomonas*). Cayston, developed by Gilead Sciences, Inc., is the first CF drug to advance — from beginning to end — through the Foundation's Therapeutics Development Program. You can learn more about Cayston, other therapies and the Foundation's Therapeutics Development Program at www.cff.org.

New drugs must be tested in people with CF in clinical trials and approved by the FDA before a doctor can prescribe them. Therefore, people with CF play a critical role in developing new drugs, by volunteering to take part in clinical trials.

Talk to your care center to learn about clinical trials you or your child can join. You can "Find a Clinical Trial" by using the search tool on the Foundation's website at www.cff.org/research/ClinicalResearch/Find.

THERAPY USE IN THE CF POPULATION

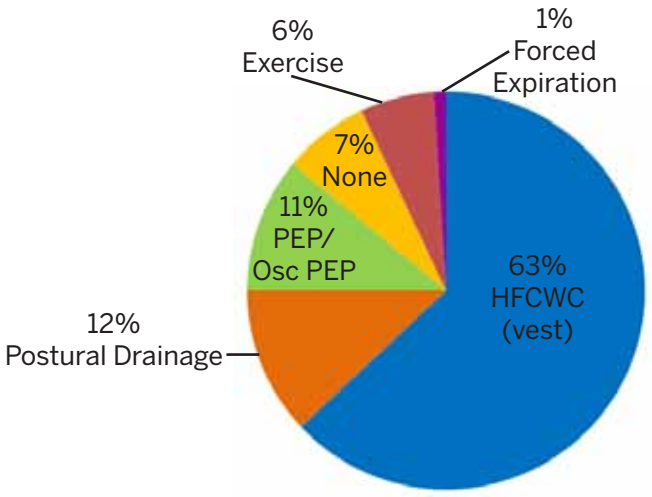
Percentage (%) of people who fit criteria for the medication and have had it prescribed

Chronic medication	Eligible patients using medication (%)	Criteria for prescribing medication
Inhaled tobramycin (TOBI ®)	68.7	• *P. aeruginosa* in cultures • at least 6 years old
Dornase alfa (Pulmozyme ®)	77.5	• at least 6 years old
Macrolides (azithromycin or Zithromax ®)	67.5	• at least 6 years old • weight over 25 kg (55lb) • FEV_1 over 30% predicted • no *B. cepacia* present • no liver disease
Ibuprofen	3.6	• 6-12 years old • FEV_1 over 60% predicted
Hypertonic saline	44.6	• at least 6 years old

Taking medicine is only part of the picture to 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 basic ACT is coughing. It is a reflex that clears mucus with high-speed airflow. However, in CF, the mucus cannot be cleared by coughing alone. There are many different ACTs that people with CF can use to help get the mucus out of their lungs.

In 2007, the CF Foundation set airway clearance guidelines for CF care (www.cff.org/treatments/CFCareGuidelines/Respiratory). The guidelines state that people with CF should do airway clearance to keep their lungs healthy, even when they are not sick. Which ACT a person uses can vary, but it is important that it be done every day. The chart on the next page shows the percentage of people with CF and which ACT is used. You can learn more about airway clearance at www.cff.org/treatments/Therapies/Respiratory/AirwayClearance and by watching the CF Education Web casts "Airway Clearance Techniques (ACT)" and "Help Your Respiratory or Physical Therapist Help You Thrive" on the CF Foundation's website.

Main Method of Airway Clearance Therapy (ACT)*



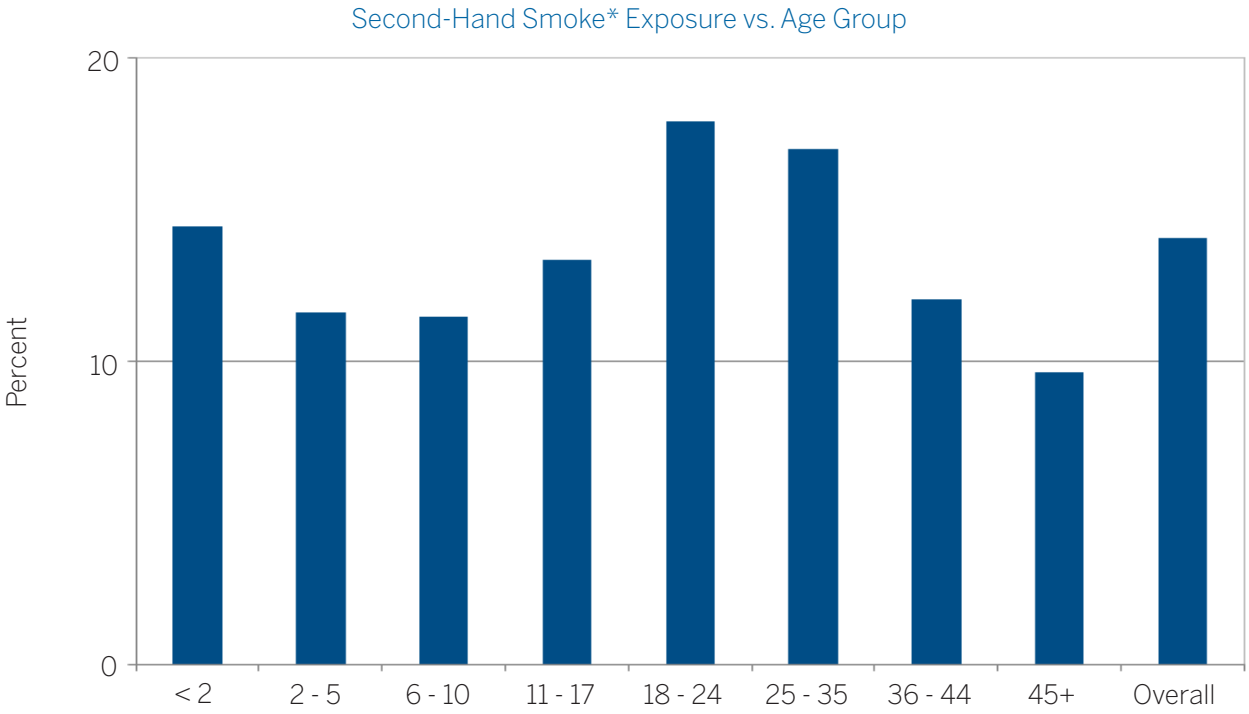
* The "Cystic Fibrosis Pulmonary Guidelines: Treatment of Pulmonary Exacerbations" recommends that airway clearance therapy be done more often when sick (www.cff.org/treatments/CFCareGuidelines/Respiratory).

Each time a person with CF has a respiratory illness, there may be more lung damage.

Here are some things you can do to prevent or lessen the chance of getting sick:

- Do airway clearance every day 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 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; and
  - cleaning and disinfecting nebulizers after every treatment.
- Avoid tobacco smoke.

The following graph shows, by age group, the percentage of people with CF who breathe in secondhand smoke. Second hand smoke is from burning tobacco in cigarettes, cigars, pipes and the smoke breathed out by people who smoke. Research has 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 U.S. Surgeon General has concluded that breathing even a little secondhand smoke can be harmful to your health.



* This includes daily or occasional exposure.

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;
- 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;
- Choose restaurants and other businesses that are smoke-free;
- Thank businesses for being smoke-free; and
- 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 best way to protect yourself and your family from secondhand smoke is to quit smoking. In the meantime, only smoke outside and away from others. Keeping your home and car smoke-free can also help you quit smoking. In addition, ask your doctor to help you quit smoking.

For more information, visit the "Smoking and Tobacco Use" section of the Centers for Disease Control and Prevention (CDC) website (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 starts to feel ill (e.g., 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 about lung care and therapies for people with CF, 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 watch archived Web casts about CF lung health and disease and how to partner with your care center (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 *Pseudomonas aeruginosa* (*Pseudomonas*) and *Burkholderia cepacia* (*B. cepacia*) complex.

The best way for anyone to avoid germs is to:

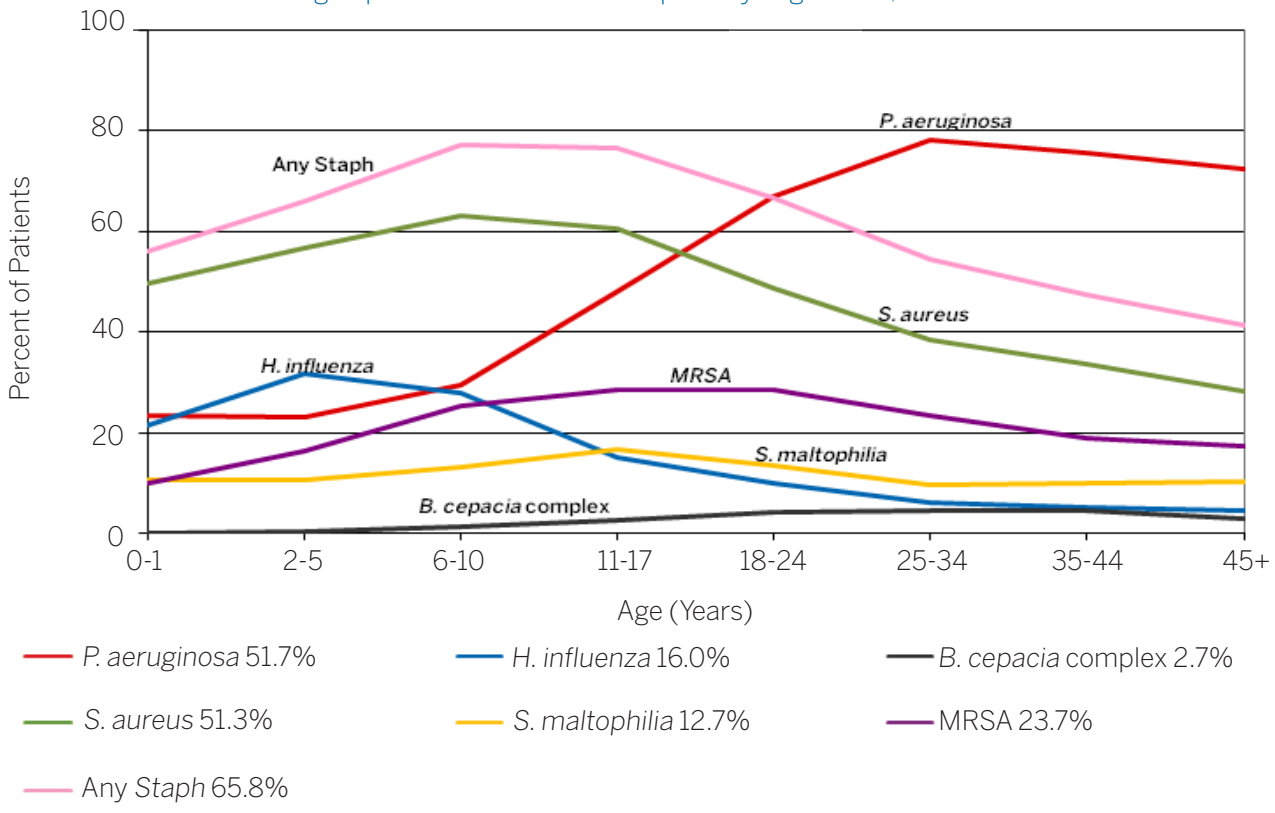
- Clean your hands often with soap and water or alcohol-based hand gels;
- Use a tissue when coughing or sneezing, then clean your hands;
- Avoid touching your eyes, nose or mouth to keep from spreading germs to yourself and others; and
- If you are ill, stay away from others.

Repeated lung infections are a concern for people with CF. Repeated infections and the inflammation from them damage the lungs.

This damage causes lung function (FEV_1) 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 (Influenza, MRSA, *B. cepacia*, etc.) and Web casts ("Germs, Infection Control and People With CF") are available at www.cff.org/LivingWithCF/StayingHealthy. You can also watch "Put Your Hands Together" at www.cdc.gov/CDCTV/HandsTogether.

Age Specific Prevalence of Respiratory Organisms, 2009



COMPLICATIONS 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, such as cystic fibrosis-related diabetes (CFRD). It is different from diabetes in people without CF. CFRD is common due to the damage the disease causes to the pancreas. Anyone with CF, 10 years of age and older, should be tested every year for CFRD.

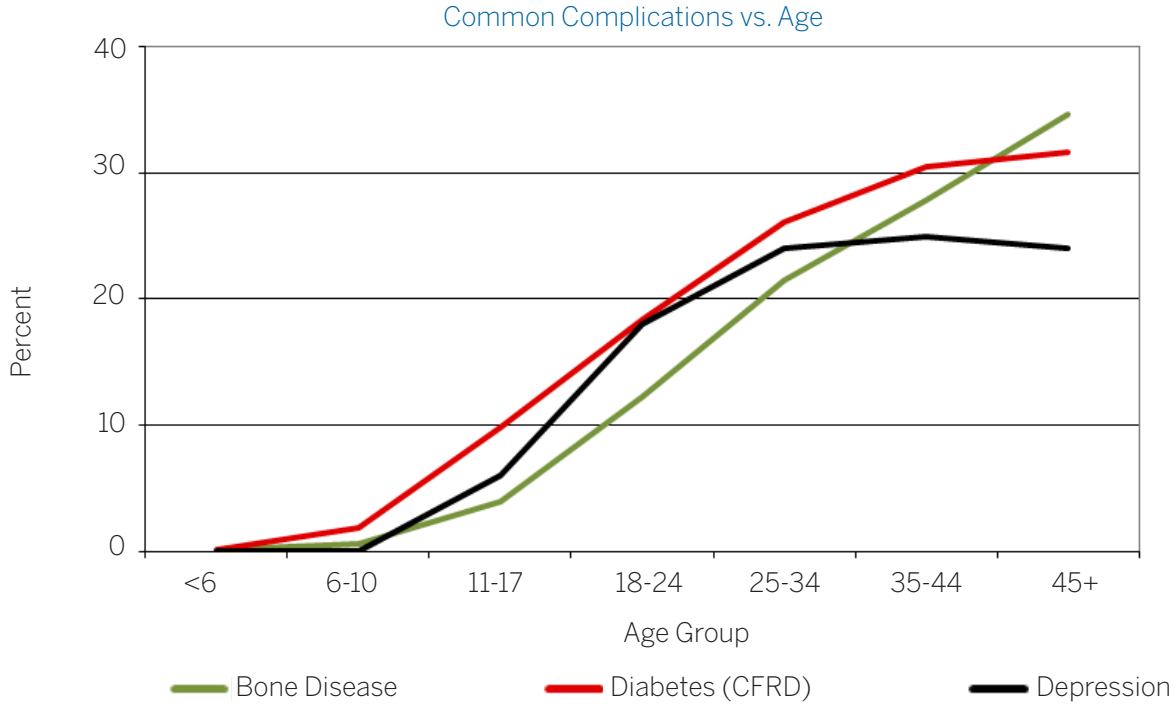
The Patient Registry data shows that early diagnosis and treatment of CFRD results in better nutrition and weight and, thus, better health. You can learn more about CFRD, how to manage it and the guidelines for CFRD care on the Foundation's website (www.cff.org/treatments). In addition, the Foundation continues to fund CFRD research; visit www.cff.org/research to learn more.

The Registry data also shows other complications of CF. It is estimated that about 12% of people in the U. S. have at least one sinus infection (acute sinusitis) in a year. About 24% of the people with CF in the Patient Registry report symptoms of sinus problems. Chronic sinusitis occurs when symptoms, such as headaches, dental pain, and facial congestion or fullness, last for eight or more weeks. If you or your child is having symptoms of chronic sinusitis, talk with your CF doctor.

Another common complication of CF is bone disease. Data from the Patient Registry show that about 20% of people with CF had bone disease in 2009. Preventing 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 more than 21% of adults and more than 2% of children with CF have signs of depression. This is common in people with other chronic diseases as well. People with CF, their families and caregivers need to be aware of this problem so it can be diagnosed and treated early. People often respond well to treatment for depression.

Other problems tracked by the Patient Registry include gastroesophageal reflux (GERD) and asthma. The CF Foundation is working to find ways to prevent or treat these complications of CF.



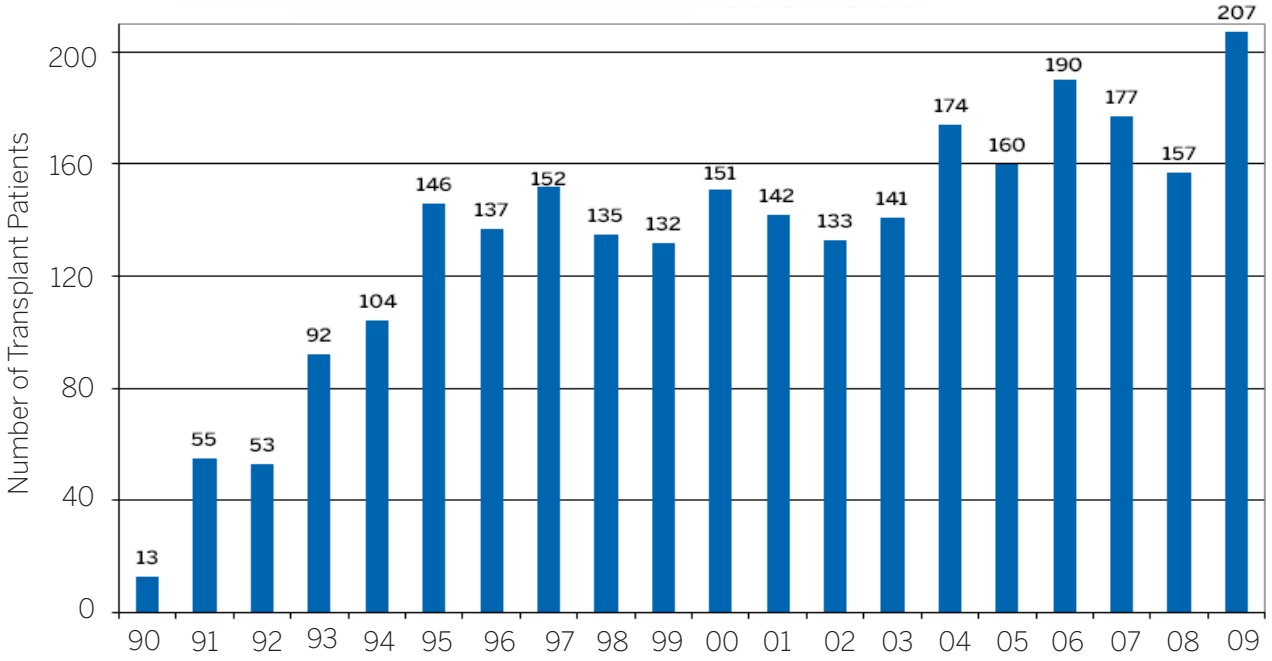
TRANSPLANTATION AND END-OF-LIFE CARE

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. A lung transplant is risky, however, and the supply of good donor lungs for transplant is limited. It is important to know who can benefit from a lung transplant and the best times for a transplant.

Research based on data from the Patient Registry has been used to help find out who is most likely to benefit. To learn more about lung transplants and organ donation, watch the CF Web cast "Partnering for Care" series and visit www.cff.org/treatments/LungTransplantation or the United Network for Organ Sharing (UNOS) at www.tranplantliving.org or www.OrganDonor.gov.

Number of CF Patients Undergoing Lung Transplantation Each Year, 1990-2009



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

There are a number of programs available to help people with CF afford the medical care and medications they need. The Cystic Fibrosis Patient Assistance Foundation (CFPAF), a 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 website (www.cfpaf.org) or call toll-free (1-888-315-4154) to learn more.

In addition, the CF Services Pharmacy, a full-service, mail-order pharmacy and wholly-owned subsidiary of the CF Foundation, works hard to make CF drugs available to everyone. It runs CF-specific patient assistance programs to help people with CF get their medications. Visit www.cfservicespharmacy.com to learn more.

Data from the Patient Registry shows that drugs like Pulmozyme® and TOBI® are available to people with CF, regardless of 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 at www.cff.org/LivingWithCF/Webcasts.

For information about the race and age of the people in the Patient Registry, "A Summary of the 2009 Data" can be found on page 21.

Insurance Coverage 2009

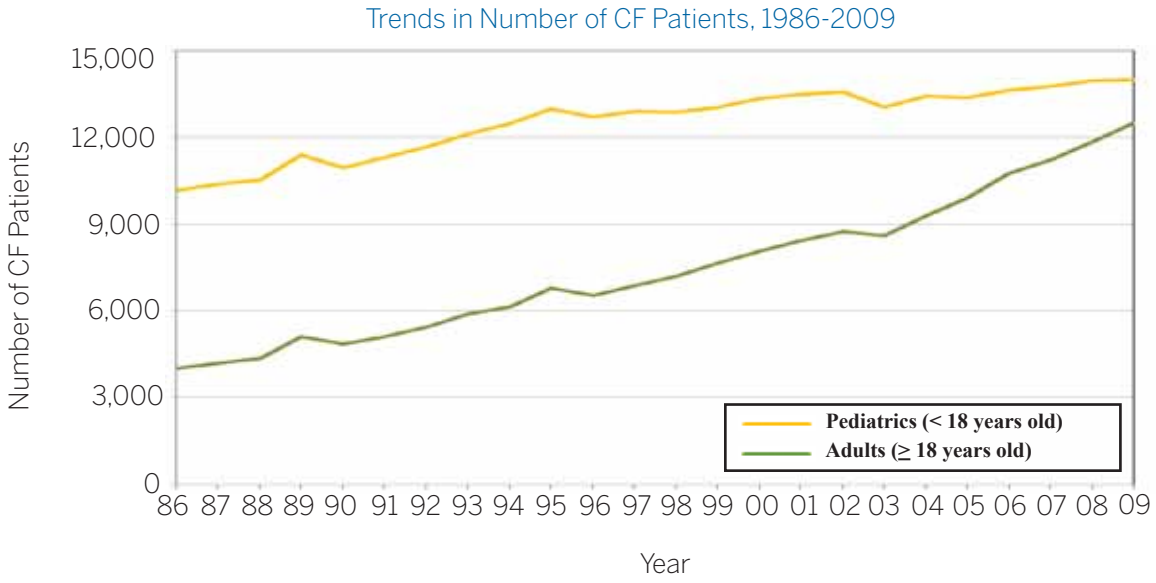
Type of Insurance*	Children <18 Years (%)	Adults ≥ 18 Years (%)
No Insurance	2.3	4.2
Private/HMO	58.4	64.3
Medicaid/State	49.6	34.1
CHAMPUS	2.0	1.5
Federal	1.0	13.0
Other	1.7	2.9

*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 website at www.cff.org/LivingWithCF. If you have trouble paying for health care, the Foundation urges you to 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.

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. This means 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 2009, more than 47 percent of people with CF in the Patient Registry were adults, and that number keeps growing.

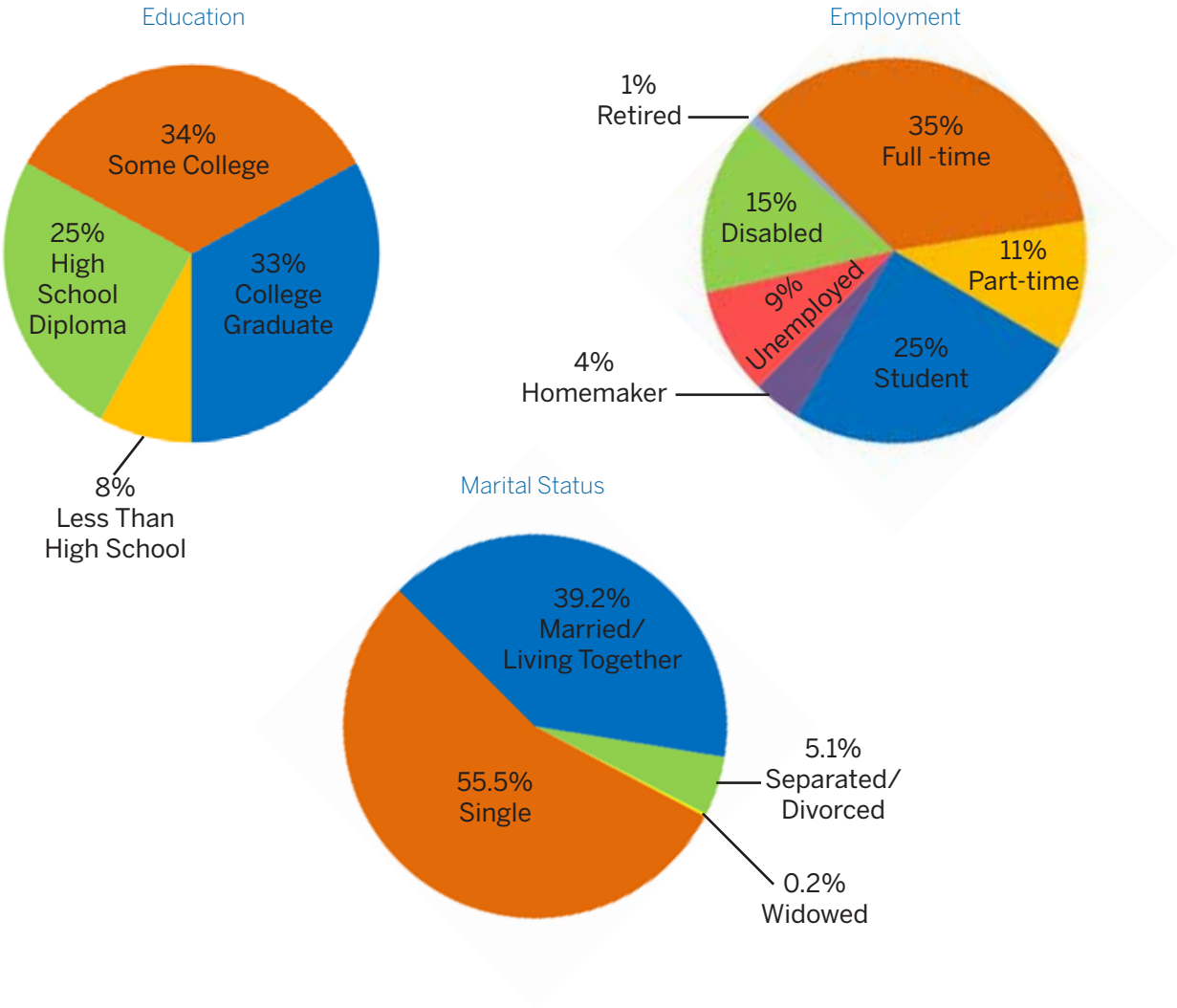
Because of the growing number of adults with CF, the CF Foundation developed guidelines for the care of adults with CF (www.cff.org/treatments/CFCareGuidelines). One of the goals of health care in general, is to give care that fits a person's age regardless of the disease or condition. This means that an adult gets care from a doctor and other healthcare professionals who are trained to care for adults.

To meet this goal, the CF Foundation supports clinics for adults with CF and gives grants to help more doctors train in the care of adults with CF to meet the growing numbers.

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.

You can learn more about CF adult care and transition into adult clinics by watching the "Partnering for Care" Web cast series (www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts).

Characteristics of Adult CF Patients ≥ 18 Years Seen in 2009



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 and baby. In 2009, the Patient Registry reported that 226 women with CF were pregnant. Advances in fertility medicine have given men with CF the option to father children. 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, under "Managing Life."

WHO ARE THE PATIENTS IN THE CF FOUNDATION'S PATIENT REGISTRY — A SUMMARY OF THE 2009 DATA.

CF patients (number)	26,477
Newly diagnosed patients in 2009 (number)	960
Patients diagnosed by NBS (%)	49.8
Age at diagnosis (median)	5 months
Age range	0 to 89 years
Total number of deaths	440
Predicted median survival	35.9 years
Patients 18 years and older (%)	47.2
Race/Ethnicity (%)	
Caucasian	94.4
Hispanic (black or white)	7.3
African American	4.3
Males (%)	51.8
Genotyped (%)	90.7
Home Therapy	
IV Antibiotics	20.6
Oxygen Use	11.3
Supplemental Feeding	
Tube	10.9
Oral (by mouth)	38.8
Pancreatic enzyme supplements (%)	89.1
Clinical trial participation (number)	4,029
FEV_1 % predicted (mean)	76.3
Increased respiratory symptoms or a pulmonary exacerbation (%)	34.7
Median BMI percentile for patients 2-20 years* (%)	48.7
Median BMI for patients ≥ 21 years* (%)	22.0
Respiratory cultures positive for (%)	
P. aeruginosa	51.7
B. cepacia complex	2.7
*S. aureus***	51.3
S. maltophilia	12.7
MRSA	23.7
Complications (%)	
Diabetes (CFRD)/glucose intolerance	22.0
Bone disease	10.2
Liver disease	10.8
Nasal polyps requiring surgery	24.0
Distal intestinal obstruction syndrome	3.8
Depression in adults	21.6
Transplants (numbers)	
Lung:	
Bilateral	205
Lobar-Cadaveric	7
Liver:	15
Therapies**	
TOBI® (tobramycin)	68.7
Pulmozyme® (rhDNase)	77.5
Ibuprofen	3.6
Macrolides (azithromycin)	67.5
Hypertonic saline	46.2
Pregnancies	226
Live Birth Rate	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/healthyweight/assessing/bmi.

**This is the percentage of patients who are eligible for a therapy and had it prescribed at least once in 2009.

***Does not include MRSA

NUMBER OF PATIENTS BY STATE IN THE CF PATIENT REGISTRY

State	Number	Percent
Alabama	433	1.64
Alaska	57	0.22
Arizona	397	1.5
Arkansas	233	0.88
California	2,085	7.87
Colorado	532	2.01
Connecticut	312	1.18
Delaware	66	0.25
District of Columbia	21	0.08
Florida	1,314	4.96
Georgia	765	2.89
Hawaii	6	0.02
Idaho	165	0.62
Illinois	1,004	3.79
Indiana	602	2.27
Iowa	384	1.45
Kansas	352	1.33
Kentucky	507	1.91
Louisiana	323	1.22
Maine	224	0.85
Maryland	508	1.92
Massachusetts	839	3.17
Michigan	1,018	3.84
Minnesota	582	2.2
Mississippi	233	0.88
Missouri	692	2.61
Montana	112	0.42

State	Number	Percent
Nebraska	222	0.84
Nevada	168	0.63
New Hampshire	200	0.76
New Jersey	667	2.52
New Mexico	119	0.45
New York	1,623	6.13
North Carolina	812	3.07
North Dakota	70	0.26
Ohio	1,464	5.53
Oklahoma	279	1.05
Oregon	309	1.17
Pennsylvania	1,424	5.38
Puerto Rico	1	0.0
Rhode Island	96	0.36
South Carolina	357	1.35
South Dakota	101	0.38
Tennessee	575	2.17
Texas	1,461	5.52
Utah	352	1.33
Vermont	135	0.51
Virgin Islands	0	0.0
Virginia	715	2.7
Washington	583	2.2
West Virginia	229	0.86
Wisconsin	641	2.42
Wyoming	49	0.19
Foreign	59	0.22

SOURCE OF DATA:
Cystic fibrosis patients currently under care at CF Foundation-accredited care centers in the United States, 2009

SUGGESTED CITATION:
Cystic Fibrosis Foundation Patient Registry
2009 Annual Data Report
Bethesda, Maryland

Cystic Fibrosis Foundation

Adding *tomorrows* every day.

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