

Daily Self Care Guidelines

The Cystic Fibrosis Foundation (CFF) has identified several daily activities that promote long-term health in patients with CF. We strongly recommend that you perform these activities daily, and become knowledgeable and expert in how these benefit your health. We will help you learn the best techniques for you, and why they work. We recognize that doing everything everyday will be challenging, but the more you do, the better you will be.

Airway Clearance Techniques

Airway clearance techniques (ACT) are important because they help both normal and abnormal secretions move through (up and out) the airways. Techniques include CPT, positive expiratory pressure (PEP), oscillating PEP (flutter and Acapella), autogenic drainage (AD), active cycle breathing technique (ACBT), the vest, intrapulmonary percussive ventilation (IPV), and vigorous exercise.

Recommendation: ACT should be done twice a day when you are well, and 4 times a day when you have increasing respiratory symptoms (e.g. cough, wheezing, congestion, etc.)

Goal: Develop a working knowledge of several types of ACT's and know when to increase their use from 2 to 4 times a day. With time, become expert in use of ACT.

Nutrition

The CFF has found that maintenance of nutrition and a healthy weight is associated with better health, better lung function, and longer life. Many people with CF have lower than normal weight. Because you need more calories than those people without CF, you need to eat more calories—150% - 200% more than what other people eat. You also need more vitamins and salt than people without CF. It is important that you eat a high-calorie diet, as well as a balanced diet (meats/beans, grains, fruits, vegetables, and dairy). To get the 150-200% more calories needed, be sure to eat 3 meals and 3 snacks every day.

The CFF recommends that we calculate weight-for-length in young children, and Body Mass Index (BMI) in those people 2 years and older. If your BMI is below the optimal level, or if you are not gaining weight appropriately, the dietitian will work with you to help plan for improving your nutrition and growth. Often, the dietitian will suggest high calorie shakes/drinks to make sure you get enough calories. In some cases, tube feedings at night are used if you cannot eat enough calories to gain weight. The dietitian will help you manage tube feedings and fit them into your schedule.

CFF recommend children 2-20 years maintain a BMI at or above the 50%tile for age.

Children under 6 months old should maintain a weight-for-length of 50%tile.

Children 6 months- 2 years should maintain a weight-for-length of 75%tile.

The OSF Saint Francis Medical Center Cystic Fibrosis center developed the following categories to help you better understand your child's level of nutrition.

Optimal nutrition:	Greater than or equal to 50% BMI for age
Sub-optimal Nutrition:	25%-49% BMI for age
At Risk:	10%-24% for age
Urgent Need:	Less than 10% BMI for age.

CFF defines Nutrition for adults Greater than 20 years old as follows:

Women should have a BMI of 22 or above.

Men should have a BMI of 23 or above.

Recommendation: Eat 3 balanced meals and 3 snacks each day. Take your pancreatic enzymes, if prescribed, with meals and snacks.

Drink plenty of fluids daily and be sure to drink water every day.

Take age appropriate CF specific Multivitamin daily.

Add salt to meals and snacks as directed by the dietitian.

Goals: Develop knowledge of nutrition, including types and amounts of food you need to eat.

Understand why your nutrition needs are different than someone without CF.

Understand the relationship between your goal BMI and your lung function.

Understand the need for and dosing of pancreatic enzyme replacement therapy (PERT).

Know your BMI and level of nutrition.

Exercise

Regular exercise has many benefits for all people. It is especially important for people with CF.

There is evidence that physical fitness enhances the immune system and helps a person fight infection. People who are physically fit have less severe and less frequent episode of infection.

Exercise may also be a valuable part of airway clearance techniques (ACT).

Recommendations: Thirty minutes of exercise daily. Cardio/aerobic exercise, (stamina and endurance exercise) should be done on most days. Examples include brisk walking, biking, jogging, swimming, and use of aerobic exercise equipment. Strength building exercises, for older children and adults, should be done 2-3 times weekly. People may use stack weights systems at a fitness center or inexpensive dumbbell weights at home. Your physical therapist can help you develop a personal exercise program for you.

Goals: Develop a working knowledge of types of exercise. Achieve and maintain a state of aerobic fitness. Achieve and maintain a better than average level of strength.

Medications and Treatments

Medications may be prescribed to maintain your health for a variety of reasons, including prevention of vitamin deficiency, enzyme replacement therapy, inhalers for asthmatic breathing,

nebulizers for mucous clearance, and nebulized antibiotics. Specific examples include vitamins, pancreatic enzymes, inhaled albuterol, inhaled corticosteroids (ICS), pulmozyme, hypertonic saline nebulized, nebulized tobramycin, nebulized colistin, and azithromycin (Zithromax).

Recommendations: Use medications as prescribed.

Goals: Use medications on a regular basis and understand the reason for the use of each medication and know the dosing.

CF-Related Diabetes

CF-related diabetes (CFRD) is common in people with CF. It is detected by doing an oral glucose tolerance test (OGGT), which is recommended yearly beginning at age 10. If CFRD is treated, you may be more susceptible to infections and may take longer to recover from infections. You may also have difficulty maintaining your goal weight if CFRD is not well controlled.

Recommendations:

Get yearly Oral Glucose Tolerance Test (OGGT) beginning at 10.

If you have CFRD:

Monitor your blood sugar as instructed by your diabetes provider

Take your insulin as prescribed.

Goals: Know how diabetes can affect your growth and your health

Know proper treatment for CFRD.

Maintain your goal weight.

Respiratory Infections/Exacerbations

Acute respiratory infections occur periodically in people with CF. If these infections are not treated quickly, they may trigger a lot of secretions (mucus) and pus, and eventually cause damage to the bronchial system. Repeated infections over the years result in progressive loss of lung function. We refer to acute infections as Pulmonary Exacerbations (PEX). We developed a scoring system to help you and us detect and treat PEX quickly and aggressively. The CF centers whose patients have the best lung function treat PEX often and quickly. We want you to call at the first sign of lung infection, so that we can treat you promptly. Symptoms to watch for include increase in sputum amount and/or color, increased cough or wheeze, shortness of breath, blood in the sputum, sinus congestion, feeling ill, low energy, fever, chills, sweats, poor appetite, weight loss, chest pain, poorly controlled sugar levels in those with CFRD.

Recommendations: Know the symptoms of Pulmonary Exacerbation (PEX), and call the center early if any new symptoms develop. At the first sign of a respiratory infection, remember to increase your airway clearance techniques (ACTs) to 4 times a day. **We would like you to call if you have any new respiratory symptoms.**

Goals: Identify and treat your Pulmonary Exacerbations early to maintain optimal lung function.

