Health Maintenance Guidelines

A patient's guide to the Cystic Fibrosis Foundation Clinical Practice Guidelines for Individuals with Cystic Fibrosis

Daily Health Maintenance

The most important behaviors for maintaining optimal health are those you do daily:

- **At least twice daily airway clearance techniques (ACT) (e.g. Vest, Acapella, CPT, etc)**
- **Regular exercise.**
- **A well-balanced, high calorie diet.**
- **Early medical attention to respiratory symptoms of increased cough and sputum.**
- **Consistent use of prescribed preventative therapies and medications.**

Quarterly Health Recommendations (At **least** 4 times a year)

CF Clinic Visits: Quarterly clinic visits allow you and the CF team to meet and evaluate your health maintenance plans and implement new strategies when needed to optimize your health. It also provides opportunity to review standard treatments and learn about new available therapies. **We believe that this is one of the most important guidelines**, and would strongly recommend that you come to clinic when you are feeling well at least 4 times a year.

Sputum and throat cultures: Cultures from the respiratory tract give valuable information about how to manage lung infections. Bacteria in the lungs can change over time, so having a recent culture is the most accurate way to treat lung infections when they occur. Cultures should be obtained at an accredited CF laboratory and processed using special techniques to identify organisms common in CF. At least once a year the sputum will also be cultured for tuberculosis.

Pulmonary Function Test (PFT)*: Frequent monitoring of PFT's can detect subtle changes in lung health before symptoms become noticeable and/or troublesome. The CFF recommends PFT's twice a year, and we have opted to exceed this minimum guideline in order to detect and respond to any decline in lung function sooner. We may also obtain PFT's at times of illness and to help evaluate the lung's response to various treatments.

Respiratory Care Review*: At each clinic visit, your respiratory therapist will teach and review airway clearance techniques, inhaled or nebulized medication administration, and respiratory equipment care and cleaning.

Physical Therapy Review*: Your Physical therapist will evaluate your development and upper body positioning for improved breathing. She will also help you individualize an exercise program to optimize your strength and endurance.

Nutritional Evaluation*: Your dietician will assess your growth, nutrition, dietary habits and nutritional therapies at each clinic visit. At least once a year, she will include a measure of body muscle and fat compositions. This allows you and the dietician to discuss changes in your

nutritional health, and make any necessary adjustments to your diet and pancreatic enzymes. Good nutrition is very important for good lung function.

Counseling Assessment*: Chronic illness has a major impact on the entire family, and you will meet with your CF Counselor at each clinic visit. This is an excellent time to address concerns such as coping, childhood behavior and development, school, or other issues important to your family.

*We exceed the CFF guidelines in order to better care for you.

Annual Laboratory Recommendations

These tests are essential in monitoring for complications in CF. Yearly lab evaluations should include:

Complete Blood Count (CBC): Checks for anemia (Adequate red blood cells), clotting dysfunction (platelets), and the body's response to infection (type and number of white blood cells).

Complete Metabolic Profile (CMP): Checks for salt imbalance (electrolytes), kidney and liver dysfunction. Patients with CF may have a salt imbalance or CF-related liver disease. Some of the medicines used to fight infections can affect the kidney, making it important to check kidney function periodically.

Vitamin levels: Patients with CF do not absorb fats in general very well, which affects some vitamin absorption. Checking the levels of fat-soluble vitamins (A, E, D, and K), looks for inadequate absorption of these that can lead to complications such as vision loss or poor bone health.

Immunoglobulin E (IgE): Checks for an allergic response of the body and screens for a condition called allergic bronchopulmonary aspergillosis (ABPA), which is more common in CF patients.

Oral Glucose Tolerance Test (OGGT): The OGGT tests the body's ability to process sugars and is the preferred test for CF-related diabetes (CFRD). It is recommended once a year beginning at10 years of age.

Periodic Tests

Chest Xray: Chest Xrays are generally ordered every 2 to 4 years depending on other symptoms of lung health. Chest Xrays give a limited picture of the lungs, and can be helpful in tracking changes over time.

Chest Computed Tomography (Chest CT): A chest CT gives an in depth three dimensional picture of the lungs and can be helpful in distinguishing early or subtle lung damage, individual anatomic variations, and other lung problems.

Dexascan: This scan is a simple radiological exam to evaluate bone health, recommended at regular intervals for all individuals with CF over 18 years of age.

Stool Elastase: This test of usually done only once, and tests the pancreas's ability to produce enzymes that are important in digesting foods. This tells if a person with CF is Pancreatic Insufficient (PI) (their pancreas doesn't produce enough enzymes), or Pancreatic Sufficient (PS) (their pancreas does produce enzymes). About 15% of people with CF produce enough pancreatic enzymes and are Pancreatic Sufficient.

Vaccines

It is very important to prevent any lung infections that we can in people with CF. There are several potentially serious lung infections that we can prevent easily. We recommend these vaccines for you along with your regular immunizations:

Influenza: This should be given during the Fall/Winter (flu season) each year, beginning at 6 months of age. The first year you receive this vaccine if you are less than 9 years old, you will receive 2 immunizations, at least one month apart. The entire family needs to receive the flu vaccine in order to further prevent you from becoming infected with the influenza virus.

Prevnar (PCV_7): This is part of the regular vaccine series given to all children beginning at 2 months of age. (given at 2, 4, 6, and 12 months) This prevents infections caused by the bacteria *Streptococcus pneumoniae,* which causes meningitis, pneumonia and many ear infections.

Pneumovax (PPV_{23}): This is a different vaccine to protect you against infections caused by the bacteria *Streptococcus pneumoniae,* which is one of the most common causes of bacterial pneumonia. If you had 4 doses of Prevnar, you should get Pneumovax 3 to 5 years after your last dose of Prevnar, and then again five years later.

Hib: This is also part of the regular vaccine series, and begins at 2 months of age. It prevents infections caused by the bacteria *Hemophilus Influenza type B,* which causes meningitis, blood infections, ear infections and pneumonia.

DTaP and Tdap: The DTaP, (Diphtheria, Tetanus and acelluar Pertussis), is part of the childhood series of vaccines, and prevents tetanus, diphtheria, and whooping cough. The Tdap is a similar vaccine, which was added in 2005 as a booster to further prevent whopping cough in teens and adults. You should get the Tdap booster at 5^{th} grade entry. If you did not receive it then, you and your adult family members should get it once. After the Tdap booster, every adult needs to return to the Td booster every ten years.

Infants and Young Children

Although most of the above recommendations apply to all people with CF, there are areas where the guidelines differ. Infants and young children follow the regular guidelines with the following exceptions:

CF Clinic Visits: If you are less than one year old, your CF team would like to see you every month in order to monitor your growth, your lung function, and your development. After you turn one, you will be seen for health maintenance visits in CF clinic every 3 months, unless your growth or lungs need to be monitored more closely.

PFT's: Children less than 6 years old are usually unable to perform PFT's, so their lung function is evaluated by the entire CF team at each clinic visit by looking at signs of illness (cough, wheezing, breathing hard or fast), breathing patterns, and upper body/chest development.

Sputum Cultures: Children younger than 6 years old are usually unable to cough up sputum. Because it is important to know what bacteria are growing in your lungs, we will culture the throat at least every three months. This will help us to know what treatment to start if you get sick with coughing or wheezing. Cultures may also be obtained more frequently if you are sick.

Our Pledge to Patients and Families

In showing our commitment to the constant improvement of quality and length of life for children and adults living with cystic fibrosis, and in accordance with Cystic Fibrosis Foundation goals, we promise to:

1. Treat children and adults with CF and their families as partners in care through timely and open communication.
2. Provide care that is respectful of patients' needs, preferences and values.
3. Provide medical and nutritional care that is aimed at achieving normal growth and good nutrition.
4. Prescribe therapies that have been proven to slow the progression of lung decline and decrease the frequency of pulmonary exacerbations.
5. Accurately diagnose and aggressively treat pulmonary exacerbations to minimize the loss of lung function.
6. Work with our clinics, hospital, patients and families to minimize the chances of spread of infection in both health care and community settings.
7. Monitor for complications of CF, especially CF-related diabetes mellitus, and treat them with the best-proven therapies when they arise.
8. Assure that all individuals with CF receive appropriate treatment regardless of race, religion, education or insurance coverage.
9. Provide timely, complete and compassionate guidance for difficult decisions such as lung transplantation.
10. Protect the privacy of all patients and families.

Accreditation of CF Care Centers

The Cystic Fibrosis Foundation accredits the Cystic Fibrosis Center at Children's Hospital of Illinois at OSF Saint Francis Medical Center as a Care and Teaching Center. Accredited CF Care Centers receive oversight and funding from the CF Foundation, and undergo periodic review by the CF Foundation Center Committee to ensure the highest standards of CF care are met in keeping with current CFF Practice Guidelines.