Cystic Fibrosis Interventions

Cystic fibrosis (CF) is a life-threatening, genetic disorder. In patients with CF, secretions found in the lungs, pancreas, intestines and other organs become abnormally thick and sticky. Interventions are necessary to restore lung function and to correct nutritional deficiencies caused by malabsorption of nutrients. Breathing treatments and chest physical therapy are necessary to rid the lungs of mucus, while dietary modifications help to maintain proper nutrition and electrolyte balance. As the child grows, pancreatic insufficiency and pancreatic fibrosis develops leading to CF-related diabetes, which is treated with insulin.

PLAY PICMONIC

Bronchodilators

Broccoli-dyed-hair

Bronchodilators will facilitate opening of the airway, making it easier to breathe. These medications are also used to prevent bronchospasm. Patients with CF may also be on corticosteroids as part of their treatment regimen.

N-Acetylcysteine (Mucolytics)

N-seagull-Sistine

This medication is a mucolytic agent used to loosen or break up mucus. Though effective, N-Acetylcysteine is not used frequently due to lung irritation.

Inhaled Hypertonic Saline

Inhaled Hiker-tonic Saline-sail

Hypertonic saline is inhaled via a nebulizer. Because the concentration of salt in this solution is hypertonic, water from the body is drawn out of the cells and into the mucus in the airway. The water acts to thin the secretions, making them easier to clear. Patients receiving this treatment may require the use of a bronchodilator to prevent bronchospasm.

Early Antibiotics

Early-sun and ABX-guy

Early treatment with antibiotics can help improve patient outcomes, especially in patients with infections caused by Pseudomonas. Antibiotic therapy should be modified according to culture and sensitivity results obtained prior to administering the first dose of antibiotics.

Dornase Alfa (Mucolytic Agent)

Door-nose Alfalfa

Pulmozyme is a medication used to thin the mucus, making it easier for the patient to remove by coughing. This treatment is administered using a nebulizer and is effective in reducing lung infections and exacerbations.

Postural Drainage with Percussion

Postural Drainage-of-mucus with Percussion

Chest physical therapy (CPT) involves percussion of the chest wall to promote drainage of mucus out of the lungs. Positive expiratory pressure (PEP) therapy may also be used. With PEP, patients exhale against resistance, which creates positive pressure in the lungs. This pressure allows air to move the mucus toward the large airway where is can be more easily removed.

Fat Soluble Vitamin Supplements

Bacon Viking-ship

Supplements are necessary to correct vitamin deficiencies in patients with CF. Replacement of Vitamin D, in particular, is necessary to prevent conditions such as osteoporosis. It is important to remember that water-miscible forms of the fat soluble vitamins (A, D, E, K) are given because of diminished uptake.

Pancreatic Enzymes with Every Meal

Pancreas Enzymes with Every Meal

Pancreatic enzymes should be taken with every meal to aid in the digestion of food and absorption of nutrients. Forgetting to take these enzymes can result in delayed growth and poor weight gain. Typically, 1-5 enzyme capsules are administered with meals.

High Protein, High Calorie Diet

Up-arrow Mr. Protein & Pancakes

In CF, nutrients from food are not properly absorbed by the body. Many valuable nutrients are excreted, and for this reason, it is recommended that CF patients eat a high calorie, high protein, and high carbohydrate diet. A diet rich in these nutrients will help patients with CF maintain optimal health and weight. On average, these patients should consume nearly 30 to 50 percent more calories than that of a healthy individual.

Added Dietary Salt

Adding Salt to Diet

Patients with CF should add salt to their diet to compensate for the loss of large amounts of salt in their sweat. This is especially true if the patient is in a hot environment, or if he/she is participating in an activity that causes increased perspiration.

Glucose Monitoring

Glue-bottle Monitor

A patient with cystic fibrosis is prone to developing diabetes and for this reason, glucose should be monitored regularly.