Cystic Fibrosis (CF) is a genetic disease that affects approximately 33,500 children and adults in the United States and Canada. CF is caused by a fault in a gene called the cystic fibrosis transmembrane conductance regulator (CFTR) gene. This gene makes a protein that controls the movement of salt and water in and out of cells in the body. In CF, the gene does not work properly and causes the body to produce abnormally thick and sticky mucus and very salty sweat. The thick sticky mucus clogs up the lungs, leading to life-threatening lung infection and breathing difficulties. The mucus can also clog the ducts of the pancreas preventing the digestive enzymes from reaching the intestine where they normally help break down food for absorption. This leads to the development of nutritional problems. Thickened secretions may cause liver problems in patients with CF. Bile secreted by the liver to aid in digestion may not drain adequately leading to liver damage. Over time, this can lead to scarring of the liver.

Common symptoms in patients with CF are acute or persistent respiratory symptoms (difficulty breathing, constant cough with thick mucus), excessive appetite with weight loss, malnutrition, failure to thrive, abnormal stools (greasy and bulky) and intestinal obstruction.

Good nutrition is crucial for people with CF. It begins at birth and continues throughout life. Up to 85% of patients with CF cannot produce the enzymes necessary to digest and absorb fats, proteins and starch (referred to as pancreatic insufficiency). The failure to digest (maldigestion) and absorb food, nutrients and vitamins (malabsorption) leads to poor outcomes associated with poor nutrition in patients with CF.

**Why is nutrition important in CF?**

Most CF patients take pancreatic enzyme replacements. These help patients with pancreatic insufficiency digest and absorb their food better. These enzymes come in small capsules containing many small beads with digestive enzymes. There are many different brands of enzyme. Your CF doctor and nutritionist will help you find the right enzyme and dosage. These enzymes will improve absorption of food and nutrients and help you gain weight. They may improve liver function in those patients who have associated liver disease.

**What are the main nutritional problems in CF?**

- Poor growth or poor weight gain, especially in the pediatric age group.
- Vitamin deficiencies, especially the fat-soluble vitamins A, D, E, K.
- Poor bone health – due to malabsorption of Vitamin D and calcium. This increases one’s risk for broken bones and osteoporosis later in life.
- Cystic Fibrosis – related Diabetes (CFRD)
- Increased salt loss
- Essential fatty acid deficiency

**What can be done to help CF patients digest and absorb their food better?**

Enzymes should be swallowed whole just before meals or snacks. In an infant or small child who can’t swallow capsules, the capsule can be opened and the contents mixed with a small amount of applesauce in a spoon and then given.

**How and when to take the enzymes?**

Enzymes are needed with all meals and snacks. Some foods that just contain simple sugars don’t need pancreatic enzymes. These include fruits, juice, soda, popsicles, Pedialyte, gum, jelly beans, hard candy, tea and coffee (without milk or cream).
Signs of maldigestion/ malabsorption that appear when one is already taking enzymes may indicate that the dose of enzyme needs to be adjusted. Consult with your physician or nutritionist before changing your enzymes since there could be other reasons for your symptoms.

Enzymes work for about one hour after taking them.

No, generic enzymes generally have less activity than brand enzymes and don’t work as well. Do not use generic pancreatic enzymes.

No, pancreatic enzymes are important, but good nutrition is just as critical for CF patients at all stages of their life in order to maintain growth, normal weight and height, and to help keep the body strong to fight infection and optimize liver function.

A balanced high calorie, high protein, high fat diet with added salt is generally recommended.

Many patients with CF need to take about 25-50% more calories and 50% more protein than others their own age.

3 meals and 3 snacks per day are generally recommended.

Supplement meals and snacks with high caloric shakes

It is sometimes impossible to eat all the calories necessary to support proper growth and weight gain to stay healthy. In this case, your physician or dietician may recommend supplemental tube feeds to help you ingest the calories required. Tube feeds are used to supplement calories but not to replace oral feeds. They are often given at night while the patient sleeps. A long thin tube can be passed through the nose (Nasogastric tube) or inserted directly into the stomach (Gastrostomy tube).

Vitamin supplements are crucial for patients with CF, especially supplementation with fat soluble vitamins A, D, E, K which are often poorly absorbed. Even when a patient is on pancreatic enzyme supplementation, vitamin supplements are needed to prevent vitamin shortages.

Different age groups have different nutritional needs. Your CF physician and dietician will help with all your questions and concerns regarding CF and nutrition throughout all phases of your life.

For more information or to locate a pediatric gastroenterologist in your area please visit our website at: www.naspghan.org

Additional information on CF can be obtained from the websites listed below.

www.cff.org.
www.ccff.ca/

IMPORTANT REMINDER: This information from the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) is intended only to provide general information and not as a definitive basis for diagnosis or treatment in any particular case. It is very important that you consult your doctor about your specific condition.