GI Kids
Help & Hope for Children with Digestive Disorders
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Cystic Fibrosis and Nutrition

Cystic Fibrosis (CF) is a genetic disease that causes the body to produce a thick mucus. This mucus leads to respiratory symptoms and infections.

Many CF patients lack the enzymes needed to digest and absorb fats. Therefore, good nutrition is important for CF patients to stay healthy. To learn more about how nutrition plays a role in treating CF in children, download the GIKids Fact Sheet on Cystic Fibrosis & Nutrition.

What is Cystic Fibrosis?

CF is a genetic disease that affects approximately 35,000 children and adults in the United States and Canada.

CF is caused by a faulty gene called the cystic fibrosis transmembrane conductance regulator (CFTR). This gene makes a protein that controls how salt and water move in and out of cells in the body.

In CF, the gene does not work properly and causes the body to produce very thick and sticky mucus and very salty sweat. The mucus clogs up the lungs, leading to life-threatening lung infection and breathing difficulties.

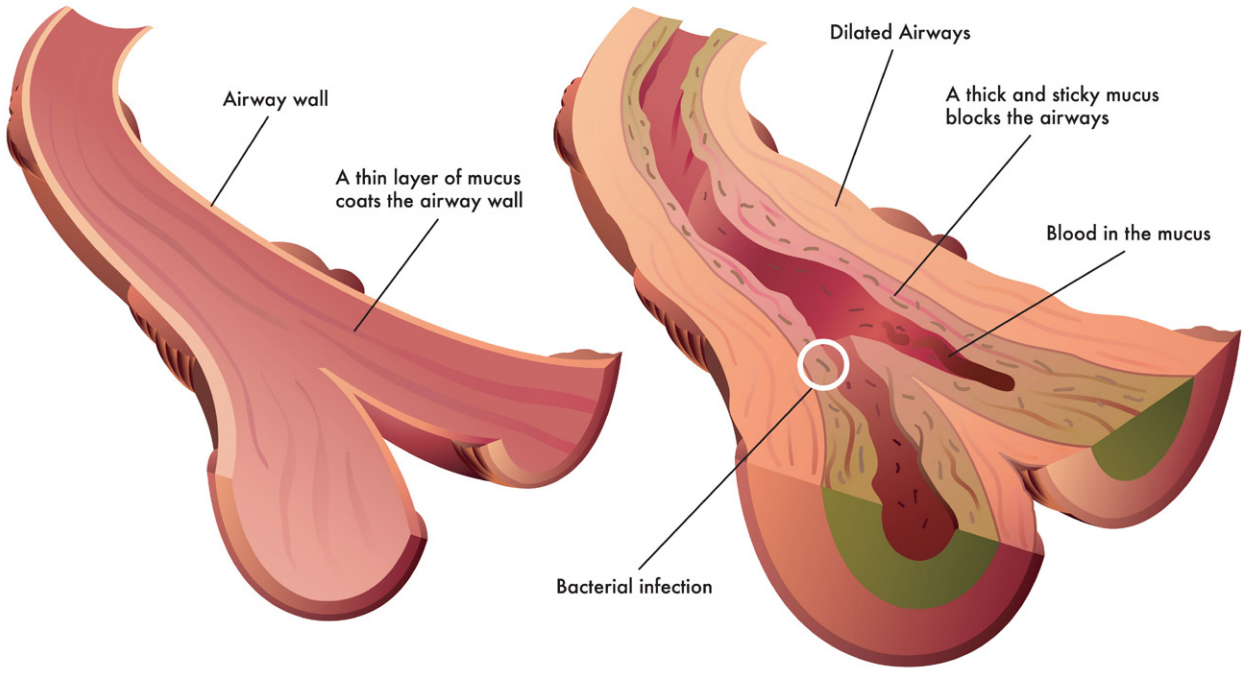
The mucus can also clog the ducts of the pancreas. This prevents digestive enzymes from reaching the intestine, where they normally help break down food for absorption. This leads to nutritional problems and poor growth.

Thickened secretions may also cause liver problems in patients with CF. Bile secreted by the liver to aid digestion may not drain adequately, leading to liver damage. Over time, this can scar 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.

How is Cystic Fibrosis diagnosed?

CF can be diagnosed many ways. It is often a combination of a complete clinical evaluation, genetic testing and a sweat chloride test. Most children are screened for CF at birth through the new born screening test. Genetic tests can be done either during pregnancy or after birth to diagnose CF.



Normal Airways

Airways affected by cystic fibrosis

Newborn screening and genetic testing are good tools, but sometimes they result in either positive test results in patients who do not have CF (false positive) or negative test results in patients who do have CF (false negative).

The sweat chloride test is a safe, painless and effective way to diagnose CF. This test measures the amount of a chemical element called chloride in the sweat. Patients with CF have high levels of chloride in their sweat compared to those without CF.

Why is nutrition important in Cystic Fibrosis?

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 needed to digest and absorb fats, proteins, and starch (called pancreatic insufficiency). The failure to digest (maldigestion) and absorb (malabsorption) food, nutrients, and vitamins leads to poor outcomes in patients with CF.

Symptoms of maldigestion and malabsorption may include:

- Poor weight gain despite a good appetite
- Abdominal pain, gas, and bloating
- Frequent loose, foul-smelling stools

Is maldigestion/malabsorption the only reason why Cystic Fibrosis patients have problems with nutrition?

Maldigestion and malabsorption are the most important reasons but not the only reasons. Patients with CF require more calories than other patients. This is usually because of increased energy demands due to coughing, increased work to breathe due to chest problems, and repeated infections. Because of these reasons, the body needs more calories to breathe and fight infections.

What are the main nutritional problems in Cystic Fibrosis?

- Poor growth or poor weight gain, especially in children
- Vitamin deficiencies, especially fat-soluble vitamins A, D, E, and K
- Poor bone health, due to malabsorption of vitamin D and calcium (this increases the risk for broken bones and osteoporosis later in life)
- CF-related diabetes (CFRD)
- Increased salt loss
- Essential fatty acid deficiency

What can be done to help Cystic Fibrosis patients digest and absorb food better?

Most CF patients take pancreatic enzyme replacements that assist in the digestion of carbohydrates, fats and proteins. These enzyme supplements are taken with every meal and snack to help patients with pancreatic insufficiency digest and absorb their food better. These enzymes come in small capsules containing beads with digestive enzymes.

There are many different brands of enzyme. Your doctor and nutritionist can help find the right enzyme and dosage for your child. These enzymes improve absorption of food and nutrients and help your child gain weight. They may improve liver function in CF patients who have associated liver disease.

How and when are the enzymes taken?

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. The capsule's contents can then be mixed with a small amount of applesauce in a spoon and fed to the child. The pill should not be crushed, chewed or taken with hot drinks as this might make it less effective.

Are enzymes required with all meals and snacks?

Enzymes are needed with all meals and snacks, milk, formula, and breast milk that contain fat, protein, and starch. Some foods that contain only simple sugars don't need pancreatic enzymes, including fruits, juice, soda, popsicles, Pedialyte, gum, jelly beans, hard candy, tea, and coffee (without milk or cream).

Can there be signs of maldigestion/ malabsorption even when taking enzymes?

Signs of maldigestion/malabsorption that appear when taking enzymes may indicate that the dose needs to be adjusted. Talk to your doctor or nutritionist before changing enzymes, since there could be other reasons for your child's symptoms.

How long do enzymes work?

Enzymes work for about one hour after taking them.

Are generic enzymes just as good as brand-name enzymes?

No. Generic enzymes generally have less activity than brand-name enzymes and don't work as well. Do not use generic enzymes.

Are enzymes all that is necessary to improve nutritional status of Cystic Fibrosis patients?

No. Enzymes are important, but good nutrition also is critical for CF patients at all stages of life. Good nutrition helps maintain normal growth, weight, and height. Good nutrition also helps keep the body strong to fight infection and maintain healthy functioning of many important organs including the lungs, pancreas and liver.

What is the recommended diet for patients with Cystic Fibrosis?

- A balanced high-calorie, high-protein, high-fat diet with added salt is generally recommended.
- Many patients with CF need to intake about 25%–50% more calories and 50% more protein than others of the same age.
- Three meals and three snacks per day are generally recommended.
- Meals and snacks can be supplemented with high-calorie shakes.
- Do all patients gain weight with a high-calorie, high-fat diet?

It is sometimes impossible to eat all the calories needed for proper growth and weight gain to stay healthy. In this case, your physician or dietitian may recommend supplemental tube feeds to provide required calories to your child.

Tube feeds are used to supplement calories but do not replace eating. They are often given at night while your child sleeps. This is usually either through a long thin tube passed through the nose (nasogastric or NG tube) or inserted directly into the stomach (gastrostomy tube or G tube).

Are vitamin supplements necessary?

Vitamin supplements are crucial for patients with CF, especially fat-soluble vitamins A, D, E, and K because they are often poorly absorbed. Even when a CF patient is taking enzymes, supplements are needed to prevent vitamin shortages.

Is nutrition the same at all ages?

Different age groups have different nutritional needs. Your doctor and dietician will help answer questions and concerns regarding CF and nutrition throughout all phases of your child's life.

What are the effects of Cystic Fibrosis on the liver?

Treatment of the respiratory complications of CF has improved along with life expectancy. Now, CF-related liver disease is increasingly important and is the third-leading cause of death in CF patients.

Patients usually have signs of obstruction of the bile ducts. This can progress to inflammation and scar the liver.

Liver injury symptoms include malnutrition and fat-soluble vitamin (A, D, E, and K) deficiency, yellow discoloration of the eyes and skin (jaundice), enlargement of the liver or spleen, and development of gallstones. In advanced cases, patients can develop liver scarring (cirrhosis), which can progress to liver failure and require transplantation.

Quick facts:

- CF is a genetic disease that disrupts how salt and water move in and out of cells. This leads to development of thick mucus and secretions, which can cause problems in many areas of the body.
- Typical symptoms of CF are frequent respiratory infections and difficulty breathing; poor digestion and absorption of food, leading to poor weight gain and loose stools; liver disease; and pancreatic problems, including insufficient digestive enzymes and diabetes.
- A pediatric gastroenterologist and nutritionist can be very important to help manage nutrition and avoid calorie and nutrient deficiencies with CF.

Links

Cystic Fibrosis Foundation (USA)

Cystic Fibrosis (Canada)

MedicinePlus

Locate a Pediatric Gastroenterologist

IMPORTANT REMINDER: *This information from the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) is intended only to provide general educational information 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.*

NASPGHAN
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