



OTHER CF DIGESTIVE SYSTEM PROBLEMS

Sometimes people with **cystic fibrosis*** (CF*) can have several **digestive system*** problems beyond **malabsorption***. (See the CF FEP module *Managing Nutrition and Digestive Problems* for more information on malabsorption.) “OTHER CF DIGESTIVE SYSTEM PROBLEMS” gives you an overview of different types of digestive problems. Your CF health care team can help you watch for other digestive or **gastrointestinal*** problems in your child. If your child develops one of these problems, the CF health care team will give you more detailed information.

MECONIUM ILEUS

Meconium* is the name for the first **bowel movements*** that a baby has after birth. A baby who has **meconium ileus*** has a **blockage*** in the **intestines*** from thick, abnormal meconium. The blockage prevents the baby from passing a stool. Meconium ileus is usually caused by malabsorption. Meconium ileus starts even before the baby is born. The **small intestine*** can rupture (also called **perforation***), which can be very serious. Not only is the small intestine blocked, but the **colon*** or **large intestine*** also may be smaller than normal. Infants with meconium ileus will often need surgery soon after birth.

CONSTIPATION

Constipation* means a person does not have stools or bowel movements regularly. Most people have at least one stool a day. A person who is constipated may

not have a bowel movement for several days. Abnormal stool or reduced motion of the muscles in the walls of the intestines can cause constipation. People can get constipated from not drinking enough fluids or not getting enough fiber in their diets. Children with CF may have constipation more often than other children.

WATCH AND DISCOVER †

Constipation Problems

Watch for **signs*** and **symptoms*** of constipation in your child.

- Fewer bowel movements than normal—usually less than once a day
- Harder stools than usual
- Stomach pain or cramps
- Decreased amount of stool compared to that in the past
- A feeling of fullness in **abdomen*** (or feeling full or not eating because of a distended or bloated belly)

A person with constipation can have stools that look like **diarrhea***. This is not really diarrhea but stools that are more liquid that are leaking around a stool blockage.

THINK AND ACT †

Managing Constipation

Talk to your doctor if you think your child is constipated. Your doctor may recommend medicines that will help soften the stool and move it through the **bowels*** (intestines). Some people with CF need to use **laxatives*** or stool softeners regularly for constipation. Other ways to help

prevent constipation are to have your child:

- Drink enough fluids (at least 4 to 8 cups or 32 to 64 ounces per day).
- Eat a high-fiber diet (your CF dietitian can give you tips on high-fiber foods, such as raw vegetables; fruit with skin, including pears and prunes; and whole wheat or high-fiber bread).
- Use fiber such as psyllium (one brand name is Metamucil®). Talk with your child’s doctor or dietitian to see if psyllium might help. Drink at least 2 cups of water with the psyllium.
- Get regular exercise. Exercise can help move stool through the intestines. (See “EXERCISE AND CF” in Appendix 3.)

DISTAL INTESTINAL OBSTRUCTION SYNDROME

Distal intestinal obstruction syndrome (DIOS)* is a more severe form of constipation and is caused by a buildup of stool and **mucus*** in the intestines. Children with CF can have DIOS with malabsorption or with **dehydration***. Sometimes DIOS can happen when children are ill with a **respiratory infection*** and are less active. Distal intestinal obstruction syndrome is also called **meconium ileus equivalent*** because it is similar to meconium ileus.

Your child’s doctor will treat DIOS by giving your child laxatives and fluids to flush out the blockage. Some children need surgery if the intestine ruptures or cannot be cleared. You can help prevent

† See the CF FEP module *Becoming a CF Manager* to learn more about Watch and Discover and Think and Act.

*See CF Words to Know Glossary.



DIOS by managing malabsorption and taking proper steps to prevent constipation. Some children will benefit from taking laxatives or stool softeners daily.

INTUSSUSCEPTION

Intussusception* is another problem that can cause a blockage in the intestine. Intussusception is when one part of the intestine collapses or telescopes into another part. Most often the last part of the small intestine (the **ileum***) slides into the colon. Children with CF are at greater risk of intussusception than children without CF. Intussusception is not a common problem and is usually seen in very young children during their first few years of life.

Intussusception can happen as a **complication*** of a **viral infection***, malabsorption, or DIOS. Symptoms include severe cramps or abdominal pain (the pain may come and go), vomiting, and decreased number of stools. Blood may be seen in the stool. Intussusception is treated by giving a special **enema*** done with **X-ray*** imaging. If this does not relieve the blockage, the child may need surgery.

GASTROESOPHAGEAL REFLEX (GER)

Gastroesophageal reflux* (also called GER or GE reflux) is a condition in which stomach acid and partially digested food can flow from the stomach back up into the **esophagus*** (the tube from the throat to the stomach). You may also hear this called heartburn (or **acid reflux***). If acid reflux happens often it can cause problems. Many infants have GER, but can outgrow it. Gastroesophageal reflux can appear at any age. In CF, coughing can make reflux worse. Acid reflux

happens more at night when a person is lying down or when a person's stomach is full.

WATCH AND DISCOVER

Symptoms of GER

Watch for these symptoms of GER:

- Repeated vomiting or spitting up
- Frequent burping
- Stomachaches or heartburn
- Decreased appetite or feeling full after eating only a small amount
- Slow or no weight gain from not wanting to eat, or losing food and nutrients with vomiting

Gastroesophageal reflux can also sometimes lead to coughing and **wheezing***. Some children may have only one symptom of GER; other children may have several.

THINK AND ACT

Managing GER
If you think your child may have GER, talk with your doctor. With your doctor, you can decide if your child needs tests or medicine for GER. Several medicines are used to treat GER, including acid blockers to reduce stomach acid so it does not irritate the esophagus, and other medicines to help the stomach empty better. Besides giving medicines, you can help control reflux by having your child:

- Eat smaller amounts more often—your CF dietitian can help you plan small meals that are high in **calories***.
- Avoid spicy foods and foods that have a lot of acid such as tomatoes, citrus fruits, or fruit juices.
- Avoid carbonated drinks such as sodas, and drinks with caffeine (colas, tea, and coffee).
- Do **airway clearance*** before eating.

- Avoid lying down right after eating a meal (eat the evening meal at least 3 hours before bedtime).
- Play quietly for 30 minutes after eating.
- Sleep with the head of his or her bed raised 6 to 8 inches, especially if problems with acid reflux continue at night.

IMPAIRED GLUCOSE TOLERANCE AND CF-RELATED DIABETES (CFRD)

Carbohydrates* from the food we eat are broken down into **glucose*** for the body to use as energy. Glucose enters the blood and the **blood glucose*** (sugar) level goes up. A rise in blood glucose level sends a signal to the **pancreas*** to release **insulin*** into the blood. Insulin is a **hormone*** that helps lower blood glucose levels by moving glucose into the **cells*** to be changed into energy. Insulin also plays a role in how cells in the body use fat and protein.

Sometimes a person with CF can have too little or no insulin to move glucose into the cells for energy. In fact, over time the pancreas of some people with CF may actually stop making insulin. Blood glucose problems in people with CF can vary. Many older children and adults with CF will have some problems with high blood glucose levels, and 15 percent or more will have more severe glucose problems. The problem may be worse when a person has an infection or is taking certain medicines, such as **corticosteroids***. Blood glucose problems can make it harder for people with CF to gain weight and also make them more prone to lung infections.



When a person has a mild problem with blood glucose levels, it is called **impaired glucose tolerance**.* People with impaired glucose tolerance may have symptoms of high or low **blood sugar*** levels at times or may have no obvious signs or symptoms.

Impaired glucose tolerance can lead to a more serious problem called **CF-related diabetes (CFRD)**.* CFRD is treated with insulin.

WATCH AND DISCOVER Symptoms of Impaired Glucose Tolerance and CFRD

- Weight loss or problems gaining weight
- Tiredness
- Unexplained worsening of **lung function***
- Lung infections more often
- Increased thirst and frequent drinking, which leads to the person **urinating*** often

The doctor uses blood tests to diagnose impaired glucose tolerance and CFRD.

- 1) **Blood glucose level:** The level of glucose can be measured in the blood. This often is done while fasting (having not eaten for at least 12 hours) in the early morning. If this level is too high, other blood tests need to be done.
- 2) **Oral glucose tolerance test***: Looks at how a person's blood glucose level changes after drinking a certain amount of glucose. For an oral glucose tolerance test (OGTT for short), the person must fast (not eat for at least 12 hours) before the test. If blood glucose levels are too high, the body may have a problem handling glucose and

it could mean the person has CFRD. There are recommended guidelines when annual glucose tolerance testing should start in children with CF.

THINK AND ACT Managing Impaired Glucose Tolerance and CFRD

If you see symptoms that suggest your child may have blood glucose problems, Act and talk with your CF health care team about having your child tested. If your child is diagnosed with either impaired glucose tolerance or CFRD, you will work closely with your CF health care team and/or a diabetes doctor (**endocrinologist*** or pediatric endocrinologist) and learn how to manage it. This may include (1) learning how to check blood glucose levels at home, (2) closely watching how many carbohydrates your child eats, and (3) learning how to measure and give insulin.

High Blood Glucose Caused by Medication or Infection

People with CF can have problems with high blood glucose levels when they are sick or are taking certain medications. In these cases, the person will have to watch blood glucose levels and the CF health care team will decide if insulin is needed. If insulin is needed, it may only be for a short time. The CF health care team will help figure out what is causing the high blood glucose levels and what the best treatment will be to get levels back to normal.

LIVER AND GALLBLADDER DISEASE

Some people with CF can have problems with the **liver*** or **gallbladder**.* The liver is an organ that sits in the upper right side of the abdomen. The gallbladder is

attached to the liver and helps store extra **bile*** made by the liver. The liver is important in digestion. It stores **nutrients*** for the body, helps break down medicines, and filters the blood.

Liver disease in people with CF varies from one person to the next. Many people with CF can have some fat buildup in the liver. The liver can become larger and **liver function test*** levels may go up. This happens most often to people who have malabsorption and poor nutrition. Another problem with the liver is a buildup of abnormal bile. In CF, bile can be too thick and does not flow well through the liver and gallbladder to the small intestine. **Gallstones*** can develop in the gallbladder with the abnormal bile and may cause problems. The much more serious problem, however, is that while the bile builds up, the liver is damaged. This liver damage is called **cirrhosis**.* **Chronic*** infections (such as hepatitis C) or alcohol abuse can also cause cirrhosis. Like the lungs, the liver can continue to function even though it is damaged; however, over time it can fail.

Cirrhosis in CF can also lead to high blood pressure in the veins connected to the liver (called **portal hypertension***), enlargement of the **spleen***, and **blood clotting*** problems. Portal hypertension can cause veins around the esophagus to swell and weaken (called **esophageal varices***). These veins can bleed into the esophagus. Between 15 and 30 percent of people with CF can have cirrhosis.

*See CF Words to Know Glossary.



WATCH AND DISCOVER



CF-Related Liver Problems

People with CF may not know their liver is having problems because they have little or no symptoms. Blood tests help detect liver problems. The CF Foundation recommends that people with CF have liver function tests once a year. The tests are the best way to find a problem early so treatment can be started before there are symptoms of liver damage. If liver disease or gallstones are suspected, the doctor will do other tests to find the liver problem.

Signs and Symptoms of Liver Disease

- Liver and spleen are larger than normal
- Indigestion and stomachaches (from gallstones or fat malabsorption)
- Easy bruising (from blood clotting problems)
- Vomiting up blood (from esophageal varices)
- Poor weight gain or weight loss

THINK AND ACT



Managing CF-Related Liver Problems

Although there is no cure for liver disease, treatments are important to help make sure a person with CF has good nutrition and his or her malabsorption is well managed. A medicine called ursodiol (or ursodeoxycholic acid, brand name Actigall®) helps with bile flow and may help with gallstones. If a person is having a lot of symptoms from gallstones, he or she may have the gallbladder removed during surgery.

CF-RELATED BONE HEALTH

Our bones depend on good nutrition to develop properly and remain strong. Vitamin D and **calcium*** are important to build and maintain bones. Regular

exercise also helps bones stay healthy. In CF, sometimes the bones don't have enough **minerals*** so they are less dense and can become weak and break more easily. Older women may have this condition called **osteoporosis***. Osteoporosis is also seen most often in adults with CF and in people who have severe nutrition and/or lung problems. Good management of CF can help prevent bone health problems in the future.

Some medicines can also affect the density and strength of a person's bones. Talk with your CF health care team about medicines and supplements your child is taking. The CF health care team can help you watch for early signs of bone health problems or things that may put your child at risk for future bone problems.

WATCH AND DISCOVER



CF-Related Bone Health Problems

Bone problems happen very slowly and often a person does not notice any symptoms (unless they are severe) until he or she has a break or fracture. Fractures hurt. If a person breaks a rib, it can be hard to breathe and cough, and airway clearance may be painful.

Osteoporosis Tests

Vitamin D and calcium blood levels are checked every year—and more often if a person has low levels or has problems with **malnutrition***.

The CF health care team may notice a change in bones on a chest X-ray, especially if the person's **bone density*** is low. Bone density can be measured with equipment that takes a special kind of X-ray. The CF Foundation recommends all people with CF have a bone density study, such as a **DEXA scan*** (dual-energy X-ray absorptiometry) by 18

years old. Low bone density often means a person is at higher risk for broken bones.

THINK AND ACT



CF-Related Bone Health

As your child grows, you can do several things to help keep his or her bones strong:

- Make sure your child gets regular exercise that can help keep bones strong. Walking, running, and weight lifting are great exercises to help keep bones strong and healthy. Exercise early in life can build bone density that will help in the future.
- Have your child eat a balanced diet, including enough **vitamins*** and minerals vital to good bone health. Alcohol and caffeine lower bone density and should be avoided. Have your child drink whole milk and other products that contain calcium, vitamin D, and calories.
- Make sure your child avoids smoking and **secondhand smoke exposure***. Tobacco smoke hurts the bones and lungs.
- Help your child keep his or her lungs healthy and avoid lung infections. Healthy lungs will contribute to healthy bones as well.

The CF health care team may prescribe calcium and/or vitamin D if your child's levels are low. The team may also prescribe some medicines that can help strengthen bones. Decide with your CF doctor if medicine would be helpful for your child.

To learn more, you can get a copy of "Bone Health and Cystic Fibrosis" (including a list of foods high in calcium) from the CF Foundation's Web site: <http://www.cff.org> or call 1-800-FIGHT-CF.