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# **Adult Guide to Cystic Fibrosis**

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# INTRODUCTION

Welcome to the Adult Guide to Cystic Fibrosis — whether you're an adult, a teen preparing to transition to an Adult CF Program, a parent of a child looking ahead, or a friend or relative of someone with CF, you'll find answers and insights here.

We asked medical experts and others living with CF to share the very best information, then gathered more from adult CF programs and the Cystic Fibrosis Foundation. The result: pages packed with answers, tips, tricks, and tons of resources related to your health and well-being.

But we didn't stop there. Being an adult with CF brings on some pretty distinct challenges. Relationships, travel, and social support — we cover all these topics too.

Let us know how helpful you found this resource and if you have ideas on how to make it more useful. Email [info@cff.org](mailto:info@cff.org) or complete this short [survey](#).

The Adult Guide is an easy reference for you to browse topics of interest. The information is intended to help you manage cystic fibrosis in partnership with your **CF care team**. It is not intended to replace treatment advice from medical professionals.

If you have questions about anything you have read here, your CF care team can answer those questions or discuss any concerns specific to your individual health care needs.

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# Basics of Lung Care

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Although cystic fibrosis affects many parts of the body, for most people with CF it's the lung disease that causes the most problems. Sometimes it's hard to give our lungs a lot of love, especially when they may seem more like mucus makers than the body's essential gear for breathing.

But with the right kind of attention from you — and the help of medications and your CF care team — your lungs can do the vital work they were meant to do. Giving you the facts, tips, and resources to make that happen is what this guide to lung care is all about.

## IN THIS SECTION:

- ▶ **Learn about your lungs** to help you keep them clear.
- ▶ **Work with your CF team** to keep your lung function as high as possible.
- ▶ **Use the right therapies** and airway clearance techniques so you can breathe easier.
- ▶ **Manage your time** so you can do all the other things you need or want to do.
- ▶ **Get help when you are sick** — it can save your life, especially when you have exacerbations.

## Basics of Lung Care

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### LEARN ABOUT YOUR LUNGS

Cystic fibrosis is caused by abnormalities in a protein called CFTR (cystic fibrosis transmembrane conductance regulator). This protein normally ends up on the surface of cells in the lungs — and in the pancreas and other organs — where it helps with the basic “housecleaning” of the cells.

When CFTR works correctly it helps move chloride, water, and other small molecules out of the cells and onto the surface of the airways.

But with CF, this protein doesn’t get to the cell surface, or it’s there but doesn’t function properly. The chloride, water, and other molecules do not move into the airways and the cells cannot do their normal microscopic housecleaning. Without the body’s typical cleaning process, the airways get into a cycle of inflammation (swelling) and infection. This causes mucus and bacteria to build up in the lungs of people with CF.

Monitoring and minimizing this buildup of mucus and bacteria are two key jobs of your CF care team. For more information, read [Work With Your CF Team](#).

### BEYOND THE BASICS

The key protein in CF is CFTR. The genetic “blueprint” for CFTR, and for all proteins in the body, lies in the DNA code that determines our body’s basic makeup. In people with CF, the instructions for the CF gene have been altered, creating a defective gene. That in turn produces a faulty CFTR protein.



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**The relationship between a person’s CF mutation (“genotype”) and the way that the disease affects that person’s body (“phenotype”) is complex.**

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More than 2,000 mutations in the CF gene have been identified. The relationship between a person’s CF mutation (“genotype”) and the way that the disease affects that person’s body (“phenotype”) is complex.

In some mutations, no CFTR protein is made at all. In others, CFTR is made but then gets “stuck” and doesn’t move from the center of the cell to the surface. Some mutations make the protein and it gets to the cell surface, but it works too weakly to do its job.

## Basics of Lung Care

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Different therapies may be useful for different cystic fibrosis mutations. If you have not been “genotyped” or tested to find out which CF mutation or mutations you have, talk about this with your CF care team.

### WORK WITH YOUR CF TEAM

Monitoring and minimizing the buildup of mucus and bacteria in your lungs are two of the key jobs of your CF care team at your care center.

When you come for a check-up, your CF care team will likely have you take pulmonary function tests (PFTs). They will ask you about your breathing, coughing, and the amount of sputum you are making. They’ll also ask for a sample of your sputum, which is tested to see what kinds of bacteria are growing and which antibiotics might fight them. This is called a “sputum culture.”



Center for Cystic Fibrosis at Keck Hospital of USC Keck Medical Center of USC

Your PFT results will probably look like a sea of numbers and maybe a line graph. The most important PFT is the FEV<sub>1</sub>, which helps to measure lung function. Your FEV<sub>1</sub> is the amount of air you can blow out in the first second of a forceful exhalation. Your care team may seem totally obsessed with keeping your FEV<sub>1</sub> as high as possible. Well, that’s their job.

Keeping your FEV<sub>1</sub> as high as possible is also the job of your meds or pulmonary therapies. See [Use the Right Therapies](#) for more detail on current therapies, as well as some potential new ones.

### ADDITIONAL RESOURCES

- › [Learn](#) how adults with CF can be full members of their CF care teams.

### USE THE RIGHT THERAPIES

Right now, there are three categories of pulmonary therapies for CF. One helps clear sputum out of your lungs; another helps control bacteria; and the third reduces inflammation.

## Basics of Lung Care

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### CLEARING OUT SPUTUM

A powerful cough is your lungs' way of getting rid of sputum. The less sputum, the less your airways get plugged, the fewer bacteria grow, and the better you'll breathe. But your regular cough often needs a boost, since CF sputum is thick and sticky and there's usually a lot of it.

Some therapies "jiggle" the sputum, making it easier to cough out. These are called airway clearance techniques (ACTs). They include manual chest physiotherapy (CPT), when another person claps on your chest with their hands; vibrating devices and vests; devices that create a fluttering pressure as you blow through them; and good old-fashioned exercise. All of these ACTs work like hitting a ketchup bottle to get ketchup out.

Of course, you can also thin ketchup. Other clearing therapies thin the sputum so it's easier to cough out. They include DNase (Pulmozyme®) and hypertonic saline. These meds work by different means, so they cannot replace one another or be considered as alternatives. A person can use both therapies. Because they have to be inhaled using a nebulizer, both take a lot of time.

### CONTROLLING BACTERIA

To minimize the bacteria in your lungs, you may be inhaling an antibiotic, such as tobramycin (TOBI®) or aztreonam (Cayston®). They are usually inhaled with a nebulizer, so they may be time-consuming. Newer antibiotics and better ways to deliver them are being developed.

### REDUCING INFLAMMATION

Some medications seem to decrease the inflammation in the lungs. When the lungs are less inflamed, they make less sputum. Some children with CF have taken ibuprofen (such as Advil®) to reduce inflammation in the airways. But ibuprofen therapy has not been shown to help the lungs of adults with CF. If your pediatric CF Care Team gave you ibuprofen, talk with your adult CF Care Team about it and about any side effects you may be having.

The antibiotic azithromycin seems to reduce inflammation and boost lung function (FEV<sub>1</sub>). Oddly, it is not the antibiotic effect but some other factor that appears to reduce inflammation.



## Basics of Lung Care

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### ASTHMA

Asthma can be a kissing cousin to CF. Some people seem to have both, so you may use or need to use asthma inhalers and oral medication.

At times, you might need to take a course of corticosteroids if your asthma suddenly gets worse. These meds help reduce inflammation. They are usually taken by mouth. Prednisone is the most commonly prescribed.

These corticosteroids are different than the steroids that some athletes have used illegally. They are generally safe in the short run. But if taken for more than a couple of weeks, they may have serious side effects. Speak with your doctor about the possible side effects of corticosteroids.

### ADDITIONAL RESOURCES

- › **Exercise regularly:** Jog, walk, ride a bike, swim, jump rope, use a trampoline, stretch, do yoga.
- › **Avoid germs:** Wash your hands often, stay at least 6 feet away from anyone who is ill, and get a yearly flu vaccine.
- › **Eat well-balanced meals:** If you use enzyme supplements, take them!
- › **Avoid tobacco smoke:** Do not smoke and keep away from secondhand smoke.
- › Read more about [ACTs](#).
- › Find more information in the CF Foundation's in-depth [respiratory care guidelines](#).

## Basics of Lung Care

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### MANAGE YOUR TIME

Let's face it: CF care takes up a lot of time, especially the inhaled medications. But they work. The key is finding the best ways to make the time for your CF care. We've got some ideas:

- › **Plan ahead.** If an upcoming event or outing is likely to conflict with your medication schedule, make time to do your treatments before or after the event. Get a converter so you can use your nebulizer in the car.
- › **Stay organized.** Put a daily reminder in your phone or on your calendar so you don't forget when to take your meds.
- › **Multi-task.** Use the time when you are doing your treatments to catch up on email or the news, check in with friends on Facebook, browse a magazine, or make a grocery list.
- › **Make it a habit.** Clean and disinfect your nebulizer at the same time every day so it becomes a part of your routine.

### AT SCHOOL OR WORK

At times, you might feel you are bothering those around you — maybe when you're coughing, using an inhaler, or doing a form of airway clearance. But you should never feel uneasy about doing what you need to do to take care of yourself.

Let your teachers or supervisors know you have special health needs, and then set up a plan to make sure that you have the time and space to do what you need to do.

Keep in mind: you are protected legally against any school or workplace discrimination related to having CF.

### ADDITIONAL RESOURCES

- › Learn about [CF and School](#).
- › Watch [Patient Advocacy: Issues and Answers](#), a Virtual CF Education Day webcast.

# Basics of Lung Care

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## GET HELP WHEN YOU ARE SICK

Smart people know when to ask for help. With CF, you need to call your CF care team when you have an exacerbation.

### WHAT ARE THE SIGNS OF AN EXACERBATION?

- › More sputum and coughing, and maybe some blood in your sputum.
- › More shortness of breath than usual.
- › Fevers and loss of appetite.
- › Falling lung function and decreasing weight.

### TREATMENT OF EXACERBATIONS

During an exacerbation, you may be asked to take a course of **oral antibiotics** or **intravenous (IV) antibiotics** and to increase your overall therapy, including **airway clearance**.

If you had an exacerbation as a child, you know the routine. If it is your first one, you might find it upsetting. Remember that treating these exacerbations quickly and aggressively results in better maintenance of your overall lung health. Work with your CF care team to make a treatment plan.

Patients whose lung function (or FEV<sub>1</sub>) has become very low may find it harder and harder to do things that used to be easy. You may require more “routine” therapy at home and may need more frequent treatment of exacerbations. You may be given extra oxygen to ensure your body gets what it needs.

This may sound very frightening; that is a perfectly normal reaction. Share your fears and concerns with members of your care team or with a psychologist. They will have worked with other people who have had similar experiences and will be able to help you make sense of your feelings. Ask at your care center whether there is a support group for people who share your concerns.

Keep in mind that new therapies are always being explored and developed to treat CF. They will make a difference!

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# Sinusitis and CF

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Cystic fibrosis can affect many parts of the body. The involvement in the lungs is usually of greatest concern, but it can also affect the sinuses. The sinuses are air-filled spaces in the skull around the face and the eyes. They are lined with the same kind of cells that are found in the lung. In fact, the sinuses are considered part of the upper respiratory tract.

Just as the lining of the lungs can become chronically inflamed in CF, so can the lining of the sinuses. As a result, similar processes occur in the sinuses that can lead to significant and long-term problems. Some of these issues can be severe and require a lot of time and attention. But even mild degrees of sinus disease can cause discomfort. Some aspects of sinus care are simple, but others can require a major time investment.

Some people with CF will have minimal sinus issues, whereas others might suffer from more problems. Many treatment options are available to make dealing with the sinus issues more bearable. With awareness and attention, you and your CF team can help minimize the effects the sinus issues may have on your body and your lifestyle.

## Sinusitis and CF

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### IN THIS SECTION:

- ▶ **Learn about your sinuses** to understand what they are and how they can cause problems.
- ▶ **Work with your CF team** to evaluate your sinuses and treat symptoms when they arise.
- ▶ **Use the right therapies** to decrease the frequency, duration, and severity of infections, and avoid potential setbacks.

### LEARN ABOUT YOUR SINUSES

CF is caused by a missing or abnormal protein that normally sits on the surface of cells in the lungs (and other parts of the body — the pancreas, the bile ducts of the liver, the sweat glands). It turns out the sinuses, which are part of the respiratory tract, have the same absent or abnormal protein as the lungs. So it's no surprise that many people with CF have sinus problems.

The sinuses consist of air-filled spaces in the face and skull. They have special names: maxillary (cheek), frontal (forehead), ethmoid (between the eyes), and sphenoid (behind the eyes) sinuses. The exact function of the sinuses is not completely known, but they are thought to lighten the skull and protect the eyes and brain from injury. Imagine how heavy your head would be if it was one large block of bone! Your neck would be really sore!

The lining of the sinuses constantly produces mucus. In the normal state, this mucus provides some protection from the external environment. The mucus is swept through small pathways from the sinuses into the nose and throat, where it is generally swallowed or coughed out. When the CFTR protein is not functioning well, mucus becomes thick and dry, and cannot be swept away. This contributes to chronic inflammation and creates an environment in which bacteria and viruses can grow, causing inflammation and infections.

Symptoms of sinus infection and inflammation include discolored nasal discharge, nasal obstruction or blockage, nasal congestion, pressure/pain/headache, decreased sense of smell, and postnasal drip. The term "sinusitis" refers to an infection in the sinuses. In addition to the symptoms noted above, it can also cause fatigue, ear fullness, and cough.

## Sinusitis and CF

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### BEYOND THE BASICS

Nearly all people with CF have some degree of inflammation in the sinuses, although the amount varies. It is unclear why, but some people will not have noticeable sinus problems or will develop them much later in life. Those people with CF who have sinus symptoms in childhood often have more severe sinus disease as adults and tend to require more sinus attention. Over 50 percent of people with CF are thought to have nasal polyps, which are swollen, fluid-filled tissues that grow from the sinuses and can block the nasal passages. Many people with CF have recurrent sinus infections, and may grow the same bacteria as is often found in the lungs — staph or *Pseudomonas*. Research suggests that during infections, the same bacteria found in the sinuses may often be found in the lungs.

### ADDITIONAL RESOURCES

**Sinus Disease in Cystic Fibrosis Webcast:** [www.youtube.com/watch?](http://www.youtube.com/watch?)

[www.entnet.org/healthinformation/noseandmouth.cfm](http://www.entnet.org/healthinformation/noseandmouth.cfm)

[www.american-rhinologic.org/](http://www.american-rhinologic.org/)

[emedicine.medscape.com/article/232791-overview](http://emedicine.medscape.com/article/232791-overview)

[emedicine.medscape.com/article/862538-overview](http://emedicine.medscape.com/article/862538-overview)

[www.uptodate.com/contents/patient-information-chronic-sinusitis](http://www.uptodate.com/contents/patient-information-chronic-sinusitis)

### WORK WITH YOUR CF TEAM

Stay aware of any changes in your nose and your sinuses — and let your care team know about these changes. Often it is difficult to know if cough, drainage, congestion, and shortness of breath are related to the sinuses or the lungs, or both. Headache can also be a difficult issue to sort out, as there are many causes. When you see your CF care team, you should discuss your symptoms with them.

At some point, your CF team may order a computed tomography (CT) scan of your sinuses to get an idea of the extent to which the sinuses are inflamed, how much fluid has accumulated in them, and if polyps are present. Depending on the severity of your symptoms, your team may suggest that you be evaluated by an otolaryngologist head and neck surgeon (an ENT), a surgical specialist who deals with disorders of the ears, nose, and throat.

After inquiring about your symptoms, the ENT may perform a nasal endoscopy using a flexible scope to peek up into the inside of your nose. This is a relatively quick and simple procedure done in the clinic. With the use of a small telescope, the otolaryngologist will look for a deviated septum, polyps, infection, and signs of

## Sinusitis and CF

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inflammation. If infection is noted, a sample can be taken to test for bacteria and guide your care team to use the best antibiotic.

If a CT scan has not been done yet, the ENT surgeon may ask for one to evaluate areas that are not accessible by endoscopy. This scan can show if there is infection or inflammation in the sinus cavities. A scan may also be needed if surgery is planned. Your otolaryngologist should communicate with your CF team when any changes in therapies are instituted, and particularly if you are considering surgery. Remind them to let the core CF team know what is going on.

### USE THE RIGHT THERAPIES

#### SALINE RINSES

On a day-to-day basis, clearing out thick mucus can be difficult. Saline (salt water) rinses are helpful to treat allergies, clear out environmental debris such as dust, and mobilize stagnant mucus.

High-volume rinses, either with a Neti Pot or a squeeze bottle, appear to be better than simple sprays. With these techniques, saline goes in one nostril, behind the nasal bone, then drips out the other nostril. It may take some practice getting the technique down, but once you do, it is pretty easy.

Hypertonic saline is a solution that is very salty. It can help reduce swelling and may thin out mucus, but may cause a burning sensation when rinsing, and the beneficial effects are temporary. Most people with CF prefer regular saline (usually 1 to 3 percent), which is just about as salty as normal human tissue. Your CF team will help you get the right kind of saline for your sinuses.

Sometimes ENT surgeons or CF care teams recommend rinses with antibiotics that are aimed at specific bacteria that may be affecting your sinuses.

### CONTROLLING INFLAMMATION

Steroid medications work to reduce inflammation. Nasal steroid sprays are not absorbed well by the nasal tissue, and daily use should not carry the side effect risks of steroids taken by mouth for many days or weeks. If you have allergies, antihistamines and other anti-inflammatory therapies can also be helpful.

### PROPER USE OF NASAL SPRAY MEDICATION

#### CHECKLIST OF STEPS

- 1 Blow nose gently.
- 2 Shake nasal spray gently.
- 3 Remove cap.
- 4 Hold nasal spray upright, thumb beneath the bottle, fingers on either side of the nozzle.
- 5 Tilt your head forward slightly.
- 6 Close one nostril with your finger.
- 7 Place the nozzle in other nostril.
- 8 Point the nozzle away from the center ridge of your nose, toward the inside corner of your eye on the same side.\*
- 9 Breathe in gently through your nose and press the applicator down firmly between your fingers and thumb.
- 10 Remove the nozzle from the nostril and breathe out gently through mouth.
- 11 Switch nasal spray to other hand and repeat steps 5-10 for the other nostril.
- 12 Wipe the nozzle.
- 13 Replace the cap.

\*You might find this easier if you hold the spray in your opposite hand.

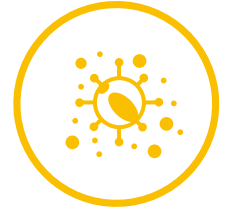
*From the National Asthma Council of Australia*



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# Nontuberculous Mycobacteria (NTM)

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For most people with cystic fibrosis, bacterial infections in the lungs are common. You probably know the names of the usual suspects found in CF lungs, such as “staph” (*Staphylococcus aureus*) or “Pseudomonas” (*Pseudomonas aeruginosa*).

But some people with CF have lung infections caused by a different type of bacteria. They are in the same family of bacteria that cause tuberculosis (TB), but they are not TB. This group of bacteria is referred to as nontuberculous mycobacteria (NTM).

NTM lung disease is not new, but it may be causing more infections in people with CF. This section provides some key information about how NTM may affect you, how it is diagnosed, and how, when needed, it is treated. Click on any of the basics below for more information.

## IN THIS SECTION:

- ▶ **Learn about NTM.** These bacteria are found in the environment and can get into the lungs of people with CF and other lung diseases.
- ▶ **Work with your CF team.** Your CF team will check your sputum for NTM at least once a year.
- ▶ **Get help when you need it.** If you find that treatments that usually make you feel better are not working so well, one reason could be an infection with NTM. Make sure to get a sputum sample to your CF team to be tested for NTM.

## Nontuberculous Mycobacteria (NTM)

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- ▶ **Use the right treatments for NTM.** If NTM is found in your sputum, several sputum samples and often a CT scan of your chest are used to decide if treatment is needed. The treatment is usually three or more antibiotics and may last up to a year or more.

### LEARN ABOUT NTM

**Nontuberculous mycobacteria** are a group of bacteria that have been found in the sputum of growing numbers of people with CF since the 1990s. These bacteria live in soil, swamps, and water sources. We know that there are over 100 types of NTM, and more are being found every year. NTM are very hardy and can survive many disinfectants and severe environmental conditions.

The group of bacteria that make up the different NTM are “cousins” to the bacteria that cause TB. These were often called “atypical mycobacteria” in the past. Despite this relationship, NTM are distinctly different and cause a different disease than TB.

- ▶ **Slow growers:** NTM grow much slower than other types of bacteria, making them much harder to diagnosis as they may not show up on laboratory cultures for up to 8 weeks. This slow growth makes the infection harder to treat as well.
- ▶ **Almost always in the lungs:** NTM can cause infections in many other diseases and medical conditions besides CF. For people with CF, the infection is nearly always in the lungs. NTM can enter the lungs directly through exposure from the environment. However, everyone with CF needs to be careful to not expose other people with CF or with problems with their immune system to respiratory infections through direct close contact. You can learn about avoiding germs in the “[Germs and Staying Healthy](#)” section on [CFF.org](#).
- ▶ **Diagnosed through sputum:** NTM lung infections are diagnosed by special cultures of your sputum. Sputum samples can be collected from coughing, from sputum induction (with saline), or from bronchoscopy (when a fiberoptic tube is passed into your lungs to get samples). The sample needs to be processed in a certain way to remove *Pseudomonas aeruginosa* and other usual bacteria. The final results may not be available for several weeks. The NTM infection can be diagnosed only if your doctor orders these special cultures. If cultures for NTM are repeatedly positive, your doctor may get a high-resolution chest CT scan to help confirm the diagnosis and find out the extent of the lung disease.

# Nontuberculous Mycobacteria (NTM)

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## BEYOND THE BASICS

There are many types, or species, of NTM. The most common types that cause infection in people with CF are *Mycobacterium avium* complex (MAC) and *Mycobacterium abscessus*.

To learn more about NTM and NTM lung disease in people with CF and in others, check out these sites and scientific articles:

- › CF Foundation webcast, "[Nontuberculous Mycobacteria and CF](#)"
- › CF Foundation [NTM Clinical Care Guidelines](#)
- › An overview of NTM:  
[www.nationaljewish.org/healthinfo/conditions/ntm/index.aspx](http://www.nationaljewish.org/healthinfo/conditions/ntm/index.aspx)
- › Information on *Mycobacterium avium* complex (MAC) lung disease:  
[www.maclungdisease.org](http://www.maclungdisease.org)
- › Centers for Disease Control and Prevention (use search term "Nontuberculous Mycobacteria"): [www.cdc.gov](http://www.cdc.gov)
- › Griffith DE, Aksamit T, Brown-Elliott BA, et al. An official ATS/IDSA statement: diagnosis, treatment, and prevention of nontuberculous mycobacterial diseases. *Am J Respir Crit Care Med*. 2007;175(4):367-416
- › [NTM Info & Research](#)

## WORK WITH YOUR CF TEAM

Your doctor or members of your CF team are probably already screening for NTM at your regular checkups. But if you are noticing symptoms or other changes in your body, you should feel comfortable bringing up the possibility of NTM with your CF team. The laboratory will not routinely check your sputum for NTM unless your doctor orders the test.

If you are diagnosed with NTM lung disease, you will likely be asked to visit the clinic more often so that your response to treatment can be checked regularly. You will be closely watched for any side effects of the treatments. You may be asked to start treatment when in the hospital; some people are able to start at home.

Since NTM disease mainly affects the lungs, it will be helpful to work with your respiratory therapist or physical therapist to make sure your airway clearance is as effective as possible. They can also help keep track of your lung function tests. Your dietitian can help you if your symptoms include weight loss or lack of appetite. Consider taking advantage of support from your social worker to help manage the costs of treatment and time away from work.

# Nontuberculous Mycobacteria (NTM)

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## GET HELP WHEN YOU NEED IT

It can be hard to diagnose NTM lung disease in CF because other CF-related infections cause similar symptoms on a daily basis.

### LUNG SYMPTOMS:

- › Cough
- › Sputum production
- › Blood-tinged sputum
- › Shortness of breath

If you already have some of these symptoms, the most important thing to think about is if these symptoms are changing or increasing in severity — especially if you and your CF team feel that your lung disease is becoming worse, even though you are taking good care of yourself and are doing all available treatments for typical with CF.

Clues that NTM may be in the lungs include worsening pulmonary function tests (PFTs); not getting well or back to your regular health after IV antibiotics or after a hospital stay; or needing oxygen when you didn't before. Another hint might be new "constitutional" symptoms, or a general level of feeling that is not your normal. People with NTM lung disease may show all, some, or none of the symptoms below — making NTM even more difficult to diagnose!

### GENERAL SYMPTOMS:

- › Fever
- › Chills
- › Night sweats — usually enough to leave your pajamas damp
- › Loss of appetite
- › Weight loss
- › Fatigue or lack of energy

Your doctor is likely screening for many of these symptoms at your regular checkups. But if you are noticing any of them or other changes in your body, you should ask your doctor or another member of your CF team if it might be NTM. If you are diagnosed with NTM, the symptoms will be watched to find out if the disease is getting better, worse, or staying the same.

# Nontuberculous Mycobacteria (NTM)

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## USE THE RIGHT TREATMENTS FOR NTM

### ANTIBIOTICS

NTM lung disease is hard to treat, especially in people with CF. Like typical CF-related infections from *Pseudomonas* and staph, NTM can chronically infect the airways. It can be very difficult to clear from the sputum. Plus, NTM is naturally resistant to many antibiotics.

Once your doctor and CF team decide that you should be treated for NTM lung disease, you will typically be started on several antibiotics at the same time. You may need to take these antibiotics for 12 months or longer. The goal is to “clear” the lungs of infection. This means having a sputum culture that no longer grows NTM.

The medications are usually taken by mouth. But there are times that an intravenous (IV) or inhaled antibiotic may be needed for a short period of time as well. The exact combination of antibiotics that your doctor prescribes will depend on the type of NTM. Tests will tell which antibiotics are the best at killing the NTM. More tests are done to watch for side effects or allergies that you might develop during months of treatment.

You will likely be asked to provide blood samples routinely in order to monitor levels of the drugs in your body. You may also need to have your hearing and vision tested routinely. This is to check for side effects of the NTM antibiotics your team is using.

### WHAT ELSE YOU CAN DO

In addition to the drugs, it is very important to continue your airway clearance. Often, people with CF and NTM need to do more airway clearance than others. Of course, regular exercise, staying away from secondhand smoke (tobacco, marijuana, etc.), and not smoking are always important. Good nutrition, care for your sinuses, and treatment of CF-related diabetes (if you have this diagnosis) are also essential.

### STAY TUNED

New techniques are being developed to better find the types of NTM. This may lead to more knowledge about NTM care. Many doctors and scientists are specifically studying NTM lung disease and treatment in CF, so there may be new approaches to treatment coming soon.

Talk to your doctor or CF team about new research and possibilities of participating in research studies related to NTM and CF.

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# Lung Transplantation

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Lung transplantation is a treatment option when your lungs can't fight back any longer. You may have had some discussions about transplantation along the way. But when your CF care team recommends that it is time to consider transplantation, it may be a difficult and personal decision.

You may think "I won't ever need a transplant." The unfortunate reality is that CF is a progressive disease, so there may come a day when the damage to your lungs is too great and your lungs begin to fail. Remember that sticking to your treatment is the best way to slow the progress of CF as much as possible. As new CF therapies come down the pipeline, the need for transplants will be delayed longer and longer. We hope someday better therapies will make lung transplant for CF unnecessary.

Lung transplant is a treatment option, not a cure. New lungs will not have CF but the rest of your body still will. With a lung transplant, you are trading one set of problems for another. Your goal should be to keep your own lungs as healthy as possible for as long as possible.

Some people with CF explore the transplant option but decide against it. No matter how you feel about transplantation, your CF team can provide important information to help you think about it.

# Lung Transplantation

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## IN THIS SECTION:

- ▶ **Learn about lung transplantation.**
- ▶ **Learn when is the right time for referral.**
- ▶ **Know the criteria for lung transplantation.**
- ▶ **Understand what is involved in the pretransplant evaluation.**
- ▶ **Know what will be expected of you before transplantation.**
- ▶ **Know what you can expect after lung transplantation.**

## LEARN ABOUT LUNG TRANSPLANTATION

**Lung transplantation** is a surgical procedure that removes both of your lungs and replaces them with healthy lungs from a recently deceased donor. This is a complex, major operation. You and the donor have to have matching blood type and tissue type, and be of similar size. A lung transplant carries serious risks, but it can extend life expectancy and enhance your quality of life when you are at end-stage CF lung disease.

People who have mild or moderate lung disease are not ready for lung transplant because the risks are greater than the benefits. Approximately half of all people (regardless of their disease) who receive lung transplants survive at least 5 years after the procedure. (For more information about lung transplantation, read the **Highlights** report containing CF patient registry data or see the United Network for Organ Sharing website at [www.unos.org](http://www.unos.org).)

The main risks are problems during the surgery and rejection of the new lungs. Because the transplanted lungs come from someone else, your body's immune system will see your new lungs as a foreign body and attempt to neutralize it. That process is called rejection.

Transplant rejection is a primary concern — immediately after the surgery and continuing throughout your life. To prevent transplant rejection, which can damage the new lungs, you must take a variety of medications that slow down rejection. These are called immunosuppressive drugs. This is a lifelong commitment and should be part of your daily care routine. (That means taking your medications as prescribed, keeping appointments, and doing what your transplant center wants you to do.)

## Lung Transplantation

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### LEARN WHEN IS THE RIGHT TIME FOR REFERRAL

When your CF team determines that your lung disease is severe enough to affect your quality of life and your life expectancy without transplantation is short, they will start the discussion of lung transplant and referral to a transplant center. This usually occurs when your FEV<sub>1</sub> is below 30 percent, you have increased shortness of breath with normal activity, exacerbations occur more frequently, you are using oxygen most of the day, you are requiring more frequent and longer hospitalizations, and you are unable to do what you want.

You need to communicate your thoughts and feelings about transplantation to your CF team. If you have concerns, fears, or questions, make sure you share them. Maybe your CF team hasn't mentioned transplant as a treatment option for you yet, and you are wondering if it is time. Bring it up with your CF team for discussion. It may be too early for a referral, but asking will give you information to be considered later.

### KNOW THE CRITERIA FOR LUNG TRANSPLANTATION

Just as CF care is offered at designated CF centers across the country, transplants are offered at specialized medical centers. Some medical centers have both, but if lung transplants are not offered at your CF center, your team will refer you to one that is close by and approved by your insurance.

Each transplant center sets its own criteria for transplant candidates, but certain criteria are generally agreed upon by all transplant centers:

- › Candidates must have end-stage lung disease.
- › Candidates must have exhausted other available therapies without success.
- › Candidates must have no other chronic medical conditions such as heart, liver, or kidney problems.
- › Candidates must have no current infections or recent cancer. In people with CF, pre-existing infection is unavoidable. Only a few centers in the country are accepting people with *Burkholderia cepacia* in their sputum, owing to the difficulty in treating this organism.
- › Candidates must not have HIV or hepatitis.
- › Candidates must not abuse alcohol or drugs.
- › Candidates must not smoke. Some centers may reject people who chew tobacco because of the increased risk of oral cancer.
- › Candidates must be within an acceptable weight range.



## Lung Transplantation

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- › Candidates must have an acceptable psychological profile.
- › Candidates must have a social support system.
- › Candidates must be able to comply with posttransplant medications and treatment recommendations. They must be willing to adhere to a lifetime regimen of medications and continuing medical care.

### UNDERSTAND WHAT IS INVOLVED IN THE PRETRANSPLANT EVALUATION

If you are being considered for lung transplant, your CF team will send your medical records to the transplant center. The center will review your records and arrange for you to have an evaluation at the transplant center. You will undergo an extensive series of medical tests to evaluate your overall health status and suitability for the transplant surgery.

- › Blood typing: Your blood type must match that of the donor. A mismatch of blood type can lead to a strong response by your body's immune system and can result in rejection of your new lungs.
- › Tissue typing: Ideally, your lung tissue will also match as closely as possible with the donated lungs, but the desire to find a compatible donor must be balanced against the urgency of a transplant.
- › Chest x-ray: To determine the size of your chest cavity
- › Pulmonary function tests
- › Oxygen assessment: Called a 6-minute walk test
- › CT scan of your chest
- › Bone mineral density scan
- › Cardiac blood pool scan
- › Cardiac stress test
- › Ventilation perfusion scan
- › Electrocardiogram (EKG)
- › Cardiac catheterization
- › Echocardiogram

This list of medical tests is not all-inclusive. The transplant center may want additional testing.

## Lung Transplantation

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Your evaluation will also include a meeting with the transplant social worker, financial coordinator, and dietitian, as well as a physical therapist or respiratory therapist. Depending on what is discovered during your evaluation, the transplant team may recommend that you meet with an anesthesiologist, psychologist, hematologist, or an infectious disease specialist.

You will also need to have your routine immunizations up to date, such as yearly flu shots, hepatitis A and B vaccines, tetanus, and pneumonia vaccines. Women will need to have a gynecological exam and mammogram, if appropriate. You'll also need to have a dental check-up. Best to start regular flossing now!

The transplant team will be able to provide you with a list of additional things you will need to do before a transplant.

The transplant center will have dedicated team members whose focus is you. Each person on the team is an expert in a different area of transplantation:

- › **Pulmonologists** are doctors who specialize in lung disease and help manage your condition before and after transplant.
- › **Transplant surgeons** are doctors who specialize in transplants and will do your surgery.
- › **Nurse coordinators** organize all areas of your care before and after your transplant. They provide education and will coordinate your diagnostic testing and follow-up care.
- › **Social workers** are professionals who help you and your family deal with many issues that arise, including lodging, transportation, finances, legal issues, and psychosocial issues. They have access to community resources and can provide you with individual and family counseling when your illness causes anxiety, depression, family conflict, bereavement, or changes in your physical or mental capabilities.
- › **Financial coordinators** have extensive experience arranging and clarifying the financial aspects of your care before, during, and after transplant.
- › **Pharmacists:** Dedicated pharmacists, who have extensive experience working with transplant patients, will be part of your team. They monitor your medicines while you are in the hospital and at your clinic visits.
- › **Dietitian:** A dietitian helps manage your nutrition before and after your transplant.

## Lung Transplantation

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Together, the team pools their expertise, knowledge, and understanding to take care of you. Like the CF team approach, you get better care when the transplantation team coordinates your care.

### **KNOW WHAT WILL BE EXPECTED OF YOU BEFORE TRANSPLANTATION**

Once it is decided you are a candidate for lung transplant, you will be placed on a regional waiting list. The length of time you may wait is impossible to predict. Your position on the list is determined by a Lung Allocation Score (LAS). The LAS system looks at the severity of your disease and your likelihood for survival and assigns you a score between 0 and 100. People with higher scores are considered the sickest and placed at the top of the waiting list. Scores are usually evaluated every 6 months.

Waiting for a transplant can be frustrating and stressful. It may be frightening at times. The social worker, transplant coordinator, and support groups can help you cope. You should use all resources available to help you get through this stressful time. Many transplant centers have support groups made up of other transplant patients and caregivers who have been through the lung transplant process. They can offer you and your family support and share their experiences. Many transplant centers have a chaplain on their team who can give you spiritual care and support as well. Your CF team can also help you deal with the stresses that often go along with being on a transplant waiting list.

During the time before your transplant, you still need to do all of your prescribed CF medicines and therapies. The transplant center may require you to enroll in a formal pulmonary rehabilitation program. They will give you the information you need to enroll. The transplant team will also give you all the information you need about the process of what happens when donor lungs are found for you.

### **KNOW WHAT YOU CAN EXPECT AFTER LUNG TRANSPLANTATION**

Once your transplant operation is over and you have your new lungs, it will be time to prepare to go home from the hospital. The lung transplant coordinators, the doctors, and nurses will begin to give you the information you need.

You will learn a great deal about medical care after a lung transplant, and you may be overwhelmed with the amount of information you need to learn. Don't worry; most everyone is.

## Lung Transplantation

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The medications used to stop or slow rejection creates a condition called immunosuppression. Once you are immunosuppressed, you will require careful attention and specialized medical care for the rest of your life. You will be given specific information on how to take care of your new lungs:

- › What medicines to take, at what time, in what amount and for what reason
- › What signs and symptoms to watch for that could indicate problems with your transplant and how to report them
- › How to monitor your health at home and keeping a logbook

Your transplant coordinator will give you information on what you will need to check when you monitor your health, preventing infection, pulmonary rehabilitation, nutrition, exercise and recreation, resuming sexual activity, dealing with emotions, returning to work or school, and handling emergencies.

You will need to know every detail of your care before you leave the hospital. Take notes and take it all in. Your attention to detail and ability to keep in good communication with the transplant center are extremely important.

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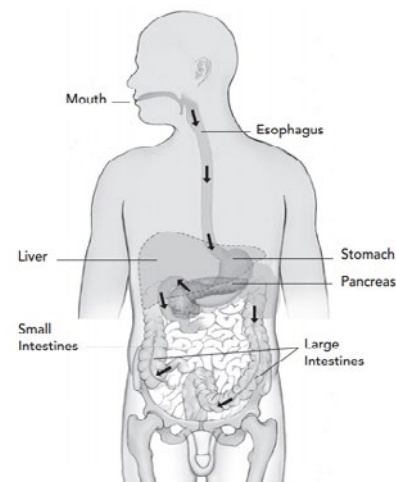
# Basics of the Digestive Tract

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If you have cystic fibrosis, you have to pay more attention to what you eat and how you digest food than most people do. And the focus is often on maintaining or even increasing weight — far different from the struggle to lose weight some of your friends probably face.

That’s because the digestive tract works a little differently when you have CF. This section gives you a kind of “owner’s manual” for your digestive tract. In the five links below, we’ve tried to give you the essential things you need to know and do when it comes to eating and keeping your digestive tract on track.



## IN THIS SECTION:

- ▶ **Learn about the digestive tract**, including the ways CF can decrease pancreatic enzymes and increase constipation.
- ▶ **Work with your CF team**, including the dietitian who can guide you on diet, pancreatic enzymes, vitamins, fluids, and even constipation medications.
- ▶ **Use the right gastrointestinal medications**, including pancreatic enzymes, replacement vitamins (especially vitamins A, D, E, and K) and, when you need them, stool softeners.

## Basics of the Digestive Tract

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- ▶ **Manage your time** — take the time to prepare good meals, eat well, and stay hydrated, and pick up cooking and shopping tips for people with CF.
- ▶ **Get help when you need it** — find out who to call if you have abdominal pain, esophageal reflux, constipation, diarrhea, or weight loss.

### LEARN ABOUT THE DIGESTIVE TRACT

Your digestive tract, which may also be called the “gastrointestinal tract” or “GI tract,” is the route your food follows once you put it in your mouth. The GI tract is where the food you eat is turned into the energy your body needs to do the things you want to do. It starts with your mouth, teeth, and tongue.

Once you swallow, the food goes down your esophagus and into the stomach, where it is turned into mush. From your stomach, the food mush moves into the small intestine, where the pancreas and liver help digest the food further so your body can use the energy. The last part of the GI tract is the large intestine, rectum, and anus — this is where the unusable bits of food you ate move out of your body.

### Beyond the Basics: A Tour of Your Digestive Tract

#### PANCREAS

To extract these nutrients, the body needs the help of the pancreas, an organ attached to the small intestine by a tube. The pancreas makes fluid — actually, a lot of fluid, over a liter each day — that mixes with the food.

One part of that fluid is a product that neutralizes the acid from the stomach. It is called “bicarbonate.” Also in that fluid are chemicals called “pancreatic enzymes” that mix with the carbohydrates, proteins, and fats in the food to break them into small bits the body can take in. These pancreatic enzymes are very, very important in getting the nutrients, calories, and vitamins into our bodies; they are the heart of good nutrition and health.

In addition to playing these important roles in the digestive process, the pancreas is also a maker of important hormones, including insulin. Insulin works in your bloodstream, not in your intestines. Insulin helps move the sugar and fuel from the blood into the cells so they can do their work.

For many people with CF, the pancreas doesn’t produce enough insulin or the body doesn’t let insulin do its job, leading to [CF-related diabetes](#) (CFRD).

## Basics of the Digestive Tract

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One way to simplify this is to think of the pancreas as two separate organs — one is a digestive organ that makes pancreatic enzymes for the intestines, the other is a hormone organ that makes insulin for the body.

To learn more about diabetes and CF, see "[Managing Cystic Fibrosis-Related Diabetes \(CFRD\): An Instruction Guide for Patients and Families](#)"

### BASICS OF THE PANCREAS

- › The pancreas lies behind your stomach, is shaped like a small fish, and is crucial for proper digestion.
- › The normal pancreas is an organ that:
  - Makes special chemicals called “pancreatic enzymes” to digest food as it moves from the stomach into the intestines
  - Makes the body’s natural antacid, called bicarbonate, to mix with food coming from your stomach into the intestines
  - Squirts fluid into the intestines to help flush food along its path through the digestive tract
  - Makes important hormones, including insulin
- › About 90 percent of people with CF have a pancreas that doesn’t make enough enzymes to digest food. Undigested food in your intestines can cause pain.
- › Everyone with CF (including people who don’t need enzymes) has a pancreas that does not make enough bicarbonate to neutralize stomach acid. This can contribute to pain.

### LIVER, GALLBLADDER, AND SMALL INTESTINE

The next organ on our tour is the liver, which, among its many functions, makes a fluid called “bile” that helps the body absorb fat. Bile travels through small tubes in the liver and is stored in the gallbladder, which can empty the bile into the small intestine.

For some people with CF, bile gets thick and flows very slowly. It can even get so thick that it forms stones in the gallbladder, which is sometimes removed. You can still have normal digestion if your gallbladder has been removed.

As the mixture of food, pancreatic fluid, bile, and pancreatic enzymes moves along the small intestine, the important nutrients make their way into the body through the walls of the intestines.

# Basics of the Digestive Tract

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## LARGE INTESTINE

After the small intestine comes the large intestine. A few nutrients are taken in by the large intestine, but its more important function is to keep the balance of fluid just right in the mass of what remains of the digested food.

The very end of the large intestine is called the “rectum.” This is where the mass of fully digested food leaves your body in the form of poop, or a bowel movement. (There are a lot of other names for poop, but we will not go over them here!) As you probably know by now, your care team will usually ask you about your bowel movements.

## MYTH BUSTERS

- › “Not taking enzymes is a good way to treat constipation, because your stools get looser when you don’t take enzymes.”

**NOT TRUE!** Not taking your enzymes puts you at INCREASED RISK of having constipation or a bowel obstruction. If you are getting constipated or straining to move your bowels, keep taking your enzymes but add fiber to your diet and get some medicine to keep your stools loose and bulky.

- › “I have bowel movements every day, so I can’t be constipated.”

**NOT TRUE!** People with CF who are constipated almost always have bowel movements every day, often more than once a day.

- › “My doctor told me I have a working pancreas (pancreatic sufficient) so I have a normal GI tract.”

**NOT TRUE!** You may still be prone to constipation and you should eat a high-fiber diet. You may need medication on occasion. Also, your pancreas is still sensitive to injury, so binge drinking of alcohol can change you from someone with a working pancreas to someone whose pancreas doesn’t work right.



# Basics of the Digestive Tract

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## WORK WITH YOUR CF TEAM

### ALL THE TIME:

- › Talk about your poop.
- › Talk about your stomachaches.
- › Let your team know if there's been any change in your pattern of digestion or bowel habit.
- › Develop and review a plan to prevent or manage constipation.

### AT LEAST ONCE A YEAR:

- › Meet with the team dietitian to review your diet, supplemental vitamins, and your use of enzymes.
- › Set a clear goal regarding your weight — or, in the terminology of your dietitian, your body mass index (BMI). Have a clear plan regarding weight gain or weight loss.
- › Review your blood levels of vitamins D, E, and A. Remember these are “fat” vitamins, and **MUST** be taken at a time you are taking pancreatic enzymes.
- › Ask about having a glucose tolerance test to check for CFRD.

## USE THE RIGHT GASTROINTESTINAL MEDICATIONS

Here are some of the most important things to know and remember about your GI medications.

### ENZYMES: TAKE THEM EVERY TIME YOU EAT

Most people with CF need to take **pancreatic enzyme** capsules with every meal and snack.

Look on the side of your enzyme bottle. It lists the amount of lipase, protease, and amylase. Lipase digests fat, protease digests protein and amylase digests starch. Unless a snack is pure sugar (such as a clear popsicle, sucking candy, or fruit juice), you need to take your enzymes so the body can digest the nutrients.

Many people have a fixed dose — for example, a certain number of capsules with each meal, half that number with a snack. Others increase their dose if they eat a meal that has lots of fat in it, such as a pepperoni pizza and chicken wings in blue cheese sauce.

## Basics of the Digestive Tract

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If you don't digest food properly by taking pancreatic enzyme pills, the poorly digested fat or protein or starch sits in your intestines, and there it encourages gas to form, which can cause pain and may cause unpleasant aromas later.

### WHAT ELSE YOU CAN DO

- › Don't put your enzymes where they can get hot, such as on a sunny windowsill or in the glove compartment of a car in warm weather. This causes the enzymes to spoil and stop working.
- › If you are away from your house, you'll need to think about ways to have enzymes available no matter where you are. Should you keep them in a drawer at work? In your backpack or purse? In your car in a thermal lunch carrier? Enzyme capsules don't need to be kept cold, they just shouldn't get too hot. Do what works for you.

### GASTROESOPHAGEAL REFLUX DISEASE OR "ACID REFLUX" TREATMENTS

The stomach makes acid. This acid can leak back from the stomach into the esophagus. This is called heartburn, acid reflux, or gastroesophageal reflux disease (GERD in medical talk).

Coughing can make this worse, so people with CF may be more likely to have GERD. The acid reflux can cause injury to the esophagus. It can also trigger cough or asthma.

Your CF care team may prescribe one of a couple types of medications for this problem to help decrease the amount of acid in your stomach. One type is called a proton pump inhibitor or PPI. **PPIs work best when you take them on an empty stomach, first thing in the morning or 30 minutes before a meal.** Another type of medication that suppresses acid is an H2 blocker. Some examples of these medications are in the table below. Your CF team might prescribe one type or the other or both if needed.

Common Medications	
PROTON PUMP INHIBITORS	H2 BLOCKERS
Omeprazole (Prilosec®)	Ranitidine (Zantac®)
Esomeprazole (Nexium®)	Nizatidine (Axid®)
Lansoprazole (Prevacid®)	Famotidine (Pepcid®)
Pantoprazole (Protonix®)	Cimetidine (Tagamet®)

Old-fashioned over-the-counter antacids, like aluminum hydroxide and magnesium hydroxide (Maalox® or Mylanta®), or H2 blockers, may be used along with, not instead of, the PPIs.

## Basics of the Digestive Tract

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### OTHER WAYS TO TREAT GERD:

- › Not eating big meals just before lying down.
- › Avoiding certain spicy foods, alcohol, and mints.
- › Lifting the head of your bed with blocks to keep your body sloping down.

There is also a surgical procedure that tightens the spot where the esophagus meets the stomach, called a Nissen fundoplication. Your physician may want to refer you to a stomach specialist (gastroenterologist) if GERD becomes an ongoing problem.

### CONSTIPATION TREATMENTS

In addition to making the digestive juices, the pancreas normally squirts 1 to 2 quarts of fluid into the intestines every day. That's right, 1 to 2 quarts! The production of this intestinal fluid is often impaired in people with CF, even if they make a normal amount of the pancreatic enzymes.

As in the lungs, the intestines in CF can make thick mucus. If you have this thick mucus but not enough fluid in the intestines, you can get constipated or even develop a bowel obstruction. This can be a serious problem and may lead to surgery if it persists.

Keep these problems in mind if you are having abdominal pain and you are having a hard time moving your bowels. Drinking plenty of fruit juices or electrolyte-containing beverages or sports drinks (not just water) may help. The over-the-counter medication polyethylene glycol is very effective in reversing hard stools and constipation. This is available at most pharmacies and grocery stores. Brand names of polyethylene glycol include MiraLAX®, GoLYTELY® and NuLYTELY®. Generics are also available.

### TWO CF CONDITIONS THAT MAKE IT HARD TO MOVE YOUR BOWELS

**Constipation** is when thick poop sits in your large intestine. People with CF can be having bowel movements every day and still have LOTS of poop backed up in their large intestine. Extra fluids, more fiber in your diet and regular exercise can help prevent this. If you have constipation, talk to your CF doctor about using a daily medicine like polyethylene glycol to keep fluid in your intestines.

**Distal intestinal obstruction syndrome (DIOS)** is a blockage or bowel obstruction, seen only in people with CF, when thick poop sits right at the spot where the small intestine joins the large intestine. This can be severe enough to totally block your intestines or it can be "low grade," when you have pain but aren't super sick.

Mild cases of DIOS may be treated at home with polyethylene glycol. Severe DIOS needs to be treated in the hospital.

## Basics of the Digestive Tract

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Treatments can include an NG tube (NG stands for nasogastric), which goes through your nose and into your stomach to drain your intestine. Sometimes special enemas are used. Other treatments could include large-volume medicines such as polyethylene glycol, which may either be consumed or given through an NG tube.

These medicines suck water into the intestines wash through the intestines to move blockages. If you have ever had DIOS, you probably need to be on a regular medicine every day to help your intestines move poop along.

### "GOOD BACTERIA" TREATMENTS

Everyone's intestines have bacteria, and most of them are in the large intestine. We normally live in harmony with the bacteria. But CF can change that balance.

With CF, instead of having a wide variety of healthy bacteria, the intestines sometimes have only one or two strains. Also, instead of staying in the large intestine, bacteria may be up in the small intestine where they can compete with our bodies for food. The bacteria can create gas and cause inflammation to the lining of the intestine, leading to pain or weight loss.

Your CF doctor may want to do a special test to look for "bacterial overgrowth" in the small intestine or may want to treat you with a special antibiotic to fix this problem (which can come and go). You may also want to consider daily use of over-the-counter products called "probiotics." These are good bacteria that come in pill form or are added to foods such as yogurt.

### STAY TUNED:

- › Believe it or not, enzyme capsules are such an old medicine that they were on the market before the Food and Drug Administration (FDA) existed! The FDA has reviewed pancreatic enzyme supplements, and many are now better and safer than ever before.
- › New technologies are being developed to study acid in the intestines. There is now a sensor pill that you can swallow that measures the acid, and it may help us understand what's going on in the GI tract of people with CF. In the past we just had to make guesses.

## Basics of the Digestive Tract

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### WHAT ELSE YOU CAN DO:

Things you can do to keep your GI tract health as good as possible:

- › Exercise regularly — it's good for your lungs and for your digestion.
- › Think actively about how to have enzymes available every time you eat.
- › Be sure to take **CF-specific vitamins** every day — best to take them with meals, so the pancreatic enzymes will help the vitamins get into your body.
- › Eat well and take time to enjoy eating.
- › Stay hydrated!

### MANAGE YOUR TIME

Successfully managing a “CF belly” really depends upon three things — input, processing, and output. It's that simple. Prepare to eat, have what you need to process (digest) what you've consumed, and anticipate the outcome. You have lots of ways to make this easier:

**Plan ahead.** Keeping yourself fed can be a lot of work and is time-consuming, whether you are feeling great or you're ill and disinterested in food.

- › Plan your menus so you have what you need on hand and don't have to play the “What's for dinner?” game or wander aimlessly around the grocery store. Many cooking websites provide creative ideas for menu planning, such as themed meals (Taco Night or No-Meat Mondays). Thinking ahead ensures more nutritious and satisfying options.
- › Use the slow cooker. For breakfast, overnight oatmeal is a hearty start to the day. If you find that you have more energy earlier in the day, make dinner in the slow cooker then so that at 5 p.m. you don't settle for pizza or fast food.
- › Even if you are only cooking for one or two people, look for magazines and recipe books designed for smaller portions so food doesn't go to waste. Or make a full portion and freeze part of it for when you're not feeling up to cooking.

**Carry a snack.** You never know when you're going to be held up or stuck somewhere. Easy-access snacks are very important for those with **CFRD**.

- › Energy bars, crackers prefilled with peanut butter or cheese, or a baggie with favorite nuts or dried fruit are easy to carry in any weather.
- › At home, string cheese or other pre-cut cheeses make great grab-and-go snacks. Keep hard-boiled eggs in the fridge for a quick breakfast or snack.

## Basics of the Digestive Tract

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**Pour pills.** Get your enzymes ready once a week rather than as needed. This is also a great way to monitor if you've taken them, because you may think you're taking all your meds but could be missing a few.

- › If necessary, use three or more pill cases (morning, afternoon, evening) and keep them where you will remember to take your meds — on the bedside table for morning, kitchen counter for evening, etc.
- › For enzymes, pour what you think you need for the week in larger containers and keep them in accessible places (on the dinner table or in your pocketbook/book bag). Consider leaving a bottle at a relative's or friend's house or in your car's glove compartment (providing it's not too hot or cold).
- › When brown-bagging it for lunch or school, pop enzymes into the bag so no effort is needed to search for them.

**Go to the bathroom before you leave.** Don't hold it! Inevitably, if you decide to wait, you'll get caught in traffic or stuck on a subway or worse.

- › If you are having a particularly "bad belly" day, think about your route ahead of time. If you are driving, prepare to stop at rest stops you know are clean. If you are in a big city, head for the public restrooms in hotel lobbies or upscale department stores.

**Be proactive with probiotics.** When on antibiotics, especially IV antibiotics, take acidophilus, yogurt, or probiotics to help your body balance the good/bad bacteria. Know your "normal" so that if your lifestyle or eating habits have changed, you can adjust enzymes to prevent diarrhea or take stool softeners to prevent constipation and obstruction and keep things moving.

**Don't strain.** Use natural fiber, polyethylene glycol, or a stool softener if you have any constipation. Beat it rather than wait to treat it!

### AT SCHOOL OR WORK

If there are times when you are having more stomachaches or gas, you may want to spend some extra time in the bathroom. Some people like to keep a small can of air freshener with them to minimize the smell.

If you are having more stomach pain or gas than usual, call your CF care center for advice. You should *never* feel ill at ease about doing what you need to do to take care of yourself.

# Basics of the Digestive Tract

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## GET HELP WHEN YOU NEED IT

Call your CF care team if:

- › You have not had a bowel movement in more than a day or two.
- › You are not feeling hungry or have no appetite.
- › You have severe stomachaches.
- › You are losing weight.
- › You see any blood in your stool.
- › You are vomiting.

GI/Nutrition Webcasts

[www.youtube.com/playlist?list=PLhoQ6vyZhggo9F9-5ANhpr3H4MtSZudt](https://www.youtube.com/playlist?list=PLhoQ6vyZhggo9F9-5ANhpr3H4MtSZudt)

Meal and Snack Ideas

[www.youtube.com/playlist?list=PLhoQ6vyZhggo9F9-5ANhpr3H4MtSZudt](https://www.youtube.com/playlist?list=PLhoQ6vyZhggo9F9-5ANhpr3H4MtSZudt)

Color Your Calories

[CFF.org/PDF-Archive/Color-Your-Calories.pdf](https://www.cff.org/PDF-Archive/Color-Your-Calories.pdf)

Pancreatic Enzyme Replacement

[CFF.org/Living-with-CF/Treatments-and-Therapies/Nutrition/Taking-Care-of-Your-Digestive-System/Enzymes/](https://www.cff.org/Living-with-CF/Treatments-and-Therapies/Nutrition/Taking-Care-of-Your-Digestive-System/Enzymes/)

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# Distal Intestinal Obstructive Syndrome (DIOS)

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Cystic fibrosis can cause changes in your gastrointestinal (GI) tract. GI problems often take “second place” to lung problems in CF, but they are very important. GI problems can be the source of pain, poor appetite, and weight loss. Paying attention to your gut and working with your doctor to address your GI problems can make you more comfortable, help you keep up your weight, and, in the long run, even help your lung function.

## IN THIS SECTION:

- ▶ **Learn about DIOS** — find out how constipation and DIOS are different, and how to know which you have. And how do you say “DIOS,” anyway?
- ▶ **Work with your CF team** to collect information about your problem that will make it easier for your doctors and caregivers to help you.
- ▶ **Get help when you need it** — Know the warning signs that mean you should see your doctor right away or go to an emergency room.
- ▶ **Use the right treatments** — Find out what works for DIOS and how to keep it from happening, or from happening again.



# Distal Intestinal Obstructive Syndrome (DIOS)

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## LEARN ABOUT DISTAL INTESTINAL OBSTRUCTIVE SYNDROME

CF causes changes in your GI tract that may lead to problems such as DIOS.

This is because:

- › The secretions in the gut of people with CF are drier than those in a normal gut. It is as hard to move things through the gut because of dryness as it is to move sputum in the lung. The mucus of the intestine is the lubricant that keeps food and old cells moving along.
- › The intestine in people with CF has slow motility. The normal gut is in constant motion, moving food and fluid toward the rectum. These movements are both slowed and irregular in the gut of people with CF. At this time, we don't fully know why.
- › If you have pancreatic insufficiency (you take enzymes with your meals and snacks), you have some undigested food in your gut. Even if you have the best enzyme dose and take them perfectly faithfully, they do not completely reproduce what the normal pancreas does.

So, as you can imagine, an intestine full of dry, poorly moving feces with undigested food in it could cause some problems. Two of these problems are constipation and DIOS.

The European Society of Pediatric Gastroenterology, Hepatology, and Nutrition (Houwen, 2010) defines the differences between constipation and DIOS. These definitions may not perfectly describe everyone's problems, but they will help you see how constipation and DIOS are different.

### CONSTIPATION

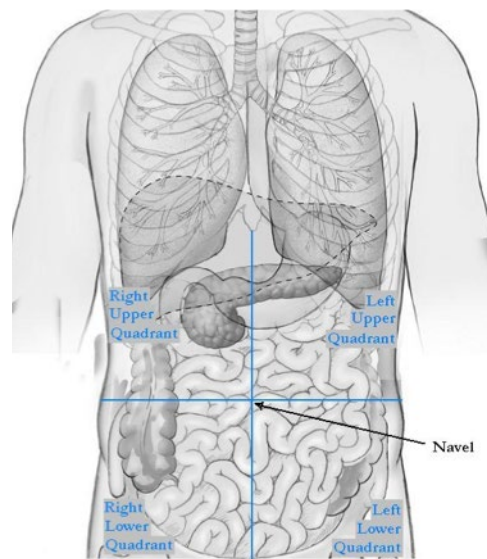
- › Constipation comes on slowly — over weeks or months (although if you get sick or dehydrated it could come on pretty fast).
- › Constipated people have fewer stools over the last weeks or months, and the stools they have are smaller and harder. The stool is more difficult to pass. When constipated people poop, the stools can look like small, hard pellets or be so gigantic they clog the toilet (Not turning around to look at your poop? If you're having trouble going to the bathroom, it's time to start looking — your gastroenterologist does!)
- › People with constipation can have abdominal pain (usually around their navel) and their bellies can look full.
- › If you are constipated, your doctor or care team may suggest taking a laxative like polyethylene glycol (MiraLAX®). If this is the first time you're having this problem, be sure you ask for advice.

# Distal Intestinal Obstructive Syndrome (DIOS)

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## DISTAL INTESTINAL OBSTRUCTIVE SYNDROME

- › DIOS usually comes on pretty fast — over a few days.
- › There is complete or partial intestinal obstruction — you may or may not be vomiting. The vomit material could be stained with bile (green or yellow). If you're not vomiting, you'll probably feel nauseated or sick to your stomach.
- › You will likely have pain — often in your right lower quadrant, and you may be able to feel a lump there. This is the same place that often hurts when you have appendicitis. It can be hard to tell the difference between DIOS and appendicitis.
- › Your abdomen may be bloated, look bigger, or feel more full.
- › Pooping may not make you feel better or completely relieve the discomfort. If you felt a lump in your right lower quadrant, it might not go away. This lump is stool that has become so hard and dehydrated that it is causing some blockage even if you have pooped.



If your symptoms fall somewhere in between, you could have very severe constipation or be moving slowly toward DIOS. The early treatment of both problems is similar. If you are worried about your symptoms, it is time to call your CF doctor and make sure you stop a major problem before it starts. Your care team will be just as happy to help you with your belly as with your lungs.

- › [View a webcast about constipation and DIOS](#)

## PRONOUNCING "DIOS"

OK, every doctor disagrees. Some say "dee-ose," some say "die-ose." The good news is you can hardly go wrong — just call for help if you're having troubles.

# Distal Intestinal Obstructive Syndrome (DIOS)

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## WORK WITH YOUR CF TEAM

Your care team will want to know if there are changes in your health or the way you are taking your medications that make DIOS more likely. Some examples:

- › Not taking your enzymes appropriately. This might cause diarrhea but can also lead to constipation (not to mention the loss of nutrients and vitamins).
- › Not drinking enough fluids, particularly in very hot weather or during exercise or hard work.
- › Taking narcotics for pain.
- › Getting sick — When you are sick for almost any reason, your body needs more water and will steal it from your gut if it needs to. Your intestines may also slow down when you are sick.

Even if you think it's "just constipation," your CF doctor should know about it because it is handled in a special way if you have CF.

## TREATING CONSTIPATION AND DISTAL INTESTINAL OBSTRUCTIVE SYNDROME

Your CF doctor will evaluate you with a history and physical exam. If you are constipated, your doctor may suggest drinking more fluids or starting either a laxative medication or one to increase the fluid in your intestinal tract, such as polyethylene glycol. If your symptoms suggest DIOS, you may get a "bowel cleanout," which is a larger dose of polyethylene glycol. One common thing to do before or after treatment of DIOS is an x-ray of your abdomen. This can help tell if a lot of stool is backed up and also help make sure that it's all cleared out once your treatment is done.

Not every case of DIOS is simple. Sometimes the first treatments of drinking more fluids, using stool softeners, or having a GI cleanout don't work. Many treatments may be done at home. Just like therapies for your lungs, though, if it's not successful at home, sometimes hospitalization is necessary. If you are nauseated or can't hold down lots of fluids or enough polyethylene glycol to clear you out, more polyethylene glycol can be given through a nasogastric tube (a tube that a doctor or nurse puts through your nose to your stomach). Large volumes of polyethylene glycol can be put through this tube to your stomach to try to get the stool moving.

For some cases, enemas (fluid inserted into your rectum) may be necessary — to try to get at the problem from both ends, so to speak. Some can be given by nurses in the hospital (or at home if you have a mild case). Others, such as a diatrizoate meglumine and diatrizoate sodium (Gastrografin®), can be given by a radiologist using an x-ray, trying to get the liquid as close to the blockage as possible to break it up.

## Distal Intestinal Obstructive Syndrome (DIOS)

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If none of these therapies work, your doctor may order a CT scan of your belly to make sure there isn't another problem such as an infection, an abscess, or a mass that is not just stool. A last resort is surgery to remove the blockage. This is not often used with people who have CF, but if your case is severe, a surgeon might be on standby in case the nonsurgical options don't work.

As you can see, there are a lot of ways to treat DIOS. Some take time, even several days. The good news is that the treatments are usually very effective. The treatment that your doctor prescribes will be specialized to what you need. After you are treated for your constipation or DIOS, you have to do some work to keep up your GI health by staying hydrated, taking your enzymes, and sometimes taking daily polyethylene glycol or stool softeners. It's not fun, but it's better than being backed up again.

### GET HELP WHEN YOU NEED IT

Red flags — any of these problems should make you head for the emergency room, or, at least, call your doctor right away:

- › Nausea and vomiting — signs of obstruction
- › Severe abdominal pain that wakes you up, keeps you from sleeping, or keeps you from doing things you otherwise would want to do
- › Blood in your stool or vomit
- › More difficulty breathing
- › Weight loss

### USE THE RIGHT TREATMENTS

You might be thinking that you could treat constipation and mild early DIOS on your own. If this problem is new for you and you have not talked with your CF care team about what actions to take, it is important to call. Some centers want you to initiate an action plan if you have no bowel movement in a 24-hour period.

If you are feeling pretty good and have experience with managing constipation related to your CF, you may want to try the following (if they have been recommended by your CF team).

- › Start drinking **a lot** more — especially salt-containing beverages like Gatorade® or Powerade®. This will give your dehydrated poop a chance to get softer and move along.

## Distal Intestinal Obstructive Syndrome (DIOS)

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- › Start taking polyethylene glycol, 1 to 2 doses a day. This may make things move forward and help you pass some stool.
- › If your doctor is prescribing narcotic pain medications for you, ask for a bowel management program at the same time you pick up your prescription for narcotics. Get it started right away.
- › DON'T try to give yourself diarrhea by taking less or no enzymes — this can make the problem much worse!
- › DON'T ignore symptoms that could turn into a bigger problem. Call if things aren't right with your GI tract, just like you would for your lungs.
- › If you have abdominal pain and are not sure why, make sure you call your CF care team. Problems other than constipation and DIOS can cause abdominal pain.

### PREVENT THE PROBLEMS

If you became constipated or had DIOS once, it can and most likely will happen again. Some tips for prevention:

- › Drink lots of fluid.
- › Take your enzymes regularly and ask your care team if you're taking the right dose.
- › Increase the fiber in your diet.
- › Take polyethylene glycol every day or more often if recommended.

### SPEAKING FROM EXPERIENCE

"After my episode with DIOS, I will do anything I can to avoid that at all costs. I was at another local hospital and they were ready to open me up and perform surgery to "fix" me. I called and requested a transfer to be where my CF doctors were. At first the GoLYTELY® solution was not taken so lightly by my body. Eventually, after two Gastrografin® enemas and two days of continued GoLYTELY® down my NG tube, the medicine worked and I was able to avoid surgery. I truly appreciate how my CF team took their time to help me heal instead of cutting me open. I try to keep all systems flowing daily with a little MiraLAX®, plenty of liquids, and the right mixture of other medications. The two seconds that it takes daily is worth every second of not being in the hospital." – Ben, Age 35

### REFERENCE

Houwen, RH. "Defining DIOS and constipation in cystic fibrosis with a multicentre study on the incidence, characteristics, and treatment of DIOS." *J Pediatr Gastroenterol Nutr.* 2010; 50 (1): 38.

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# Gastroesophageal Reflux in CF

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**Gastroesophageal reflux (or “reflux”) is very common in the general population — you have seen all the ads for reflux medication on TV. This may make you think that heartburn or feeling food come back up after you swallow is OK, but it is not. Reflux is a concern for people with cystic fibrosis for two reasons: First, heartburn is a source of pain, as it is for anyone, but if you have CF, and have so much pain that you eat less and lose weight, that can be bad for your lungs and overall health. Second, for people with lung disease, the chance of aspirating (breathing in) some of the refluxed stomach contents is dangerous.**

## **IN THIS SECTION:**

- ▶ **Learn about gastroesophageal reflux disease (GERD)** — how to know whether you have reflux.
- ▶ **Work with your CF team:** Collect information about your problem that will make it easier for your doctors and caregivers to help you.
- ▶ **Use the right treatments:** Find out what works for GERD and how can you keep it from happening, or from happening again.
- ▶ **Get help when you need it:** Know the warning signs that mean you should see your doctor right away or go to an emergency room.

# Gastroesophageal Reflux in CF

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## LEARN ABOUT GASTROESOPHAGEAL REFLUX DISEASE

Reflux occurs when stomach contents (food, saliva, and acid) come back up the esophagus. The material may come part way up, or come all the way into your mouth (doctors call this “water brash” or “acid brash” — it tastes like vomit).

During the day, reflux may cause heartburn — a vague, burning pain in the middle of your chest. It can be severe enough that people think they are having a heart attack (and if you think that, go to the emergency room). Heartburn can be very painful and unpleasant. It can keep you from eating foods that you like and that are good for you, such as citrus fruits and tomatoes, because more acidic foods may hurt more as they go down.

The problem is even more serious at night. During the day, when food and acid come back up our esophagus, we feel it happen and swallow hard to wash the material back down. We may take sips of water to help. At night, when we are asleep, we depend on “unconscious swallows” to get this material back down — and they are not as powerful as our conscious swallows.

At night we are also lying flat in bed, so we don’t have gravity working in our favor. The refluxed material can more easily get all the way up our esophagus, and, instead of swallowing it, we may breathe it in (aspirate it) down into our lungs. This can cause coughing and choking, and may cause pneumonia. You can even have silent aspiration. That means you don’t notice the reflux happening but lung damage and scarring can take place. As you can see, reflux at night is very risky for people with CF.

You may have heartburn and be very suspicious that you have reflux. Other clues: You hiccup a lot, burp a lot, or have burps that taste bad. You may wake up at night with chest pain, or you may wake up in the middle of the night coughing and choking. You may have really bad breath during the day. (Many people have bad breath first thing in the morning — that is probably not reflux.)

## WORK WITH YOUR CF TEAM

Keep a record of your symptoms to share with your doctor. Write down what time of day you are having pain (particularly if it happens at the same general time each day), where on your body you feel the pain, and how the pain feels to you. Your clinician may ask what you would do to give him or her the same pain. Think about that — is the pain like pins sticking into you, or burning, or someone standing on you? Describe however it feels, and that will help them figure it out! Write down any other symptoms you notice, like nausea or vomiting. You may think you will remember all this, but under the stress of an office visit, a written record can really help.

## Gastroesophageal Reflux in CF

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Your doctor may do any of the following: first, they might give you an acid-suppressing medication such as an H2 blocker (like ranitidine or famotidine) or a proton pump inhibitor (PPI, like omeprazole, lansoprazole, pantoprazole, or esomeprazole). If you are already on one of these meds, they might try a higher dosage. Second, they may want to refer you to a gastrointestinal (GI) specialist. That specialist may suggest other therapies or do tests to look for the presence of reflux and see how serious it is.

One test is a pH probe, in which a sensor is placed through your nose down into your esophagus to sense how acidic it is and if stomach contents (which are very acidic!) are found where they're not supposed to be. Another common test is an endoscopy, a test where they look down into your esophagus and stomach to see how much damage the reflux has done, if any.

Talk to your CF doctor before an endoscopy, to make sure you are in good shape as far as your breathing is concerned or to "tune up" your lungs before you get the sedation for the procedure. A tune-up might mean increasing your airway clearance or going on a course of oral or IV antibiotics. Depending on your symptoms and the exam, other things the GI doctor might do is draw blood to look for other causes of your symptoms, do an ultrasound of your abdomen, or take an x-ray of your stomach (an "upper GI series").

### USE THE RIGHT TREATMENTS

If you think you have reflux, you can try a few things before seeing a doctor (but see the list of "red flags" under "Get Help When You Need It" and always call your CF team with questions).

#### TRY ACID SUPPRESSION

Calcium carbonate — antacids (such as TUMS®) — can be safely tried at home for symptoms of reflux. If you need them less than once or twice a week, this might be the answer for you. Antacids are often not powerful enough and do not last long enough to be a good long-term treatment, although they may offer short-term relief. If you're using them every day or many times a day, see your doctor for something stronger. You might be covering up a problem that needs to be addressed more seriously.

Your doctor might suggest taking an over-the-counter acid suppression medication such as omeprazole (a PPI) or famotidine (an H2 blocker) every day for 2 weeks to see if your pain goes away. A PPI should be taken in the morning 30 minutes before breakfast or anything else in your stomach. An H2 blocker should be taken at night, before you go to bed. Ask for advice from your doctor and pharmacist before trying meds on your own.



## Gastroesophageal Reflux in CF

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If one of these drugs does not relieve your pain in 2 weeks or the pain gets worse, call and see a doctor. If your pain does go away, you may use the medicine for 6 to 8 weeks and then stop. If the pain returns after stopping or a medicine stops working as well as it used to, call and see your CF team.

Some people with CF are prescribed an H2 blocker or PPI not because they have reflux, but because these meds can help their enzymes work better. Enzymes work best at a certain pH, and these meds can help raise the pH in your GI tract to allow the enzymes to do their job well. You can still have reflux if you are on these meds for this reason. If you do, you might need a higher dose or to see a GI specialist for further testing. Regardless, tell your CF care team about this at your next appointment.

### GET GRAVITY WORKING FOR YOU AT NIGHT

Another thing you can do, particularly if you have symptoms at night, is to put your bed at an angle. Use something such as slabs of wood to lift the section of your bed where your head would be. The goal is to have your head be a little bit higher than your feet while your body is still straight. You should be able to see the angle when you look at your bed, but when you get in bed, you shouldn't feel like you are going to slide to the bottom. If you have to hang onto the top of the mattress to keep from falling off the other end, you have put the bed too high!

If this seems silly, remember that before acid suppression drugs came along, raising the head of the bed at night was one of our only and most powerful tools in fighting reflux! Don't take a short-cut — putting extra pillows under your head can bend your body in a way that can make reflux worse.

### AVOID HABITS THAT CREATE MORE ACID AT NIGHT

We want people with CF to eat frequently, but if you are eating 1 to 2 hours before bedtime, it increases your risk of having reflux problems at night. Try not to eat just before you go to bed. Also, some food and drinks, such as drinks containing alcohol and caffeine, are known to stimulate reflux. Tobacco is just plain bad all the way around and stimulates reflux as well. Try to avoid caffeine after noon. If you want an alcoholic drink in the evening (and you are of age!), try to have it early, before dinner, to reduce its effect on your nighttime reflux.

Besides drinks containing alcohol and caffeine, here are other foods and drinks that often make reflux worse: garlic, onions, pepper or spicy foods, mint beverages or peppermints, chocolate, citrus, carbonated beverages, and milk. Some fatty foods can make reflux worse too, but you don't want to avoid fatty foods and cut calories to correct your reflux problem. Keeping your weight up is very important. Talk with your CF team for help maintaining a good balance between eating enough and controlling reflux.

# Gastroesophageal Reflux in CF

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## PAY ATTENTION TO TUBE FEEDS

Reflux can complicate tube feeds. You might need to use some of the strategies in the previous text if you have a G-tube and are doing feeds overnight to help keep your calories up. If you are having trouble with your feeds, feeling too full in the morning, or feeling sick, this might be a reflux problem that needs to be fixed. Instead of giving up on feeds, talk to your doctor so you can get the most out of your feeds. For more information, read [Supporting Nutrition: Understanding Tube Feeding](#).

## GET HELP WHEN YOU NEED IT

Red flags are things that should have you going to the emergency department or calling or seeing your doctor right away. Don't fool around trying to make these things go away on your own:

- › Vomiting blood or having blood in your stool (it can be red, maroon, or black)
- › Severe pain that keeps you from doing things you enjoy or makes you fearful that you are having a heart attack
- › Vomiting
- › Losing weight
- › More pain when you eat fatty foods (this might suggest your problem is your gallbladder, not reflux)
- › A lot of coughing at night, or awakening many nights with the taste of vomit in your mouth — our lungs are too important to take any chances

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# Improving Nutrition With Tube Feedings

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Many adults with cystic fibrosis use tube feedings to support their nutrition. This section will help you understand why and how tube feeding can help you.

You have heard from your CF care team, your parents, and others all your life about how important it is to increase the calories you eat to help with weight gain and girth. You know all the tricks to adding calories to your meals. Often the amount you need to eat is greater than how much you can eat. Many people with CF need to take in double the calories of their friends and family who don't have CF. This is where support from tube feeding can be helpful and help ease the burden of needing to eat so much. Extra nutrition by a tube feeding can help you meet your nutrition goals determined by you and your CF care team.

Extra nutrition can not only increase your weight but it can also lead to improved ability to fight infection and improve your energy.

It is important to discuss all changes in your care with your CF care team. This section will help you educate yourself and understand how tube feeding can ultimately help you.

# Improving Nutrition With Tube Feedings

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## IN THIS SECTION:

- ▶ **Learn about tube feeding** — when is it time for a feeding tube; what kinds of tubes there are.
- ▶ **Learn how to manage your feeding tube.**
- ▶ **Work with your CF team** on proper supplements and insurance coverage.

## LEARN ABOUT TUBE FEEDING

### WHEN DO I REALLY NEED A TUBE?

There are many personal and medical reasons why you may choose to have a feeding tube placed. You may have a goal of wanting to gain weight or just maintain a certain weight.

There are many reasons for having healthy weight goals. Good evidence from population-based studies shows that normal ranges of weight and height are associated with better pulmonary function. This translates into a BMI (body mass index) of 22 for women and 23 for men. Your dietitian can explain what your BMI should be. More weight gain can increase your energy, possibly reduce exacerbations of your lung disease, and allow you to have better exercise workouts to increase your endurance and strength. To calculate your BMI, go to [nhibisupport.com/bmi](http://nhibisupport.com/bmi).

Your health care team wants to work with you to keep you healthy. They will help you get the most out of your nutrition through supplements and appropriate use of pancreatic enzymes.

Some factors may get in the way of you being able to achieve adequate weight:

- ▶ **Poor appetite.** If your appetite is always poor or you can't physically eat the amount of food you are used to (which leads to weight loss, muscle loss, and getting sick more often), extra nutrition through a feeding tube might be right for you. Your appetite can be reduced for many reasons but when you do not have the desire to eat, your calorie intake goes down. CF lung disease may make your lungs expand and hinder your stomach from stretching after a meal; therefore, eating large portions may be difficult for you.
- ▶ **Nausea or vomiting.** If you often vomit from coughing or have severe heartburn, you may want to consider taking in extra calories with a feeding tube.

## Improving Nutrition With Tube Feedings

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- › **Worsening lung function.** If you are unable to regain weight that was lost when you were sick or you are trying to reach a good weight for lung transplantation, tube feeding is an option.

### WHAT KINDS OF TUBES ARE THERE?

**Gastrostomy tube (G-tube):** The most common feeding tube is the gastrostomy tube, also called a G-tube or a percutaneous endoscopic gastrostomy (PEG) tube. It is a flexible tube that is inserted directly into the stomach through an incision in your abdomen. This makes it possible to deliver nutrition directly into your stomach.

**Low profile "button" G-tube:** After a few months of healing from the G-tube placement, your doctor may recommend replacing the tube with a "button." This device is flatter and lies against the skin of your abdomen. It can easily be placed once the G-tube site is healed, usually about 8 to 12 weeks after you have had the G-tube placed. This time may vary at your hospital. The button can be opened for feedings and closed in between feedings or medications. For many, the transition to a button makes tube feedings and care easier and more convenient. Some hospitals may place the button without inserting the G-tube first.

**Nasogastric tube (NG tube):** An NG tube is a thin, flexible tube that goes in your nose, down your throat, and into your stomach. This is the least invasive type of feeding tube because inserting an NG tube does not require a surgical incision. These tubes need to be inserted each night and then taken out in the morning for overnight feedings.

**Jejunostomy Tube (J-tube):** A J-tube is inserted through an incision in the abdomen directly into the section of your small intestine called the jejunum. It bypasses the stomach. J-tubes are used if a person cannot tolerate feedings directly into the stomach.

**Gastrostomy-jejunostomy Tube (GJ-tube):** A GJ-tube is inserted through the abdomen into the stomach, but does not stop there. The GJ-tube passes through the stomach into the jejunum, delivering the feedings directly into the small intestine, just as they are with a J-tube.

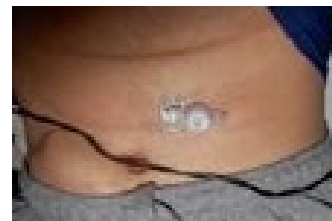
### WHAT WILL THE TUBE LOOK LIKE ON ME?



▶ Tube



▶ Button



▶ PEG Tube

# Improving Nutrition With Tube Feedings

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## LEARN HOW TO MANAGE YOUR FEEDING TUBE

### WHAT GOES THROUGH THE TUBE?

Usually the same supplements your health care team has prescribed for you to drink can go through the tube to nourish you. You can obtain both brand-name products and generic supplements over the counter or through your home care or medical equipment company. There are also tube feeding products that are specialized for a person's different needs.

### HOW DO I GIVE THE FEEDINGS?

Everything is individual! You will work with your CF care team to make a schedule that works best for you. Some choose to give tube feedings during the daytime, at nighttime, only between meals, or all day. Some feedings can be delivered with a pump, a hanging bag, or a large syringe. Overnight feeding can last anywhere from 6 to 18 hours each night.

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**You will need to take pancreatic enzymes with your tube feeding to help you absorb the nutrients: protein, fat, vitamins, and minerals. Your dietitian will discuss with you the best way to take your enzymes. The main goal is to prevent malabsorption (which presents as oily, multiple, light-colored stools) and promote weight gain.**

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### WILL THERE BE ANY PROBLEMS?

With any new therapy, concerns may come up. Your team can help you troubleshoot them.

# Improving Nutrition With Tube Feedings

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## NAUSEA AND VOMITING

### › Possible causes:

- Medications can cause nausea and vomiting.
- Something is blocking the intestine.
- Tube is not positioned correctly.
- Tube feeding formula, rate, or amount may not be ideal.

### › What to do:

- Reduce the rate of the tube feeding.
- Space the tube feedings farther apart.
- If nauseated, hold feedings until the symptoms subside.
- Check the amount of liquid you have in your stomach before feeding. This is done with a syringe. Your CF care team will give you guidelines on when to hold your tube feeding.
- If you feel you will vomit, leave the tube open to drain.
- Call your health care professional if nausea and vomiting prevent you from getting your full feedings for more than one day.

## CONSTIPATION (DECREASED FREQUENCY OR HARD TO PASS STOOLS)

### › Possible causes:

- Inadequate fluid intake or skipping feedings
- Physical inactivity
- Not enough fiber
- Medications

### › What to do:

- Check that you are taking the prescribed amounts of tube-feeding formula and water flushes.
- Do not skip feedings.
- Participate in physical activity (walking).

## Improving Nutrition With Tube Feedings

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- Try ½ cup prune juice flushed with ½ cup water down the tube one or two times per day.
- Call your health care professional if you are constipated for more than 3 days.

### DIARRHEA (LOOSE, WATERY, FREQUENT STOOLS)

#### › Possible causes:

- Tube feeding rate that is too fast (especially if you have a J-tube or small bowel tube)
- Not enough pancreatic enzymes
- Tube not positioned correctly
- Lack of fiber in diet or tube feeding
- Medications
- Bacterial contamination of feedings, virus, or other infection

#### › What to do:

- Decrease the tube feeding rate.
- Increase water flushes by 2 to 3 cups per day to replace losses in the stool.
- Keep unused, opened formula covered in the refrigerator for only 24 hours; discard it if not used.
- Use clean techniques for storing and giving feedings.
- Discontinue use of laxatives or stool softeners.
- Call your health care professional if diarrhea occurs more than six times a day.

### CLOGGED TUBE

#### › Possible causes:

- Tube older than 2 years
- Tube that has a small diameter or tube that is very long
- Not flushing the tube after feedings or medications
- Not dissolving medications well
- Not flushing the tube after checking residuals



## Improving Nutrition With Tube Feedings

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### › What to do to prevent a clogged tube:

- Replace old tubes as directed by your doctor.
- Flush with at least 60 mL of warm water after feedings, medications, and residual checks.
- Do not put any solids or foods through your tube.
- If your feedings begin to run in slower, irrigate the tube with water more often.

### › What to do if your tube clogs:

- Push 30 mL of warm water through the tube using a pumping motion with the syringe (you may need to do this three to five times before clog begins to move).
- Try to move the contents of the tube by pulling and pushing the syringe barrel several times.
- Pump air through the tube using the syringe (you may need to do this three to five times).
- You may use small amounts of carbonated beverages to unclog the tube; however, speak with your dietitian or health care professional before using products other than warm water.
- Call your health care professional if none of the above methods work to unclog your tube.

The tube should flush with some resistance, but you should be able to give feedings, water flushes, and dissolved medicines without difficulty.

### SKIN IRRITATION AROUND THE TUBE

- › If the area is red and sore, clean it with soap and warm water. Rinse around the area with plain water and pat dry. You may use an antibiotic ointment around the site.
- › If any areas appear crusty, gently soak or scrub the crusty areas with soap and warm water on the skin and tube itself. If you prefer, you may use a solution of ½ hydrogen peroxide and ½ water applied with a cotton swab to help clean these areas. After cleaning, rinse with plain water and pat dry. You may use an antibiotic ointment around the site.

## Improving Nutrition With Tube Feedings

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### FEELING FULL IN THE MORNING — UNABLE TO EAT BREAKFAST

- › Wait about 2 hours after you stop your tube feeds before eating breakfast.
- › Slow down the rate and start the tube feedings earlier in the evening, lengthening the time of the feedings.

### HOW QUICKLY WILL I GAIN WEIGHT?

You will work with your dietitian and CF team to set calorie and weight goals. These should be based on your daily schedule and your overall health goals. Unfortunately, when you lose weight you may be losing muscle along with body fat. Safe weight gain should include muscle gain and not just body fat gain. This will happen best when you continue to exercise and maintain muscle with activity.

### WORK WITH YOUR CF TEAM

Everyone plays their part in answering questions and helping you through this process:

- › Your physician can discuss the benefits and overall health goals, and can also direct the kind of tube placement you may need.
- › Your nurse can assist you with equipment and help troubleshoot any problems. They will help educate you on the care of your tube. They can also help you with insurance questions.
- › Your dietitian will help you find the correct formula, calorie level, and schedule for you. They can also help educate you on the care of your tube.
- › Your social worker helps with insurance questions and can discuss your feelings about this process.

### WILL INSURANCE COVER ANY COST?

Working with your CF care team and knowing what your health insurance plan will cover is key. Some insurance companies cover the equipment that is needed but not the tube feeding product. This will need to be taken into consideration as you are deciding whether a feeding tube is right for you.

# Improving Nutrition With Tube Feedings

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## HOW CAN MY FAMILY AND FRIENDS HELP ME?

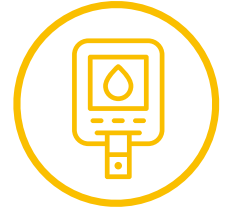
Education for your family and friends on the entire process will help you become successful. When others understand why you are making this decision, it is easy for them to help.

Your team is happy to educate anyone who will be instrumental in your care. Making the decision to have a feeding tube placed is a personal decision. Information and assistance from your CF care team will help you make the right decision to improve your health.

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# CF-Related Diabetes (CFRD)

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You've likely heard that cystic fibrosis often comes with its own version of diabetes — CF-related diabetes (CFRD). And you might have thought, hey, CF is complicated enough on its own.

But the truth is, CFRD can be managed if you are smart about it. If you recognize and deal with CFRD early, your overall health will improve and you will feel better. Being smart about diabetes starts with being smart about screening and diagnosis. We've put together these links to help.

## IN THIS SECTION:

- ▶ **Learn about CFRD** — the common signs and how it develops.
- ▶ **Work with your CF team** to screen for and diagnose CFRD.
- ▶ **Know what to do if you've been diagnosed with CFRD.**
- ▶ **Know the CFRD terms** — a glossary with helpful definitions.
- ▶ **Living with CFRD** — hear from an adult with CFRD.

# CF-Related Diabetes

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## LEARN ABOUT CFRD

Cystic fibrosis-related diabetes (CFRD) occurs in approximately 20 percent of teens and 40 to 50 percent of adults with CF. Women are diagnosed with CFRD more frequently than men. And while CFRD has some features in common with type 1 diabetes (usually childhood onset) and type 2 diabetes (usually adult onset), it also has important differences that require a different management approach. So, it's good to know the specifics of CFRD.

Let's start with the most common warning signs of diabetes in CF:

- › Having to urinate (pee) more often
- › Being very thirsty and needing to drink often
- › Feeling very tired
- › Losing weight without trying or having a hard time gaining weight
- › Losing lung function that you and your CF care team can't explain

If you are having a hard time keeping your weight up, have lost weight without explanation, or have an unexplained decline in lung function, you should talk with your CF team. They may consider screening you for diabetes.

## WHAT EXACTLY IS CFRD?

CFRD is primarily caused by not having enough insulin. Insulin is a hormone that helps your body metabolize food nutrients, so it can absorb all of the calories from the carbohydrates, protein, and fat in the food you eat. Insulin is made in the pancreas in special cells called beta cells.

CF can cause the cells in the pancreas to scar and not make insulin normally. When there is not enough insulin available, people lose weight without trying (mostly muscle and fat stored in the body). Lack of insulin can also make lung function worse as high blood sugars can make lungs more inflamed. Be on the lookout for weight loss, since unexplained weight loss and a drop in lung function have been shown to occur 6 to 24 months before being diagnosed with diabetes.

The other symptoms (increased thirst, urinating more often, feeling tired) are caused by high blood glucose levels. It's easy to miss these symptoms because people with CF often drink more (and then use the bathroom more) because of dry mouth.

Undiagnosed and untreated diabetes may shorten survival. Fortunately, treatment with insulin reverses muscle loss and helps with weight gain. It also improves survival.

## CF-Related Diabetes

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### MYTH BUSTERS

#### **“I can prevent CFRD if I watch my diet and don’t eat sugar.”**

CFRD is not like type 2 diabetes, which typically runs in families and occurs more commonly in people who are inactive and overweight. People with a family history of type 2 diabetes can prevent or delay diabetes by being active, eating healthy, and keeping their weight in a healthy range. CFRD is not caused by diet, weight, or lifestyle, and cannot be prevented. This is why you need regular screening to detect CFRD as early as possible, before it can cause problems with your weight and lung function.

### WORK WITH YOUR CF TEAM

#### YEARLY SCREENINGS — TAKING THE ORAL GLUCOSE TOLERANCE TEST

It’s crucial that you work with your CF team to get tested every year for CFRD. Early diagnosis and treatment will improve your overall health, your nutritional status, your lung function, and your survival.

The best way to diagnose CFRD is an oral glucose tolerance test (OGTT). This screening test is usually started at age 10 and done once a year when your health is stable (that is, you are not sick with a flare-up).

The OGTT is done in the morning. You’ll fast (no food or beverages, except water) for at least 8 hours beforehand. You should eat your usual diet for 3 days before the test. You will be given a sweet beverage to drink containing a high carbohydrate load (high glucose) dissolved in water. Then you’ll sit or lie quietly for 2 hours. Your blood sugar will be measured before you drink the beverage and again 2 hours later.

Your CF team will make a diagnosis of CFRD when:

- › Your fasting blood glucose (sugar) is more than or equal to 126 mg/dL on two separate days.
- › A blood glucose taken at any time of day is greater than or equal to 200 mg/dL and you have some of the warning signs listed in “What Exactly Is CFRD?”
- › Your 2-hour OGTT blood glucose is greater than or equal to 200 mg/dL.

## CF-Related Diabetes

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### OTHER TIMES TO SCREEN — WHEN YOU'RE SICK OR PREGNANT

CFRD can also be diagnosed if you become sick, are on IV antibiotics, are taking steroids, or are pregnant. These conditions cause the body cells to become resistant to the action of insulin, not allowing it to be absorbed into the cell. This is called “insulin resistance.” Insulin resistance, combined with not enough insulin being made by the body, causes blood glucose to rise. CFRD can also be diagnosed when people with CF are on continuous tube feedings.

### SCREENINGS WHEN YOU ARE SICK

When you are sick and in the hospital, your blood glucose is often checked for the first 48 hours after admission. It will be checked when you are fasting (before you eat) and 2 hours after you eat.

If you are getting IV antibiotics or high-dose steroids at home, you should check your blood glucose both when fasting and after meals with a blood glucose meter as instructed by your health care team. They would have to confirm a diabetes diagnosis by a lab test at the hospital.

### SCREENINGS RELATED TO PREGNANCY

If you are planning on becoming pregnant, your CF team or your obstetrician may recommend an OGTT to rule out diabetes first. If you should become pregnant and haven't had an OGTT within the last 6 months, you should have an OGTT early in your pregnancy. If it is negative for diabetes, you should have another OGTT at 12 to 16 weeks and again at 24 to 28 weeks. After delivery, you should have your blood glucose checked again within 6 to 12 weeks with an OGTT. Your obstetrician may be the one who orders these screening tests. If so, have the results — whether they are normal or abnormal — sent to your CF care team.

### OTHER TIMES TO SCREEN FOR CFRD

High blood glucose can occur with the use of high-calorie tube feedings. So people who need tube feedings should have their blood glucose checked every so often, either at home with a glucose meter or while in the hospital during the tube feeding, to check for CFRD.

People without CFRD who are listed for a lung transplant and have not been screened for CFRD within the last 6 months should be screened. Blood glucose should be monitored after transplant while in the hospital. After being discharged, people who have had a transplant should have regular screening every year for CFRD if they have no symptoms.

## CF-Related Diabetes

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### FAQS ABOUT DIAGNOSING CFRD:

#### › Can CFRD be diagnosed with a hemoglobin A1c (HbA1c) test?

The HbA1c test is not a good screening test because it may miss people who have diabetes. The test is used in people with CFRD to follow how well they are doing with the management of their blood glucose.

#### › Can diabetes be diagnosed with a fasting blood glucose test (FBG)?

An FBG finds people with CFRD who have high fasting blood sugars but does not find people with CFRD who may have normal FBG but very high blood glucose after eating. This is why an OGTT is recommended. It is the most accurate test for diagnosis of CFRD.

#### › Can CFRD be diagnosed using other tests?

Other tests such as fructosamine, urine glucose, and random glucose levels are not as accurate as an OGTT to test for CFRD.

#### › Can I wear a continuous glucose monitor (CGM) to diagnosis diabetes?

A CGM is not a tool that can be used to diagnose diabetes. No studies have shown that a CGM can give your doctor the information needed to diagnose CFRD in the way that the OGTT does.

## KNOW WHAT TO DO IF YOU'VE BEEN DIAGNOSED WITH CFRD

### JUST ONE MORE THING TO MANAGE

You have handled lots of challenges up to now, and you will handle this one too. Remember, you can look forward to feeling better and having better control of your lung disease with good CFRD management. Rely on your CF team to help you. You will want to get to know the endocrine team (diabetes specialists) and begin the important process of learning how to manage your blood sugars with medication — usually insulin — and your diet.

If you have CFRD, every 3 months you should see an endocrinologist and a multidisciplinary team with expertise in diabetes. There should be good communication between the diabetes team and your CF team.



## CF-Related Diabetes

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### MAKE THE LINK

If you have just been diagnosed with CFRD, this manual of care should be the FIRST publication that you study to learn about how to manage CFRD: [Managing Cystic Fibrosis Related Diabetes \(CFRD\), An Instruction Guide for Patients and Families](#). Your endocrine and CF teams may also have a copy of this resource for you.

You can also learn more about CFRD in this CF Education Webcast:

#### › [Diagnosis and Screening](#)

### WHAT ELSE YOU CAN DO

Make a list of questions to ask your endocrine care team when you have your first appointment with them. Here are a few you may want to think about:

- › Is my insurance going to cover my new medications and supplies for testing?
- › I love sweets! Is this the end of being able to eat them?
- › I have heard about insulin pumps. Would a pump be right for me?

### ADDITIONAL RESOURCE:

Moran A, Brunzell C, Cohen R, et al. Clinical Care Guidelines for Cystic Fibrosis-Related Diabetes. A position statement of the American Diabetes Association and a clinical practice guideline of the Cystic Fibrosis Foundation, endorsed by the Pediatric Endocrine Society. *Diabetes Care*. 2013 33, (12): 2697-2708.

### KNOW THE CFRD TERMS

Here are definitions of some common terms that will help you to understand what your CF doctor is telling you about your CFRD test results. You may want to print these definitions for reference when you talk with your doctor.

### DEFINITIONS

**CFRD with fasting hyperglycemia:** A “fasting” blood glucose level of at least 26 mg/dL (7.0 mmol/L) and/or a blood glucose 2 hours after an oral glucose load of at least 200 mg/dL (11.1 mmol/L) during an OGTT. This type of CFRD will likely cause the classic diabetes symptoms and is always treated with insulin. Now referred to as CFRD.

## CF-Related Diabetes

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**CFRD without fasting hyperglycemia:** A “fasting” blood glucose less than 100 mg/dL (5.6 mmol/L), but when the 2-hour blood glucose after an oral glucose load during an OGTT is at least 200 mg/dL (11.1 mmol/L). You may or may not have classic diabetes symptoms with this type of diabetes. This type of CFRD is always treated with insulin. Now referred to as CFRD.

**Fasting blood glucose (FBG):** A blood glucose level measured after not eating any food and not drinking for 8 hours. Normal FBG is less than 100 mg/dL (5.5 mmol/L).

**Hemoglobin A1C (HbA1c):** This test shows how much sugar is bound or “stuck” to your red blood cells. It shows the average blood level over the preceding 3 months. It is not as accurate of a test to diagnose CFRD as the OGTT; however, if your HbA1c is 6.5 percent or greater, you have CFRD.

**Indeterminate glucose tolerance/indeterminate glycemia (INDET):** A blood glucose level measured in the middle of an OGTT that is at least 200 mg/dL (11.1 mmol/L) in the presence of normal fasting and 2-hour blood glucose levels. Indeterminate glucose tolerance can also be detected when testing blood glucose with a glucose meter randomly at home or by using a continuous glucose monitor system (CGMS).

**Impaired fasting glucose (IFG):** A fasting blood glucose that is above normal; 100-125 mg/dL (5.6-6.9 mmol/L).

**Impaired glucose tolerance (IGT):** A fasting blood glucose of 100-125 mg/dL (5-6.9 mmol/L) and/or a blood sugar of 141-199 mg/dL (7.8-11.1 mmol/L) 2 hours after an oral glucose load during an OGTT.

**Insulin insufficiency:** When the pancreas doesn’t make enough insulin.

**Insulin resistance:** A condition in which the pancreas produces insulin, but the cells in the body cannot not absorb the insulin properly; therefore, the blood glucose is high. More insulin is needed to lower blood glucose.

**Oral glucose tolerance test (OGTT):** This test is used to diagnose not just CFRD but also the different types of abnormal glucose tolerance in CF. You must fast (nothing to eat or drink) for 8 hours. Your blood is then drawn to measure your “fasting” glucose level. You are then asked to drink a glucose-containing beverage. Your blood glucose is measured again 2 hours later. Sometimes, glucose is measured at 1, 2, and 3 hours later. If your blood glucose after 2 hours is 200 mg/dL (11.1 mmol/L) or higher, you have diabetes.

**Postprandial glucose level:** Blood glucose level 2 hours after a meal.

## CF-Related Diabetes

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### PUTTING THE DEFINITIONS TO WORK

Now let's use some of these definitions to understand the different levels of glucose tolerance that usually lead to CFRD.

### ABNORMAL GLUCOSE TOLERANCE

Few people with CF have "normal" glucose tolerance. Glucose tolerance abnormalities can be thought of as the following:

- › Impaired fasting glucose (IFG)
- › Impaired glucose tolerance (IGT)
- › CFRD without fasting hyperglycemia (CFRD FH-)
- › CFRD with fasting hyperglycemia (CFRD FH+)

Many people have normal fasting and 2-hour blood glucose levels, but they have a high blood glucose in the middle of the OGTT. This is called indeterminate glucose tolerance or indeterminate glycemia (INDET). INDET can also be seen when testing blood glucose with a glucose meter randomly at home or by using a continuous glucose monitor system (CMGS).

What does impaired glucose tolerance and indeterminate glucose tolerance mean in CF? We don't know. They are common and they measure mildly abnormal glucose tolerance. In children with CF before puberty, both of these have been linked with an early onset of CFRD. Everyone with abnormal glucose tolerance should have an OGTT every year or have it done earlier if symptoms occur.

### LIVING WITH CFRD

Hear from an adult with CFRD.

"Are you kidding me? You want me to manage another chronic illness on top of my CF?" Those were my thoughts after I was given the diagnosis of CFRD more than 25 years ago.

It's not easy to manage CFRD, especially at the beginning. But, as you learn how your body responds to insulin, the payoff in terms of better health and improved energy are worth it!

Before my diagnosis of CFRD, I had lost a lot of weight and couldn't gain it back despite all I ate; my lung function had also dipped. I had to stay off work because I had no energy and I wasn't healing. All of that changed when I started insulin therapy — it was a wonder for me. I began to gain back the weight I had lost, felt more energized and less tired, and I started to heal. I went back to work and resumed my life, while managing CFRD.

## CF-Related Diabetes

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### HERE'S WHAT I'VE LEARNED:

- › **Accept the diagnosis.** Being in denial about CFRD doesn't make it go away. Learn about the basics of managing insulin, food, and exercise. Like any skill, managing CFRD is difficult and awkward at first, but you get better at it with practice. It will become second nature. I continue to learn about how my body responds to insulin at different times. Rather than get frustrated, I remember that "it's hard to be a pancreas!" When I'm successful at figuring out how much insulin to take with a meal, making adjustments for my blood glucose level and exercise plans, I feel like a winner because my blood glucose is in the normal range.
- › **Accept help and support from your CF and endocrine teams.** Find a diabetes nurse educator you can work with. They are the ones who can really help you with the nitty-gritty of CFRD. They can also help you troubleshoot when you're having a hard time getting things right for you. Like CF, CFRD requires a lot of self-management and discipline. But because of our CF, we've already been refining those skills. Seek out other people who may be managing CFRD or type 1 diabetes for support. They can share helpful tips about managing diabetes daily. I was fortunate to have a very close friend with type 1 diabetes who showed me the ropes. She helped me fit diabetes into my life and not turn my life upside-down for diabetes.
- › **Be prepared to handle times of low blood sugars.** Always carry a simple carbohydrate juice or candy snack. It's easy to slip candy or small packets of fruit snacks into a pocket or purse. I usually carry a small juice box in my purse or knapsack, too: I think I'm the only adult who sometimes drinks from a juice box! Even if I'm going to take a walk around the block, I stick something in my pocket just in case my blood sugar gets low. By always having something with me, I don't get stressed if I need to treat a low blood sugar.
- › **Don't be afraid.** Testing your blood sugar doesn't have to fill you with dread. The meters for testing are small, silent, and quick. It hurts less if you prick the sides of your fingers (not the fleshy part of your fingertip) where there are fewer nerve endings. Plus, the amount of blood needed for testing keeps getting smaller and smaller. Taking insulin is very different from getting flu shots or other typical immunizations. The needles are very thin and short. They are made to go just under the skin (subcutaneous or 'sub q'). I can honestly say that I barely feel the needle. There are also insulin pumps and pens that may be easier and more discreet.

## CF-Related Diabetes

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- › **Take good care of yourself.** Although CFRD has an impact on daily life, it doesn't have to limit what you want to do. I still work, go out to eat with friends, travel, run and bicycle, and eat all sorts of foods. CFRD just adds another layer of things I need to do to live life to the fullest. I do what I have to do to stay healthy so that I can do the things I want to do in my life. By taking care of my CF and CFRD, I can have the best lung health and energy level and work to reach my goals.

Don't let CFRD stand in your way — you can manage it successfully!

You can hear from another adult living with CFRD in a CF Education Webcast:

- › [Living With CFRD](#)

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# Basics of Infection Control

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As a person with cystic fibrosis, your lungs are more prone to chronic infections because of thick and sticky secretions, which allow germs to thrive and grow. So, prevention of infections is a top priority for your lung health. Viruses, bacteria, and fungi (molds) are three categories of germs that can cause respiratory problems in people with CF. People with CF can also spread germs to each other.

You can help reduce your risk of exposure to germs — by being consistent with your daily treatments, washing your hands carefully, properly cleaning your respiratory equipment, and avoiding direct contact with others with CF.

Thinking ahead and being prepared can help you handle social situations that may put you at risk for being exposed to more infection.

## IN THIS SECTION:

- ▶ **Learn about controlling infection** to protect yourself and others.
- ▶ **Follow recommended ways to limit germs.**
- ▶ **Work with your CF team** to make your health care as safe as possible.
- ▶ **Manage sensitive social situations** by following these tips.
- ▶ **Get help when you need it** — and learn what you can do to avoid getting or spreading germs.

# Basics of Infection Control

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## LEARN ABOUT CONTROLLING INFECTION

### BASICS OF CONTROLLING INFECTION

Bugs (such as bacteria) find a good place to live in the lungs of people with CF. They come from lots of places in the world you live in, including warm, damp dirt; around and in water; and through close contact with other people.

You can't control everything or go live in a bubble, but you should try to control these important things:

- › Keep bugs away from your nose and mouth by washing your hands **OFTEN** and avoiding touching your hands to your mouth and nose as much as possible.
- › Avoid close contact with others with CF.
- › Regularly and carefully clean the nebulizer equipment you use to put medications into your lungs.

You probably also want to know how to best handle some, shall we say, delicate situations like being out on a date. A couple of quickies to remember:

- › You **CAN'T** get these bugs from kissing someone who doesn't have CF.
- › If you kiss someone who has CF, you **CAN** give your bugs to them and you can get their bug.

So be careful who you kiss ... or share food and drink with ... or get close to.

You may also want to know how to handle some common life situations. Let's say you have an office mate who has a cold and is coughing. You can get [information from the Centers for Disease Control and Prevention](#) (CDC) or a [poster from the Cystic Fibrosis Foundation](#) on how to cover your cough and stop the spread of germs. Try displaying the chart or poster in the restroom you and your co-worker use. Put hand sanitizer and tissues on prominent display on your desk. If you feel comfortable, let your office mate know that respiratory viruses are harder for you to handle than the average Joe and ask him or her to help you — by limiting personal contact and by using email or texting with you for at least three days when he or she has a cold.

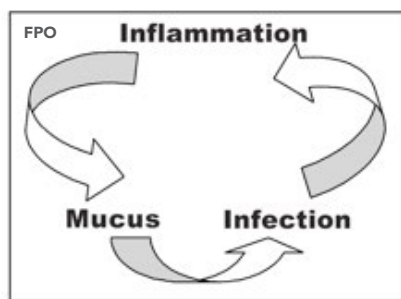
## Basics of Infection Control

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### WHY ARE PEOPLE WITH CF MORE SUSCEPTIBLE TO GERMS?

A gene mutation causes a problem with the **CFTR protein**. This protein helps your body move fluids in and out of the cells. This movement helps regulate the fluids for digestion, sweat, and mucus. When CFTR is not working right, the mucus in your lungs does not have enough fluid for it to flow and move out of your body. In CF, this is what causes the mucus to become thick and sticky (think paste vs. slime).

This thick mucus then interrupts one of the normal ways the lungs stay clean. It pushes down on cilia, hair-like projections on cells in your nose and lungs that normally trap and sweep away debris out of the lungs. So the mucus, combined with the interruption of the cilia's job of cleaning, creates a perfect environment for germs to thrive and thus for infections to grow.



The infections that grow in this environment are chronic (or long-lasting). As the germs multiply, your body tries to fight the infection by sending white blood cells to the lung. This creates inflammation, which leads to swelling and increased secretions and mucus. Inflammation and chronic infections lead to scarring and changes in the lungs (the medical term for this is bronchiectasis).

This process repeats itself in CF and is called the “vicious cycle” of infection and inflammation.

Unfortunately, bacterial infections are the major cause of lung health getting worse in people with CF. At this time, the best therapy is prevention or infection control.

### HOW DO I COME IN CONTACT WITH THESE GERMS IN THE FIRST PLACE?

There are three ways that germs get picked up: contact, droplet, and airborne transmission. The two we are most concerned with in CF are contact and droplet.

- › **Contact transmission** is the most frequent type. It includes both direct contact and indirect contact. Common germs that are spread through contact transmission include *Pseudomonas aeruginosa* and *Burkholderia cepacia* complex.
- › **Droplet transmission** is when germs are passed short distances through coughing, sneezing, or talking. An example of a germ spread by droplet is seasonal influenza (the flu).



## Basics of Infection Control

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- › **Airborne transmission** is when germs are carried through the air on dust particles and are inhaled. Tuberculosis and *Aspergillus* are spread this way.

For more details on how germs spread, visit [CFF.org](https://www.cff.org).

### WHICH GERMS SHOULD I BE WORRIED ABOUT?

Respiratory germs that can cause respiratory infections in CF fall in three categories:

- › **Bacteria** are microorganisms that are all around us (in soil, water, on our skin, on surfaces, etc.). Most are harmless and not a threat to your lungs. But some are common in CF and can cause chronic problems. These include *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and *Burkholderia cepacia* complex
- › **Respiratory viruses** are microorganisms that multiply inside our living cells. Examples are influenza (the flu) and rhinovirus (the common cold). Viruses are often seasonal and are spread between people.
- › **Fungi or molds** are sometimes also called yeast. One example of fungus seen in CF lung disease is *Aspergillus fumigatus*.

Many germs are especially dangerous for people with CF and may lead to a faster decline in lung function. Medical studies show that people with CF are at particular risk of spreading certain germs among others with the disease. This is called cross-infection.

Some of these germs include:

- › ***Pseudomonas aeruginosa*** (*P. aeruginosa*)
- › **Methicillin-resistant *Staphylococcus aureus*** (MRSA)
- › ***Burkholderia cepacia* complex** (*B. cepacia*)
- › ***Aspergillus***
- › **Nontuberculous mycobacteria** (NTM)
- › **Influenza**, commonly called the flu

You can learn more about these germs [here](#) on CFF.org.

## Basics of Infection Control

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### I WAS TOLD I HAVE A MULTIPLE-DRUG-RESISTANT ORGANISM.

#### WHAT DOES THAT MEAN?

Multiple-drug-resistant organisms (MDRO) in CF are bacteria that have become resistant to multiple types of antibiotics.

Resistance means that the antibiotic does not work to kill or weaken the bacteria. Some bacteria are naturally resistant to antibiotics. For others, resistance happens when the bacteria is exposed to antibiotics over time. With each exposure, the bacteria can make changes to outsmart the antibiotics.

You can also get this type of germ by coming into contact with a person infected with the resistant bacteria or equipment that has been contaminated (has the germ on it). Drug-resistant bacteria can be a problem in CF, as they can make treating your lung infections and exacerbations more difficult.

This brings us to a very important point: The misuse of antibiotics is one of the leading causes of antibiotic resistance. So when you are prescribed antibiotics, you need to take the prescription as it was ordered and finish it unless you are told otherwise by your health care provider. Sometimes we forget or we start to feel better and decide we'll save the antibiotic for "when we really need it." But that is never a good idea! It can lead to the antibiotic not working any more to fight infections.

### FOLLOW RECOMMENDED WAYS TO LIMIT GERMS

#### INFECTION CONTROL IN DAILY LIFE

Infection control extends to all areas of your life, such as your home, your community (work, school, shopping), the outpatient clinic, and the hospital when you're admitted. It's also important to remember that you can get germs from others who don't have CF.

Viruses and bacteria that are common to many people can be especially hazardous to a person with CF. Good hygiene habits at home or work can help keep you at your healthiest.

## Basics of Infection Control

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### THE CENTERS FOR DISEASE CONTROL AND PREVENTION (CDC) SAYS YOU SHOULD CLEAN YOUR HANDS...

- › Before, during, and after preparing food
- › Before eating food
- › After using the toilet
- › After changing diapers or cleaning up a child who has used the toilet
- › After blowing your nose, coughing, or sneezing
- › Before and after your inhaled medicines and airway clearance
- › Before and after caring for someone who is sick
- › After touching an animal or animal waste
- › After touching garbage
- › Before and after treating a cut or wound

### WHAT IS THE RIGHT WAY TO WASH YOUR HANDS?

- › Wet your hands with clean, running water (warm or cold) and apply soap.
- › Rub your hands together to make a lather and scrub them well; be sure to scrub the backs of your hands, between your fingers, and under your nails.
- › Continue rubbing your hands for at least 20 seconds. Need a timer? Hum the "Happy Birthday" song from beginning to end twice.
- › Rinse your hands well under running water.
- › Dry your hands with a clean towel or air dry them.
- › Turn off the water with a paper towel, then throw it away.

### TIPS FOR REDUCING INFECTION:

- › Keep up with your daily treatments:
  - Do your daily airway clearance.
  - Take your medications to help clear secretions by either thinning them with dornase alfa (Pulmozyme®) or rehydrating them so they are easier to cough out with hypertonic saline. Bacteria need mucus in order to thrive and grow. When you clear mucus out, you slow down the process!

## Basics of Infection Control

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- Inhale your prescribed antibiotics deeply to fight infection and decrease the amount of bacteria in your lungs, for instance tobramycin or aztreonam (TOBI<sup>®</sup>, Cayston<sup>®</sup>).
- Take your medications that reduce the amount of inflammation or swelling in your lungs (ibuprofen, azithromycin).
- › Take antibiotics as prescribed. These are medications that kill or slow the growth of certain germs.
- › Clean and disinfect or sterilize respiratory equipment properly.
- › Keep hands clean — wash them! — at home and in public places.
- › Avoid sharing items that can have germs from household members, whether or not they have CF. This includes toothbrushes, eating utensils, drinking cups, and respiratory equipment.
- › Get your yearly flu shot and other immunizations including pneumococcal and pertussis (whooping cough).
- › Remind your loved ones, close contacts, and caregivers to keep up with their yearly flu shots and other immunizations too!
- › Avoid using humidifiers or misting systems in your home. If not disinfected well, these systems can be a great place for bacteria and fungus to grow.

### MYTH BUSTERS

- › **Myth:** Using vinegar solution is a good way to clean your respiratory equipment.
- › **Fact:** Vinegar is not effective in killing the germs most common in CF. Always follow the CF Foundation guidelines in your daily [cleaning of respiratory equipment](#).
- › **Myth:** If others aren't sick, they can't spread germs.
- › **Fact:** Even if a person isn't visibly ill, they can still spread germs to other people.

# Basics of Infection Control

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## ADDITIONAL RESOURCES

CF Foundation webcasts and information on infection control:

- › **Infection Prevention and Control Clinical Care Guidelines:**  
[CFF.org/For-Caregivers/Clinical-Care-Guidelines/Infection-Prevention-and-Control-Clinical-Care-Guidelines/Infection-Prevention-and-Control-Clinical-Care-Guidelines/](https://www.cff.org/For-Caregivers/Clinical-Care-Guidelines/Infection-Prevention-and-Control-Clinical-Care-Guidelines/Infection-Prevention-and-Control-Clinical-Care-Guidelines/)
- › [www.youtube.com/playlist?list=PLhoQ6vyZhggqs0rosMBCZFQBpY02yTAK4](https://www.youtube.com/playlist?list=PLhoQ6vyZhggqs0rosMBCZFQBpY02yTAK4)

## FOR MORE INFORMATION ON HAND WASHING:

- › [www.cdc.gov/cleanhands/](https://www.cdc.gov/cleanhands/)

## FOR MORE INFORMATION ON IMMUNIZATIONS:

- › [www.cdc.gov/vaccines/](https://www.cdc.gov/vaccines/)

## WORK WITH YOUR CF TEAM

### INFECTION CONTROL IN THE HOSPITAL OR CLINIC

Though we are not always sure how or where a person becomes infected with a germ, we do know that germs in the respiratory sputum of those with CF can be passed on to others. We know this by doing a test called molecular typing. Bacteria have their own “fingerprints,” and this testing can actually look at the DNA found in bacteria from two patients to see if their bacteria are from the same source.

This testing has been done in the past when there have been outbreaks of certain



strains of bacteria in camps, social gatherings, and hospital and clinic settings. Because of this testing, we know that hospitals and clinics can be a place for bacteria to spread. Health care workers and equipment can be a way for germs to get from one patient to the next.

# Basics of Infection Control

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## TIPS FOR STAYING PROTECTED IN THE HOSPITAL OR CLINIC

- › **Keep your hands clean.** Make sure you have alcohol-based antibacterial hand gel readily available when in public places, especially in common areas such as the clinic waiting room or the sign-in and checkout desk.
- › **Avoid indirect or direct contact with others with CF.** This includes spending limited time in common areas — the waiting room, check-in and check-out areas, and the clinic bathroom.
- › **Use disposable tissues** to keep secretions to yourself when coughing or blowing your nose. Keep tissues handy in your purse, pocket, or backpack, then clean your hands.
- › **Go to your CF center** at least once every 3 months **for regular follow-ups** and to have regular sputum cultures done.
- › **Understand and follow your CF center's infection control practices.**

## SOME PRACTICES AT CF CARE CENTERS FOR INFECTION CONTROL:

- › Hand cleaning before contact with all patients
- › Gloving for all patients
- › Gowning with all or some patients
- › Cleaning the clinic or hospital room after patients are discharged home
- › Creative scheduling to reduce wait time in clinic
- › Scheduling patients with certain germs in separate clinics
- › Eliminating or limiting face-to-face socializing between patients with CF while in the clinic or hospital and at camps and education days
- › Having all patients use respiratory masks when in the clinic or on hospital grounds

## Basics of Infection Control

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### IF YOU DON'T HAVE SOAP AND WATER:

The Centers for Disease Control and Prevention (CDC) says that washing hands with soap and water is always the best way to reduce germs. But if that's not possible, use an alcohol-based hand sanitizer

(at least 60 percent alcohol), which can quickly reduce — but likely not eliminate — the number of germs on hands. Note also that hand sanitizers are not effective when hands are visibly dirty.

### HOW TO USE HAND SANITIZERS:

- › Apply the product to the palm of one hand.
- › Rub hands together, and rub the product over all surfaces of your hands and fingers until hands are dry.

For more information on hand washing, see:  
[www.cdc.gov/handwashing/](http://www.cdc.gov/handwashing/)

### MYTH BUSTER

- › **Myth:** There is too much of a fuss made about CF infection control. Hanging out with my friends with CF won't hurt anything.
- › **Fact:** There is documented evidence that germs spread between people with CF. Some of these cases have led to a serious health decline of the people involved. Using other ways to communicate — over the internet and by email or telephone — with your friends with CF will help protect you and your friends from getting or spreading germs to others with CF.

# Basics of Infection Control

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## HOW DO I KNOW WHAT INFECTION CONTROL PRACTICES MY CF CENTER FOLLOWS?

The CF Foundation has infection control recommendations that list ways to minimize the spread of germs. Even with these guidelines, some practices may be slightly different from center to center.

One example would be having all people with CF use a mask when on hospital grounds. This has been widely debated, and some centers may enforce this rule, while others may not. If you have questions or concerns about the infection control practices at your center, discuss them with your doctor, nurse, or care team member. Remember, it's OK to ask questions!

Many CF centers also have a parent/patient advisory council that may be able to help with your concerns. You may even want to consider getting involved with the work they are doing.

## MANAGE SENSITIVE SOCIAL SITUATIONS

You may have concerns about how to handle social situations that may pose a risk to your health. Be prepared for these situations so that when the time comes you can pull it off without a second thought.

If you notice that a health care worker forgets to wash her hands or forgets to glove before touching you, you might say, "I'm sorry, but could you put on a pair of those gloves over by the sink while you examine me?" You could then add, "With all the patients you take care of every day, I'm sure it gets tiring to be constantly gloving and washing your hands." Or perhaps you notice not everyone is gowning and gloving when entering your room during your hospital stay. A gentle reminder is all that is needed and is much appreciated most of the time.

It's important to remember that the goal of infection control is to reduce the risk of exposure to germs in your environment, not eliminate it. There is a balance between ignoring infection control guidelines and the other extreme — becoming obsessed with germ control. Find a balance in your daily routine by laying down good infection control habits while still having an active lifestyle. That is ultimately the recommended approach.



## Basics of Infection Control

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### GET HELP WHEN YOU NEED IT

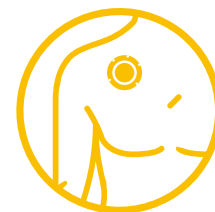
If you are not sure about how you can avoid germs in your everyday life:

- › Ask your doctor or nurse.
- › Watch the CF Education Day webcasts [on infection control, germs, and everyday life](#).
- › Read about how to avoid germs and the spread of germs on the CF [Foundation's website](#).
- › Visit the Centers for Disease Control and Prevention's website and read about how to be [a germ stopper](#).
- › Watch a [video](#) to see how germs spread and how to avoid them.
- › Watch the Share With Those Who Care video: [www.youtube.com/watch?v=8K\\_O-7CWa64](http://www.youtube.com/watch?v=8K_O-7CWa64)

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# Vascular Access Devices (a.k.a. PICCs/Ports)

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Sometimes you need antibiotic treatments for more than just a few days in a row (like during an exacerbation). Regular IVs can cause irritation to the veins, can get infected, or can fall out after a few days.

That's where vascular access devices (VADs) come in. They're designed to be used for days, weeks, or months; they are made of material that doesn't get contaminated with germs too easily; and, if treated with respect, they won't fall out.

Read on to learn more about the different kinds of VADs.

## IN THIS SECTION:

- ▶ **Learn about VADs** — what the different types are and where they're placed.
- ▶ **Work with your CF team** to choose the right device for you.
- ▶ **Get help when you need it** so that your VAD doesn't cause complications.
- ▶ **Speaking from experience** — hear from others with CF.

## Vascular Access Devices (a.k.a. PICCs/Ports)

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### LEARN ABOUT VASCULAR ACCESS DEVICE

VADs are made of flexible, strong plastic tubes and come in many sizes. They can be split into channels (called “lumens”) to accommodate more than one drug, but for routine CF care, a single lumen is usually enough. That’s good because it means you’ll only need a small VAD. Although the complication rate is very low with these devices, they can become contaminated and can cause clots to form in the veins. Your CF care team should discuss these concerns with you before any device is placed.

### TYPES OF VASCULAR ACCESS DEVICES — EXPLORE THE DIFFERENCES

Although there are many different types and brands of VADs out there, the two main categories are based on how long the device will be left in place. Short-term VADs (usually left in a vein for weeks) are called “PICCs” and long-term VADs (usually months, often years) are called “ports.”

#### PICCs

Peripherally inserted central catheters, or PICCs, are intended for temporary use — usually for a number of weeks. They’re a good choice for people who need therapy only once or twice a year. You’ll typically receive one round of antibiotic treatments, then have the device taken out.

A PICC is about the same diameter and length as a piece of cooked spaghetti, and just as flexible. It’s inserted into a vein in the crook of your elbow or in your upper arm (with a short piece of catheter and an injection cap showing), then secured to your skin with an adhesive device, an internal securing devices or stitches. Sometimes it’s placed through a vein in the side of the neck or near your collarbone. The PICC is then covered by a small dressing. Afterward, you’ll probably have an x-ray taken to make sure the catheter is in the right place.

You may hear the term “midline” catheter. It is a type of catheter in the PICC family — it is a bit shorter and usually goes through a vein in the upper arms.

The dressing is important, because PICCs must stay clean and dry at all times (which can be a challenge in warm weather). You can do most normal activities (except swim) with a PICC by protecting the site with a plastic covering. You should also avoid any type of activity that might irritate or dislodge the catheter — very strenuous use of your arm, like weight-lifting or serving a tennis ball. You’ll get coached in how to keep the site dry and what activities to avoid.

The PICC must be flushed at regular intervals. This means a small volume of sterile fluid (like saline) is injected into the PICC to keep it flowing freely. Your CF health care team can show you how to flush the PICC.

## Vascular Access Devices (a.k.a. PICCs/Ports)

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Some patients find this hard to do. Don't worry, if you can't do it yourself, a nurse in the hospital or even a nurse who comes to your home can help you with flushing. Many patients want to learn to do this themselves to be as independent as possible — but you must learn to do this flushing in a very clean way to reduce the chance of any infection at the site of the catheter.

### PORTS

Ports stay in place even when you're not getting any IV antibiotics. They're hidden beneath the skin, with only a small bump to indicate where they are.

The procedure to put in a port is more invasive than the one used for a PICC but still quite simple and usually done as an outpatient. Typical places for ports include the upper chest area just below the collarbone and inside the upper part of your arm. The team placing the port will mildly sedate you, numb the area, make an incision about 1 inch long, then place the port in a skin pocket. Once it's healed, you'll have a bump and a small scar. If the port needs to be removed, the incision would be re-opened.

To start infusions, your team will swab an antiseptic over the port, then pierce the middle part just underneath the skin (called the "septum") with a special type of needle called a Huber needle. Once the needle is in place, you can receive medications or have blood drawn for up to a week before the needle needs replacing.

Ports are helpful because their placement under the skin means less maintenance, although you'll still have some: ports should be flushed every 4 weeks when not in use. In general, people with CF decide to get a port if they are needing frequent courses of IV antibiotics or if they have had a hard time having PICCs place in the past. Talk to your CF care team if you have questions or any curiosity about a port.

Once a port has been placed and the small incision has healed, there are no restrictions on your activities — you can swim, bathe, and do sports with no restrictions.

### PLACING THE VASCULAR ACCESS DEVICE

Different hospitals have different approaches to placing VADs. They are always placed under very clean ("sterile") conditions: your skin is cleaned carefully, the person placing the line will be wearing sterile gloves and a gown, and, of course, the catheter itself is germ-free. These lines can be placed by physicians, nurses, radiology technicians, physician assistants, or nurse practitioners — it depends on your hospital. Whoever places the line will have received special training in doing the procedure.

In some hospitals, PICCs can be placed at the bedside with local anesthesia to numb the skin. Other hospitals may take you to the radiology suite — the place where

## Vascular Access Devices (a.k.a. PICCs/Ports)

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x-rays are taken — and have the line placed there. Usually your veins are located by using an ultrasound machine or a fluoroscopy machine (x-ray projected onto a TV screen).

Ports are usually placed in the radiology department or in an operating room. A small incision, or nick, is made in the skin where the port is placed. Your skin will be numbed and you may receive some general sedative first. Rarely, general anesthesia is used. The end of the port is hidden beneath the skin, so while there may be a “bump” under the skin, the end of the catheter is not visible. It is a very simple procedure.

Once a port is placed, some special care of the site will be needed for a few days while the incision heals. Your care team will give you pain medication if it is necessary.

### WORK WITH YOUR CF TEAM

While we are talking about PICCs and ports together, let’s look at some of the differences.

The decision to place a PICC is a relatively easy one — if you need IV antibiotics to treat a CF exacerbation, you will likely get a PICC line placed. They are much more practical than regular IV catheters placed in the crook of the arm or the back of the hand. A PICC can be left in place for several weeks or even months, which may be necessary if you are on long-term antibiotics.

Your CF health care team may suggest a port if you need IV antibiotics frequently or if placing a PICC has been difficult because it is hard to locate a vein or the PICC has been painful (you may have heard someone describe that as a “tough stick”). One advantage of having a permanent device is that it only has to be placed once to be available over a long period of time when you need it (and there is little maintenance required between infusions).

Of course, take your time in deciding to get a port. It requires minor surgery and means there is a device hidden beneath your skin that stays in place, even when you are not receiving any medication through it. You may have some self-consciousness about how visible the small bump will be. Ask questions of your team. You may want to speak to a member of the team that will place the port.

There’s no right or wrong to this, so be realistic about how you feel. It’s your body and you can decide what device is best for you. If you need help making a decision, ask the team of specialists who place VADs to give you information about the procedures and share their experiences.

## Vascular Access Devices (a.k.a. PICCs/Ports)

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### GET HELP WHEN YOU NEED IT

Managing the upkeep of PICCs and ports — and monitoring them for complications — is generally simple but should be done regularly and with the help of your care team.

Both PICCs and ports need to be flushed with a sterile fluid like saline after an infusion of medication to prevent the catheter from getting plugged by a blood clot. The frequency of flushing will be up to your CF health care team, but you can expect it to be anywhere from several times a week (for PICCs) to about once a month (for ports). If you are currently receiving IV therapy, you or your nurses will flush after each dose of medicine.

Your VAD should never be painful or uncomfortable to use. If it is, notify your CF health care team immediately — it could be the beginning of an infection or clot. And remember: though your port will be sore for a few days after it's inserted, it should be free of discomfort after that.

Any swelling, redness, or puffiness around your VAD is not normal. Contact your health care team right away if you notice this because it could be an early sign of an infection or a clot. Do not try to treat this yourself with hot or cold packs.

Although rare, VADs can become contaminated with germs. Contact your care team if you have fever, chills, or other signs or symptoms of infection. Your team will need to sort out if your symptoms are related to the VAD or something else. They will probably draw blood and perform cultures to help figure this out.

Contact your CF health care team if you see these other problem signs:

- › **Leaking from the catheter or site.** Leaking may mean a hole or break in your tubing — or blockage in the catheter. Ask your CF health care team to examine your VAD, and know that it can always be exchanged for a new one (usually at the same site).
- › **Blood backing up in your infusion tubing.** This should not happen. If it does, there may be pressure at the catheter tip that can lead to clots.
- › **Resistance to flushing.** Feeling resistance may mean you've got a clot, which often can be treated at home or in the clinic using clot-busting drugs called thrombolytics.

### SPEAKING FROM EXPERIENCE

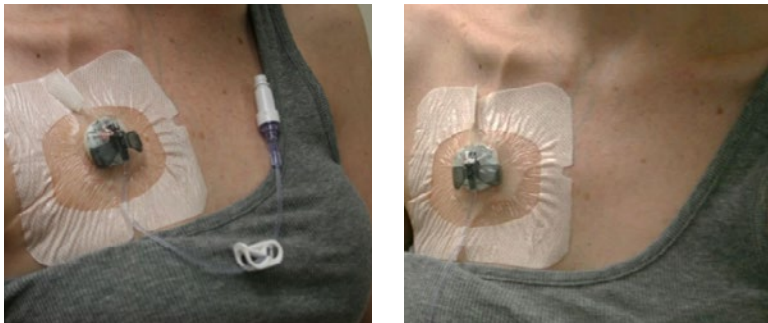
“The decision to have a port placed can be a difficult one. Every patient has to weigh the risks and benefits together with their family and their CF team. Ports are not for everyone. After many years of receiving PICC lines for my IV antibiotic cleanouts, my skin became very sensitive to the dressings and the skin cleaner. Even a 2-week course would leave my skin irritated. In addition, although we in the CF community are quite used to being poked and prodded, being stuck with a large-bore needle is never a fun experience.

“As my IV antibiotics were becoming more frequent and of longer duration, my wife and I made the decision to speak with the CF team about having a port placed. We discussed the risks, most notably of infection, and the benefits of having the procedure. For me, it was a clear choice to have the port. The procedure was completed, and I recovered with very little pain. The area where the port was placed was somewhat tender for a day or two only. After the site healed over, the only reminder I had that there was a port in my upper chest was the small bump I could see protruding when I looked in the mirror. I could not “feel” it at all. I received some teaching from my home health agency and then began accessing the port myself. I feel this is one of the main advantages of having the port.

“When it was time to start an antibiotic course, many times I would access the port myself at home after getting the delivery of supplies from the home health company without the need for having to go the hospital or have a nursing visit. Once a month, I would also have to access the port to do a saline flush to keep the port clear. Of course, like everything else with CF care, this requires a lot of compliance. Many CFers may feel intimidated by their own needle sticks, which is perfectly understandable, and will continue to need a nurse to help them access the port and change the dressings every week while on antibiotics.

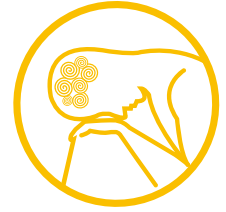
“I took my time to make the decision about a port and, of course, talked it over with my wife and the CF team. I have had the port for several years now and it has made my care easier and has helped maintain my independence. It was a good decision for me.”

– John M., Age 39



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# Anxiety and Depression



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Everyone feels sad, down or worried at some point. Feeling this way is a normal reaction to loss, life's struggles, or stress. And dealing with cystic fibrosis can be stressful.

But if these feelings become more than just a "bad mood" — if they last a long time, get in the way of your daily life and sleep, or just seem too much to handle — then it's time to ask for help.

Click on any of the "basics" below to get more in-depth information, tips, and additional resources.

## IN THIS SECTION:

- ▶ **Learn about anxiety and depression** — how it's often linked with CF and when it's time to ask for help.
- ▶ **Avoid substance abuse**, including drinking too much, taking illegal drugs, and even taking more medications than prescribed.
- ▶ **Work with your CF team** to receive effective treatments and make a plan.
- ▶ **Manage your time** with tips for staying in control of your life, and myth busters.
- ▶ **Get the help you need** with additional resources.



# Anxiety and Depression

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## LEARN ABOUT ANXIETY AND DEPRESSION

### DEPRESSION

It's more than just feeling sad. It may include:

- › Difficulty concentrating, remembering details, and making decisions
- › Feeling tired all the time and having low energy
- › Feeling guilty, worthless, or helpless
- › Feeling hopeless or negative
- › Trouble sleeping
- › Feeling irritable
- › Loss of interest in activities or hobbies that were once enjoyable
- › Changes in eating (overeating or loss of appetite)
- › Constant aches or pains, cramps or digestive problems that do not ease up
- › Constant sad or anxious feelings
- › Thoughts of suicide or suicide attempts

People with depression do not all experience the same symptoms. Some people may have all of the above symptoms while others may have only a few.

### WHEN TO ASK FOR HELP

Call your CF care team right away if you suspect you might be suffering from depression or are thinking about suicide.

### THOUGHTS ABOUT SUICIDE

Sometimes depression or anxiety can be too much to handle and you might start thinking about suicide. These thoughts are serious, and you need to discuss them with your health care provider:

- › **It's a signal you should ask for help.** Thinking about suicide may mean you have more pain and sadness than you can cope with and you need help. So ask for help. You don't need to feel this way.
- › **Talk it out with a person you trust.** Talk about your feelings and thoughts. Voicing concerns and getting support can help you find solutions. Call a trusted friend, family member, doctor, or therapist — or a member of your CF care team.

## Anxiety and Depression

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- › **Ask someone to help you set up a plan to keep yourself safe.** Consider making a contract with your doctor or therapist. Ask someone to help remove any dangerous objects or weapons from your home. Ask someone to take care of your medicines and give them to you one day at a time.
- › **If you do not know who to turn to:** Call the [National Suicide Prevention Lifeline](#) at **800-273-TALK (8255)** or the [National Hopeline Network](#) at **800-SUICIDE (800-784-2433)**. These toll-free crisis hotlines offer 24-hour suicide prevention and support. Your call is free and confidential.

### ANXIETY

We all have a lot of things to worry about in life. Anxiety is more than just being worried about regular stuff. At times, your worrying may get in the way of doing things you normally would do, or you may experience panic symptoms that begin to stop you from doing your normal activities.

Symptoms of anxiety can include

- › Too much worry most of the time for at least 6 months
- › Finding it difficult to control the worry
- › Feeling restless
- › Being easily fatigued
- › Difficulty concentrating
- › Muscle tension
- › Sleep disturbances

Sometimes anxiety can cause a panic attack. A panic attack is an event of extreme fear or discomfort that lasts more than 10 minutes and may include these symptoms:

- › Heart palpitations
- › Sweating
- › Trembling or shaking
- › Feeling short of breath
- › Feeling like you are choking
- › Chest pain or discomfort
- › Nausea or abdominal distress

# Anxiety and Depression

- › Feeling dizzy or lightheaded
- › Fear of losing control, going crazy, or dying
- › Numbness
- › Chills or hot flashes

## ANXIETY, DEPRESSION, AND CF

Anxiety and depression are common in CF and can disrupt your life. People with a chronic illness are more likely to have anxiety and depression than people without a chronic illness. In turn, anxiety and depression can then make it even harder to care for CF.

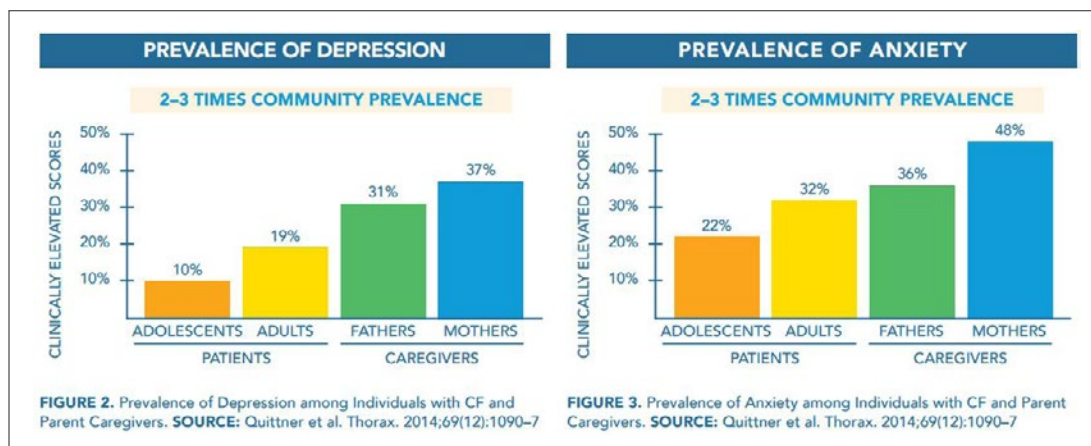
With CF, you have a lot of things to handle — from treatments and clinic appointments to eating the right foods and getting exercise. On top of these you might have concerns about your future. So it's not surprising that research has shown that having any chronic illness puts people at higher risk for anxiety and depression.

## TEENAGERS AND YOUNG ADULTS ARE MORE AT RISK

A study in nine countries (the TIDES study) showed that depression and especially anxiety were elevated in people with CF and in parents of children with CF (Figures 2 and 3 in graphic below).

Certain people are at a higher risk for anxiety and depression. Teenagers and young adults are most at risk compared with younger children (under age 12) or adults over age 30. And women are more likely to feel depressed or anxious than men.

**The International Depression/Anxiety Epidemiological Study** (TIDES) is the first epidemiological study on the rates of depression and anxiety in people with CF. The study was conducted in nine countries, including 45 CF care centers throughout the United States.



## Anxiety and Depression

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### DEPRESSION AND ANXIETY CAN AFFECT YOUR HEALTH — AND LOWER YOUR QUALITY OF LIFE

Your health can be affected if you are depressed or anxious. Depression can make it harder to do CF treatments. People who are depressed sometimes do not have the energy or motivation to take care of themselves by following a healthy diet, exercising, and managing their disease.

In addition, people who are depressed or anxious may have trouble focusing and remembering to do their treatments. They may feel hopeless and think that doing their treatments will not help, and they may stop doing them.

Symptoms of depression and anxiety can also lower your quality of life. A study showed that adults with CF who had moderate or severe lung disease and more depressive symptoms reported worse quality of life than those with moderate or severe lung disease and no depressive symptoms. In other words, *depression can make living with CF a lot harder to handle.*

### AVOID SUBSTANCE ABUSE

Substance abuse is linked to anxiety and depression. When you are feeling anxious or depressed, you are more likely to misuse substances such as alcohol, tobacco, illegal drugs, or even prescription medications.

While drinking more or getting high — or taking more medication than is prescribed — may make you feel better in the short term, over time this kind of abuse can actually make depression or anxiety worse. And it often leads to more substance abuse — even addiction — resulting in a downward spiral that leaves you feeling far worse than you felt in the beginning.

There has not been much research on the impact of substance use on people with CF. We do know that tobacco use and smoking are particularly harmful to people with CF and should be avoided to prevent damage to the lungs. You should also avoid exposure to secondhand smoke.

Too much alcohol use can damage your immune system and may make you more likely to get infections. Use of alcohol and other substances to the point of intoxication may leave you with a hangover. When people are hung over, they are less likely to do the things they need to do to take care of themselves, such as CF treatments.

## Anxiety and Depression

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People with CF often experience lots of pain. Physicians may prescribe pain medication to help manage the pain and improve quality of life. Most people who take prescription medications use them responsibly. But when abused — that is, when a person takes medicine more often or in higher doses than prescribed or uses someone else’s medication — prescription medications can produce serious negative health effects, including dependence or addiction.

### COMMON SIGNS AND SYMPTOMS OF SUBSTANCE ABUSE

The following common signs and symptoms may mean your substance abuse has gotten out of hand and you need help. Call your CF care team if:

- › You have built up a tolerance and need to use more substances to get the same feeling.
- › You often use more than you planned, even though you told yourself you would not.
- › You take substances to avoid or relieve withdrawal symptoms. If you go too long without using them, you experience withdrawal symptoms such as nausea, restlessness, insomnia, depression, sweating, shaking, and anxiety.
- › You are using substances under dangerous conditions, such as driving drunk, or engaging in risky behaviors when high.
- › You continue to use substances, despite knowing the abuse is causing major problems in your life — blackouts, infections, mood swings, depression, paranoia.
- › You ignore responsibilities at home, work, or school.
- › You are having problems with your relationships, such as fights with your partner or family members, your boss not happy with your work or the loss of friendships.
- › You drop activities you used to enjoy, such as hobbies, sports, and socializing.
- › You’re getting into legal trouble, such as arrests for disorderly conduct, driving under the influence, or stealing to support a drug habit.

### WORK WITH YOUR CF TEAM

If you are feeling sad or overwhelmed, it is important to talk with your CF care team. Your **care team may offer an annual screening** for anxiety and depression. There are many effective treatments for depression and anxiety.

Anti-depressant and anti-anxiety medications can help people with chronic illnesses

## Anxiety and Depression

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and depression or anxiety. Talk therapy, such as cognitive behavioral therapy (CBT), with a trained therapist also helps. For most people, **combining** medication and talk therapy works the best.

### MAKE A PLAN WITH YOUR TEAM

Your CF care team or primary care doctor can help you come up with a plan. They can also prescribe medications, provide referrals to counselors, or just listen to understand what you are going through.

At your CF center, the social worker or psychologist may be able to provide you therapy or offer referrals to specialists in the community who have experience treating anxiety, depression, or substance abuse in adults with a chronic illness such as CF. At many CF centers, particularly smaller ones, psychologists are available by referral and the social worker is the team member most likely to be your “first stop” for help.

If you decide to see a mental health specialist, it is important to talk with your health insurance plan. Mental health treatment such as therapy or seeing a psychiatrist is often covered under a special mental health “carve-out.” This means that your claims for mental health services are processed and paid by a different company than claims for your medical services.

We suggest that you talk with your health insurance plan before starting services to find out which providers are covered and what expenses you will be expected to pay.

If you do not have insurance or cannot afford treatment, you may be eligible for treatment at publicly funded mental health centers and through other mental health programs. These programs often work on a sliding scale based on your ability to pay. Use the resources guide at the end of this section for help in finding treatment.

### MANAGE YOUR TIME

#### TIPS FOR STAYING IN CONTROL OF YOUR LIFE (PREVENTION)

It is true that some forms of depression may not be preventable. That's because depression may be triggered by a chemical malfunctioning in the brain.

But the latest medical studies confirm that depression may sometimes be prevented with good health habits. Taking care of yourself by following these good health habits may help you both physically and emotionally:

- › Eating well
- › Exercising regularly
- › Taking time out for fun and relaxation
- › Thinking positively, replacing negative thoughts with more positive ideas

# Anxiety and Depression

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## WHAT ELSE TO DO WHEN FEELING SAD OR WORRIED

- › Talk with someone every day, preferably face to face. Though you feel like retreating and being alone, ask trusted friends and acquaintances to spend time with you.
- › Spend time with people who are not depressed or substance users. This can make you feel better.
- › Avoid alcohol and other drugs. They will only make you feel worse in the long run.
- › Make a schedule for yourself every day and stick to it, even if you don't think you have the energy or aren't in the mood.
- › Do not skip meals or any of your CF therapies, and get at least 8 hours of sleep each night.
- › Get outside or into nature for at least 30 minutes a day.
- › Make time for things that bring you joy. Spend more time doing things that you enjoy.
- › Include at least 30 minutes of exercise in every day.

## MYTH BUSTERS

### **Myth #1: There are no cures for depression, anxiety, and substance abuse.**

**Fact:** Treatment can help. Studies show that there are many highly effective treatments. But people respond differently to different treatments. What might work for some may not work for others. This is why it's important to seek professional help to determine the right treatment program for you.

### **Myth #2: Depression, anxiety, and substance abuse are not real medical problems.**

**Fact:** These are all real and serious conditions. The medical community has recognized the seriousness of these disorders. So it is important to let your medical provider know if you are having trouble with any of these issues.

### **Myth #3: Depression and anxiety are something that strong people can "snap out of" by being positive.**

**Fact:** No one chooses to be depressed or anxious — just like no one chooses to have CF. People with depression or anxiety cannot just "snap out of" their problems.

## Anxiety and Depression

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### **Myth #4: Depression and anxiety will just go away on their own.**

**Fact:** While for some people depression or anxiety may go away without treatment, this is not usually the case. Without treatment, symptoms of depression can continue for weeks, months, or even years. Depression can lead to suicide, the third-leading cause of death for 18- to 24-year-olds, highlighting the importance of seeking treatment. The good news is that most people do get better with treatment.

### **Myth #5: Medications will change your personality.**

**Fact:** Taking medicine that changes your brain chemistry can be scary. However, anti-depressant and anti-anxiety medications are designed to change only certain chemicals that cause the symptoms of depression or anxiety. They do not change your personality. Most people who take anti-depressants are actually happy to feel like themselves again, instead of feeling like a different person when depressed. It is best to speak with your doctor about the side effects of any medication.

### **Myth #6: Talking about depression or anxiety only makes it worse.**

**Fact:** It is easy to understand why someone might be worried about discussing depression. But being alone with your thoughts is even more harmful when facing these disorders. If you are hesitant to discuss difficulties you might be facing with a close family member or friend, think about other people in your life. Spiritual leaders or school or employee health counselors will be willing to discuss your struggles with you. If, at any point, you feel so overwhelmed by feelings of sadness and hopelessness that you are considering hurting yourself, call the [National Suicide Prevention Lifeline](#) at **800-273-8255** for help.

### **Myth #7: Treatment for depression, anxiety, or substance abuse should be a one-shot deal.**

**Fact:** Like many other illnesses, these disorders are usually long term. Many people need more than one session of therapy to treat these conditions. You may need just a few sessions to feel better and a few months later need more counseling to get through another rough patch.

### **Myth #8: There should be a standard treatment program for everyone.**

**Fact:** One treatment method is not always right for everyone. The best programs develop an individual treatment plan based on a full understanding of your current problems and history. The plan may combine a variety of methods adapted to address your specific needs. A plan may include behavioral therapy (such as counseling, cognitive therapy, or psychotherapy), medications, or a combination of any of these. Referrals to other medical, psychological, and social services may also be important parts of treatment.



# Anxiety and Depression

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## GET HELP WHEN YOU NEED IT

Remember, you can call your CF care team if you think your anxiety, depression, or substance abuse has gotten out of hand and you need help. Here is a list of resources to help you:

- › American Counseling Association — [www.counseling.org](http://www.counseling.org)
- › Anxiety and Depression Association of America — [www.adaa.org](http://www.adaa.org)
- › American Psychological Association — [www.apa.org](http://www.apa.org) or **800-374-2721**
- › CF Foundation’s Screening & Treating Depression & Anxiety Guidelines — [CFF.org/For-Caregivers/Clinical-Care-Guidelines/Screening-Treating-Depression-and-Anxiety-Clinical-Care-Guidelines/](http://CFF.org/For-Caregivers/Clinical-Care-Guidelines/Screening-Treating-Depression-and-Anxiety-Clinical-Care-Guidelines/)
- › Depression and Bipolar Support Alliance — [www.dbsalliance.org](http://www.dbsalliance.org) or **800-826-3632**
- › Freedom From Fear (for anxiety and depression) — [www.freedomfromfear.org](http://www.freedomfromfear.org) or **718-351-1717**
- › National Association of Social Workers — [www.socialworkers.org](http://www.socialworkers.org)
- › Mental Health America — [www.nmha.org](http://www.nmha.org)
- › Partnership for Drug-Free Kids — [www.drugfree.org](http://www.drugfree.org) or **855-DRUGFREE**
- › Substance Abuse and Mental Health Services Administration — [www.samhsa.gov](http://www.samhsa.gov)

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# Basics of CF and Male Infertility

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## IN THIS SECTION:

- ▶ **There are two basic facts about CF and male infertility.**
- ▶ **Beyond basics.**
- ▶ **Starting a family.**

## Basics of CF and Male Infertility

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### THERE ARE TWO BASIC FACTS ABOUT CF AND MALE FERTILITY:

**Fact #1:** Yes, CF affects a man's ability to have children.

**Fact #2:** No, CF does not affect his ability to have sex or a normal orgasm.

### HERE'S A QUICK PUBLIC SERVICE ANNOUNCEMENT AND ANOTHER IMPORTANT FACT:

**Fact #3:** Men with CF are just as much at risk as everyone else for getting or giving a sexually transmitted disease, so use a condom! If you are unclear about proper use of a condom, ask your physician.

- › Most men with CF (over 95 percent) don't have any sperm in their ejaculate. This is a condition physicians call "azoospermia." That's because they are missing the tubes (vas deferens) that normally carry sperm from the testicles into the ejaculate. Thus, they cannot get a woman pregnant through normal intercourse. In case you want to impress your friends, the technical term for the missing tubes is congenital bilateral absence of the vas deferens, or CBAVD. (See Figure 1)

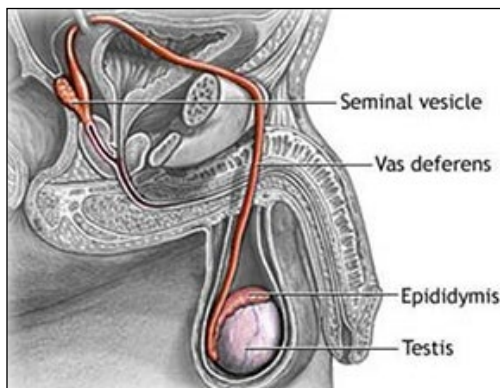


Figure 1  
Normal Male Genitourinary Anatomy

The male reproductive system has three primary parts: the testicles, the epididymis and the vas deferens."

- › If you are interested in starting a family, here's some more good news: most men with CF make normal sperm in their testicles. It just doesn't have a way to reach its destination. By using assisted reproductive technology (ART), a urologist can collect sperm directly from the testicle via testicular sperm aspiration (TESA) or microsurgical epididymal sperm aspiration (MESA). From there, an infertility specialist can use the sperm to fertilize the woman's egg (a procedure called intracytoplasmic sperm injection [ICSI]; see Figure 2) and place it in her uterus.

## Basics of CF and Male Infertility

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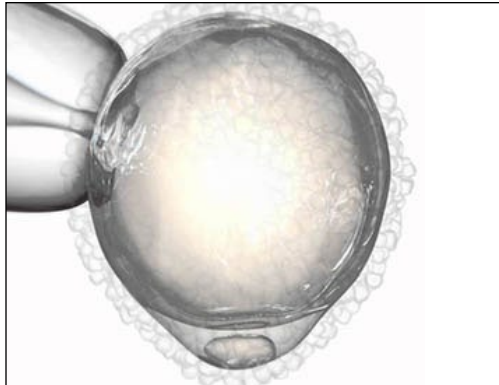


Figure 2  
Intracytoplasmic Sperm Injection (ICSI)

- › When thinking about starting a family using this technique, it is important to find out if the woman is a CF carrier. If she is, there's a 50/50 chance a child conceived like this will have CF.
- › Assisted reproductive technology (ART) is not the only option for men with CF who want to start a family. Some couples opt to use donated sperm or they decide to adopt a child.

### AND

- › Since a small number of men with CF are fertile (that is, there are sperm in their ejaculate), the smart move is not to make any assumptions! Talk to your CF team about checking whether you are fertile.

**Remember Fact #3:** Men with CF are just as much at risk as everyone else for getting or giving a sexually transmitted disease, so use a condom!

In your late teens and early adulthood, you may sometimes have confusing emotional and personal concerns about sex and relationships that have nothing to do with reproduction. Starting serious personal relationships with someone can be challenging with or without having CF to worry about. But for many people, the CF (a lot of coughing, a lot of pills, daily treatments, etc.) makes it more challenging. Please ask your CF care team questions about sex, emotional commitment, LGBTQ issues, and long-term relationships. They might not have specific answers but they should offer a safe place to talk about these issues.

## Basics of CF and Male Infertility

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### BEYOND THE BASICS

If you were diagnosed with CF as a child, you may have been told about CF-related reproductive issues. If you were diagnosed as an adult, some of this may be new to you. Your CF team spends a lot of time focusing on your lung and digestive health, so sometime issues related to sex and reproduction get lost in the shuffle.<sup>1</sup> As with all aspects of CF, it's important to be aware of how CF affects your body, and in this case, how it affects your partner and your intimate life as well.

Let's get into some biology, shall we?

Male reproduction needs three main "parts": the testicles, the epididymis, and the vas deferens. Sperm develop in the testicles. The epididymis is a series of tubes that sit behind the testicles and serve as the place where sperm are stored until they are made available at ejaculation. The vas deferens is a long tube that connects the epididymis to the ejaculatory ducts, so mature sperm can come through the penis during ejaculation.

Nearly all men with CF are missing the part of the male reproductive tract that carries the sperm into the ejaculate, which brings us back to CBAVD.<sup>2</sup> Again, the tubes are missing, but the sperm are not. Sperm production in the testicles is normal in 90 percent of men with CF and CBAVD.<sup>3</sup>

Nobody knows for sure what causes CBAVD. But some think the same gene mutations that cause the lung and pancreas problems with CF likely cause these problems.

Interestingly, 1 to 2 percent of infertile men who do not have signs of the lung disease or digestive problems so common in CF have CBAVD.<sup>4</sup> It's thought that CBAVD is based on similar genetic patterns found in typical CF but that it involves milder mutations that cause only the CBAVD and not the typical, full-blown CF.

That's why it's important for any man with CBAVD to be tested for CF.<sup>5</sup> CBAVD does not affect sexual performance or the ability to have intercourse. In fact, most people don't know they have CBAVD until they can't conceive a child and seek care from a urologist.

### STARTING A FAMILY

The decision to start a family is a big one, particularly when a couple is working hard to make peace with a chronic condition like CF. If you are eager to start a family, first talk to your CF team about it. (Well, maybe your partner first, then the experts.)

Some couples explore the adoption route. Others opt for sperm donation. There are a lot of technical and financial issues involved, and a wife or partner should get genetic counseling and testing. But a fair number of men with CF have created families by using their own sperm. If you are interested in this, consult a male reproductive expert first.

# Basics of CF and Male Infertility

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## Learn More

### REFERENCES

1. Sawyer SM, et al. Sexual and reproductive health in men with cystic fibrosis: consistent preferences, inconsistent practices. *J Cyst Fibros*. 2009;8:264-269.
2. Kupker, W, et al. Genetics of male subfertility: consequences for the clinical work-up. *Hum Reprod*. 1999;14 (Suppl 1):24-37.
3. Meng MV, et al. Impaired spermatogenesis in men with congenital absence of the vas deferens. *Hum Reprod*. 2001;16:529-533.
4. Stuhmann M, Dork T. CFTR gene mutations and male infertility. *Andrologia*. 2000; 32:71-83.
5. Phillipson GT, Petrucco OM, Matthews CD Congenital bilateral absence of the vas deferens, cystic fibrosis mutation analysis and intracytoplasmic sperm injection. *Hum Reprod*. 2000;15:431-435.

### ADDITIONAL RESOURCES

- › How Does CF Affect the Male Reproductive System?  
[CFF.org/Living-with-CF/Reproductive-Health-and-Fertility/How-Does-CF-Affect-the-Male-Reproductive-System/](https://www.cff.org/Living-with-CF/Reproductive-Health-and-Fertility/How-Does-CF-Affect-the-Male-Reproductive-System/)
- › Living a Healthy Adult Life With CF: Fertility & Family: Male Fertility  
<https://www.youtube.com/watch?v=S-Rzn1KGQHg&index=16&list=PLhoQ6vyZhgqoJAJpmmZeTvm9KDPH0SuvM>

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# Birth Control for Women With CF

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You may have heard that it might be hard for you to get pregnant because you have cystic fibrosis. We are here to tell you that you can get pregnant. Thick mucus may make it harder for sperm to travel through the cervix, but pregnancy is possible. Many women with CF have become pregnant — you could, too!

Planning pregnancy for a time when you are healthy is important. If you do not want to be pregnant now, it is important to use birth control.

Is this a time when having a baby would be good for you?  
If not, read on!!

## IN THIS SECTION:

- ▶ **Learn about birth control methods.**
- ▶ **Work with your CF team** to help you choose the right method of birth control that works best for you.

# Birth Control for Women With CF

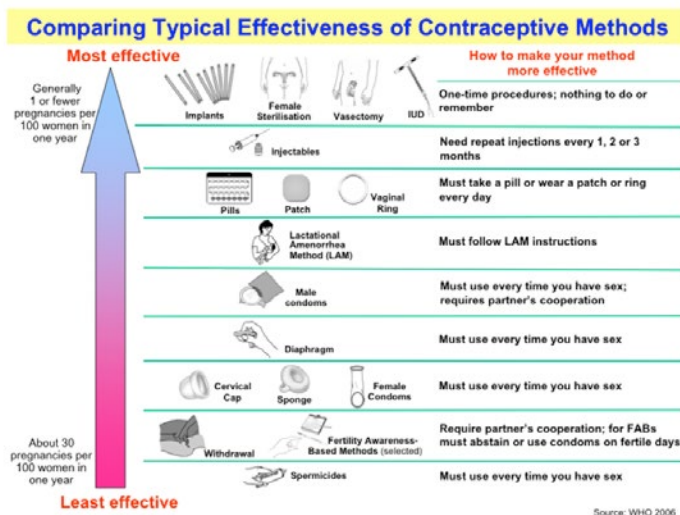
## LEARN ABOUT BIRTH CONTROL METHODS

There are a lot of good methods of birth control for women with CF. Use this information and talk with your partner and CF care team to choose a method that you are happy with and that which supports your health. How well the method works is also important.

The table below shows the effectiveness of birth control methods. For the most protection from pregnancy, use a method correctly all of the time. If you have a new partner or want to protect yourself from sexually transmitted diseases (STDs) — yes, they are out there! — use a condom.

This discussion groups birth control methods according to the way that they work, beginning with the most effective ones:

- › **Sterilization:** vasectomy, tubal ligation, or contraceptive tubal occlusion device and delivery system (Essure®)
- › **Intrauterine contraception:** copper intrauterine device (Paragard®) or levonorgestrel intrauterine device (Mirena®)
- › **Hormonal methods:** birth control pills, norelgestromin and ethinyl estradiol patch (Ortho Evra®), etonogestrel/ethinyl estradiol vaginal ring (NuvaRing®), medroxyprogesterone acetate (Depo Provera®), etonogestrel implant (Implanon®), Nexplanon®, and levonorgestrel tablet (Plan B®)
- › **Barrier methods:** male condom, female condom, nonoxynol-9 sponge (Today Sponge®), diaphragm, cervical cap, and vaginal barrier contraceptive devices (FemCap®)
- › **Behavioral methods:** abstinence, natural family planning, and calendar-based methods (CycleBeads®)





## Birth Control for Women With CF

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### STERILIZATION

*I've decided that I don't want to be pregnant in the future.*

Many women with CF choose sterilization for themselves, or their partners choose vasectomy. Sterilization is permanent birth control.

**Vasectomy** is for men and **tubal ligation** is for women. These methods prevent pregnancy by cutting or tying the vas deferens (in men) or the fallopian tubes (in women). This keeps sperm from fertilizing an egg. Vasectomy is an easier and less expensive procedure than tubal ligation.

Vasectomy is usually performed in a medical office. After a vasectomy, no sperm will be in the man's semen (ejaculate). It is important to have a follow-up check to be sure that the operation was successful.

Tubal ligation is usually done in a hospital or surgery center. The fallopian tubes are tied or cut, blocking the pathway for sperm to reach an egg.

**Contraceptive Tubal Ligation Device and Delivery System (Essure®)** for women is a non-surgical sterilization procedure. Small coils are placed in the fallopian tubes during an office visit. Within three months, the fallopian tubes become blocked. Systemic steroids such as prednisone may keep this birth control method from working to block the tubes.<sup>1</sup> If you want to use this method, choose a time when you are not taking these medications to start.

### INTRAUTERINE CONTRACEPTION

The IUD is a T-shaped plastic device placed in the uterus by a health care provider. It keeps sperm from traveling through the uterus to fertilize an egg.

*I've heard that women cannot use IUDs if they haven't had a child.*

IUDs are safe and effective for women who have not had children. Using an IUD will not keep you from getting pregnant after it is taken out.<sup>2</sup>

The intrauterine copper contraceptive prevents pregnancy for 10 years. There is a thin copper wire on it. Except for sterilization, this is the most effective method that does not use hormones. Some women experience more bleeding and cramping with their periods when using the intrauterine copper contraceptive.

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<sup>1</sup> Retrieved from <http://www.sfwomenshealthcare.com/services/essurer> on September 6, 2009.

<sup>2</sup> Contraceptive Technology. Ardent Media. 2007.117.

## Birth Control for Women With CF

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The levonorgestrel-releasing intrauterine system (Mirena®) prevents pregnancy for 5 years. It is also a hormonal method and the progestin in it thickens the cervical mucus and blocks the cervix. Also, like the intrauterine copper contraceptive, the levonorgestrel-releasing intrauterine system keeps sperm from moving through the uterus to the fallopian tubes. Women using this method of birth control often have little or no menstrual bleeding and less cramping.

### HORMONAL METHODS

These methods use hormones like those in a woman's body, estrogen and progestin (synthetic progesterone). The hormones thicken cervical mucus and prevent ovulation. Menstrual periods can be shorter and lighter. Some women may have little bleeding or no period at all.

Methods that combine estrogen and progestin follow this pattern: hormones for 21 or more days followed by a few to 7 days of no hormones. During the hormone-free days withdrawal bleeding, or a period, occurs.

- › **Oral Contraceptives** ("the pill"): Women take one pill each day.
- › **The Patch** is placed on the skin, like a band-aid — one patch each week for three weeks. During the fourth week no patch is used. The hormones are in the glue. Estrogen levels with the patch are higher than with birth control pills.
- › The vaginal ring (**NuvaRing**) is made of silicone that contains the hormones estrogen and progestin. The ring is placed in the vagina for three to four weeks, then removed for up to 7 days. The vaginal ring uses fewer hormones than oral contraceptives and passes through the liver only once. Some women may have more vaginal discharge when using the vaginal ring.

## Birth Control for Women With CF

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**Progestin-only methods** do not use estrogen. If you can't use estrogen, these methods might be right for you. Because there is no hormone-free time, women may have irregular periods, spotting, or no bleeding at all.

- › **The progestin-only pill** is taken every day. It is very important to take these pills at the same time every day.
- › Medroxyprogesterone acetate (**Depo Provera®**) is a shot of progestin given every three months. Medroxyprogesterone acetate may cause a loss of bone density but bone density levels return after stopping. Because women with CF have a higher risk of osteoporosis, this method may raise concerns. To support bone health, health care providers can prescribe estrogen for women using medroxyprogesterone acetate.
- › The implant (**Implanon®**) is a silicone rod containing progestin that is placed beneath the skin. It lasts for 3 years.

### EMERGENCY CONTRACEPTION

*My partner and I use condoms. Sometimes a condom will break or slip off.*

- › Emergency contraception, levonorgestrel tablet (**Plan B One Step®**), is one progestin pill. It lowers the chance of pregnancy if taken within 5 days after unprotected sex. The tablet prevents or delays ovulation and is 85 percent effective in preventing pregnancy. Note that this is considered emergency contraception and should not be your only method of birth control!

### THINGS TO WATCH OUT FOR — BIRTH CONTROL ISN'T PERFECT

Estrogen and progestin are drugs and can affect other medications and health issues. For women with CF who use hormonal birth control, there are some concerns:

- › There may be increased blood sugar with estrogen.
- › Women with active gallbladder disease should not take estrogen.
- › There is a possibility of bone density loss with use of medroxyprogesterone acetate (Depo Provera®).
- › Progestin in drospirenone and ethinyl estradiol (YAZ® and Yasmin®) increases salt and fluid retention, so women who have kidney, liver, or adrenal disease should not use these pills.<sup>3</sup>

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<sup>3</sup> Retrieved from [www.yaz-us.com](http://www.yaz-us.com) and <http://yasmin-us.com/index.html> on May 10, 2009.

## Birth Control for Women With CF

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- › There is more risk for vaginal yeast infection.
- › There is increased risk of developing gallstones.
- › Less hormone may be absorbed in the intestines.
- › There is a higher risk of blood clots with estrogen-containing products.

### WILL ANTIBIOTICS MAKE MY BIRTH CONTROL METHOD LESS EFFECTIVE?

When taking antibiotics and some antifungals, hormonal birth control can be less effective. Use another method such as a condom when taking antibiotics and for 7 days after you finish your medication. This is very important for women who are taking rifampin or griseofulvin.<sup>4</sup> If you are taking antibiotics all of the time, such as azithromycin, you need a second method during the first two weeks after changing antibiotics.<sup>5</sup>

### BARRIER METHODS

*I want a method without hormones.*

*We don't have sex very often. I want a method that I use only when I need it.*

Barrier methods prevent pregnancy by keeping sperm from traveling to the fallopian tubes, where eggs are fertilized. You can get many of these methods without a prescription.

**Condoms** are important for preventing sexually transmitted infection and HIV. They are also a good choice for birth control. There are many sizes and shapes of condoms, and most are made of latex. Some condoms are made from soft plastic or sheep intestines. To keep from getting a sexually transmitted infection, use condoms made of latex or plastic. Lambskin condoms prevent only pregnancy — not infection. And condoms lubricated with spermicide are not more effective.

*I'm allergic to latex and can't use condoms.*

If you or your partner is sensitive to latex you can use a non-latex condom. Some condoms are made from soft plastic, and prevent pregnancy and sexually transmitted infection. Lambskin condoms prevent pregnancy but not infection.

Some women or their partners react to spermicides. They can be in the condom lubricant or the contraceptive cream or jelly used with the diaphragm and cervical cap. If you or your partner has a reaction, try another brand or use another method.

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<sup>4</sup> *Contraceptive Technology*. Ardent Media. 2007:237.

<sup>5</sup> The sexual health of adolescents with cystic fibrosis, *J Roy Soc Med*. 2005;98(446):10.

## Birth Control for Women With CF

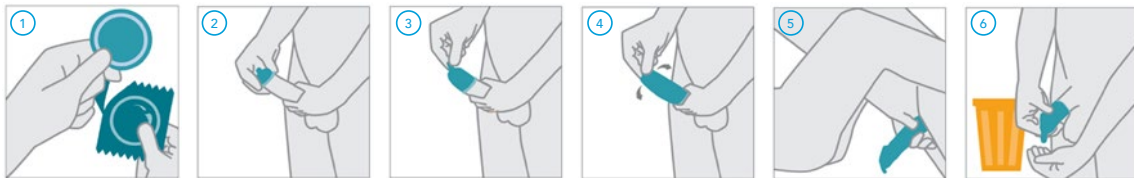
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### HOW TO USE A CONDOM

- › Talk with your partner about using condoms to prevent pregnancy or infection.
- › Be sure the package is not broken.
- › Apply before genital contact.
- › Use water-based lubricant — a drop in the inside of the condom — to enhance sensation for the man wearing the condom.
- › Pinch the tip.
- › Unroll onto erect penis.
- › Use water-based lubricant on the outside of the condom, if needed.
- › Enjoy intercourse.
- › Hold onto base of condom (either partner).
- › Withdraw while penis is still erect.
- › Discard in trash — not the toilet!

### HOW TO PUT ON AND TAKE OFF A MALE CONDOM

1. Carefully open and remove condom from wrapper.
2. Place condom on the head of the erect, hard penis. If uncircumcised, pull back the foreskin first.
3. Pinch air out of the tip of the condom.
4. Unroll condom all the way down the penis.
5. After sex but before pulling out, hold the condom at the base and withdraw the penis.
6. Carefully remove the condom and throw it in the trash.



## Birth Control for Women With CF

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Content source:

[National Center for HIV/AIDS, Viral Hepatitis, STD, and TB Prevention, Centers for Disease Control and Prevention](#)

**The female condom** is a plastic pouch. It is placed in the vagina before sex. It is less effective than a condom worn by a man. Female condoms should NOT be used with a male condom.

**Vaginal spermicides (foam, cream, gel, film, or suppository)** are placed in the vagina before sex. They destroy sperm. Vaginal spermicides and male condoms used together are very effective. You can buy vaginal spermicides at a family planning clinic, pharmacy, or grocery store.

**The contraceptive sponge**, is a small, soft, round plastic device with spermicide. It is placed in the vagina, covering the cervix, before sex. It can stay in the vagina for 24 hours. Wait for 6 hours after sex to remove the sponge. Women may have more vaginal discharge when using the sponge. If you think you might have a vaginal infection after using the sponge, it is a good idea to visit a health care provider.

**The diaphragm and cervical cap** are made of latex or silicone. These devices are placed in the vagina, covering the cervix — the opening to the uterus — before sex. They work with spermicide to keep sperm from getting through the cervix. Wait for 6 hours after sex to remove the diaphragm or cervical cap .

Might the thicker cervical mucus of a woman with CF create a problem for keeping these devices in place? Use the diaphragm, sponge, or cervical cap with another method (e.g., pill, vaginal ring, or condom) in the first month of use to see if the device stays in place over the cervix during sex.

The **diaphragm** is a latex, dome-shaped device that holds spermicide over the cervix. It must be used with spermicide. The diaphragm comes in different styles and sizes, and is fitted by a health care provider. Have your diaphragm checked if you gain or lose weight or become pregnant.

A cervical cap (**FemCap**) is made of silicone and comes in three sizes. It is a good choice for women who are allergic to latex. Check with your doctor or nurse practitioner about using this method. You need a prescription to get a cervical cap.

# Birth Control for Women With CF

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## Fertility Awareness-Based Methods

### BEHAVIORAL METHODS

**Abstinence** means having no sexual activity that can cause pregnancy or infection. This is the surest way to keep from getting pregnant. Many couples have times when they don't have sex.

*My partner and I are using the "pull-out" method.*

**Withdrawal** is when the man pulls his penis out of his partner's vagina before he ejaculates. Couples use withdrawal by itself or with other methods. Withdrawal is less effective than the male condom in preventing pregnancy. It does not protect you from a sexually transmitted infection.

**Fertility awareness** or **natural family planning** helps women and their partners know when a woman is fertile. If a woman has regular cycles (from the beginning of one period to the day before the next period begins), she and her partner can use this method well with careful use and support from an experienced teacher. This approach can also help women who wish to become pregnant.

One of the ways to learn about fertility during a woman's cycle is to take her temperature each morning with a **basal body thermometer**.

**Cervical mucus** changes during the cycle. A woman can learn about and chart these changes to learn when she is fertile and when she is not.

**Calendar-based** contraception, or using **CycleBeads**<sup>®</sup> also helps women and couples understand fertility. The color-coded string of beads shows when a woman could get pregnant. Women whose cycles are between 26 and 32 days can use this type of method. This is a great way to learn about changes that happen during the menstrual cycle.

### WORK WITH YOUR CF TEAM



The best method is one that works well that you will use carefully. Talk about birth control choices with your partner. Your health care team can help you find a method that supports your health. You will be able to choose a method that is right for you.

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# Pregnancy in CF

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**Good news! As people with cystic fibrosis are living longer and healthier lives, pregnancy among women with CF is becoming more common.**

**Before you make the decision to have a baby, you need to know the pros and cons. Others will need to be involved to help you make the right decision — your spouse or significant other, your family, and, most important, your CF care team.**

## **IN THIS SECTION:**

- ▶ **Learn how CF can affect getting pregnant.**
- ▶ **Work with your CF team** to plan for a safe pregnancy.
- ▶ **Understand the potential complications** that could occur.
- ▶ **Understand the importance of managing your care** to include possible additional therapies needed for a safe pregnancy.
- ▶ **Get help** to maintain your health during your pregnancy.



## Pregnancy in CF

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### LEARN HOW CF CAN AFFECT GETTING PREGNANT

Good outcomes with pregnancy are associated with optimal nutrition, adequate glucose control, and appropriate lung function before pregnancy. With proper management, women with CF can experience uncomplicated pregnancies that result in healthy babies.

### PLAN YOUR PREGNANCY

Women who plan their pregnancies tend to have better outcomes. Making sure your overall health is in the best shape possible before pregnancy is the most important thing you can do for yourself and your baby. Discuss with your CF care team if any of your meds or therapies are harmful to you or your baby during pregnancy.

### SHOULD I HAVE MY PARTNER TESTED FOR CF PARTNER?

If your partner is a carrier, this may have an effect on your decision to have a child. Deciding whether to have your partner tested for CF gene mutations is your own personal choice. Talk it over with your partner and with your health care provider. Get as much information as you need to decide what's right for you and your family.

The only way to get CF is to have two genes that cause CF — one from the mother and one from the father. A CF carrier has only one CF gene and has no CF symptoms. Approximately 1 in 30 Americans are symptomless carriers of the CF gene. CF carrier testing is done on blood or saliva. It checks to see if parents-to-be have the abnormal gene that causes CF. The test can help determine if you're at increased risk for having a child with CF.

If you have CF and your partner is found to have no abnormal CF genes, your baby will likely only be a carrier. If your partner is found to be a carrier, there is a 50-50 chance that your child will have CF. It is important to understand that a negative test does not guarantee that your child will not have CF. The most common types of CF gene mutations are tested for, but your partner could still carry an uncommon one that was not found on testing. Approximately one in eight carriers have a mutation that is not in the standard CFTR mutation panel.<sup>1</sup>

If test results show that you are at risk for having a baby with CF, your health care provider can test the baby in utero, though this can be costly and imperfect.<sup>2</sup> If the baby tests positive for CF, you and your partner can prepare in advance and make contact with a pediatric CF care center.

The American Congress of Obstetricians and Gynecologists (ACOG) recommends that health care providers make the CF carrier screening test available to all couples.

## Pregnancy in CF

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### IS IT HARD TO GET PREGNANT?

Women with CF may have thicker cervical mucus, making it harder for sperm to move through the female reproductive tract.<sup>3</sup> Other reasons for trouble conceiving can be poor nutrition, leading to disruptions in ovulation. However, the majority of women with CF become pregnant without difficulty. Up to 70 percent of women with CF are able to conceive a child.<sup>4</sup>

If you do have difficulty, assisted reproductive techniques such as intrauterine insemination (IUI), in-vitro fertilization (IVF), and intracytoplasmic sperm injection (ICSI) are viable options, as is adoption and surrogacy. Your obstetrician can provide you with information to help with your decision.

### WORK WITH YOUR CF TEAM

Working with your CF team is imperative to ensure you have a safe pregnancy. It is highly recommended that you have an obstetrician who specializes in high-risk pregnancies and whose practice is at the same facility as your CF care center. Your care team and your obstetrician will need to work together to ensure you and your baby remain healthy during your pregnancy.

### AM I HEALTHY ENOUGH TO HAVE A BABY?

**Nutrition:** In women with CF, a BMI (body mass index) of  $\geq 22$  is considered adequate. If your BMI is below 18, improving your nutritional status before you get pregnant is the best thing you can do for yourself and your baby. Your CF care team can assist you. The increased nutritional demands of pregnancy place a woman with CF at risk for inadequate weight gain or weight loss.<sup>5</sup> Historically, pregnancy in women with CF occurred in those who were pancreatic sufficient and nutritionally stable. More recently, women who are pancreatic insufficient have become pregnant because weight is more important than pancreatic status in achieving the best outcomes for you and your baby. Therefore, if you maintain an adequate weight, you can have a successful pregnancy.

**Lung function:** Several studies have suggested if your FEV<sub>1</sub> is above 50 percent, your chances of having good outcomes are much better. One study found that women who were pancreatic sufficient, did not have *Burkholderia cepacia* colonization, and had an FEV<sub>1</sub> above 50 percent did better than others.<sup>6</sup> Other studies have reported that having diabetes and a pre-pregnancy FEV<sub>1</sub> below 50 percent were associated with worse outcomes in both mother and infant.<sup>7</sup> Some studies have shown that children of women with CF have an increased chance of being born prematurely or have a lower birth weight when the mother has an FEV<sub>1</sub> below 50 percent.<sup>8,9</sup> Overall, outcomes for you and your baby are likely better when your FEV<sub>1</sub> is greater than 50 percent before you get pregnant.

## Pregnancy in CF

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**Cirrhosis:** CF can be associated with severe liver disease such as cirrhosis. If you have liver disease, you need to discuss the implications of pregnancy and your liver disease with your care team.

### UNDERSTAND THE POTENTIAL COMPLICATIONS

**Nutritional deficiency:** It is important to make sure your nutritional health is in good shape before, during, and after pregnancy. Optimally, your BMI should be 22 or higher. Your nutritional demands will increase while pregnant and put you at risk for inadequate weight gain or weight loss.<sup>5</sup> Many people are advised to drink supplements. Aim for higher-calorie diets, but focus on maintaining and gaining weight while pregnant.

**Constipation:** During pregnancy, all women are at risk for constipation but CF puts you at higher risk. If you notice a decrease in the number or amount of your normal stools, call your physician. To prevent this, you can try to drink more water, eat more vegetables and fruits, and add fiber to your diet. Your CF care team may recommend the use of stool softeners or laxatives while pregnant.

**Diabetes:** Diabetes and glucose intolerance occur often in people with CF. Becoming diabetic during pregnancy (called gestational diabetes) can occur in women who have CF and those who don't. You will need to be monitored closely for diabetes. During pregnancy, diabetes can raise your risk of a worse outcome. If you already have CF-related diabetes (CFRD), monitoring and controlling your blood sugars is imperative. It is ideal to have your sugars optimally controlled before getting pregnant. If you are taking insulin to control your diabetes, your insulin requirements may change when you are pregnant.

**Exacerbations:** You may experience more exacerbations when you are pregnant. To stay as healthy as possible, you should maintain your respiratory therapies throughout pregnancy. Your care team will discuss with you if any of your therapies should be stopped or changed. Chest physiotherapy (CPT) is an important part of your therapies. Many people are concerned that CPT shakes the baby. There is no evidence to suggest that CPT is harmful for your baby and it clearly improves your health, so you should continue to do this or some other form of CPT while pregnant. Ideally, you should maintain these therapies to avoid or minimize exacerbations. If antibiotics are required, though, you will need to discuss their risks and benefits with your CF care team.

**Vitamin A (retinol) toxicity:** Careful monitoring of your vitamin A levels is necessary because high levels, especially in the first 3 months of pregnancy, can be associated with developmental anomalies in your baby. If you are taking vitamin A supplementation, your obstetrician will have you stop doing so during your pregnancy. Do not stop your other CF vitamins without discussing it with your care team and your obstetrician.

## Pregnancy in CF

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### UNDERSTAND THE IMPORTANCE OF MANAGING YOUR CARE

#### WHAT MEDICATIONS SHOULD I TAKE OR NOT TAKE WHILE I'M PREGNANT?

The medical regimen for women with CF is very complex. It often includes pancreatic enzymes, antibiotics, mucolytics, and anti-inflammatories. Many of these meds have not been tested during pregnancy, so safety information is based on animal studies, clinical experience, or knowledge about how they work. Risks and benefits should be weighed against the trimester of pregnancy and your health in discussion with your CF care team.

#### WHAT ARE THE OUTCOMES FOR MOTHERS WITH CF AND THEIR BABIES?

Several studies have evaluated outcomes of CF pregnancies. The majority result in live births, and the statistics do not appear to vary from those of women without CF. There is no clear increased risk of fetal demise or birth defects. The most common complication is preterm delivery, which in one study was associated with low pre-pregnancy lung function. Diabetes was also reported to be more common in women who delivered prematurely. Overall, adequate nutrition, well-controlled sugars, and reasonable pre-pregnancy FEV<sub>1</sub> are associated with better outcomes.

#### SHOULD I BREASTFEED?

You can breastfeed, and your milk should contain sufficient amounts of protein and sodium for your baby. Be aware that your energy needs are 110 to 200 percent greater than for women without CF your age.<sup>5</sup> If you do choose to breast-feed, ensure your own nutritional health is adequate and meet with your nutritionist or dietitian to develop a plan that will provide enough extra calories for you and your baby.

#### PLANNING FOR THE FUTURE

It is important to think about your long-term plan as a parent. Despite advances in CF care, CF is still a life-shortening disease. You should discuss this with your partner and make plans for the future.

#### GET HELP

You will have many questions about getting pregnant and what to expect during pregnancy. Work with your CF care team and your obstetrician to ensure a safe pregnancy. Support from your spouse or significant other, family, and friends to help you maintain your health during and after your pregnancy is imperative to ensure you will be able to enjoy motherhood. It is important to remember that you need everyone's help.

## Pregnancy in CF

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Being a new mother without having CF can be overwhelming. Add in the time for your CF therapies to the time it takes to care for a baby, and you can see that you will need help. Don't be afraid to ask!

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# Menopause in CF



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**We are going to talk about menopause. Isn't that great!?**

As women with cystic fibrosis get older, they will have menopause. Like puberty, menopause is a natural hormonal event with physical changes. The average age of menopause is 51 in the general population. For women with CF it may happen a few years earlier. Perimenopause is the time of transition from before menopause starts until one year after it is complete. This time may also occur earlier and last longer in women with CF — from a few to several years.

“The change,” as it is often called, describes the menopause experience. Heralded by decreasing hormones and menstrual cycle changes, menopause affects women in many ways. Like having CF, each woman will have a unique experience. But women with CF may wonder, “Is it menopause or is it CF?”

This article will help you learn about menopause, work with your health care providers and CF team, and maintain your best health during your menopausal years.

## **IN THIS SECTION:**

- ▶ **Learn about menopause.**
- ▶ **Work with your CF team.**
- ▶ **Get the help you need.**
- ▶ **Take care of yourself.**

## Menopause in CF

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### LEARN ABOUT MENOPAUSE

The word menopause literally means “end of monthly cycles.” Menopause is a time when a woman’s menstrual cycles usually spread out and gradually stop. The last menstrual period — considered the one that is followed by no bleeding for 12 months — marks the end of menopause. Perimenopause (“peri” means “around”) is the time right before menopause and one year after menopause is complete.

Changing levels of hormones cause the menstrual cycle and all sorts of other things to change. This can happen over time or all at once. Cycles may be shorter or longer, and bleeding can be heavier or lighter. Women may skip periods or have more spotting or bleeding than usual. During this crazy time of change, women may be fertile so birth control should be used if pregnancy is not desired.

As hormone levels change, women can experience many symptoms:

- › Hot flashes
- › Night sweats
- › Sleeping problems
- › Fatigue
- › Vaginal changes (dryness, thinning, less flexibility)
- › Incontinence
- › Urinary tract infections
- › Mood changes or depression
- › Change in sexual interest and response
- › Osteoporosis
- › Trouble concentrating or focusing
- › Achy joints
- › Heart sensations of skipping or racing
- › Variable and less predictable blood sugar levels

You may already have some of these problems and wonder what will happen for you. Just like your CF, your menopause will be different from that of other women.

## Menopause in CF

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Much has been written about menopause, but we know little about the experience in women with CF. Thanks to Anna Tsang, R.N., M.S.N., N.P., and her colleagues at the Toronto Adult CF Center, who studied the experience of women with CF, we have some information. Thirty-three women, averaging 42 years of age, answered questions about their menopause experiences. Women with CF in the survey had:

- › Menopause 2 to 3 years earlier than women without CF
- › Most notably, difficulty with sleep and depression
- › Increased respiratory symptoms

In a small survey like this, you can't get all the answers, but some important points were raised. The authors encourage CF care providers to learn about and discuss menopause with their female patients around age 40 and to work with other health care providers such as gynecologists, psychologists/psychiatrists, and primary care providers to support women during the transition.<sup>1</sup>

### WHAT ABOUT HORMONE REPLACEMENT THERAPY?

In the 1960s, doctors began giving hormone replacement therapy (HRT) for menopausal symptoms. It was thought that replacing decreasing levels of some hormones could reduce hot flashes, osteoporosis, urinary problems, heart disease, cancer, and even Alzheimer's. However, there were questions about the safety of taking hormones after menopause. In 1991, the Women's Health Initiative started clinically observing the effects of HRT on women. One part of the study looked at the effects of taking one specific hormonal combination (called Prempro®), the most common form of HRT. This part of the larger study was halted in 2002 when the risks (heart disease, stroke, blood clots, and breast cancer) were shown to be greater than the benefits.

HRT is now used with caution. It can be helpful for some women to reduce symptoms of moderate to severe hot flashes and to treat weak, fragile, or painful vulvar and vaginal tissues. It may also help boost sexual desire, concentration, memory, and a sense of well-being. Current guidelines are to use the lowest effective dose for the shortest time to ease debilitating symptoms. Women with CF, and all women for that matter, should carefully consider the risks and benefits of HRT and choose the best treatment plan with their gynecologist and care team.

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<sup>1</sup> Tsang A, et al. Perimenopause in CF: assessment of onset and symptomatology. Poster presented at: North American Cystic Fibrosis Conference; October 2007.



# Menopause in CF

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## RELATED LINKS

### [Toronto Adult CF Centre](#)

Perimenopause and menopause - [www.torontoadultcf.com/cf-information/hot-flashes-sleeplessness-nervousnessperimenopausal-symptomswhat-expect](http://www.torontoadultcf.com/cf-information/hot-flashes-sleeplessness-nervousnessperimenopausal-symptomswhat-expect)

## BEYOND THE BASICS – RELATED LINKS:

Women’s Health Initiative - [www.nhlbi.nih.gov/whi/](http://www.nhlbi.nih.gov/whi/)

## WORK WITH YOUR CF TEAM

You may be a “menopause pioneer” at your CF clinic. Questions may arise as you learn about and go through menopause. What can you do to work with your CF Team?

- › Continue your regular care and follow-up.
- › Your ongoing CF care monitors some concerns associated with menopause, such as osteoporosis. Bone density screening and vitamin and calcium levels are important to check and treat.
- › Let your care team know if you’re having new symptoms that you’re not used to — things like incontinence, sleep difficulty, fatigue, and mood changes might be perimenopausal changes.
- › Do some research and discuss what approaches you are considering with your CF team.
- › Ask your gynecologist and primary care doctor to communicate with your CF team.
- › Tell your CF team about any new medication, including herbs or over-the-counter supplements, that you are interested in taking or have been prescribed.
- › Try to keep a personal health record — a menopause tracker might be helpful for you. Here is an example from womenshealth.gov (see next page):

## Menopause in CF

**HEALTH TOOLS**

### Menopause Symptom Tracker

Use this chart to track your menopause symptoms.

Date	Symptom	How much this bothers me	Ideas I've tried to feel better	Questions for my doctor	New ideas to try

U.S. Department of Health and Human Services, Office on Women's Health

### GET THE HELP YOU NEED

There is a lot of information about menopause for the general population in books, articles, and the Internet. Some resources will be helpful, others not.

To get started, talk to your gynecologist and primary care provider. If necessary, you might need to switch health care providers to someone who is willing to learn about CF and who will communicate and work with your CF team.

Yearly gynecological exams are important. They should include pelvic and breast exams with pap tests every 1 to 3 years. Your gynecologist will do pap tests when needed and advise you when to have mammograms.

Your mental and emotional health are important. Some women find they can use some help during perimenopause. This may mean working with a clinic social worker or therapist, or joining a support group. Keep up your social support from friends and family. The activities you already enjoy can be especially important through menopause and beyond.

### RELATED LINKS

General information - [www.womenshealth.gov/menopause](http://www.womenshealth.gov/menopause)

Symptom tracker - [www.womenshealth.gov/files/assets/docs/charts-checklists-guides/menopause-symptom-tracker.pdf](http://www.womenshealth.gov/files/assets/docs/charts-checklists-guides/menopause-symptom-tracker.pdf)

## Menopause in CF

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### TAKE CARE OF YOURSELF

Many things that help with CF also help with menopause. Keep some of these things in mind in general and especially around the time of menopause.

Focus on nutrition and hydration — this is advice that helps keep you healthy in general and during menopause it's no less true. Good food choices, enough calories, and enough fluids will help fuel your body. Pay attention to foods (spicy) or drinks (alcohol) that might trigger hot flashes.

- › **Get exercise!** Your lungs will thank you and your hormonal body will too. Weight-bearing exercise (walking, dancing, sports, weight training) boosts bone density and strength, which helps to reduce the risk of osteoporosis.
- › **Get enough rest and prepare for hot flashes.** Hot flashes can interrupt your day and night. Dressing in layers and wearing natural fiber clothing may help. At night, a fan or open window may help. Doing your exercise during the day (not the evening) and keeping a regular bedtime routine may also help. If you are not getting sleep and your hot flashes are too hot to handle, talk to your doctor.
- › **If incontinence is a problem, here are some things to think about:** Avoid alcohol, caffeine, and acidic foods that can irritate the bladder. Beware during airway clearance, as coughing can cause bladder leakage which can increase after menopause — if you have to go, go before chest PT. Many women wear pads or panty liners for incontinence. Other strategies that help with urinary continence include Kegel exercises, physical therapy, vaginal support such as a pessary and weight loss (only for women who are overweight).
- › **Stay sexually active if you wish.** Vaginal changes from having less of the estrogen hormone can make intercourse uncomfortable. Personal lubricants can be helpful. Some women find that hormone therapy in the vagina (tablet or ring) helps. Discuss these and other options with your gynecologist.

Complementary or alternative treatments can ease the menopause transition. The following approaches can make a difference for some women:

- › Yoga
- › Meditation
- › Massage therapy
- › Acupuncture

Maintaining health and well-being can be challenging during perimenopause and menopause but look how far you've come! Like every challenge with CF, knowing what works for you, staying in tune with changes in your body, and communicating with your CF team and other doctors can make menopause a little easier.

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# Day-to-Day Time Management

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Adult life is awesome — and busy. With hobbies, school, work, relationships, and family demands, it can be a real drag fitting in all the cystic fibrosis stuff.

It's easy to make excuses and say you're too busy or to procrastinate on doing your treatments. But even if you're feeling well, you still have to do treatments as an investment in the long haul.

Whether you're trying to get more done in a day or just looking to chill out more, this section discusses ways to manage your time so your CF care is more doable and your life is more enjoyable.

## DID YOU KNOW ...

...About 50 percent of people with CF do not follow their doctor's treatment recommendations completely (G. Latchford, et al. 2009). That's probably because of time management challenges.

...We live in a rushed society: some 19 percent of meals in the United States are eaten in an automobile (CBS News Healthwatch, Car Cuisine, 2/11/09).

## Day-to-Day Time Management

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### IN THIS SECTION:

- ▶ **Learn time management tips** that keep you healthy while living a full adult life.
- ▶ **Work with your CF team** to coordinate your daily schedule.
- ▶ **Make decisions that help you use your time wisely.**
- ▶ **Forgive yourself**, because you simply can't do and be all things — but you can promise to do your best.
- ▶ **Get help before it's too late** and you run into a crisis.
- ▶ **Speaking from experience** — hear from other adults with CF.

## Day-to-Day Time Management

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### LEARN TIME MANAGEMENT TIPS

People with CF are required to spend a great deal of time doing treatments, eating well, exercising, and taking care of their medical needs. That time can take away from things you enjoy — like work, school, hobbies, friendships, and family.

Organizing your time to fit in your health needs can help make your CF health care seem less overwhelming and help you stick to your treatments. And the more time you spend taking care of yourself, the more time you'll have to do all the other stuff you want to do for many years ahead.

These time-management tips can help you stay healthy while living a full life:

- › **Multitask when possible.** While doing your nebulizer, put out your meds and clothes for the next day, fold laundry, etc. You can move your air compressor to the kitchen (or better yet, get a second machine for the kitchen), and do your nebulizer while packing tomorrow's lunch or washing your other nebulizer's parts.
- › **Shift your chores online when possible.** Order meds by email or use an auto-refill option, and go online to send questions to your health care providers, pay bills, order stamps, and shop — you can do all these chores while doing a breathing treatment.

“**I've resorted to auto-refill services. It's a major load off ... going to the local pharmacy 1 to 3 times every week.**”  
- **Eric, Age 34**

- › **Use technology tools.** Set your cell phone alarms, or use an online calendar such as Google or Outlook Calendar, to remind you about appointments or when it's time to order your meds. Set an alarm when it's time to start a treatment or get ready for bed. Regular alarms can help you get into a routine.

**There are very cool pager-like devices, computer programs, and vibrating pillboxes** that can help remind you to take medications and start treatments. Ask your CF health care providers about these or search online for “adherence management tools.”

## Day-to-Day Time Management

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- › **Don't waste time waiting.** When going to your CF clinic or other appointments where you'll know you'll be waiting, bring homework, a good book, paperwork from your job, or a laptop to make the most of your time.
- › **Learn to love lists.** Keep lists of reminders and things you need to do. As questions pop into your head for your CF health care providers, jot them down on a sticky note in your calendar on the date of your next appointment. This saves the time you'd need to call back with questions after the visit.
- › **Combine errands.** If you have a clinic appointment, maybe you can also pick up meds at the pharmacy, get your IV line flushed and then go to the store for tissues, calcium, or an oral supplement.
- › **Just say no.** When you have CF, you can't push yourself to do everything or your body will feel it. You have to know your limits and know when to say no. It's more courageous to admit you can't do something than to blindly endure and pay the price.

## WORK WITH YOUR CF HEALTH CARE TEAM

The folks at your CF care center are a great resource. Use them. Talk to them if you're struggling with the time it takes to do your treatments. Be honest. You're not a kid who is going to get in trouble.

Your CF health care team will understand how hard it is to fit it all in as an adult with a full life. They may have suggestions about ways to rearrange your treatment schedule to fit everything in. Ask them if you could you use a metered-dose inhaler instead of an aerosol inhalation; go for a jog instead of using the vest; or maybe substitute an hour of clarinet or drumming for airway clearance.

Here are some other points to talk about with your CF health care team:

- › Ask them what are **the absolute priorities in your medical care**. That way, if you do run out of time, you can focus on the most important treatments and your team will know that is what you are doing.
- › Ask if you can get a **portable air compressor** to use in the car. Also, ask if you can combine doing your aerosols with your chest vest. Be sure you are clear on which medications cannot be mixed together or used in certain devices.

## Day-to-Day Time Management

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- › Your CF health care team can offer **community resources to save your time**. If you qualify for a disabled parking placard, ask for the paperwork. If you are struggling to make time for health care, this may be a good time to talk to your social worker about working fewer hours or taking fewer classes or maybe even quitting your job or school and going on disability for a while. Focusing on your health is the same as focusing on your future. You can return to school or work when your health stabilizes.
- › If you are feeling unmotivated, lethargic, or hopeless, and those feelings are causing you to misuse time or not take the time to do treatments, **be honest with your CF health care team**. They understand how hard it is to live with CF. Sometimes having a real conversation about your **emotional barriers** can lead to ideas and help you get the emotional support you need.

### MAKE DECISIONS THAT HELP YOU USE YOUR TIME WISELY

You may not always realize it, but you have the power to manage your time. A lot of this power comes from the decisions you make every day.

- › **Set time goals.** For example, tell yourself, “Today, I’m going to do three treatments and 1 hour of exercise, plus spend 4 hours on this project.” Then schedule your eating, phone calls, and emails around those three priorities.
- › **Try a time journal.** Write down everything you do for several days, minute-by-minute. Look at where you are wasting time. What can you change? Where do you see a window of opportunity? For example, can you squeeze in 30 pushups and jumping jacks before getting in the shower? Can you start laundry before you start a treatment?
- › **To plan ahead, map out your whole day or week with tasks and priorities.** Plan from the moment you wake up until you go to sleep. What matters most to you? How can your time reflect your priorities? Set your priorities by reciting a mantra, like, “Health comes first, then school or work, then comes everything else.”

Since everyone with CF is different, you have to decide what’s most important to your health. Do your lungs come first, or exercise, nutrition, or mental health? Postpone anything on your to-do list that’s not urgent. Better yet, if there’s no space in your calendar, let go of things you “should” do or even want to do.

- › **Establish routines and stick with them.** Charles C. Noble said, “First we make our habits, then our habits make us.” So true! Once you know the schedule of your day or week, find a routine that works for you. Maybe a 9 a.m. exercise class that you pay for will motivate you to wake up at 7 a.m. and do a treatment, grab a quick bite, and head out the door. With practice and habit, you can look at the clock at 8 a.m. and know it’s time to start your treatment.



## Day-to-Day Time Management

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- › **Decide on time limits for tasks.** A trip to the grocery store should take 30 minutes max. Limit emails to 30 minutes in the morning and night. An aerosol should take about 20 minutes. If you find yourself distracted and notice your aerosol has been sputtering for 15 extra minutes, it might be helpful to set a timer when you start the treatment. Also, if you are feeling too busy to do a treatment, try doing an aerosol for 10 minutes. Commend yourself for just doing something. CF care is like exercise. It's best to do 30 minutes of exercise three times a week, but if you manage 10 minutes here and there it all adds up and is better than nothing.
- › **If you find yourself procrastinating at the start of a treatment, or going to bed, assess the situation.** Remember to do your "I hates" first. Ask yourself what you are avoiding. What are you getting out of delaying what you need to do? What are the consequences, positive and negative, of procrastination? What tools (friends, rewards, alarms) can you use to help out? When you are finally doing what you need to do, recite this quote over and over: "Heroism ... is endurance for one moment more." (George F. Kennan)
- › **Schedule breaks!** Life is so full when you are an adult. You need to make time to chill out. If you need to block out 2 hours of your calendar to nap or lie down and stare at the ceiling, that's OK! Your mind and body will be grateful for some stillness.

## FORGIVE YOURSELF

Don't be so hard on yourself. You simply can't do and be all things — and that's OK. You can only promise to do your best. Hopefully, these hints and ideas will help you avoid overload.

- › **Remember that CF takes more time and energy than almost any other disease.** You are being asked to be superhuman just to live. It's not fair, and you have the right to feel overwhelmed. If you can't do it all or if you slip on your routine, don't beat yourself up. Sometimes sleep is most important. You are probably not being lazy. If you are being lazy, do some soul searching as to why. You have reasons for doing what you choose to do. Just gently coax yourself back on track. It's never too late. Tell yourself how important your future is and how managing your time today will help add to your tomorrows.
- › **Plan mini-vacations.** Sometimes taking a road trip or weekend getaway helps break us from our monotonous routine. The change of scenery re-energizes us to recommit to our daily rituals. Doing your CF stuff at the beach, at a friend's house, or in a nice hotel can feel positively different.

## Day-to-Day Time Management

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- › **It's very normal for people with CF to feel a sense of urgency.** We want to see, do, and experience as much as we can. Deep inside, we may fear that our time on earth will be short. If that's the case, discovering your own time-management strategies now is critical. Remember that quality is more important than quantity. Doing one thing well and unrushed — to stay in the moment — is much more enjoyable than having scattered thoughts of what's next.

A more manageable schedule improves quality of life and increases time for health care, increasing quantity in turn. And with new treatments on the horizon, it's helpful to stay optimistic that you'll be around for a while. Memorize the quote, "Hope is patience with the lamp lit!" (Tertullian)

- › **CF can be a selfish disease.** Sometimes we may want to spend our time for others, but our health needs do not allow that. Accepting this is a process. The vast majority of your supporters will understand that you have to make yourself your priority. They'll want you to, so they can keep loving you for the long haul.
- › **CF invites us to "subvert the dominant paradigm," or, in plain English, challenge social norms.** American culture says, "An idle mind is the devil's workshop." Whether you have CF or not, is that really true? Why? It's OK for us to schedule downtime, rest, and relaxation without guilt. We're lucky to learn that lesson. It's good for the soul and the body!

## GET HELP BEFORE IT'S TOO LATE

Sometimes all of the treatments you need to do can feel completely overwhelming. There are ways to get help to lighten your load. Here are just a few:

- › **Get the gift of time.** Instead of material gifts for holidays and birthdays, ask your friends or relatives to give you money for a cleaning service and similar services that save you time.
- › **Designate or outsource.** The best way to save time is to give some tasks to others. Ask your friends if they can grab some groceries for you on their way to visit. Ask a co-worker to take on a little more of a project when you have medical appointments. Can you hire a dog walker? Can a partner wash your nebs or order meds for you? If you volunteered to bake cookies for a function but feel too tired, ask neighborhood kids to come help you out.
- › **Think about the return.** A good rule of thumb is to check your return on investment (ROI) before doing something. The ROI tells you how much you get back for how much you put in. If you get back less, it's time to give the task to others or drop it.

## Day-to-Day Time Management

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- › **Build your skills.** Consider taking a class on time management or organization, or go see a life coach who can help you structure your time in the healthiest way possible.
- › **Learn from the experts.** Contact other people living with CF online, through your CF care center education days or by telephone, and find out what they do. Only people with CF can give you real-life scenarios and tools that work. But remember, everyone with CF is different. Just because someone else can manage some things a certain way doesn't mean you have to do the same.

“**Half our life is spent trying to find something to do with the time we have rushed through life trying to save.**”  
- *Will Rogers*

### SPEAKING FROM EXPERIENCE

“With all there is to do for CF, make sure you do something normal every day. It's good for your spirit.” – *Anna, Age 29*

“I am a procrastinator by nature. But I've found having a set routine up to the very minute is helpful. Every day my alarm goes off at 8 a.m., and I don't allow myself to get out of bed past 8:05. I start my treatment no later than 8:20 a.m. and make sure I finish by 9:30.” – *Adult with CF*

“This is how my whole day goes. I do not let myself start my afternoon or evening treatment after a certain time, otherwise the rest of my day is off. The more routines I set, the more I am able to manage normal things like socializing and spending time with my fiancé.” – *Maggie, Age 23*

“I use a routine/medication checklist. I print the list and put it in a sheet protector. Each day I use a dry erase marker and slowly check off each part of my routine. In late-stage CF, there is a lot to remember. This checklist helps me relax and not have to worry about what I am forgetting.” – *Kristina, Age 35*

“I was working part time for a few years and returned to full time almost 2 years ago. It helps to be regimented. On weekdays, my schedule is usually wake up and do a sinus rinse immediately (it's a quicker alternative to coffee), then saline and vest. Then I'll eat breakfast since I don't like vesting on a full stomach. Then TOBI® or Cayston®, inhalers, then on to normal getting-ready stuff, like taking a shower, etc.”  
– *Adult with CF*

## Day-to-Day Time Management

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"I've gotten into the habit of taking all my BID [twice a day] pills with breakfast and dinner. This supposedly helps the absorption since it's combined with food and enzymes. I'll take an inhaler throughout the day at work when I start feeling extra tight or congested, and can luckily close my office door and blinds for 5 minutes while I take Cayston® in the afternoon." – *Adult with CF*

"I always keep enzymes and snacks in my desk, and I'll send emails to my personal email account when I need to bring in a new bottle. At night, I'll usually get my treatments and vest in after my dinner feels digested, or before I eat if I have something time-consuming later in the evening." – *Adult with CF*

"I've come to realize the extra time it takes for CF stuff is not that dramatic when I compare it with people with kids and long commutes. TOBI® months can be tough because it's an extra half-hour or so in the morning, but it's manageable. Though sometimes this can be too rough if I'm really not feeling well. I plan to work part time from home again one day when I'm really not feeling up to the CF and work challenges." – *Eric, Age 34*

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# Eating on the Go

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Chances are, most people you know are trying to keep pounds off. But you need more calories and nutrients than they do just to keep the pounds on. That's because cystic fibrosis makes it hard for the body to absorb fat and nutrients. But keeping your weight — and sometimes increasing it — is key to fighting infection and keeping your lungs strong.

We've put together this section to make it easier for you to "eat on the go." It turns out that small changes in your routines can make a big difference for your weight.

Click on any of the basics below to get more information.

## IN THIS SECTION:

- ▶ **Follow these planning and organizing tips** — including ways to organize your kitchen and your shopping.
- ▶ **Use these ideas for "grab 'n' go" breakfasts, mini-meals, and snacks.**
- ▶ **Work with your CF team** — to get advice on nutrition and keeping a healthy weight.
- ▶ **Check out these 15 proven recipes** — quick and easy shakes, smoothies, and frappés.
- ▶ **Get advice from other adults with CF** — including recipes and smart buys.
- ▶ **Check out these sites for more ideas.**

## Eating on the Go

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### FOLLOW THESE PLANNING AND ORGANIZING TIPS

#### FAST WAYS TO PLAN AND PREPARE MEALS

Quick planning and fast preparation are key to having meals and snacks ready to “grab ‘n’ go.” Here are some tips to help get you started:

- › **Before you go to sleep:** Think about the busy day ahead. Where will you be spending your time? Where can you easily stash food? Is there a refrigerator nearby? Microwave? How about a place to eat?
- › **Think about “packability”:** Plan meals or snacks that you can carry in your backpack, purse, or briefcase and store in your desk drawer, locker, or a cooler in your car. Buy a variety of foods so that you don’t get tired of eating the same foods every day.
- › **Cook once to eat three times:** When cooking, make enough to pack a meal for tomorrow’s lunch or use plastic containers to freeze meals that you can easily grab ‘n’ go.

#### ORGANIZE YOUR KITCHEN

- › **Keep ‘em within arm’s length:** Organize your kitchen so that everything you need for grab ‘n’ go snacks and meals is within arm’s reach. Keep in stock things like paper bags, plastic bags, napkins, and food containers.
- › **Give them a shelf of their own:** Create a shelf in your kitchen and/or refrigerator just for your grab ‘n’ go favorites.
- › **Pack in plastic:** Keep plastic containers on hand to store meal-sized portions in the refrigerator or freezer. In the morning, just grab a filled container to take to work or school or wherever you go.

#### ORGANIZE YOUR SHOPPING

- › **Buy single-packet servings:** Buy peanut butter, jelly, cream cheese, and other foods in single-packet servings you can just toss in your bag. If you have trouble finding these in your grocery store, check out the countless varieties available online at restaurant supply stores. (Use the search phrase “restaurant supplies condiments.”)
- › **Buy in bulk to save:** You can save money by buying in bulk at discount stores and individually wrapping foods yourself in plastic wrap, plastic bags, or foil.
- › **Read food labels:** Food labels will help you learn to choose foods that meet your goals. For more on food labels, see the [U.S. Food and Drug Administration website](#).

## Eating on the Go

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- › **Keep an eye out for new ideas:** Just look around you. Grab 'n' go options are everywhere. Check out the selection in convenience stores, vending machines, corner markets, food stands, even bookstores and sporting events.

### REMEMBER YOUR ENZYMES

Be sure to always have your enzymes with you so that you can eat at every opportunity.

## USE THESE IDEAS FOR "GRAB 'N' GO" BREAKFASTS, MINI-MEALS, AND SNACKS

### GRAB 'N' GO BREAKFAST

The morning routine can be stressful. Leaving the house hungry isn't a good idea. With a little planning, you'll be able to make meals you can just grab 'n' go.

- › Scramble an egg with cheese, wrap it in a tortilla, and off you go.
- › Microwave a breakfast sandwich while you are dressing.
- › Keep containers of shakes, yogurt drinks, and other high-calorie beverages in your book bag or briefcase.
- › Buy giant muffins in bulk, wrap each in its own bag, and freeze.
- › Fill a water bottle with your favorite beverage or shake each night before you go to bed. Grab it before you head out in the morning.
- › Make a batch of French toast or pancakes, wrap individual servings, and freeze. In the morning, pop a serving in the microwave.
- › Keep single-serving oatmeal (in a packet or a prepackaged insulated bowl) in your pantry or cupboard. Just add hot milk and take it with you.
- › Buy cold cereal in individual containers (bowls or boxes) or pour your favorite cereal into a plastic container. Take along single servings of boxed liquid milk (the kind that does not need to be refrigerated).
- › Keep a bowl of fruit by your house or car keys. Grab a banana, orange, or apple on your way out the door.

## Eating on the Go

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### GRAB 'N' GO MINI-MEALS

- › Top bagel halves with spaghetti sauce and shredded cheese.
- › Make sandwiches (PB&J, ham, turkey) at the beginning of the week and freeze them. Toss one in your bag and let it thaw during the day. You can also look for frozen sandwiches in the grocery store.
- › Think about refrigerated wraps or burritos. Buy microwavable burritos, so you can wrap one in a paper towel, heat it, and run.
- › Pack microwavable instant soup, instant noodles, and fun-size containers of spaghetti and meatballs and macaroni and cheese.
- › Try a tuna kit (comes with tuna fish, crackers, and mayo).

### GRAB 'N' GO SNACKS

Keep these snacks cool with an ice pack in an insulated lunch bag or in a cooler:

- › High-fat deli meat and cheese “roll-ups”
- › Cheese sticks and single servings (peel-and-eat varieties such as gouda, cheddar, and string cheeses)
- › Single servings of whole-milk cottage cheese
- › Whole-milk yogurt and yogurt drinks
- › Hummus in a small container and pita bread cut into triangles
- › Single-serving canned pears, peaches, or fruit cocktail

### KEEP THESE ANYWHERE:

- › Trail mix
- › Granola, protein, and snack bars
- › Fig bars
- › Cheese and cracker packs
- › Shakes, canned or bottled
- › Individual peanut butter packets
- › Muffins
- › Graham crackers, vanilla wafers, gingersnap cookies, animal crackers



## Eating on the Go

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- › Nuts (peanuts, cashews, almonds, walnuts, macadamia nuts)
- › Sunflower seeds
- › Raisin bread
- › Pretzels or chips
- › Cereal
- › Single-serving juice or milk boxes
- › Dried fruit
- › Bottled coffee drinks
- › Pudding snacks (some brands do not have to be refrigerated)
- › Hot cocoa mix
- › Fresh fruit

### WORK WITH YOUR CF TEAM

Life is so busy. You may have school or work and all of the responsibilities that go with everyday life. You also need to find time to fit in all of your lung treatments, plus eat enough to maintain a healthy weight.

Just how much should you weigh? That answer will vary, but the CF Foundation recommends that women maintain a body mass index (BMI) of at least 22 and men a BMI of at least 23.

[Calculate your BMI with this online BMI calculator from the Centers for Disease Control and Prevention](#). You can also ask about your BMI the next time you visit your CF center.

**To gain weight, you need about an extra 500 calories every day.**

Spend some time thinking about what your weight gain goal should be. Talk with your center's dietitian (nutritionist) and ask about ways you can reach your BMI goal. Your CF team can also suggest some simple ways to add calories to the food you already eat.

## Eating on the Go

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We're not talking about anything out of the ordinary. You can get those extra 500 calories from foods like these:

- › A grilled ham and cheese sandwich
- › A small bowl of spaghetti with sauce and cheese
- › A cup of macaroni and cheese
- › A large order of fries
- › A large muffin

As you can see, nothing on this list would cost much to make or take much effort. In other words, you can get your 500 extra calories eating on the go. And be sure to have your enzymes with you so you can eat at every opportunity.

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**[Learn more about nutrition for adults with CF from CF Education Webcasts.](#)**

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### CHECK OUT THESE 15 PROVEN RECIPES

## FORTIFIED MILK

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**SERVES: 4**

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### INGREDIENTS

- Quart whole milk
- 1 cup instant non-fat dry milk

### DIRECTIONS

Pour liquid milk into deep bowl. Add dry milk and beat slowly with mixer until dry milk is dissolved (usually less than 5 minutes). Refrigerate. The flavor improves after several hours.

Calories: 209  
Protein: 14 g  
Carbohydrate: 20 g  
Fat: 8 g  
Calcium: 461 mg

## PUDDING SHAKE

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**SERVES: 4**

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### INGREDIENTS

- Quart whole milk
- 1 cup vanilla ice cream (slightly softened)
- 1  $\frac{3}{4}$  ounce package instant banana cream, vanilla, or chocolate pudding and pie filling mix (or substitute your favorite flavor)

### DIRECTIONS

Place all ingredients in blender container. Cover and blend at medium-low setting until smooth and thick, about 30-45 seconds. Refrigerate unused portion.

Calories: 294  
Protein: 12 g  
Carbohydrate: 35 g  
Fat: 12 g  
Calcium: 389 mg

### BANANA SHAKE

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**SERVES: 2**

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#### INGREDIENTS

- 1 cup fortified milk (well chilled)
- ½ cup vanilla ice cream
- 1 small ripe banana, sliced
- ½ teaspoon almond flavoring (optional)

#### DIRECTIONS

Combine all ingredients in a blender. Blend for 10-15 seconds. Serve immediately or chill in freezer until ready to serve.

Calories: 249

Protein: 9 g

Carbohydrate: 32 g

Fat: 10 g

Calcium: 277 mg

### COCOA-NUT SHAKE

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**SERVES: 2**

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#### INGREDIENTS

- 1 cup fortified milk
- ½ cup smooth peanut butter
- ½ cup chocolate ice cream
- 2 teaspoons vanilla

#### DIRECTIONS

Place all ingredients in a blender. Blend for 15 seconds until thick. Serve immediately or chill in freezer until ready to serve.

Calories: 570

Protein: 24 g

Carbohydrate: 32 g

Fat: 41 g

Calcium: 297 mg

### INSTANT BREAKFAST

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**SERVES: 1**

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#### INGREDIENTS

- 1 cup fortified milk
- 1 package instant breakfast (flavor of choice)

#### DIRECTIONS

Empty contents of 1 package of instant breakfast into a large glass. Add 1 cup of fortified milk and stir.

Calories: 339  
Protein: 19 g  
Carbohydrate: 48 g  
Fat: 8 g  
Calcium: 711 mg

### STRAWBERRY BANANA SHAKE

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**SERVES: 1**

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#### INGREDIENTS

- 1 package instant breakfast (strawberry)
- 1 cup fortified milk
- 1 small ripe banana

#### DIRECTIONS

Place all ingredients in a blender. Blend for 10-15 seconds.

Calories: 444  
Protein: 20 g  
Carbohydrate: 74 g  
Fat: 8 g  
Calcium: 645 mg

### PINEAPPLE SURPRISE

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**SERVES: 2**

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#### INGREDIENTS

- 1 package instant breakfast (vanilla)
- 1 cup fortified milk
- ½ cup pineapple-orange juice

#### DIRECTIONS

Place all ingredients in a blender. Blend until smooth, about 20 seconds. Chill.

Calories: 202

Protein: 10 g

Carbohydrate: 32 g

Fat: 4 g

Calcium: 443 mg

### COCOA-NANA BREAKFAST DRINK

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**SERVES: 1**

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#### INGREDIENTS

- 1 cup fortified milk
- 1 package instant breakfast (chocolate)
- 1 small banana (cut up)

#### DIRECTIONS

Combine all ingredients in a blender. Blend for 10-15 seconds.

Calories: 454

Protein: 19 g

Carbohydrate: 74 g

Fat: 10 g

Calcium: 717 mg

### CHOCOLATE-PEANUT BUTTER MILKSHAKE

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**SERVES: 2**

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#### INGREDIENTS

- 2 cups ice cream (chocolate or vanilla)
- 2½ tablespoons peanut butter
- 3 tablespoons chocolate syrup
- ⅓ cup half-and-half
- 1 teaspoon vanilla flavoring

#### DIRECTIONS

Place all ingredients in a blender. Blend about 15 seconds.

Calories: 617

Protein: 12 g

Carbohydrate: 57 g

Fat: 39 g

Calcium: 225 mg

Optional: Top with Oreos and add 53 calories per cookie.

### PEACH YOGURT SHAKE

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**SERVES: 2**

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#### INGREDIENTS

- 1 cup sliced peaches
- 1 cup plain yogurt
- 1 cup whole milk
- 1 tablespoon honey

#### DIRECTIONS

Combine all ingredients in a blender and blend until smooth.

Calories: 230

Protein: 8 g

Carbohydrate: 33 g

Fat: 8 g

Calcium: 272 mg

Variations: Instead of peaches, use 1 cup sliced bananas, fruit cocktail, strawberries, raspberries, or blackberries.

### INSTANT BREAKFAST FRAPPÉ

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**SERVES: 1**

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#### INGREDIENTS

- ½ cup fortified milk
- ½ cup vanilla ice cream
- 1 package instant breakfast (flavor of choice)

#### DIRECTIONS

Place all ingredients in a blender. Blend until thoroughly mixed.

Calories: 419  
Protein: 15 g  
Carbohydrate: 54 g  
Fat: 16 g  
Calcium: 568 mg

### STRAWBERRY- BLUEBERRY SMOOTHIE

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**SERVES: 1**

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#### INGREDIENTS

- ¾ cup blueberry yogurt (do not buy “light” yogurt)
- ⅓ cup strawberries (fresh or frozen)
- 3 tablespoons whole milk
- 2 teaspoons sugar

#### DIRECTIONS

Combine all ingredients in a blender and whip to desired thickness.

Calories: 254  
Protein: 8 g  
Carbohydrate: 51 g  
Fat: 3 g  
Calcium: 208 mg



### APPLE PIE À LA MODE

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**SERVES: 2**

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#### INGREDIENTS

- 1 cup apple pie filling
- ½ cup whole milk
- 1 cup vanilla ice cream
- Dash of cinnamon

#### DIRECTIONS

Mix in blender.

Calories: 336

Protein: 5 g

Carbohydrate: 49 g

Fat: 14 g

Calcium: 156 mg

### MARIA'S OREO MILKSHAKE

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**SERVES: 2**

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#### INGREDIENTS

- 2 double-stuffed Oreos
- 2 tablespoons marshmallow fluff (cream)
- 1½ cups vanilla ice cream
- ½-1 cup whole milk (vary depending on desired consistency)

#### DIRECTIONS

Mix in blender.

Calories: 429

Protein: 7 g

Carbohydrate: 49 g

Fat: 24 g

Calcium: 196 mg

# SHERBET SHAKE

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**SERVES: 1**

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## INGREDIENTS

- $\frac{3}{4}$  cup nonfat milk
- 1 cup sherbet (flavor of choice)

## DIRECTIONS

Combine all ingredients in a blender.  
Blend about 10-15 seconds.

Calories: 262

Protein: 8 g

Carbohydrate: 53 g

Fat: 3 g

Calcium: 245 mg

Note: This is a lighter shake than a typical milkshake and is also suggested for a fat-restricted (or cholesterol-restricted) diet.

## Eating on the Go

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### GET ADVICE FROM OTHER ADULTS WITH CF

Christian, a 19-year-old college student with CF, offers these suggestions for other students:

1. Get the unlimited meal plan — this is very important.
2. Always keeps easy-to-prepare foods in your room, such as canned pasta (Chef Boyardee®), high-calorie frozen dinners, and crackers for peanut butter.

"I regularly eat chicken breasts and lean proteins to keep myself healthy. I also eat a lot of foods that have omega-3s in them such as salmon and other natural fatty acids. I like to get a lot of my nutrition from whole foods like whole wheat grains. Complex carbs are absorbed better and you have more body fuel throughout the day without spiking your blood sugar, which can be an issue with people who have CFRD.

"I generally don't eat too much junk food and have a minimal soda intake. I usually stick to water or a sport drink on a daily basis, which has made a huge difference in my health. Eating nuts and fruits is also a huge factor in staying healthy. The way I look at it, food is medicine and we need to take our medicine. We need to eat proper diets and not necessarily loads of junk food. However, I find sometimes you have to cheat a little bit or else you'll not want to eat the proper foods on a daily basis. One of the things that inspired me to eat healthily was to get out of the monotonous grocery stores and go to local markets, where you can see all the healthy food together. Having a good sense of what you are supposed to be eating just makes you feel better about what you're doing."

– Frank, Age 23

### CHECK OUT THESE SITES FOR MORE IDEAS

These sites can provide more ideas and help when it comes to eating on the go:

Columbia University Medical Center:

[www.hdny.org/recipes.html](http://www.hdny.org/recipes.html)

TeensHealth:

[kidshealth.org/teen/recipes/cf\\_recipes/about\\_cf\\_recipes.html](http://kidshealth.org/teen/recipes/cf_recipes/about_cf_recipes.html)

Great list of high-calorie recipes.

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# Exercise With CF

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Regular exercise is tough, and we all find reasons to avoid it. But regular exercise makes sense, for some pretty basic and powerful reasons:

- ▶ **Better overall health:** Our bodies were made to be active. When we become inactive all sorts of bad things happen, like increased risk for heart disease, diabetes, and cancer.
- ▶ **More energy:** When you improve your heart, lung, and muscle function, you have more energy for daily tasks, as well as daily cystic fibrosis therapy.
- ▶ **Better lung function:** Regular exercise can improve your ability to get mucus out of your airways and cough it out.
- ▶ **More social engagement:** Taking part in physical activities with others is a good way to motivate yourself and a great way to maintain social relationships.

## Exercise With CF

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Click on any of the “basics” below to get more in-depth information, tips, and tricks.

### IN THIS SECTION:

- ▶ **Learn about exercise in CF** — why it makes sense and how to get started.
- ▶ **Work with your CF team** to develop the best program for you.
- ▶ **Follow these management tips** to track your progress.
- ▶ **Use the right approach** — don't let excuses hold you back from better health.
- ▶ **Speaking from experience.**

# Exercise With CF

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## LEARN ABOUT EXERCISE IN CF

### THE BASICS

- › **It's a journey:** Fitness and good health are not destinations, but rather a lifelong journey you travel by doing regular exercise.
- › **Lots of benefits:** There is clear scientific and clinical evidence that regular exercise provides multiple benefits for those with CF. These benefits are more than better lung function — it also helps in managing diabetes and heart disease.
- › **Active but not uncomfortable:** Do some exercise on purpose at least three to four days a week at an intensity that allows you to talk during the activity. You don't want to be so out of breath when you exercise that you can't talk. Any exercise is better than none, but it is possible to overdo it. As a guide, stay comfortably active by doing a variety of activities that last more than 10 minutes (aerobic exercise), even if done with some rest pauses. On one or two of these days, include some resistance activities (like weight lifting).
- › **Make it sustainable:** The three keys to developing an exercise program you'll keep doing are selecting a variety of activities that you know you will enjoy, finding a social setting in which to participate (partners), and talking with a person who knows CF and exercise to help develop a plan for you.

### BEYOND THE BASICS: WHY EXERCISE MATTERS

There is growing agreement in the medical community on the role of exercise both for preventing new diseases and for managing disease in many people. Just as the use of diet, medications, and therapy have a place in CF health, so too does exercise.

For everyone, the benefits of regular exercise and physical fitness have become well known. Recent research has shown that these benefits are not just from vigorous activities like marathon running and other competitive sports. Even moderate amounts of physical activity — like brisk walking and bicycling — can help extend life and help people stay active and independent. The benefits of regular activity are seen in women and men, older and younger adults, and in those who have health conditions, including CF.

The U.S. Department of Health and Human Services has federal [guidelines on physical activity for Americans](#). The guidelines encourage all adults to take part in at least 150 minutes per week of physical activity that is at least moderately intense. This activity is over and above the usual actions of daily living.

## Exercise With CF

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An example might be 30 minutes of brisk walking (moderate intensity) on three days, plus 20 minutes of jogging (vigorous intensity) on another day of the week. As physical fitness improves, adults are encouraged to do an activity longer or at greater intensity to get more health benefit. The guidelines also recommend that adults do resistance exercise, like weight lifting, two days per week. This type of activity provides more benefits that you can't get by aerobic exercise alone.

### WHY YOU, WHY NOW?

So, how does all of this apply to you? People with CF can safely exercise if done properly. It is clear that regular exercise will give you the same benefits as it does people without CF.

Even when you are in the hospital you should try to move about as much as possible. This will help you maintain your fitness, and you will do better when you are discharged. When you are in the hospital, ask your CF team to get an order written so someone can help you exercise while you're there.

The bottom line is that our bodies were made to move. Being inactive is not natural for your body and can lead to disease and disability. A car that sits unused in a garage for a long time likely will not function at peak performance, and the same is true for a human body that has long periods of being inactive. The good news is that most people can quickly fix inactive living through simple and low-cost lifestyle changes. Now, let's start exercising!

### WORK WITH YOUR CF TEAM

We often say one must walk before running. And it's smart to start by taking small steps toward an exercise program and a more active lifestyle. Start by thinking about your interests and abilities, then plan an exercise program, with help from your CF care team.

To learn about how a physical therapist can help, watch the [Partnering for Care webcast](#) on how a physical therapist can help you thrive.

Before you begin an exercise program, here are some basic steps to take:

- › **Determine your starting place.** Becoming physically active is a big goal. Too often, people start an exercise program but quickly quit. Change doesn't happen all at once, nor does it happen at the same rate for different people. Start by finding your "stage of readiness." A member of your care team can help you do this and start your exercise program.
- › **Get cleared for exercise.** People with medical conditions, including CF, should talk with their health care provider before beginning an exercise program. Your care provider may want you to do some tests first, depending on your level of activity and health.

## Exercise With CF

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- › **Test your current physical fitness.** Next, find out your level of physical fitness with the help of a member of your care team. Usually, it's smart to measure both aerobic fitness (cardiovascular or endurance fitness) and muscular fitness (muscular strength and endurance). Fitness testing may use simple tests like walking certain distances and doing calisthenics, or may be done in a laboratory with special exercise equipment. These tests help to:
  - Find your baseline exercise ability, which you will aim to improve or maintain.
  - Find the right exercise intensity for you and your goals.
  - Gauge your progress. At different stages of your program, you can repeat the test to see how far you've come.
  - Show if your oxygen levels decrease during exercise and, if so, at what intensity. This will help check the success of your exercise program. It can also give you and your CF team information about your CF lung disease, much like pulmonary function, nutritional status, and other clinical tests.
- › **Set goals.** Goal setting is one of the keys to success in changing your habits. The clearer and more realistic your goals, the more likely you are to achieve them (see the "Step It Up" section for more information on setting goals).
- › **Find out your exercise prescription.** You and your care provider will find out the specific amount of energy expenditure (calories burned) you want for your exercise routine. Energy expenditure may also be referred to as "exercise dose"; think of it like a dose of a medication. To find your desired exercise dose, you will consider type, intensity, duration, and frequency:
  - The **type** of exercise should be dynamic and use many major muscle groups. Examples are walking, jogging, swimming, dance, and stair stepping. When done at enough intensity and for enough time, these activities result in positive health benefits.
  - The **intensity** should be at least moderate. A simple way to monitor intensity is the "talk test." For moderate intensity, you should be able to converse comfortably while doing an activity that's raising your heart rate and breathing rate, and maybe even making you sweat a little. Vigorous intensity brings large increases in heart rate and breathing, definitely results in sweating, and also means you are not able to converse. Another common approach is to use a simple heart rate monitor. See the "Step It Up" section for more ideas.
  - The **duration** and **frequency** of exercise depend on two factors: your initial fitness level and the amount of exercise dose that you aim to achieve.



## Exercise With CF

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Your exercise dose is a function of the intensity, duration, and frequency of activity. It can be increased by performing exercise at higher intensity while keeping length of time and frequency constant or by performing exercise for longer times or at more frequent intervals while keeping the intensity constant.

For resistance exercise, the prescription lays out the type, repetitions, intensity, sets, and frequency of muscle-strengthening activities.

- › The **type** of activity generally involves free weights, weight machines, rubber exercise bands, or callisthenic exercises that use large, multiple muscle groups (e.g., bench press, leg press, push-up, lunge).
- › The **repetitions** are the number of times you can perform the activity at a given intensity.
- › The **intensity** is the amount of resistance (e.g., the amount of weight) that you work against. For free weights and weight machines, this is most often the amount of weight you can lift for a given number of repetitions (e.g., 8-12 times). You can determine resistance-exercise intensity from the resistance-exercise fitness test with the help of your care team. Other times, your body weight may serve as the resistance. Examples are when doing calisthenic exercises like push-ups, pull-ups, sit-ups, and lunges.
- › The **sets** are the total numbers of times you repeat a given lift (e.g., two sets of eight repetitions each).

Your health care provider can help you develop an exercise program tailored for your level of fitness, your exercise and health goals, and the equipment available.

You can learn more about exercise and physical fitness from MedlinePlus.

[www.nlm.nih.gov/medlineplus/exerciseandphysicalfitness.html#cat48](http://www.nlm.nih.gov/medlineplus/exerciseandphysicalfitness.html#cat48)

### ASK YOUR CF CARE TEAM:

A regular exercise test can help your care team find out the effect of CF on your lungs, cardiovascular system, and muscles. Regular testing, along with more planned exercise advice, should improve your care and your quality of life.

## Exercise With CF

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### STEP IT UP:

#### SETTING PROCESS AND PRODUCT GOALS

There are two major types of goals, product and process:

- › **Product goals** are the overall objectives of your exercise program.
- › **Process goals** are the in-between steps needed to get there.

Your overall goal may be to move from an inactive lifestyle to doing regular physical activities at levels recommended for health (e.g., 150 minutes per week of moderate to vigorous activity). To reach this long-term goal, you may set a series of process goals. For example, you might start with two brisk 10-minute walks on five days a week for two weeks; then do three 10-minute walks, five days per week in the next two weeks; and move on to a 30-minute walk, three days per week, plus a 10-minute jog, one day per week during the next two weeks, and so on.

The point is, you don't simply say, "I plan on exercising more every two weeks." Instead, you set a specific goal of exercises with specified intensities, duration, and frequency. You also plan for realistic time intervals (e.g., every Monday, Wednesday, and Friday) that can be monitored and modified as needed. Even if you don't fully reach your product goal, an exercise program still may produce meaningful changes in health through reaching the process goals.

### FOLLOW THESE MANAGEMENT TIPS

#### KEEP RECORDS TO TRACK YOUR PROGRESS

A critical part of the exercise program is keeping records of your daily activity habits. This will help you know if you are meeting both process and product goals (see "Step It Up"). Keeping records will be helpful if, every so often, you need to change your exercise if you are injured or when your progress is stalled.

One approach is to keep a written (or typed) exercise diary. A step counter, or pedometer, is an easy-to-use monitoring device. Low-cost, A step counter, or pedometer, is an easy-to-use monitoring device. These devices are usually portable and record the number of steps that you take throughout the day. Your aerobic exercise program can include a target number of daily steps, often set at 10,000 steps per day, which may help motivate you to reach your exercise goals.

## Exercise With CF

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### GET PERIODIC RETESTING

Repeating the physical fitness testing you did with your care team before starting your exercise program will give you important information on how exercise is helping your body. You can use this information for modifying the exercise prescription during the program and for evaluating the overall success of the exercise program. The results from repeat fitness testing also give your health care provider information to use in assessing your disease status. It may help you and your health care provider decide on whether to begin new CF therapies or modify your current therapies.

### WHAT KIND OF EXERCISE IS BEST?

In the past, many people assumed that only vigorous activities such as running would benefit health and physical functioning. Scientific research has debunked this myth. It has been shown that the type of activity is less important than doing regular exercise.

Select an activity that you enjoy and thus are more likely to regularly do. Walking, swimming, jogging, running, ice-skating, snowshoeing, elliptical strider, and tennis are examples of aerobic activities that are at least moderate intensity and, if done long enough, will result in benefits for your health.

### WHAT ABOUT STRETCHING?

As part of your cooling-down period after exercise, think about doing some gentle stretches. Not only will these stretches help you wind down after exercise, they may also improve your flexibility. This [slideshow from the Mayo Clinic](#) demonstrates some of the more important stretches to include in your routine. The CF Foundation webcast "[Exercise and CF](#)" also shows you some simple stretches.

### IS EXERCISE SAFE?

Regular exercise is a good health habit. But the benefits can only be felt after thinking about and talking with your doctor or physical therapist about any risks that may result.

The most common risk during exercise is musculoskeletal injuries, such as sprained ligaments, strained muscles, and overuse injuries. Injury risk is higher in those with a history of previous musculoskeletal injury and is associated with exercise intensity. The risk of exercise-related cardiovascular complications (e.g., cardiac arrest) is quite low. Cardiac events during exercise are most likely to occur in people with existing cardiovascular disease and in those who are inactive and out of shape.

## Exercise With CF

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### **The risks of exercise can be reduced through these sensible habits:**

- › Warm up before and cool down after exercise.
- › Slowly increase exercise amount and intensity toward your goal. You don't want to start walking three miles (your goal) when you haven't even walked four blocks regularly.
- › Pay attention to sensations or responses during exercise that may be a sign of injury.

### **Talk to your doctor or physical therapist before exercising if you have:**

- › Cor pulmonale (failure on the right side of the heart) or pulmonary hypertension.
- › Symptomatic heart disease.

### **Modify your exercise program when you have:**

- › An exacerbation or acute respiratory infection.
- › A musculoskeletal injury or condition that limits mobility.

### **WHAT ELSE YOU CAN DO:**

- › Get the most out of your exercise program by paying close attention to your diet. Eat a balanced diet that has enough calories to meet your energy needs.
- › Seek ways to engage with others in your activity; join a hiking club, fundraising walks, and organized runs.
- › Reduce your inactive time — sitting at the computer, playing video games, watching TV — and replace that time with more active pursuits.

Remember, the severity of your lung disease as well as certain medications may change how your heart rate responds to exercise. You should always talk to the physical therapist or exercise specialist on your care team to be sure you are monitoring your exercise intensity correctly.

## Exercise With CF

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### WANT TO CHALLENGE YOURSELF EVEN MORE?

If you are thinking of doing a competitive sport, keep these things in mind:

- › Find an environment where you will get positive feedback. This means that the activities are supervised or refereed properly and that the competition does not pose special risks.
- › Avoid competitive environments that put pressure on you to continue activity when your lungs or body might be telling you to stop.
- › Winning is OK, but not at all costs.

Many adults with CF are able to participate in strenuous sporting events, including marathon running. Talk about these activities with your health care team before starting on an ambitious exercise program.

### USE THE RIGHT APPROACH

#### DON'T LET EXCUSES HOLD YOU BACK FROM BETTER HEALTH

Let's talk about how to get over one of the biggest hurdles to exercising: excuses. We have all heard just about every excuse imaginable — we may have even used them ourselves. So, to cut to the chase, just pick the excuse that works most often for you!

#### ***"I don't have enough time."***

You do need to take time out of each day for exercise, but it is likely that you already have developed good daily time-management skills for your regular CF therapy. You should think of regular exercise as an extension of your plan to stay healthy with CF.

#### ***"It hurts too much."***

When overdone, exercise can hurt. But remember: you don't have to become fit in one week. Take the long view and improve your level of exercise over a longer period. This will help you avoid the exercise intensity that causes muscle ache and pain.

If you do little exercise right now, start with 5 to 10 minutes of easy walking, three to four times a week. Then, over a period of several months, increase the walking time to 30 minutes, five to six times a week. Although a bit more exercise than this may provide additional health benefits, be careful not to exercise to the point that you become overly breathless. It is important to talk with your care providers about your exercise plan.

## Exercise With CF

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### ***“I simply don’t like to exercise.”***

Well, you share this opinion with others. There is no question that exercise does take some effort. But so does taking your daily CF medications or doing your other health care tasks. We bet you don’t really like to do those either. But you do them anyway because you know they are good for you. Exercise is no different.

But ... you have some choices with exercise, and this is an important difference from your other health care needs. You can choose from a wide variety of activities, or do only those that you like to do, or do multiple activities. Maybe you can walk several days a week through a pleasant park or woods and then play tennis or some another game on other days. Don’t limit yourself. Be creative and ... HAVE FUN!

### ***“I can’t find anything I like to do.”***

OK, now you are really pushing the excuse limit. There must be something you like to do. Do you have a friend that you like to talk with regularly? Go for a walk and do your talking — either by walking together or by calling your friend on your cell phone. Join your local gym and try a variety of activities. You are likely to find one or two things that you enjoy and can do at different times in the week for some variety.

### ***“The dog ate my exercise program.”***

Of course, this is a pretty lame excuse, but it does bring up an important fact: you should consult with someone who understands CF and exercise so they can help you set up an exercise program.

## STEP IT UP

Still trying to convince yourself that regular exercise will be good for you? Check out these facts.

- › Exercise stimulates coughing. Don’t try to suppress this cough as it can help you get rid of excess mucus — a good thing!
- › Regular exercise helps keep CFRD in better control and can help control and prevent other diseases, like bone disease.
- › Exercise strengthens your breathing muscles so you are stronger if and when you get sick.
- › Exercise helps you build muscle and have stronger bones.

**Remember:** Stay hydrated! Since you will lose more salt in your sweat when you exercise than people without CF do, you need to pay particular attention to hydration. Consider adding ½ teaspoon of salt to a quart of water or ¼ teaspoon of salt to a quart of sport drink.

## Exercise With CF

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### SPEAKING FROM EXPERIENCE

"I was diagnosed with CF at the age of 4. My physicians told my parents to treat me as a "normal" child, and prepare for the day when I would be much sicker. As a result, I was enrolled in T-ball, town soccer, ice skating, and swimming lessons, and learned how to ski.

"When I reached high school and was cut from the school's soccer team, I began running cross-country and track. I was not fast. My apparent lack of talent did not deter me. Given that I had no chance of winning races or even scoring for my team, all of my running goals focused on self-improvement.

"From the very earliest days, I remember focusing on setting personal records. By my senior year of high school, I had greatly improved, and ran a respectable 5K at a little over 17 minutes. Not record breaking, but fast enough to make running at the NCAA Division III level a possibility. I continued running through college, gradually improving.

"Throughout my adolescence and early adulthood, the desire to run faster or make a team resulted in me wanting to do my medication routine.

**“When I look in the mirror today, I don’t see a sick person. I see an athlete, a law school graduate, and a husband.”**  
**– Chris, Age 31**

"Running competitively taught me how to set attainable short-term and long-term goals, and how to do the work required to accomplish them. I was able to develop a self-image centered on ability in the face of obstacles. I learned that CF does not prevent me from reaching my goals — be they athletic, academic, or professional.

"I am now an avid cyclist. I have completed a half-ironman triathlon and cross-country ski marathons, ridden my bike 800 miles through the Canadian Rockies, and completed dozens of 100-mile rides. When I look in the mirror today, I don't see a sick person. I see an athlete, a law school graduate, and a husband.

"Setting goals around physical activities motivates me to do my other daily treatments and reduces the anxieties I feel from living a life with CF. I want to be able to ride my bike. I'm willing to work incredibly hard to do what I can to maintain that ability.

## Exercise With CF

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"I focus on doing what I can do today, and what I'm going to do tomorrow. As long as I take one day at a time, I don't become consumed by the unknowables of a future clouded by CF, and in the process, live an incredibly rich and full life."

– Chris, Age 31

"Regular exercise has become an important part of my CF (life) care. Depending on my body's needs and where I am at the time, my choice about what activity to do can have a wide range.

**The continued push to find joyful activity that I CAN do keeps me healthier, grateful, and gives me a reason to smile each day.**  
– Kori, Age 32

"Sometimes it just needs to be a lap around the hospital unit; sometimes it's a walk around the block or housework with music. Sometimes it can be 20 minutes of dancing or even an hour of hula-hooping with friends. I also love to bike, swim, longboard, snowboard, and hike. I like to mix it up, doing as much as I can. The continued push to find joyful activity that I CAN do keeps me healthier, grateful, and gives me a reason to smile each day."

– Kori, Age 32



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# Having a Fabulous Social Life

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Whether it's a best friend from college, that group that goes out every Friday night, or the teammates you work out with, friends are people who make daily life rich and meaningful.

Fortunately, the vast majority of adults with cystic fibrosis enjoy normal and fulfilling social lives. But when you have CF, interactions with friends can sometimes seem particularly supportive or stressful. We have to face the fact that we are different than our healthy peers, and we don't want to be.

When CF makes us different, it can feel bad. It can be hard to explain CF to your healthy friend or to know when to turn down invitations that might be unhealthy for you.

But don't worry. This section of the Adult Guide is packed with insights and ideas for making the most of your social life.

## IN THIS SECTION:

- ▶ **Learn how to manage your social life** and how you can benefit by being open with your friends about your CF.
- ▶ **Let go of negative peers** who don't understand you and encourage you to participate in unhealthy behaviors.
- ▶ **Work with your CF team** to learn about ways your friends can support you.

## Having a Fabulous Social Life

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- ▶ **Get help when you need it** — it's more than OK to depend on those around you.
- ▶ **Make time for your social life** by planning and scheduling.
- ▶ **Identify what to say and resources** for social disclosure.
- ▶ **Speaking from experience** — hear from other adults with CF.

### LEARN HOW TO MANAGE YOUR SOCIAL LIFE

Friendships can play a major role in your well-being. But CF can make some social situations tricky or awkward.

While it may not always be easy or natural for you, talking about CF with your friends is a great way to connect with them on an even deeper level. And, like many things in life, a little humor can go a long way in making everyone more comfortable!

Here are some tips and thoughts about taking control of your social life and reaping the rewards:

- ▶ **Get comfortable about opening up.** Although the teenage years are typically the time when people with CF are most insecure, adults of all ages may think twice about letting people know about their disease. Choosing to tell your peers about your CF is an individual choice. There are no rules. You have to follow your heart and trust your instincts.

It helps to look at the pros and cons of telling your social group about your CF. Think about what you are afraid of. Would it be a relief that you wouldn't have to hold in a cough, hide an IV (intravenous line), or have people make harsh comments after you come out of the bathroom? Or is it better to be just one of the gang who won't be treated any differently? Are you a private person generally? Sometimes it depends on the person you are telling.

- ▶ **Find relief in telling your CF story to others.** You can stop explaining yourself or making excuses. You can show them who you really are, and that you do not just take time off, flake out, or arrive late to dates because you are careless. And your friends can support you in your CF journey — like helping you with treatments or joining you at a doctor's appointment.

It can feel really good to joke and share laughter about all the stuff that goes along with CF — for example, warning them "Do not go in there!" Chances are, your friends who know about your CF are inspired that you are living a full life.

## Having a Fabulous Social Life

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Since not very many people have CF, you can act as a CF ambassador — educating people about what this disease really is. Inviting your peers to a [Great Strides walk](#) can be a nice way to start the conversation. They can help find the cure!

- › **Build stronger, deeper friendships.** Believe it or not, sharing your CF story with others can open the doors to even better relationships. You'll be educating healthy peers about diversity and disability. Many people appreciate the closeness that comes from talking about life challenges with each other. Besides, you also are inviting your friends to share something personal about themselves with you!
- › **Beware of unhealthy situations.** There will be times when you have to make quick decisions. Would you visit a friend who has a sick baby at home? Can you pull an all-nighter on the town? Will your friends pressure you to smoke, drink, or use illicit drugs?

Make choices that are in your best long-term interests. Charm and humor can help get you through difficult social situations. You'll develop your own communication style with practice.



**I've seen and met angels wearing the disguise of ordinary people living ordinary lives.**

**– Tracy Chapman**



### LET GO OF NEGATIVE PEERS

Sometimes friends just won't get it. When there's conflict in a friendship, adults with CF may tend to think their disease caused the tension. If a friend grows distant, it may or may not be due to your CF. Either way, it's not your fault. Friends come in and out of everyone's life. You deserve healthy, supportive, and loving friends who are there for you at every step of your life's journey. Put the problem in the other person's court. The unsupportive friend may need some time to figure things out, grow up, and get some perspective before he or she can be the kind of friend you deserve.

Unfortunately, not all friends will understand your health needs, like when you have to say no to outings. Some will get irritated waiting for you to finish a treatment. These are normal human reactions to situations that they can't control or don't understand. But nobody wants to be judged or rejected.

Experiences with unsympathetic friends can be tremendously hurtful. You may want to figure out your own way to explain your CF limitations to them ahead of time. You can practice what you'd say in your mind, out loud in front of a mirror or by writing a letter.

## Having a Fabulous Social Life

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Remember, your time and energy are very valuable. You deserve compassionate friends. Gradually move away from any unsupportive peers. Look instead for people who are caring, understanding, and helpful. You won't have to look far. You'll find them at work, school, faith communities, and music or art clubs — and in your neighborhood.

There are times when only people with CF can truly relate to what you are going through. Why not meet new CF friends online? There are numerous CF chatrooms and groups on Facebook, Yahoo groups, and other websites. The Internet is a great resource and place to find support and understanding from other people with CF while avoiding cross-infection risks.

### WORK WITH YOUR CF TEAM

Your CF team is a great resource to guide you through life's challenges and opportunities. Talk to your team if you are struggling with unsupportive peers. Your social worker can offer suggestions on how you can help them understand CF.

Your team may also have some handouts you can give your friends.

It's important to be honest with your CF team about the kinds of peer pressure you are facing. Your CF team can give you good advice on ways to balance your health with certain high-risk social situations. Feel free to ask them things like:

- › What should I do if all my best friends are smokers?
- › How much alcohol can I drink — and how do my meds interact with alcohol?
- › How do I manage my insulin if I drink with my friends?

Your CF health care team might also welcome another set of eyes and ears on your visits. If you find these visits overwhelming, ask a friend to tag along. He or she may appreciate being invited into your special world of CF.

### GET HELP WHEN YOU NEED IT

American culture values independence and self-reliance. But having CF can limit your independence and make you feel almost childlike at times. Try not to get discouraged. Everyone needs the help of others at some point — CF or no CF. It's perfectly OK to rely on others for help when you need it. Remember — it takes a village to live well with CF.

Keep these things in mind as you consider reaching out:

- › **Be clear with your friends** about what you need and want. Sometimes you may want no special treatment at all. Or you just need a supportive ear. Other times you may need a ride home from the hospital or help picking up medicine at the pharmacy.

## Having a Fabulous Social Life

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- › **Return the favor.** When you are feeling stronger, offer to do favors for your friends.
- › **Know your limits.** Most people with CF don't want to let their disease get in the way of life. It's a great attitude, but it can cause problems if your body needs a break. Don't push yourself because you are too embarrassed to say no or you want to be seen as reliable by others. Explain what's going on to those around you. Tell them you really want to do something but you can't right now for health reasons.
- › **Be on the lookout for [depression](#).** It is normal to feel lonely sometimes. The worse CF gets, the more it tends to remove you from places where you can meet friends. Loneliness can lead to sadness, not keeping up with treatments, and losing the will to live. Similarly, if you ever find yourself staying away from friends for long periods of time on purpose, this may be a sign of depression. A life totally away from others is not good for your mental health — and can actually worsen your physical health. Seek out someone to talk to whenever you are sad or lonely, or if you think your thoughts and behaviors aren't normal. Your CF health care team will be able to help you find someone to talk with.

### MAKE TIME FOR YOUR SOCIAL LIFE

Don't kid yourself: Having quality interactions with friends takes time and energy, like anything else in life. When you have CF, sometimes those two things are hard to find.

It might help to explore ways to mix your health care needs with your desire for a full social life. For example, have friends over who can cook a high-calorie meal with you or go out to exercise together. If you are under the weather, perhaps a short phone call, email, or visit can give you comfort and boost your spirits.

Always make sure you prioritize your health and set limits on doing stuff with friends. Learning to say no can be life-saving!

You can also strategize by turning your cell phone off when you do your treatments or take a nap. Ask your friends if you can hang out later in the morning after you finished your treatments, or if you can all go out earlier in the evening so you have energy to do your treatment later. Explain your needs clearly and your friends will understand.

### WHAT TO SAY

Here are a few key phrases that may come in handy in your social life, whether breaking the ice or mending fences. Learn them, practice them, use them!

## Having a Fabulous Social Life

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- › “I want to share with you a part of my life that I usually just share with my closest friends...”
- › “I know this sucks, but I can’t go out with you tonight because I need to take care of my health. I’m really glad you invited me, though, and hope it works out next time.”
- › “It’s disappointing and hurtful to me when you say things that seem to imply you don’t understand how important my health is to me.”
- › “I know I used to go out partying with you guys. But I’m changing, and growing, and it’s just not something I feel like doing much anymore. I’m trying to focus more on my health.”
- › “I wonder if you are upset with me because I have to cancel our plans often. Or, am I just putting thoughts into your head?”

### IDENTIFY WHAT TO SAY AND RESOURCES

To tell or not to tell — that is the question. One helpful resource is [The 411 on Disability Disclosure: A Workbook for Youth with Disabilities](#). The workbook is designed to help people make informed decisions about whether or not to disclose their disability and understand how the decision may impact their education, employment, and social lives. The workbook treats disclosure as a very personal decision, and helps people think about and practice disclosing their disability.

To learn more, read [“To Disclose or Not Disclose: Lawyers with Disabilities Speak Out.”](#)

### SPEAKING FROM EXPERIENCE

“I think this is one of the harder aspects of CF — balancing mental, emotional, and physical health. It’s really important to not only fight for days of life, but for quality of life. It can be hard to keep up with my healthy peers. I try to be honest and open with them so we can find unique and rewarding ways to enjoy each other’s company and support one another, while still respecting the time and energy restraints of my CF. I have found being open about my CF allows my friends to help me be as strong as I can be. This has improved and strengthened our friendships overall.”

– Kristina, Age 35

## Having a Fabulous Social Life

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“Socializing can sometimes be tough when you have CF. The main thing is to always have some enzymes at the ready. I usually use old, smaller, empty pill bottles that can easily fit in my pocket. This is one of the major challenges, because it needs constant refilling. If I'm going out at night and will be back somewhat late, I'll try to get neb and vest treatments in before I leave. I usually can manage to sneak enzymes in my mouth either quickly or away from people. It can be frustrating trying to blend in sometimes when CF is on the mind, but most of the time I can just enjoy myself like anyone else. Sometimes it's hard turning things down in the week or having to go home earlier than others do because treatments and sleep are higher on my priority list. I just have to be more mindful than most of not sacrificing my health.”

– Eric, Age 34

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# Traveling With CF

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Just because you have cystic fibrosis doesn't mean you have to limit your traveling. You just need to prepare ahead of time.

This section includes ideas, hints, and important reminders to help you travel. As a general rule, it's best to discuss the details of your destination, trip, health needs, and precautions with your CF health care team. You should also travel with at least one other person. And it's a good idea to have a home base of sorts wherever you go so you can store all your supplies.

## IN THIS SECTION:

- ▶ **Learn the basics of traveling with CF** and how to prepare for your trip.
- ▶ **Work with your CF care team** to plan for a safe and happy travel experience.
- ▶ **Tips on traveling with meds and managing diabetes.**
- ▶ **Learn helpful tips** for cleaning equipment, compressor use, and oxygen use.
- ▶ **Maintain your health and infection control** while away from home.
- ▶ **Speaking from experience** — hear from other adults with CF.



## Traveling With CF

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### LEARN THE BASICS OF TRAVELING WITH CF

What's the best way to have a good trip? Be prepared. Start with this checklist of things to do 3 to 6 months before you travel.

- › Check in with your CF center before booking your trip to make sure you get a travel note and to assess your medical status. If you are not feeling well, you may need to be treated prior to your trip. You should also discuss a plan for treatments if you become sick and any impact that time zone differences may have on timed medications.
- › Check on immunizations required for international travel.
- › Make lists of medication and supplies you will need on your trip ahead of time. (Check and recheck this list!)
- › Make sure you request a refrigerator at the place you will be staying for medications that need to be kept cold.
- › Count all medications well ahead of your trip and get refills (order a month or so in advance if you use a mail-order pharmacy). You may need to talk with your insurance provider for refills needed prior to your trip; some will provide a "vacation allowance."

### WHAT TO PACK

- › Prepare a list of emergency contacts in case you become sick or have problems with luggage.
- › Bring a list of doctors who specialize in CF in the areas where you'll be traveling so you can get appropriate care if needed. Check [CFF.org](http://CFF.org) (national travel) or [CFWW.org](http://CFWW.org) (international travel) for locations of CF centers.
- › Bring your insurance information in case you need to visit a CF center.
- › Pack alcohol-based hand sanitizer with you for good hand hygiene.
- › Bring a prepaid phone card to contact your own CF center for advice in an emergency. These cards are also good to have if your cell phone does not work for some reason. You may also want to explore the following options:
  - Pay-as-you-go phones
  - International roaming plans from your phone company that can be turned on and off to suit travel dates
  - Unlocked phones, in which you can insert an inexpensive SIM card bought in the destination country

## Traveling With CF

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- › Bring instructions for your medical equipment in case of a malfunction (e.g., problems with your insulin pump).
- › Bring a calendar of times and details on taking your medications to help those who will be with you if needed.

### WORK WITH YOUR CF TEAM

- › Ask them to help you [find CF centers](#) and doctors near your destination(s). Discuss your trip plans with your CF care team and make sure you have the right medications with you — not just for daily use, but also for a possible increase in cough and congestion. Also, bring extra medication in case your trip is extended because of flight delays or other events.
- › Ask your doctor for extra written prescriptions in case your supplies run out. (Local pharmacies may have to order some CF-specific medications and this can take one business day or longer.) If a local pharmacy cannot fill the quantity of your prescription due to their limited supply, remember that you can always request less than the amount written on the prescription.

**Important Note:** When traveling in foreign countries, U.S. prescriptions may not be valid and U.S. medications may not be available. Plan ahead and bring enough supplies to last.

- › If you need oxygen, talk with your local oxygen company for resources to take on your trip. Make sure your mode of transport allows oxygen on board. Work with your CF health care team to identify the right concentrator and portable compressor to bring. Check with the [Transportation Security Administration \(TSA\)](#) for information about security screening of equipment.

### TIPS ON TRAVELING WITH MEDS AND MANAGING DIABETES

People with CF can't exactly pack light. That goes double if you also have CFRD. Here are some helpful insights on taking medications with you.

- › When traveling by plane, keep your medications with you in a travel bag that's easy to access for you and the [Transportation Security Administration \(TSA\)](#). If your medications do not fit in a plastic quart-size bag, they must be declared to the transportation security officer. Be sure to retrieve all of your belongings after TSA screening.
- › Ask for a travel note from your CF center that states your diagnosis, medications (and the need for refrigeration), and medical supplies you need to carry with you (like your compressor, nebulizer cups, airway clearance device, and/or diabetic supplies, liquids for low blood sugar, and ice packs). Check with the [TSA](#) for information related to security screening of equipment.

## Traveling With CF

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- › Keep medications in their original pill containers and/or carry a copy of your original prescriptions. Bring the prescription label for liquid medications (nebulized solutions, insulin, etc.) and for syringes or other medical supplies.
- › Carry your enzymes with you on the plane, where the climate is controlled. If they get hot in your suitcase they can be damaged and not work.
- › Pack medications that need to be refrigerated in travel coolers or cool packs. Use an insulated picnic or lunch bag and small “blue ice” packs or medical ice packs. You can find cooler bags online. It’s also important to bring a thermometer to maintain proper temperature. Check medication package inserts for the allowed temperature ranges.
- › Request a refrigerator at your hotel ahead of time (there is not always an extra charge if you say it’s for your medications). In hotels that do not offer refrigerators or mini-bars in the rooms, ask to use the hotel/restaurant refrigerator to keep your medications cool and to refreeze ice packs.

Be sure to label your medications with your name, hotel room number, cell phone number, and the words **“DO NOT FREEZE!”**

### TRAVELING WITH DIABETES (CFRD)

- › Keep insulin and supplies with you. And carry your prescriptions for all your diabetes supplies.
- › Make sure you bring snacks on the trip and have access to food or snacks at all times.
- › If you take once daily, long-acting insulin, contact your CF center or endocrinologist to make any adjustments for visiting or traveling through different time zones.

You can find more hints for traveling with diabetes at the [American Diabetes Association website](#) and the [Transportation Security Administration website](#).

## Traveling With CF

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### LEARN HELPFUL TIPS

#### KEEPING YOUR NEB CUPS CLEAN

- › Bring dishwashing detergent to clean your nebulizer cups.
- › Make sure your lodging has a kitchenette or microwave, or bring a portable device (like a steam sterilizer) to disinfect nebulizers. Check the condition of the water supply in advance and plan to use bottled water or other sources if needed.

#### CLEARING YOUR AIRWAYS

- › Sometimes, a form of airway clearance used at home is not the most ideal for travel. For example, a vest unit may be too heavy to travel with, or the person who performs the treatments may not be available. Talk with your CF center respiratory or physical therapist about airway clearance methods or smaller, more portable devices. Learn about different airway clearance methods at [CFF.org/treatments/Therapies/Respiratory/AirwayClearance/](https://www.cff.org/treatments/Therapies/Respiratory/AirwayClearance/).

#### USING COMPRESSORS

- › Check with your CF center and manufacturer to make sure you can use your compressor in other countries. Some compressors work only with 110-volt/60-Hertz (voltage/current) electricity, which is not available in all countries. Even with an adapter and converter, the compressors may be damaged and the warranty may be void if they are used internationally.
- › If you're traveling by car, get an adapter so you can do some of your nebulizer treatments while getting to your destination.

#### TRAVELING WITH OXYGEN

People with CF may wish to travel while on oxygen. This requires planning with your CF health care team, the transportation company, your place of destination, your insurance provider, and your oxygen company.

##### **If you will be traveling with oxygen:**

- › Review the guidelines for traveling with oxygen put out by the [Transportation Security Administration](#) and the [American Lung Association](#).
- › When making reservations, ask the transportation company about oxygen and let them know you will need oxygen while traveling. You will need to provide a letter from your CF center or complete a form from the transportation company.

## Traveling With CF

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Talk with your health insurance company about traveling with oxygen ahead of time. You may need different equipment on an airplane that your insurance may or may not cover.

- › You will need to contact your local oxygen supply company for help with using oxygen when traveling. Your oxygen company can give you contacts for other companies to make sure you are prepared in case you need additional supplies or have problems with oxygen during your trip.

### MAINTAIN YOUR HEALTH AND INFECTION CONTROL

There are many steps you can take to keep yourself healthy and reduce infections while traveling. Here are some of the main ones:

- › Clean your hands with alcohol-based hand gel or soap and water. Clean them often, such as after pumping gas.
- › Use a tissue when coughing or sneezing. Then throw it away and clean your hands.
- › Keep at least 6 feet away from others who appear ill or are coughing. If you have CF, keep at least 6 feet away from others with CF.
- › If the person next to you on a plane is sick, ask to move to another seat to minimize your exposure. People who do not have CF can better withstand getting a cold without it becoming a serious respiratory infection.
- › Bring oral antibiotics to have on hand — just in case.

### WHAT TO DO DURING AN EMERGENCY OUTBREAK

If a local, national, or international health issue occurs while you are traveling (like H1N1, also known as the swine flu), make sure you can check the [Centers for Disease Control and Prevention](#) website for more information. Also, keep handy the contact information for your CF center or a local CF doctor in case you need more information about CF and the health emergency.

### SPEAKING FROM EXPERIENCE

#### SUZANNE GOES ABROAD

"I've traveled overseas all my life. My family traveled a lot when I was a kid, but we always camped so my mom could make sure my CF health care and diet needs were met.

"Now, as an adult with CF and CF-related diabetes, I travel often for business and pleasure. I've been across the U.S. often, since I have family on both coasts. I've also been to Europe several times and to Australia — and I hope to go back! I'm already planning my next trip abroad to celebrate my 50th birthday!

"The key to traveling for me is to make sure I have enough medications to last the trip. I also find it very helpful to travel with a companion. Not only is it more fun, but a companion can help you keep up with your health care needs, carry your bags or get you a soda if you've worn yourself out.

"I also bring a folder or envelope with me to hold all of my travel papers. This includes information on flights and hotels, as well as emergency contacts, CF centers, new prescriptions, current prescription labels from refrigerated medications, and the medical travel letter from my CF doctor allowing me to carry my medical supplies. I keep this information at the ready for TSA if needed.

"Staying as healthy as possible, including when I travel, allows me to live each day to the fullest. Although my abilities change as I age, I still want to do my best and not let CF stop me. With proper planning and good company, traveling can open new horizons and give you great memories for years to come! Bon Voyage!"

– Suzanne, age 49

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# Parenting as an Adult With CF

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Adults with cystic fibrosis are able to meet many of life's milestones. One of the choices is whether or not to become a parent. Becoming a parent can be a challenging and rewarding time in your life. It will be a balancing act, as it is with everyone. You have the added responsibility of caring for your CF, so you can be there through many years of parenting.

## IN THIS SECTION:

- ▶ **Learn about parenting with CF** to help you plan and think ahead.
- ▶ **Work with your CF team** to help you stay adherent to your therapies and be able to enjoy time with your child.
- ▶ **Manage your time** so you will be able to balance your self-care and your parenting role.
- ▶ **Get help when you need it** so you will stay well and be effective in your role as a parent.
- ▶ **Discuss CF with your child** at an age-appropriate level.

## Parenting as an Adult With CF

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### LEARN ABOUT PARENTING WITH CF

For those with CF, becoming a parent may occur naturally, via MESA (microsurgical epididymal sperm aspiration), or require other reproductive assistance. You may choose to adopt or use a surrogate.

**If you are a man**, you should already be aware that 95 to 97 percent of men with CF have congenital bilateral absence of their vas deferens (CBAVD) and cannot father a child naturally. Only 2 to 3 percent of men with CF are able father a child naturally. If you would like to have children who are genetically yours, you and your partner will need to go to a fertility specialist. You would need to undergo a sperm aspiration by a procedure called MESA or TESA (testicular sperm aspiration). Sperm is retrieved from the male; the mother of your child would then need to go through in-vitro fertilization.

CF does not necessarily affect a **woman's reproductive health**.

Perhaps you have decided to adopt. This is also a wonderful option and your CF care team may need to write a letter of support for you. This will depend on your adoption agency.

You will have other issues to consider and plan as you think of becoming a parent. It is important for you to think through some of the following questions and have a plan prior to becoming a parent.

- › How will you fit in your daily treatments and also care for a child?
- › What will you do on the days you don't feel well?
- › Who will help you?
- › Who will get up during the night with the baby or toddler?
- › What will you do when your child is sick?
- › What is your plan should you need IV antibiotics and/or hospitalization?

You have had CF all your life, so you should be used to planning ahead and being flexible. The issue of parenting is really no different. With some forethought, you should be able to deal with family planning as well as child rearing.



**Making the decision to have a child is momentous.  
It is to decide forever to have your heart go walking  
around outside your body.**

**– Elizabeth Stone**





## Parenting as an Adult With CF

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### WORK WITH YOUR CF TEAM

Your CF care team is an excellent source for help when you need support, advice, and tips. They can be a wonderful resource for you before, during, and after your child is born. In order to enjoy your family life, it will be important for you to sustain your daily therapies. It is very common for parents to put their children before themselves. It is very easy to get busy with your child and forget to do treatments. This is a temptation you need to resist. It is of utmost importance to make your health a priority. Your CF care team can help you assess what support you will need to maintain your health and enjoy your family.

Perhaps finances become an issue with your growing family. Talk with your team social worker. He or she is there to help you take advantage of the various programs that might be available to you.

You, your child, significant other, or other family members may need some help coping with the added stress of parenting and all it entails. This may change during your child's various developmental stages and your changing health status. Perhaps you think your spouse/partner or other family members are feeling an emotional impact of having to "pick up the slack" of parenting. Your CF care team can help you. Don't be afraid to admit that parenting is causing you and others stress. As your child grows, he or she will need a realistic image of health and wellness. You will have various levels of wellness. Sometimes you will need to go to the CF care center for well visits, and other times you may have an exacerbation. You may need to be hospitalized for treatment. Your CF care team can help prepare you and your child for the separation that will be occurring while you are hospitalized. Your children will need reassurance that you may only be gone for a short time, and someone will be there to care for them, maybe their other parent or a grandparent or other family member. Your child's response may be unpredictable. You need to be realistic about what is going on and give them enough age-appropriate information so they can remain hopeful and not expect the worst.

**A healthy attitude is contagious but don't wait to catch it from others. Be a carrier.**

**– Tom Stoppard**

## Parenting as an Adult With CF

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### MANAGE YOUR TIME

Don't isolate yourself; CF is part of your life and part of your family life as well. Depending on your child's age, they can help you with this task. This is a way they can feel special and useful and will help alleviate fears. It could be as simple as getting your water for you to take your oral medication. Just make sure they understand they are not to take the medication, and keep it out of their reach. Perhaps exercise would be good for you; when your child is young, look into a bilateral. Put your young child into the stroller and take a walk. These are all ways you can get your child involved in your care when they are young, while keeping it a natural part of your everyday life.

Sometimes you might have to revise the expectations of your family life. You will have to deal with good and bad days. It is important to have a plan for those bad days, alternate plans will need to be discussed and agreed upon by others involved in your child's care before they become issues.

You may want or think you can do it all. But you can't! It is important to know your limitations. Balance your abilities and limitations. When you are not feeling well, you may have to alter what you do. Perhaps you will not be able to go to the park, but you could spend time with your child on the couch, read a book to them, or do a puzzle with them. Remember that CF will not get in the way of love, hugs, teaching, and spending time with your child. CF should not consume your family life. However, it should be a normal part of the household. CF is part of your life — it is not your life.

It is important to be honest with yourself and know your limitations. What treatments do you struggle with? What sabotages your treatments? For example, do you get too busy and forget or run out of time to do your treatments? You may need to develop a schedule for your treatments that helps you complete your treatments so you can then focus your attention and time on your parenting responsibilities. Push yourself during your "better" times. Your CF care team can help you develop a schedule to manage your time so that you can complete your treatments and enjoy family time.

**“Boundary setting is really a huge part of time management.”**  
– *Jim Loehr*

## Parenting as an Adult With CF

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### GET HELP WHEN YOU NEED IT

It is important to realize that you do not (and should not) do everything yourself. Don't be afraid to ask for help from your child's other parent, your family, your friends, and your CF care team. For example, if you are hospitalized, or too sick to attend a school function, consider asking a grandparent, favorite aunt, or friend to take your place. Maybe they can record the function for you so you can sit and watch with your child. This can still be a very special time for you to share in your child's life.

It is important not to feel that it is a sign of weakness to ask for help. This will show your child that it is OK to ask for help from others in your community. You can ask your child to help out at times. However, it is very important to make sure you let your child be a kid, and not your caretaker. Your child's teacher, guidance counselor, and school nurse will be important allies to assess how your child is dealing with your illness. It is important to have good communication with the staff at school. Your positive attitude can also help your child realize that having a disability or illness does not affect a person's worth.

It is important for you to always make CF your first priority. Just as they tell you before a plane takes off, parents should put on their masks before tending to their children. It is the same with CF and parenting. To be the best parent you can be, it is important for you to remain healthy, so take care of yourself, before tending to your child. Caring for yourself first is not being selfish. Caring for yourself is a gift you are giving to your family so you can be as healthy as possible and able to enjoy family time, both in quality and quantity.

Some parents have found journaling to be very therapeutic. You can write down things your children said and did, special times you spent together and also how these events made you feel about them. Your child will treasure these books forever, and you will be able to look back over these memories and realize how much a part of your child's life you are.

“**Each day of our lives we make deposits in the memory banks of our children.**”  
– *Charles R. Swindoll*

## Parenting as an Adult With CF

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### DISCUSS CF WITH YOUR CHILD

It is important to discuss CF with your child. Through the different stages of their lives they will have different levels of understanding, and the information you give them will have to change as they grow and develop. Your child will have questions, and it is important to remember to only answer the question they are asking. Don't shield your children from what is happening to you. If it is appropriate for their age, include them in discussions. This will help them cope with what is happening and create a family bond; be honest about your health. If you don't know what to say, ask your CF care team to help you. Perhaps you can practice your discussion with someone from the team.

**Preschool:** Infants and toddlers like routine. They can be very sensitive to the emotional environment in your home. If you are hospitalized, it will affect their routine, and perhaps their sense of security. They will need plenty of reassurance by touching, hugging, loving, and repeated verbal assurance.

**TIP:** If you are hospitalized, it will be important to have your spouse, a grandparent, or aunt help care for them so their routine is not disrupted too much. It is important to think ahead and have a plan ready should the need arise.

Preschoolers may feel they caused your illness. It is important to make sure they do not feel guilty for your health. They also need to realize they cannot catch this disease. It may help them explore their feelings by getting them a doctor's kit.

**Ages 6-12:** Children in this age group will start to ask more questions and become more curious. They may start asking, "Why do you cough so much?" They will also start to realize that their friends' parents don't cough as much as you do. This may cause a little bit of embarrassment when they are with their friends. This age group will become aware of their emotions, but not comfortable with them. It is important to make sure they talk about their feelings. They also may start to be aware of death and may ask questions about dying and heaven. They are at a stage where they can help you with simple tasks, like getting your water for your medication. This will help them feel special and useful. These tasks may also help reduce their fears of your illness. Provide them information in simple terms that they can understand. At this age, they should know how to dial 911 in case of an emergency.

**Adolescents:** This is a time of physical and emotional change. They will need a good support system. It is important for you to make sure they have permission to go out and enjoy themselves, and not always have to stay home and help you. They will expect you to be honest with them. It is common for teens to think abstractly. They may look into things for meaning.

## Parenting as an Adult With CF

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**TIP:** If you have to stay in the hospital emergently, your child may feel it important to see you right away. You may want to make a deal with your children that the non-CF parent will come get them, wherever you are and bring them to the hospital to see you. Your teen may have the need to talk with someone throughout your health crisis. This might be you, the other parent, or other family members, If that is not possible, find someone your teen can open up and talk to.

Throughout your child's life, it will be important for you to assess if your child is having difficulty dealing with issues:

- › Are they starting to withdraw or act out?
- › Is there any change in eating habits?
- › Do they complain of frequent stomach or headaches?
- › Have they regressed to former behaviors?
- › Do they isolate themselves?
- › Are they experiencing new or irrational fears or increased anger?
- › Has there been a change in school performance?

You might have to deal with preparing your child for your death. This will be very difficult, but how you handle the issue will help them have the tools and confidence to cope with this challenging part of life.

### TIPS

- › Create memorable times with your child — make the most of the time you have together.
- › Consider making a video or special photo album.
- › Consider writing letters to them that they will be able to open at special events during their life, in case you are not there with them.
- › Make a memory box filled with special items.

In the book, *Now That I Have CF*, Karen Mackle gives her personal perspective of having a parent with CF. She says that it never made a difference to her. Her mother doing treatments, or IVs, was just something her mother did. She suggests that children be involved in their parent's care so CF will be viewed as part of the daily routine of the family.

### ADDITIONAL RESOURCES

[www.cffinfo.org](http://www.cffinfo.org)

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# Finding Community Support

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**“Life is 10 percent what you make it and 90 percent how you take it.”** Composer Irving Berlin’s famous quote could easily apply to the attitude required to manage the unique practical, financial, and emotional needs that come with living with cystic fibrosis.

Tracking down the information to help you can be exhausting, even when you are feeling your best. You may have to fill out long applications, copy important documents, spend hours on the phone or online, speak to many people, and, most important, wait patiently to find exactly what you need.

But don’t despair. You belong to an extremely motivated, supportive, and organized community. Thanks to dedicated family members, volunteers, and friends, people with CF have access to thousands of resources, including college scholarships and free tuition, help with exercise costs, and public assistance such as food stamps and disability income.

You and your close supporters can begin your resource search today. Consider the helpful tips in this section as you navigate the available services in your community.

## Finding Community Support

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### IN THIS SECTION:

- ▶ **Learn how to find resources** in your community.
- ▶ **Work with your CF team** to tap into existing resources.
- ▶ **Become the best investigator you can** for finding support.
- ▶ **Follow time-management tips** to find resources efficiently.
- ▶ **Get support before there's a crisis.**
- ▶ **Learn to accept help graciously.**
- ▶ **Check out these key CF resources.**
- ▶ **Speaking from experience** — hear from other adults with CF.

### LEARN HOW TO FIND RESOURCES

Sometimes just figuring out where to start your search can seem overwhelming. But it need not be. Start with a list of the agencies and institutions you are already a part of or know about — from the department of motor vehicles and utility companies in your area to your hospital and workplace, your grocery store and gym.

Now think about the services these places offer that could help ease your CF care burdens. For example, utility companies often reduce rates for medical needs; gyms offer discounts to members with disabilities; workplaces provide free employee counseling.

Next, tap into any available public service resources. Whether it's Social Security Disability, Supplemental Security Income, Medicare, Medicaid, or other forms of financial and medical support, these services are there to help. Call your local **Social Security Administration office**. And don't forget to explore your state, county, and city **health department websites** for similar support.

## Finding Community Support

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As you work, keep a list of contacts — people in the CF community who know its many resources — close at hand. You may already know role models at your hospital or have favorite online experts, and you'll no doubt find others in your research. In the meantime, here are a few to add to your list:

- › Beth Sufian heads the [CF Legal Information Hotline](#).
- › Tiffany Christensen speaks on patient empowerment with [Sick Girl Speaks Inc.](#)
- › Jerry Cahill coordinates programs with the [Boomer Esiason Foundation](#).
- › Ronnie Sharpe facilitates [Cystic Life](#), a social network for the CF community.

Now that you're off to such a strong start, you're ready to branch out:

- › **Speak up.** One of the easiest, and most effective, ways to find existing resources is by word of mouth. Simply mention what you're looking for in casual conversations. You never know what your friends, colleagues, and other contacts can refer you to.
- › **Get online.** CF websites like this one, as well as hundreds of others, provide access to the latest information on everything from treatment options to clinical trials. If you do not have Internet access, call organizations like the CF Foundation (**800-FIGHT-CF**) for printed information.
- › **Connect with community.** Remember, you're not alone. Reach out to CF peers virtually for moral and educational support. You can also learn about the latest medications and devices by meeting with CF pharmaceutical and medical equipment sales representatives. The CF Foundation has a **wealth of resources** to help you.
- › **Think outside the CF box.** Expand your search to other community health organizations, such as those for **diabetes, transplant,** and lung health (the [American Lung Association](#) is a great one). They feature ideas and resources you may not find in the CF community. Check out non profit organizations that specialize in chronic illness or disability needs, too.
- › **Become a groupie.** You may be surprised to find out what general community groups — fraternal organizations, such as Rotary, Lions, and Elks, or churches can offer to help you. So ask around.
- › **Advocate. You can make a difference for people with CF by engaging key decisions makers. Get involved and contact your local chapter.**



## Finding Community Support

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### WORK WITH YOUR CF TEAM

Every member of your CF care team — from social workers to nurse coordinators to respiratory therapists — should know about available resources in your community. Turn to them for phone numbers, websites, and expert references that have worked best for other patients with CF.

- › **Share online health information with your CF care team.** Many websites, like PubMed from the National Institutes of Health (NIH), offer articles on the latest medical research, complete with ideas for new treatments. Talk to your CF team members about what you learn and get their opinion about these options.
- › **Follow the paper trail.** Keep copies of any forms you need CF providers to sign and/or fax in case they get lost. Always give a deadline and be polite — but persistent — if you need to follow up. Finally, don't hesitate to get a second opinion if a CF care team member refuses to sign a form.
- › **Recruit others to your CF health care team.** Ask about other hospital staff who might help you. Financial counselors or patient advocates, for example, can offer a unique perspective. Check out Patient Relations or Guest Services at your hospital to see what they can offer.
- › **Make sure you have a two-way relationship.** Your pediatric CF health care team probably took care of more things for you than your adult CF health care team does. They will not do everything for you, but rather point you in the right direction so you can solve problems on your own. Some adults with CF get frustrated, saying things like, "They didn't call me back. They must not care." The truth is, the only one who is in charge is YOU. Try not to judge your providers, but realize that they may be teaching you to be self-reliant. Watch the [Partnering With Your Center for CF Care](#) webcast series to learn more.

### BECOME THE BEST INVESTIGATOR YOU CAN

When it comes to tracking down the right help, the best sleuths have learned how to ask for services. So before you pick up the phone or visit that agency, make sure you can answer this question: What are you looking for — information, practical support, emotional support, financial assistance?

Now be specific about what you need, clearly describing your problem. Use assertive phrases like, "I think, I need, I would like, I feel." But make sure to be polite and patient. Making notes or preparing questions in advance will help you cover every point. Or write a script to use when making calls. You can even enlist a support person to listen in and jot down notes for you.

## Finding Community Support

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Here are a handful of other tips for a successful investigation:

- › **Find or ask for a case manager.** Establish one contact within your hospital, insurance company, or public agency. When this person becomes familiar with you and your case, you are likely to have an ally who will add to your voice.
- › **Outsmart the system.** Most agencies have an appeal process — so plan to use it. Keep meticulous records, find alternatives, talk to supervisors, and get confirmation. It may seem labor-intensive, but the appeal shows you're serious, committed, and justified to have your case re-examined.
- › **Learn how to follow up.** Keep a detailed log of your phone calls, including the name of the person you talked to, the date and time of the call, the outcome, and any follow-up required. Above all, be persistent.
- › **Stay organized.** Maintain a file of scanned or photocopied applications, income tax statements, and other important paperwork in a designated place. Also keep your emails in tidy electronic folders and archive emails when they become irrelevant.
- › **Don't give up.** Norman Vincent Peale said, "It's always too early to quit." When you hit a roadblock, try another person within the organization. If you encounter a closed door, try another organization. It takes initiative to be persistent.
- › **Keep your cool.** It's normal to get frustrated when you are stuck on hold, get disconnected, or receive unsatisfactory service. But anger isn't a good starting point when expressing your needs. Take a deep breath and re-check your perspective. Having CF gives us access to resources that are a privilege — not an entitlement.
- › **Know your rights:** If you think you've been wronged, discuss your concerns with a lawyer. CF Foundation *Compass* can connect you with resources for legal information by calling **844-COMPASS (266-7277)**. You may also want to file a grievance with your state if appropriate.

## FOLLOW TIME-MANAGEMENT TIPS

- › **Mark your calendar.** Use follow-up reminders, application deadlines, and other schedule prompts to keep everything — including you — in the right place at the right time.
- › **Make research routine.** A set schedule will help you make the most of your research time. Browse the Internet and write emails during your 9 a.m. treatment. Call agencies after lunch. Put your phone on speaker if you are put on hold so you can cross other tasks off your list.

## Finding Community Support

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- › **Always ask for deadlines.** Chances are if you leave something open-ended, it will drag out. So push for closure. When do I need to call back? How long will it take to deal with this matter? Ask for an anticipated callback date, and follow up if you don't get it.
- › **Fill out applications immediately — or ASAP.** Be sure to ask for the application deadline. And if you foresee a problem, ask what happens if you can't get it in on time. Then do your part; get any applications or forms in as soon as possible for the best service.
- › **Confirm appointments.** Ever show up for an appointment only to be told you're not on the schedule? No one has time for that. Be sure to check in and confirm one week in advance by email, or call the office the day before.

### GET SUPPORT BEFORE THERE'S A CRISIS

The time to identify available help in your community is before you need it — not when you are in the middle of a health care or other life crisis. So resolve to prepare now for whatever may come your way. Search for community services long in advance of when you may need them. Start your reference file of helpful services today!

- › **Prepare for change.** Plan for major life transitions such as high school graduation, college graduation, and marriage. These are times when you can lose insurance or encounter financial struggles that require extra support.
- › **Mind your medications.** Never let your CF get worse because you don't have medications! You can get help — just be proactive, persistent, and honest with your CF health care team. If that doesn't work, consider calling another CF clinic for a second opinion. Check out specialty pharmacies and learn more.
- › **Do your homework.** You can't expect your CF care team to know about every available resource — there are too many to know them all. So share the responsibility for finding the right resources for you. Then share them with your team so others with CF can benefit.
- › **Volunteer to learn.** What's the best way to learn about community resources? Become a volunteer in the CF community. Join a board or committee at your local CF Foundation chapter or CF non-profit and you'll soon be the expert.
- › **Make the connection.** If you don't have Internet access, call local community centers or 411 and ask for agencies related to your need. You can also go to your local library or hospital health library.

## Finding Community Support

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### LEARN TO ACCEPT HELP GRACIOUSLY

Asking for — and accepting — support can be hard. Some people with CF feel bad about using community resources. They'd rather not "impose on the system." Some cling to independence, too proud to look for a "handout." Others fear the phone calls, forms, and frustrations that can come with the search for resources.

The next time you hesitate about seeking or accepting assistance, consider these points:

- › **Return the favor.** It takes a village to live well, and you are an active member of the village. Whatever help you receive, you can use that to improve your health so you can contribute to society in other ways.
- › **Accept the mission.** Humbly recognize that non-profits want you to tap into their services — their mission is to serve.
- › **Pat yourself on the back.** When you receive help, don't just be grateful: be proud. You've successfully navigated the resource maze and found your way to the prize at the end!

### CHECK OUT THESE KEY CF RESOURCES

Looking for more? Here are some more great resources to explore in your ongoing search for the right combination of CF services in your community.

- › [Vocational Rehabilitation Services](#) help people who have been out of the workforce due to disability with either retraining or assistance in getting adaptive equipment that lets them re-enter the active workforce. Vocational rehabilitation services are state-based. Search online by entering your state name after the term "Vocational Rehabilitation." Your social worker may also know about the availability of vocational rehabilitation services in your state.
- › [CF Roundtable](#) is a newsletter for adults with CF published by the **United States Association of Cystic Fibrosis Adults**.
- › [Boomer Esiason Foundation](#) offers athletic scholarships.
- › [Elizabeth Nash Foundation](#) offers scholarships.
- › [The Living Breath Foundation](#) provides community support with some grant funding.
- › [AbbVie](#) offers college scholarships (ask your social worker for details).
- › [Blooming Rose Foundation](#) provides community support and education.

## Finding Community Support

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All of the following sites are other places to find information and support:

- › [CF Foundation](#)
- › [Genetic Alliance](#)
- › [CF Living](#)
- › [Cysticfibrosis.com](#)

### BOOKS

*Cystic Fibrosis: A Guide for Patient and Family* by David M. Orenstein, M.D. (Lippincott Williams & Wilkins; 4th edition, 2011).

*Cystic Fibrosis in Adults* by James Yankaskas, M.D., and Michael Knowles, M.D. (Lippincott Williams & Wilkins; January 1999).

*Sick Girl Speaks: Lessons and Ponderings Along the Road to Acceptance* by Tiffany Christensen (iUniverse, Inc., 2007).

### SPEAKING FROM EXPERIENCE

"I love having CF community support, but it's limited for me because I cultured MRSA. I have attended one CF education event, which was an amazing experience that far exceeded my expectations. Now, when I need to reach out to the CF community, I turn to [cysticfibrosis.com](#). I've also gotten involved in writing a *CF Roundtable* article and creating images working with *The Breathing Room*."

– Eric, Age 34

"I find both careful, conscientious CF education gatherings and official CF support groups to be very helpful. Facebook and other online CF groups are really great, too. I also write a blog or online journal for my friends and family and that allows me to draw a lot of strength from them."

– Kristina, Age 35

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# Managing Home Care

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There's no place like home. You get to sleep in your own bed, you're not as exposed to super-bugs, you're more active, and you can enjoy your favorite foods. That's why so many adults with cystic fibrosis opt for home care when they need antibiotics.

Just as when you go to the hospital if your CF flares up or gets worse, the goal of home care is to help you finish treatments, feel better, AND keep you in your best physical shape.

Sometimes, that means starting a course of treatment in the hospital, such as IV (intravenous) antibiotics, and finishing at home, or starting a treatment like home oxygen or tube feeding at home for an unknown period of time. No matter the situation, the responsibility of home care can be scary, especially when you start off feeling lousy or depressed about being sick and needing treatment.

But remember: Managing your own home care — and becoming an expert at treating yourself — is a very rewarding experience.

## IN THIS SECTION:

- ▶ **Learn ways to manage home care and your life.**
- ▶ **Make your home care work** as well as possible.
- ▶ **Work with your CF team and home care agency** to get extra support.

## Managing Home Care

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- ▶ **Manage your time** to prioritize rest and health care, and learn how to balance your medical schedule with everything else in your life.
- ▶ **Get help when you need it**, because it's just not safe or possible to do everything on your own.
- ▶ **Give yourself a little love**, because home care is not always easy.
- ▶ **Speaking from experience** — hear from other adults with CF.

### LEARN WAYS TO MANAGE HOME CARE AND YOUR LIFE

Most adults with CF will have to use home care in some form. This might include IV antibiotics, home physical therapy, tube feeding, IV nutrition, supplemental oxygen, or assisted ventilation. Managing home care well will likely reduce your complications and hospital stays.

If you choose to do your treatments or tune-ups at home rather than in the hospital, you should be ready for the time and work that they require. That said, home care can become as routine and manageable as your daily airway clearance therapies. With practice, you can find out your own best way to make home care work.

As an added bonus, home care technology is always getting better. Be sure to ask your health care providers about any new access devices or delivery devices (pumps, oxygen tanks, etc.) that could improve your quality of life and make home care even easier.

Here are some good tips and thoughts to keep in mind for managing your home care successfully:

- › Designate one “**clean corner**” of a table, shelf, or counter for getting your medication ready. This is where you can hook up your IVs, prepare your meds, draw up syringes, and do other medical work that requires good hygiene. You may want a clean corner in your fridge as well.
- › **Be hygienic** — use paper towels and anti-bacterial soap religiously.
- › **Organize** your home care supplies to make sure you have enough to get through your treatment without stopping. You may want to plan ahead by counting out your daily supplies and setting them aside for a week.

## Managing Home Care

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- › When bathing or washing, follow your nurses' guidelines on **covering your IV line**, implanted port, or button. Consider washing your hair in the sink every day and showering every other day. Get a showerhead with a hose to make showering easier when you can't get one part of your body wet. Use a plastic cover on your arm if you have a **PICC** (peripherally inserted central catheter).
- › Keep IVs and other home care products **safely away from young children** and pets in your home.
- › Don't leave IVs in the sunshine or the car, or anywhere the medication is subject to **extreme temperatures**.
- › **Get out and enjoy life.** Our mental health affects our physical health. It's important to deal with the emotions that come with the extra burden of care and disability, and what that means to you. Going out in the sunshine, taking a walk with friends or just going out for a meal while on home treatment boosts your quality of life.
- › **Be creative.** Figure out what works for you. If you have a gravity-drip IV bag, placing it on a hanger or on a tack on the wall is sometimes more convenient than pushing a pole around.
- › If you have a **hectic home life**, with roommates or family members who come in and out in an unruly way, or you are in the process of moving, talk to your CF health care team to see if you can receive care in the hospital rather than at home.

## MAKE YOUR HOME CARE WORK

When you need home care, your body is usually fighting an exacerbation (where your lungs are worse than usual). Therefore, **your CF care demands more time and attention for you to get better.** Balancing your home care needs with greater requirements for nutrition, fluid intake, airway clearance, and sleep is a challenge for everyone with CF.

- › Having an exacerbation that takes extra energy is like drawing money out of your nutritional bank account. Make sure you get good nutrition while doing IVs. Try preparing large amounts of food so you can have leftovers, or ask your medical team for some extra nutritional supplements. Home care is a great excuse to treat yourself — to get take-out or have food delivered, order groceries online, ask friends to bring you food, or buy frozen meals that are easy to make.



## Managing Home Care

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- › **Take the time during an infusion of IV medication to do airway clearance.** As long as you're stuck getting an IV, you might as well use that time to inhale your medications and do chest percussion. Try to multitask more. Maybe you can use your [Oscillating PEP](#) device while doing email or cooking or when you are a passenger in a car (never drive while doing treatments!). Airway clearance is more important than ever when you are on IVs. Just think of all the germs that are being killed — you want to get the dead stuff out!
- › **Even if you aren't feeling up to it, exercise is very important when you are having an exacerbation and need home IVs.** Make time to maintain your exercise routine, but give yourself permission to reduce intensity or duration when you are on home IVs. If you sweat a lot, ask your home care agency for extra supplies for frequent dressing changes.

Ask your CF care team if you need to adjust your activities while on home care. For example, playing football might be a bit risky if you have an implanted port accessed! You may also want to plan your exercise around your dressing changes. For example, you can schedule a swim between your doses when your implanted port is de-accessed. Even when you're using home oxygen, exercising with oxygen can help improve your baseline.

- › **Do home care for the right reason.** If you're doing home IV infusions so you can continue going to school or work, make sure you're getting a break somehow, with extra sleep or airway clearance. During IVs, your full-time job should be your health — and you may have to humbly admit that you can't do it all. Perhaps you can ask your co-workers to donate vacation days/paid time off or ask your professors to give you an extension on assignments.
- › If you must work when on home care, **is it possible to work from home?** Maybe you can work part-time or flexible hours during the course of your treatment.
- › **Sleep is still the best medicine**, and when you are having an exacerbation, you'll likely need more sleep. Try to go to bed earlier. If your IV is due at 11 p.m., set your alarm for 11 p.m. and go to bed at 9 p.m. Then when the alarm rings, wake up, connect your IV, set the alarm again, and go back to sleep until the infusion is finished. Then you can flush and return to sleep. Let yourself sleep in or take naps to make up for lost sleep.

## Managing Home Care

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### WORK WITH YOUR CF TEAM AND HOME CARE AGENCY

It's very important to have regular phone contact with your CF health care team during home care. You also need regular follow-up visits to your CF clinic. (Hopefully you can make late morning or afternoon appointments so you can sleep in!) You'll want to check your pulmonary function and weight to see if the treatments are working. Your CF health care team can help with problem-solving tips and ideas.

Here are some other ways to work with your CF health care team and a home care agency to get extra support:

- › **Ask your CF health care team about different types of IV access** (heparin locks, **peripherally inserted central line, implanted ports**, or central lines), oxygen systems (liquid or gas tanks or a concentrator), or tube feeding devices. Having choices that fit your lifestyle gives you some control over your quality of life.
- › **Tell your CF health care team if you feel at all uncomfortable doing home care.** Maybe you'd feel better visiting an outpatient infusion clinic for the first few doses. Your team may also be able to order a visit by a nurse to help you get started, be available for problem solving, and provide extra education.
- › **Be compliant with lab draws!** Getting your blood drawn for peaks and troughs (drug levels) of certain medications can make a difference in whether you save your hearing or kidneys. If you're too tired or busy for labs, ask your CF health care team if they can order a visiting nurse to draw labs at your home.
- › **You may have choices in home health care agencies.** Choose one that is close and offers the highest-quality service. Call your insurance provider to see what your choices are. If you prefer an agency that's familiar with CF treatments and protocols, your CF nurse probably has some recommendations. If you have concerns about the care provided by your home care company, contact your CF health care team.
- › **Ask your home care agency for the most portable and convenient IV pump possible.** You can infuse with disposable bottles, portable pumps, push syringes, or IV poles with pumps. Try to let them know clearly why you need one version over another to help you do your medicines as planned.
- › **Ask your respiratory therapist for tips on using supplemental oxygen.** Are there new and more comfortable cannulas or masks? How do you flush your sinuses when using oxygen? How do you monitor your flow rate, and when do you need a flexible refill schedule? This is all good information to have.

## Managing Home Care

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- › **Don't always take "no" for an answer.** If you find a pharmacy or home care agency that says, "No, that's not our policy," or "No, we don't carry that," always ask to speak to the supervisor. You can talk with your CF care team or look up information online and give that to your agency to challenge their rules and policies. Maybe you can give the agency an article about a new delivery device you find at an education event, on the Internet at [PubMed](#), or through your CF care team or a friend. Maybe they'll be convinced to order that device!
- › **Don't do home IVs just to save money.** If money is your concern, talk to your social worker to see if there are other options for hospital coverage.

PubMed provides free access to over 21 million medical and research articles, including information published about CF. It is a part of the National Institutes of Health (NIH).

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[Learn how to use PubMed by using their Quick Start guide.](#)

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## MANAGE YOUR TIME

As with everything else involving CF, time management plays a big role in successful home care. Try these tips to help you focus on feeling well and balancing your medical schedule with everyday life:

- › **Take your IV medication out of the fridge an hour before your infusion.** When you're going to sleep, place your IV into a cooler bag (with or without an ice pack, depending on how long until your infusion). It will warm up before it is time to infuse. This saves you a trip to the fridge.
- › **Set alarms.** Try using a conventional alarm clock, cell phone alarms, or kitchen timers to remind yourself when to start your IVs or flush your line.
- › **Try to stay on schedule with your medication.** If your infusions are every 6 hours, a 6 a.m.–12 p.m.– 6 p.m.–12 a.m. schedule might work best. Or, if it's every 8 hours, a 7 a.m.– 3 p.m.–11 p.m. routine might work. Be flexible and rearrange all of the priorities in your schedule. Anything that's not health-related might have to wait.
- › **Slow down.** Errors happen when we rush. Talk yourself through medication or IV preparation so each step is clearly followed. Try to schedule your routine so you won't feel rushed, and stick to that schedule.

## Managing Home Care

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- › If you can't be home for a delivery, **ask if your home care agency can deliver your supplies to your work or a neighbor's home.** Ask if you can place your oxygen tank outside your door and have it refilled when you're gone.

### GET HELP WHEN YOU NEED IT

- › **See if a friend or family member can help you with one infusion a day,** such as your evening infusion, so you can go to sleep. Doing IVs all alone takes a toll on your body and mind. IV antibiotics are more effective when you're rested. You might even ask a family member or friend to help you with one meal a day.
- › If you're always very tired or feeling completely overwhelmed by home care, **it may not be best for you to continue treatment at home.** This is especially true if you live alone and have no support.
- › Some people with CF can tell when their lungs need a tune-up. **Try planning a home IV tune-up when you have extra support,** for example, when family is visiting or over the holidays. If this isn't possible, maybe a friend or family member can stay with you during the time you are receiving treatment to help out.
- › **Avoid crises.** If you have signs of infection, an allergic reaction, or bleeding from the sites of IV lines or G-tubes (feeding tubes), call your CF health care team or home agency immediately, or call 911 in an emergency.
- › **Have a back-up plan.** Ask for extra oxygen tanks in case the power goes out or the concentrator breaks. Always call your home care agency if you're not sure what to do.

### GIVE YOURSELF A LITTLE LOVE

- › Home care can disrupt your sense of normalcy. It's normal to feel upset about being on home IVs or using oxygen. These are reminders that CF is progressing. You can also feel anxious and wonder if the treatment will work this time.
- › During these times, stay in the moment. Remember that some people with CF use oxygen, tube feeding, or positive pressure ventilation (BiPAP®) only during exacerbations. Also, many people with CF do regular IV therapy for decades. Home care is part of your long-term investment in health.



#### Home Care Mantras:

"This too shall pass."

"This is nothing compared to the alternative."

"I am the master of my fate:  
I am the captain of my soul."  
— "Invictus," William Henley

"A stitch in time saves nine."

"Start by doing what's necessary; then do what's possible; and suddenly you are doing the impossible."  
— St. Francis of Assisi

## Managing Home Care

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Here are a few more helpful reminders to ease your stress:

- › Physical devices (like oxygen by nasal cannula, an accessed implanted port, or a G-tube button) can take some adjustment. But just think: they are helping you live longer and better.
- › You're in serious fight mode during home care. Charles Dickens wrote, "It was the best of times; it was the worst of times." Your CF fight is tiring and unwanted, but these are the times when you may notice silver linings: you can explore your inner strength and spirituality, gain confidence in your abilities, and cultivate loving, compassionate relationships, so you don't feel like you're all alone.
- › Allow yourself to feel sad, angry, fearful, frustrated, bummed out, even sorry for yourself. Those are real feelings, and they are normal, especially now that you are required to do even more at home. It helps to talk with those around you about how you're feeling.

Doing home care is very hard work. Try to focus on one day at a time. Commend yourself for being such a strong, bright, organized person. Most healthy people can't handle what you are managing!

- › Practice gratitude when you are using home care. You are both caregiver and care-receiver: thank your higher power for the chance to get healthier at home, for the technology we have, and for your own abilities.

### SPEAKING FROM EXPERIENCE

"When I'm on IV meds lately, I'll work part time from home, since it's an added chore in my life that can take a lot of time. I used to go to work and would 'hook up' to the meds in a bathroom stall with lots of alcohol being used to make sure everything is clean. The doctors will usually prescribe one drug for just once a day to cut down on my morning obligations.

Sometimes, for the late day doses, I'll set my phone alarm to go off because I would often fall asleep. It can get complicated when on three or more meds, and unfortunately I'm usually doing this all solo."

– Eric, Age 34

"I think this is the hardest part of CF care. I manage the intense IV schedule with a routine/medication checklist, and my whole family and I keep an eye on this schedule to get it all done. Sadly, nearly every minute of my day ends up planned and scheduled while on IVs. To pass the time, my family and I watch movies, play cards or dice games, and find reasons to laugh ... and laugh hard!"

– Kristina, Age 35

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# Changing Care Centers

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Sometimes you experience life changes that will have an effect on other aspects of your life. Maybe you are moving to a new city to attend college. Maybe you are starting a new job. Maybe a family member you live with such as a parent, spouse, or significant other has a job change requiring a move. Moving to a new location may mean you have to change your cystic fibrosis care center. A change in your health insurance may mean you have to change care centers, especially if there is more than one care center where you live.

Changing care centers can be a smoother transition than you may think.

## IN THIS SECTION:

- ▶ **Learn how to find a new CF care center** — know what you need to do to establish care at your new center.
- ▶ **Work with your CF team** to make the transition smoother.
- ▶ **Manage your time** — things to do before you make the move.

## Changing Care Centers

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### LEARN HOW TO FIND A NEW CF CARE CENTER

While you are contemplating a change in where you live, you need to find out where the closest CF center is to where you will be living. The Cystic Fibrosis Foundation's website, has a list of [accredited care centers](#). Is the CF center within a reasonable driving distance? If not, you may want to reconsider whether relocating to that area is a good idea.

Even if your move is for college and you will be returning to your current CF center for care, you still need to establish care at a center close to your college — in case you get sick while in school and can't get back to your current center.

Talk to your current CF team about your move. They can help you with deciding which care center might be best for you. They can also help make sure your medical records are sent to your new care center. You will need to sign a release of information form so your records can be sent.

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**It is important to call the new CF center before you make your move so that you can establish care shortly after your move. The new center would prefer to see you in your normal state of health rather than seeing you for the first time when you get sick.**

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### WORK WITH YOUR CF TEAM

Call the CF center before you move and ask to speak to the care center coordinator.

#### Questions to ask:

- › Do they accept your health insurance?
- › What days are clinic?
- › How many physicians do they have, and how do they decide who your physician will be?
- › What happens if you are hospitalized — what is the procedure and how is your care managed while you are in the hospital?
- › Who do you call if you are sick?
- › Who do you call after office hours?
- › To what address should your records be sent?
- › From how far back do they need records?

## Changing Care Centers

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- › Contact your current CF center and give them the information about where your medical records should be sent.
- › Obtain a copy of your most recent chest x-ray or CT scan on a CD to take with you to your new center.
- › Before you move, make an appointment at the new center for a time after you have moved.
- › Give the new center the name and phone number of your current nurse coordinator, so the new center can contact him or her for any questions.

You might have the idea to see if you can go by the new center and have a “meet and greet” with the staff, and see how things are run before you make your decision. Although that may seem like a good idea, it doesn’t tell you how the team will manage your care. The only way to find out is to have a clinic visit. If you decide that you don’t feel comfortable with how your care will be managed after your initial appointment, it is OK to go to another center.

## MANAGE YOUR TIME

### Things to Do Before or Shortly After Your Move

- › Make sure you have an adequate supply of all your medications. Your new center won’t be able to refill your prescriptions until your first appointment with them.
- › If your prescriptions are filled through mail order, call the mail order pharmacy with your new address.
- › If you fill your prescriptions at a retail pharmacy, find a pharmacy in your new location and ask your pharmacy to transfer your prescriptions to the new one.
- › Contact the care center you are leaving to make sure your medical records have been sent.
- › Contact your new care center to make sure they have received your medical records.

Managing and coordinating a move to a new place as well as changing care centers can be stressful. While doing all of the things needed so your move goes smoothly, you also need to make sure you find time to do your CF therapies. If you put your therapies on the back burner during your move, you may get sick — which will add additional stress to your move.



## Changing Care Centers

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Changing care centers does not have to be traumatic. Look at it as another chapter in your life as an adult.

### CHECK LIST FOR MOVING TO A NEW CENTER

TASK	DONE
Locate care center at new location.	
Talk to current CF team.	
Sign release of information form at current center.	
Call new care center to get information about the center.	
Get address from new care center where medical records should be sent.	
Give address of new care center to current care center.	
Make sure medical records are sent to new center.	
Make sure medical records are received by new center.	
Obtain copy of chest x-ray or CT scan on CD.	
Make appointment at new center.	
Get prescriptions filled before the move.	
Find new pharmacy.	
Ask current pharmacy to transfer prescriptions.	

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# Participating in CF Clinical Trials

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We all want to make a difference in the fight against cystic fibrosis. Taking part in clinical trials is one of the surest ways you can have an impact. In fact, many of the CF medications you are likely using — including dornase alfa (Pulmozyme<sup>®</sup>), tobramycin (TOBI<sup>®</sup>), azithromycin, aztreonam (Cayston<sup>®</sup>), and ivacaftor (Kalydeco<sup>®</sup>) — were approved on the basis of clinical trials done at CF centers across the country.

In clinical trials, people with CF just like you test new medications and therapies. People with CF participate in clinical trials for many reasons:

- ▶ To take a more active role in their own health care
- ▶ To get access to a new drug being studied before it is available to the public
- ▶ To help those newly diagnosed with CF
- ▶ To help find a cure for CF
- ▶ To help find new therapies and drugs to improve the quality of life for everyone with CF

If you are thinking about volunteering in a clinical trial, you should gather as much information as possible. We'll help you do just that in this section, where you'll learn about what is involved in clinical trials, how to participate, and what questions to ask before you enroll.

## Participating in CF Clinical Trials

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### IN THIS SECTION:

- ▶ **Learn the basics about clinical research.**
- ▶ **Learn about participating in clinical trials.**
- ▶ **Work with your CF team** to see if clinical trials are right for you.
- ▶ **Get information when you need it** to learn about clinical trials.

### LEARN THE BASICS ABOUT CLINICAL RESEARCH

Finding a new drug usually begins in a lab setting. Once a potential drug is developed in the lab, it must be tested to see how it will work in people with a disease (in this case, CF). Testing is done through clinical trials, or clinical studies. Participating in a clinical trial means you volunteer to take the new drug (or a placebo in some cases) and then have a research team observe how you respond.

Below are some common questions and answers about clinical trials:

- › What are the different types of clinical studies for people with CF?
- › What are the phases of clinical trials?
- › What are the steps in a clinical trial?
- › Who is involved in clinical research?

### WHAT ARE THE DIFFERENT TYPES OF CLINICAL STUDIES FOR PEOPLE WITH CF?

The Cystic Fibrosis Foundation's Clinical Trial Finder search tool lists two types:

- › In an **observational clinical study**, researchers observe certain aspects of routine care and track health outcomes. These studies are very important for developing new ideas about CF and how the disease might best be treated.

A perfect example is the CF Foundation's [Patient Registry](#) Annual Data Report. For those of you who get your care at one of the CF Foundation's [accredited care centers](#), you may well be participating in this very important study. This ongoing study pools information about the health and care of people with CF in the United States to help improve care nationwide.

- › In an **interventional clinical trial**, participants receive one or more treatments

## Participating in CF Clinical Trials

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(interventions) or a placebo so that researchers can evaluate the effects on the participant's health. In addition to tracking changes in your physical health like your breathing and nutrition, some studies also track how the therapy affects your overall well-being.

### WHAT ARE THE PHASES OF CLINICAL TRIALS?

It may take anywhere from 10 to 14 years for a new drug to move from discovery in a lab to testing on human participants to approval by the U.S. Food and Drug Administration (FDA) for regular use. This lengthy process occurs in phases. Each phase carries a risk that the drug might not work or may have unwanted side effects. On average, only one in five drugs tested on humans ever becomes approved for use.

- › **Phase 1 trials:** Researchers test an experimental drug or treatment in a small group of people to learn if it is safe, find a safe and tolerable dosage range, decide how to administer it (orally, intravenously, inhaled, etc.), and learn the side effects. Volunteers for Phase 1 trials do not necessarily have a disease or other health problems.
- › **Phase 2 trials:** The experimental drug or treatment is given to a larger group of people with the targeted disease (in this case, CF) to see how well it works and to keep testing its safety. At this stage, the results do not provide enough information to know whether a drug works to treat an illness, but people may report some benefits. If a drug is found to be dangerous in this phase, or in any phase for that matter, it is withdrawn from further research.
- › **Phase 3 trials:** The experimental drug or treatment is given to large groups of people to continue testing how well it works, determine dosage amounts, watch for side effects, compare it with a commonly used treatment or a dummy drug (placebo), and collect information regarding its safety. The FDA usually approves a drug for general use after Phase 3 trials are successfully completed and there is clear evidence that the drug is working.
- › **Phase 4 trials:** These provide more details about a drug that has been approved to track how it works over a long period and how it affects people's quality of life. They also compare a drug's costs with its effectiveness and may measure the drug against other new and standard treatments.

Watch a webcast about [CF clinical trials](#) to learn more about the basics of clinical research.

# Participating in CF Clinical Trials

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## WHAT ARE THE STEPS IN A CLINICAL TRIAL?

### PROTOCOLS

A **protocol** is a detailed plan for the clinical trial developed by the principal investigator (the main researcher) or the sponsor. It specifies how the study will be conducted, who can and cannot participate, what procedures will occur and when, the number of visits, and the length of the study. Protocols make sure that all participants get the same level of care and that their health remains protected during the study. To maintain safety and ensure accurate data collection, everyone involved in the study, both you and the research team, must follow the protocol exactly as written. Any deviation means that data will not be used in the drug's final analysis.

### RECRUITMENT OF STUDY PARTICIPANTS

This step involves making the public aware of the clinical trial. Sponsors develop information about the specific trial that participating care centers give to people with CF. They also post clinical trial information on [www.clinicaltrials.gov](http://www.clinicaltrials.gov). This information provides an overview of the trial, inclusion and exclusion criteria, and contact information.

The CF Foundation has also developed tools, including [email alerts](#) and a [Clinical Trial Finder](#) to help you find clinical trials for which you may be eligible. If you are interested in a study, contact your center and talk with them to see if it is the right fit for you. Your center team may also approach you about a clinical trial.

### INFORMED CONSENT

If you are interested in participating in a specific clinical trial and meet many of the eligibility criteria, you will receive detailed written information about the study called an informed consent form that explains all parts of the clinical trial: its purpose, what will happen, how long it will last, the possible risks and benefits, your responsibilities, and your right to withdraw at any time. That is a key principle in all of these research trials — even if you agree to take part in a study, you can drop out at any time.

The research team will explain this information and answer all your questions. After you sign the form, you will have a screening visit. But remember, informed consent is more than signing a form — it is a learning process that continues throughout the study. Informed consent makes certain that you can ask questions and get answers before, during, and after the trial. You should receive a written copy of the consent form for review.

You can learn more about informed consent at:

[CFF.org/Our-Research/Clinical-Trials/Clinical-Trials-101/Informed-Consent-Your-Rights-Are-Protected/](http://CFF.org/Our-Research/Clinical-Trials/Clinical-Trials-101/Informed-Consent-Your-Rights-Are-Protected/)

## Participating in CF Clinical Trials

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### SCREENING

The screening process determines if you meet all the eligibility criteria to enroll in the clinical trial.

**Inclusion criteria** are traits everyone must have in order to be in a certain trial. For example, a trial for a new CF medication would require that all participants have a confirmed diagnosis of CF. **Exclusion criteria** are traits that would prevent people from being in a certain trial. For instance, a trial for medication to clear mucus from the airways of young children would exclude adults.

Determining eligibility usually involves the researchers reviewing your medical history, asking a series of questions, and conducting screening tests and a physical exam. If you pass the screening process, then you will be scheduled for the next study visit.

### STUDY VISITS

The number of visits and types of procedures done at the research site and at home vary. Each trial's protocol describes the time and frequency of the research appointments and what goes on at every research visit. Some study protocols require tests, such as undergoing lung function or breathing tests, having blood and urine samples taken, or completing a survey about how you feel. It is important to the study's success that participants do everything expected of them and stay in close touch with the research team.

## WHO IS INVOLVED IN CLINICAL RESEARCH?

### THE SPONSOR(S)

Any number of organizations or individuals can sponsor clinical research in part or entirely. For example, medical institutions, universities, foundations, voluntary groups, drug companies, and federal agencies, such as the National Institutes of Health, all sponsor research. The sponsor usually has these responsibilities:

- › Develop the study protocol.
- › Choose a principal investigator to run the trial.
- › Provide all the information to conduct the trial.
- › Monitor the study's progress and data collected.
- › Report adverse events to the FDA.
- › Analyze the data and report results.

# Participating in CF Clinical Trials

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## THE STUDY TEAM

- › **Principal Investigator:** The principal investigator (PI) is the main doctor overseeing the study at the care center. This person has the background and training in science and research to lead the study. The PI follows the protocol as he or she recruits participants, sees them at study visits, monitors each participant's safety and health throughout the study, and works with the research coordinator on administrative aspects of the trial.
- › **Research Coordinator:** The research coordinator (RC) assists the PI in conducting a clinical trial. The RC recruits participants, follows the protocol to ensure participant safety, coordinates the trial's day-to-day activities, and collects and manages data. From an administrative standpoint, the RC works with the institutional officials on contract with the sponsor and often prepares and negotiates the budget with the sponsor.
- › **Research Participant (also called research subject or volunteer — that's you!!):** The research participant is the person who volunteers for the study. The research participant is an active part of the team with clearly defined rights.

## LEARN ABOUT PARTICIPATING IN CLINICAL TRIALS

So, you may be interested in joining a clinical trial. You could play a key role in the advancement of CF treatments. But what exactly are you getting yourself into?

Here are answers to some commonly asked questions about participating in clinical trials:

- › How can I find out about trials in which I might participate?
- › What are the benefits and risks to participating in clinical trials?
- › How is my safety protected in clinical trials?
- › Does participating in a clinical trial replace visits to my care team?
- › Will I receive reimbursement for participation?

You can also watch a [webcast about CF clinical trials](#) to learn more.

# Participating in CF Clinical Trials

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## HOW CAN I FIND OUT ABOUT TRIALS IN WHICH I MIGHT PARTICIPATE?

Sponsors — the organization or individuals sponsoring the research — post their clinical trial information on [www.clinicaltrials.gov](http://www.clinicaltrials.gov). You'll see an overview of the trial, inclusion and exclusion criteria, and contact information.

The CF Foundation has also developed materials to explain general clinical trial information, and you can learn more about clinical trials [here](#). There are also materials available at your care center. If you are interested in a study, contact your center to learn more.

Your center team may also approach you about a clinical trial. After initial recruitment, there will be a screening of all potential study participants.

## WHAT ARE THE BENEFITS AND RISKS TO PARTICIPATING IN CLINICAL TRIALS?

### **Possible benefits:**

- › Helping others by contributing to medical research
- › Taking an active role in your own health care
- › Gaining access to new treatments not available to the public
- › Getting expert medical care at leading health care facilities
- › Receiving a treatment that works for you

### **Possible risks:**

- › Side effects or adverse reactions to medications or treatments
- › Receiving a treatment that doesn't work for you
- › Being required to travel to the study site, receive treatments, stay in the hospital, or follow complex dosage requirements



## Participating in CF Clinical Trials

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### HOW IS MY SAFETY PROTECTED IN CLINICAL TRIALS?

The U.S. government has very strict guidelines and safeguards to help protect clinical research participants. The CF Foundation goes beyond these guidelines to keep risks as low as possible and to help ensure that the risks are worth any possible benefits.

In every CF clinical trial, there are many layers of protection:

- › The FDA is the federal agency that must approve all clinical trial protocols. The FDA monitors studies to ensure that all guidelines are followed as the trial goes on. Upon completion of the trial, the FDA determines whether the new drug or therapy will be approved for public use.
- › The study sponsor is the company or individual that creates the clinical trial protocol. Study sponsors make sure that the protocol includes a guide for how safety will be monitored. During the study, the sponsor provides a medical expert who reviews adverse events in real time.
- › The Therapeutics Development Network (TDN) is a group of care centers across the country that conduct CF research studies. Experts from the TDN, along with CF community members, review clinical trial protocols to judge the merit of the study drug, study design, and safety.
- › **The Data Safety Monitoring Board (DSMB)** is an independent committee of experts in CF that review safety data before a study begins and make a plan (also known as a charter) on how the study will be monitored for safety. Once a study begins, the DSMB monitors the study for possible problems or unwanted side effects. The CF Foundation is the only voluntary health organization to organize a DSMB whose members are experts in CF, completely independent, and not involved in any way with trials or their participants. In this way, the CF Foundation does its best to keep participants safe throughout the clinical trial.
- › The institutional review board (IRB) of the participating hospital or university is a committee usually made up of doctors, other health care providers, and the public. The committee looks at the trial's protocol to make sure participants' rights are protected and the trial does not cause unnecessary risk.
- › The PI at your site is responsible for overseeing the study. Before you enroll in a study, the PI will review your health to see if you can safely participate. During the study, the PI will continue to monitor your health and can pull you out of the study if your health becomes a concern.

## Participating in CF Clinical Trials

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### DOES PARTICIPATING IN A CLINICAL TRIAL REPLACE VISITS TO MY CARE TEAM?

No. If you are participating at a CF care center other than where you receive care, you still need to see your CF care team for regular clinical care visits in addition to the research visits. If your research team notes any worsening of your health during the trial, they will notify your care team and the study sponsor. They must decide if the change in your health was related to the study drug. Many studies of new medications will include scheduled visits after you have stopped taking the drug, to monitor your reaction.

### WILL I RECEIVE REIMBURSEMENT FOR PARTICIPATION?

Some clinical trials offer research participants money to support their participation. How much, if anything, is up to the trial sponsor. The amount of payment often depends on the kind of trial and what will be asked of you. When money is given, it is not meant to bribe you into participating but rather to help pay for your time and travel.

Under the [Ensuring Access to Clinical Trials Act \(EACT\) of 2015](#), clinical trial participants who receive Supplemental Security Income (SSI) or Medicaid benefits may accept up to a certain amount in research compensation without losing government medical benefits. To qualify for the exemption, SSI recipients must report any compensation received for participating in a qualifying clinical trial to the Social Security Administration.

You will receive information about compensation during the informed consent process before the study begins.

### HOW LONG DOES IT TAKE TO LEARN ABOUT RESULTS?

Usually participants will learn the results of a study in a timely manner. But there are several reasons why trial results may be released either very slowly or not at all:

- › Participants may be recruited over several years and getting the results back after the final volunteer has been tested can take a long time.
- › It may take a while to analyze the results of the trial to find out if it helps people with CF.
- › Typically, only one in three drugs ever makes it to Phase 3 of a clinical trial. Results for drugs that do not make it are usually not published. When results can be published, a strict review process slows down the reporting. Publication in a medical journal may not happen for a year or more after the trial is over.

## Participating in CF Clinical Trials

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All of these factors make it difficult to communicate the results to the CF community. Even so, the CF Foundation is committed to improving the communication of results to trial participants. It will post results as they are available on the [Clinical Trial Finder](#).

### WORK WITH YOUR CF TEAM

To participate or not to participate — it's your personal decision. But you can definitely rely on your care team to help you gather the facts and weigh the advantages and disadvantages.

When you speak with your CF physician or the research coordinator at your care center about a clinical trial, consider asking the following questions:

- › What is the purpose of the study?
- › What will be asked of me?
- › Who will be in charge?
- › Will the study benefit me and others?
- › Why do researchers think that this particular drug or treatment might work?
- › What current treatments, if any, do I have to come off during the trial?
- › What kinds of tests and experimental treatments are involved?
- › How do the possible risks, side effects, and benefits compare with my current treatment?
- › How long will the study last?
- › Will hospitalization be necessary?
- › Who will pay for my participation in the trial? Will I be reimbursed for any expenses?
- › How will I know if the experimental drug is working?
- › Will I receive results of the studies?
- › What type of long-term follow-up care will be required?
- › How will I fit the study schedule into my daily life along with school, work, and treatments?

## Participating in CF Clinical Trials

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If you attend a care center that is not participating in clinical trials, or in the clinical trial you are interested in, use the [Clinical Trial Finder](#) to find the right trial for you. You can search for trials choosing filters for age, mutations, FEV<sub>1</sub>, therapeutic approach, and even the distance you are willing to travel. Also, you can sign up to get [email updates](#) when new trials and results are posted.

If you are interested in a particular study, contact information can be entered into a pre-populated email and sent to the participating site's research coordinator. There is also the option to print the study description page, or email it to your care provider or care center nurse, so you can discuss it with your care team at your next clinic appointment.

**REMEMBER: Participation in a research study is ALWAYS voluntary.**

### GET INFORMATION WHEN YOU NEED IT

Want to learn more about participating in CF clinical trials? Try these resources:

- › Information on the CF Foundation's website, featuring people with CF, their families, and their research teams talking about clinical trial participation: [CFF.org/Research/Developing-New-Treatments/Clinical-Trials/Clinical-Trials-101/Why-Participate-in-a-Clinical-Trial/](https://www.cff.org/Research/Developing-New-Treatments/Clinical-Trials/Clinical-Trials-101/Why-Participate-in-a-Clinical-Trial/)
- › The FDA at: [www.fda.gov/default.htm](https://www.fda.gov/default.htm)
- › The National Institutes of Health: National Institute of Child Health and Human Development: [www.nichd.nih.gov/health/clinicalresearch/Pages/index.aspx](https://www.nichd.nih.gov/health/clinicalresearch/Pages/index.aspx)

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On behalf of the Cystic Fibrosis Foundation, we would like to thank the following members of our community for lending their time, expertise, and insights into creating the Adult Guide.

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