Understanding cystic fibrosis

Walgreens

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What you need to know about cystic fibrosis

Learning how to manage cystic fibrosis (CF) might feel like a challenge, especially at first. But understanding your diagnosis can help you take control of your health. Staying on track with treatment and adopting key lifestyle changes can help you control symptoms and improve your overall health. This booklet provides information about CF, what to expect after diagnosis and how to keep up with your treatments to live a full and active life.

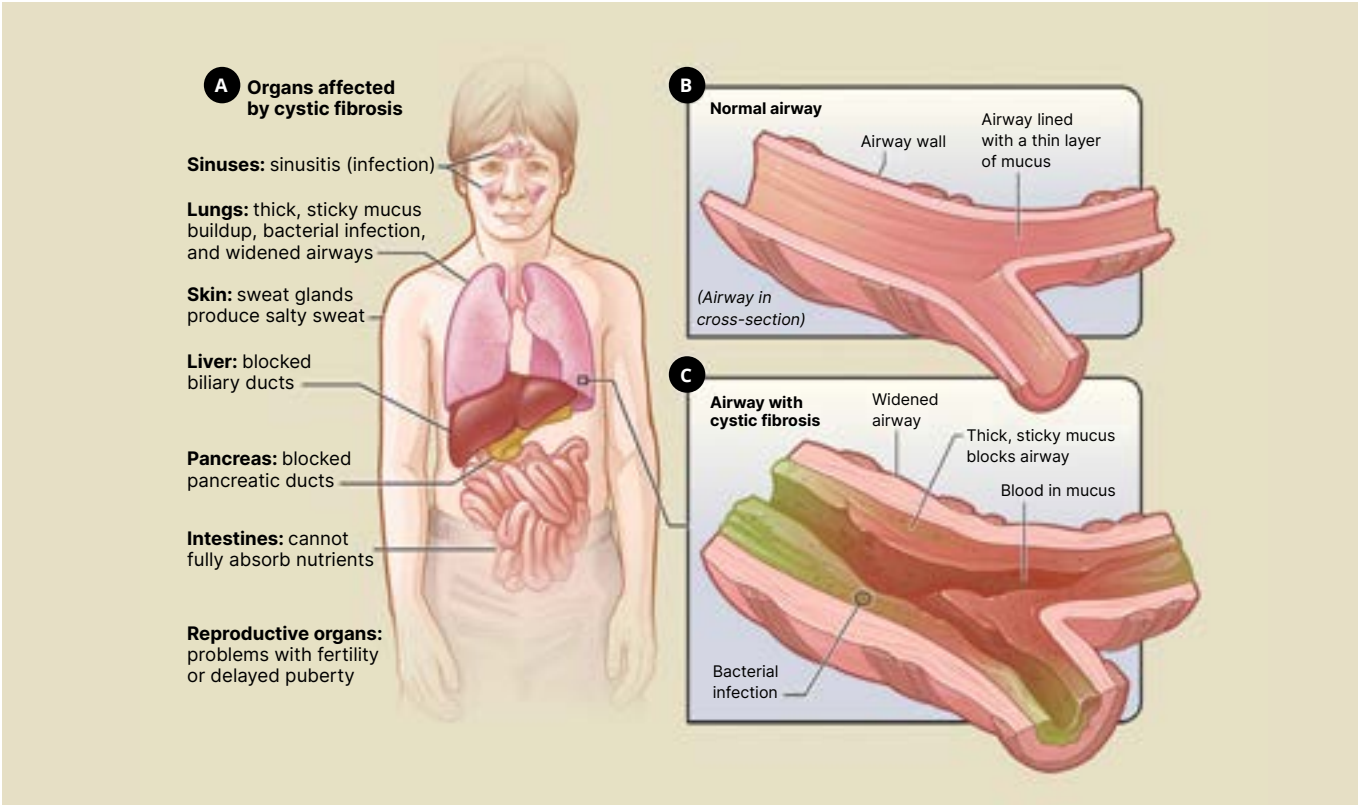
Overview

CF is a genetic disease that affects how the body produces mucus. Normally, mucus in the body is slippery and protective. In CF, mucus is thick and sticky. This can cause problems in the way the body works.[1,2]

Thick, sticky mucus can cause blockages and trap bacteria. This raises the risk for infections. This can also cause inflammation. In some cases, it can damage the organs.[1,2]

CF mainly affects the respiratory and the digestive systems. It can cause problems with the pancreas, liver and kidneys. It can also affect the reproductive system.[1,2]

Figure 1. Affected body systems in CF



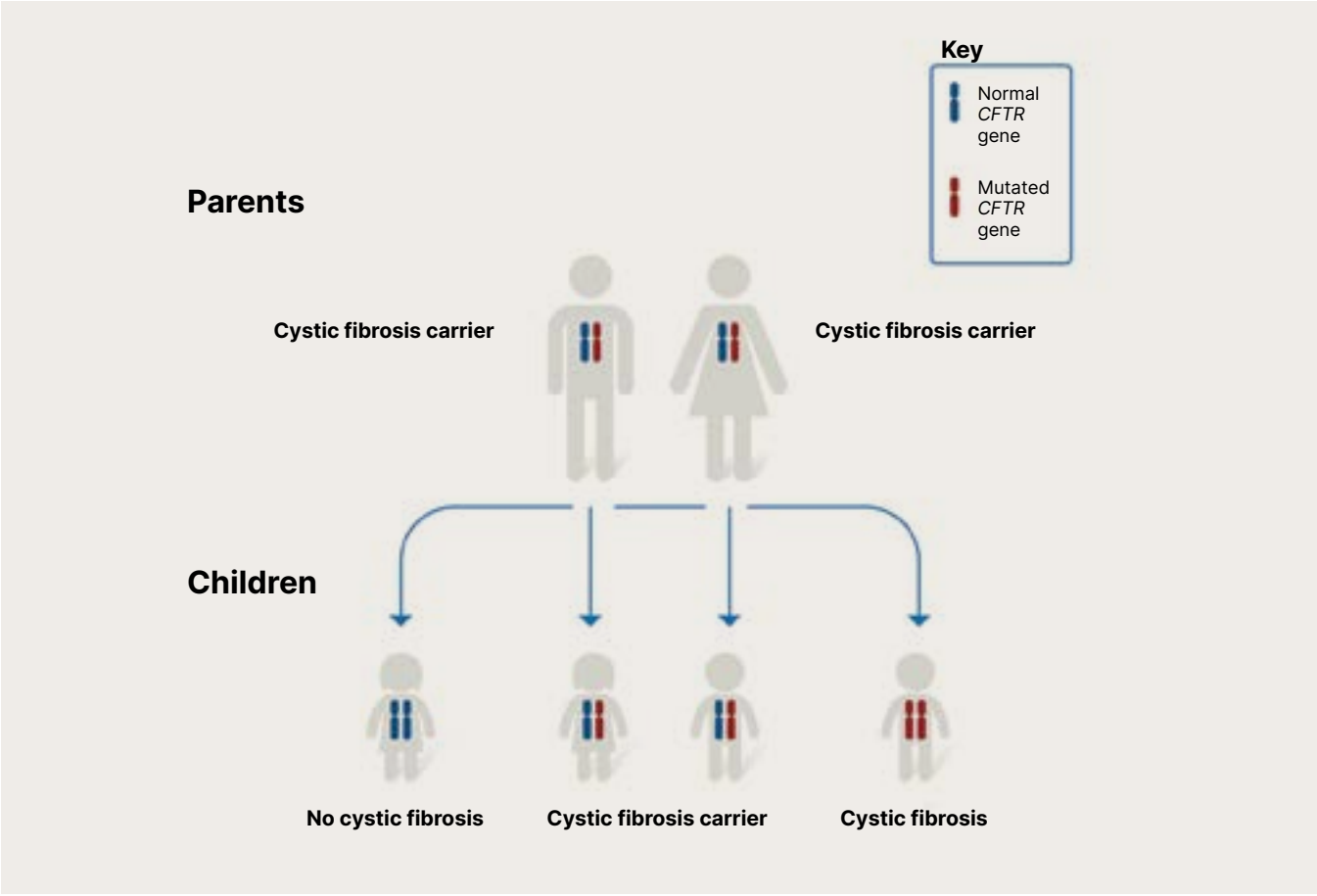
Source: National Heart, Lung, and Blood Institute; National Institutes of Health; U.S. Department of Health and Human Services.

Causes and risk factors

CF runs in families. It is caused by a problem with the cystic fibrosis transmembrane conductance regulator (CFTR) gene.[1] This gene has a role in how the body makes mucus. In CF, there is a mutation, or defect, in this gene.

People with CF received two copies of the mutation — one from each parent.[1,2] Some people have only one copy of the defective gene. They do not have CF and are called carriers. They can still pass the gene mutation on to future children.[2] An estimated 10 million people in the United States carry the CFTR gene mutation but do not know it.[1] Figure 2 shows how CF is passed down in a family.

Figure 2. CF inheritance pattern



Source: National Heart, Lung, and Blood Institute; National Institutes of Health; U.S. Department of Health and Human Services.

Having a parent with CF or with the CFTR mutation raises the risk of CF. The risk is higher for people whose siblings, half-siblings or first cousins have CF. The disease is also more common in people of Northern European descent.[1,4]

Diagnosis

Confirming a CF diagnosis can include many steps. Your diagnosis or your child's diagnosis might have come after a number of tests and evaluations, such as:

- Prenatal testing[1]
- Newborn screening[2,4]
- Sweat chloride testing[1,2]
- Genetic or carrier testing[1,2]
- Clinical evaluation[2]

Genetic testing can find what types of CFTR mutations affect a specific person. There are currently more than 2,000 recognized CFTR defects. They are grouped into seven different classes based on how they affect CFTR protein.[3] Knowing which ones affect you can help your doctor build the right treatment plan.[3]

CF by the numbers

Approximately 40,000 people in the United States have CF.[2]

More than half of the CF population is **age 18 or older**.[2]

More than 75% are diagnosed **by age 2**.[2]

About **1,000 new cases** are diagnosed **each year**.[2]

Symptoms

Most people living with CF have symptoms that affect their daily lives. Some people have minor symptoms or none at all. In other cases, symptoms can become severe or life-threatening.

Common symptoms can include[1,2]:

- Diarrhea, loose stools or constipation
- Frequent lung infections
- Persistent cough, sometimes with phlegm or blood
- Poor or delayed growth
- Stomach pain
- Wheezing or trouble breathing

Other symptoms can include but are not limited to[1,2]:

- Fertility problems in men
- Muscle and joint pain
- Salty skin and saltier than normal sweat

Complications

CF most commonly causes problems in the lungs and in the pancreas. It can also cause problems in other parts of the body. This can lead to serious health issues, such as[1,4]:

- Arthritis and weak bones
- Cancers of the digestive tract
- Diabetes
- Heart failure
- Kidney problems
- Liver disease or liver failure
- Lung damage requiring lung transplantation

Living with cystic fibrosis

There is not yet a cure for CF. However, several therapies can help people manage CF and improve their overall health. The goals of CF treatment include[1,2]:

- Clearing the airways
- Correcting how the CFTR protein works
- Improving nutrition
- Preventing complications

Your doctor will build a treatment plan that is right for you. Most CF patients are prescribed a combination of airway clearance therapies (ACTs) and medications.[1] These treatments are combined with healthful eating and regular physical activity.[1,2]

Airway clearance therapies

ACTs use breathing, movement and coughing to loosen and clear lung mucus.[1] These techniques can include[5]:

- **Active cycle of breathing technique:** Controlled breathing to expand the chest, then forcefully cough out mucus
- **Autogenic drainage:** Varied speeds of breathing to move mucus through the small, medium and large airways to be coughed out

- **Percussion and postural drainage:** Varying positions to allow gravity to drain mucus, often paired with clapping or vibrating the chest to loosen mucus
- **High-frequency chest compression:** An inflatable vest that compresses and vibrates the chest to loosen mucus
- **Positive expiratory pressure:** Breathing through a device that makes it harder to exhale, which opens the airways and moves mucus

No one method is preferred over the others. You can work with your doctor to choose which ones are best for you. Finding something that fits your daily life can help you keep up with ACT.[5]

Medications

Along with ACT, medications help clear your lungs and make it easier to breathe. They can also fight infections and improve the way the CFTR protein works. Table 1 lists some common CF medications.

Table 1. CF medications[1,2]

Medication	Action
Antibiotics	Prevent or treat infection and improve lung function
Anti-inflammatory drugs (e.g., ibuprofen, corticosteroids)	Reduce inflammation
Bronchodilators	Relax and open the airways
CFTR modulators	Help defective CFTR protein work better
Mucus thinners	Make it easier to clear mucus from the airways
Pancreatic enzyme replacement therapy (PERT)	Help digestion and improve how the body absorbs nutrients from food

Staying on track with treatment

Keeping up with CF therapy is important. It can help you control your symptoms and improve your health. No matter which drugs and therapies are part of your treatment, you should use them exactly as your doctor prescribes. This means keeping up with daily ACTs. It also means taking medications at the right times and the correct doses.

CF treatment can take a good amount of time each day. Planning therapies and building them into your day will help you stay on track. This might include [6,7]:

- Doing treatments while checking email, reading or watching TV
- Setting reminders on your calendar, computer or phone
- Scheduling treatments and equipment cleaning for the same times each day

Some days, it might feel hard to keep up with your treatment routine. Other times, you might feel like it isn't helping. Do not stop treatment on your own. Tell your care team your concerns. They can help you adapt your treatment schedule so it works best for you.

Treatment away from home

It is important to keep up with CF treatment every day. You will need to continue your therapy even when you are at school or work and while you are traveling. Planning ahead can make it easier to stay on track.

School and work [7,8]

- Explain your medical needs to your school or employer.
- Work with administrators, teachers or colleagues to plan for your needs.
- Arrange for any time or space you might need for your care during the day.
- Make treatment a priority and schedule it into your day, even if it is before school or work.

Travel[7]

- Ask your doctor for signed documents that list your health summary, medications, equipment and supplies.
- Pack extra medication or extra prescriptions.
- Plan to keep your medication and equipment where you can access them easily.
- Have a way to keep medications cool.
- Bring supplies to power and clean your equipment.

These are just some of the ways you can plan for activities away from home. Talk with your care team about other things you might need to consider.

Lung transplantation

CF is a progressive disease. This means it can get worse over time. For some, CF can cause severe lung damage. It can also cause respiratory failure.

In these cases, lung transplantation might be a treatment option.[1] This replaces damaged lungs with healthy ones. It can improve a person's quality of life. It might also help extend their lifespan.[7]

A lung transplant is complex. It requires careful preparation and lifelong care. Thinking about such a major procedure can feel scary. You can talk openly with your care team about lung transplantation long before it might ever be needed. This can help you better understand what is involved in the transplant process.[7]

Lifestyle changes

Along with your treatment regimen, healthy lifestyle changes can help you control CF. Healthy habits can help slow the disease process. They can also help lower the risk for serious health problems. Staying active, eating well, avoiding infections and managing emotional health are important parts of staying well with CF.

Physical activity

Regular exercise can help improve overall health. It can also strengthen the bones and muscles.[1] In CF, exercise can improve how the lungs work.[1] It can induce coughing and help get rid of extra mucus. It can also help control other health conditions that occur with CF, such as diabetes.[7]

Current guidelines (US Department of Health and Human Services) recommend adults with chronic conditions get at least 150 minutes of moderately intense activity each week. Children and adolescents should get about 60 minutes of movement a day. Talk to your doctor about how much activity is right for you.[9]

Daily activity can be broken up into smaller sessions throughout the day. Workouts should be spread out over the week. Movement should include both aerobic and muscle-strengthening exercises.[9]

Strengthening exercises might include weightlifting, resistance band work or body weight exercises like push-ups or planks.[9] Moderate-intensity aerobic exercise might include[9]:

- Ballroom or line dancing
- Brisk walking
- Cycling
- Doubles tennis
- Swimming
- Water aerobics
- Yard work or home repairs
- Yoga

Think of physical activity as a prescription. It is important to keep up with it to improve your health. Several steps can help you get started and stay on track[7,9]:

- Check your fitness level. Your care team can measure how much you can do. They can also help you track how activity might affect your oxygen levels.
- Choose activities that match your abilities. Find activities you enjoy. If you haven't already been active, start out slowly. Choose simple routines you can maintain.
- Set detailed goals. Be specific about how long, how often and how hard you will exercise. Outline how you will meet your goals.
- Keep track of your progress. Record how much you are doing each day. This might mean logging things like minutes or steps. Watching your progress toward a goal can keep you going.

Optimal nutrition

Good nutrition is important for good health. For those with CF, a healthful diet is critical. In CF, the pancreas does not make enough enzymes to help with digestion. This makes it hard for the body to absorb nutrients from food. At the same time, the thick, sticky mucus of CF can cause digestive problems. These problems can affect growth and overall health.[1,4,7]

A CF diet will focus on helping you reach and maintain a healthy weight. Your target weight is based on your body mass index (BMI). This is found by dividing your body weight in kilograms by your height in meters squared. Clinical guidelines for those with CF recommend a BMI of at least 22 for women and at least 23 for men.[7, 10]

A healthy weight is important for fighting off infections and keeping up energy.[7] Better nutrition can mean better lung function.[10] Each person's nutritional needs are different. These can also change over time. A dietitian can help you build a diet that gets you the calories and nutrients you need.

Food choices

CF can pose many nutritional challenges. A person with CF usually needs more daily calories, sodium, protein and fat than a person without CF. Table 2 lists some ways to meet these needs. You can work with your dietitian to choose specific foods that boost your diet.

Table 2. Meeting CF nutritional needs[10,11]

Nutritional need	Example sources
Additional calories and fat	• Fatty fish, such as salmon or mackerel • Full-fat dairy products, alone or mixed into other foods • Healthy fats in nuts, avocados, olive oil
Fat-soluble vitamins (A, D, E and K)	• Eggs, dairy and fatty fish • Fortified breakfast cereals • Leafy green vegetables and sweet potatoes • Vegetable oils, nuts and seeds
Calcium	• Calcium-fortified beverages • Collard greens and kale • Full-fat dairy • Salmon with soft bones
Sodium	• Fluids with added salt, such as electrolyte sports drinks • Salt for seasoning food
Iron	• Beans, lentils and beef • Fortified breakfast cereals • Leafy greens • Oysters
Zinc	• Beans • Dairy products • Eggs • Meats • Shellfish

It is also important to get enough water. Staying hydrated helps with digestion. It can also help prevent constipation.[7] Your daily intake of water comes from everything you eat and drink. This might include milk, juice, fruits or soup.[10] How much water you need can depend on many things, like your age, weight or level of activity.[7,10]

Plan well, eat well

A little bit of planning can help make it easier to eat right, even when things get busy. Small steps can help you save time and stay on track[7]:

- **Prep menus:** Plan your meals for the next few days. This can prevent last-minute searches for healthful food.
- **Make meals ahead of time:** Try to cook or prep double servings of meals. Freeze or save the extra servings for later.
- **Pack snacks:** Bring nutrient-dense, high-calorie snacks with you when you leave home. This might include trail mix, yogurt or granola bars.
- **Remember supplements:** Always keep enzymes with you. Pack enzymes with meals and snacks. Keep them away from heat.

Dietary medications and supplements

Certain medications and supplements are also part of a CF diet plan. Some help with digestion. Others help boost nutrition. Table 3 lists some common examples.

Table 3. Dietary medications and supplements[7]

Type	Action
Antacids	Reduce stomach acid
CF-specific vitamins (A, D, E and K)	Replace vitamins that are hard to absorb with CF
H2 blockers	Reduce stomach acid
PERT capsules	Aid digestion of nutrients from food
Polyethylene glycol	Reverse hard stool and constipation
Probiotics	Improve intestinal bacteria
Proton pump inhibitors	Reduce stomach acid

Each person has different needs. Talk with your CF team about what will support your diet. They can help you understand why, when and how often to take certain medications and supplements.

Supplemental nutrition

Sometimes, it can be hard to eat enough food to meet a daily calorie goal for CF. Some people need supplemental nutrition. This is also called tube feeding.[7] With these feedings, liquid supplements are taken directly through a tube. This can provide nutrients and calories beyond what a person eats each day. Tubes can be placed in different ways[7]:

- **Gastrostomy tube (G-tube):** Inserted through the abdomen and into the stomach and can be fitted with a flatter "button" that can be opened and closed for use
- **Jejunostomy tube (J-tube):** Inserted through the abdomen and into a section of the small intestine called the jejunum
- **GJ-tube:** Inserted through the abdomen to pass through the stomach and end at the jejunum
- **Nasogastric tube:** Inserted in the nose, down the throat into the stomach, and inserted/removed for nightly feedings

If you are having trouble eating enough and keeping a healthy weight, ask your CF care team if tube feeding is an option for you.

Cystic fibrosis-related diabetes (CFRD)

A hormone called insulin helps the body process glucose from food and move it into the cells for energy. In CF, problems with the pancreas make it hard to make insulin. When this happens, too much glucose stays in the bloodstream. This can cause CF-related diabetes (CFRD).[7]

CFRD develops in about 20% of teens and in 40% to 50% of adults with CF.[7] It can cause serious health problems if left untreated. The lack of insulin makes it hard to keep a healthy weight. It also makes lung function worse.[7]

Some early signs of CFRD might include[7]:

- Constant thirst
- Fatigue
- Frequent urination
- Unexplained loss of lung function
- Weight loss or difficulty gaining weight

Tell your CF care team if you notice any of these symptoms. Your ongoing care should also include regular screenings for CFRD. In the event of a positive diagnosis, changes in diet, exercise, supplements and medications can help keep CFRD under control.[7]

Family planning and pregnancy

Though CF treatment focuses largely on the lungs and digestive system, CF care also includes particular attention to reproductive health. For adults living with CF, family planning and pregnancy require special considerations.

Those who wish to avoid pregnancy should use an effective form of birth control. The chosen method can depend on a person's CF treatment plan and overall health. For example, some types of hormonal birth control can worsen CF-related issues like bone problems and kidney or liver disease. Hormonal birth control might also be less effective because of poor absorption in the intestines or as a consequence of ongoing antibiotic treatment.[7] Ask your doctor what type of birth control is safe for you.

Those hoping to get pregnant will have different considerations. CF and its related health issues might make it hard for a woman to get pregnant or stay pregnant without medical support.[7] Many men, about 95%, have a condition called azoospermia, which is the absence of sperm in the ejaculate.[7] For these men, medical interventions may be needed when planning a family.[7] Prospective parents also need to consider how CF might affect future children. Preparing for pregnancy might include genetic testing. This can assess the likelihood of having a child with CF.[4,7] Prospective mothers will also need to assess their health status to prepare for the physical demands of pregnancy and breastfeeding. This might include[7,12]:

- Improving lung function
- Improving nutrition
- Planning a diet with enough additional calories
- Preventing or controlling blood glucose or diabetes
- Reaching a healthy BMI of 22 or higher
- Switching to treatments or therapy schedules that are safer for pregnancy or breastfeeding

Share your plans and any concerns about reproductive health with your care team. They can provide guidance and support. They can also refer you to specialists if needed.

Avoiding infections

In CF, thick mucus in the lungs can make it easy for germs to take hold. This can cause long-term infections and respiratory problems. When lungs with CF try to fight off infection, they become inflamed. This produces even more sticky mucus. All of this can damage the lungs.[7]

Precautions against infection are important. Germs like bacteria, fungi and viruses can spread through direct contact. They can spread through droplets, like sneezes or coughs. They can also spread through the air.[7] Some healthy habits can help you limit infections. Keep these basic practices in mind[1,7]:

- Avoid close contact with others who have CF. Germs spread more easily between people with CF.
- Clean your CF equipment thoroughly.
- Do not share personal items like cups, utensils or toothbrushes.
- Keep your hands clean. Wash your hands with soap and water for at least 20 seconds. If you don't have water, use an alcohol-based hand sanitizer.
- Keep your hands away from your nose and mouth.
- Stay current with immunizations. This includes vaccines for the flu, coronavirus, pneumonia and whooping cough. Your close contacts should also be up to date with vaccines.
- Consider wearing a mask, especially in large indoor settings or on public transportation.
- Take your medications as prescribed.

It's important to stick to these habits wherever you are. Encourage others to follow these precautions as well. This includes those who care for you at home and at medical appointments. Preventing and avoiding infections is crucial for your health.

Emotional health

Life with CF can be demanding. Fitting in daily treatments might sometimes feel stressful. Lifestyle changes might feel too hard to maintain. All of this can make you feel anxious, mad or sad. This is normal. You can find healthy ways to cope[7,13]:

- Avoid alcohol, caffeine, cigarettes or drugs.
- Create a daily schedule.
- Do not skip meals or treatments.
- Get enough sleep each night.
- Go outside every day.
- Join a CF support group.
- Learn and use relaxation techniques.
- Make time for things you enjoy.
- Move your body.
- Spend time with people who make you happy.
- Talk to someone about your feelings.

Some days will be better than others. If you try these things and it still feels too hard to get through each day, it's important to get help. People with chronic conditions like CF are more likely to have anxiety and depression.[7] Improving your mental health will support your overall health.

Talk to your care team if you have any of these symptoms of anxiety[7]:

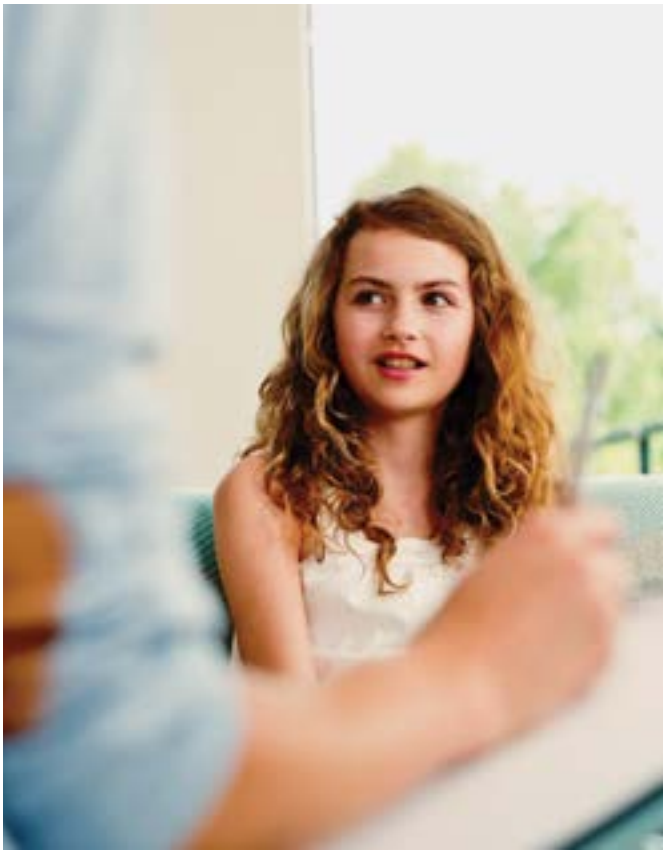
- Difficulty concentrating
- Frequent, excessive worrying for at least half a year
- Muscle tension
- Restlessness
- Sleep problems
- Trouble controlling worry

Tell your care team if you have any of these symptoms of depression[7]:

- Aches, pains, cramps or digestive problems that don't go away
- Trouble with concentration, memory or decisions
- Eating too much or too little
- Fatigue and low energy
- Feeling guilty, worthless or helpless
- Feeling hopeless, negative or irritable
- Feeling sad or anxious most of the time
- No interest in things you used to enjoy
- Sleep problems
- Thoughts of harming yourself

If you have thoughts of suicide, call 911 or your local emergency services number. You can also call a doctor, mental health professional, crisis center or hotline for help.

Every person is different. Some have many symptoms of depression or anxiety. Others have just a few. Ask your care team about things that could help, like counseling or medication. For some people, talking with a therapist is useful. For others, taking antidepressant or anti-anxiety medications can help. Many people use counseling and medication together to successfully improve their mental health.[7]



Ongoing care

Work with your care team to manage your CF treatment. Regular checkups will help you and your care team keep track of your condition and your overall health. You can talk about how well your treatment regimen and lifestyle changes are working. Your doctor can also adjust your therapies as needed.

We provide this information because the more you know about cystic fibrosis — the better you'll be able to manage it.

Additionally, the Walgreens Specialty360 Therapy Team is here to support you with dependable, personalized service to help manage your medication side effects and stay on track with your prescribed therapy.

We look forward to being a member of your healthcare team and helping you get the best results from your treatment.

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Resources

You might find it helpful to contact these organizations for additional support and resources.*

Cystic Fibrosis Foundation

www.cff.org
800-344-4823
Facebook: www.facebook.com/cysticfibrosisfoundation
Twitter: www.twitter.com/CF_Foundation

The CF Foundation is a donor-funded nonprofit that supports ongoing research for CF therapies and the mission of finding a cure. Its website features educational materials for patients and families, information about care centers and updates on the latest research developments. The CFF also offers case management and guidance regarding insurance, financial and legal information through its Compass program.

Cystic Fibrosis Research, Inc.

www.cfri.org
855-237-4669
Facebook: www.facebook.com/cfri.org
Twitter: www.twitter.com/CysticFibrosisR

CFRI is a nonprofit charitable organization that funds CF research and offers education, advocacy and support programs for CF patients and their families and caregivers. The CFRI website features educational materials and registration information for a host of CF support programs and wellness classes.

National Heart, Lung, and Blood Institute

www.nhlbi.nih.gov
1-877-NHLBI4U (1-877-645-2448)
Facebook: www.facebook.com/NHLBI/
Twitter: www.twitter.com/nih_nhlbi

The National Heart, Lung, and Blood Institute oversees the research, training and education for the prevention and treatment of heart, lung and blood disorders. Its website features educational resources for patients and clinicians and information about awareness efforts and clinical trials for a variety of disease and conditions, including cystic fibrosis.

*The referenced organizations are provided for informational purposes only. They are not affiliated with, and have not provided funding to Walgreens for, this booklet. Walgreens does not endorse or recommend any specific organization.

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