

# Pain

**People with cystic fibrosis (CF) often report that pain is a troubling problem. Acute pain usually has a clear source and a foreseeable end. It may result from a CF-related injury, such as rib trauma caused by intense coughing, or from an unrelated problem, such as migraine or acute low back pain. Acute pain also occurs after surgery or other medical interventions.**

People with CF also can develop chronic pain, which comes back frequently or is constant for months or more. Chronic pain can be related to CF or to a separate condition, such as fibromyalgia.

## What are Some Causes of Pain in CF?

Acute or chronic pain can be directly related to the effect CF has on multiple organs in the body, for example:

- CF sinus disease can cause headache, or pain in the eye or ear.
- CF lung disease, with cough and rapid breathing, can put stress on the chest wall and produce chest or back pain.
- CF disease in the gastrointestinal system—the gallbladder, liver, pancreas, or intestines— can result in various types of pain.
- Chronic inflammation from CF may lead to joint pain in the arms or legs.

## Potential Concerns Caused by Chronic Pain

Whether or not chronic pain is directly related to CF— indeed, whether there is any diagnosis that explains why pain is so persistent—it may damage your physical and emotional health, day-to-day functioning, and quality of life. Some people develop depressed or anxious mood or trouble sleeping. Some reduce their physical activity,

becoming deconditioned. Mood changes and physical limitations may lead to more pain with movement or exercise, less social interaction, and trouble with self-care. It can be harder to feel like doing CF treatments, such as airway clearance. These consequences can make it difficult to work, go to school, play, or do other activities that are important to you. Chronic pain usually needs treatments focused specifically on the pain and the impact it has on you and your family.

## If Pain Occurs, What Should I Do?

If you experience a new and unfamiliar type of acute pain, or if pain is not going away, the first step is to talk to your CF care team.

Treatment must always be based on a careful assessment. You should be prepared to describe the pain in detail, including its impact on your mood and function, and list treatments that have been tried (see Self-Assessment Guide). In some cases, the CF team will ask for tests to identify the cause of the pain in the hope that treating the cause will reduce the pain.

## Self-Assessment Guide

- Keep track of your pain with a daily log. Things to track include:
  - Location of the pain
  - When did the pain start? How long did it last?
  - Did the pain occur at rest or with activity?
  - Describe the pain: sharp, dull, burning, throbbing.
  - On a scale of 0 (no pain) to 10 (worst possible pain), rate the pain

0	1	2	3	4	5	6	7	8	9	10
No Pain				Moderate Pain						Worst Possible Pain

- What makes the pain better?
- What makes the pain worse?

- What medications or strategies did you use for the pain, and did these help?
  - Is the pain interfering with your life? Is it preventing you from doing things you planned to do? Is it affecting your family or loved ones?
  - What are your expectations for pain treatment? What in your life would change if your pain were to significantly improve?
- Discuss this with your CF care team to determine the next course of action. They will assess if you need an urgent or routine evaluation.
  - Call your care team immediately if your pain worsens and is uncontrollable.

When turning to specific treatments for the pain, there are many options:

- Self-monitoring** can be helpful to keep track of pain, both for yourself and your care team, and to recognize what makes it worse and what helps. You can use the Self-Assessment Guide for monitoring.
- Self-management approaches** include relaxation strategies and deep breathing exercises to reduce muscle tension and improve mood and anxiety. These can help improve pain, well-being, and feelings of self-mastery and personal control.
- Medical therapies** include many approaches your care team may prescribe, most commonly drug therapy.
- Psychosocial approaches** include numerous strategies to reduce stress, improve coping, and enhance the ability to complete activities.
- Alternative medicine approaches** combine self-management, psychosocial approaches, and medical therapies.

## What Can I Expect if I Use Highly Effective CFTR Modulators?

We are still learning about CFTR modulator effects on pain and other symptoms. Pain may improve based on your response to the medication. For example, chest and lung pain may reduce because you have fewer exacerbations as a result of a CFTR modulator. Better sinus health may lessen headaches and sinus pain. Some people experience changes in bowel habits, mood, or anxiety. Report all changes to your doctor to determine if any medication changes may be needed.

## Examples of Treatments Used to Manage Acute or Chronic Pain in CF

- Psychosocial approaches** aim to promote relaxation. Pain can cause muscles to tense. People in pain may hold their bodies in awkward positions to protect themselves from feeling pain without even noticing. Stress and anxiety also lead to muscle tension. Two commonly used psychosocial approaches are passive and progressive muscle relaxation. Among the relaxation techniques that are easily learned are deep breathing and guided imagery.

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## Relaxation Approaches

### • Deep breathing

- Make yourself comfortable.
- Breathe in through your nose (or mouth if your nose is congested).
- Lay your hands over your abdomen (**not your chest**) and feel your hands gently rise and fall.
- Relax your abdominal muscles.
- Take slow, deep breaths.
- As you gently breathe out, release your tensions and feel these flow out.

### • Guided imagery

- Find a quiet place to yourself.
- Comfortably pace your breathing—slowly and deeply.
- Close your eyes and imagine a peaceful or happy scene in which you feel relaxed and safe. Include the sounds, smells, and colors.
- Repeat this every day for 5 minutes.

Progressive muscle relaxation also can be learned.

## Progressive Muscle Relaxation

Progressive muscle relaxation involves gradually tensing, and then relaxing one area of the body at a time, eventually bringing relaxation to your entire body. This involves tensing groups of muscles, then releasing the tension and focusing on the relaxation you experience. Do not tense to the point of feeling pain, and be careful about tensing areas where you may have an injury.

### Preparation:

Find a comfortable, quiet place where you will not be disturbed for 15-20 minutes. Sit or lie down, wear comfortable clothes, and make sure you are comfortably warm before starting the technique.

### Instructions:

1. Focus your attention on one part of the body at a time.
2. Breathe in as you tense the muscles of that body part.
3. Hold the tension for 5-7 seconds.



4. Release the tension and relax the muscles.



5. Focus on the muscles relaxing for 10-20 seconds before moving to the next body part.
6. You can start by tensing and relaxing your facial muscles, followed by your neck and throat, shoulders and back, arms, hands, chest, abdomen, thighs, legs, and finally feet.

- **Movement-based approaches** can help relieve stress and anxiety while also promoting physical fitness. Examples include yoga, muscle-strengthening, stretching, aerobic exercise, and physical therapy.
- **Cognitive behavioral therapy (CBT)** for pain is a psychotherapy approach focused on developing and using a set of proven pain management strategies in collaboration with a supportive therapist. CBT can reduce pain, distress, and pain's interference with activities. It is also a highly effective treatment for depression, anxiety, and sleep problems, which often co-occur with pain.
- **Medical treatments for pain** are numerous. These treatments must be individualized to meet your needs and monitored over time for safety and effectiveness.
  - Some over-the-counter medical treatments for pain include acetaminophen (Tylenol®) and NSAIDS (non-steroidal anti-inflammatory drugs, such as ibuprofen).
  - Non-opioid prescribed therapies that can help with pain include drugs in many classes, such as antidepressants, anticonvulsants (such as gabapentin or pregabalin), and topical therapies.
  - Opioid therapies, such as morphine, are often used to treat acute pain and rarely used to manage chronic pain.
  - Medical cannabis also may help treat pain. Be aware that its use is subject to state regulations and federal limitations.

## What Else May Help?

It is helpful to try 'pacing' or balancing your activities with rest. With chronic pain, it's important to stay active and engaged in life, but pacing is key. Both under- and over-activity can increase pain. Try to break down activities by time (rather than task completion) and schedule periodic breaks. Increase the time involved in activity gradually to increase your success and build stamina.

Reach out for support from family, friends, your CF care team, a therapist, or the CF community. One source of support can be the CF Foundation Peer Connect, which is a one-to-one peer mentoring program for adults with CF to share their experiences with each other. Visit the following site for more information: [cfpeerconnect.com](https://cfpeerconnect.com).