Patient Education Series

Dyspnea and Cough

Dyspnea is the medical term for difficulty breathing, or an abnormal shortness of breath. It is not the shortness of breath that occurs after vigorous exercise, but rather, is a symptom that may occur out of proportion to activity or even when a person is at rest. Dyspnea may be associated with chest tightness or difficulty catching one's breath. It may or may not be accompanied by cough or wheezing.

Cough, like shortness of breath, is a normal phenomenon, but can be abnormal when it is very frequent or persistent, or easily triggered. When cough occurs, it may be a dry cough or associated with secretions.

Both dyspnea and cough can be very distressing symptoms in people with cystic fibrosis (CF). They may worsen anxiety, make it difficult to work or attend school, interfere with relationships, cause social embarrassment, and prevent a good quality of life. Some people with CF have dyspnea or cough that interferes with sleep, which may cause fatigue.

What Can Cause Dyspnea and Cough in CF?

Shortness of breath and cough are both respiratory symptoms and may be due directly to the CF or its complications, or to other respiratory disorders separate from CF. For example:

- Lungs affected by CF tend to produce thick mucus that make it difficult to breathe or trigger cough as the body tries to expel the secretions.
- Patients with CF are predisposed to get bacteria in the lungs, which may be the cause of pulmonary exacerbations requiring antibiotics. Viral infections, such as influenza and coronavirus, also could occur and cause acute breathing problems and severe cough.
- Some people with CF have seasonal allergies or sensitivity to inhaled pollutants, vaping, or secondhand smoke; sometimes cough is due to drainage from the sinuses or acid reflux.
- Other common disorders such as asthma may occur in people with CF and increase the likelihood of dyspnea or cough. Some people experience these symptoms as a result of anxiety or stress.

What Can I Do to Help?

When troubling dyspnea or cough occurs, it is important to tell your CF care team so that the symptoms can be investigated if needed, and medical therapies can be started.

The CF team usually emphasizes the importance of interventions that should be done routinely to manage the CF lung disease and reduce the likelihood that dyspnea or cough will become a problem. These interventions, some of which are specifically prescribed by your CF provider, include:

- Airway clearance therapies, such as manual or VEST chest physiotherapy or positive expiratory pressure devices (Aerobika® or flutter device)
- Nebulized therapies, such as albuterol and mucus-thinning agents
- Using a pulse oximeter to check your oxygenation, or a home spirometer device to check your lung function, so that you have information available when you contact your CF provider.

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- An exercise program to condition your muscles, like resistance training, including weightlifting and resistance bands, and endurance training, such as walking, jogging, or cycling. Good posture is important, and a physical therapist can evaluate and teach you exercises to help you stretch and improve your posture. At times, your CF care team will consider a specific pulmonary rehabilitation program.

Some of the approaches recommended by your CF care team may be learned and used by yourself when needed.

- Pursed-lip breathing helps to reduce the work of breathing. Try this when exercising, climbing stairs, bending, or lifting.

Pursed Lip Breathing Approach

1. Relax your neck and shoulder muscles. Breathe in slowly through your nose for 2 seconds.
2. Purse your lips as if you are about to whistle or blow out a candle.
3. Breathe out gently through your lips **for at least 4 seconds**.
   Always breathe out for longer than you breathe in. This allows your lungs to empty as much as possible.

- The Calming Hand Breathing Approach can help you relax and recover from an episode of breathlessness.

The Calming Hand Breathing Approach

The Calming Hand Breathing Approach incorporates five strategies while focusing on the five fingers of one's hand. Practice it daily to improve overall wellbeing, or repeat as many times as necessary until a breathlessness episode passes.

Thumb:	**Recognize** — Squeeze your thumb as you recognize that this episode will pass.
Pointer finger:	**Sigh Out** — Let your shoulders drop down and exhale out.
Middle finger:	**Inhale Gently** — Take a slow and gentle breath in through your nose.
Ring finger:	**Exhale Slowly** — Slowly breathe out.
Pinky finger:	**Stretch Hand and Relax** — Gently stretch your hand out, which can help you relax.

- Cognitive and behavioral strategies can help you cope and manage distressing symptoms on your own—at home, work, or school. These strategies usually are taught by a mental health professional. They may include:
  - Focus on relaxing; take a moment to stop, sit, or lie down and rest.
  - Yoga and meditation can help you remain calm and focus on your breathing.
  - Using a fan to create a breeze on your face can help.
  - Consider whether you can reduce or remove environmental triggers, such as secondhand smoke and things in the environment that trigger allergy.

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  - An additional resource is 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.

What Can My CF Care Team Do to Help?

Your CF care team can examine your symptoms to see if you need immediate care. They may order tests to determine causes of shortness of breath and cough. The results may determine the best treatments for you to feel better. Some tests include:

- Pulmonary Function Test (PFT) to check your lung function
- Oxygen saturation measurement at rest, walking, or while sleeping if indicated
- Blood-gas test to measure your carbon dioxide level
- Sputum culture to monitor for new bacteria or changes in old bacteria—these changes may determine the most appropriate antibiotics to treat you
- Chest X-ray or CAT scan of your lungs
- Bronchoscopy (a test to inspect your airways and sample secretions while under anesthesia)
- Viral swab to check for viral illness such as influenza or COVID-19

What Are Some Medical Treatments?

Your CF care team may suggest medical therapies to treat the underlying causes of dyspnea or cough:

- If you have asthma, new inhalers may help. People with severe asthma may require monthly injections of biologic therapies to control their symptoms.
- A new airway clearance regimen may be recommended to help reduce secretions in the airways.
- Inhaled or systemic antibiotics will be prescribed if an infection is suspected.
- Sinus treatments, sometimes based on the recommendations of an Ear-Nose-Throat (ENT) doctor, may focus on sinus discharge as a cause of cough or dyspnea.
- Acid reflux treatments may be suggested if this is suspected to be a cause of cough.
- Seasonal allergies can be addressed with new allergy medications.
- Occasionally, when dyspnea is severe, your CF provider will prescribe oxygen therapy.
- Rarely, severe dyspnea is treated with an opioid therapy.

What Should I Tell My CF Care Team at My Office Visit?

Describe your symptoms and possible triggers. *How long do they last? What time of day do they occur? What are you doing when they happen? What have you tried to help the symptoms?*

Call your CF team for significant, sudden changes in your symptoms. Additional concerning symptoms include: shortness of breath associated with chest pain, fever, lightheadedness, or lip color changes. Cough-related symptoms that include bloody sputum and cough that is associated with fever or shortness of breath are of particular concern.

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

CFTR modulators have been associated with a reduction in dyspnea and cough, less sputum production over time, and both improved lung function and fewer pulmonary exacerbations.

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