The 1s, 2s, and 3s of Bronchiectasis
The COPD Foundation’s mission is to prevent and cure chronic obstructive pulmonary disease and to improve the lives of all people affected by COPD.

The Bronchiectasis and NTM Initiative aims to meet the unmet needs of the bronchiectasis and nontuberculous mycobacteria (NTM) communities. The Initiative offers a global online platform for community members to connect and interact, educational resources for patients, and research programs meant to assist in accelerating therapeutic solutions and cures for both, bronchiectasis and NTM.

The 1s, 2s, and 3s of Bronchiectasis was created by the COPD Foundation.

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Learning about bronchiectasis (brawn-key-eck-tay-sis)

What is bronchiectasis?

Bronchiectasis is a chronic lung disease. In this disease, the tubes (airways/bronchial tubes) that carry air to the lung(s) become widened, scarred, and inflamed. This can make it difficult to breathe. Also, the tiny hair-like structures (cilia) in the lung(s) that would normally help move mucus in healthy lungs, don’t function as well with bronchiectasis. These changes in the airways can cause mucus to pool in the lung, allowing germs to grow, resulting in an increased number of lung infections. The collection of mucus in the lung can also lead to cough with large amounts of mucus production.

Image courtesy of NIH/NHLBI
**Causes of bronchiectasis**

The most common cause of bronchiectasis is repeated or chronic lung infection. Infection causes two out of five cases of bronchiectasis. However, in up to half the cases of bronchiectasis, doctors are unable to determine the cause. This is referred to as ‘idiopathic’ (id-ee-oh-pa-thick) bronchiectasis. Repeat, uncontrolled infections may lead to more advanced bronchiectasis over time.

**Severe lung infections:** Infections that cause bronchiectasis include: pneumonia, nontuberculous mycobacterial (NTM) infection, influenza, tuberculosis, whooping cough, and measles. At the time of infection, the airways become inflamed and damaged, which can lead to bronchiectasis.

**Injury:** Lungs can become injured by food and liquids “going down the wrong tube.” This is called aspiration. When this happens often over a period of months or years, the food and liquid can cause inflammation and damage to the airways that can result in bronchiectasis. Other injuries to the lungs, such as smoke inhalation, can also lead to bronchiectasis.

**Immune deficiencies (im-yoon dee-fi-shin-sees):** When your immune system is weak, you are more likely to get lung infections that can lead to bronchiectasis. Examples of these deficiencies include Human Immunodeficiency Virus/Acquired Immune Deficiency Syndrome (HIV/AIDS) and immunoglobulin deficiency (lack of proteins your body makes to fight infection).

**Inflammatory diseases:** Some diseases that cause inflammation throughout the body can also cause inflammation in the airways and lead to bronchiectasis. Examples of these diseases include ulcerative colitis, Crohn’s disease, rheumatoid arthritis and Sjögren’s syndrome.
Inherited (genetic) disorders: Some people are born with genetic variants that lead to bronchiectasis. These include cystic fibrosis and primary ciliary dyskinesia (dis-kin-ee-sha). In these diseases, there are problems with mucus clearance that lead to bronchiectasis.

**Symptoms**

The main symptom of bronchiectasis is cough with varying amounts of mucus production. Many people have symptoms for months, even years, before a diagnosis is made. This is especially true for people who have mild symptoms that come on slowly.

**Cough:** Nearly all individuals with significant bronchiectasis cough every day for months, and sometimes, years. Some individuals may also cough up blood. Coughing up blood is referred to as hemoptysis (hee-mop-tih-sis).

**Sputum production:** Most people with bronchiectasis cough up mucus (sputum) every day. Some people with bronchiectasis may cough up nearly a cup of mucus a day. Not many people with bronchiectasis cough up little or no mucus.

**Shortness of breath with exertion:** In bronchiectasis the airways become widened, scarred, and inflamed. This can lead to difficulty breathing while walking, climbing stairs, or doing other activities.

**Frequent lung infections:** People with bronchiectasis get more lung infections because mucus pools in the lungs. When this happens, bacteria stays in the lungs instead of being coughed out. Repeat pneumonias in the same part of the lung are common in bronchiectasis.

**Tiredness (fatigue):** This is also very common among individuals with bronchiectasis and can limit daily activities.

**Chest pains:** People with bronchiectasis may experience chest pains that get worse when taking deep breaths.
Bronchiectasis and NTM Facts

- Among all ages it has been estimated that about 25 people per 100,000 have bronchiectasis.
- For those over 74 years old, this number increases to 272 per 100,000.
- Over half the cases of bronchiectasis have no known cause or association.
- It is estimated that 30-35% of bronchiectasis cases follow a lung infection that damages the airways for the first time.
- In 2001, it was estimated that the annual medical cost of care per person with bronchiectasis in the U.S. was greater than that of the person with heart disease or Chronic Obstructive Pulmonary Disease (COPD).

**Getting tested**

Medical tests help determine if you have bronchiectasis and, if you do, what caused it. X-ray tests are the most commonly used tests to diagnose and/or confirm bronchiectasis. A common type of x-ray test is a computed tomography (CT) scan of your chest. Another common test for detecting bronchiectasis is the pulmonary function test (PFT).

**Chest CT**

A CT scan of the chest is a special kind of x-ray that shows a very detailed picture of your lungs. Your doctor can see the widening of the airways on this type of scan and know how much of your lungs are affected.
What should I do to prepare for a chest CT scan and what should I expect?

• Wear comfortable, loose-fitting clothing.
• You may be allowed to keep your clothes on or take something off and wear a gown. You will be asked to remove metal objects such as necklaces and glasses.
• The CT scanner looks like a huge donut.
• You will be lying on a movable exam table with a padded surface, shaped to keep you from falling.
• The technician will help you get into the position that will provide the clearest images.
• You will most likely be lying on your back.
• The table will move you through the donut hole to determine the correct starting position for the scans. Then it will move again through the donut hole to take the actual scans.
• You may be asked to hold your breath during scanning.
• The actual CT scanning takes less than 30 seconds and the entire process is usually completed within 30 minutes.

Pulmonary Function Test (PFT)

A pulmonary function test (PFT) is a breathing test that measures how much air moves in and out of your lungs and how fast it moves. This test helps your doctor understand how well your lungs are working. It may also help determine why you have shortness of breath.
What should I do to prepare for a PFT and what should I expect?

- You will be asked to sit in a chair near the testing machine or on a seat in a clear glass booth – something like a phone booth.
- Before starting, the technician will place a soft clip on your nose.
- You will breathe through your mouth into a tube to perform the testing.
- The technician will show you and coach you on how to breathe.
- For some parts of the test, you will be asked to breathe normally. For other parts of the test, you may be asked to breathe fast and deep.
- For some people, breathing fast and deep makes them cough, feel dizzy, or feel short of breath. Don’t worry. Tell the technician what you feel and he or she will give you time to rest and try again.

Sputum samples
The mucus you cough up may be tested for germs, including bacteria and fungus. You will be asked to cough up mucus into a special container. This sample will be sent to the laboratory to determine which germs will grow (culture) in the mucus.

Blood tests
Blood tests may be ordered to determine why you have bronchiectasis. This may include immunodeficiency, inflammatory, and genetic testing.
Living with bronchiectasis

Treatments

Although bronchiectasis cannot be cured, there are effective treatments. Your treatment plan depends on the severity of your bronchiectasis symptoms as well as the cause of your bronchiectasis. Almost all people with bronchiectasis should use some method of airway clearance every day.

Airway clearance: Airway clearance is an essential part of treatment for bronchiectasis. Airway clearance helps remove the mucus that has pooled in the airways. The goal is to loosen this mucus so you can cough it up more easily. There are several ways to do this.

There are many methods of airway clearance. It is critical to find the method that works best for you. This is usually done through trial and error.

Handheld device: Your doctor may prescribe a handheld positive expiratory pressure (PEP) device that causes vibrations inside your airways when you exhale. These vibrations shake mucus loose so you can cough it out. Each treatment with a handheld device should take 5-15 minutes. Therapy may be repeated several times a day.
**Percussive vest:** A percussive vest is connected to a machine that inflates and deflates it quickly. This causes vibrations in the chest that help loosen mucus.

**Inhaled medications:**
Your doctor may prescribe inhaled medications to help with your airway clearance. Hypertonic saline is a sterile salt solution that is inhaled through a nebulizer. It helps to thin the mucus so it is easier to cough up. Nebulizer treatments with a bronchodilator (brawn-coe-die-lay-ter) might also be prescribed along with airway clearance. Nebulizers are sometimes preferred over handheld inhalers/metered-dose inhalers (MDIs) because the aerosol adds moisture and because this method of treatment gives you more time to inhale the medication.

**Oral antibiotics:** Oral medications are pills or liquids you put in your mouth and swallow. Depending on the cause of your bronchiectasis, antibiotics may be used. The goal is to get rid of the infection that is causing a flare-up of bronchiectasis. Some antibiotics, such as azithromycin, are also used regularly for their anti-inflammatory effects.

**Inhaled antibiotics:** In some cases, antibiotics inhaled via nebulizer can be used to help decrease the growth of bacteria in the lungs. Inhalation delivers the antibiotic directly to the airways where it is needed. In some cases, this results in fewer side effects than when swallowing antibiotics as a pill or liquid.
**Intravenous (IV) Antibiotics:** IV antibiotics are used in more severe situations, such as during an exacerbation that has not gotten better with oral antibiotics. Also, IV antibiotics are used to treat certain bacteria that are resistant to oral antibiotics.

**Oxygen:** In some cases of bronchiectasis, supplemental oxygen may be required when blood oxygen levels are low.

**Surgery:** Lung resection (removal of part of the lung) may be performed in some cases of bronchiectasis where it is found in only one part of the lung.

**Recognizing Flare-ups – Exacerbations**

Sometimes your lung symptoms suddenly get worse. These “flare-ups” of your bronchiectasis are called “exacerbations” (x-saa-cer-bay-shuns). They are often caused by an infection.

### Warning Signs of an Exacerbation
- Change in color, thickness, odor, or amount of mucus
- Increased coughing
- Increased shortness of breath
- Increased tiredness that lasts more than one day
- Low grade fever that doesn’t go away
- Decrease in lung function by at least 10%
- Increased use of rescue/fast-acting medications

**These symptoms call for immediate action! Ask your doctor now what you should do if you see these signs:**
- Coughing up blood
- Blue color in lips or fingers
- New or increased ankle swelling
- Chest pain
It’s important to watch for early warning signs of an exacerbation and contact your doctor right away when they occur. Delaying treatment may result in a more serious exacerbation and a longer recovery time or only partial improvement. Your doctor may prescribe antibiotics and increase how often you do airway clearance. Sometimes a more serious exacerbation needs to be treated in the hospital with IV antibiotics or steroids.

In addition to exacerbations, other factors such as sinus infections and gastroesophageal reflux disease (GERD) may make your bronchiectasis symptoms worse. It is important to call or visit your doctor if you suspect that something is causing your usual symptoms to be worse.

**Making changes**

Living with a chronic lung disease such as bronchiectasis can be challenging. Performing airway clearance and taking your medications goes a long way toward improving your breathing. Making changes in your lifestyle can also help. Here are some examples of how making changes can improve your breathing.

**Exercise:** Exercise can help clear mucus from the airways and can improve quality of life for those with bronchiectasis. Your doctor might encourage you to do some exercise each day, such as walking, bicycling, swimming, or yoga. Pulmonary rehabilitation, an exercise program for people with chronic lung disease, may also be an option.

**Do not smoke:** Smoking, or being around cigarette smoke, can make your symptoms worse. If you smoke, you should stop smoking. You should also ask others not to smoke around you.

**Vaccinations:** Get a flu shot every year and talk to your doctor about a pneumonia shot. These vaccinations will help prevent common lung infections.
Eat to maintain a healthy weight: If you are overweight, losing weight can help with your breathing. If you are underweight, gaining weight can help improve your strength and help your body recover from flare-ups.

Schedule and keep appointments with your doctor: See your doctor on a regular basis even if you are feeling well. Your doctor will want to know if you have any symptoms such as cough, shortness of breath, or chest pain while doing activities. Your doctor will also want to know if you have any side effects from your medications. Take notes in between appointments and share them with your doctor.

Keep track of your treatments: Keep a list of the medications you are taking and any medication allergies or other allergies you have. Keep this list in your wallet and take it with you whenever you go out.

Keep important numbers handy: Make sure you have a list of phone numbers, including your doctor’s office and answering service as well as friends who can help you in an emergency.

Learn all you can about your disease: Join a bronchiectasis support group, in person or online.
Taking action for bronchiectasis

About the COPD Foundation and the Bronchiectasis and NTM Initiative

The COPD Foundation is a not-for-profit organization that began in 2004. The COPD Foundation supports research and awareness about obstructive lung diseases such as bronchiectasis. Bronchiectasis and COPD are two different diseases, but some people may have both bronchiectasis and COPD. The COPD Foundation’s Bronchiectasis and NTM Initiative aims to meet the needs of the bronchiectasis and NTM communities. It includes: BronchandNTM360social, educational materials, and research programs such as the Bronchiectasis and NTM Research Registry. For more information visit www.BronchiectasisandNTMInitiative.org.

**BronchandNTM360social:** This is an online community for individuals affected by bronchiectasis and/or NTM. It has resources for patients, family members, caregivers, and healthcare providers. It is a place to share thoughts and ideas, ask questions, and communicate with peers as well as experts. To join this community or to look around go to www.BronchandNTM360social.org.

**Bronchiectasis and NTM Research Registry:** The Bronchiectasis and NTM Research Registry is a database of individuals with non-cystic fibrosis bronchiectasis and/or NTM from all over the United States. The goal of the registry is to support collaborative research and assist in planning multi-center trials for the treatment of these diseases. To learn more, visit: www.BronchiectasisandNTMInitiative.org.
**How to get involved**

The COPD Foundation welcomes all types of support!

**Participate in research:** Research into the best ways to treat bronchiectasis is ongoing. There are clinical trials as well as survey opportunities. To learn more about research opportunities, visit www.BronchiectasisandNTMInitiative.org.

**Join the conversation:** Connect and communicate with other individuals affected by bronchiectasis and/or NTM by joining www.BronchandNTM360social.org.

**Make a donation:** You can support the COPD Foundation’s Bronchiectasis and NTM Initiative by making a tax-deductible donation. You can donate online at www.BronchiectasisandNTMInitiative.org by clicking the “Donate Now” button. To donate by mail, please mail your check or money order to:

**COPD Foundation**  
3300 Ponce de Leon Blvd.  
Miami, FL 33134
Support organization and resources

**NTM Info & Research:** An organization formed on behalf of patients with NTM disease for the purpose of patient support, medical education, and research. NTM Info & Research has many resources for individuals affected with NTM lung disease. To learn more, visit www.NTMinfo.org.
GLOSSARY

**Airways** – The tubes that carry air through the lungs. In bronchiectasis, they become inflamed and are widened and thickened.

**Airway clearance** – Removing excess mucus from the lungs. This may include using a handheld device that vibrates the airways and lungs or using a vest that vibrates the chest.

**Antibiotics** – Medications that kill bacteria.

**Aspiration** – When food or liquids are accidentally inhaled into the lungs. They have “gone down the wrong tube.”

**Crohn’s disease** – An inflammatory disease that can affect any part of the gastrointestinal tract including the mouth, esophagus, stomach, small intestine, and large intestine.

**CT scan or “CAT” scan** – CT (computerized tomography) scan combines a series of x-ray images taken from different angles to create a detailed picture. It is used to make a diagnosis of bronchiectasis.

**Cystic fibrosis** – A genetically inherited cause of bronchiectasis causing thick mucus to build up in the lungs, pancreas, and other organs.

**Exacerbation** – A flare-up of chronic disease symptoms. In bronchiectasis, this may mean an increase in shortness of breath, cough, and mucus production.

**HIV/AIDS** – Human Immunodeficiency Virus/Acquired Immunodeficiency Syndrome. This virus attacks the body’s immune system. This makes people more likely to get infections, including lung infections that can result in bronchiectasis.

**Hypertonic saline** – A concentrated salt solution that can thin mucus when it is inhaled into the lungs.

**Immune deficiency** – Any disease that makes it hard for the body to fight off infection.
Immunoglobulin deficiency – In this condition, the body cannot make specialized proteins called immunoglobulins that help fight infection.

Influenza – Commonly known as the flu, a viral infection that causes fever, cough, sore throat, and runny or stuffy nose. It can be complicated by pneumonia (a lung infection) that can lead to bronchiectasis.

Measles – A viral infection that causes rash, fever, cough, and runny nose. It can lead to pneumonia and bronchiectasis.

Mucus – A slimy substance, often called sputum or phlegm.

Nebulizer – A machine that creates a fine mist that can be inhaled.

Nontuberculous mycobacteria (NTM) – Organisms found in soil and water. When a susceptible person inhales these organisms, that person can become infected and this can lead to bronchiectasis.

Percussive vest – An inflatable vest that creates vibrations in the chest to help loosen mucus so that it can more easily be coughed out.

Pneumonia – An infection of the air sacs of the lungs.

Pulmonary function testing – A breathing test that measures how well the lungs take in (inhale) and get rid of (exhale) air. It can also measure how easy it is for oxygen to move from the lungs into the blood.

Pulmonary rehabilitation – A program of exercise, education, and support to help individuals with chronic lung disease be as active, healthy, and independent as possible, for as long as possible.

Primary ciliary dyskinesia – An inherited disorder of the cilia, the hair-like structures that help move mucus out of the lung. In this disease, the cilia do not work properly and mucus pools in the lungs, leading to bronchiectasis.

Rheumatoid arthritis – A chronic inflammatory disease that affects the joints. It can also cause inflammation in the lungs which can lead to bronchiectasis.

Sjögren’s syndrome – A chronic inflammatory disease that can cause dry eyes and dry mouth. It can also cause inflammation in the lungs which can lead to bronchiectasis.
Smoke inhalation – Occurs when breathing in large amounts of environmental smoke. Both smoke and heat can cause damage to the airways, which can lead to bronchiectasis.

Sputum – Mucus or phlegm that is coughed up from the lungs.

Ulcerative colitis – A chronic inflammatory disease of the large intestine. It can cause inflammation in the lung, which can lead to bronchiectasis.

Vaccination – An injection or “shot” that protects the body from getting an infection such as the flu or pneumonia.

Whooping cough – A respiratory tract infection that can lead to bronchiectasis. Most people are protected from it by vaccinations.