

Bronchiectasis

What is Bronchiectasis?

Bronchiectasis (pronounced bron-kee-ek'-tas-is) is a condition of the airways in the lungs. These airways (bronchial tubes) are tube-like structures that branch from the trachea into the right and left lungs. When a person has bronchiectasis the airways are permanently and abnormally widened (dilated) and inflamed. These damaged airways can no longer clear mucus and bacteria from the lung, so that exacerbations of cough, sputum production and shortness of breath can occur.

Bronchiectasis is caused by one or more infectious or inflammatory insults to the lungs. People with bronchiectasis are more likely to get lung infections. Each lung infection can make the bronchiectasis worse.

Generally the prognosis of bronchiectasis is very good. The earlier it is diagnosed, the earlier treatment and management can be initiated preventing it from worsening. Be proactive with your doctor. Remember that the gold standard to make a diagnosis of bronchiectasis is now a high resolution CT scan of the lungs and NOT a simple chest xray.



How Can You Develop Bronchiectasis?

There are many causes of bronchiectasis — some are acquired and others you may be born with (congenital). The following is a list of the most important and/or common causes.

Infections

Infections can damage the airways and cause bronchiectasis. They may also be a consequence which in turn can lead to worsening bronchiectasis.

Examples include:

- Viral infections (measles, adenovirus, influenza)
- Bacterial infections (*Pseudomonas aeruginosa*, *Staphylococcus aureus*, *Klebsiella*)
- Mycobacterial infections (tuberculosis, *Mycobacterium avium*, *Mycobacterium abscessus*) and
- Fungal infections (histoplasmosis)

Immune Diseases

People with immune deficiencies such as antibody deficiencies are more likely to have repeated lung infections which can damage the airways and cause bronchiectasis.

Aspiration

Chronic pulmonary aspiration occurs when a person inhales oral or stomach material into his/her lungs. If severe or recurrent, aspiration can lead to inflammation of the airways and causes bronchiectasis. The aspiration can occur from:

- Impaired ability to swallow (oropharyngeal dysphagia) which may cause saliva or food to enter the lung.
- Gastroesophageal reflux disease (GERD) which occurs when the valve of smooth muscle between the esophagus and the stomach does not function properly. This allows contents (acid and non-acid) to flow back up into the esophagus. The stomach contents may enter the lungs and irritate the airways. Some signs and symptoms of GERD include: heartburn or sour taste in mouth, but many (possibly up to 30 percent) of people with GERD may have no symptoms ("silent" GERD).

Autoimmune Diseases

Rheumatoid arthritis, lupus, Sjogren's syndrome and Wegener's granulomatosis are examples of rheumatologic, autoimmune or connective tissue diseases that can cause bronchiectasis.

Genetic Diseases

- Cystic fibrosis causes impaired drainage of mucus and bacteria from the airways. This leads to recurrent lung infection and bronchiectasis. Classic cystic fibrosis is obvious at birth, but there are forms of cystic fibrosis that may not be recognized until adulthood.
- Primary ciliary dyskinesia impairs the ability of small hairs, called cilia, to clear mucus and bacteria from the airways. Recurrent lung infections can occur and cause bronchiectasis.
- Alpha1 antitrypsin deficiency. Alpha₁ antitrypsin is a protein that moderates inflammation that occurs during infection. People who are deficient in alpha₁ antitrypsin or who have an abnormal protein may be more likely to have recurrent lung infections that cause bronchiectasis.

Obstruction of the Airways

Obstructed airways trap mucus and infections behind the obstruction which can damage the airways and cause bronchiectasis.

- Obstruction of the airways can be caused from a growth or tumor.
- Chronic obstructive pulmonary disease (COPD) and allergic bronchopulmonary aspergillosis are diseases that can cause obstruction of the airways.

What Happens in the Lungs with Bronchiectasis?

First, inflammation occurs in the walls of the airways from a number of causes just discussed. This inflammation causes injury to the airways. The resulting loss of the normal defenses in the lungs leads to impaired drainage of the airways. This makes the airways susceptible to infections. Repeated lung infections can worsen the damage to the airway walls.

What are the Symptoms?

Symptoms of bronchiectasis include a cough. The cough may be productive of mucus. With infections the mucus may be discolored, foul-smelling and may contain blood (hemoptysis). Shortness of breath, wheezing, weight loss and fatigue can also occur. Some people with bronchiectasis also have chronic sinusitis. This requires further evaluation since bronchiectasis and sinusitis may be due to the same underlying disease.

If left untreated, symptoms of bronchiectasis may progress. Further symptoms may include increasing shortness of breath, worsening quality of life and even heart failure.

How is Bronchiectasis Diagnosed?

A multiple step process usually leads to the diagnosis of bronchiectasis. Many factors are considered and different tests are completed. The evaluation for bronchiectasis often includes:

- A complete medical history and physical examination by a health care provider.
- A chest CT scan (a specialized X-ray which produces detailed slice-like pictures) of the lungs.
- Breathing tests, called pulmonary function tests. These determine the presence and severity of abnormal airflow out of the lungs.
- Specific screening or diagnostic tests for some of the possible underlying diseases that may cause bronchiectasis, based on the history and physical examination.

How is Bronchiectasis Managed?

Bronchiectasis management is long-term and is directed at:

- Improving the clearance of sputum, also called bronchopulmonary hygiene
- Treatment of infections
- Treatment of associated conditions (such as GERD and sinusitis)
- Improving muscle strength and endurance through pulmonary rehabilitation
- Identifying the need for surgical resection of affected segments or lobes of the lung

Your health care provider will evaluate your history and recommend the best management plan for you.

Bronchopulmonary Hygiene Therapy

Improved clearance of mucus is the cornerstone of the management of bronchiectasis and includes several components. They include:

- Inhaled medication (bronchodilator and/or inhaled steroid, saline)
- Airway clearance measures (oscillating positive expiratory pressure device, high-frequency chest wall oscillation vest).

Your health care provider may recommend one or more of them depending on your individual needs.

Inhaled Medication

Inhaled Bronchodilators - An inhaled bronchodilator medication opens the airways by relaxing the smooth muscles around the airways. This type of medication is available in a number of inhaled forms. Commonly used inhaled short-acting bronchodilators include:

- ProAir®, Proventil® HFA, Ventolin® HFA (albuterol)
- Xopenex® (levalbuterol)

Inhaled long-acting bronchodilators may also be used. They include:

- Serevent® (salmeterol)
- Foradil® (formoterol)
- Spiriva® (tiotropium)

Inhaled Steroids – Inhaled steroids reduce and prevent swelling inside the airways. Common inhaled steroids include:

- Flovent® (fluticasone)
- Pulmicort® (budesonide)
- QVAR® (becolmethasone)
- Asmanex® (mometasone)
- Azmacort® (triamcinolone)

- Aerobid® (flunisolide)

Inhaled Steroid and Long-Acting Bronchodilator Combinations

Common combinations of inhaled steroid and long-acting bronchodilator include:

- Advair® (fluticasone and salmeterol)
- Dulera® (mometasone and formoterol)
- Symbicort® (budesonide and formoterol)

Inhaled hypertonic saline may be used to loosen airway mucus for easier airway clearance.

Airway Clearance Measures

Airway clearance measures are treatments designed to clear trapped mucus from the airways.

- Oscillating positive expiratory pressure devices (OPEPD): These include devices such as the Aerobika®, Acapella® or the Flutter Valve® that help clear mucus from your lungs. These are small devices you inhale and/or exhale into.
- High-frequency chest wall oscillation vests: These include The Vest®, SmartVest® and AffloVest® are inflatable vests that you put on. The vest shakes your chest to help dislodge the mucus from the airway walls. Sometimes the Aerobika® or Acapella® is used **after** the inflatable vest. Once the mucus is dislodged, the device can help clear the mucus.
- Postural drainage and clapping use gravity to promote drainage of mucus from the lungs.



Each technique can be prescribed by your health care provider.

Correct technique using these devices is very important. Make sure a health care provider, often a Respiratory Therapist, experienced in the use of the device shows you how to use it. It is also important to have your technique checked periodically to make sure you continue to use it correctly to obtain the most benefit.

Treating Infections

Antibiotics are used to treat bacteria and other infectious organisms causing infection in the lungs to improve respiratory symptoms and prevent further damage to the airways. For example, treating *pseudomonas aeruginosa* may entail 2-3 weeks of intravenous antibiotics when symptoms are severe. Sometimes inhaled antibiotics are given to prevent exacerbations of pseudomonas. Treatment of mycobacteria may require multiple antibiotics taken for a year or longer. Rotating or chronic antibiotics to prevent infections are not encouraged because this promotes the development of drug-resistant organisms. However, long-term azithromycin may sometimes be beneficial for people who experience frequent bronchiectasis flare-ups.

Treatment of Associated Conditions

Treatment of any identified specific causes, including those listed under “Causes of Bronchiectasis” is important. Examples include:

- Treatment of chronic infections with non-tuberculous mycobacteria,
- Treatment of antibody deficiency with immune globulin if appropriate,
- Treatment of swallowing disorders and GERD that cause chronic pulmonary aspiration. The MedFacts, Gastroesophageal Reflux Disease, discusses this topic in more detail.
- Prompt treatment or removal of any foreign object, growth or tumor causing obstruction of the airways,
- Treatment of other chronic lung disease
- Treatment of chronic sinusitis. The MedFacts, Sinusitis, discusses this topic in more detail.

Pulmonary Rehabilitation may improve your overall health. A well-rounded rehabilitation program includes education, exercise and eating well and can help you stay healthy and feel good.

Resective Surgery is occasionally indicated – usually only if bronchiectasis is very localized in the lung and medical treatment and other therapies are not effective.

What about a Healthy Lifestyle?

A healthy lifestyle is important for everyone. Here are some tips to consider:

- Exercise regularly as directed by your health care provider. This helps you breathe easier by improving your muscle strength and tone and helps improve clearing the mucus from the airways.
- Eat a well-balanced diet and drink plenty of fluids.
- Give up smoking and avoid exposure to passive smoke. Ask your health care provider for techniques to help you give up smoking.
- Get a flu shot every year in the fall. Get the pneumococcal vaccines as recommended by your health care provider.

Living with Bronchiectasis

- Living with bronchiectasis is a unique and special challenge that you and your family must deal with on a daily basis. But the more you know about bronchiectasis, the better suited you are in managing the various aspects of your disease. As you take control, your quality of life will improve.
- Be sure to talk with your health care provider if you have questions or concerns about your plan. Write down any questions you have and ask your health care provider at your next appointment.

Visit our website for more information about support groups, clinical trials and lifestyle information.

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