



# medfacts

AN EDUCATIONAL HEALTH SERIES FROM NATIONAL JEWISH HEALTH™

## Bronchiectasis

### What is Bronchiectasis?

Bronchiectasis (pronounced bron-kee-ek'-tas-is) is a condition of the airways in the lungs. These airways 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/or thickened. Mucus can collect in the abnormal airways. The mucus is difficult to clear because of the damage to the normal ways the airways clear the mucus.

Bronchiectasis is caused by one or more infectious insults to the lungs. This can lead to episodes of infection and then, worsening bronchiectasis. These insults are more likely to occur if there is impaired drainage of your airways or a defect in your immune defenses.

Early diagnosis and treatment of bronchiectasis is very important.

### 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 ones.

#### Infections

Infections can be a cause of 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* complex) and
- Fungal infections (histoplasmosis).

#### Immune Diseases

Immune diseases include:

- Antibody deficiencies (e.g., common variable immunodeficiency) and
- White blood cell dysfunctions (e.g., chronic granulomatous disease).

#### Post-inflammatory Pneumonitis

Chronic pulmonary aspiration is caused by the recurrent inhalation of oral or stomach material into your lungs. This can lead to post-inflammatory pneumonia. 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 muscle band is called the lower esophageal sphincter.

Normally the lower esophageal sphincter acts as a guard to prevent stomach contents (acid and non-acid) from backing up into the esophagus. An increase of the pressure in the stomach and/or relaxation of the muscle tone of the valve may cause reflux to occur. The stomach contents may enter your lungs and irritate the airways. Some signs and symptoms of GERD include: heartburn, sour taste in mouth, or swallowing problems, but many (possibly up to 30%) of people with GERD (some with consequent bronchiectasis) may have no symptoms (“silent” GERD).

#### Autoimmune Diseases

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

#### Impaired drainage of the Airways

- Impaired drainage of secretions can be due to cystic fibrosis (both classical and non-classical/atypical adult variants).
- Impaired drainage can also be caused by diseases that affect the function of the cilia. The cilia are small hairs that line the airways and move together to clear secretions from the airways. Diseases such as primary ciliary dyskinesia, Kartagener’s syndrome and Young’s syndrome all have altered cilia function.

#### Obstruction of the Airways

- 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.

#### Alpha-1 antitrypsin deficiency or anomaly

## 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?

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?

The management of bronchiectasis is long-term and is directed at:

- Improving the clearance of sputum, also called bronchopulmonary hygiene,
- Controlling infection (both prevention and treatment),
- Identifying and treating co-morbid conditions often seen with bronchiectasis (such as GERD and chronic sinusitis),
- Improving muscle strength and endurance through pulmonary rehabilitation and
- 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) and
- 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)
- Maxair® (pirbuterol)

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® (Flovent® and Serevent®)
- Symbicort® (Pulmicort® and Foradil®)

### **Airway Clearance Measures**

If you produce a large amount of mucus your health care provider may recommend techniques to help clear the mucus.

- Oscillating positive expiratory pressure devices (OPEPD): These include devices such as the Acapella® or the Flutter Valve® that help clear mucus from your lungs. These are small devices you exhale into.



- High-frequency chest wall oscillation vests: These include The Vest™ and the SmartVest® and are inflatable vests that you put on. The vest shakes your chest to help dislodge the mucus from the airway walls. Sometimes the Acapella® or the Flutter Valve® is used after the inflatable vest. Once the mucus is dislodged, the Acapella® or the Flutter Valve® 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 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.

**Treatment of Infections:** If a specific infection, such as *Pseudomonas aeruginosa*, is found to be the cause of the bronchiectasis, then antibiotics are tailored to the underlying cause. Rotating antibiotics are not encouraged because this promotes the development of drug-resistant organisms. Therefore, your health care provider will prescribe an antibiotic based on your signs, symptoms and the results of appropriate sputum cultures. For example, you may need an antibiotic

only when you experience increased shortness of breath, cough, blood in the mucus or an increase in the amount and thickness of the mucus.

## **Treatment of Co-Morbid Conditions**

**Gastroesophageal Reflux Disease** – In some people the muscle between the esophagus and stomach may not work well. This can allow some backflow of stomach contents (acid and non-acid) into the esophagus. This reflux response may lead to inflammation and then scarring with bronchiectasis. The Med Facts, Gastroesophageal Reflux Disease, discusses this topic in more detail.

**Chronic Sinusitis** – Sinusitis is an inflammation (swelling) of the mucous membranes that line the sinus cavities. This can interfere with normal sinus drainage and cause increased mucus production. Sinusitis and bronchiectasis are often associated with each other, and exacerbation of one can lead to a flare of the other. The Med Facts, 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.

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

- Treatment of chronic infections such as non-tuberculous mycobacteria,
- Treatment of immune diseases with immune globulin if appropriate,
- Treatment of swallowing disorders and GERD leading to chronic pulmonary aspiration,
- Prompt treatment or removal of any foreign object, growth or tumor causing obstruction of the airways and,
- Treatment of other chronic lung diseases.

## **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.

## **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 a pneumococcal vaccine every 5 or 6 years as recommended by your health care provider.

## **What is the role of National Jewish Health?**

Doctors at National Jewish Health have evaluated and treated bronchiectasis for decades. Currently, scientists at National Jewish Health are investigating new treatments for bronchiectasis. They are also pursuing aggressive treatment programs designed to minimize or prevent the occurrence of bronchiectasis. You may call 1-800-222-5864 to make an appointment for further evaluation of your bronchiectasis.

Note: This information is provided to you as an educational service of National Jewish Health (1-800-222-5864). It is not meant to be a substitute for consulting with your own doctor.

©Copyright 1987, Revised 1993, 2001, 2005, 2006, 2009. NATIONAL JEWISH HEALTH.

PTE.029