

# BRONCHIECTASIS

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Bronchiectasis is defined in morphological terms as irreversible dilatation and destruction of bronchial walls. René Laënnec, who invented the stethoscope in 1816, described bronchiectasis in 1819 while observing patients with tuberculosis and pneumonia. The term bronchiectasis is from the Greek words bronchion, meaning windpipe, and ektasis, which means stretched. Although bronchiectasis is regarded as a chronic obstructive lung disease, it is typically not considered to be a distinct disease. Instead, bronchiectasis is a pathologic development stemming from pulmonary infections, inflammation, obstructions, and other insults.

## Etiology & Pathogenesis

From an etiologic standpoint, bronchiectasis is classified as either congenital or acquired. Congenital bronchiectasis can be exemplified by a condition known as Kartagener's syndrome, also known as primary ciliary dyskinesia syndrome, in which the patient displays situs inversus, chronic sinusitis, and bronchiectasis. In Kartagener's syndrome the situs inversus refers to the condition in which the organs of the chest and abdomen are positioned in a mirror image of their normal locations. Situs inversus can occur with the heart positioned on either the right side of the chest, i.e., situs inversus with dextrocardia, or with the heart located in the left chest – situs inversus with levocardia. The normal arrangement of the abdominal and thoracic organs is called situs solitus.

The link between Kartagener's syndrome and bronchiectasis focuses on the dysmotility of the cilia. Initially, the belief was that

in Kartagener's syndrome the cilia were immotile. However, studies have demonstrated that the cilia are not immotile. Instead, they beat in an uncoordinated fashion, and are simply ineffective in transporting mucus. Retained secretions develop. Pulmonary infections ensue. Recurrent infections and chronic inflammation precede the development of bronchiectasis.

The chronic sinusitis associated with Kartagener's syndrome contributes to the development of bronchiectasis because the ciliated cells in the sinuses also function improperly, and cannot clear the mucus produced at these sites. Mucus from the sinuses drains into the lungs, exacerbating the secretion problem caused by the dysfunctional mucociliary escalator. The primary ciliary dyskinesia also affects spermatozoal flagella, rendering males sterile.

Cystic fibrosis is another example of congenital bronchiectasis, accounting for approximately 50% of this form of bronchiectasis. The clinical manifestations of cystic fibrosis are pulmonary or gastrointestinal, or both. Essentially, impaired release of chloride into the extracellular fluids results in the dehydration of surrounding respiratory and intestinal mucosal linings, and impaired sodium reabsorption of the sudoriferous glands. Mucosal dehydration, coupled with inflammatory and infective byproducts, create thick, tenacious mucus that clogs and damages airways. Gastrointestinal absorption of nutrients is adversely influenced for the same reasons.

Airway obstruction caused by the inability to clear viscous secretions accounts for a significant degree of the fatal progression of cystic fibrosis. Epithelial destruction throughout the tracheobronchial tree, bronchiectasis, and chronic obstructive pulmonary disease are inevitable, because of constant pulmonary insults from inflammatory oxidants. Acquired bronchiectasis can result from frequent pneumonias, especially necrotizing bacterial infections from the Klebsiella species, and from the inhalation of noxious gases and chemicals.

The entire bronchial wall becomes deranged in bronchiectasis. The structural components of the bronchial walls such as the epithelial layer, smooth muscle, cartilage, and elastic tissue are destroyed and replaced by fibrotic tissue. The respiratory epithelial lining experiences exfoliation and desquamation. Submucosal edema (inflammation) develops. Airway obstruction predisposes the patient to infection, which causes atelectasis, airway scarring, and traction on the airway walls. Pulmonary infections, in turn, weaken airway walls, leading to bronchial dilatation. The re-configuring of the bronchial walls resembles squamous metaplasia. Early peribronchial fibrosis is also present. This series and sequence of pathologic events essentially become self-perpetuating because the compromised mucociliary function leads to continued retention of secretions, which further promotes the colonization of bacteria, leading to more pulmonary infections.

Ultimately, the bronchi demonstrate permanent widening, widespread inflammatory destruction, chronic inflammation, increased mucus production, and extensive disruption of ciliary function. These pathologic alterations produce a reduction in lung volumes, lower air flow rates, ventilation-perfusion abnormalities, and, of course, hypoxemia.

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*Bronchiectasis... Continued from page 10*

Patients with bronchiectasis often develop pulmonary hypertension because the chronic hypoxemia causes the pulmonary vasoconstriction. The chronic hypoxemia also produces an increase in red blood cell production. The pulmonary vasoconstriction and the polycythemia impose greater work on the right ventricle. With these cardiovascular developments, patients with bronchiectasis can develop cor pulmonale.

From a pathophysiologic viewpoint, three types of bronchiectasis are described – cylindrical, varicose, and cystic. With cylindrical bronchiectasis the bronchi are uniformly dilated and display no tapering. Consequently, the bronchial dilatation begins and terminates abruptly. The dilatation, however, is not extensive. Cylindrical bronchiectasis frequently occurs after a person experiences acute bronchitis, and is reversible. Cylindrical bronchiectasis is the most common among the three. Varicose bronchiectasis is characterized by dilated bronchi having irregular, bulging contours. This configuration parallels the pattern of varicose veins. Cystic bronchiectasis, sometimes referred to as saccular, features sharply reduced bronchial subdivisions and dilated bronchi terminating in cystic, pus-filled cavities. Cystic bronchiectasis is the most severe form among these three types.

### Signs & Symptoms

Most patients with bronchiectasis have chronic cough and copious purulent to mucopurulent sputum production. Hemoptysis is common among these patients because of the severity of the bronchial wall destruction and hypertrophied bronchial arteries. In fact, hemoptysis can be a life-threatening problem with these patients because the pulmonary vessels that tend to bleed are bronchial arteries, which are systemic vessels

under high pressure. Generally, patients with bronchiectasis produce from 100 to 500 ml of sputum per day. Patients with chronic bronchiectasis produce sputum, that when allowed to remain in a collection cup, separates into three distinct layers. The top layer is frothy; the middle layer appears green, while the bottom layer is an accumulation of pus. Interestingly, not all bronchiectasis patients have a productive cough. Some are asymptomatic and have what is termed "dry" bronchiectasis.

Dyspnea on exertion is common, and in severe cases occurs at rest. Digital clubbing is sometime. Auscultation of the chest frequently reveals crackles, rhonchi, and wheezes resulting from bronchial damage and the presence of underlying secretions. Other findings may include plethora, cachexia, and weight loss.

### Diagnosis

High-resolution computed tomography (CT) scanning is sensitive and specific for detecting bronchiectasis, and is regarded as the cornerstone for the diagnosis of this condition. The deranged and distorted bronchial walls are clearly visualized in cross-sectional images from a CT scan. Chest radiography is also used, but the results are frequently non-specific. A chest radiograph is normal in about 30 percent of patients with bronchiectasis. When chest radiography yields findings associated with bronchiectasis, the x-ray may demonstrate enlarged, dilated airways. However, these findings are difficult to differentiate from enlarged alveoli caused by bullous emphysema, or from honeycombing in patients who have severe interstitial lung disease. In cylindrical bronchiectasis, linear, parallel lucencies are sometimes viewed emanating from the hilum. This radiographic characteristic is described as "tram tracks."

Pulmonary function tests commonly reflect an obstructive defect, which tends not to be reversible with bronchodilator therapy. However, a restrictive defect is sometimes observed in patients who have advanced scarring, fibrosis, and atelectasis.

### Treatment

The therapeutic mainstays for bronchiectasis are antibiotics and bronchial hygiene. Amoxicillin, ampicillin, and trimethoprim-sulfamethoxazole (Bactrim) are often prescribed while results of a culture and sensitivity are pending.

Bronchial hygiene techniques such as chest physiotherapy, flutter valve, acapella, or positive expiratory pressure (PEP) are directed toward facilitating the clearance of tracheobronchial secretions, and reducing the degree of airflow obstruction.

Nutritional supplementation is an important consideration with these patients because bronchiectasis produces wasting and weight loss from an increased work of breathing. Smoking cessation is critical for smokers with bronchiectasis. Avoidance of second-hand smoke is equally imperative. Immunization against pneumonia and influenza may be beneficial. Patient with severe forms will likely require oxygen therapy.

Mucolytic agents and/or 7% saline aerosols are frequently prescribed to assist with the mobilization of secretions. Inhaled corticosteroids have proved favorable for some of these patients. Surgical resection may be useful, particularly if the bronchiectasis is confined to localized areas of the lungs.

### Prognosis

Because bronchiectasis often results from some other underlying pulmonary condition, the prognosis often depends on how well the predisposing disease can be managed. In general, the prognosis tends to be good, especially for patients who are compliant and vigilant with their therapeutic regimens.