BRONCHIECTASIS

'Bronchiectasis' is the term used for the permanent abnormal dilatation of one or more bronchi.
Bronchiectasis is characterized by chronic dilation and distortion of one or more bronchi—usually as a result of extensive inflammation and destruction of the bronchial wall cartilage, blood vessels, elastic tissue, and smooth muscle components. One or both lungs may be involved. Bronchiectasis is commonly limited to a lobe or segment and is frequently found in the lower lobes. The smaller bronchi, with less supporting cartilage, are predominantly affected. Because of bronchial wall destruction, normal mucociliary clearance is impaired. This results in the accumulation of copious amounts of bronchial secretions and blood that often become foul-smelling because of secondary colonization with anaerobic organisms. Thoracic infection and irritation may lead to secondary bronchial smooth muscle constriction and fibrosis. The small bronchi and bronchioles distal to the affected areas become partially or totally obstructed with secretions. This condition leads to one or both of the following anatomic alterations:
(1) hyperinflation of the distal alveoli as a result of expiratory check-valve obstruction or
(2) atelectasis, consolidation, and fibrosis as a result of complete bronchial obstruction.

Based on gross anatomic appearance, the long-accepted Reid classification subdivides bronchiectasis into the following three different patterns:
• Varicose (fusiform)
• Cylindrical (tubular)
• Cystic (saccular)

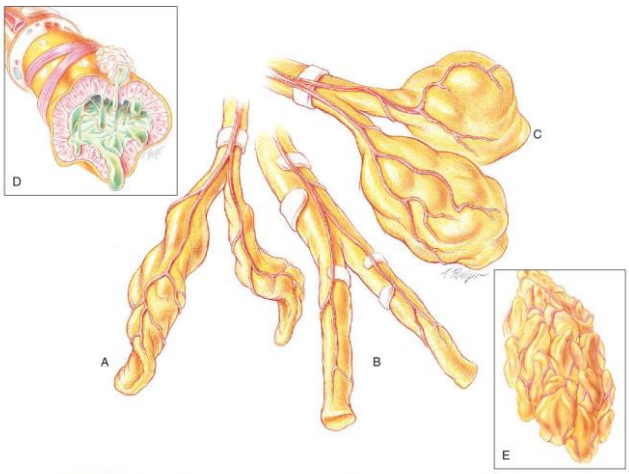

FIGURE 14-1 Bronchiectasis. **A**, Varicose bronchiectasis. **B**, Cylindrical bronchiectasis. **C**, Cystic (saccular) bronchiectasis. Also illustrated are excessive bronchial secretions (**D**) and atelectasis (**E**), which are both common anatomic alterations of the lungs in this disease.

Varicose Bronchiectasis (Fusiform Bronchiectasis)
In **varicose (fusiform) bronchiectasis**, the bronchi are dilated and constricted in an irregular fashion similar to varicose veins, ultimately resulting in a distorted, bulbous shape.

Cylindrical Bronchiectasis (Tubular Bronchiectasis)
In **cylindrical (tubular) bronchiectasis**, the bronchi are dilated and rigid and have regular outlines similar to a tube. X-ray examination shows that the dilated bronchi fail to taper for 6 to 10 generations and then appear to end abruptly because of mucous obstruction.

Cystic Bronchiectasis (Saccular Bronchiectasis)
In **cystic (saccular) bronchiectasis**, the bronchi progressively increase in diameter until they end in large, cyst like sacs in the lung parenchyma. This form of bronchiectasis causes the
greatest damage to the tracheobronchial tree. The bronchial walls become composed of fibrous tissue alone—cartilage, elastic tissue, and smooth muscle are all absent.

AETIOLOGY

The causes of bronchiectasis are commonly classified into the following categories:
• Acquired bronchial obstruction
• Congenital anatomic defects

• Immunodeficiency states
• Abnormal secretion clearance
• Miscellaneous disorders (e.g., alpha1-antitrypsin deficiency)

TABLE 14-1 Causes of Bronchiectasis—cont'd

Category	Specific Examples	Diagnostic Tests
Immunodeficiency States		
IgG deficiency	Congenital (Bruton's type) agammaglobulinemia; selective deficiency of subclasses (IgG2, IgG4); acquired immune globulin deficiency; common variable hypogammaglobulinemia; Nezelof's syndrome; "bare lymphocyte" syndrome	Quantitative immunoglobulin levels; immunoglobulin subclass levels; impaired response to immunization with pneumococcal vaccine
IgA deficiency	Selective IgA deficiency ± ataxia-telangiectasia syndrome	Quantitative immunoglobulin levels
Leukocyte dysfunction	Chronic granulomatous disease (NADPH oxidase dysfunction)	Dihydrorhodamine 123 (DHR) oxidation test; nitroblue tetrazolium test; genetic testing
Other rare humoral immunodeficiencies (CXCR4 mutation, CD40 deficiency, CD40 ligand deficiency, and others)	WHIM syndrome; Hypergammaglobulinemia M	Neutrophil count; quantitative immunoglobulin levels
Abnormal Secretion Clearance		
Ciliary defects of airway mucosa	Kartagener's syndrome; ciliary dyskinesis (formally called impaired ciliary motility syndrome)	Chest x-ray showing situs inversus; bronchial biopsy; ciliary motility studies; electron microscopy of sperm or respiratory mucosa
Cystic fibrosis (mucoviscidosis)	Typical early childhood syndrome; later presentation with predominantly sinopulmonary symptoms	Sweat chloride; genetic testing
Young's syndrome	Obstructive azoospermia with sinopulmonary infections	Sperm count
Miscellaneous Disorders		
$Alpha_1$-antitrypsin deficiency	Absent or abnormal antitrypsin synthesis and function	$Alpha_1$-antitrypsin level
Recurrent aspiration pneumonia	Alcoholism; neurologic disorders; lipoid pneumonia	History; chest imaging
Rheumatic disease	Associated with rheumatoid arthritis and Sjögren's syndrome	Rheumatoid factor; antiSSA/antiSSB; salivary gland MRI or biopsy
Inflammatory bowel disease	Crohn's disease; ulcerative colitis	History; lower gastrointestinal endoscopy; imaging studies; colonic biopsy
Inhalation of toxic fumes and dusts	Ammonia; nitrogen dioxide, or other irritant gases; smoke; talc; silicates	Exposure history; chest imaging
Chronic organ rejection following transplantation	Bone marrow, lung and heart lung transplantation; associated with obliterative bronchiolitis	History; PFT; chest CT imaging with inspiratory and expiratory views

COPD, Chronic obstructive pulmonary disease; *CT*, computed tomography; *Ig*, immunoglobulin; *MRI*, magnetic resonance imaging; *PFT*, pulmonary function test; *PPD*, percussion and postural drainage.
Modified from Wolters Kluwer Health/UpToDate.com: *Causes of bronchiectasis in children and clinical manifestations and diagnosis of bronchiectasis in adults*. Accessed March 1, 2013.

Pathophysiology

The inflammatory processes related to acute or chronic lung infection damage the cilia, which enables bacteria to remain in the airway and colonize the mucus. These microorganisms stimulate a host inflammatory response which further inhibits ciliary function, damages the elastic and muscular tissue of the bronchial walls and stimulates mucus production. The loss of elastic and muscular tissue in the airway wall leads to a dilatation of the bronchi. A vicious cycle ensues where clearance of bronchial secretion from these dilated bronchi is impaired and secretions often become chronically infected, producing a persistent host inflammatory response which results in a progressive destructive lung disease.
Depending upon the aetiology, bronchiectasis can be localized to a specific lung region or can be widespread.

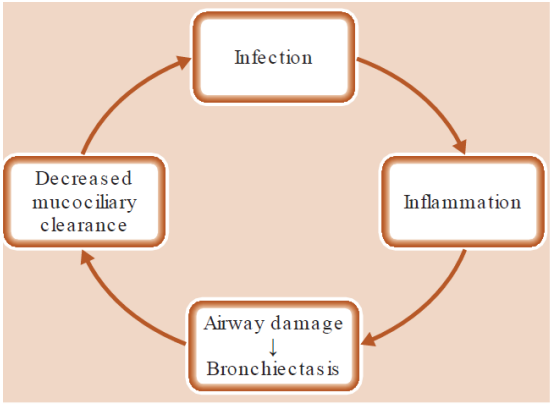

FIGURE 5-7 ■ Vicious cycle of bronchiectasis.

The following are the major pathologic or structural changes associated with bronchiectasis:
• Chronic dilation and distortion of bronchial airways
• Excessive production of often foul-smelling sputum
• Bronchospasm
• Hyperinflation of alveoli (air trapping)
• Atelectasis
• Consolidation and parenchymal fibrosis
• Hemoptysis secondary to bronchial arterial erosion.

Clinical Features

Symptoms vary according to disease severity, with some people being totally asymptomatic and others, with severe disease, having a cough productive of large amounts of purulent sputum which is sometimes bloodstained. Severe exacerbations may be accompanied by chest pain, breathlessness and fevers. The signs are non-specific and include auscultation findings of localized or widespread inspiratory and expiratory crackles with occasional wheezing. Clubbing of fingers and/or toes may occur in severe disease. In some cases of more severe disease, exercise tolerance may be reduced.

Diagnosis and Investigations

- High-resolution computed tomography (HRCT)
- Chest Radiograph
- Sputum specimens are analyzed to identify the micro-organisms and their sensitivity to antibiotics. The most common bacteria found in bronchiectatic sputum are *Haemophilus in uenzae*,

  Streptococcus pneumoniae and *Pseudomonas aeruginosa*.

- Lung function tests are used to assess severity of airflow obstruction and the degree of reversibility.

Medications Commonly Prescribed by the Physician

- Expectorants
  Expectorants sometimes are ordered when oral liquids and aerosol therapy alone are not sufficient to facilitate expectoration. Their clinical effectiveness is doubtful.
- Antibiotics
  Antibiotics are commonly administered to treat associated respiratory tract infections.

Implications for Physical Therapy Treatment

Management of bronchiectasis can include all of the following physical therapy interventions:
• Secretion clearance techniques
• Controlled breathing techniques coordinated with activity
• Strength training
• Endurance exercise
(For detailed treatment protocol refer to textbooks mentioned in references)

REFERENCES

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