"He who studies medicine without books sails an uncharted sea, but he who studies medicine without patients does not go to sea at all."
William Osler

Medicine

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Bronchiectasis

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Bronchiectasis

Definition of Bronchiectasis

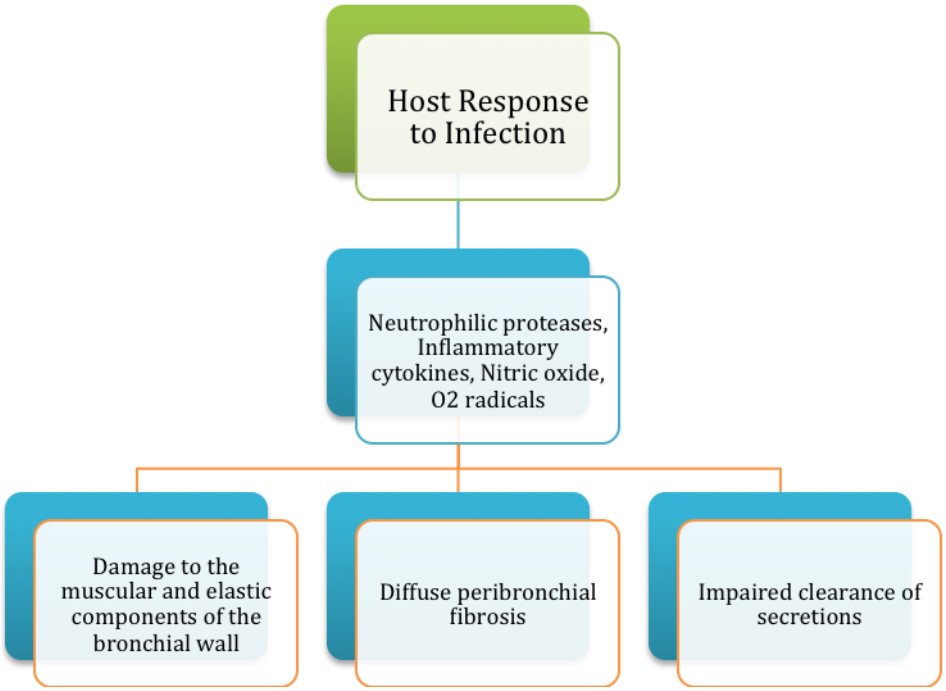
Is a chronic disorder characterized by permanent dilatation of the bronchi accompanied by inflammatory changes in their walls and in the adjacent lung parenchyma.
Bronchiectasis remains a significant cause of morbidity and mortality in the developed world

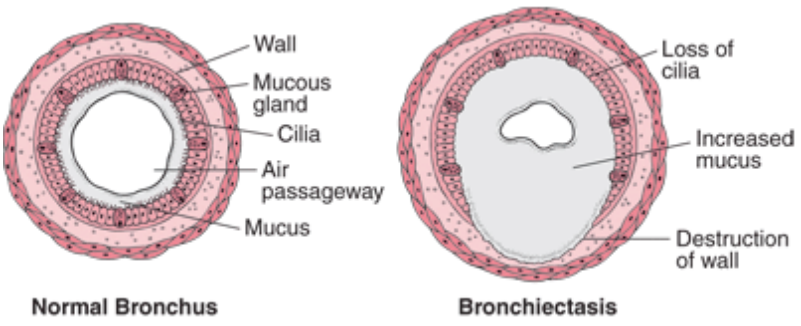
Physiology

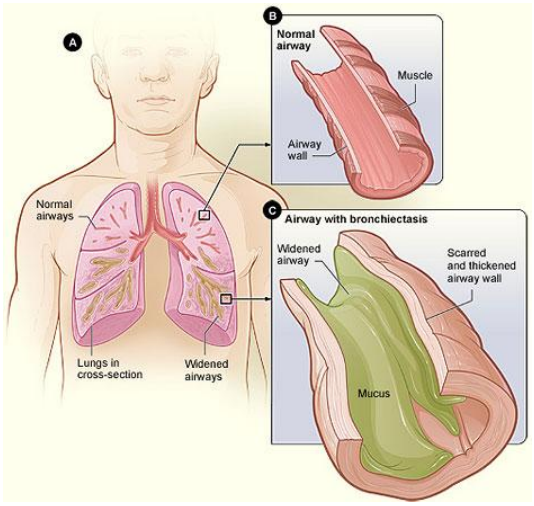
The human airways are lined with ciliated epithelium with submucosal goblet cells secreting mucus that makes up the top layer of the airway surface liquid
The lower layer being the periciliary fluid that bathes the cilia and ensures they function appropriately.
Lungs are continuously exposed to inhaled pathogens and have developed an advanced mechanism for trapping and removing them
In healthy individuals the mucus traps inhaled pathogens and the continuously motile cilia transport the mucus and its contents out of the lung.

Pathophysiology and mechanism of Bronchiectasis

Any defect in this mucociliary clearance mechanism can lead to the retention of pathogens resulting in the progression of airway infection, inflammation and ultimately bronchiectasis.







Types of Bronchiectasis

- Cystic fibrosis (*Channelopathies)*
- Non-CF related bronchiectasis (More common)

Cystic fibrosis (*Channelopathies)*

- Is the most common lethal inherited disease in white persons
- Caused by defects in the gene for cystic fibrosis transmembrane conductance regulator (CFTR)
- Decreased secretion of chloride and increased reabsorption of sodium and water across epithelial cells.
- Decreased hydration results in mucus that is stickier to bacteria, which results in infection and inflammation.
- ***Viscid secretions in*** the respiratory tract, pancreas, GIT, liver, sweat glands, and other exocrine tissues.
- Bronchiectasis associated with CF occurs secondary to mucous plugging of proximal airways and chronic pulmonary infection (frequently Pseudomonas)

*Prolonged asthma can lead to Non-CF related bronchiectasis (More than 60 years of asthma)

Etiology of bronchiectasis

Obstruction of single bronchus

- Tumors
- Foreign body

Obstructive airways disease

- Asthma
- COPD
- AAT deficiency

Defects of mucociliary clearance

Ciliary dyskinesia

- Primary ciliary dyskinesia (Hereditary/Genetic)
    - Kartagener's syndrome

- Secondary ciliary dyskinesia
    - P. aeruginosa and H. influenzae
    - Disable mucociliary clearance

Major pathogens are Staph. aureus, Pseudomonas aeruginosa, H. influenzae and anaerobes.

Fungal infections can lead to Bronchiectasis in Immunocompromized patients

Channelopathies

- CFTR dysfunction (loss of a chloride channel)
- ENaC dysfunction (Na channel hyperactivity)

ABPA: (Allergic bronchopulmonary aspergillosis)

- Aspergillus fumigatus.

Immunodeficiency

- Common variable immunodeficiency (CVID)
- (XLA) X-linkedagammaglobulinaemia -
- Antibody deficiency with normal Ig
- Secondary immunodeficiency;-
    - Haematological malignancy
    - Post-allogenic bone marrow transplant
    - Drug-induced immunosuppression

Infections

- Childhood infections
- Tuberculosis
- Pneumonia
- Measles
- Whooping cough
- Nontuberculous mycobacteria (TB)

Bronchiectasis in systemic diseases

- Inflammatory bowel disease
- Connective tissue diseases
- Yellow nail syndrome

Idiopathic bronchiectasis

Diagnosis

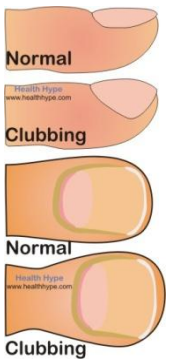
History and clinical examination

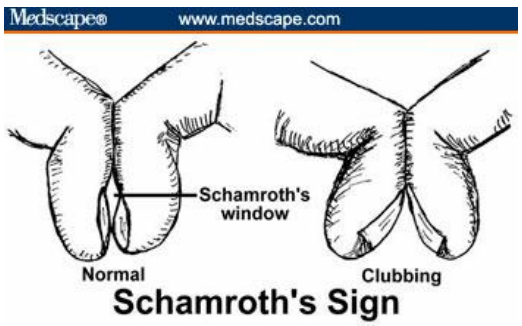
History

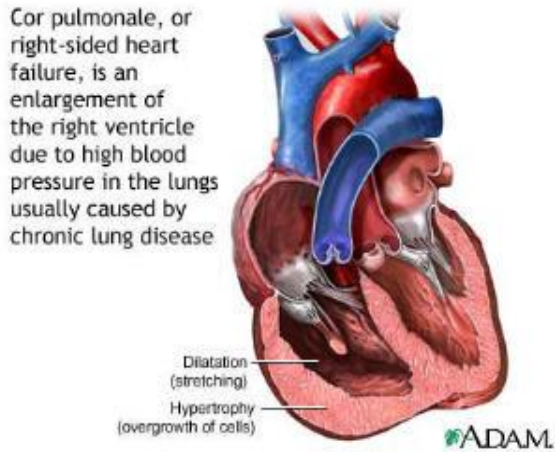
- Multiple episodes of infections
- Cough and daily production of large amounts of mucopurulent sputum
- Blood-streaked hemoptysis
- Dyspnea
- Pleuritic chest pain
- Wheezing
- Fever
- Weakness, and weight loss

Examination

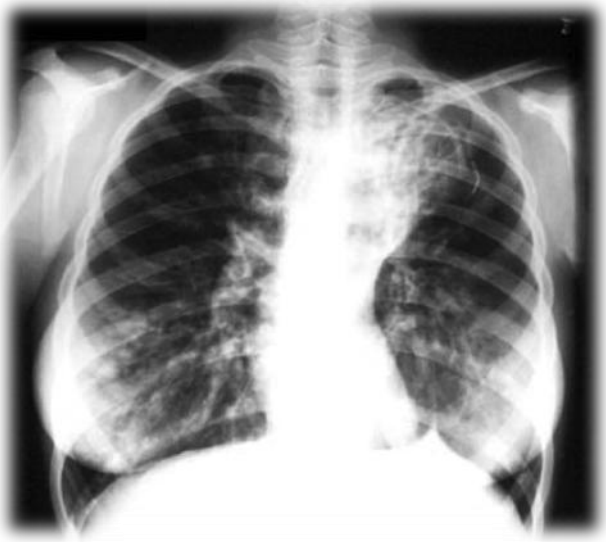
- Coarse crackles (A loud and low-pitched, discontinuous, 'explosive' crackling sound heard in patients with pneumonia, atelectasis, pulmonary fibrosis, acute bronchitis, bronchiectasis, or pulmonary oedema secondary to left-sided congestive heart failure)
- Scattered wheezing: Wheezing may be due to airflow obstruction from secretions
- Digital clubbing
- Cyanosis and plethora are rare findings secondary to polycythemia from chronic hypoxia.
- Wasting and weight loss
- Cor pulmonale. (Right-sided heart failure .)
- Respiratory failure

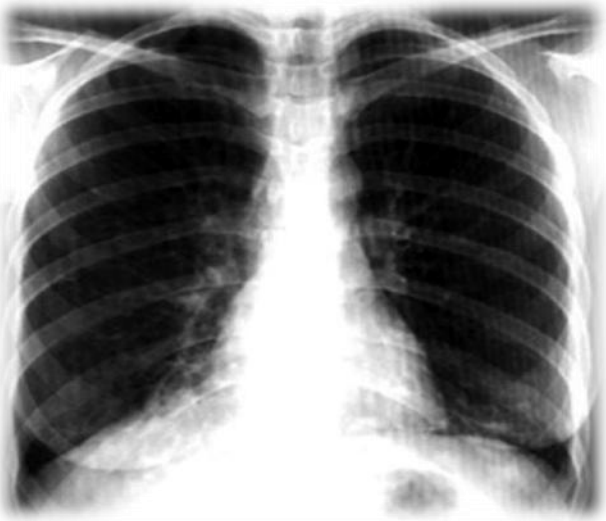






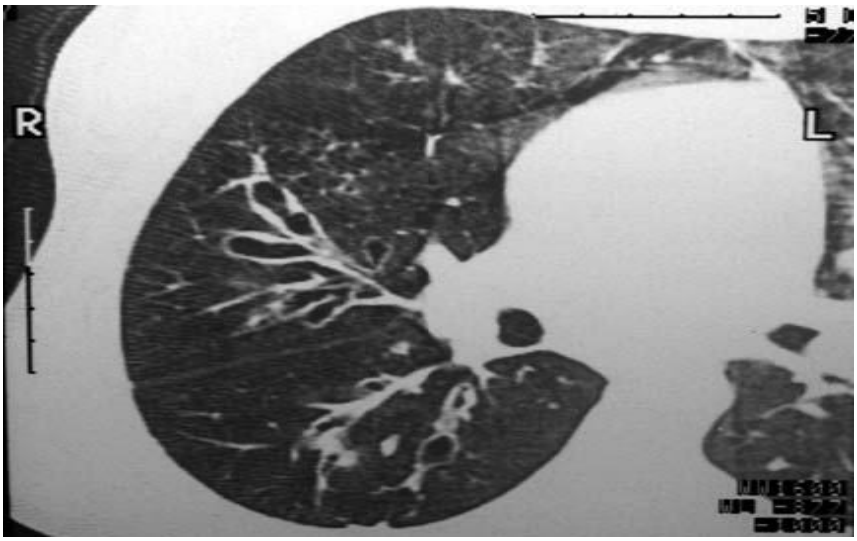
Chest X-ray (Usually nonspecific)





Standard chest x-rays may show increased bronchovascular markings from peribronchial fibrosis and intrabronchial secretions, crowding from an atelectatic lung, tram lines (parallel lines outlining dilated bronchi due to peribronchial inflammation and fibrosis), areas of honeycombing, or cystic areas with or without fluid levels.

CT Scan (High Resolution CT is the study of choice)



Laboratory Studies

A sputum analysis G stain , AFB and c/s

- CBC
- Quantitative immunoglobulin levels
- Quantitative AAT levels (measuring Alpha antitrypsin level, it will be decreased in bronciectasis)
- Sweat test
- Genetic analysis
- *Aspergillus* precipitins and serum total IgE levels
- Rheumatoid factor and/or other autoimmune screening tests
- Pulmonary function test (Reveal an obstructive pattern)

Treatment

- Antibiotic therapy
- Oral, parenteral, and aerosolized antibiotics
- Chest physiotherapy /Postural drainage
- Bronchodilator therapy
- Azithromycin

Surgical care

- Surgery should be reserved for patients who have focal disease that is poorly controlled by antibiotics.
- Massive hemoptysis (bronchial artery embolization)
- Foreign body or tumor removal

Prognosis

- The advent of effective antibiotic therapy has greatly improved the prognosis.
- Ultimately, most patients with severe bronchiectasis will develop respiratory failure.
- Cor pulmonale is another well-recognized complication.
- The three pathogens that can cause infective episodes and are difficult to eradicate are Pseudomonas aeruginosa, Aspergillus fumigatus and MAI.

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