Bronchiectasis

429 Medicine Team Notes

Blue: added notes

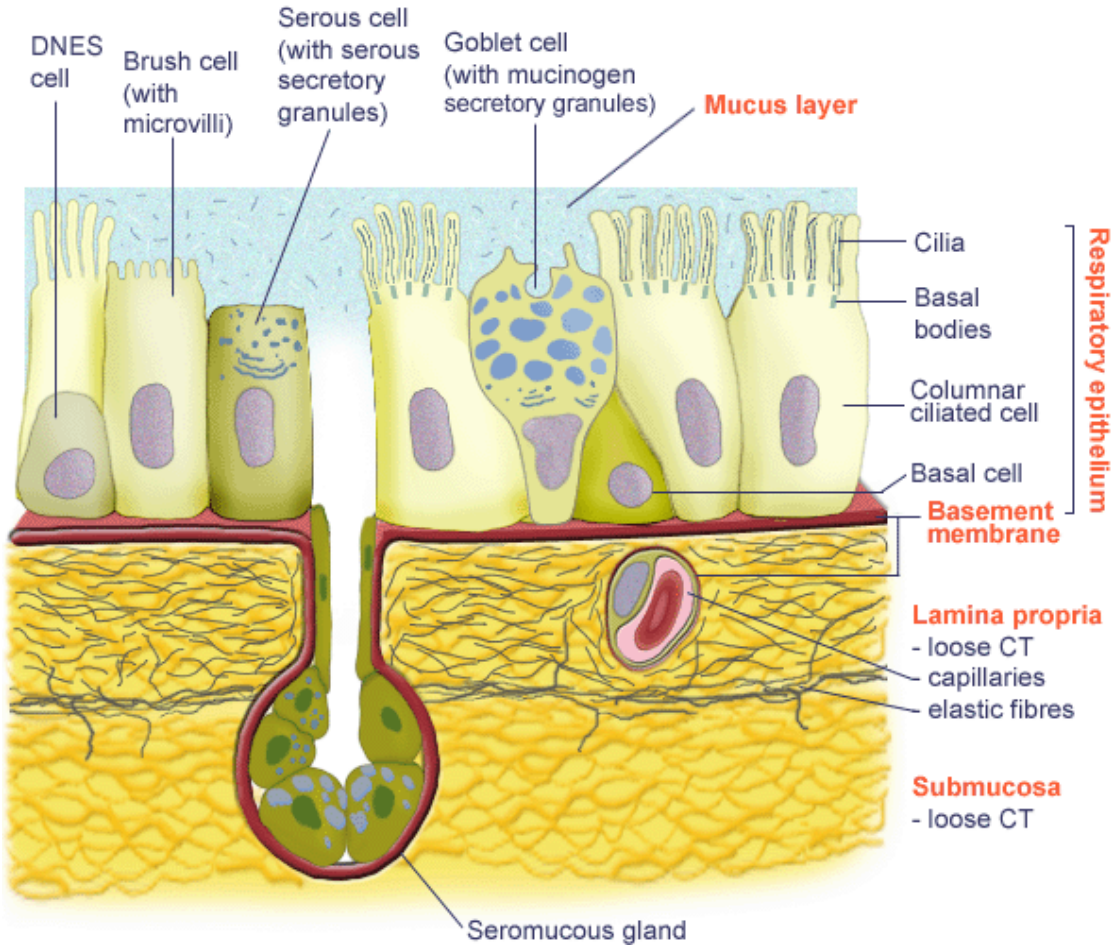
Bronchiectasis

- Is a chronic disorder characterized by thickening and 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

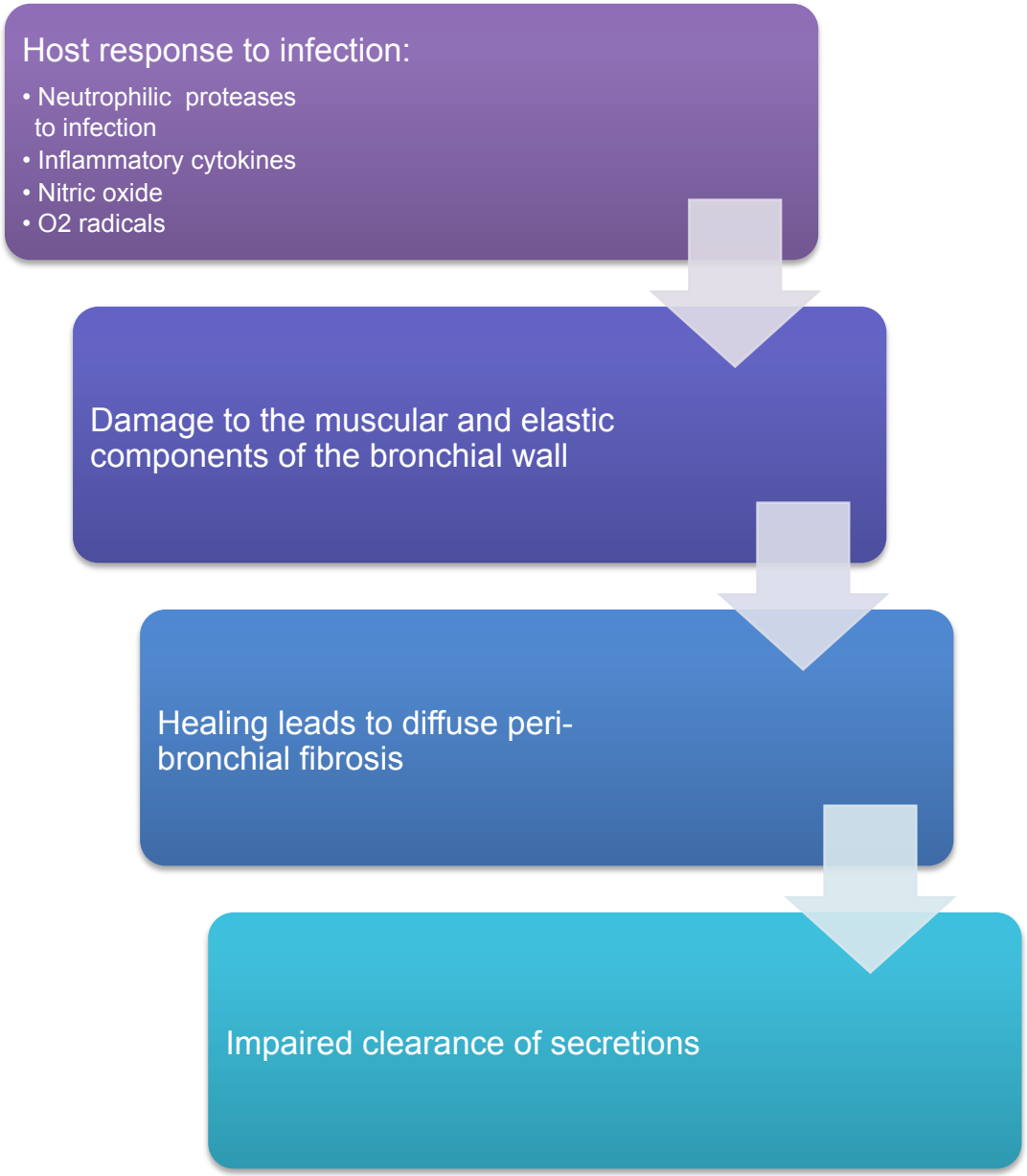
- Airways are lined with ciliated epithelium with submucosal goblet cells secreting mucus that makes up the top layer of the airway surface liquid
- The lower layer: peri-ciliary fluid that bathes the cilia and ensures they function appropriately
- Lungs are continuously exposed to inhaled pathogens 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

Histology of the Airways



Pathophysiology and Mechanism

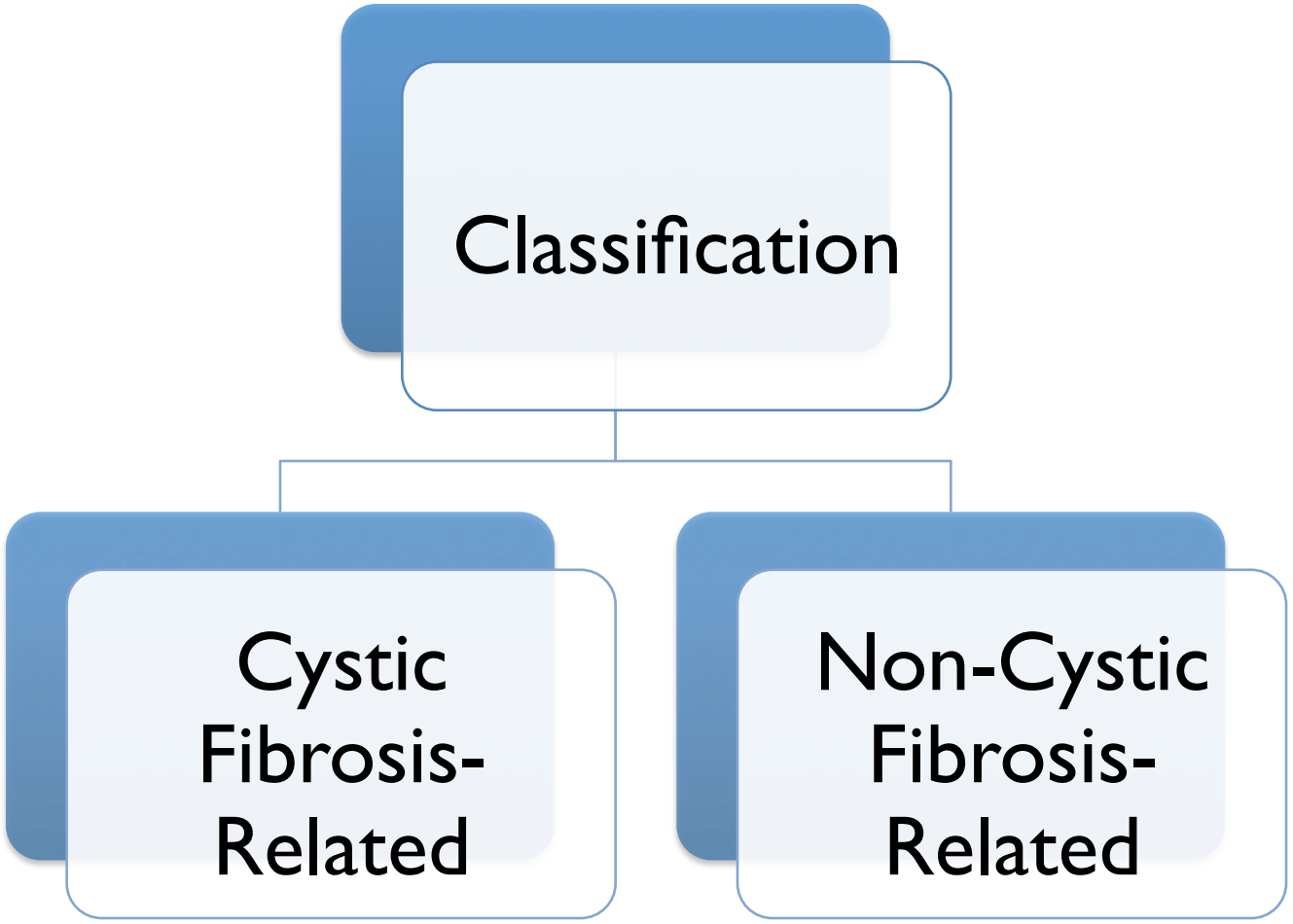
Any defect in this mucociliary clearance mechanism can lead to the retention of pathogens resulting in the progression of airway infection, inflammation and ultimately bronchiectasis.



Epidemiology and Causes

- *Bronchial dilatation can be caused by*
  - A structural defect in the wall itself
  - An effect of abnormal airway pressure on the bronchial wall e.g. coughing
  - Damage to the airway elastic tissue and cartilage as a result of bronchial wall inflammation
- *Inflammation within the bronchial wall*
  - An airway infection
  - Inhalation of injurious agents e.g. gasoline
  - An autoimmune disease

Bronchiectasis



Causes of Non-Cystic Fibrosis-Associated Bronchiectasis (in adults)

- Bacteria (major role), viruses, fungi and nontuberculous mycobacteria > inflammation
- Ciliary dyskinesias (low motility)
- Channelopathies (Na^+ retention, Cl^- channels)
- Connective tissue disease
- Inflammatory bowel disease
- Immunodeficiencies
- Autoimmune disease

Etiology of Bronchiectasis

- **Structural lung conditions/ congenital defects**
  - Williams–Campbell syndrome: absence of cartilage from lobar to first- to second-generation segmental airways that results in extensive peripheral bronchiectasis
  - Mounier-Kuhn syndrome: tracheobronchomegaly; rare disorder characterized by dilation of the trachea and segmental bronchi (central bronchiectasis)
  - Ehlers–Danlos syndrome:
    - A group of more than 10 different inherited disorders; all involve a genetic defect in collagen and connective-tissue synthesis and structure
    - Affects the skin, joints, and blood vessels
    - Lax supporting tissue may lead to bronchiectasis

Etiology of Bronchiectasis

- **Toxic damage to airways** *(leads to wall inflammation)*
  - Inhalational injury
  - Aspiration secondary to neuromuscular disease
  - Gastro esophageal reflux disease (GERD)
- **Obstruction of single bronchus (rare)** *(increased pressure)*
  - Tumors
  - Foreign body
- **Obstructive airways disease** *(increased pressure)*
  - Asthma
  - COPD
  - α_1-antitrypsin deficiency

Etiology of Bronchiectasis

- **Defects of mucociliary clearance**
  - *Ciliary dyskinesia*
    - Primary ciliary dyskinesia: immotile cilia
    - Kartagener's syndrome: clinical triad of situs inversus (the major visceral organs are reversed or mirrored from their normal positions), nasal polyps or sinusitis, and bronchiectasis in the setting of immotile cilia of the respiratory tract; congenital
    - Secondary ciliary dyskinesia: P. aeruginosa and H. Influenzae disable mucociliary clearance
  - *Channelopathies*
    - Cystic Fibrosis Transmembrane conductance R dysfunction (loss of a chloride channel)
      - ➤ blockage of the movement of salt and water into and out of cells → cells that line the passageways of the lungs produce abnormally thick, sticky mucus → obstructs the airways and glands → signs and symptoms of cystic fibrosis
      - ➤ thick mucus cannot be removed by cilia, so it traps bacteria that give rise to chronic infections.
    - Epithelial Na Channel Dysfunction (Na channel hyperactivity) causes retention of water inside the cell

Etiology of Bronchiectasis

- **Immunodeficiency**
  - Common variable immunodeficiency (CVID): a disorder that involves the following:
    - (1) low levels of most or all of the immunoglobulin (Ig) classes
    - (2) a lack of B lymphocytes or plasma cells that are capable of producing antibodies
    - (3) frequent bacterial infections
  - XLA: X-linked agammaglobulinemia
  - Antibody deficiency with normal Ig
  - Secondary immunodeficiency
    - Hematological malignancy e.g. leukemia, lymphoma
    - Allogeneic bone marrow transplant
    - Drug-induced immunosuppression

  All lead to: Recurrent infections which cause permanent damage to the bronchi and may lead to bronchiectasis

Etiology of Bronchiectasis

- **Infections**
  - Childhood infections
    - Tuberculosis
    - Pneumonia (complicating: Measles or whooping cough)
    - Nontuberculous mycobacteria
  - Adults
    - Suppurative pneumonia
    - Pulmonary TB
- Bronchiectasis in systemic diseases
  - Inflammatory bowel disease
  - Connective tissue disorders e.g. SLE, yellow nail syndrome
- Idiopathic bronchiectasis

Etiology of Bronchiectasis

- **ABPA:** Allergic bronchopulmonary aspergillosis
  - A hypersensitivity reaction to inhaled Aspergillus antigen (A. fumigatus) that is characterized by bronchospasm, bronchiectasis, and immunologic evidence of a reaction to Aspergillus species
  - Airway plugging by viscid secretions containing hyphae of Aspergillus species

Summary of Etiology

Congenital

- Cystic Fibrosis *
- Primary Ciliary Dyskinesias *
- α1-Antitrypsin Deficiency
- Anatomical Defects
  - Tracheoesophageal fistula
  - Bronchotracheomalacia
  - Tracheomegaly
  - Pulmonary sequestration
  - Yellow Nail Syndrome, Marfan Syndrome, etc

Acquired

- Infections
  - Bacterial *
  - Viral: Measles, HIV *
  - Fungal: ABPA *
- Immunodeficiency
  - Primary: Common variable immunodeficiency (CVID) *, X-linked agammaglobulinemia *, IgA Deficiency
  - Secondary
    - Hematological malignancy *
    - Chemotherapy *
- Interstitial lung disease * i.e. traction bronchiectasis (because of fibrosis)
- Autoimmune: Rheumatoid Arthritis *, Ulcerative Colitis, Sarcoidosis
- Post-surgical
- GERD

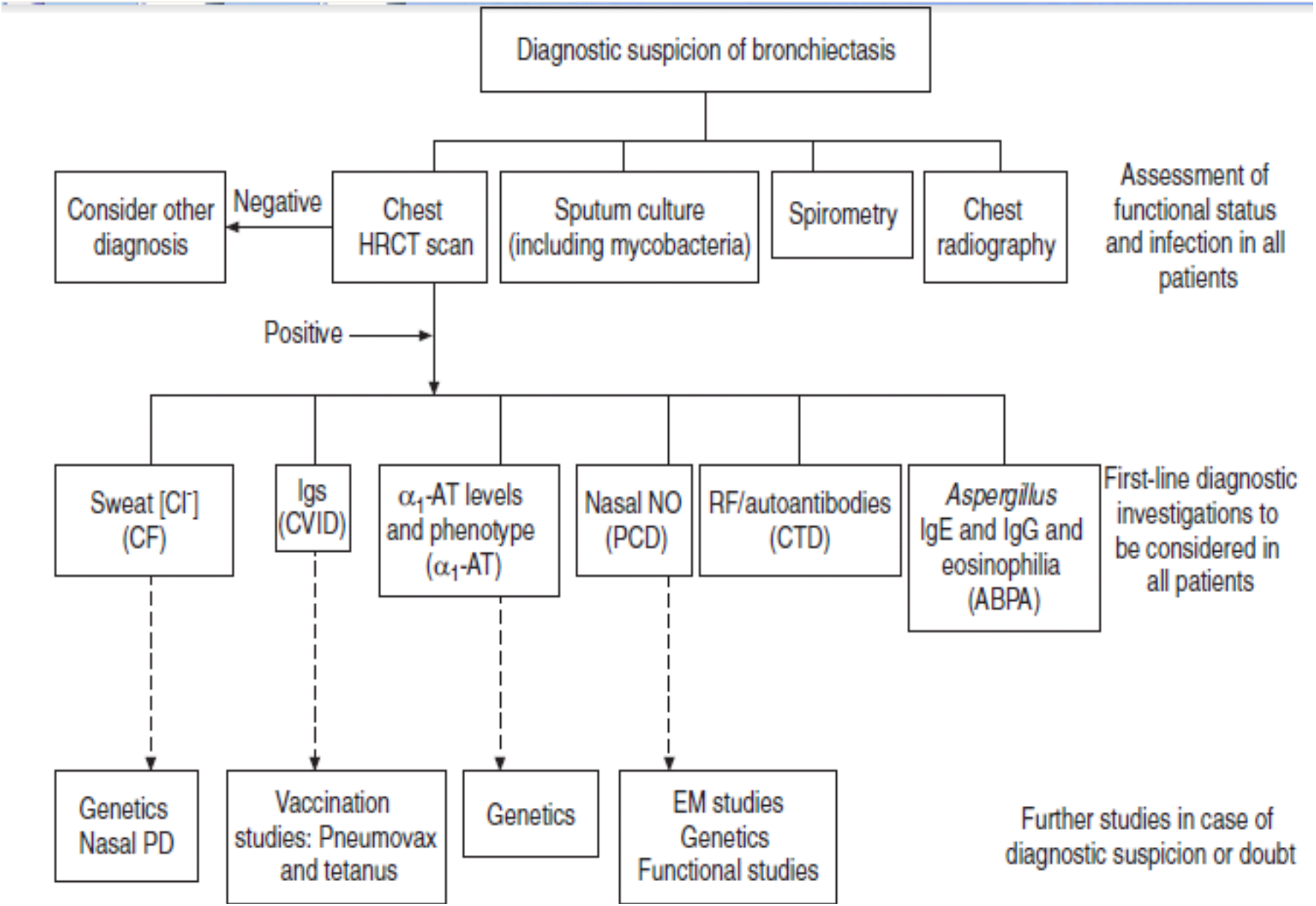
* Most common

Cystic Fibrosis

- Is the most common lethal inherited disease in white persons
- Cause: 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 occur secondary to mucous plugging of proximal airways and chronic pulmonary infection

Clinical feature

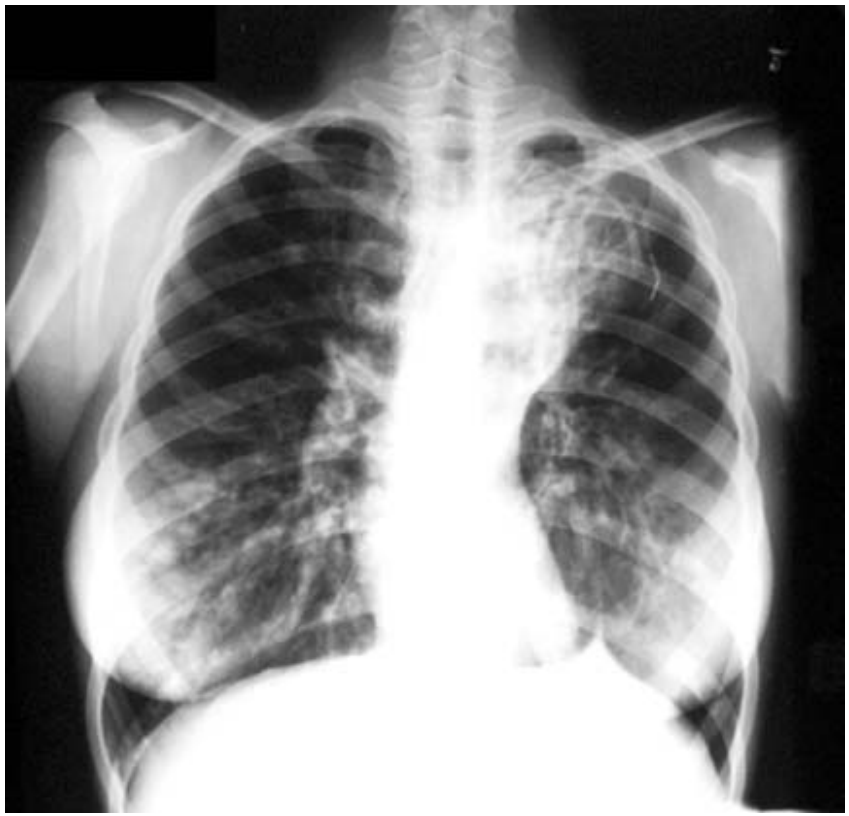
- Multiple episodes of infections
- Cough and daily production of big amount of mucopurulent sputum
- Blood-streaked hemoptysis
- Dyspnea
- Pleuritic chest pain
- Wheezing
- Fever
- Weakness
- Weight loss and wasting
- Coarse crackles
- Scattered wheezing (may be due to airflow obstruction from secretions)
- Digital clubbing
- Cyanosis and plethora (rare findings secondary to polycythemia from chronic hypoxia)
- Cor pulmonale. (Right-sided heart failure In severe cases)
- Respiratory failure

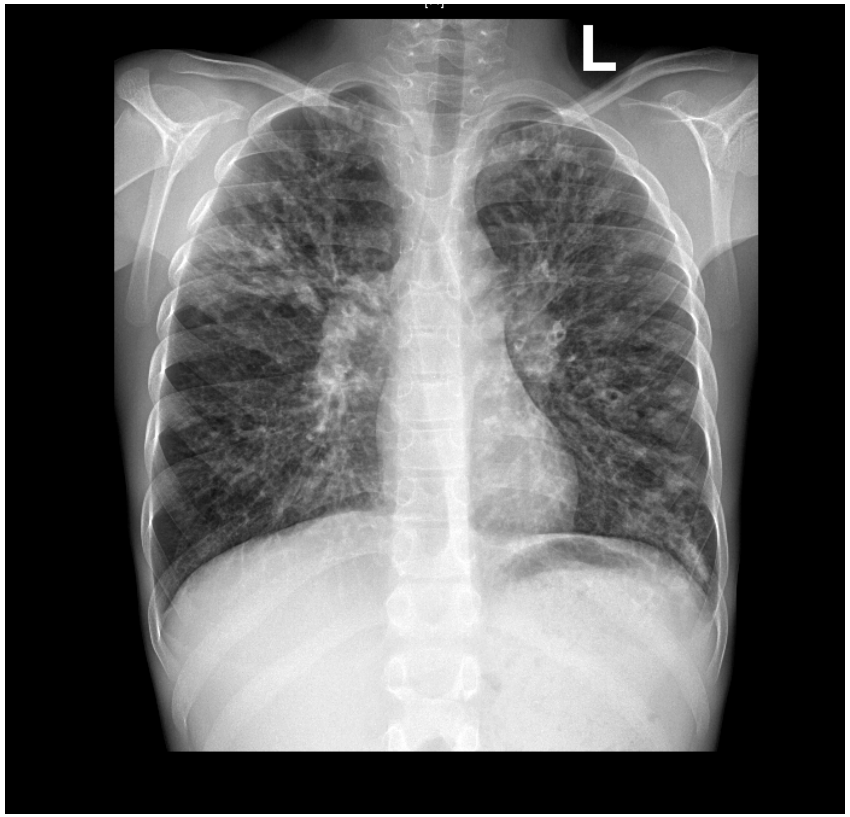

Diagnostic suspicion of bronchiectasis
Consider other diagnosis
Negative
Chest HRCT scan
Sputum culture (including mycobacteria)
Spirometry
Chest radiography
Assessment of functional status and infection in all patients
Positive
Sweat [Cl⁻] (CF)
Igs (CVID)
α_1-AT levels and phenotype (α_1-AT)
Nasal NO (PCD)
RF/autoantibodies (CTD)
Aspergillus IgE and IgG and eosinophilia (ABPA)
First-line diagnostic investigations to be considered in all patients
Genetics Nasal PD
Vaccination studies: Pneumovax and tetanus
Genetics
EM studies Genetics Functional studies
Further studies in case of diagnostic suspicion or doubt

Diagnosis

- History and clinical examination
- CXR: may be normal or may show dilated bronchi with thickened bronchial walls and sometimes multiple cysts containing fluid
- Standard: High resolution computed tomography (HRCT) shows bronchial dilatation with loss of airway tapering at the periphery, bronchial wall thickening and cysts at the end of the bronchioles

Imaging





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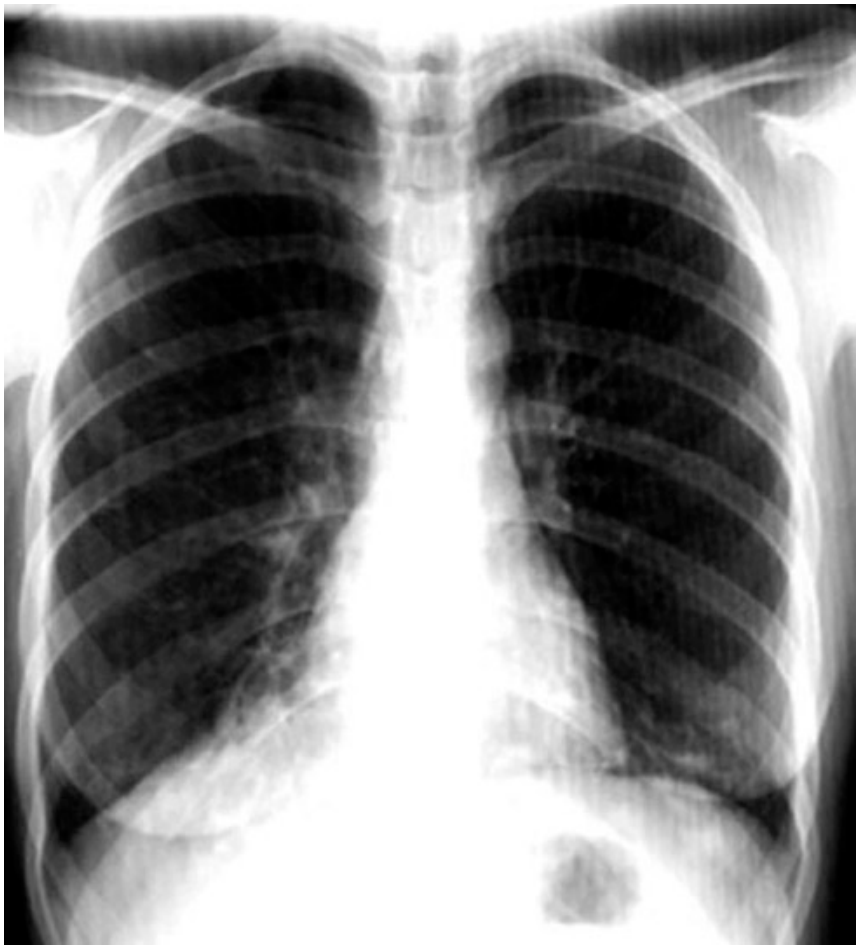

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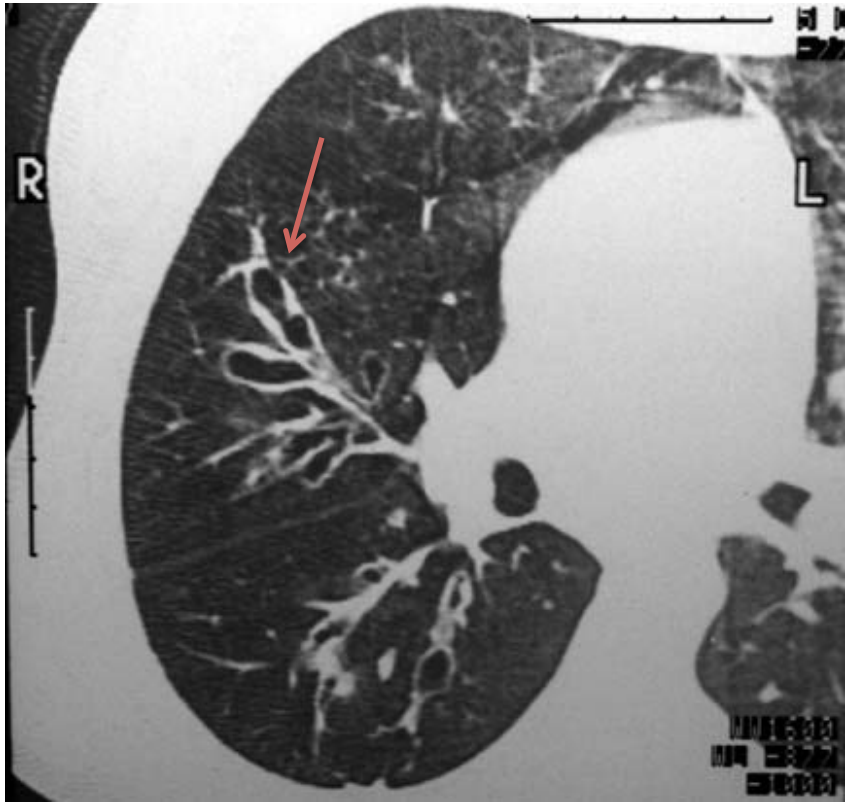

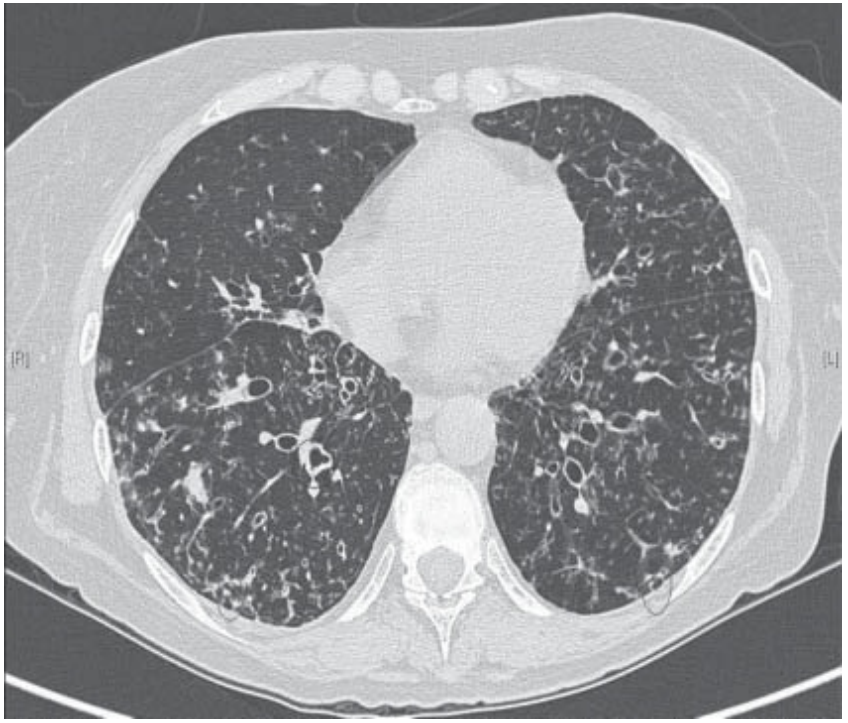

Dilated airways in both lower lobes; bronchi have ring-like appearance (because of mucus)

Laboratory Studies

Bronchiectasis

- **Sputum analysis:** Gram stain, Acid Fast Bacilli and culture/ sensitivity
- CBC: Leukocytosis, neutrophilia etc
- Quantitative immunoglobulin levels (IgA could be low)
- Quantitative AAT levels
- Aspergillus precipitins and serum total IgE levels
- Rheumatoid factor and/or other autoimmune screening tests
- Pulmonary function test

To D_x Cystic Fibrosis:

- Sweat electrolytes: for sodium concentration (should be low) ± Cl^- concentration
- Genetic analysis; blood DNA analysis of gene defect
- Motility of cilia: Mucociliary clearance (nasal clearance of saccharin) *A cube of saccharin is placed on the inferior turbinate and the time to taste measured (normally less than 30 minutes)*

Treatment

- Antibiotic therapy
  - Oral, parenteral, and aerosolized antibiotics
  - Should be guided by the microbiological sensitivities (esp. secondary infections)
  - If mild to moderate: azithromycin, amoxicillin, tetracyclines, cephalosporins (cefaclor), fluoroquinolones
  - If moderate to severe: parenteral aminoglycoside (tobramycin, gentamycin), antipseudomonal penicillins (piperacillin), cephalosporins (cefixime, ceftriaxone, cefotaxime)
- Chest physiotherapy/Postural drainage:
  - *Patients must be trained by physiotherapists to tip themselves into a position in which the affected lobe(s) are uppermost at least three times daily for 10-20 minutes.*
  - *Most patients find that lying over the side of the bed with head and thorax down is effective.*
- Bronchodilator therapy
  - if there is airflow limitation
- Azithromycin

Surgical Care

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