

Etiology (only 40% of cases have an identifiable cause)

- Underlying Problem
 - Obstructive Process (CF, Tumor, Foreign Body, Asthma, COPD, Extrinsic Airway Narrowing, etc)
 - **Middle Lobe Syndrome:** b/c the bronchus supplying the middle lobe is narrow, has a sharp angle branch, and a fish mouth opening it is more prone to obstruction especially from extrinsic sources
 - **Mounier-Kuhn Syndrome:** absence of bronchial elastin/muscle
 - **Williams-Campbell Syndrome:** absence of bronchial cartilage
 - Impaired Bronchial Drainage (CF, ABPA, Inhalational Injury, Bad GERD etc)
 - **Kartegener's Syndrome:** congenital (AR) poorly functioning cilia resulting in (1) bronchiectasis and chronic sinusitis, (2) hearing problems, (3) sperm immotility resulting in sterility, and (4) situs inversus
 - **Young's Syndrome:** acquired (Mercury poisoning) poorly functioning cilia resulting (1) bronchiectasis and chronic sinusitis, (2) hearing problems, (3) sperm immotility resulting in sterility
 - Impaired Immune System (CF, Immunodeficiency, Transplantation resulting in Bronchiolitis Obliterans Syndrome, etc)
 - Other (Connective Tissue Disease, IBD, etc)
 - **Yellow Nail Syndrome:** lymphatic obstruction resulting in (1) bronchiectasis, (2) lymphedema, (3) pleural effusion, and (4) yellow discoloration of the nails
 - **Traction Bronchiectasis:** fibrotic lung disease resulting in traction of airways
- Predisposes Pt to a Necrotizing Infection usually *Pseudomonas aeruginosa* or *Haemophilus influenza*
 - NB it used to be a disorder that begin in children b/c of just poorly treated recurrent pneumonias (Measles, Pertussis, Mycoplasma, TB) but now it is seen in adults b/c of causes noted above
- Resulting in irreversible destruction of bronchial tissue leading to permanent dilation of bronchi, bronchioles, and distal airways

Morphology

- Dilated airways that extend to pleural surface bilaterally and especially at lower lobes
- Diffuse if systemic process as cause vs Localized if prior pneumonia as cause (classically if ABPA then almost always central bronchiectasis)
- Can be classified as cylindrical/fusiform/tubular (least severe w/ preserved pulmonary arterial flow), varicosal (in between), or saccular/cystic (most severe w/ diminished pulmonary arterial flow)

S/S

- productive cough
 - w/ hemoptysis
 - w/ large amounts of foul smelling purulent sputum w/ halitosis
- dyspnea
- recurrent infections (pneumonia/bronchitis)

Dx

- CXR: 10% are normal, "tram track or toothpaste lines" (parallel lines outlining dilated bronchi) that originate from hilum, cysts
- HRCT: with this imaging modality many pts are diagnosed with bronchiectasis but do not have symptoms

Tx

- Prevention: Vaccinations, inhaled anti-psuedomonal aminoglycosides if colonized with *Pseudomonas*
- Exacerbation Tx: ABX, BD, Bronchiopulmonary Hygeine, Inhaled NSAIDs, ICS
- Definitive Tx: IR/bronchoscopic bronchial arterial embolization if hemoptysis, lobectomy to transplant