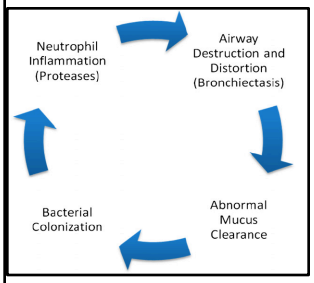
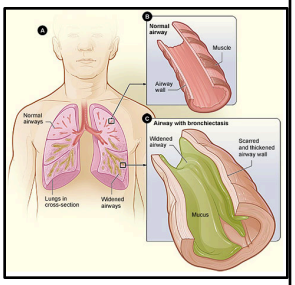
Non-CF Bronchiectasis

Clinical Features
- Chronic cough and sputum
- Hemoptysis
- Dyspnea
- Weight loss
- Recurrent pulmonary infections
- Recurrent rhinosinusitis
- Wheezing
- Fatigue

Other Suggestive Factors
- Difficult to treat asthma
- Nonsmokers diagnosed with COPD
- *Pseudomonas aeruginosa* or NTM in sputum

Pathophysiology





Differential Diagnosis

Postinfectious conditions
- Bacteria (pseudomonas, haemophilus)
- Mycobacterium tuberculosis
- Aspergillus species — Hypersensitivity → ABPA
- Virus (adenovirus, measles virus, influenzavirus, human immunodeficiency virus)

Congenital conditions
- Primary ciliary dyskinesia
- Alpha$_1$-antitrypsin deficiency
- Cystic fibrosis
- Tracheobronchomegaly (Mounier–Kuhn syndrome)
- Cartilage deficiency (Williams–Campbell syndrome)
- Pulmonary sequestration
- Marfan's syndrome

Immunodeficiency
- Primary
  - Hypogammaglobulinemia — HIV, Job's syndrome, CGD
- Secondary
  - Caused by cancer (chronic lymphatic leukemia), chemotherapy, or immune modulation (after transplantation)

Sequelae of toxic inhalation or aspiration — TEF
- Chlorine — Smoke inhalation: fire, COPD
- Overdose (heroin)
- Foreign body — Obstruction: tumor, foreign body, LAD

Rheumatic conditions
- Rheumatoid arthritis
- Systemic lupus erythematosus
- Sjögren's syndrome
- Relapsing polychondritis

Other
- Inflammatory bowel disease (chronic ulcerative colitis or Crohn's disease) — Diffuse panbronchiolitis, Constrictive bronchiolitis
- Young's syndrome (secondary ciliary dyskinesia)
- Yellow nail syndrome (yellow nails and lymphedema)

Etiology based on disease location

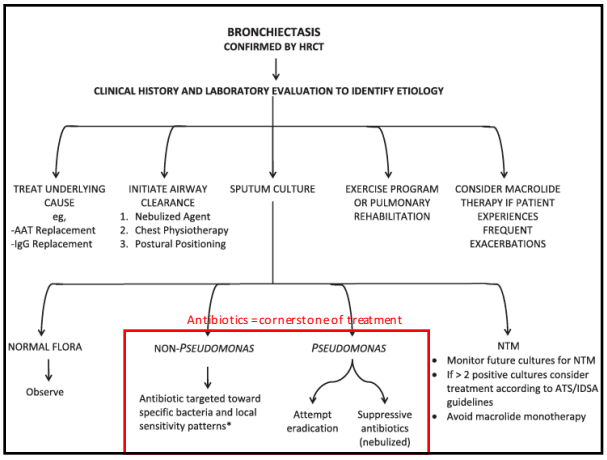
Disease Location	Possible Etiology
Focal bronchiectasis	Broncholithiasis Endobronchial neoplasm Foreign body Congenital bronchial atresia Mucus plugging
Upper lung	Cystic fibrosis Sarcoidosis Post-tuberculosis bronchiectasis
Central lung	ABPA
Right middle lobe and lingula	Atypical mycobacterial infection (e.g., MAI) Right middle lobe syndrome Immotile cilia syndrome (primary ciliary disease)
Lower lung	Chronic aspiration Usual interstitial pneumonia Hypogammaglobulinemia Recurrent infections α_1-antitrypsin deficiency

TABLE 2. DIAGNOSTIC EVALUATION OF THE PATIENT WITH BRONCHIECTASIS

Bacterial and mycobacterial sputum culture
Immunoglobulins A, E, G, and M
Titers to pneumococcal vaccine
ANA, RF, aCCP, SSA, SSB antibodies
α_1-Antitrypsin level and phenotype
In some cases:
- Bronchoscopy
- Gastrointestinal evaluation

First exclude cystic fibrosis
- Sweat test: < 40 normal, 40 – 60 borderline, > 60 abnormal
- CFTR mutation analysis (most common genes) or full sequencing
- Nasal PD, semen analysis

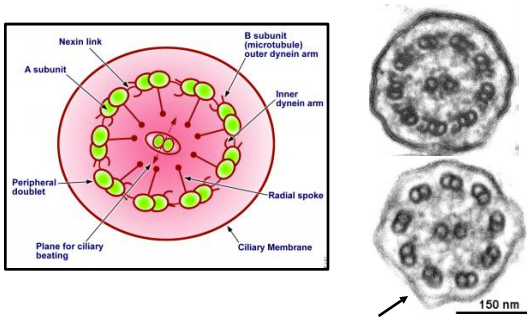
Diagnosis of PCD
- Nasal nitric oxide testing can be used as a screening test – levels will be low
- Carinal or nasal biopsy with electron microscopy examination of cilia is the gold standard for diagnosis
- Nasal scrapings at specialized research centers
- Genetic testing – mutations in the *DNAI1* and *DNAH5* genes account for up to 30 % patients, extended genetic testing encompasses 70 % patients



Management Pearls

- DNAse: patients treated for 24 weeks (BID dosing) had increased number of pulmonary exacerbations and greater decline in FEV1 – not typically used
- Corticosteroids: not routinely used unless they are part of the treatment for underlying cause (ABPA)
- Antibiotics during an exacerbation
  - Typically chosen based on prior culture data
  - Consider choosing something with activity against PA if no prior culture data
  - British Thoracic Society recommends combination antibiotics for MDR PA
  - Duration of antibiotics is not clear cut but 2 weeks is typically recommended

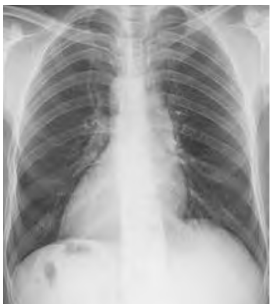
Primary Ciliary Dyskinesia



Absence of inner and outer dynein arms

Kartagener's syndrome

- Triad of bronchiectasis, rhinosinusitis, and situs inversus
- Accounts for about 50% of patients with PCD



Bronchiolar Disorders

- Diffuse panbronchiolitis
  - Idiopathic inflammatory disease
  - Most well recognized in Japan
  - Coexisting sinusitis
  - Treatment with macrolides
- Constrictive bronchiolitis
  - Post viral
  - GVHD

Tracheobronchomegaly (Mounier Kuhn Syndrome)

- Diameter of trachea > 3 cm
- Diameter of right main bronchus > 2.4 cm
- Diameter of left main bronchus > 2.3 cm
- Inefficient cough mechanism leads to impaired mucociliary clearance

Yellow Nail Syndrome

- malformations affecting the fingernails and toenails
- Pleural effusions, recurrent sinopulmonary infections, bronchiectasis
- lymphedema

Young's Syndrome

- Bronchiectasis, sinusitis, and obstructive azoospermia
- Need to exclude cystic fibrosis and PCD

Cartilage deficiency (Williams-Campbell)

- absence of cartilage in subsegmental bronchi
- Leads to distal airway collapse

Job's Syndrome

- Autosomal dominant hyper-IgE syndrome
- triad of eosinophilia, eczema, and recurrent skin and pulmonary infections

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