Bronchiectasis

- irreversible airway dilation that involves the lung in either a focal or a diffuse manner
- classically has been categorized as
  - cylindrical or tubular (the most common form)
  - varicose
  - cystic

Etiology

- Infectious or non infectious
- Pattern of involvement gives clues
- ***Focal*** bronchiectasis
  - bronchiectatic changes in a localized area of the lung
  - consequence of ***obstruction of the airway***
    - either extrinsic (e.g., due to compression by adjacent lymphadenopathy or parenchymal tumor mass)
    - intrinsic (e.g., due to an airway tumor or aspirated foreign body, a scarred/stenotic airway, or bronchial atresia from congenital underdevelopment of the airway).
- ***Diffuse*** bronchiectasis
  - characterized by widespread bronchiectatic changes throughout the lung
  - often arises from an ***underlying systemic or infectious disease process***
- ***upper*** lung fields
  - is most ***common in CF***
  - also is observed in ***postradiation fibrosis***, corresponding to the lung region encompassed by the radiation port
- ***lower*** lung fields
  - usually has its source in ***chronic recurrent aspiration*** (e.g., due to esophageal motility disorders like those in scleroderma),
  - end-stage ***fibrotic lung disease*** (e.g., traction bronchiectasis from idiopathic pulmonary fibrosis),
  - or ***recurrent immunodeficiency-associated associated infections*** (e.g., hypogammaglobulinemia)
- ***Midlung*** fields
  - Non-tubercular mycobacteria
  - Mycobacterium avium-intracellulare complex (***MAC***) is most common
  - Congenital ***dyskinetic/ immotile cilia syndrome***
- ***Central*** airways
  - association with allergic bronchopulmonary aspergillosis (***ABPA***), in which an immune-mediated reaction to Aspergillus damages the bronchial wall
  - Congenital cartilage deficiency
    - include tracheobronchomegaly (Mounier-Kuhn syndrome)
    - Williams-Campbell syndrome.
- 25-30% have idiopathic disease with no clear etiology
- ***Traction*** bronchiectasis refers to dilated airways arising from parenchymal distortion as a result of lung fibrosis (e.g., postradiation fibrosis or idiopathic pulmonary fibrosis).

TABLE 290-1 Major Etiologies of Bronchiectasis and Proposed Workup

PATTERN OF LUNG INVOLVEMENT	ETIOLOGY BY CATEGORY (EXAMPLES)	WORKUP
Focal	Obstruction (aspirated foreign body, tumor mass)	Chest imaging (chest x-ray and/or chest CT); bronchoscopy
Diffuse	Infection (bacterial, nontuberculous mycobacterial)	Sputum Gram's stain/culture; stains/cultures for acid-fast bacilli and fungi. If no pathogen is identified, consider bronchoscopy with bronchoalveolar lavage.
	Immunodeficiency (hypogammaglobulinemia, HIV infection, bronchiolitis obliterans after lung transplantation)	Complete blood count with differential; immunoglobulin measurement; HIV testing
	Genetic causes (cystic fibrosis, Kartagener's syndrome, α_1 antitrypsin deficiency)	Measurement of chloride levels in sweat (for cystic fibrosis), α_1 antitrypsin levels; nasal or respiratory tract brush/biopsy (for dyskinetic/immotile cilia syndrome); genetic testing
	Autoimmune or rheumatologic causes (rheumatoid arthritis, Sjögren's syndrome, inflammatory bowel disease); immune-mediated disease (allergic bronchopulmonary aspergillosis)	Clinical examination with careful joint exam, serologic testing (e.g., for rheumatoid factor). Consider workup for allergic bronchopulmonary aspergillosis, especially in patients with refractory asthma.[a]
	Recurrent aspiration	Test of swallowing function and general neuromuscular strength
	Miscellaneous (yellow nail syndrome, traction bronchiectasis from postradiation fibrosis or idiopathic pulmonary fibrosis)	Guided by clinical condition

Idiopathic	Exclusion of other causes

[a]Skin testing for *Aspergillus* reactivity; measurement of serum precipitins for *Aspergillus*, serum IgE levels, serum eosinophils, etc.

Epidemiology

- patients with CF develop clinical bronchiectasis in late adolescence or early adulthood
- atypical presentations of CF in adults in their thirties and forties are also possible.
- MAC infection classically affects nonsmoking women >50 years of age
- Incidence increases with age
- F > M
- May be co-diagnosed with COPD or asthma
- tuberculosis is prevalent, bronchiectasis occurs as a sequela of granulomatous infection
  - ***Focal*** bronchiectasis from ***extrinsic*** compression by enlarged granulomatous lymph nodes and/or from ***intrinsic*** obstruction as a result of erosion of a calcified lymph node through the airway wall (e.g., broncholithiasis)
  - reactivated tuberculosis, parenchymal destruction from infection can result in areas of more ***diffuse*** bronchiectasis.

Pathogenesis

- most accepted is ***Vicious cycle hypothesis***
  - susceptibility to infection and poor mucociliary clearance result in microbial colonization of the bronchial tree.
  - Pseudomonas aeruginosa, exhibit a particular propensity for colonizing damaged airways and evading host defense mechanisms
- Impaired mucociliary clearance
  - inherited conditions such as CF or dyskinetic cilia syndrome
  - Single severe infection (eg pneumonia caused by Bordetella pertussis or Mycoplasma pneumoniae) can cause airway damage and poor secretion clearance
  - bacterial mediators
- microbes incites continued chronic inflammation, with consequent damage to the airway wall, continued impairment of secretions and microbial clearance, and ongoing propagation of the ***infectious/ inflammatory cycle***
- Pathology
  - significant small-airway wall inflammation and larger-airway wall destruction as well as dilation, with loss of elastin, smooth muscle, and cartilage
  - cells in the small airways release proteases and other mediators, such as reactive oxygen species and proinflammatory cytokines, which cause damage
- Inflammation causes small airway obstruction
- α1 antitrypsin deficiency. loses protective action
- noninfectious bronchiectasis
  - include immune-mediated reactions that damage the bronchial wall
  - associated with systemic autoimmune conditions such as Sjögren's syndrome and rheumatoid arthritis

Clinical manifestations

- persistent productive cough with ongoing production of thick, tenacious sputum. is most common
- crackles and wheezing on lung auscultation
- some patients with bronchiectasis exhibit clubbing of the digits.
- Mild to moderate airflow obstruction often is detected on pulmonary function tests, overlapping with others like COPD
- Acute exacerbations
  - changes in the nature of sputum production, with increased volume and purulence
  - Typical signs and symptoms of infection like fever and infiltrates may not be present

Diagnosis

- persistent chronic cough and sputum production accompanied by consistent radiographic features.
- CXRAY
  - lack sensitivity
  - presence of “tram tracks” indicating dilated airways is consistent with bronchiectasis
- CT CHEST
  - More specific
  - Imaging of choice
  - airway dilation
    - detected as parallel “tram tracks” or
    - as the “signet-ring sign”—a cross-sectional area of the airway with a diameter at least 1.5 times that of the adjacent vessel
  - lack of bronchial tapering (including the presence of tubular structures within 1 cm from the pleural surface),
  - bronchial wall thickening in dilated airways,
  - inspissated secretions (e.g., the “tree-in-bud” pattern)
  - cysts emanating from the bronchial wall (especially pronounced in cystic bronchiectasis)

Approach to patient

- elicitation of a clinical history, chest imaging, and a workup to determine the underlying etiology.
- Evaluation of ***focal*** bronchiectasis almost always requires ***bronchoscopy*** to exclude airway obstruction by an underlying ***mass*** or foreign body
- If diffuse look for other etiologies especially CF

Treatment

- directed at the control of active infection and improvements in secretion clearance and
- bronchial hygiene so as to decrease the microbial load
- Minimize risk of repeated infection

Antibiotic

- Target causative or presumed pathogen
- Haemophilus influenzae and P. aeruginosa isolated commonly
- 7-10 days in acute exacerbations upto 14 days
- MAC
  - diagnostic criteria for true clinical infection with NTM should be considered in patients with symptoms and radiographic findings of lung disease who have at least two sputum samples positive on culture; at least one bronchoalveolar lavage (BAL) fluid sample positive on culture; a biopsy sample displaying histopathologic features of NTM infection (e.g., granuloma or a positive stain for acid-fast bacilli) along with one positive sputum culture; or a pleural fluid sample (or a sample from another sterile extrapulmonary site) positive on culture.
  - HIV negative MAC treat with macrolide and rifampin and ethambutol

Bronchial Hygiene

- hydration and mucolytic administration
- aerosolization of bronchodilators and hyperosmolar agents (e.g., hypertonic saline)
- chest physiotherapy
  - postural drainage,
  - traditional mechanical chest percussion via hand clapping to the chest, or
  - use of devices such as an oscillatory positive expiratory pressure flutter valve or a high-frequency chest wall oscillation vest)
- mucolytic ***dornase*** (DNase) is recommended routinely in CF-related bronchiectasis but not in non-CF bronchiectasis

Anti-inflammatory therapy

- small-scale trials have yielded evidence of alleviated dyspnea, decreased need for inhaled β-agonists, and reduced sputum production with ***inhaled glucocorticoids***
- No difference in lung function or exacerbations
- Risks of immunosuppression and adrenal suppression
- Anti inflammatory oral or inhaled glucocorticoids
  - ABPA
  - Active autoimmune cause
- ABPA also give long course of ***Itraconazole***

Refractory cases

- Surgery with focal resection of suppuration
- Lung transplant in advance cases

Complications

- microbial resistance to antibiotics
- Antimicrobial toxicity
- Superficial mucosal vessel injury due to infection causing bleeding and life threatening hemoptysis

Prognosis

- Depends on underlying etiology and comorbidities
- Frequency of exacerbations and pathogen involved
- P. aeruginosa has worse prognosis

Prevention

- Smoking cessation
- reverse immunodeficient state
- Vaccination for influenza and pneumococcal
- if >3 episode per year
  - suppressive antibiotics to minimize the microbial load and reduce the frequency of exacerbations
- Suppressive strategies
  - administration of an oral antibiotic (e.g., ciprofloxacin) daily for 1–2 weeks per month
  - use of a rotating schedule of oral antibiotics (to minimize the risk of development of drug resistance)
  - administration of a macrolide antibiotic daily or three times per week
    - benefit related to non-antimicrobial properties, such as anti-inflammatory effects and reduction of gram-negative bacillary biofilms
    - 6-12 month course useful
    - more risk of NTM
    - rule out
      - NTM infection
      - QT prolongation
  - inhalation of aerosolized antibiotics (e.g., tobramycin inhalation solution) for select patients on a rotating schedule (e.g., 30 days on, 30 days off), with the goal of decreasing the microbial load without eliciting the side effects of systemic drug administration;
  - intermittent administration of IV antibiotics (e.g., “clean-outs”) for severe disease or resistant pathogen