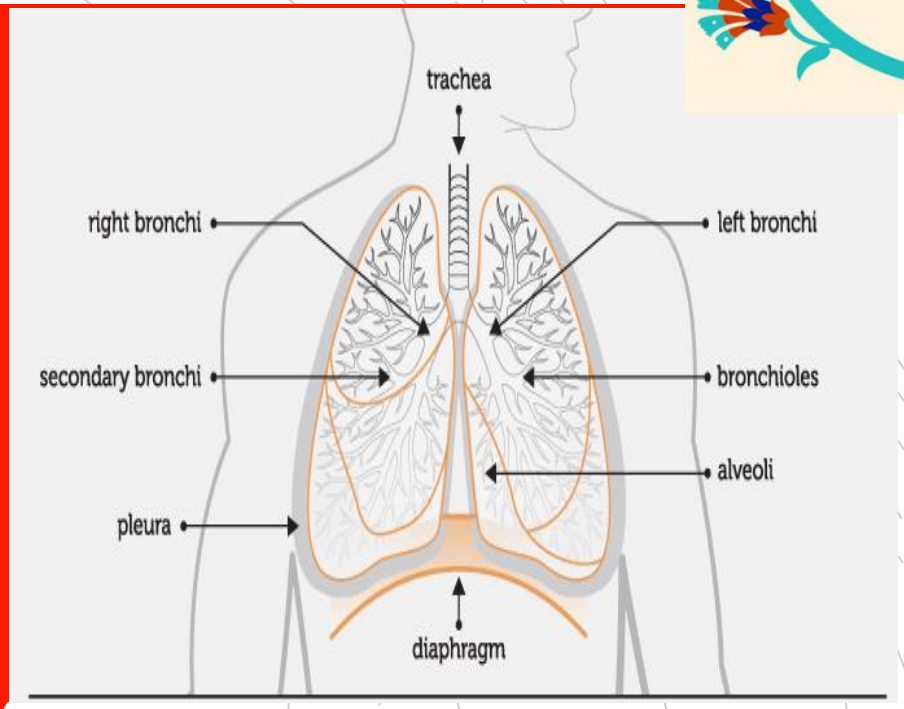
مرکز آموزشی درمانی افضلی پور

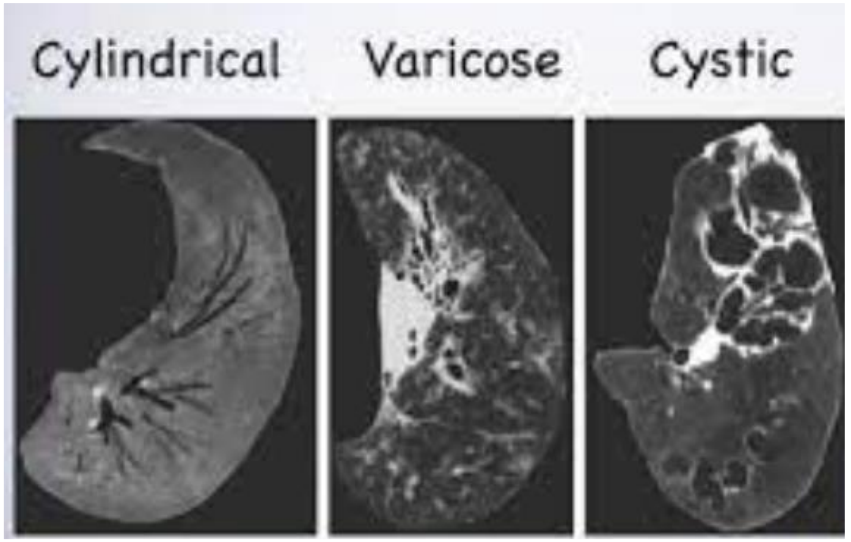
دانشگاه علوم پزشکی کرمان

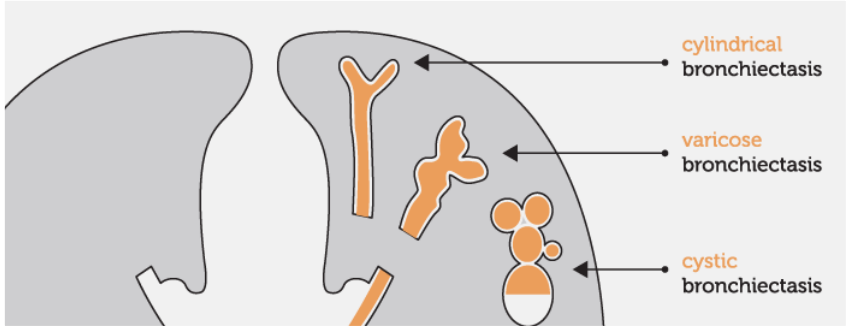
Bronchiectasis



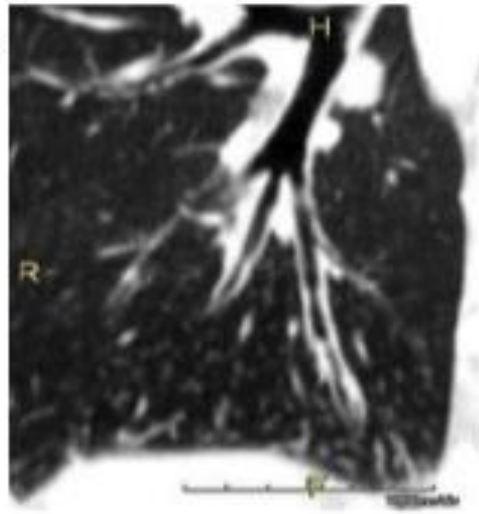
DR. SHAFIEPOUR/PULMONOLOGIST

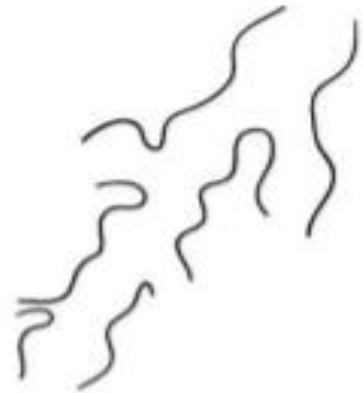
Bronchiectasis refers to an irreversible airway dilation that involves the lung in either a focal or a diffuse manner and that classically has been categorized as cylindrical or tubular (the most common form), varicose, or cystic



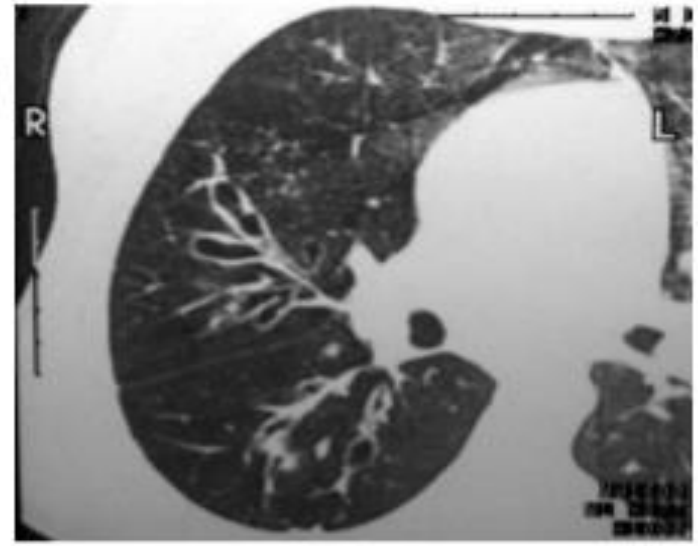


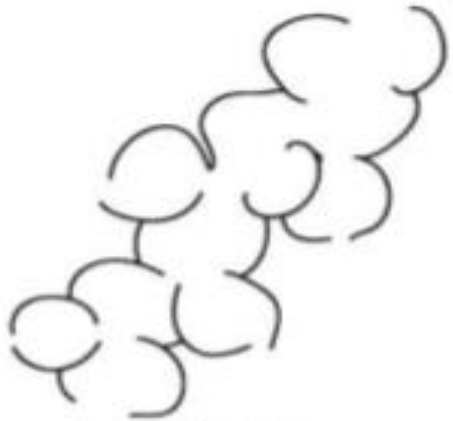
Cylindrical



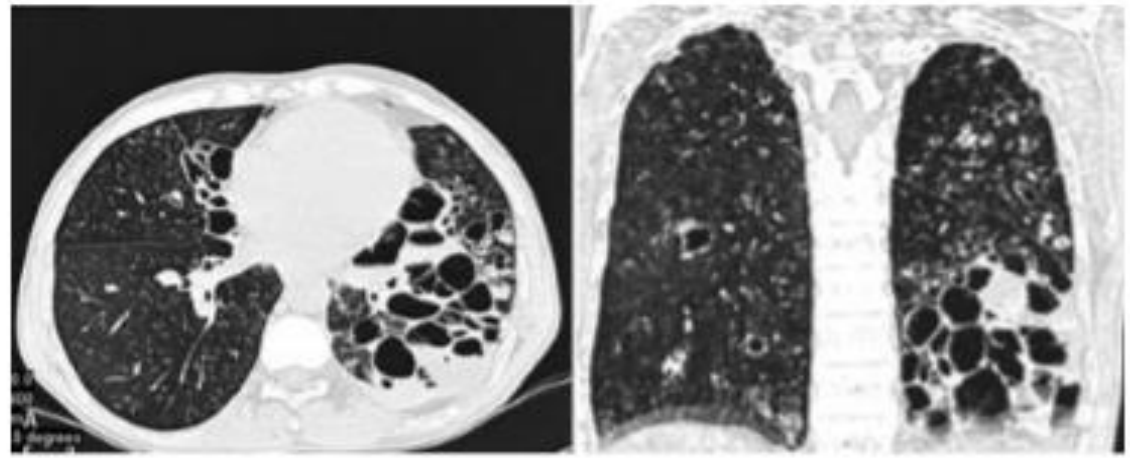


Varicose



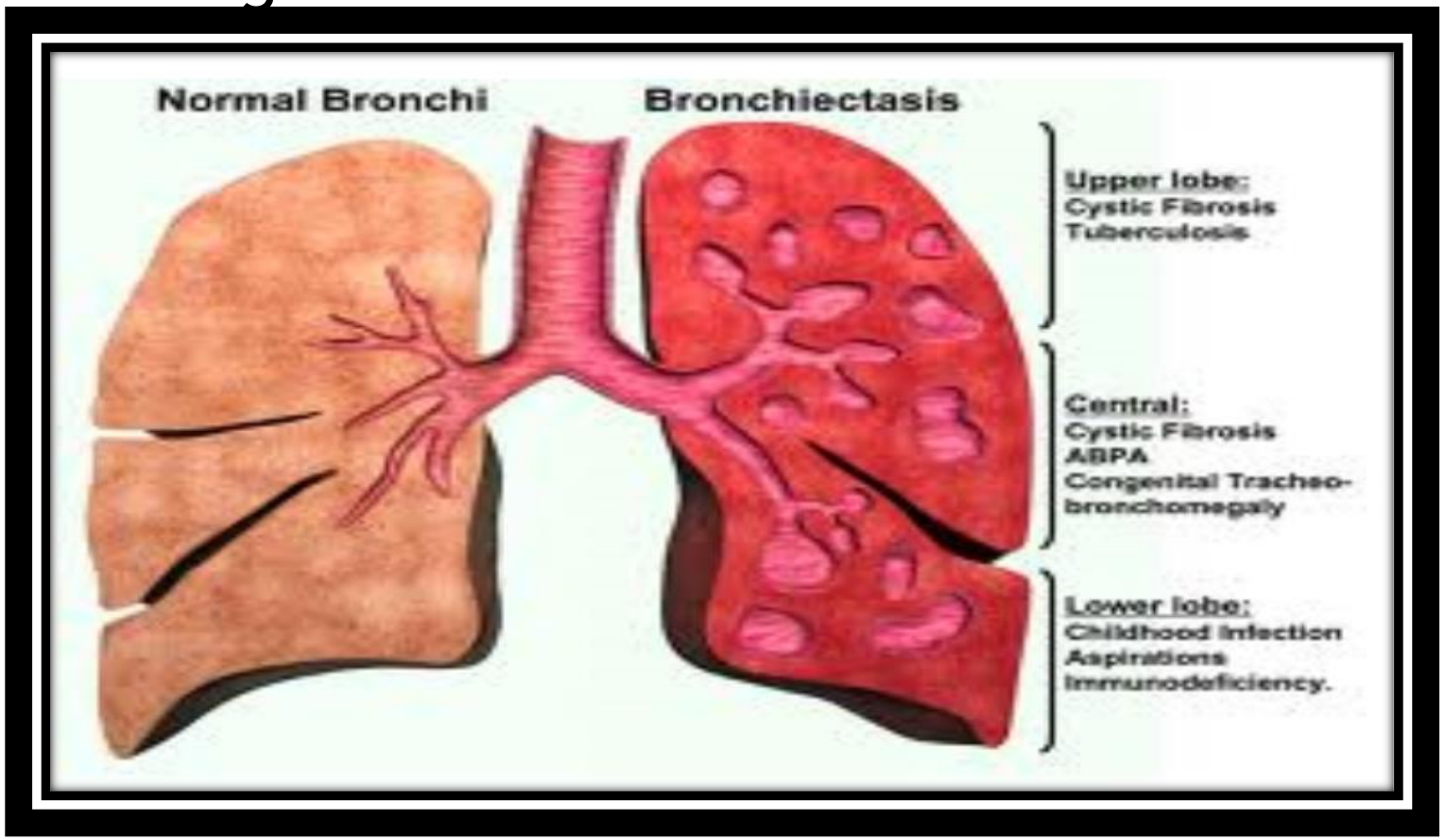


Cystic



Etiology: Bronchiectasis can arise from infectious or noninfectious causes

Clues to the underlying etiology often are provided by the pattern of lung involvement.



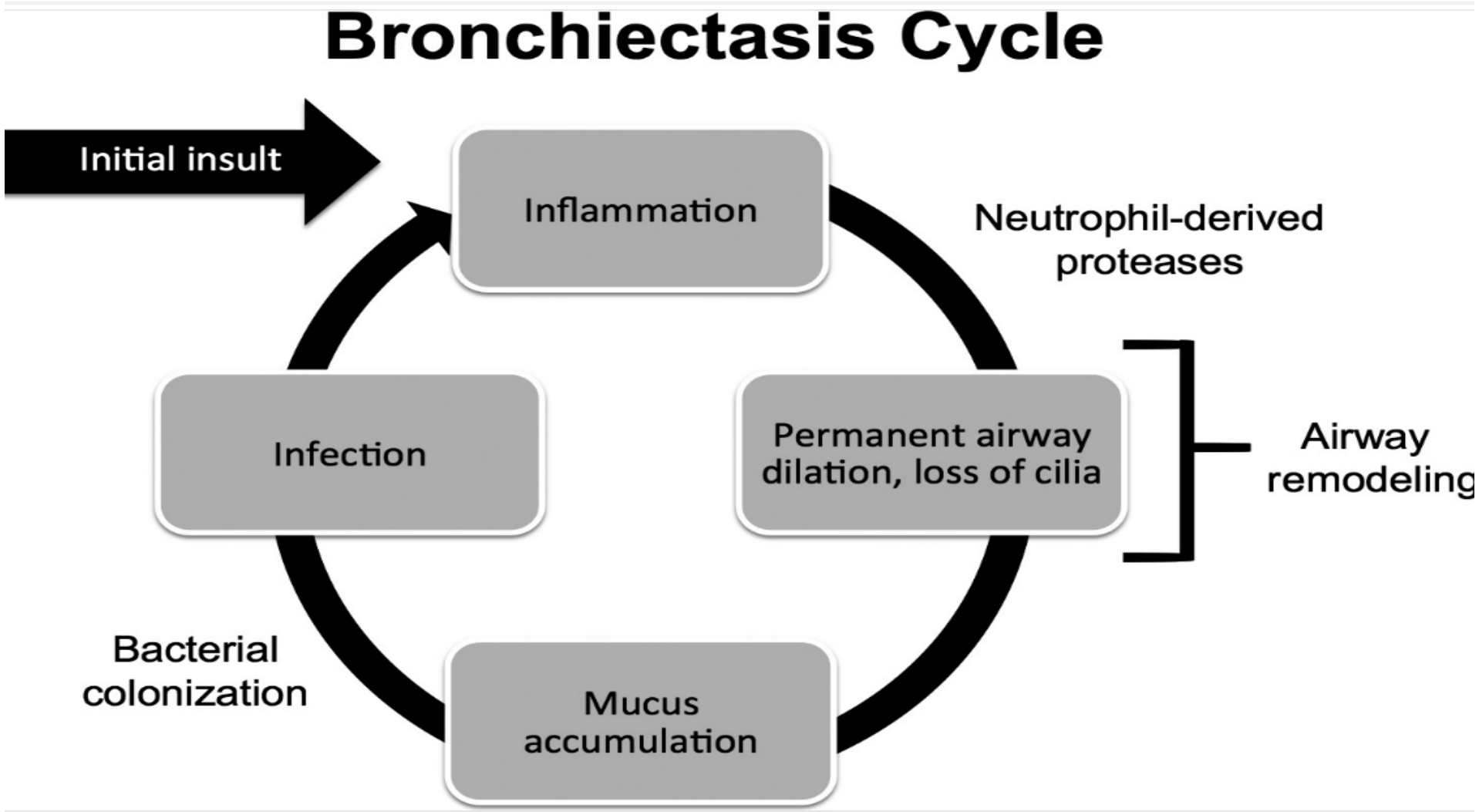
Major Etiologies of Bronchiectasis and Proposed Workup

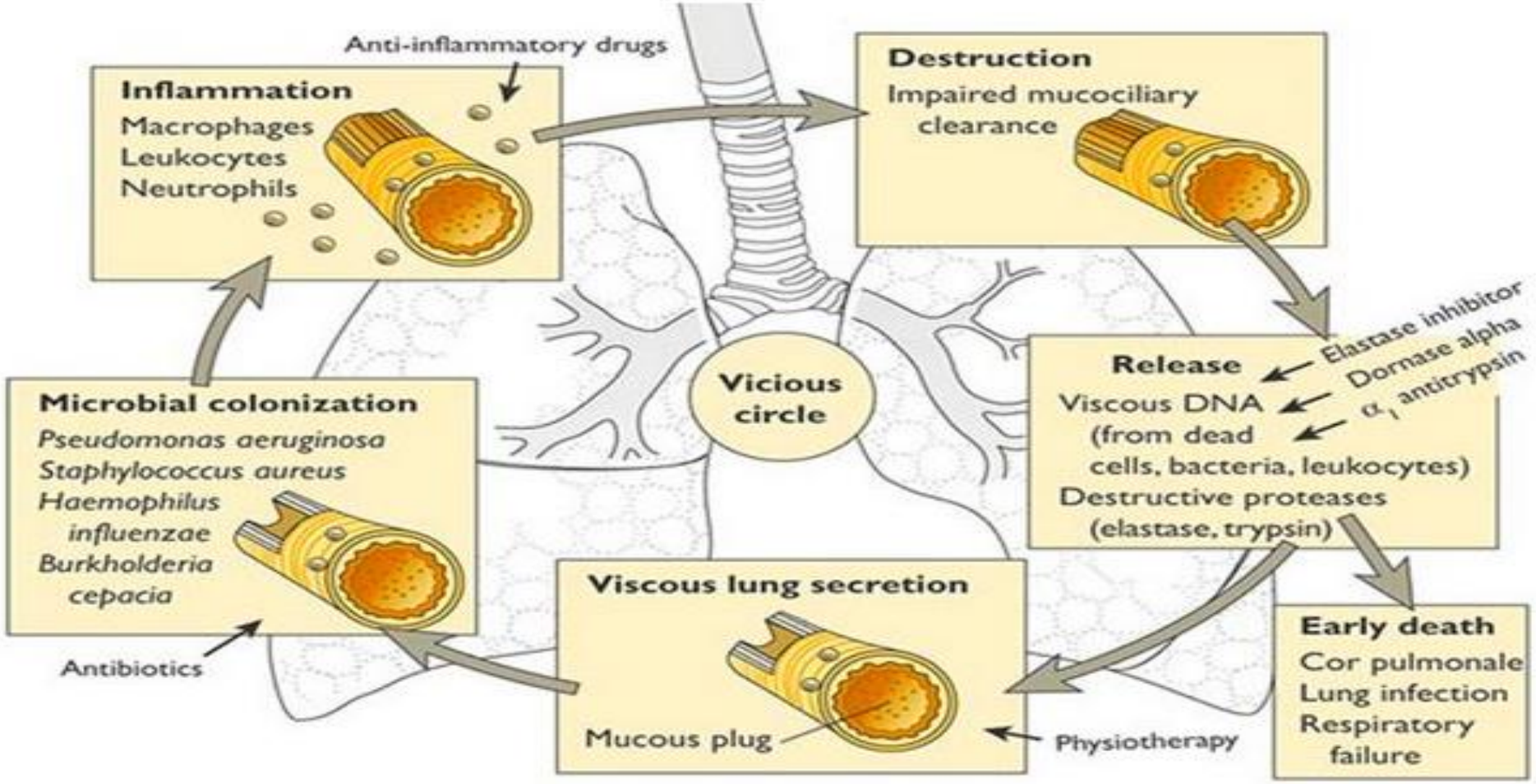
ETIOLOGY BY CATEGORY (EXAMPLES)	WORKUP
Obstruction (aspirated foreign body, tumor mass)	Chest imaging (chest x-ray and/or chest CT); bronchoscopy
Infection (bacterial, nontuberculous mycobacterial)	Sputum Gram's stain/culture; stains/cultures for acid-fast bacilli and fungi. If no pathogen is identified, 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

EPIDEMIOLOGY:

the epidemiology of bronchiectasis varies greatly with the underlying etiology. For example, patients born with CF often develop significant clinical bronchiectasis in late adolescence or early adulthood, although atypical presentations of CF in adults in their thirties and forties are also possible. In contrast, bronchiectasis resulting from MAC infection classically affects nonsmoking women >50 years of age. In general, the incidence of bronchiectasis increases with age. Bronchiectasis is more common among women than among men. Bronchiectasis may also frequently be co-diagnosed with chronic obstructive pulmonary disease (COPD) or asthma.

PATHOGENESIS AND PATHOLOGY:




Anti-inflammatory drugs
Inflammation
Macrophages
Leukocytes
Neutrophils
Destruction
Impaired mucociliary clearance
Vicious circle
Release
Viscous DNA (from dead cells, bacteria, leukocytes)
Destructive proteases (elastase, trypsin)
Elastase inhibitor
Dornase alpha
α_1 antitrypsin
Microbial colonization
Pseudomonas aeruginosa
Staphylococcus aureus
Haemophilus influenzae
Burkholderia cepacia
Antibiotics
Viscous lung secretion
Mucous plug
Physiotherapy
Early death
Cor pulmonale
Lung infection
Respiratory failure

CLINICAL MANIFESTATIONS :

The most common clinical presentation is a persistent productive cough with ongoing production of thick, tenacious sputum

Clinical presentation

- **The production of large quantities of purulent and often foul-smelling sputum.**

The volume of sputum can be used for estimating the severity of the disease

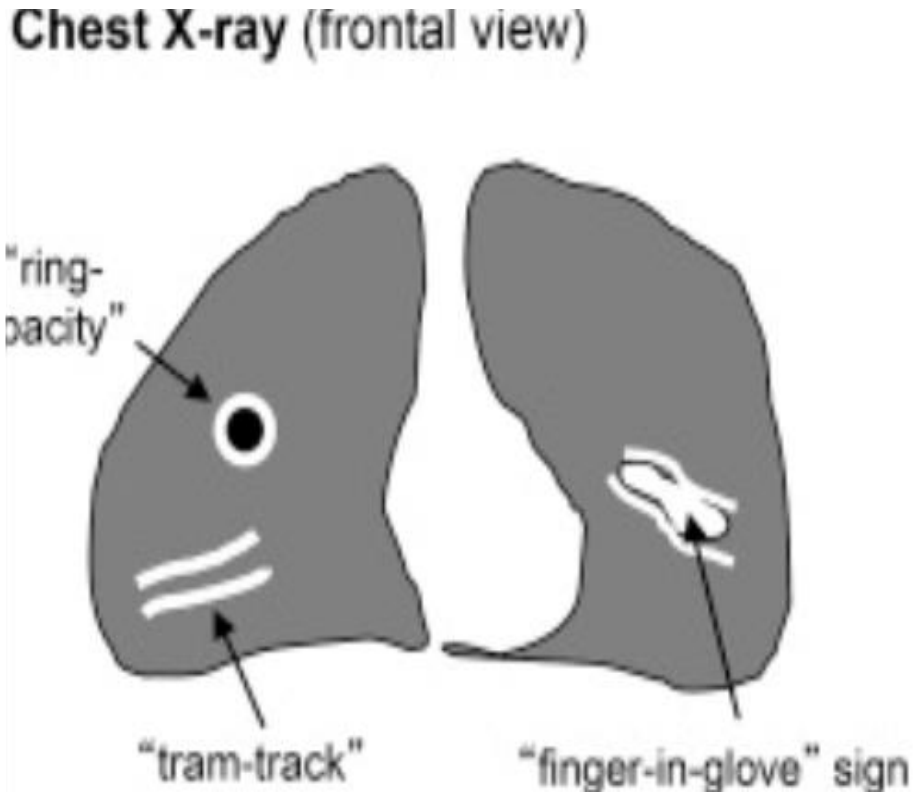
- Mild < 10 mL
- Moderate 10~150 mL
- Severe >150 mL

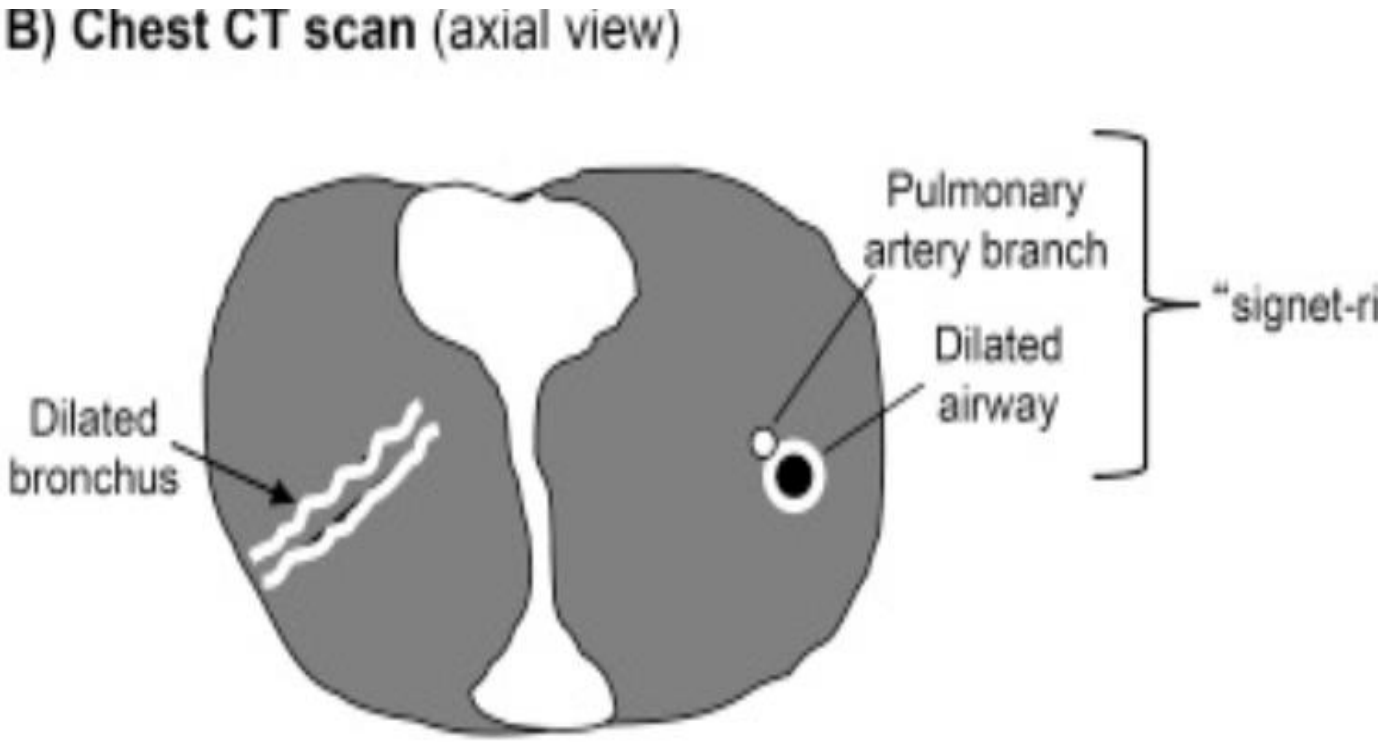
- **Dry bronchiectasis**

usually involve the upper lobes

DIAGNOSIS:

The diagnosis usually is based on presentation with a persistent chronic cough and sputum production accompanied by consistent radiographic features

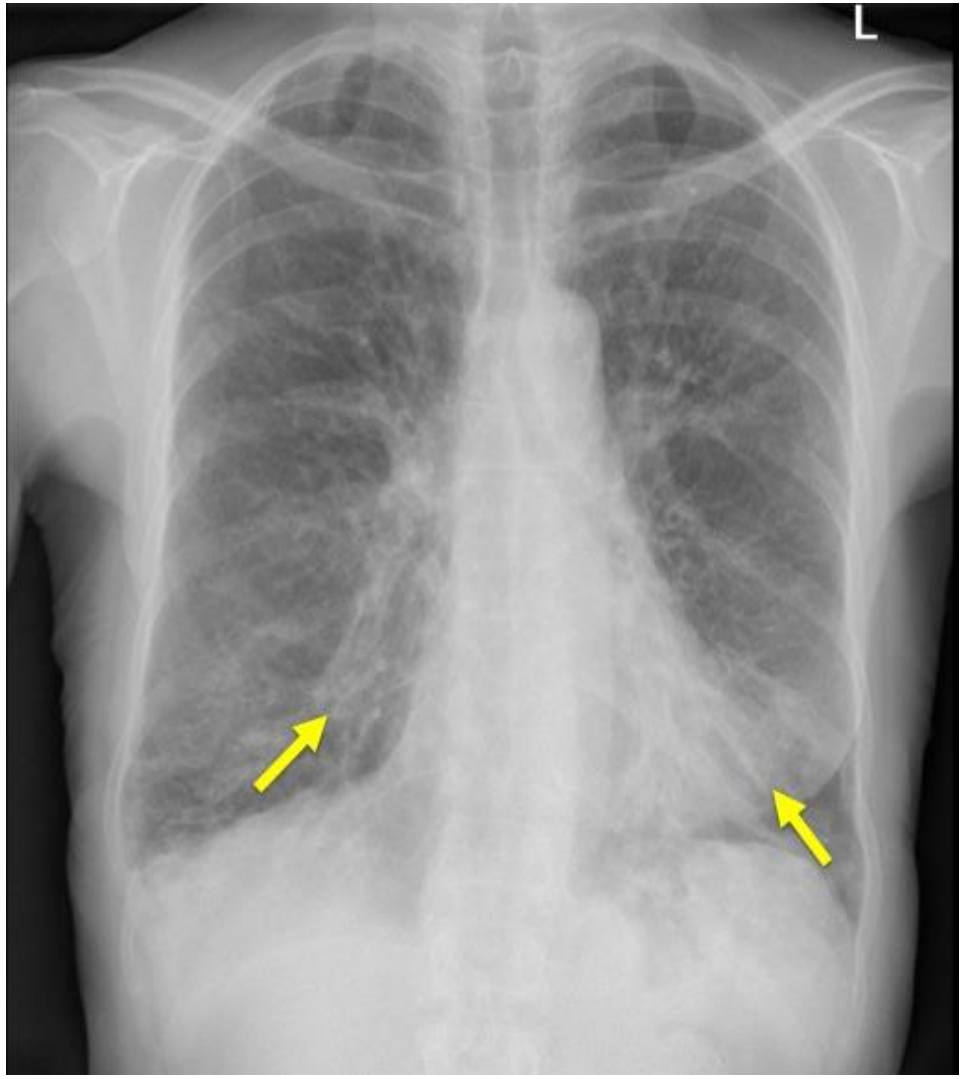




ADIL RADIOLOGY

Bronchiectasis

The chest radiograph is generally the first radiologic test requested by the pulmonologist when a diagnosis of bronchiectasis is suspected. Unfortunately, because there are key issues of sensitivity and specificity, the chest radiograph has limited value in the diagnosis of bronchiectasis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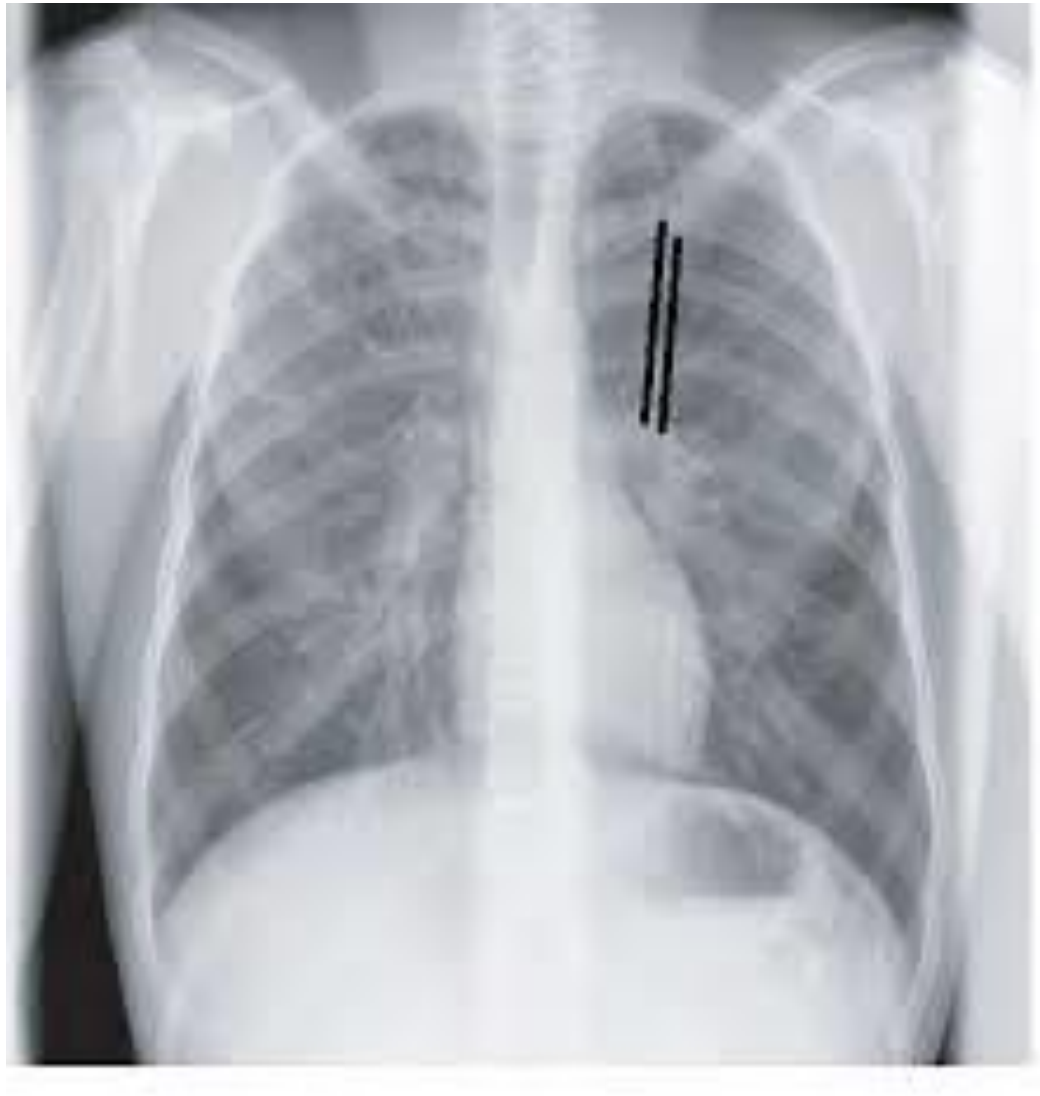


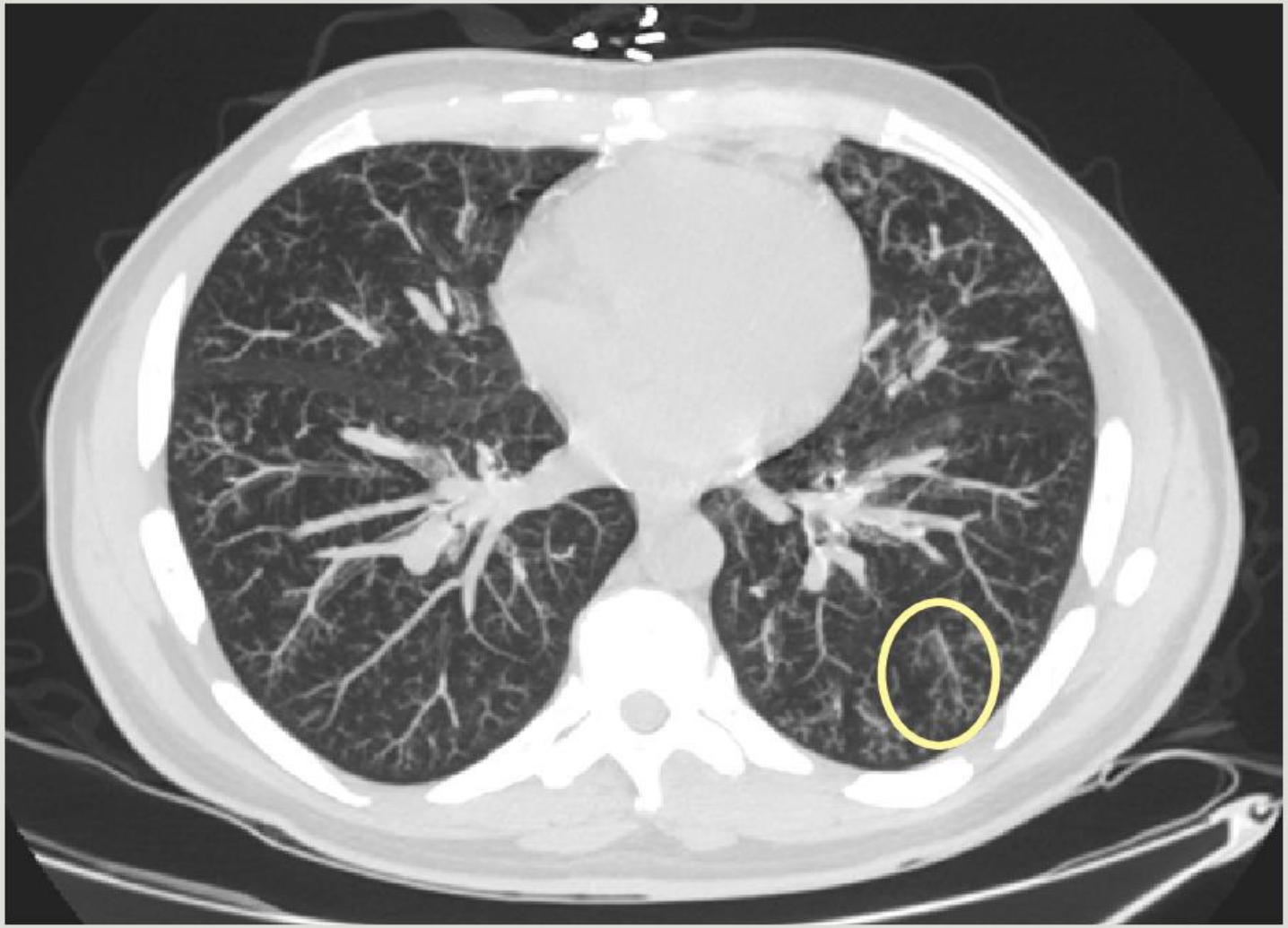


Figure 1: *CT scan with tree-in-bud pattern*

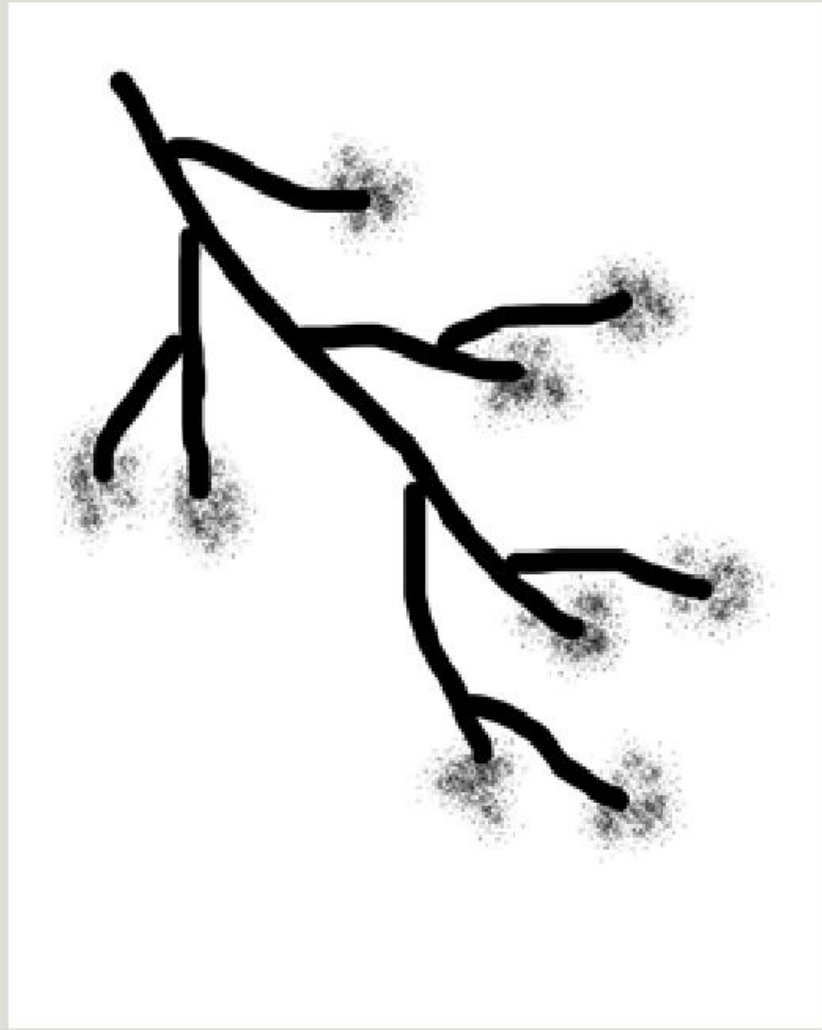


Figure 2: *Drawing rendition of tree-in-bud*

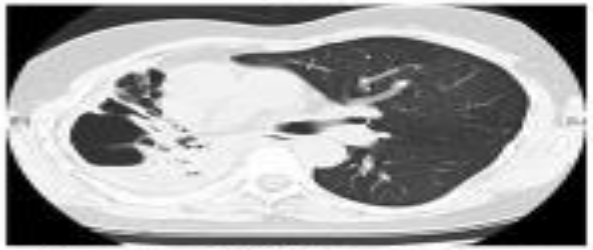
APPROACH TO THE PATIENT Bronchiectasis

Clinical suspicion of bronchiectasis

Bronchiectasis confirmed by HRCT

More *localized* bronchiectasis

Post-infectious (*e.g.*, TB)
Post-obstruction
Chronic aspiration
NTM lung disease*

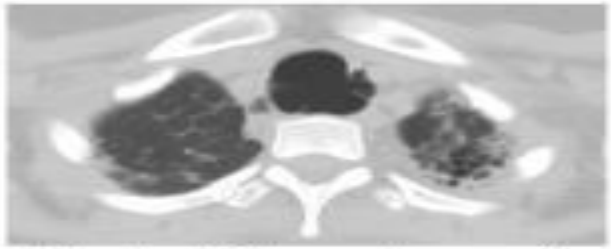


Remote TB in the right lung complicated by *M. abscessus* infection, destroying the right lung.

More *diffuse* bronchiectasis

Diagnosis highly suggestive by imaging

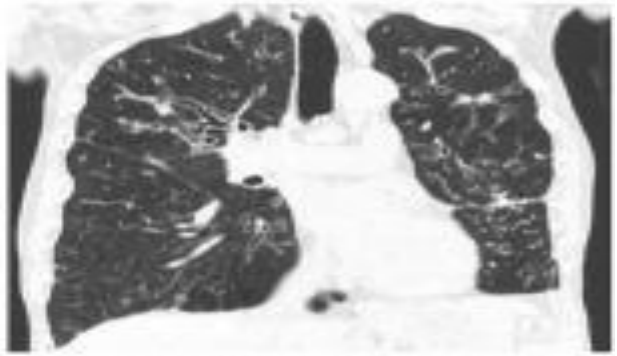
Mounier-Kuhn syndrome
Williams-Campbell syndrome
PCD with *situs inversus*



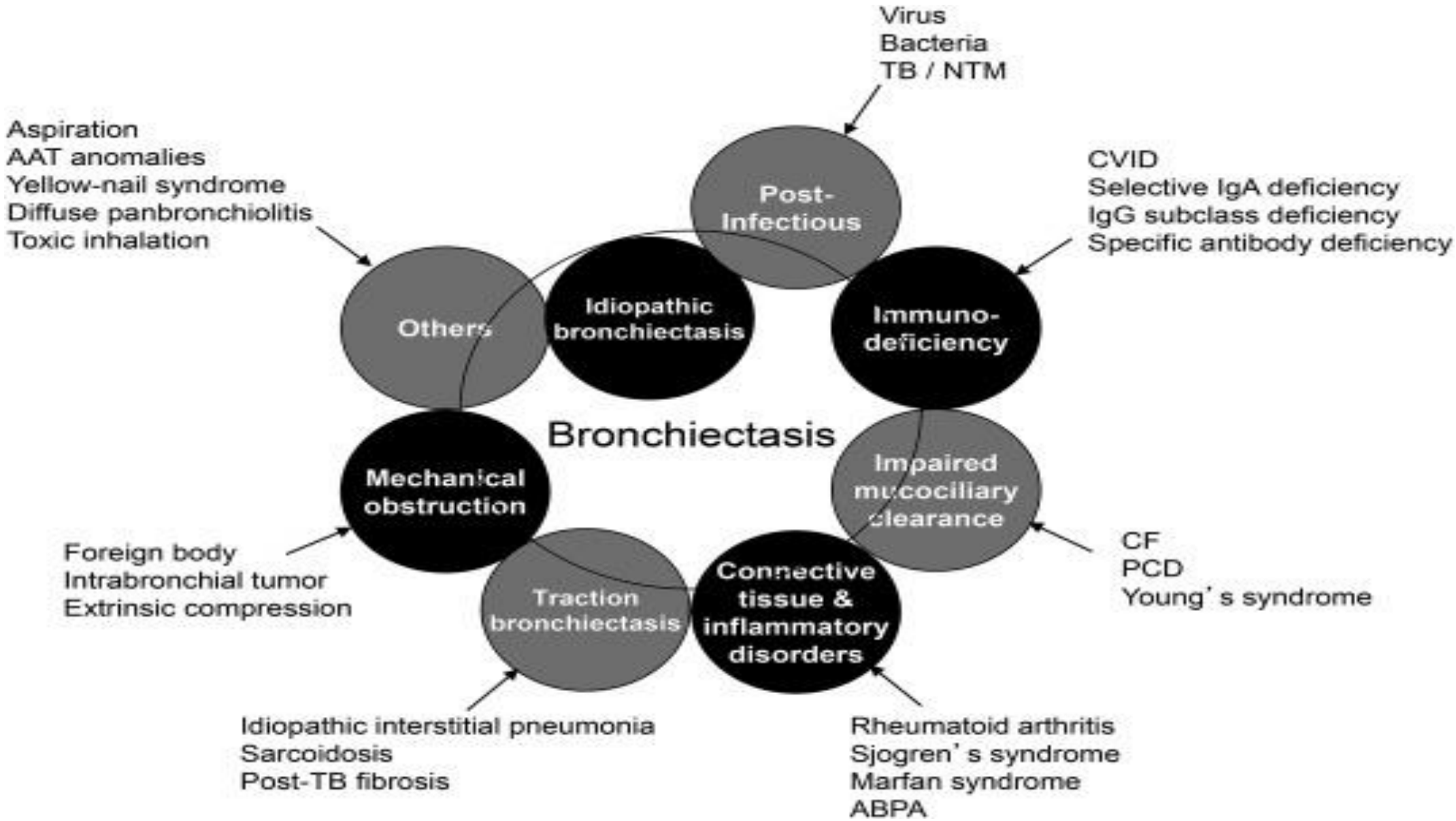
Mounier-Kuhn syndrome with tracheomegaly and tracheal diverticulae.

More specific biomarkers / diagnostics available

Cystic fibrosis
AAT deficiency
PCD, ABPA
CVID
NTM lung disease*



NTM lung disease in an adult woman with cystic fibrosis with five-lobe bronchiectasis and diffuse tree-in-bud opacities.


Virus
Bacteria
TB / NTM
Aspiration
AAT anomalies
Yellow-nail syndrome
Diffuse panbronchiolitis
Toxic inhalation
CVID
Selective IgA deficiency
IgG subclass deficiency
Specific antibody deficiency
Post-Infectious
Others
Idiopathic bronchiectasis
Immuno-deficiency
Bronchiectasis
Mechanical obstruction
Impaired mucociliary clearance
Foreign body
Intrabronchial tumor
Extrinsic compression
CF
PCD
Young' s syndrome
Traction bronchiectasis
Connective tissue & inflammatory disorders
Idiopathic interstitial pneumonia
Sarcoidosis
Post-TB fibrosis
Rheumatoid arthritis
Sjogren' s syndrome
Marfan syndrome
ABPA

Evaluation

- After the diagnosis of bronchiectasis, we need to identify
  - **potentially treatable causes**,
  - **microbiologic pathogens**, and
  - **functional assessment**

Evaluation

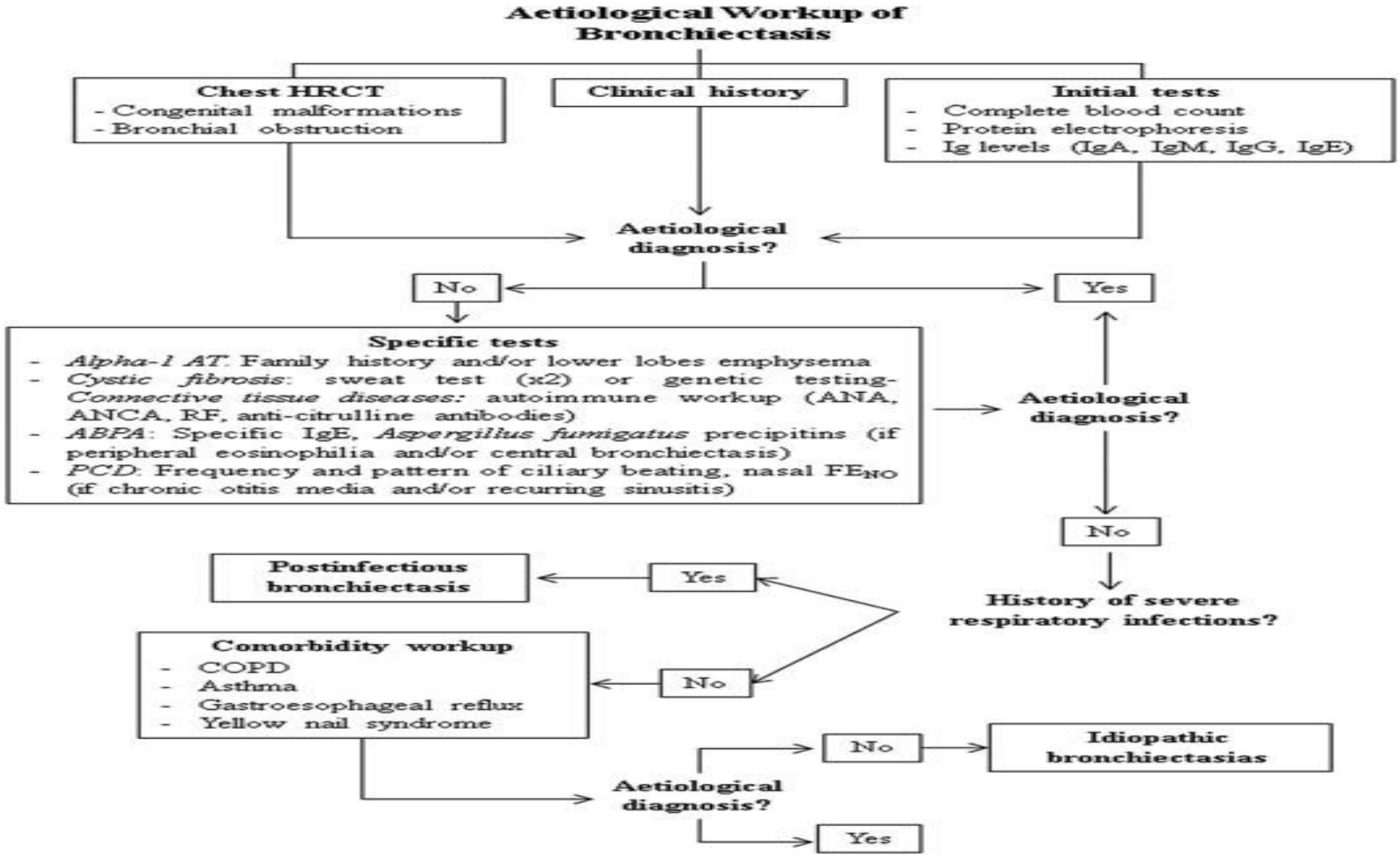
- Personal history: childhood symptoms, infertility, previous pneumonia/viral illness, gastric aspiration, asthma, joints pain or deformity
- Family history: primary ciliary dyskinesia (PCD) (**Kartagener's syndrome –** sinusitis, situs inversus and bronchiectasis), cystic fibrosis (CF)

Evaluation

- Investigations:
  - Full blood count with differential
  - Immunoglobulin levels (IgG, IgM and IgA), consider IgE when eosinophilia
  - Rheumatoid factor
  - Sputum smear and culture for bacteria, mycobacteria (NTM) and fungi (mainly *Aspergillus*)

Additional Tests

- Testing for cystic fibrosis
  - Sweat chloride (minimum 2 requirements) and/or mutation analysis of the cystic fibrosis transmembrane conductance regulator (CFTR) gene (if clinical suspicion high)
- Nasal nitric oxide test
- Alpha-1 antitrypsin level and genotype
- HIV antibody test
- Swallow study/pH monitoring
- Bronchoscopy


Aetiological Workup of Bronchiectasis
Chest HRCT
- Congenital malformations
- Bronchial obstruction
Clinical history
Initial tests
- Complete blood count
- Protein electrophoresis
- Ig levels (IgA, IgM, IgG, IgE)
Aetiological diagnosis?
No
Yes
Specific tests
- Alpha-1 AT: Family history and/or lower lobes emphysema
- Cystic fibrosis: sweat test (x2) or genetic testing- Connective tissue diseases: autoimmune workup (ANA, ANCA, RF, anti-citrulline antibodies)
- ABPA: Specific IgE, Aspergillus fumigatus precipitins (if peripheral eosinophilia and/or central bronchiectasis)
- PCD: Frequency and pattern of ciliary beating, nasal FENO (if chronic otitis media and/or recurring sinusitis)
Aetiological diagnosis?
No
History of severe respiratory infections?
Yes
Postinfectious bronchiectasis
Comorbidity workup
- COPD
- Asthma
- Gastroesophageal reflux
- Yellow nail syndrome
No
Aetiological diagnosis?
No
Idiopathic bronchiectasias
Yes

TREATMENT

ANTIBIOTIC TREATMENT

Antibiotics targeting the causative or presumptive pathogen (with Haemophilus influenzae and P. aeruginosa isolated commonly) should be administered in acute exacerbations, usually for a minimum of 7–10 days and perhaps for as long as 14 days

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.

BRONCHIAL HYGIENE

The numerous approaches used to enhance secretion clearance in bronchiectasis include hydration and mucolytic administration, aerosolization of bronchodilators and hyperosmolar agents (e.g., hypertonic saline), and chest physiotherapy (e.g., 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).

ANTI-INFLAMMATORY THERAPY

It has been proposed that control of the inflammatory response may be of benefit in bronchiectasis, and relatively small-scale trials have yielded evidence of alleviated dyspnea, decreased need for inhaled β-agonists, and reduced sputum production with inhaled glucocorticoids. However, no significant differences in lung function or bronchiectasis exacerbation rates have been observed

du**administration of oral/systemic glucocorticoids** due to certain etiologies, such as ABPA, or of noninfectious bronchiectasis due to underlying conditions, especially that in which an autoimmune condition is believed to be active (e.g., rheumatoid arthritis or Sjögren's syndrome). Patients with ABPA also may benefit from a prolonged course of treatment with the oral antifungal agent itraconazole.

REFRACTORY CASES

In select cases, surgery can be considered, with resection of a focal area of suppuration. In advanced cases, lung transplantation can be considered.

COMPLICATIONS

In more severe cases of infectious bronchiectasis, recurrent infections and repeated courses of antibiotics can lead to microbial resistance to antibiotics.

Recurrent infections can result in injury to superficial mucosal vessels, with bleeding and, in severe cases, life-threatening hemoptysis

PROGNOSIS

Outcomes of bronchiectasis can vary widely with the underlying etiology and comorbid conditions and also may be influenced by the frequency of exacerbations and (in infectious cases) the specific pathogens involved (with worse outcomes associated with P. aeruginosa colonization).

the decline of lung function in patients with non-CF bronchiectasis was similar to that in patients with COPD, with the forced expiratory volume in 1 s (FEV1) declining by 50–55 mL per year as opposed to 20–30 mL per year for healthy controls

PREVENTION

Reversal of an underlying immunodeficient state (e.g., by administration of gamma globulin for immunoglobulin-deficient patients) and vaccination of patients with chronic respiratory conditions (e.g., influenza and pneumococcal vaccines) can decrease the risk of recurrent infections. Patients who smoke should be counseled about smoking cessation.

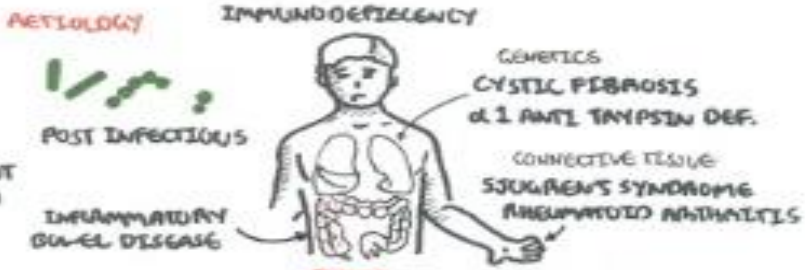
After resolution of an acute infection in patients with recurrences (e.g., ≥3 episodes per year), the use of suppressive antibiotics to minimize the microbial load and reduce the frequency of exacerbations has been proposed.

BRONCHIECTASIS

- IRREVERSIBLE DILATION OF THE AIRWAY DUE TO INFLAMMATORY DESTRUCTION OF AIRWAY WALLS RESULTING FROM PERSISTENTLY INFECTED MUCUS
- P. AURUGINOSA IS THE MOST COMMON PATHOGEN
- CLINICAL MANIFESTATION INCLUDE CHRONIC COUGH COPIOUS MUCOPURULENT EXPECTORATION
- COMMON IN PATIENTS WITH CYSTIC FIBROSIS

SIGNS AND SYMPTOMS

AETIOLOGY

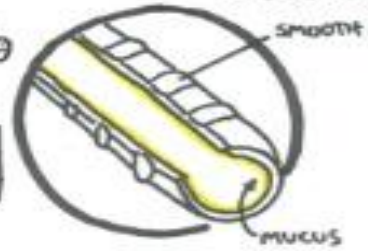


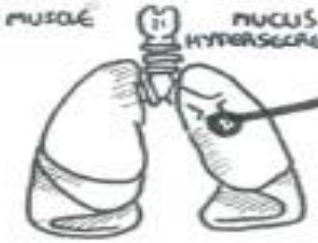
MANAGEMENT

TREAT UNDERLYING CONDITION (IE. CYSTIC FIBROSIS, COPD)

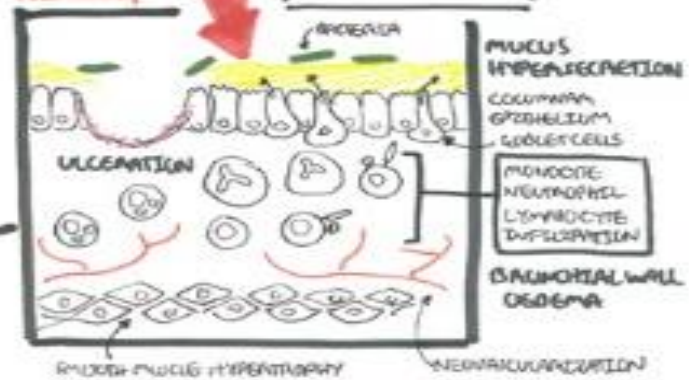
EXACERBATIONS - SEVERE

COMPLICATION



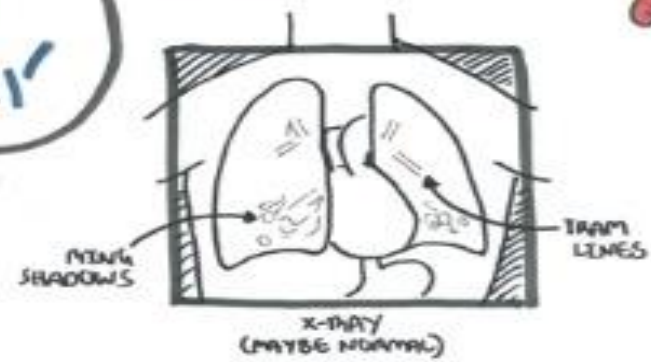


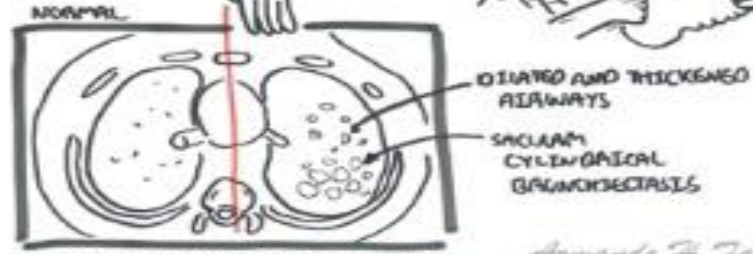
PATHOLOGY



MICROBIAL SPUTUM OF PATIENTS FREQUENTLY FOUND TO BE COLONIZED WITH PATHOGENIC MICROORGANISMS

INVESTIGATIONS





Armando H. Faigl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