Bronchiectasis

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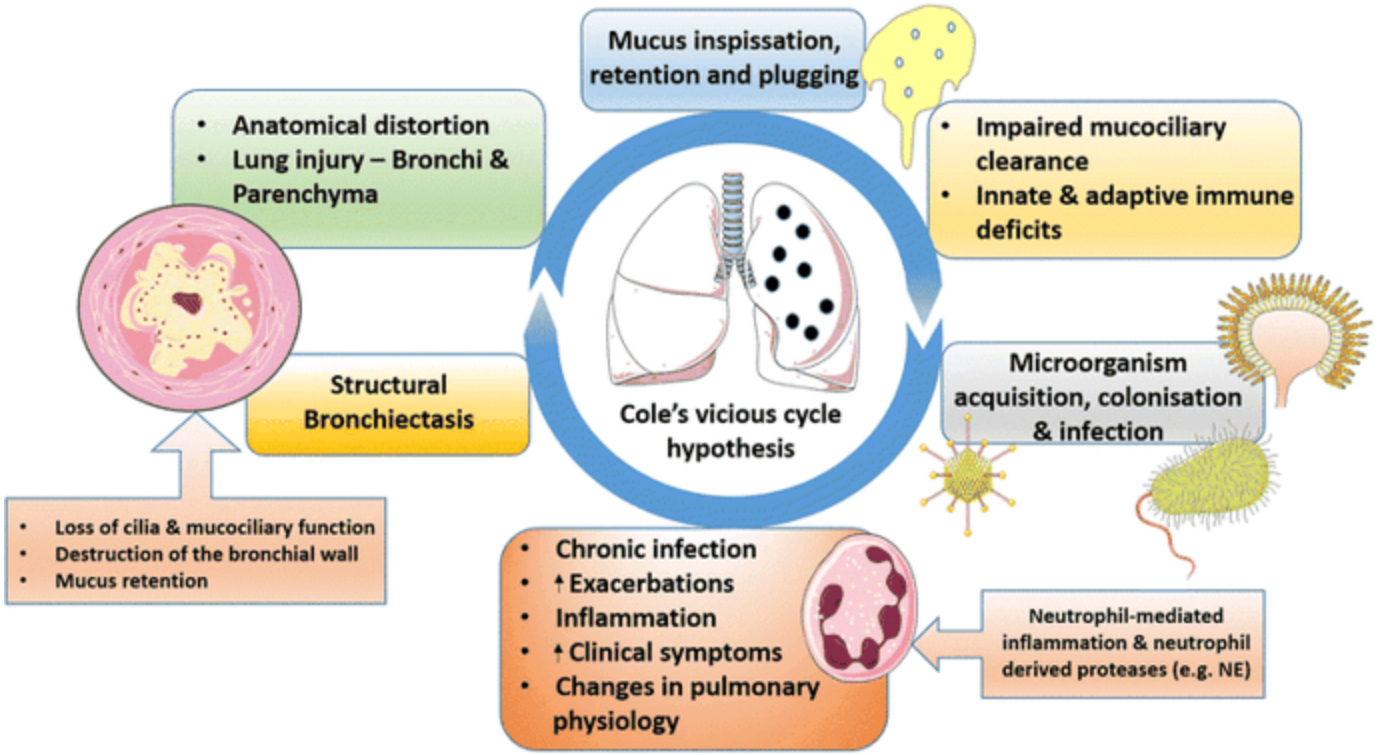
Definition

Chronic respiratory disease characterized by permanent and **abnormal dilatation** of the bronchi and bronchioli which destruct muscular and elastic components of bronchial walls.

Epidemiology

- approximately 40 /100.000

Bronchiectasis



Causes :

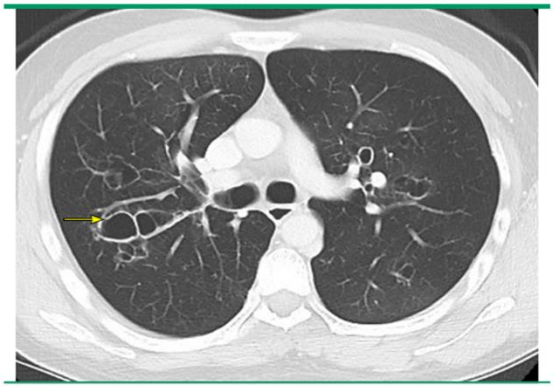
Airway obstruction

Congenital :

- bronchomalacia
- tracheobronchomegaly

Acquired

- Airway obstruction
  - foreign body aspiration
  - (benign) tumour
  - hilar adenopathy (TB, sarcoidosis)
- chronic bronchitis
- polychondritis
- mucus impaction (ABPA,



Chest radiograph of bronchiectasis in Williams-Campbell Syndrome

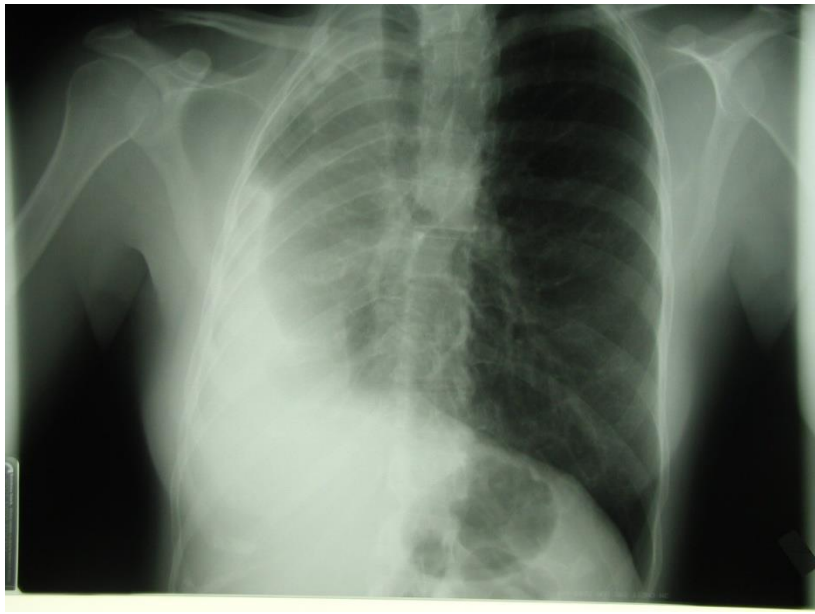
Chest computed tomogram of a patient with Williams-Campbell syndrome showing cystic bronchiectasis (arrow) and dilation of medium caliber airways. In Williams-Campbell syndrome, generalized tracheobronchomalacia is caused by deficient cartilage in the 4th to 6th order bronchi.

Causes :

H**ost defense**

- IgG deficiency (agammaglobulinemia, subclass deficiency.
- IgA deficiency.
- chronic granulomatous disease .
- AIDS / HIV

Causes
Impaired drainage / other



Impaired drainage:

- CF
- Young's sy.
- PCD
- Kartagener's sy.

Other:

- RA, Sjoegren's sy
- alpha – 1 antitrypsin deficiency
- GIT disorders (UC, Crohn, GERD)
- infections in childhood (pertussis, measles, bacterial pneumonia, TB, adenovirus, ...)
- inhalation of toxic fumes and dusts.

Clinical features :

Chronic cough and mucopurulent sputum .

Associated dyspnea, wheezing, chest pain

Recurrent bronchitis and frequent antibiotic courses

Cough	**98%**
Daily sputum	**78%**
Rhinosinusitis	**73%**
Dyspnea	**62%**
Hemoptysis	**27%**
Pleurisy	**20%**
Crackles	**75%**
Wheezing	**22%**
Digital clubbing	**2%**

*King PT et al. Respir Med 2006; 100: 2183.

Physical examination

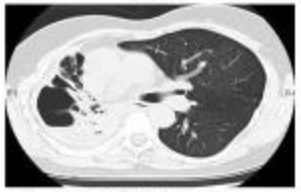
- Signs of chronic respiratory disorders.
- Signs of underlying cause .
- Biphasic crackles on auscultation .

Clinical suspicion of bronchiectasis

Bronchiectasis confirmed by HRCT

More *localized* bronchiectasis

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

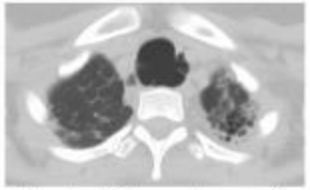


Remote TB in the right lung complicated by *M. abscessus* infection, destroying the right lung.

More *diffuse* bronchiectasis

Diagnosis highly suggestive by imaging

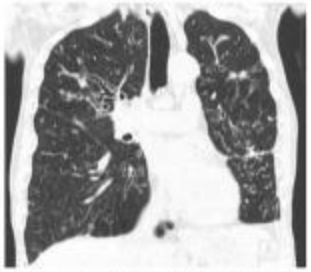
Mounier-Kuhn syndrome
Williams-Campbell syndrome
PCD with *situs inversus*



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.

laboratory testing

1. CBC, differential BC
2. immunoglobulin quantitation (levels of IgG, IgM, IgA)
3. Testing for cystic fibrosis:

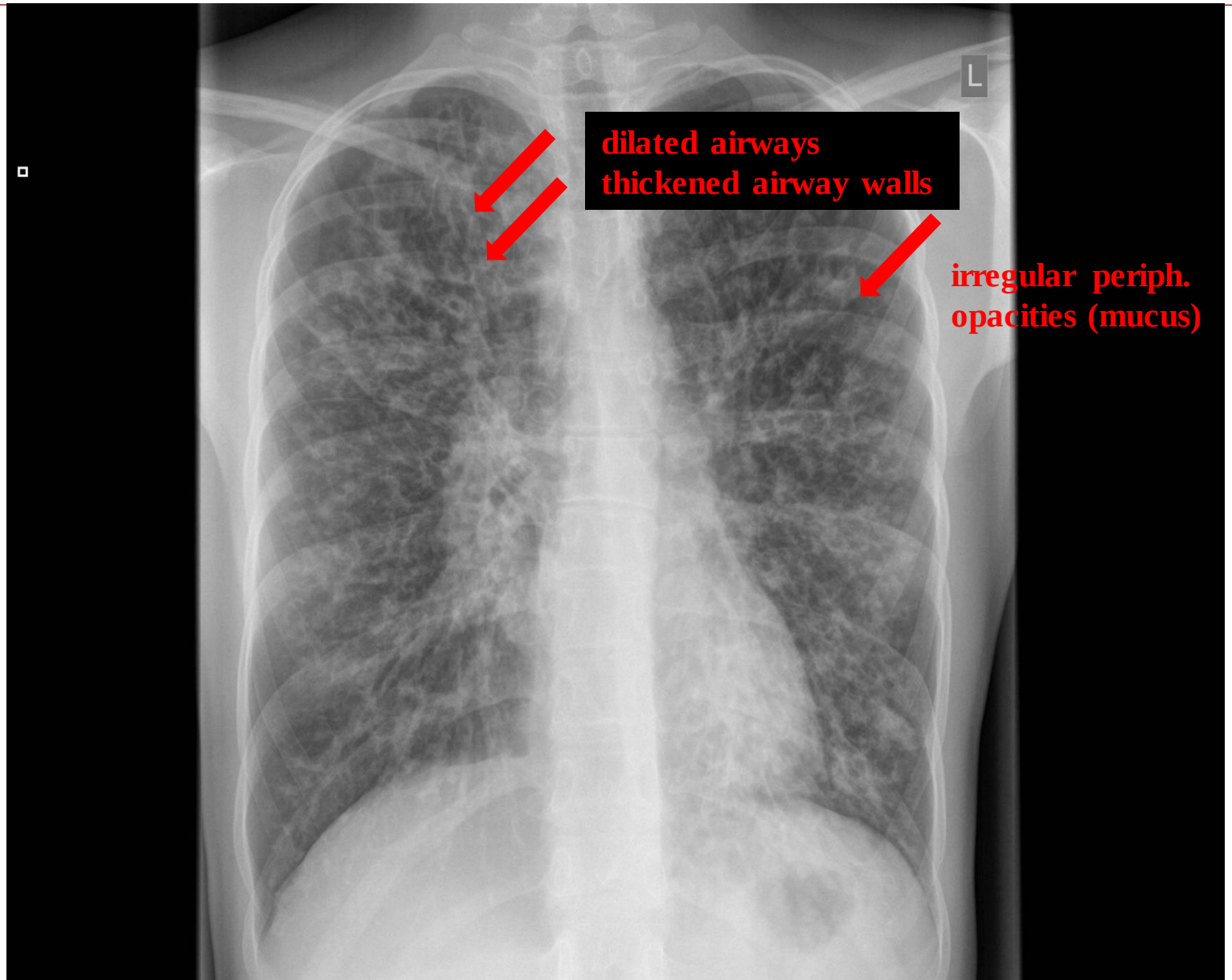
   Sweat chloride

   mutation analysis of the cystic fibrosis transmembrane conductance regulator (CFTR) gene
4. sputum culture (bact. / TBC / fungi)

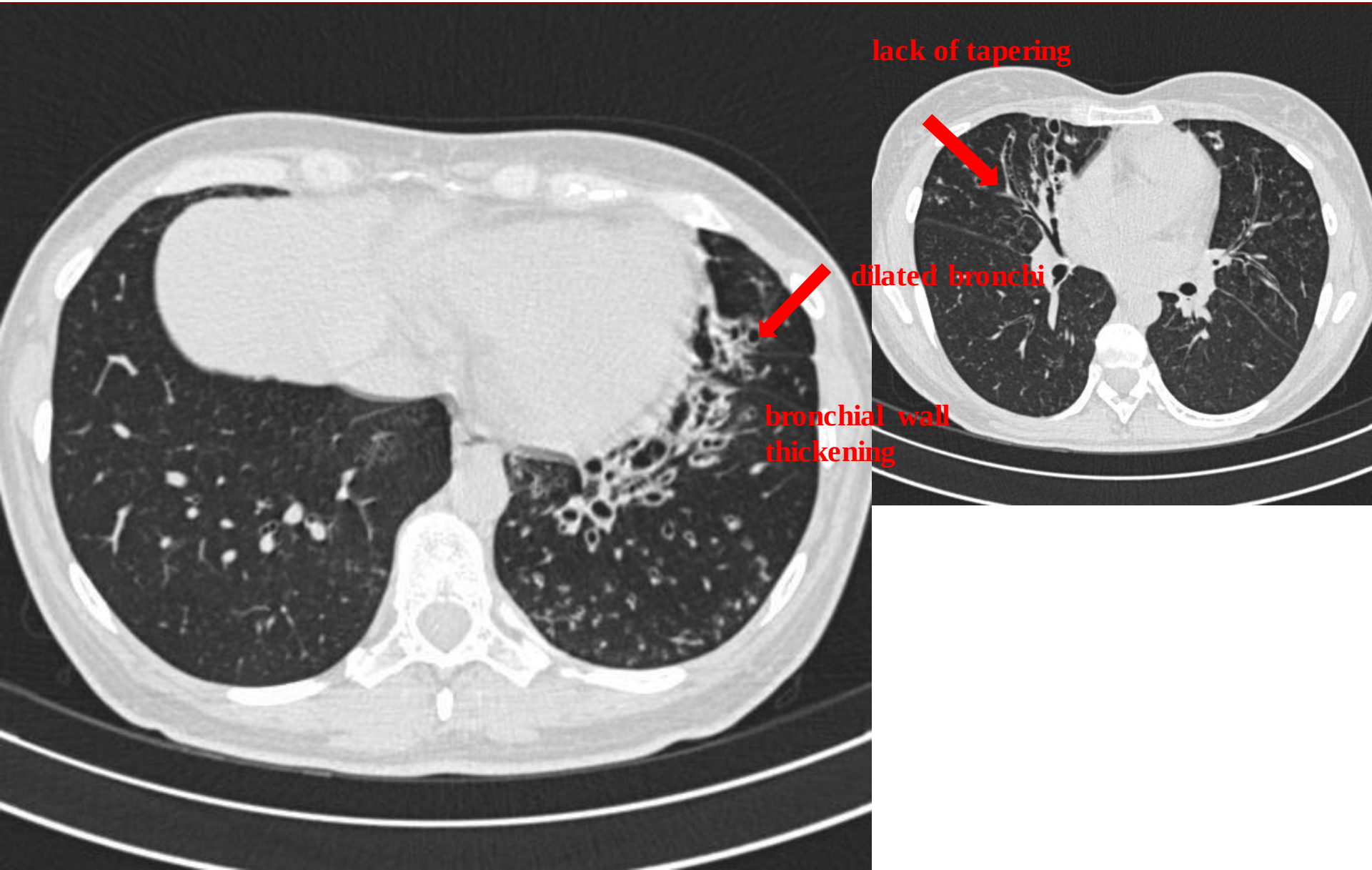
Additional testing

- Specific aspergillus IgE and IgG antibodies, total serum IgE level(Allergic bronchopulmonary asperjillosis) .
- IgG subclass levels .
- Alpha-1 antitrypsin level and/or genotype .
- Rheumatoid factor.

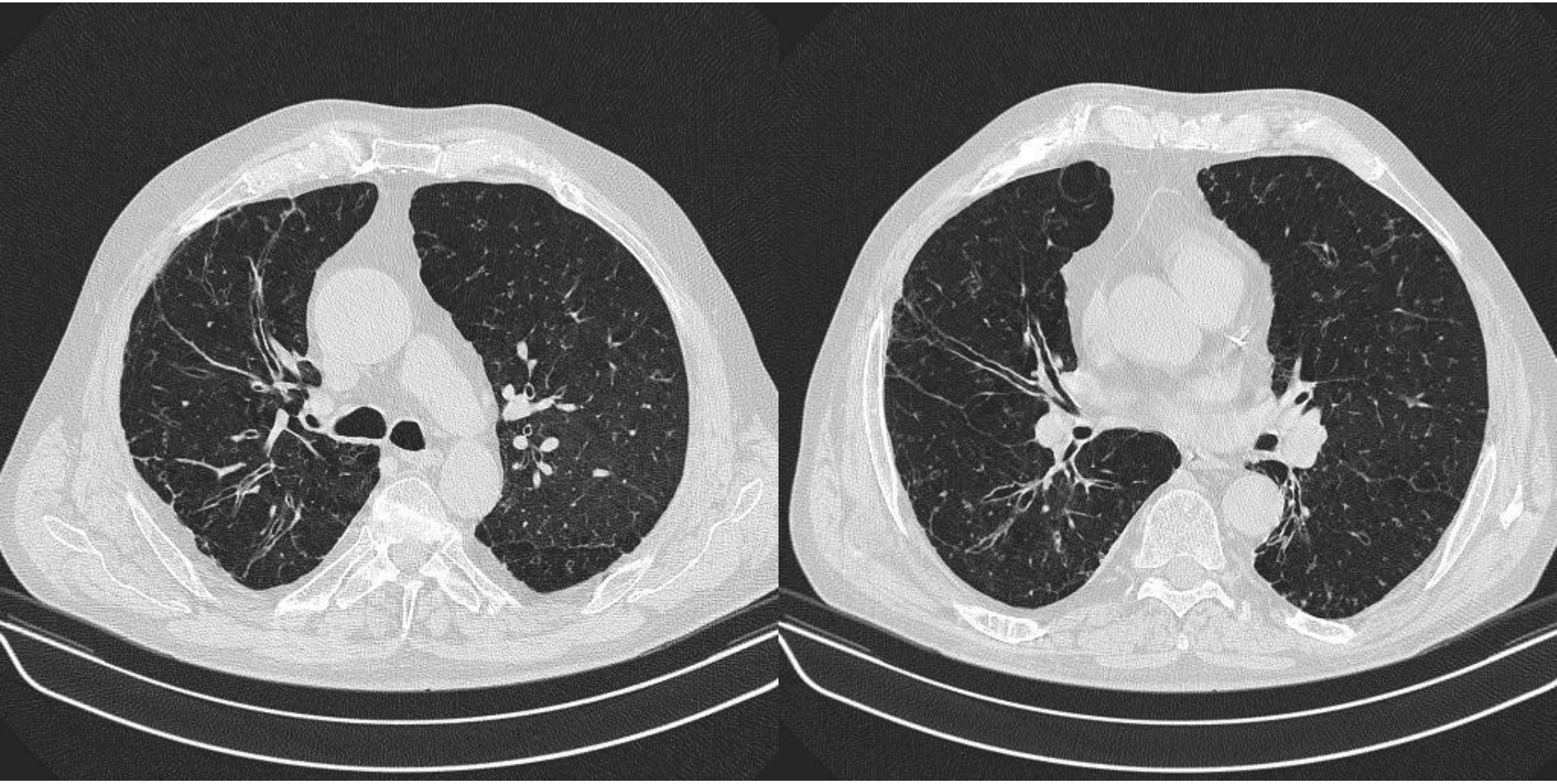
CXR


L
dilated airways
thickened airway walls
irregular periph.
opacities (mucus)

Chest CT

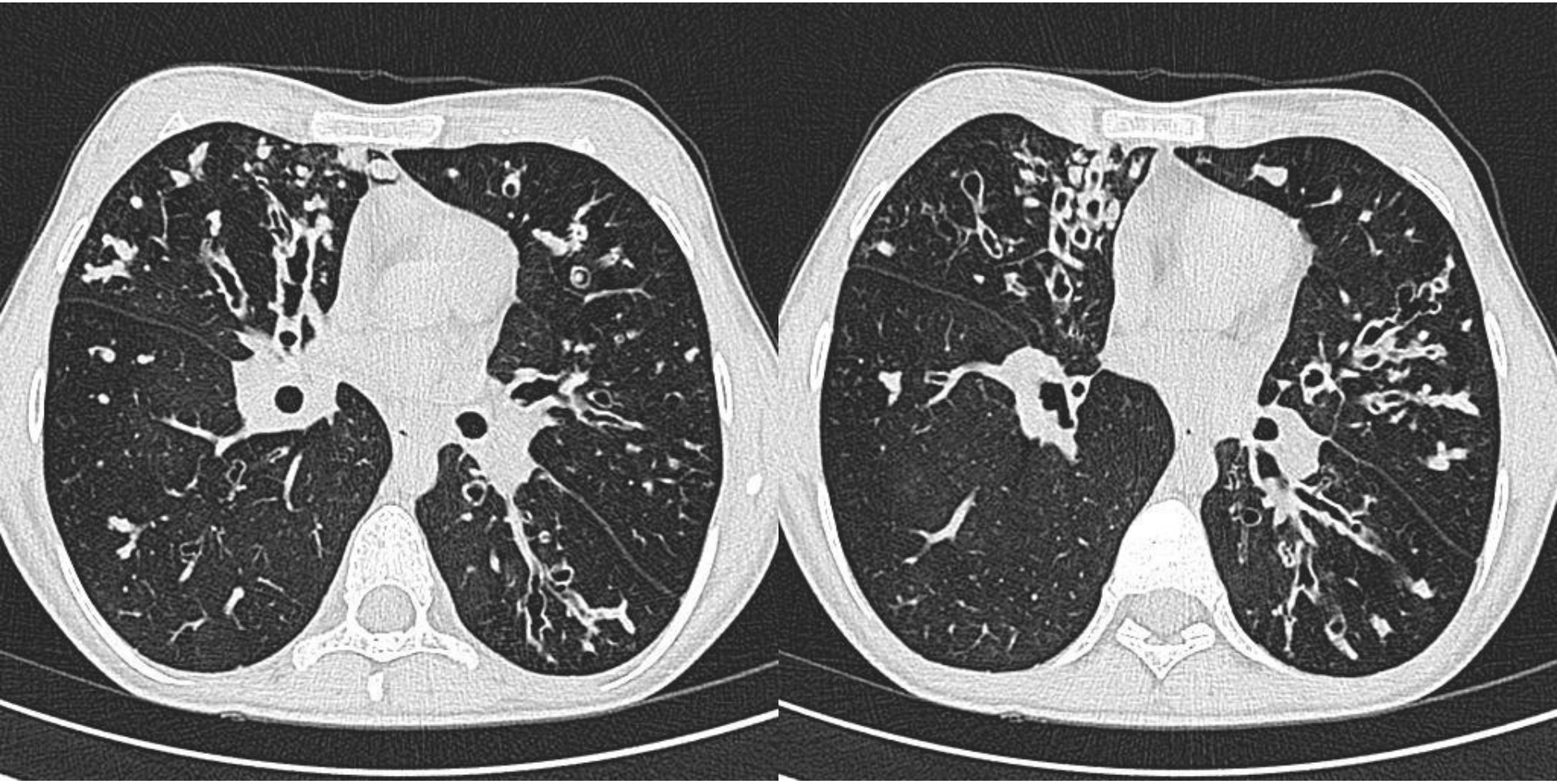

lack of tapering
dilated bronchi
bronchial wall thickening

Chest CT



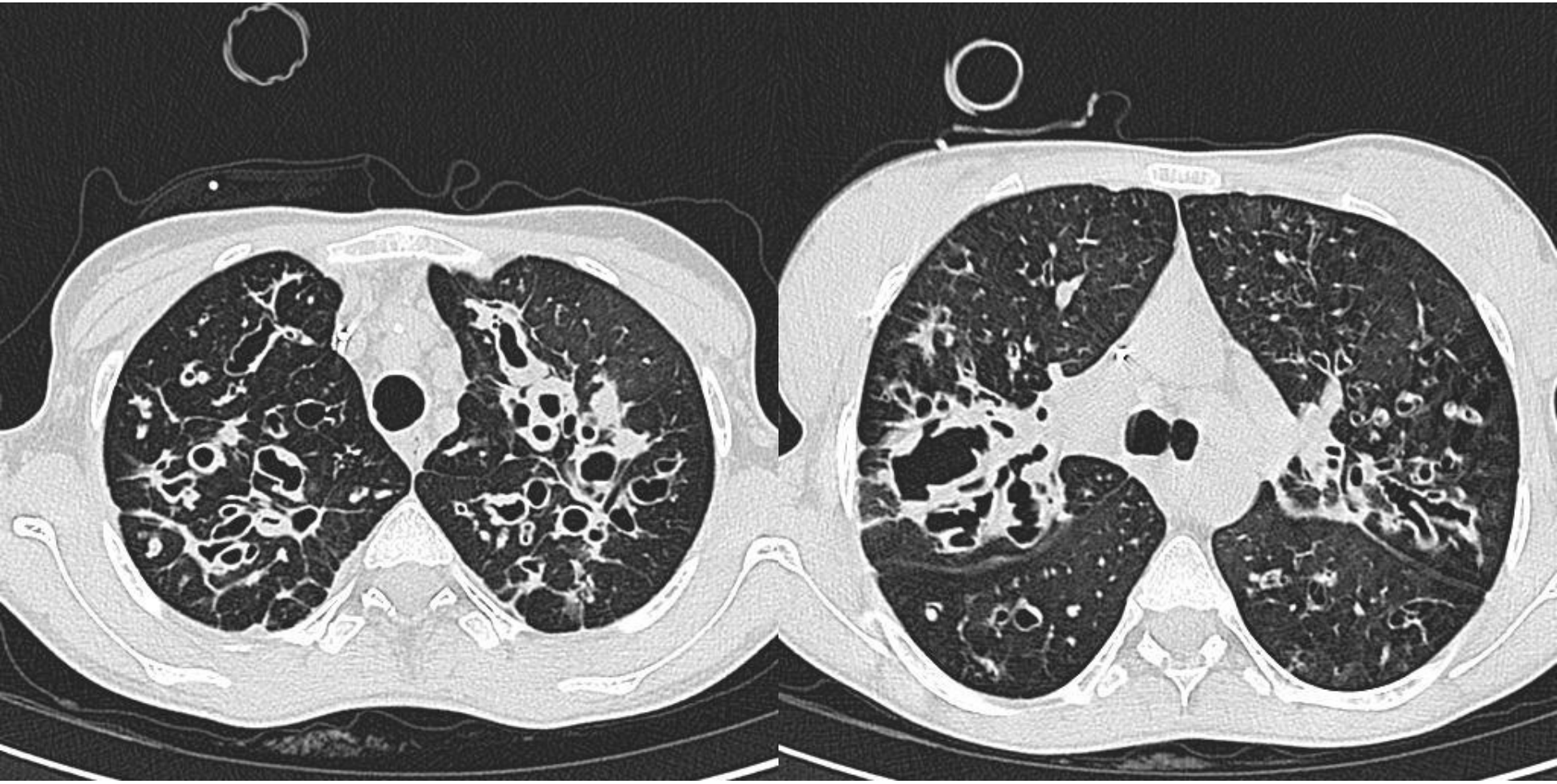
Cylindrical bronchiectasis

Chest CT



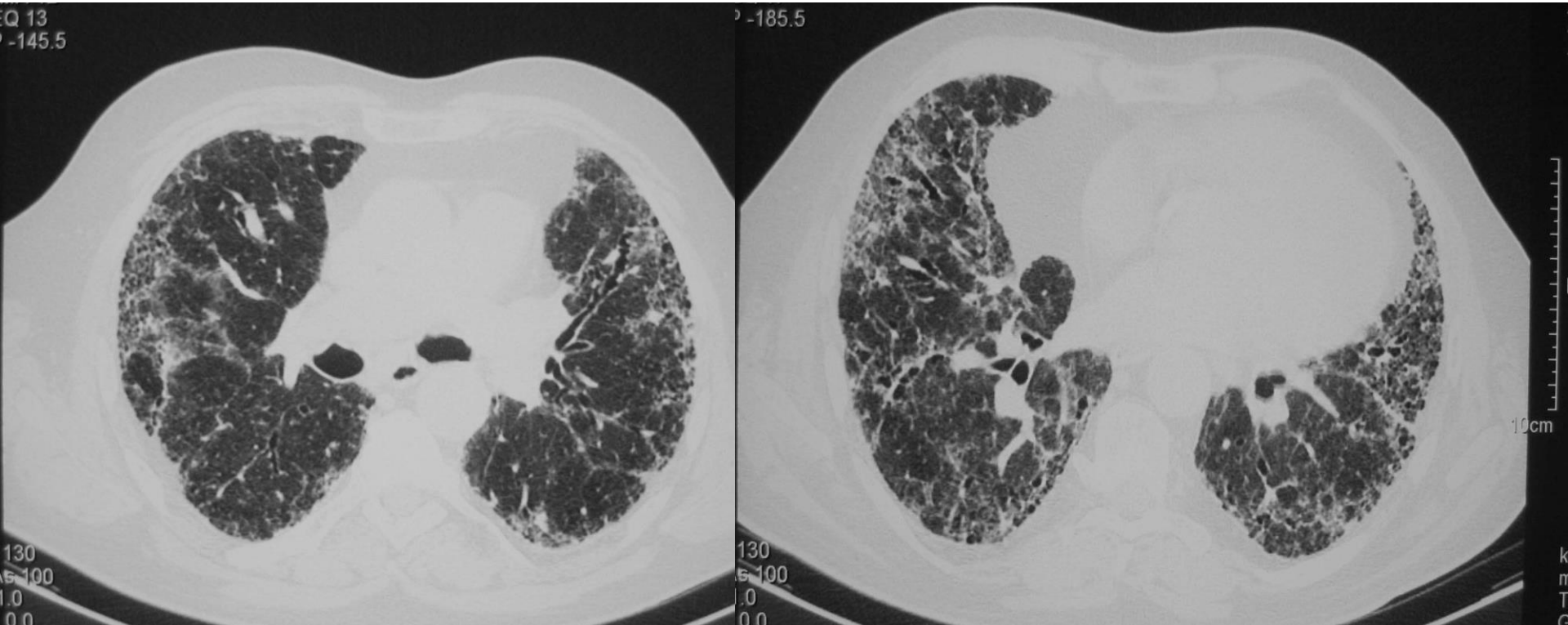
Varicose bronchiectasis

Chest CT



Cystis / saccular bronchiectasis

Chest CT



Traction bronchiectasis (fibrosis)

Pattern of distribution

1. central (perihilar)	ABPA
2. predominant upper lobe	CF, Young sy, post - TBC
3. middle /lower lobe	PCD
4. lower lobe	Idiopathic

Lung function test

- Obstructive spirometry .
- Low FVC in advanced disease

Bronchoscopy

Obstructing lesion .

Treatment

- **Treatment of the underlying disease if possible.**

Nontuberculous mycobacterial infection.

Immunodeficiencies.

Cystic fibrosis.

Recurrent aspiration.

Allergic bronchopulmonary aspergillosis.

Bronchiectasis associated with rheumatic disease.

Treatment of Exacerbation

- Antibiotics is standard therapy .
- Sputum culture should guide antibiotic choice ?
- Duration of antibiotics for hospitalized patients should be 10-14 days .
- For recurrent exacerbations…. macrolide antibiotic.
- Airway clearance techniques to remove airway secretions .
- Inhaled dornase (Dnase).
- Nebulized hypertonic saline.
- Inhaled and systemic glucocorticoids.
- Immunization .
- Pulmonary rehabilitation.
- Surgery and lung transplant.

Thank you