Bronchiectases: lecture

Ass.Prof. Nina A.Filippova

Definition

- Bronchiectasis - uncommon disease, most often secondary to an infectious process, that results in the abnormal and permanent distortion of one or more of the conducting bronchi or airways (**Medscape**).

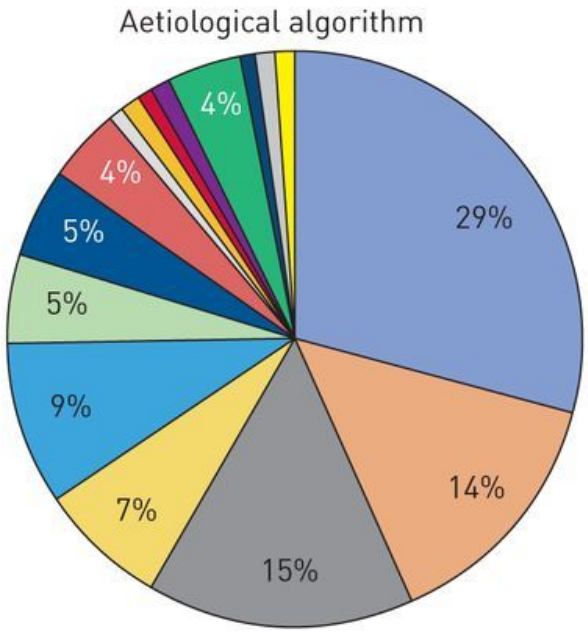
ERS guidelines for the management of adult bronchiectasis
(Eva Polverino, Pieter C. Goeminne, Melissa J. European Respiratory Society European Respiratory Journal 2017):

- Bronchiectasis is
- chronic respiratory disease
- characterised by a clinical syndrome of cough, sputum production and bronchial infection
- and radiologically by abnormal and permanent dilatation of the bronchi.
- The objectives of treatment in bronchiectasis are to prevent exacerbations, reduce symptoms, improve quality of life and stop disease progression.
- Cough and sputum production, along with breathlessness are the most frequent symptoms but rhinosinusitis, fatigue, haemoptysis and thoracic pain are also common

Classification by etiology

1. Genetic disorders (cystic fibrosis, primary ciliary dyskinesia, alpha1-antitrypsin deficiency)
2. Post infectious disease (bacteria, virus, fungi, other)
3. Immunodeficiency (congenital, acquired)
4. Aspiration (gastro-oesophageal reflux, swallowing dysfunction, tracheo-esophageal fistula)
5. Congenital structural malformations (lobar emphysema, bronchomalacia, etc.)
6. Mechanical factors (foreign body, extrinsic compression, endobronchial lesions)

Classification by etiology

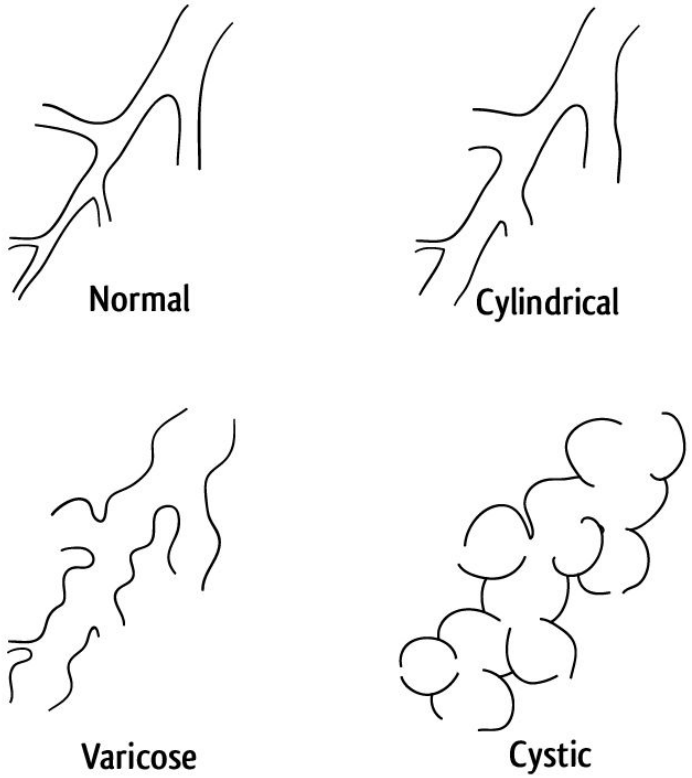


- 29% idiopathic
- 14% post-infective
- 15% - COPD (predominantly not numerous, local*)
- 7% asthma (predominantly not numerous, local*)
- 9% - connective tissue diseases (traction bronchoectases)
- 5% - allergic bronchopulmonary aspergillosis
- 5% - immune deficiency
- 4% - post Tb
- 4% - GERD (aspiration)
- Others – less than 1% any (NTM: nontuberculous mycobacteria; ; PCD: primary ciliary dyskinesia; CF: cystic fibrosis; CFTR-RD: cystic fibrosis transmembrane conductance regulator-related disease; A1ATD: α_1-antitrypsin deficiency; IBD: inflammatory bowel disease; YNS: yellow nail syndrome; DPB: diffuse panbronchiolitis)

Idiopathic | Post-infective | COPD | Asthma | CTD | ABPA | Immunodeficiency | post-TB
IBD | PCD | NTM | Sarcoidosis | GORD/aspiration | Other | Two aetiologies | A1ATD

Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneityProf Patrick A Flume, MD, Prof James D Chalmers, MBChB, Kenneth N Olivier, MD The Lancet Volume 392, Issue 10150, Pages 880-890 (September 2018)

Classification: by shape



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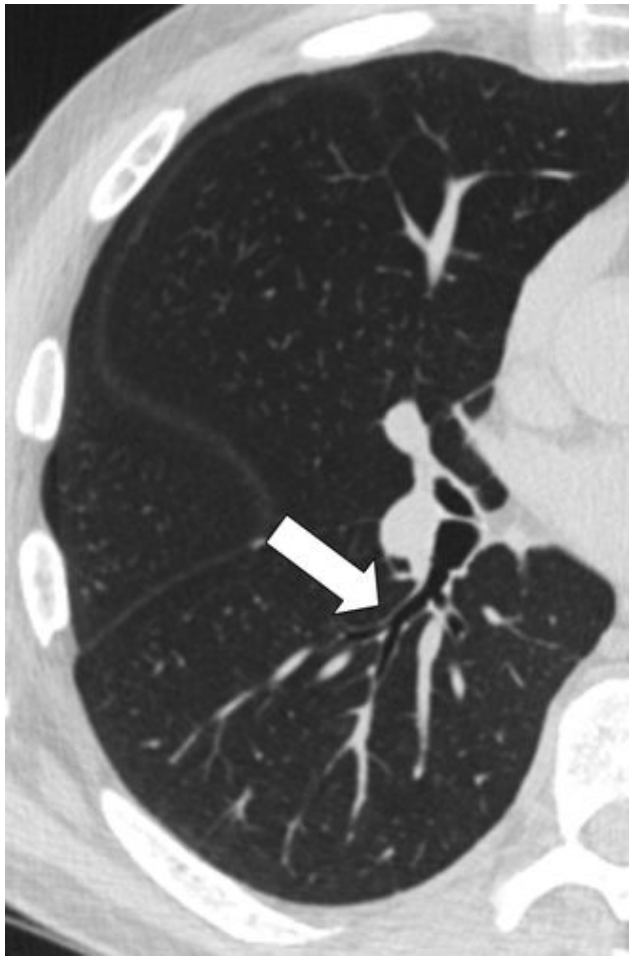
Classification by shape: Normal bronchus; no bronchoectases



American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3**
Bronchiectasis Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

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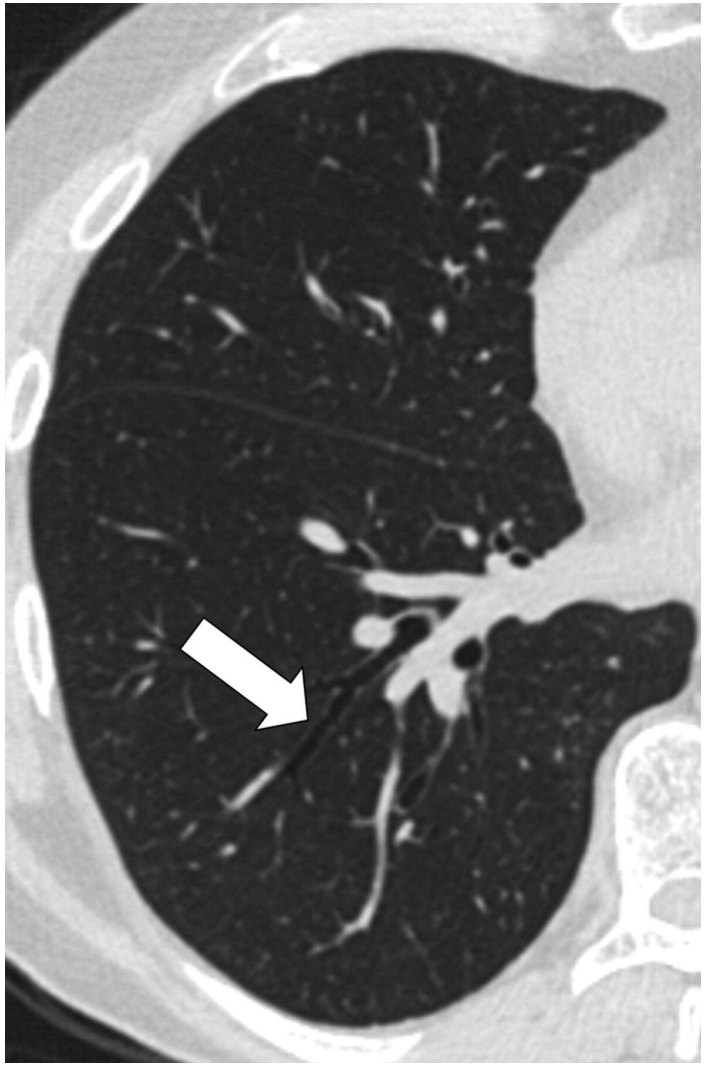
Cylindric bronchiectasis with lack of bronchial tapering



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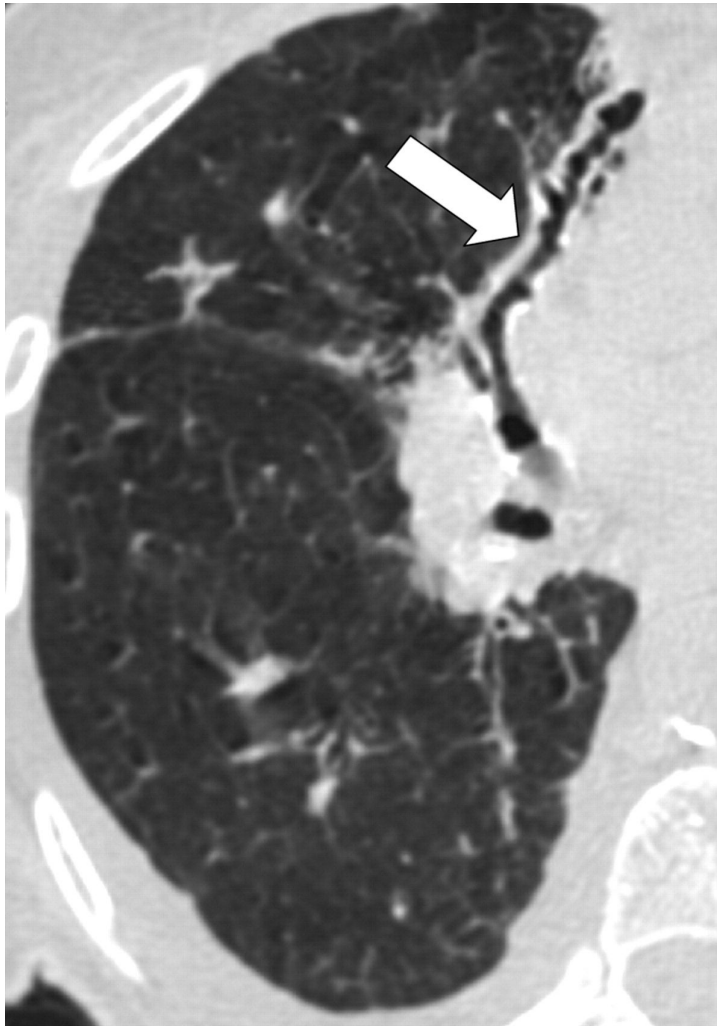
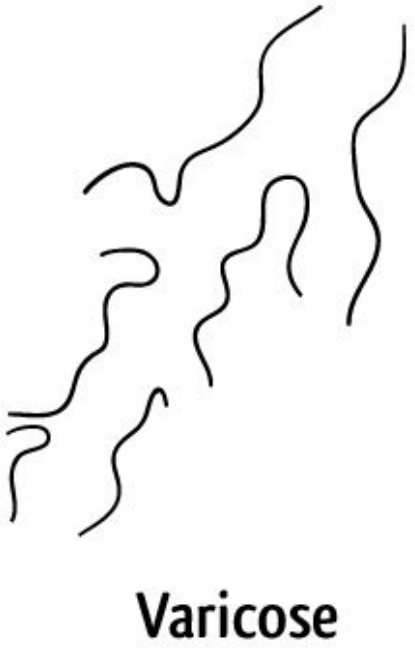
varicose bronchiectasis with string-of-pearls appearance





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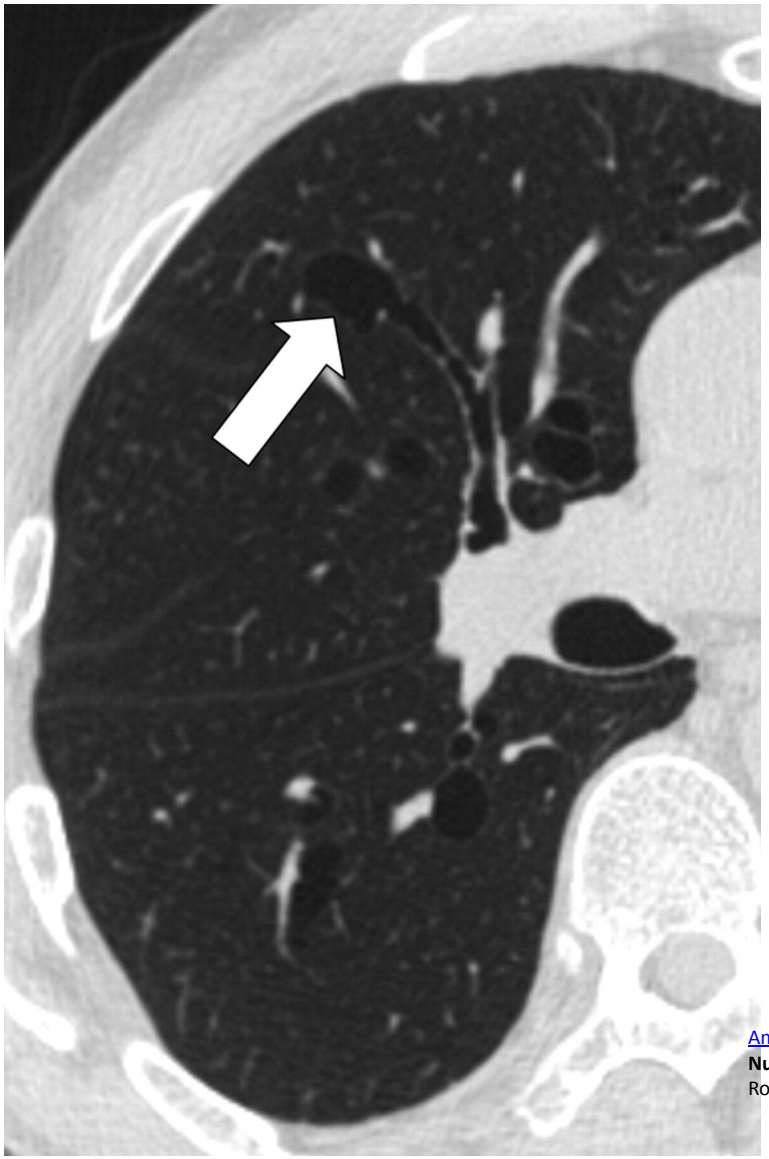
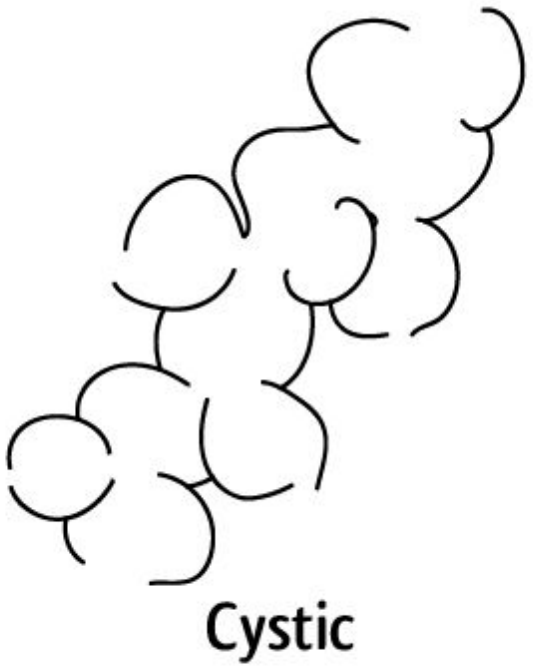
cystic bronchiectasis





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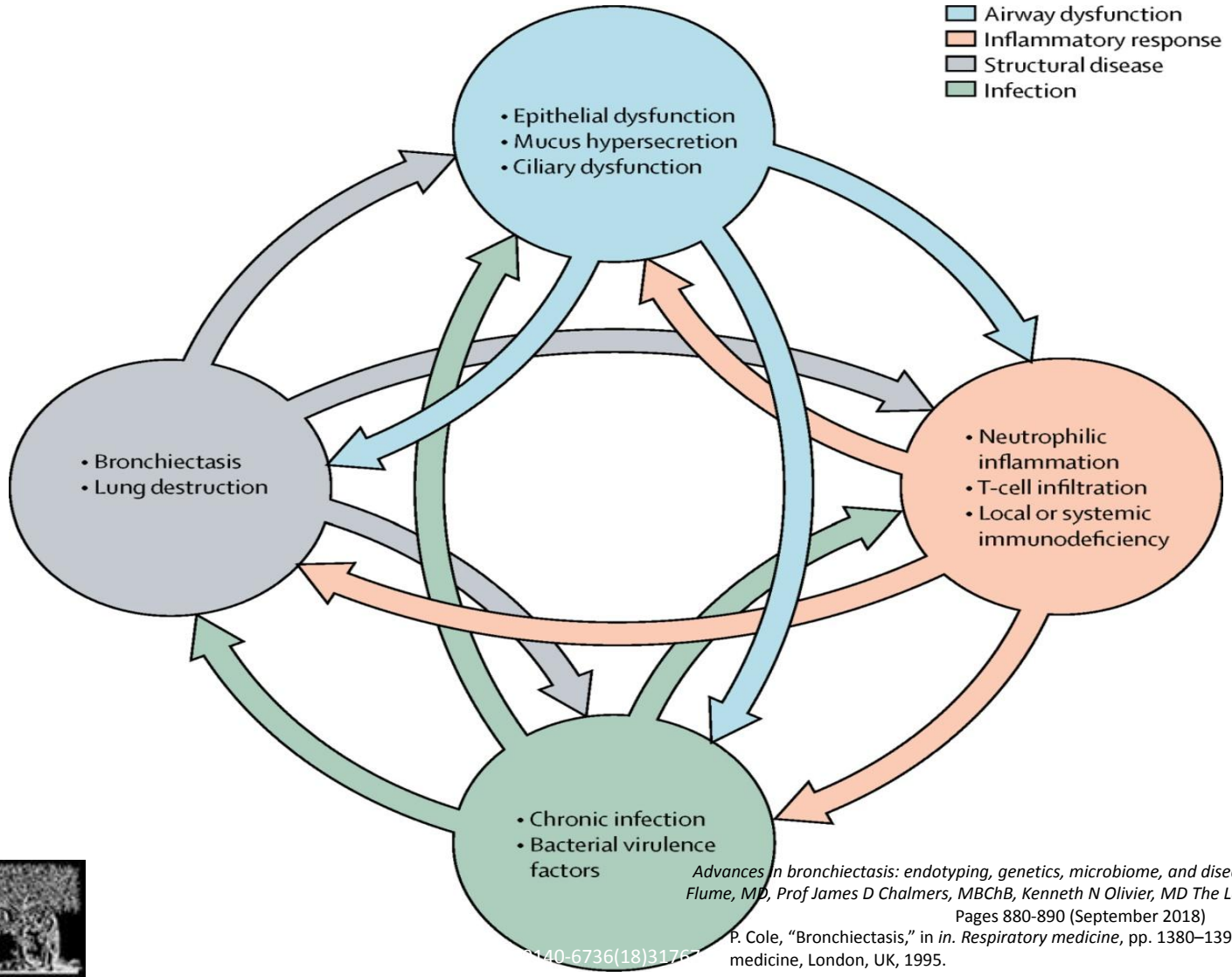
Classification: etiology and location

- **Diffuse**: Peripheral predominance
- **Upper lung** – cystic fibrosis, sarcoidosis, postradiation fibrosis
- **Lower lung** – idiopathic, postinfectious, aspiration related, fiibrotic lung disease, posttransplant rejection, hypogammaglobulinemia
- **Right middle lobe and lingular** – atypical mycobacterial infection, immotile cilia syncrome

- **Focal** (congenital bronchial atresia, extrinsic compression, extrabronchial malignancy, foreign body, broncholothiasis, airway stenosis
- **Diffuse:** central predominance
- Allergic bronchopulmonary aspergillosis
- Mounier-Kuhn syndrome
- Williams-Campbell syndrome

Pathogenesis

vicious cycle of bacterial infection, neutrophilic elastastes induced injury of the epithelium; ciliary disfunction, progression of infection and destruction of bronchial wall



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P. Cole, "Bronchiectasis," in *in. Respiratory medicine*, pp. 1380–1395, Bronchiectasis. in. Respiratory medicine, London, UK, 1995.

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Components

- **Neutrophilic inflammation** – destruction of wall by elastases
- **Ciliary disfunction** (primary or secondary) – retention of sputum and decrease of infection agents clearance
- **Sputum properties changes** (in cystic fibrosis) – retention of sputumand decrease of infection agents clearance
- **Anatomic disorders** (primary or secondary) with deformities and/or compression - retention of sputum and decrease of infection agents clearance, increase of intrabronchial pressure with promotion of deformities
- **Immune supression** – promotion of neutrophil-mediated process

Inflammation: neutrophilic

- **Neutrophils recruitment acceleration**: **degradation of elastins; increase of neutrophilic proteolytic molecules, damage and structural changes of components of bronchial wall, resulting to its dilation**
- Participants: IL-1β, TNF α, LTβ4, IL-8 (CXCL8); action of IL-8 and other CXCs through CXCR1 and CXCR2 receptors;
- **CXR1 - neutrophil degranulation and phagocytosis,**
- **CXCR2 - adhesion and chemotaxis to the site of infection**
- Results: increase of neutrophils total number and percentage; concentration of **neutrophilic proteolytic molecules** (neutrophilic elastase (NE), myeloperoxidase (MPO) and metalloproteinase (MMP)-9 at site of inflammation
- **CXCR2: important in response to** *Pseudomonas*, *Aspergillus, Nocardia*

The double-edged sword of neutrophilic inflammation in bronchiectasis
Miguel Ángel Martínez-García, Concepción Prados Sánchez, Rosa María Girón Moreno
European Respiratory Journal 2015 46: 898-900;

Importance of this mechanism for control the disease

- block of neutrophilic elastase: NE inhibitor AZD9668 in bronchiectasis patients - significant functional improvement and a trend to reduce in inflammatory biomarkers
- block of CXCR2 prevents neutrophils chemotaxis on infection site: CXCR2 antagonists: MK-7123 in COPD, non Th2 asthma; SB-656933 in cystic fibrosis; AZD-5069 – pilot study for bronchoectases (64% reduction of neutrophils in sputum in patients)

De Soyza A, Pavord I, Elborn JS, et al. A randomised, placebo-controlled study of the CXCR2 antagonist AZD5069 in bronchiectasis. Eur Respir J 2015; **46**: 1021–1032.

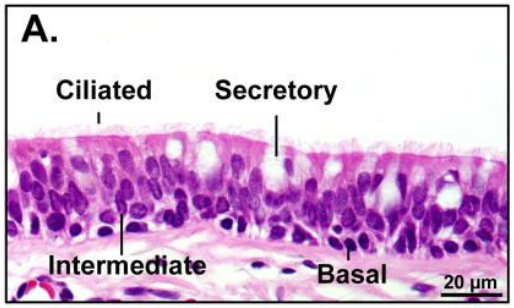
Moss R, Mistry S, Konstan M, et al. Safety and early treatment effects of the CXCR2 antagonist SB-656933 in patients with cystic fibrosis. J Cyst Fibros 2013; **12**: 241–248.

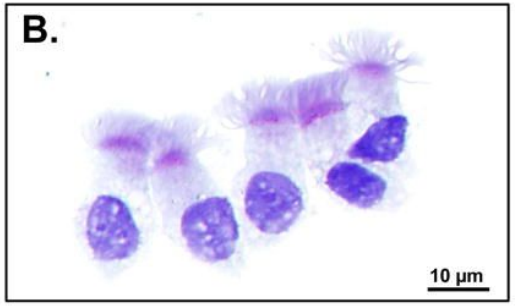
Nair P, Gaga M, Zervas E, et al. Safety and efficacy of a CXCR2 antagonist in patients with severe asthma and sputum neutrophils: a randomized, placebo-controlled clinical trial. Clin Exp Allergy 2012;**42**: 1097–1103.

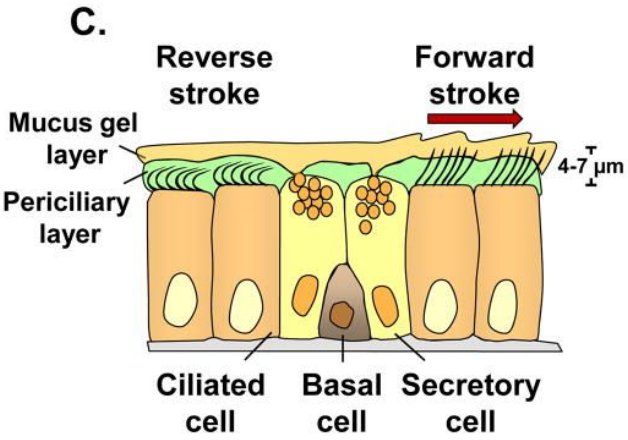
Rennard S, Dale D, Donohue J, et al. CXCR2 antagonist MK-7123. A phase 2 proof-of-concept trial for chronic obstructive pulmonary disease. Am J Respir Crit Care Med 2015; **191**: 1001–1011

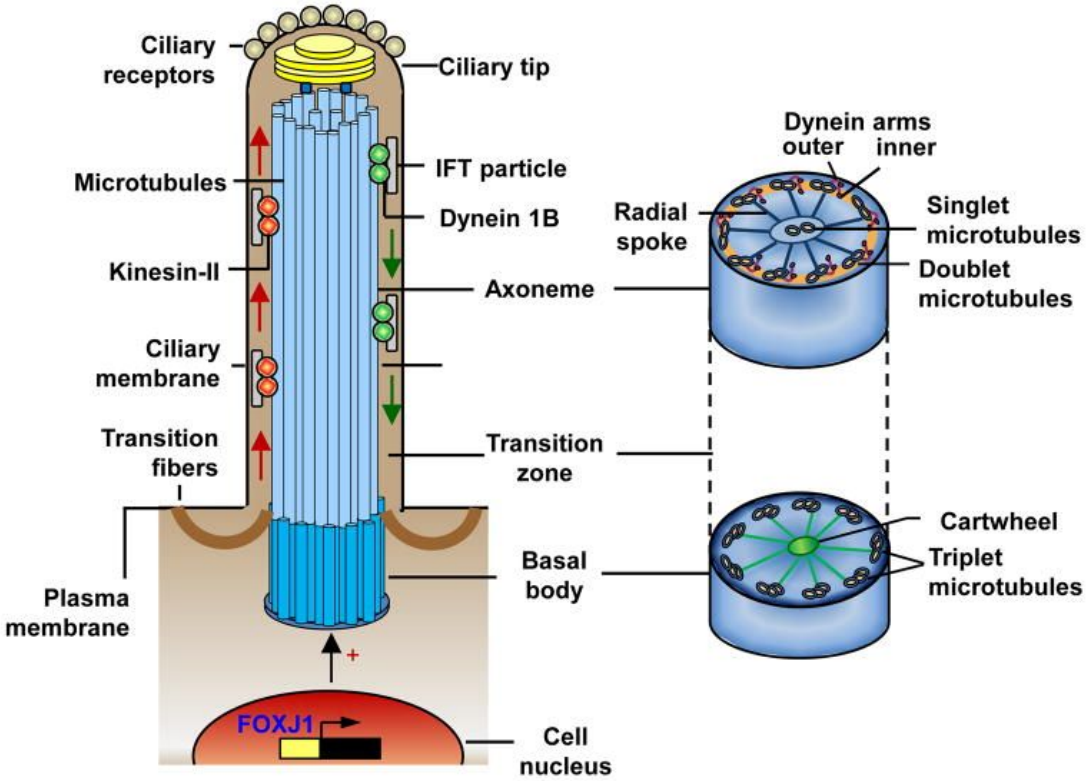
Stockley R, De Soyza A, Gunawardena K, et al. Phase II study of a neutrophil elastase inhibitor (AZD9668) in patients with bronchiectasis. Respir Med 2013; **107**: 524–533.)

Ciliary disfunction: primary and secondary









Cilia Dysfunction in Lung Disease
Ann E. Tilley,[1,2] Matthew S. Walters,[1] Renat Shaykhiev,[1] an
Annu Rev Physiol. 2015; 77: 379–406.

Genes Encoding Major Components of Airway Motile Cilia

- **Axoneme – outer dynein arm – Dyenin axoneal heavy/intermediate/ light chain genes (**DNAH5, DNAH9, DNAH11, DNAI1, DNAI2, DNAL1)
- **Dynein assembly and docking (**Dynein, axonemal, assembly factors 1-3 -DNAAF1-3 etc)
- **Tubulins and other microtubule-associated (**NME/NM23 family member 8NME8 etc)
- **Receptors, ion channels and signaling molecules (**Nitric oxide synthase 3 (endothelial cell)NOS3)
- **These genes changes predispose do development of primary ciliary dyskinesia**

Secondary ciliary disfunction

- **Viruses**
- **Bacterial mediators - *H. influenzae*, *P. aeruginosa*, *Streptococcus pneumoniae (direct damage)***
- **Smoking (direct action on cilia, down-regulation of above mentioned genes)**

Primary and secondary mucociliary clearance disturbance leads to

- airway dehydration, excess mucus volume and viscosity.
- Increase of sputum content and further infection development

Primary anatomical changes, promoting clearance disorders due to bronchi deformities or compression

- **Traction bronchoectases** – advanced pulmonary fibrosis with traction of the airways
- **PostTb** – advanced fibrotic changes and localized peribronchial lymphadenopathy squeezing bronchi and causing localised bronchial obstruction (particularly in the right middle and upper lobes) with secondary decrease of clearence and infection persistence
- **Childhood infections** - whooping cough, measles, adenovirus – increase pressure in bronchiols during paroxysmal cough, mucus plugs in bronchi
- **Mycobacterium avium complex (MAC)** in elder women cause obstruction from lymphadenopathy with right middle lobe bronchiectasis
- **Other causes** – inborn changes, bronchial and lung dysplasia, endobronchial calcifications, foreign bodies etc

Flora

- *Haemophilus influenzae* (29%–70%)
- *Pseudomonas aeruginosa* (12%–31%).
- No pathogenic bacteria (30%–40%)
- Best preserved lung function: no pathogenic bacteria isolated.
- Worst prognosis – *H.influenzae; P.aeruginosa; Morax. catarrhalis, Staph.aureus,* Enterobacter.
- Aspergillus infection (ABPA – allergic bronchopulmonary aspergillosis)
- Mycobacterial infections (in older women- mycobacterium avium complex (MAC) causing obstruction from lymphad-enopathy with right middle lobe bronchiectasis)

Int J Chron Obstruct Pulmon Dis. 2009; 4: 411–419. The pathophysiology of bronchiectasis
Paul T King

Effects of flora promoting bronchoectases

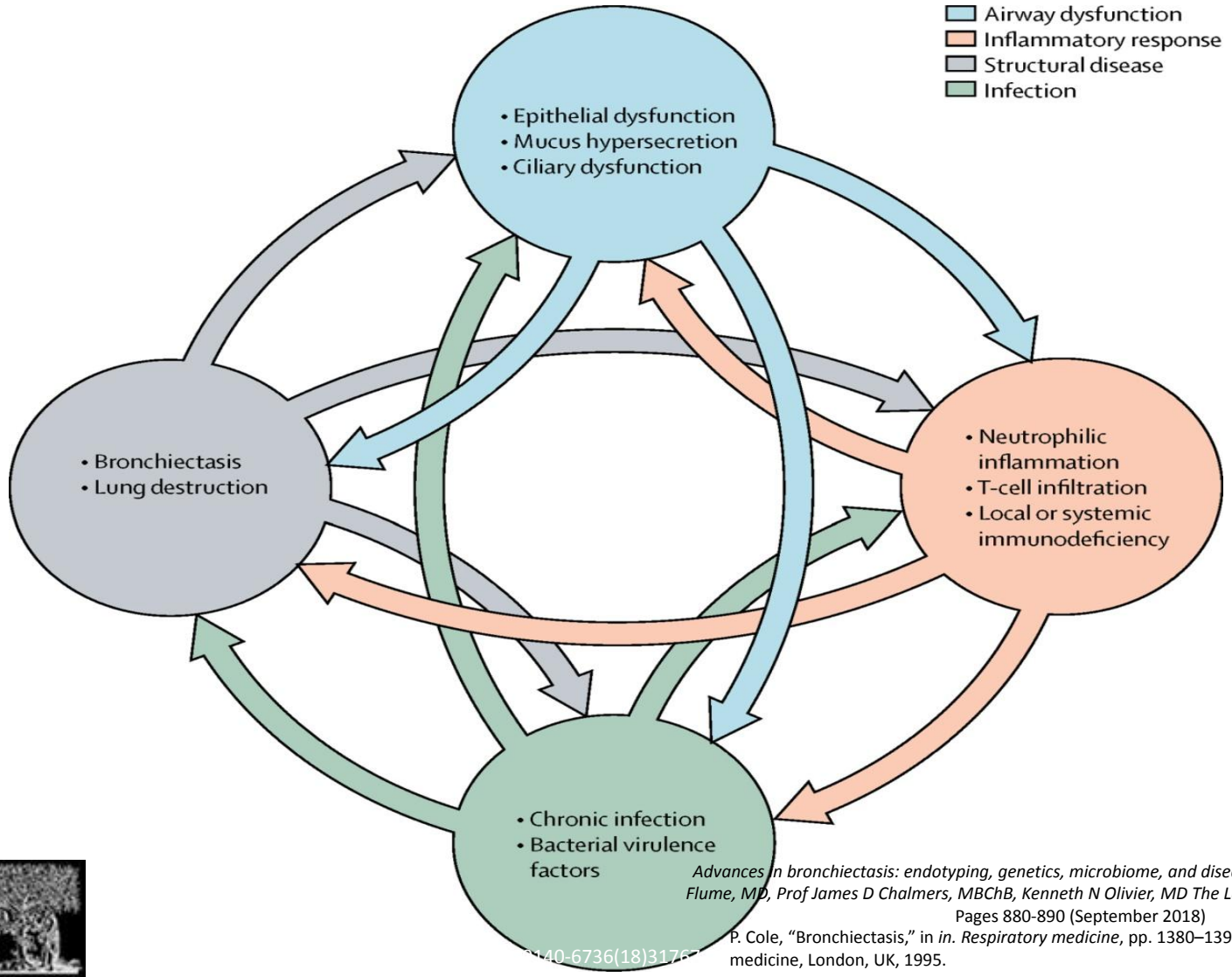
- inhibition of the mucociliary clearance: mediators of *H. influenzae*, *P. aeruginosa*, *Streptococcus pneumoniae* directly damage ciliated epithelium, and inhibit mucous transport and release glycoproteins attracting neutrophils.
- *H. influenzae* direct damage to airway epithelium; invasion into the bronchial wall and interstitium of the lung
- *P. aeruginosa* - forms biofilms, which form impenetrable matrix around bacteria *and defend it from* immune system and antibiotics

Immune dysfunction

- Malnutrition
- Extremes of age
- hypogammaglobulinemia, human immunodeficiency virus (HIV), interferon gamma receptor deficiency, type I major histocompatibility complex deficiency, late stages of lung transplant rejection, very high IgE levels without ABPA

Pathogenesis

vicious cycle of bacterial infection, neutrophilic elastastes induced injury of the epithelium; ciliary disfunction, progression of infection and destruction of bronchial wall



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Clinical manifestations

- Chronic productive cough - 98% of patients
- Sputum - produced on a daily basis - 70% - 96% of patients (4%-30% - “dry” bronchoectases.
- Sputum usually mucoid; during infectious exacerbations – greenish/yellowish purulent, may be offensive odor.
- Sputum amount usually >50 ml daily, in mild BE – 10 ml and less; in moderate 10-150 mL ; in severe more than 150 mL
- Hemoptysis - 56-92% of patients; more commonly in dry bronchiectasis; usually mild; appears from dilated bronchial arteries: massive hemoptysis – rare

Author: Ethan E Emmons, Bronchiectasis Clinical Presentation Updated: Jul 23, 2019
https://emedicine.medscape.com/article/296961-clinical#b3

- Dyspnea – 62%-72% of patients, mixed (obstruction + restriction due to fibrosis)
- Wheezing – rare (more common in asthma)
- Fatigue – 73%, in severe cases – weight loss
- Crackles – 73% (small and medium caliber); rhonchi; more rare wheezing (predominantly local if not asthma)
- Clubbing – 2-3%
- Cyanosis – in severe cases

In whom should be suspected?

- Persistent mucopurulent/purulent sputum + risk factors
- rheumatoid arthritis + chronic productive cough/recurrent chest infections.
- COPD frequent exacerbations (two or more annually)
- inflammatory bowel disease + chronic productive cough
- asthma: severe/poorly-controlled disease
- HIV-1,solid organ and bone marrow transplant, immunosuppressives; + chronic productive cough or recurrent chest infections.
- chronic rhinosinusitis – chronic productive cough or recurrent chest infections.
- connective tissue disease or inflammatory bowel disease - chronic productive
- cough or recurrent chest infections
- otherwise healthy individuals - cough that persists > 8 wks, especially with sputum production or a history of an appropriate trigger

Adam T Hill,1 Anita L Sullivan,2 James D Chalmers British Thoracic Society Guideline for bronchiectasis in adults Thorax 2019;**74(Suppl 1):1–69. doi:10.1136/thoraxjnl-2018-212463**

Diagnosis: to confirm

- baseline chest X-ray in patients with
- suspected bronchiectasis.
- Thin section computed tomography scan (CT) to confirm a diagnosis of bronchiectasis when clinically suspected.

Adam T Hill,1 Anita L Sullivan,2 James D Chalmers British Thoracic Society Guideline for bronchiectasis in adults Thorax 2019;**74(Suppl 1):1–69. doi:10.1136/thoraxjnl-2018-212463**

CT features of bronchiectasis

- bronchial dilatation as suggested by one or more of the following:
- Bronchoarterial ratio >1 (internal airway lumen vs adjacent pulmonary artery)
- Lack of tapering
- Airway visibility within 1cm of costal pleural surface or touching mediastinal pleura.
- The following indirect signs are commonly associated with bronchiectasis:
- Bronchial wall thickening
- Mucus impaction
- Mosaic perfusion / air trapping on expiratory CT

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Diagnosis: general + flora

- full blood count incl ESR
- In all patients: specific antibodies against capsular polysaccharides of *Streptococcus pneumoniae (*specific antibody deficiency). If pneumococcal antibodies are low, immunise with 23 valent polysaccharide pneumococcal vaccine, followed by measurement of specific antibody levels 4–8 weeks later.
- Sputum cultures- in all patients for routine and mycobacterial culture.

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Diff: COPD

Bronchiectases

- Sputum >50 ml, more purulent
- Hemopthisis common
- Fever more common
- Dullness zones may be
- Crackles, moist rales locally
- Pneumonias frequent, same locations

COPD

- Small amount of sputum, purulent rare, at exacerbations
- Hemopthisis possible (exclude BE, cancer, embolism)
- Fever more rare, exacerbations
- Harsh or diminished respirations
- Wheezes, rhonchi
- Pneumonias may be, usually at GCS taking patients

Diff: cancer

Bronchiectases

- Sputum >50 ml, more purulent
- Hemopthisis common
- Fever more common
- Dullness zones may be
- Crackles, moist rales locally
- Pneumonias frequent, same locations

Cancer

- Dry cough, paroxysmal, nocturnal, usual decrease amount of sputum if compared to previous COPD symptoms; increased in bronchioalveolar
- Hemopthisis frequent
- Rapid progression of dyspnea (expiratory, then inspiratory)
- Fever in case of pneumonia
- Harsh or diminished respirations, wheezes, rhonchi - in case of pneumonia complication

Diff: embolism

Bronchiectases

- Sputum >50 ml, more purulent
- Hemopthisis common
- Fever more common
- Not typical pleural pain
- PII, epigastric pulsation may be
- Dullness zones may be
- Crackles, moist rales locally
- Pneumonias frequent, same locations

embolism

- Sputum not typical
- Hemopthisis frequent
- Pleural pain typical, PII, epigastric pulsation
- Deep veins thrombosis (unilateral leg pain and edema)
- BP decrease
- Fever not typical (if more than 2 days duration – pneumonia may develop)
- Local changes (dullness, rales) in case of pneumonia

Rare syndroms (ciliary disfunction, cystic fibrosis)

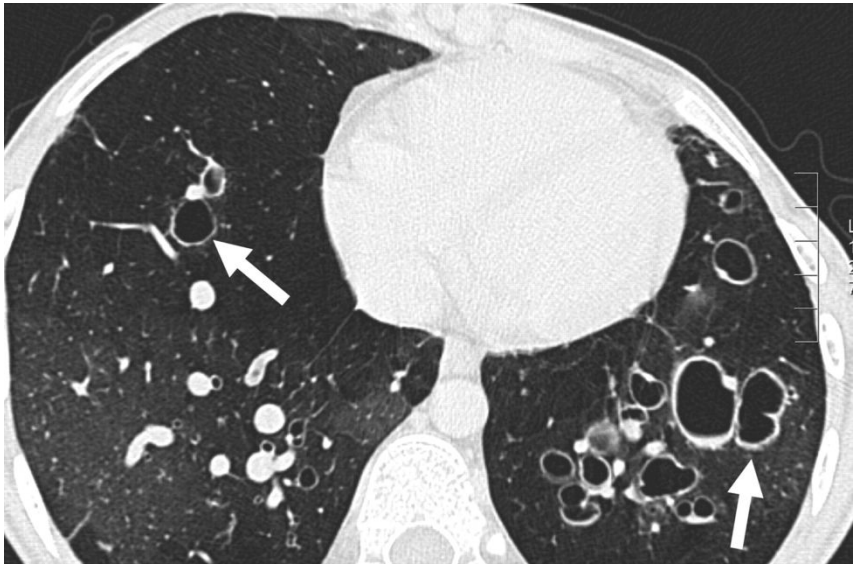
Cystic fibrosis

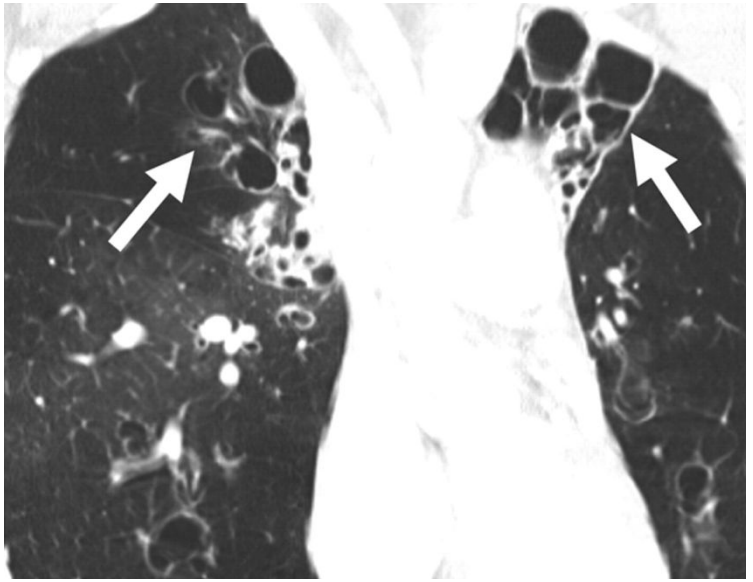
- Fetal meconium ilius may be
- Start in early life more possible
- Rhinosinusitis
- Pancreatitis, malabsorbtion, low weight gain
- Biliary liver cirrhosis (liver enlargement, jaundice)
- Viscous secretions
- High risk of Ps.aeruginosa
- Bad prognosis in case of absence of modern treatment and sever mutation
- Chloride in sweat - low
- Genetic disease – severity of disease depend only on mutation
- In adolescens and young adults – male infertility

Ciliary disfunction

- ARDS in infancy and early childhood
- Chronic otitis media
- Sinusitis
- Male infertility
- Situs viscerus inversus – Kartagener syndrome

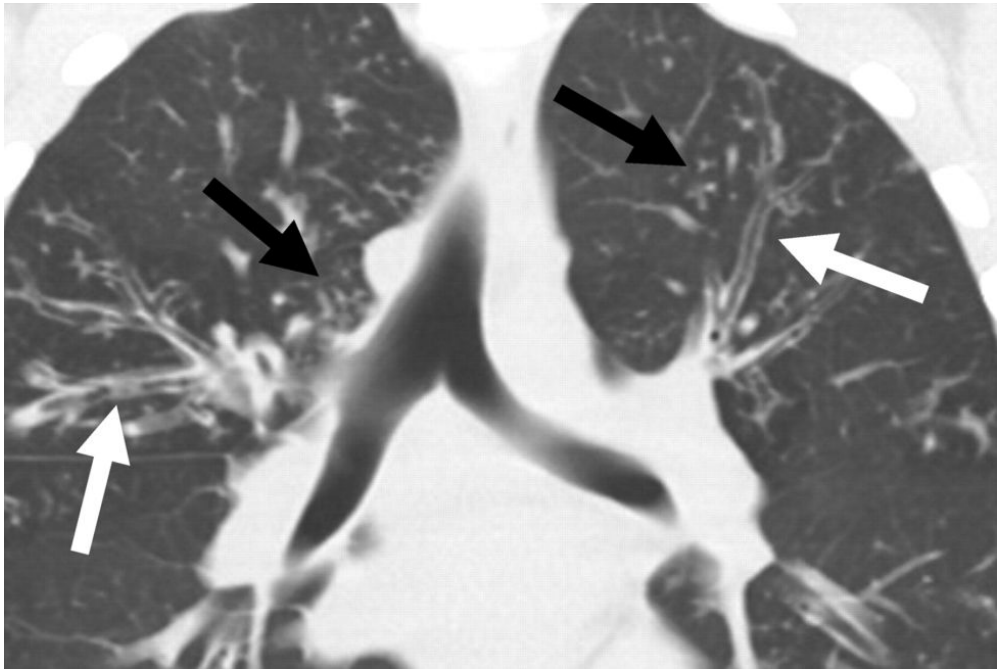
Cystic fibrosis





- predominance of cystic bronchiectasis (*arrows*)
- volume loss (fibrosis)
- diffuse heterogeneous attenuation
- enlarged lung volumes (obstruction)

Adult cystic fibrosis (milder case)



- cylindric bronchiectasis (*white arrows*)
- bronchiolitis (*black arrows*) – tree in bud syndrome

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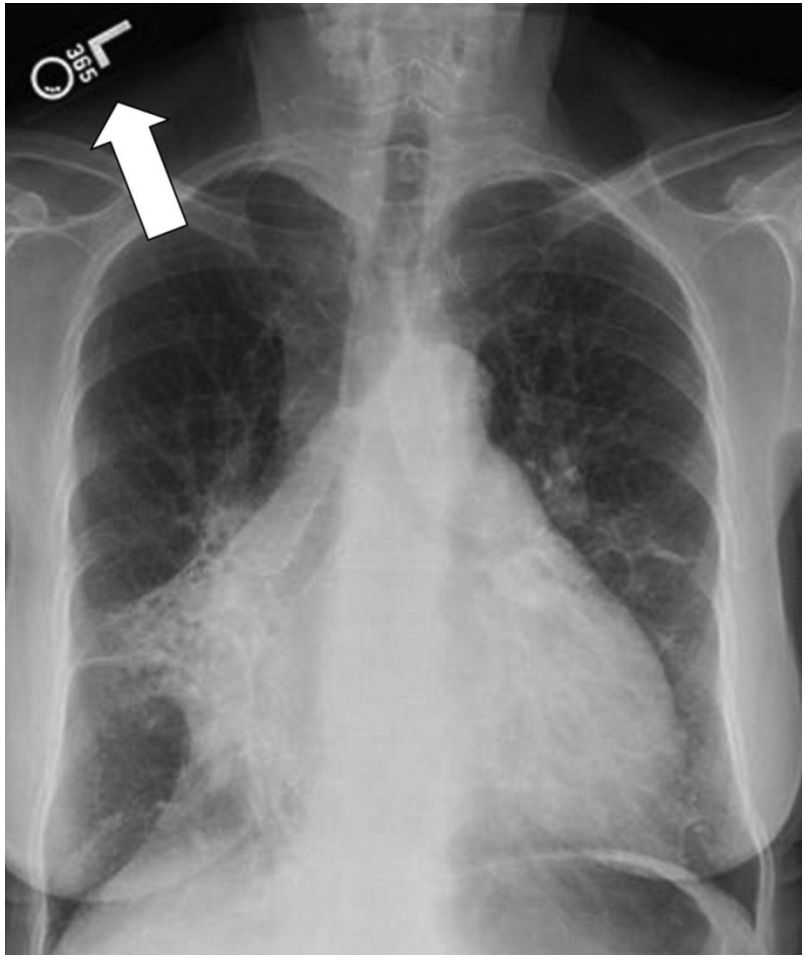
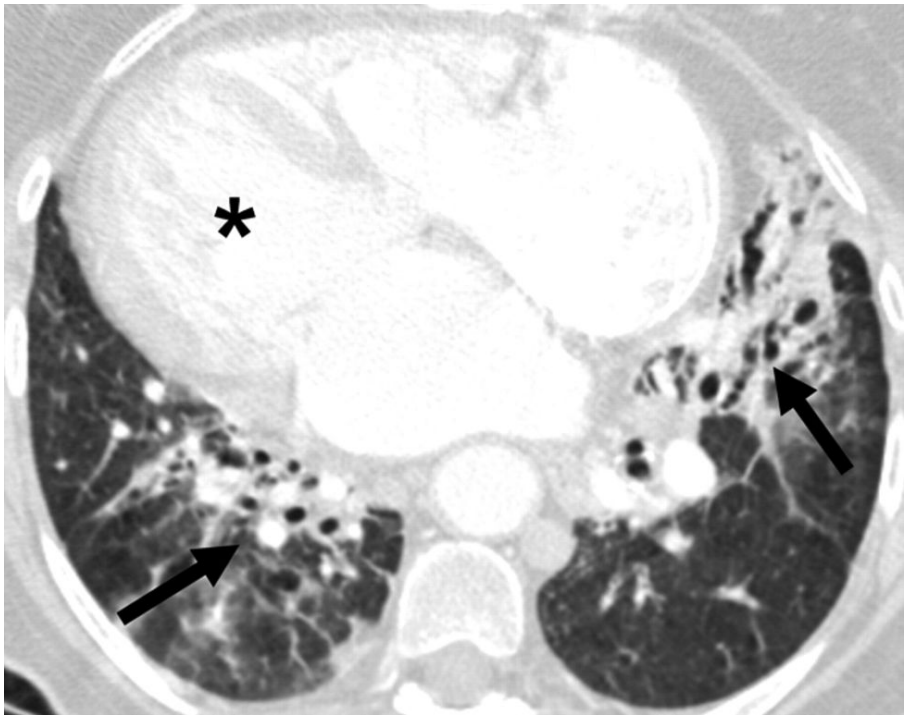
Kartagener's syndrome



- Dextrocardia (here + cardiomegaly)
- Here - left middle lobe bronchiectasis, volume loss.
- Arrow points to wrong-sided left marker



- CT confirms dextrocardia (*asterisk* is in left ventricle)
- bronchiectasis (*arrows*) predominantly midportion of lungs

Other endotypes

Alpha -1 antitripsin deficiency

- Panacinar basal emphysema in non-smokers < 30-40
- Liver cirrhosis

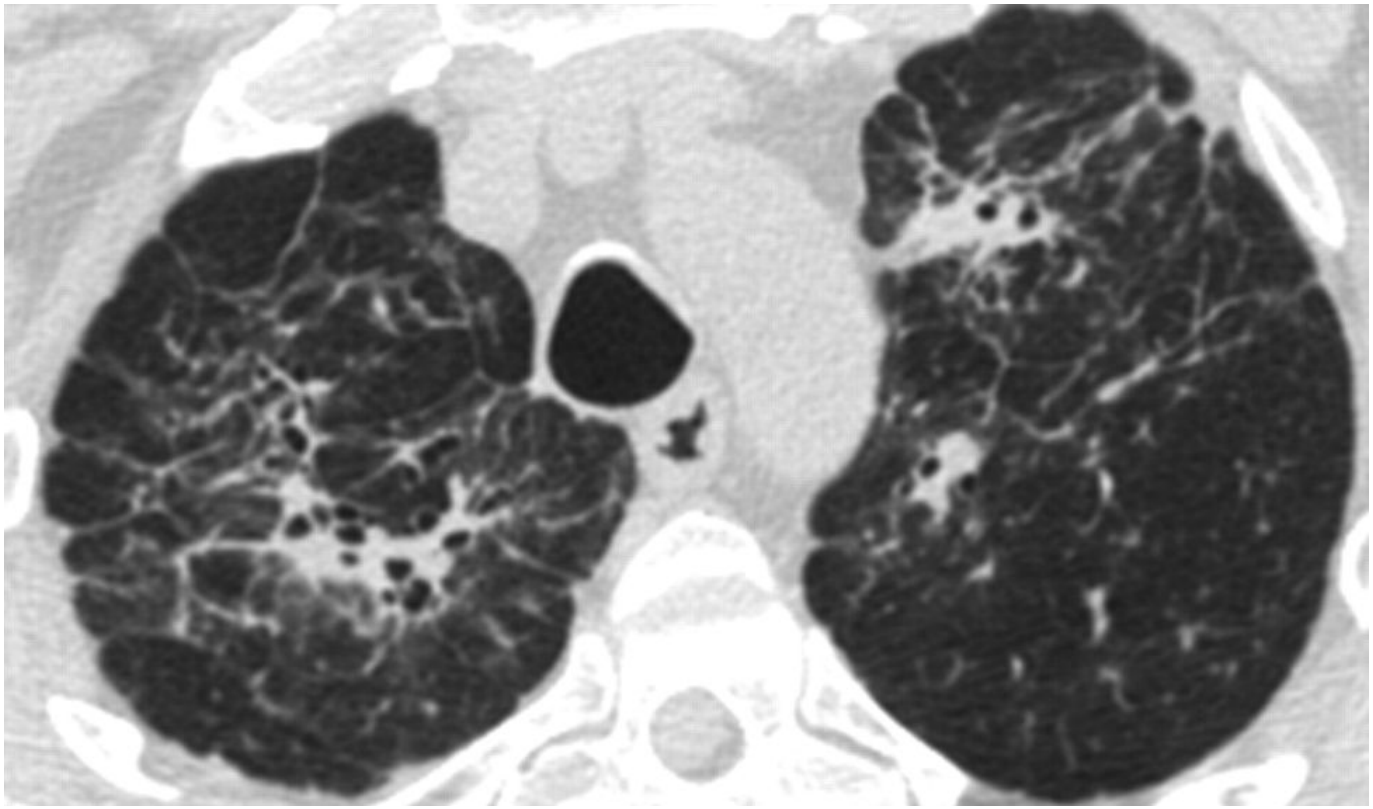
Traction bronchoectases

- Presence of idiopathic pulmonary fibrosis or lung affection due to rheumatoid arthritis, inflammatory bowel disease, connective tissue diseases, seronegative spondiloarthritis
- More common “dry” ones

Non-TB mycobacteria

- post-menopausal non-smoker females
- chronic cough, more common “dry”
- No predisposing factors
- May be CFTR mutations and ciliary dysfunction, not meeting diagnostic criteria for cystic fibrosis or primary ciliary dyskinesia.
- tall, asthenic, scoliosis, pectus excavatum, mitral valve prolapse, dural ectasia, minor features overlapping with Marfan and Ehlers-Danlos syndromes

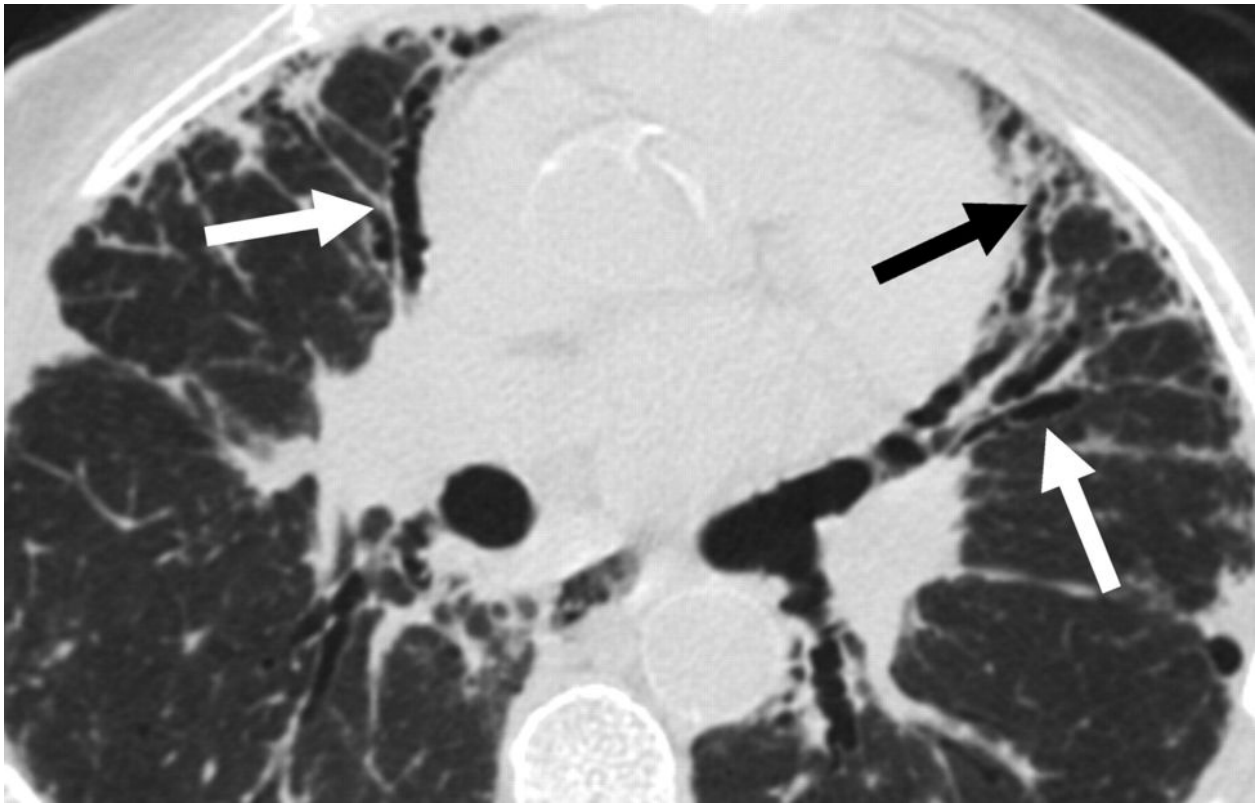
Sarcoidosis



- Diffuse fibrosis
- traction bronchiectasis (*arrows*, **B**) predominantly upper lobes.

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

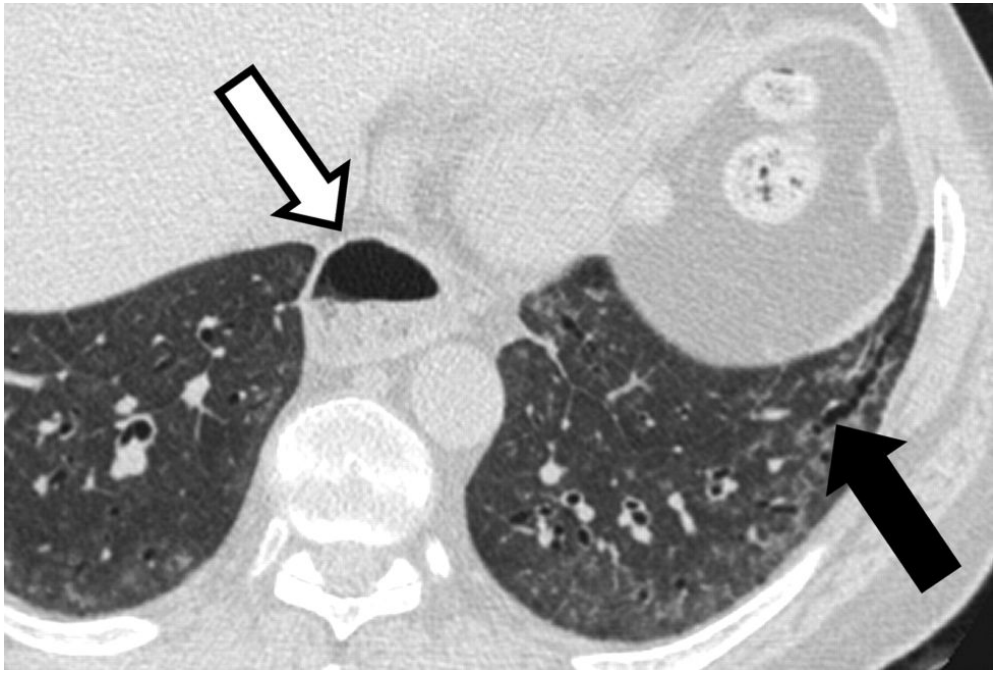
Usual interstitial pneumonia (idiopathic pulmonary fibrosis; rheumatoid arthritis, more rare Sjogren, scleroderma)



- Bibasilar and subpleural reticulation
- traction bronchiectasis in areas of fibrosis (*arrows*)

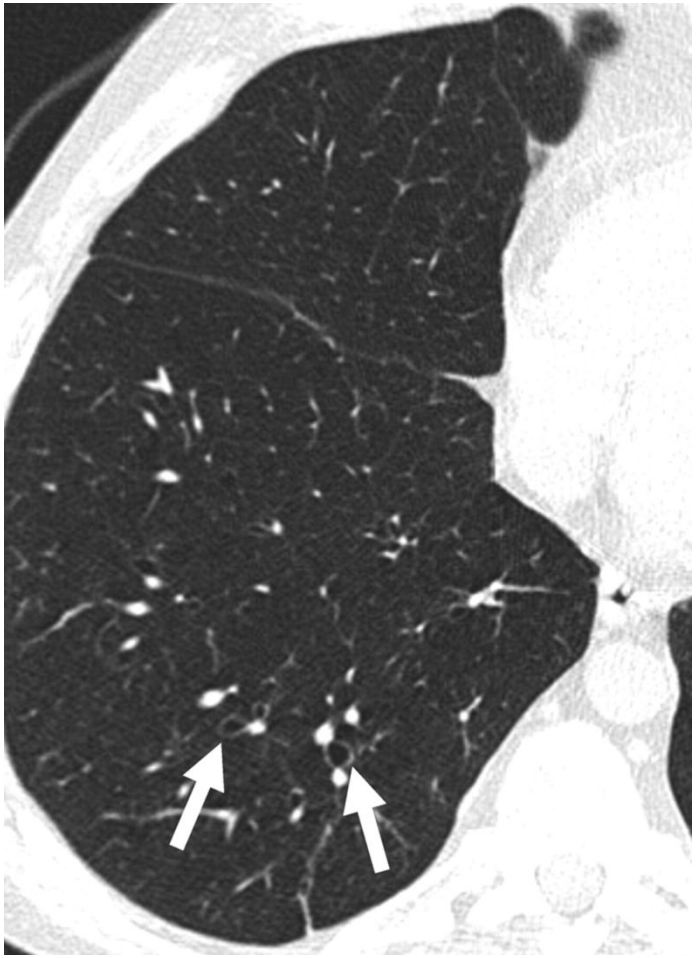
American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

Scleroderma and other connective tissue diseases – more typical NSIP



- Dilated esophagus(*white arrow*)
- ground-glass opacities (NSIP)
- recurrent aspiration with subsequent bibasilar bronchiectasis
- Black arrow points to bronchus visible in peripheral 1 cm of lung.

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053



- Bronchiolitis obliterans after lung transplantation. Transverse images of right lung in deep inspiration (**A**) and end expiration (**B**) show subtle basilar cylindric bronchiectasis (*arrows*, **A**) and widespread air trapping (*arrows*, **B**).

Other endotypes

ABPA

- Blood eosinophilia
- thick sputum with black
- Bronchial obstruction with wheeze,
- Asthma in case history
- recurrent exacerbations

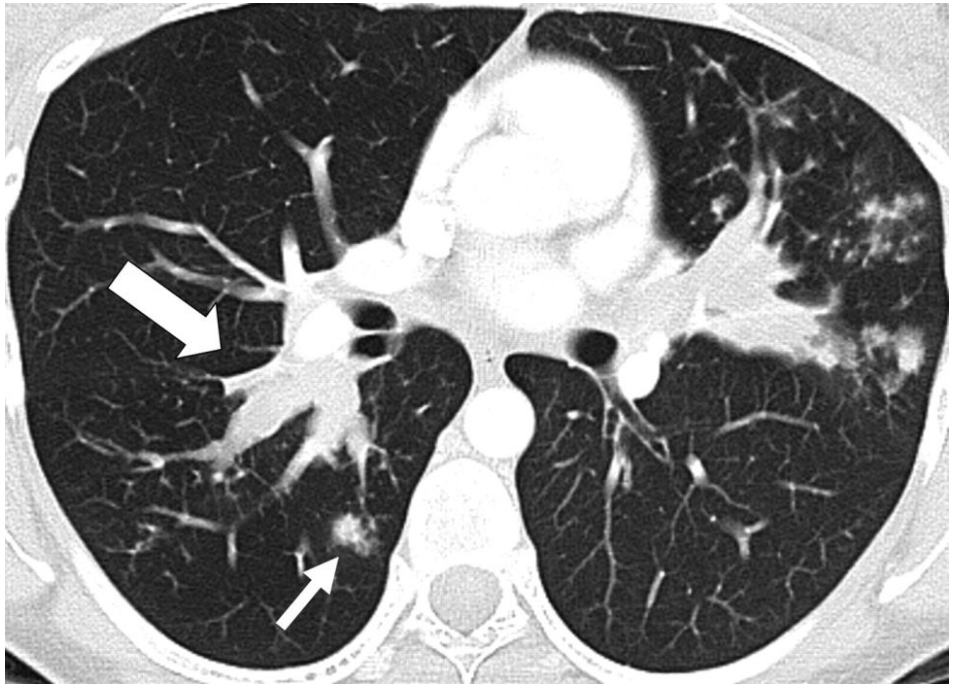
Post-infective

- ulilateral, localized
- Severe infection in case history

Immune deficiency

- Start at early age
- Infections from childhood/infancy if inborn
- Frequent exacerbations
- Pneumonias
- non-respiratory infections

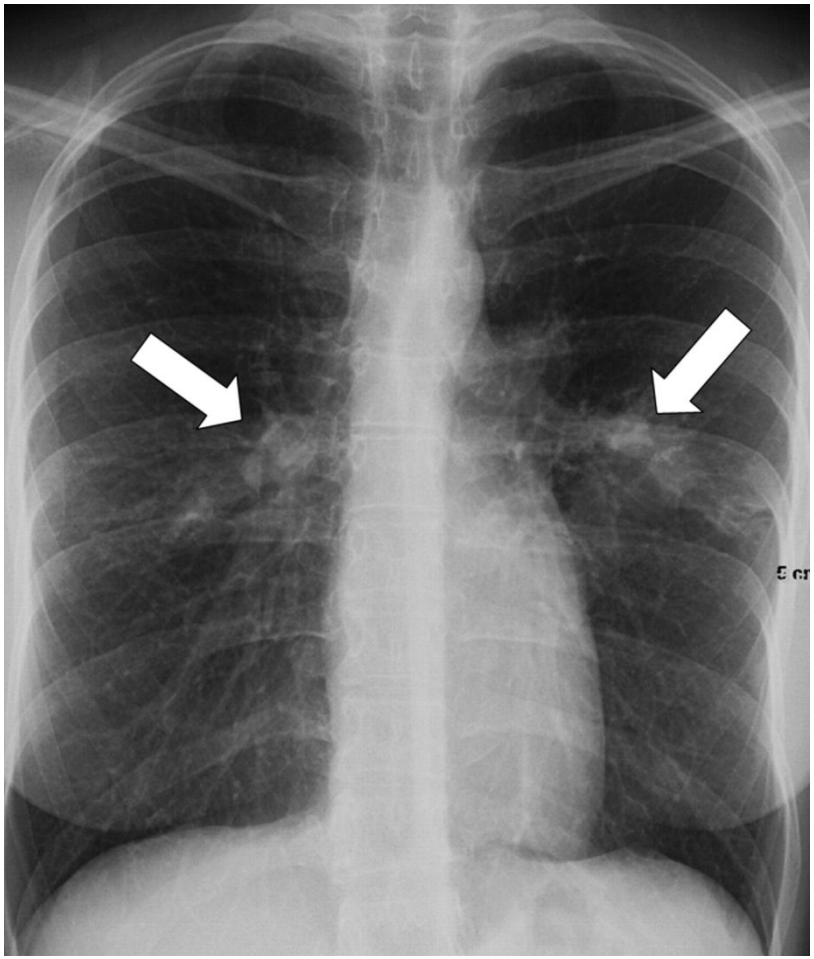
Allergic bronchopulmonary aspergillosis



- Asthma
- Presence of transient pulmonary infiltrates (fleeting shadows)
- Elevated total serum IgE
- Peripheral blood eosinophilia
- Elevated serum IgE and IgG to *Af* Immediate cutaneous reactivity to *Af*
- Precipitating antibodies against *Af*
- Central/proximal bronchiectasis with normal tapering of distal bronchi
- mucoid impaction (*large arrow*)
- distal bronchiolitis (*small arrow*)

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

Same



- central bronchiectasis and mucoid impaction, so-called finger-in-glove appearance

Other investigations: endotypes assessment

- Co-morbidities and past medical history to identify relevant and possibly causative disease
- serum total IgE and specific IgE or skin prick test to *Aspergillus fumigatus in all patients with bronchiectasis*
- Serum IgG, IgA, IgM in all patients with BE

Adam T Hill,1 Anita L Sullivan,2 James D Chalmers British Thoracic Society Guideline for bronchiectasis in adults Thorax 2019;**74(Suppl 1):1–69. doi:10.1136/thoraxjnl-2018-212463**

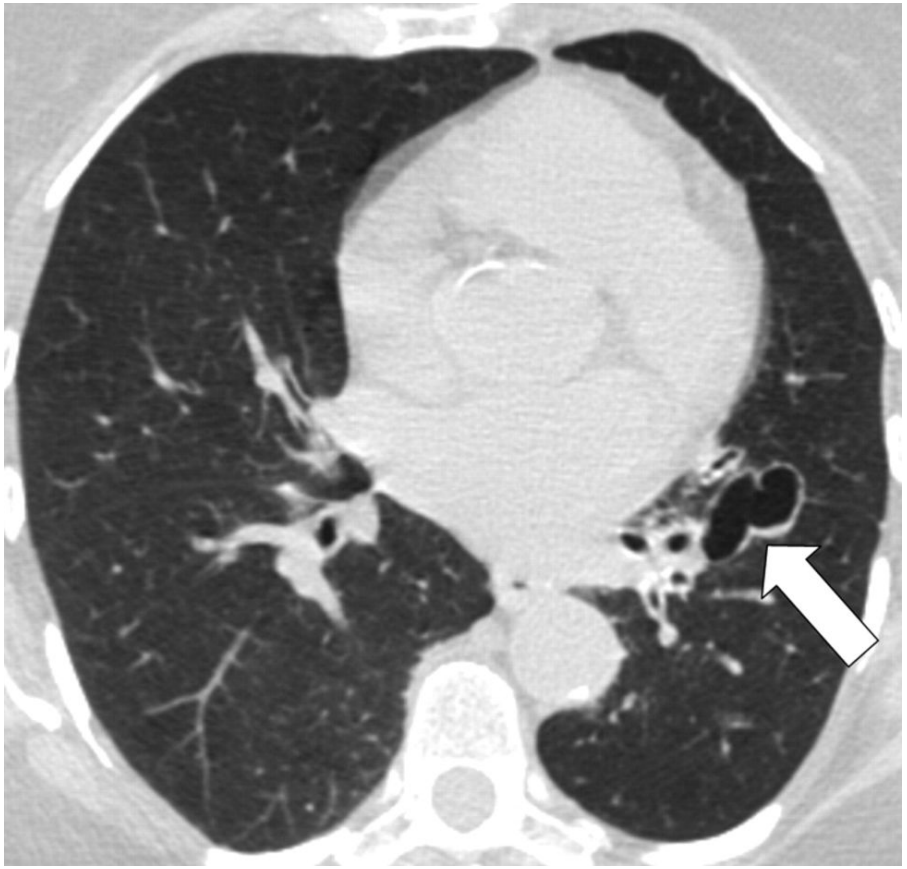
- **Tests for:**
- **cystic fibrosis** - early onset, male infertility, malabsorption, pancreatitis
- **Primary Ciliary Dyskinesia** if supporting clinical features- neonatal distress, symptoms from childhood, recurrent otitis media, rhinosinusitis, or infertility
- **arthritis, connective tissue disease/vasculitis**: rheumatoid factor, anti CCP, antinuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies
- **alpha 1 antitrypsin (A1AT) deficiency:** basal panacinar emphysema
- **reflux and aspiration:** - symptomatic patients/or other suggestive clinical features.
- **Bronchoscopy:** localised disease to rule out an endobronchial lesion or foreign body as the cause of bronchiectasis.
- **Bronchial aspiration/bronchial wash** from CT defined areas of bronchiectasis in patients with no sputum (non tuberculous mycobacteria?)
- **Serum protein electrophoresis:** bronchiectasis with raised immunoglobulins.
- **HIV-1 serology:** clinical features suggestive of increased risk of retroviral infection.

Adam T Hill,1 Anita L Sullivan,2 James D Chalmers British Thoracic Society Guideline for bronchiectasis in adults Thorax 2019;**74(Suppl 1):1–69. doi:10.1136/thoraxjnl-2018-212463**

Other investigations

- Spirogram/functional investigation of lungs, oxygen saturation, blood gases
- Daily protein loss, GFR, urine analysis – for early diagnosis of inflammatory (SAA) amyloidosis
- Other investigations if necessary

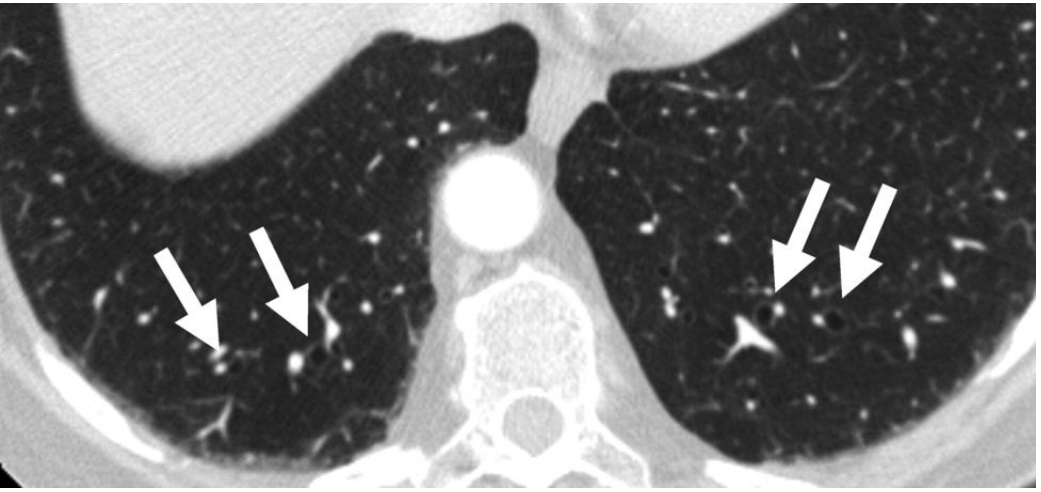
Focal idiopathic in left lower lobe



- non-TB mycobacteria
- Perimenopausal females
- Usually dry bronchoectases
- Focal bronchiectasis frequently

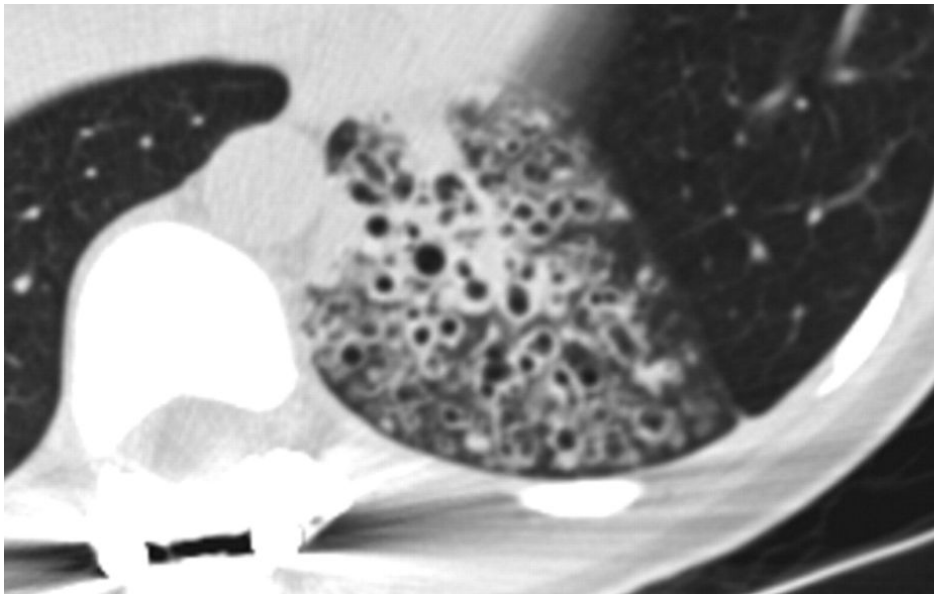
American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3**
Bronchiectasis Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

Idiopathic – lower lobe predominance, different severity



- subtle idiopathic bibasilar cylindric bronchiectasis shows signet-ring sign (*arrows*).

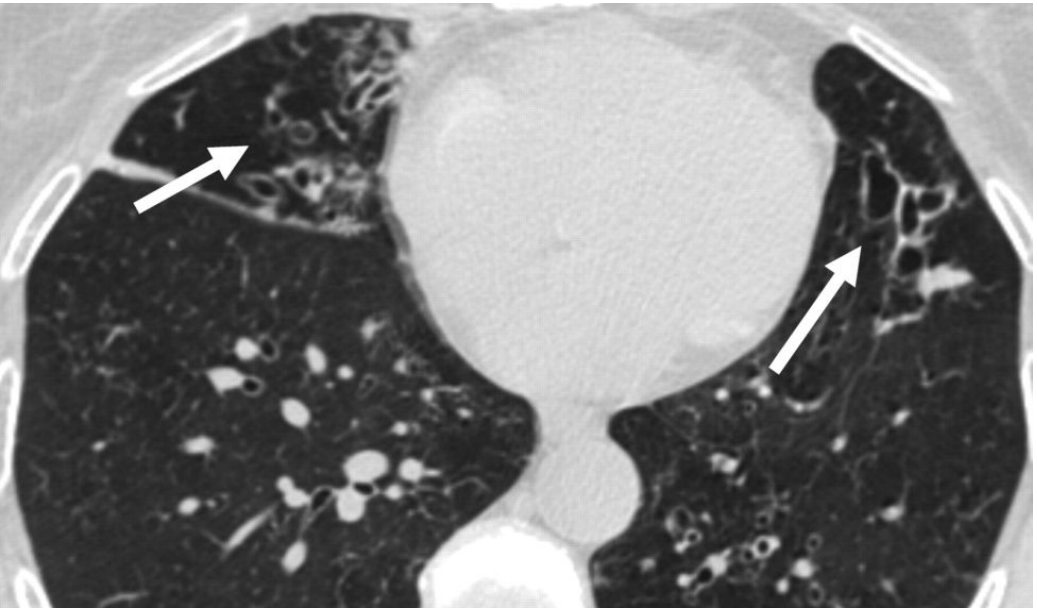
https://www.ajronline.org/doi/10.2214/AJR.09.3053



- marked idiopathic left lower bronchiectasis with volume loss, bronchial wall thickening, and diffuse opacity.

https://www.ajronline.org/doi/10.2214/AJR.09.3053

Mycobacterium avium- intracellulare infection



- Bronchiectasis (*arrows*) predominantly involves right middle lobe and lingula.

American Journal of Roentgenology >
Volume 193, Issue 3 >
Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis**
Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

Obstruction

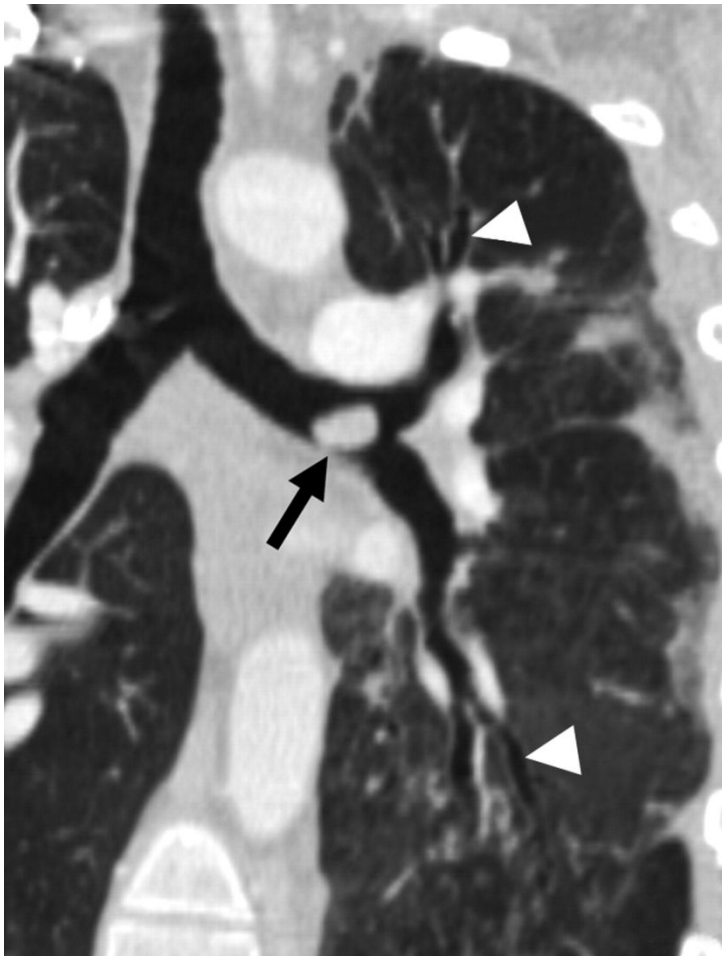
Tumor

- More gradual onset (1-3 mo)
- Dyspnea progression from expiratory to inspiratory
- Dry cough, hemopthisis
- More see “lung cancer”

Foreign body

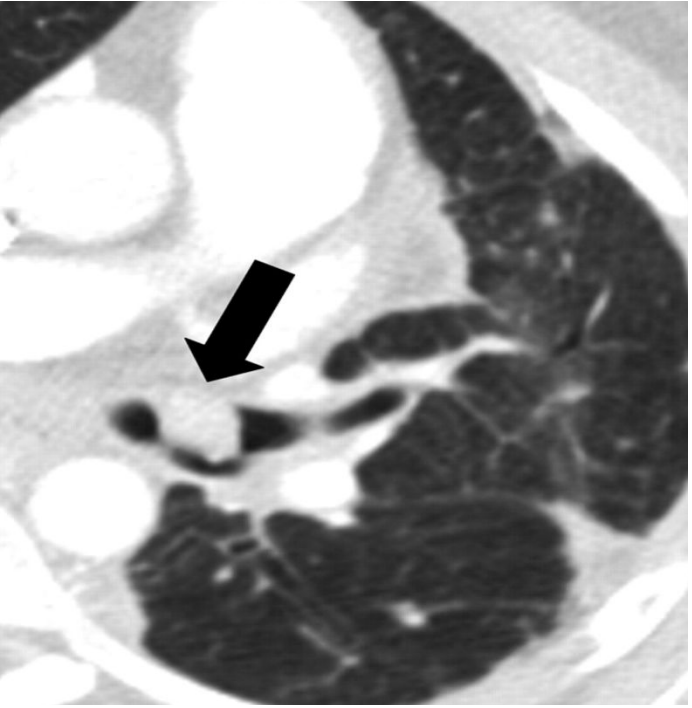
- Usually in children
- May be acute suffocation episode in case history with stridor
- Relapsing pneumonias

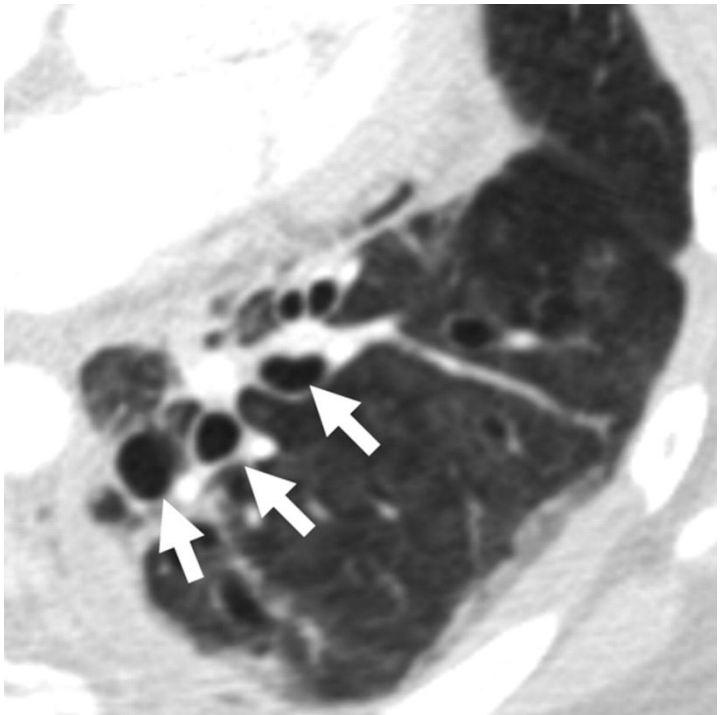
Carcinoid.



- Endobronchial growth
- May arise berofe bifurcation of lobar bronchi
- Serotonin secretion symptoms as following:
- Flushes up to 10-20 times daily
- Bronchospasm
- Restritive CMP of endomyocardial nature

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

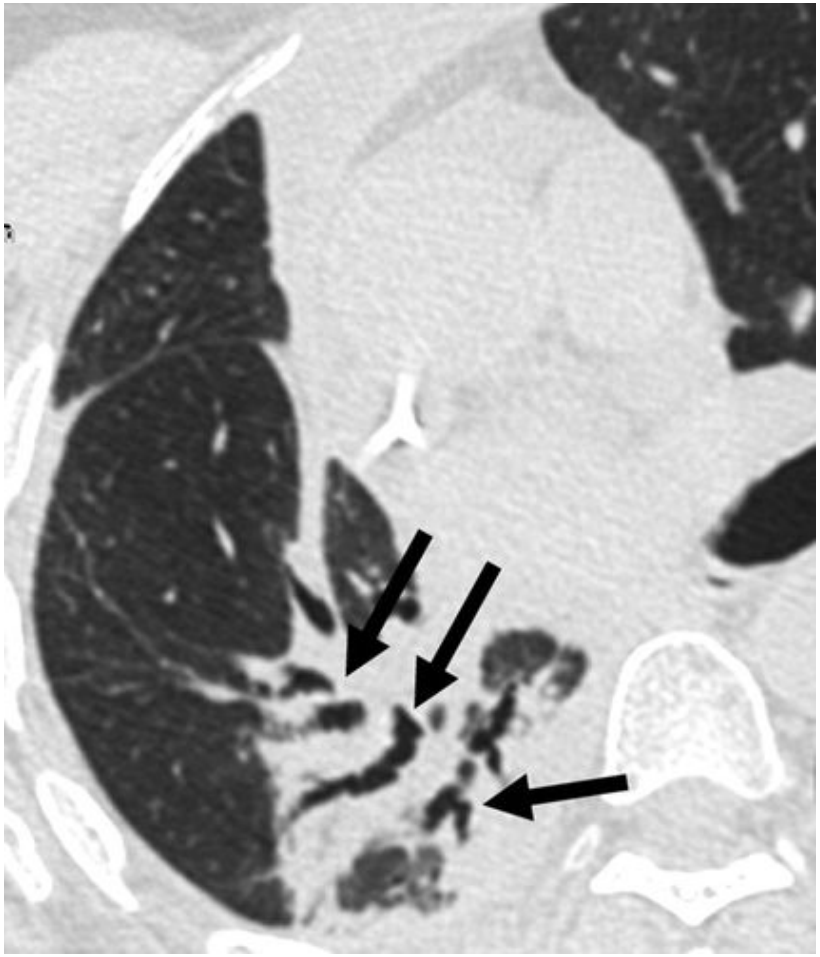




- Carcinoid
- Distal bronchiectases

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3**
Bronchiectasis Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

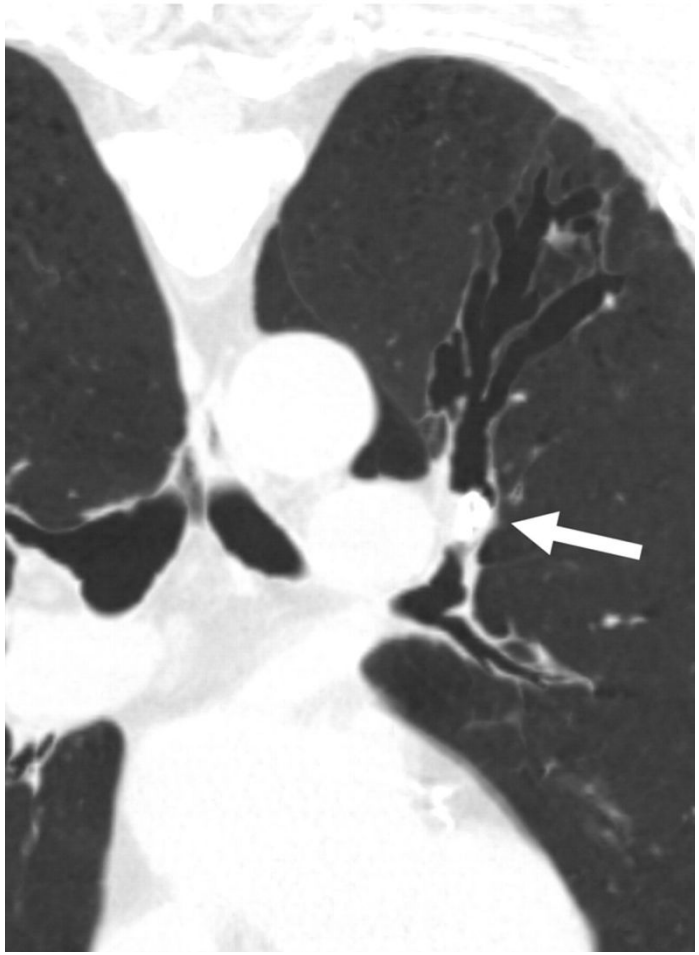
Tumors: postradiation fibrosis



- Right paramediastinal fibrotic changes, developed after treatment of lung cancer, are associated with traction bronchiectasis (*arrows*).

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

Broncholithiasis: post TB



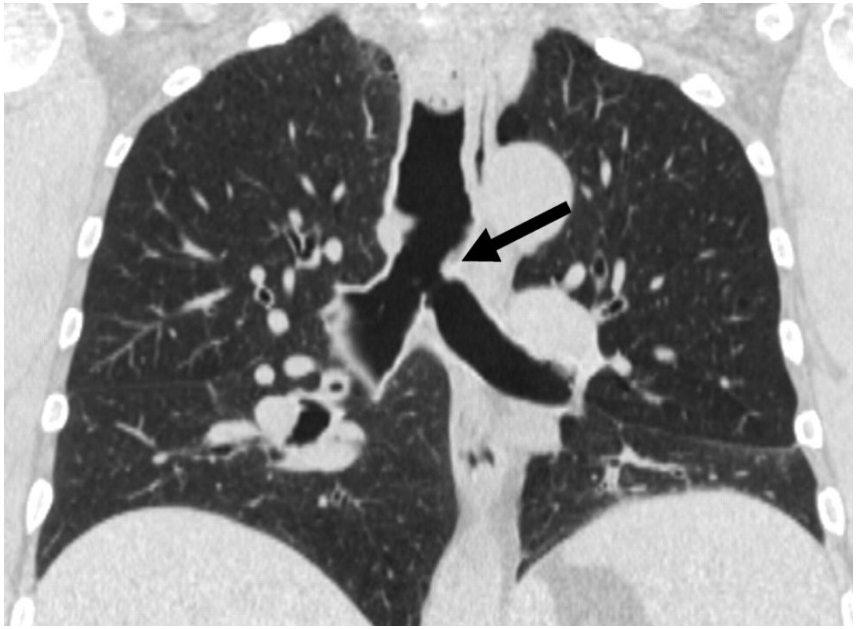
- Calcified left upper lobe endobronchial broncholithiasis
- Results of lymph node calcification (compression and erosion of calcified perigronchial lymph nodes
- Cause – TB, histoplasmosis or other granulomatous disease, more rare foreign body
- 0.1% - 0.2% of all lung diseases.

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

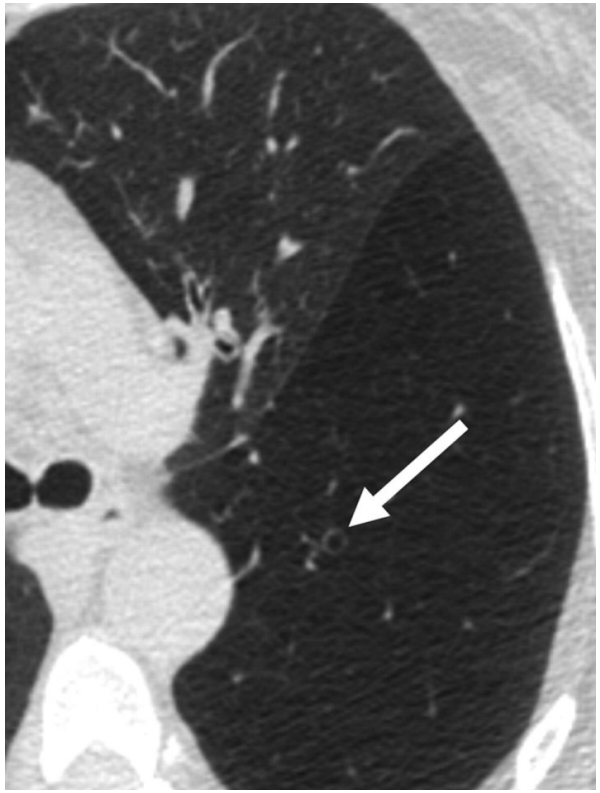
Broncholithiasis: An Uncommon Cause of Chronic Cough
Ungprasert, Patompong MD; Srivali, Narat MD; Bauer, Michael A. MD; Edmonds, Lee C. MD
Author Information Journal of Bronchology & Interventional Pulmonology: January 2014 - Volume 21 - Issue 1 - p 102-103

Congenital abnormalities

- Congenital stenosis
- (left mainstem bronchus

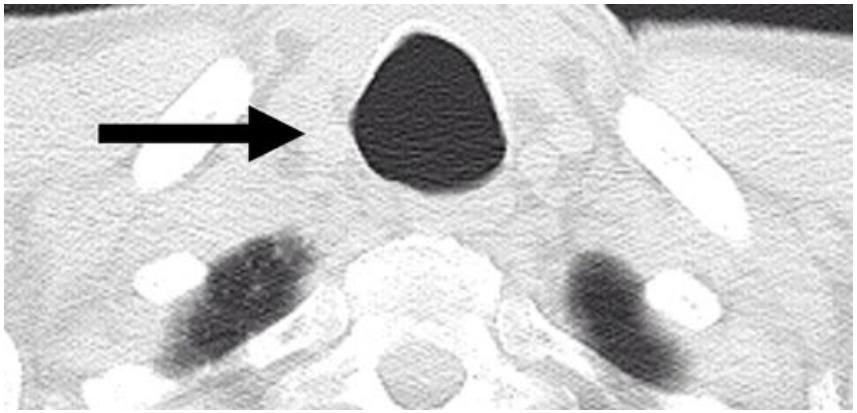


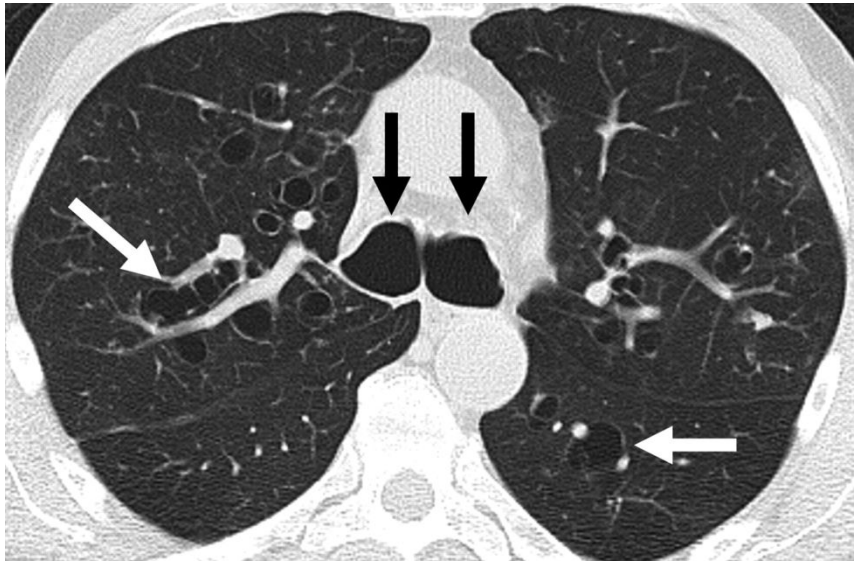
- Bronchial atresia
- focal bronchiectasis (*arrow*) distal to bronchial atresia associated with hyperlucency and hyperexpansion of left lung.



American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3**
Bronchiectasis Luce Cantin[1], Alexander A. Bankier[1] and Ronald

Other causes

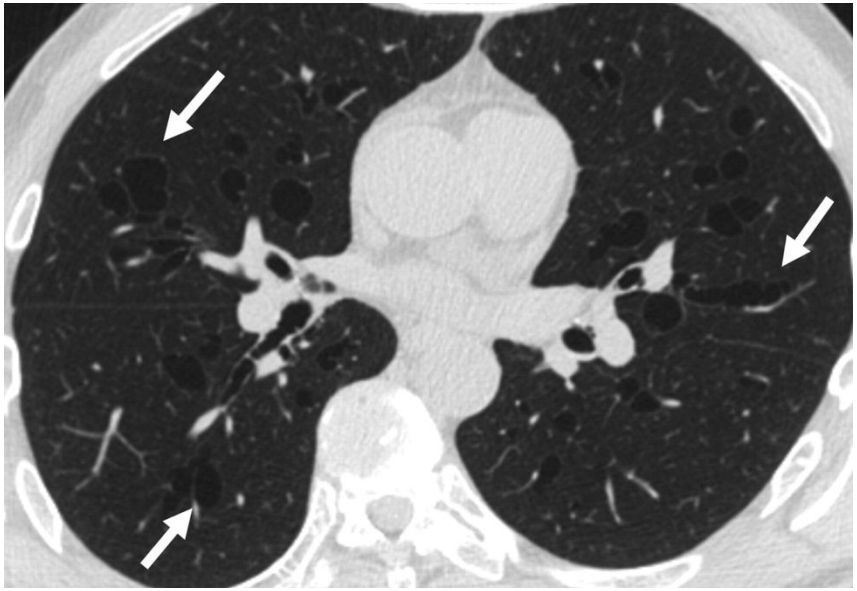




- Mounier-Kuhn's syndrome. Enlarged trachea (*arrow*).
- Enlarged mainstem bronchi (*black arrows*) and distal bronchiectasis (*white arrows*).

American Journal of Roentgenology > Volume 193, Issue 3 > Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis** Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

Williams-Campbell



- mostly varicose and cystic central bronchiectasis (*arrows*).

American Journal of Roentgenology >
Volume 193, Issue 3 >
Bronchiectasis**September 2009, Volume 193, Number 3 Bronchiectasis**
Luce Cantin[1], Alexander A. Bankier[1] and Ronald L. Eisenberg[1] American Journal of Roentgenology. 2009;193: W158-W171. 10.2214/AJR.09.3053

	Age of onset	Radiology	Microbiology	Symptoms or features	Physiology or lung function
Primary ciliary dyskinesia	Usually presents in childhood	Middle or lower lobes	*H influenzae,* any	Chronic rhinosinusitis, recurrent otitis media	Any
Chronic obstructive pulmonary disease	Smokers or ex-smokers older than 40 years	Lower lobe cylindrical bronchiectasis	Any or no bacterial infection	Recurrent exacerbations, sputum production	obstruction (BE more common with more severe airflow obstruction)[56]

Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneityProf Patrick A Flume, MD, Prof James D Chalmers, MBChB, Kenneth N Olivier, MD The Lancet Volume 392, Issue 10150, Pages 880-890 (September 2018)

	Age of onset	Radiology	Microbiology	Symptoms or features	Physiology or lung function
Inflammatory bowel disease	Any	Any lobes affected, bronchiolitis, could include other features of inflammatory bowel disease-associated lung disease	Often no pathogens isolated	Gross bronchorrhea, which is often responsive to corticosteroids	Airflow obstruction
Cystic fibrosis	Young age of onset but can present in adulthood	Upper lobes	*P aeruginosa, S aureus,* others	Rhinosinusitis, infertility, pancreatitis, malabsorption, gastrointestinal symptoms	Airflow obstruction

Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneityProf Patrick A Flume, MD, Prof James D Chalmers, MBChB, Kenneth N Olivier, MD The Lancet Volume 392, Issue 10150, Pages 880-890 (September 2018)

Cystic fibrosis

- Cystic fibrosis (CF) is an autosomal recessive disease
- Loss of function of the cystic fibrosis transmembrane conductance regulator (CFTR) at the apical membrane of airway epithelial cells

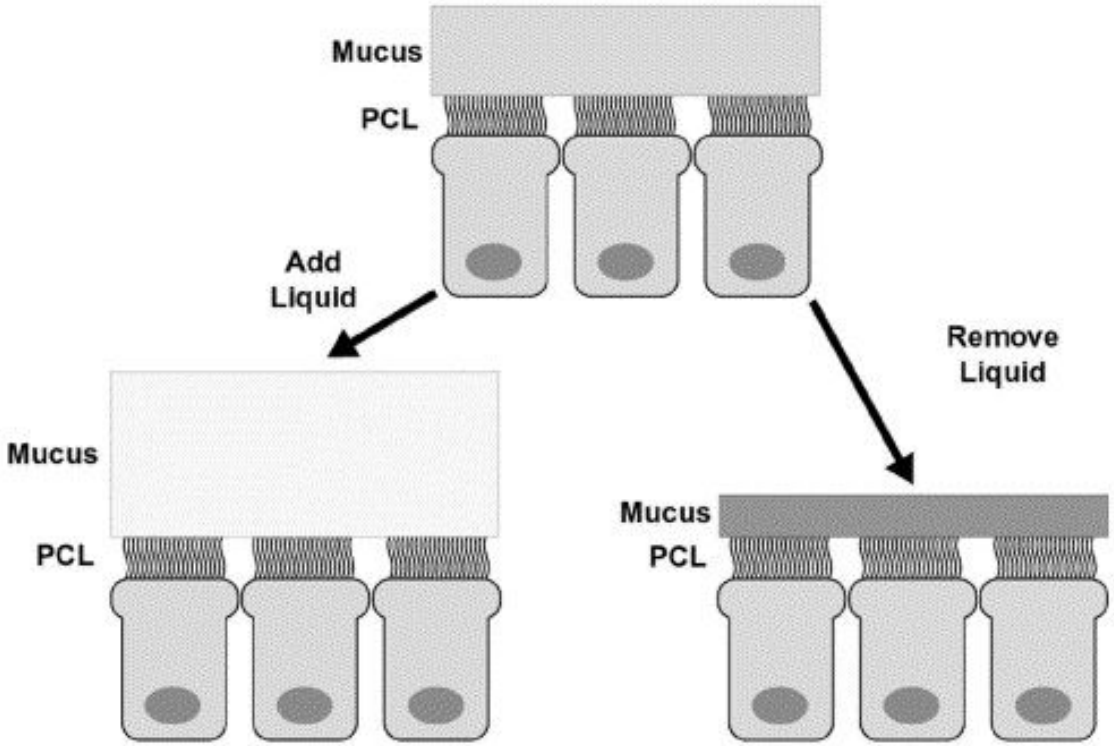
American Journal of Respiratory and Critical Care Medicine
Vol. 187, No. 7 | Apr 01, 2013
Cystic Fibrosis Pulmonary Guidelines Chronic Medications for Maintenance of Lung Health
Peter J. Mogayzel Jr.[1], Edward T. Naureckas [2],

Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneity The lancet Volume 392, Issue 10150, 8–14 September 2018, Pages 880-890

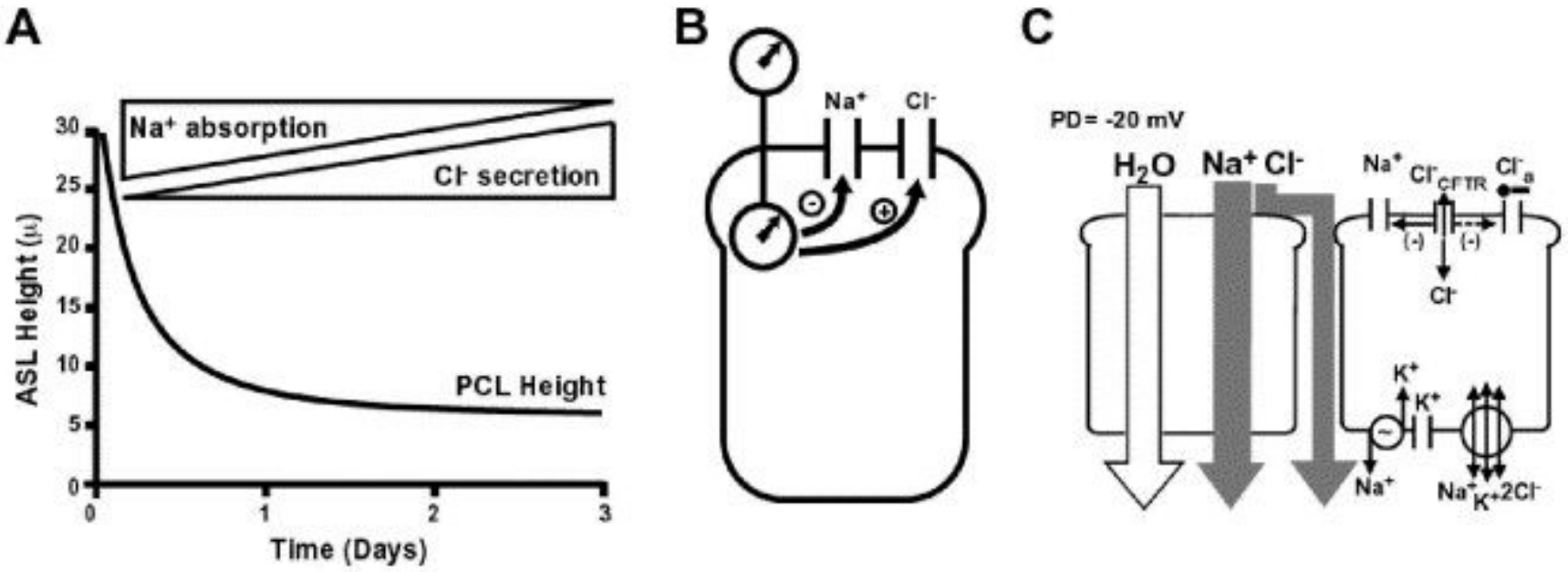
Pathogenesis: hypothesis

- **chemical shield' hypothesis:** in normal condition airway epithelium produces low salt (<50 mM NaCl) airway surface liquid, so defensin-like antimicrobial activities are performed
- importance of isotonic (i.e. ~150 mM NaCl) airway surface liquid volume normally performs efficient mucus clearance

Periciliary liquid layer

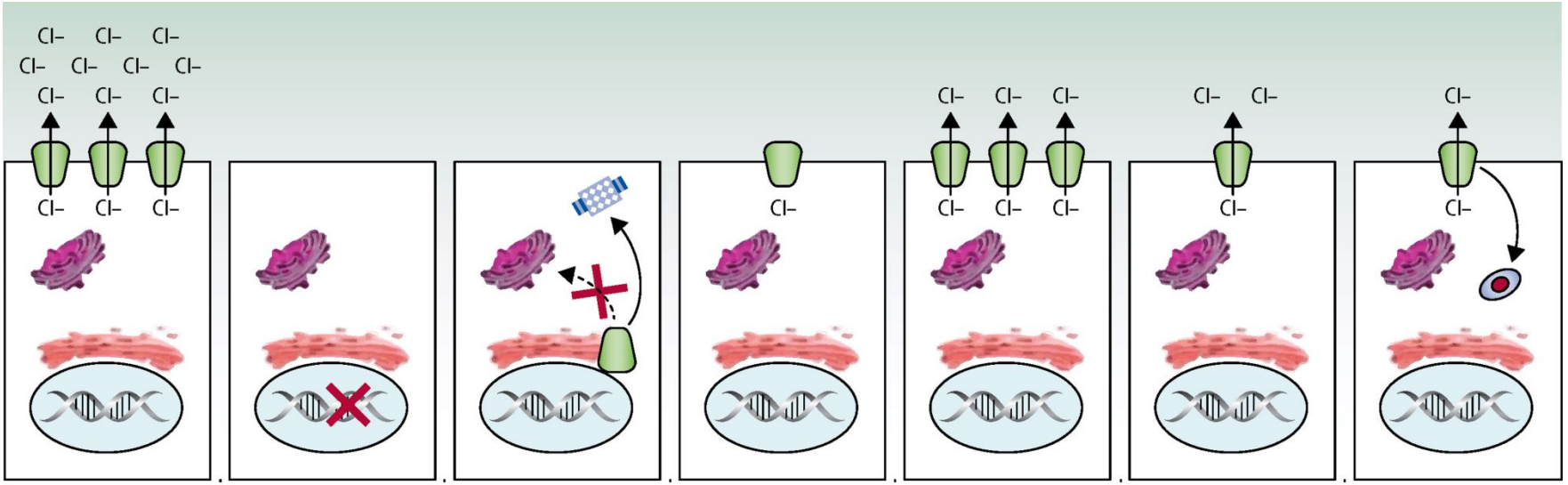


The mucus layer as a liquid reservoir.
Upper panel depicts normal geometry of mucus and periciliary liquid (PCL) layers
Lower left: additional liquid expands the mucus layer
Lower right: removal of liquid can remove ~50% of the mucus layer without perturbing PCL volume



Advanced Drug Delivery Reviews Volume 54, Issue 11, 5 December 2002, Pages 1359-1371 An overview of the pathogenesis of cystic fibrosis lung disease R.Cboucher

CFTR mutations classifications



Wildtype CFTR	I	II	III	IV	V	VI
Defect types	No protein	No traffic	No function	Less function	Less protein	Less stable
Mutation examples	Gly 542 x Arg 553 x Trp 1282 x	Gly 85 Glu Δ Ile 507 Δ Phe 508 Asn 1303 lys	Val 520 Phe Ser 549 Arg Gly 551 Asp	Arg 117 His Arg 334 Trp Ser 1235 Arg	Ala 455 Glu 1680–886 A→G 2657+5 G→A	Δ Phe 508 Gln 1412 x
Required approaches	Rescue protein synthesis	Correct protein folding	Restore channel conductance	Restore channel conductance	Maturation or correct misplicing	Promote protein stability
Approved drugs	..	Lumacaftor, Tezacaftor	Ivacaftor	Ivacaftor	..	..

Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneity The lancet Volume 392, Issue 10150, 8–14 September 2018, Pages 880-890

- Median age at diagnosis- 6-8 months; two thirds of patients are diagnosed by 1 year of age

Primary ciliary dyskinesia

- multiple genes

Idiopathic bronchiectasis associated with non-tuberculous mycobacteria (NTM)

- post-menopausal non-smoker females
- chronic cough
- No predisposing factors
- share characteristics with other endotypes, notably a high prevalence of CFTR mutations and ciliary dysfunction, but do not meet diagnostic criteria for cystic fibrosis or primary ciliary dyskinesia.
- tall, asthenic type, with scoliosis, pectus excavatum, mitral valve prolapse, dural ectasia, minor features overlapping with Marfan and Ehlers-Danlos syndromes

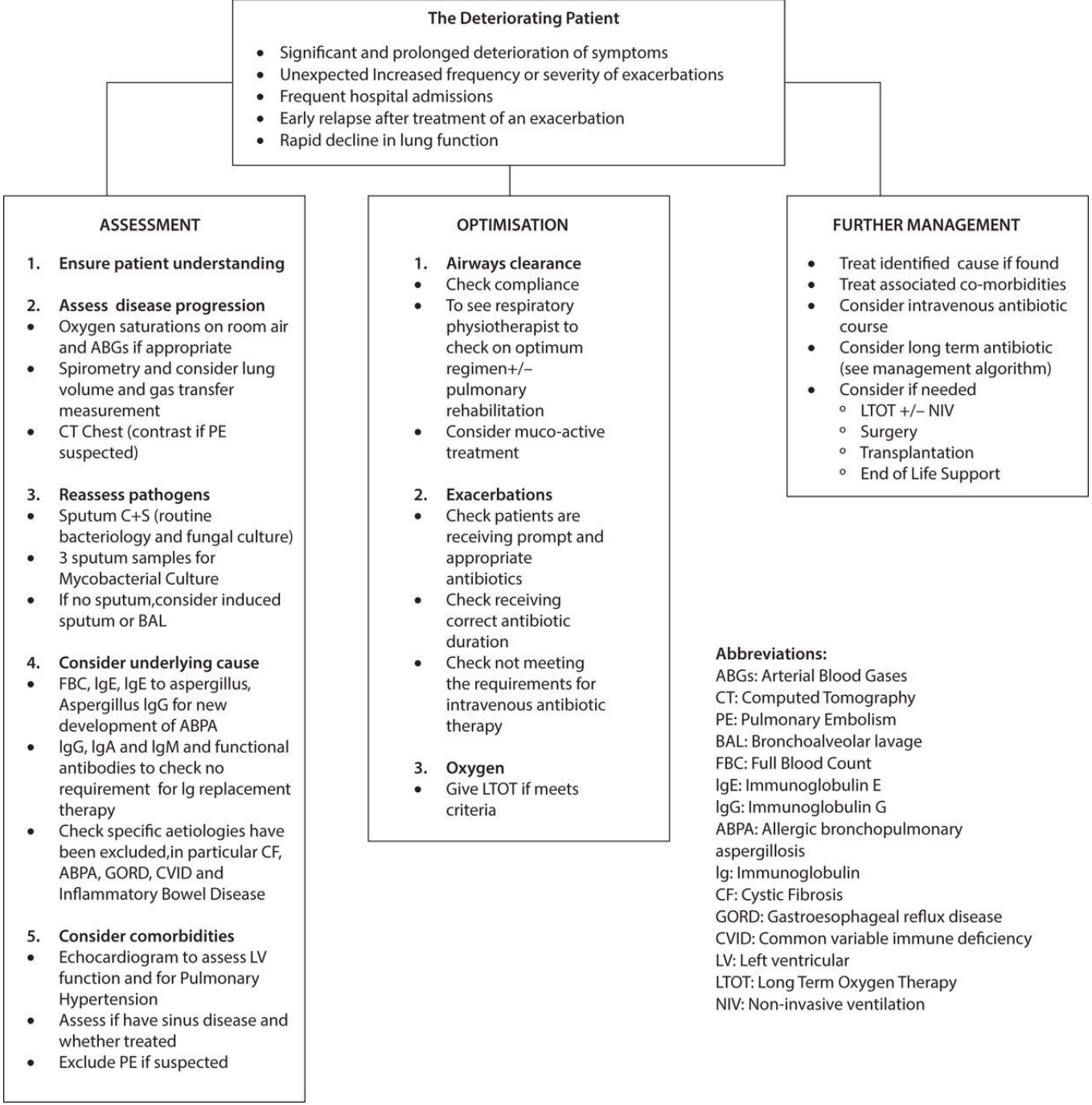
Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneity The lancet Volume 392, Issue 10150, 8–14 September 2018, Pages 880-890

	Age of onset	Radiology	Microbiology	Symptoms or features	Physiology or lung function
Idiopathic	Predom.postmenopausal women	Any radiological pattern	*Ps. aeruginosa, Haemophilus influenzae,* any pathogens or none	Any	Any
Post-infective	Any	Any pattern, unilobular	Any pathogens or none	Typically onset soon after severe infection	Any
Connective tissue disease	Any	Any	Any	Poor prognosis/ rapidly progressive, features of systematic disease[54]	Airflow obstruction (but other patterns seen)
Immune deficiency	Primary immune deficiency often at young age, secondary immune deficiency at any age	Lower lobe	Any	Frequent exacerbations, pneumonia, non-respiratory infections	Airflow obstruction

Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneityProf Patrick A Flume, MD, Prof James D Chalmers, MBChB, Kenneth N Olivier, MD The Lancet Volume 392, Issue 10150, Pages 880-890 (September 2018)

	Age of onset	Radiology	Microbiology	Symptoms or features	Physiology or lung function
Allergic bronchopulmonary aspergillosis	Any	Central bronchiectasis, infiltrates	Typically*Staphylococcus aureus*[55]	Thick sputum, wheeze, recurrent exacerbations, background of asthma	Airflow obstruction
Non-tuberculous mycobacteria	Post-menopausal at any age	Middle lobe and lingula bronchiectasis, tree in bud, nodular changes	In addition to non-tuberculous mycobacteria, can have typical bacteria such as *P aeruginosa*	Dry bronchiectasis, chronic cough, malaise, weight loss, systemic features, low body-mass index, scoliosis, pectus excavatum	Any

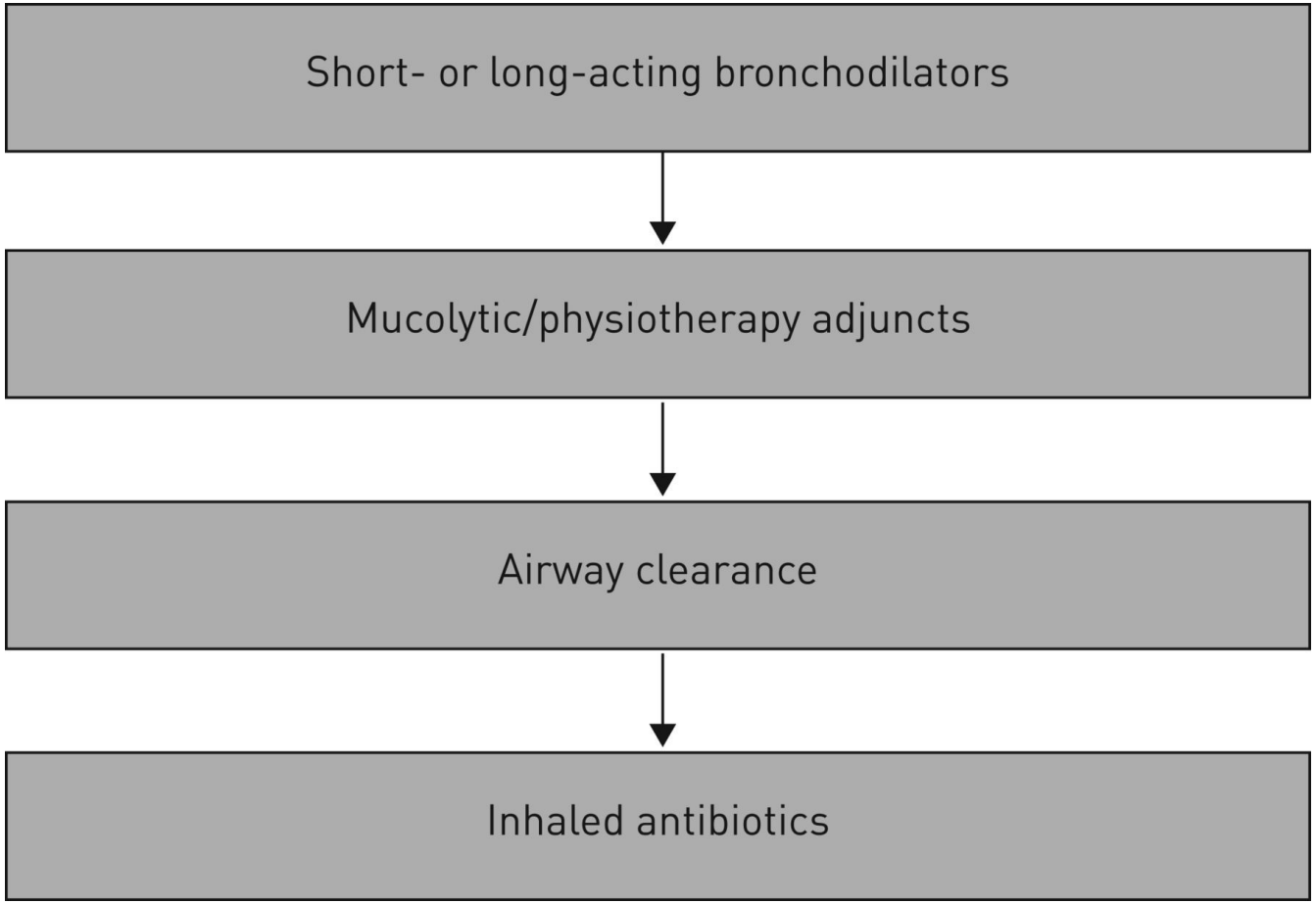
Advances in bronchiectasis: endotyping, genetics, microbiome, and disease heterogeneityProf Patrick A Flume, MD, Prof James D Chalmers, MBChB, Kenneth N Olivier, MD The Lancet Volume 392, Issue 10150, Pages 880-890 (September 2018)



Abbreviations:
ABGs: Arterial Blood Gases
CT: Computed Tomography
PE: Pulmonary Embolism
BAL: Bronchoalveolar lavage
FBC: Full Blood Count
IgE: Immunoglobulin E
IgG: Immunoglobulin G
ABPA: Allergic bronchopulmonary aspergillosis
Ig: Immunoglobulin
CF: Cystic Fibrosis
GORD: Gastroesophageal reflux disease
CVID: Common variable immune deficiency
LV: Left ventricular
LTOT: Long Term Oxygen Therapy
NIV: Non-invasive ventilation

T Hill A, L Sullivan A, D Chalmers J*, et al* British Thoracic Society Guideline for bronchiectasis in adults *Thorax* 2019;**74:**1-69.

Initial treatment

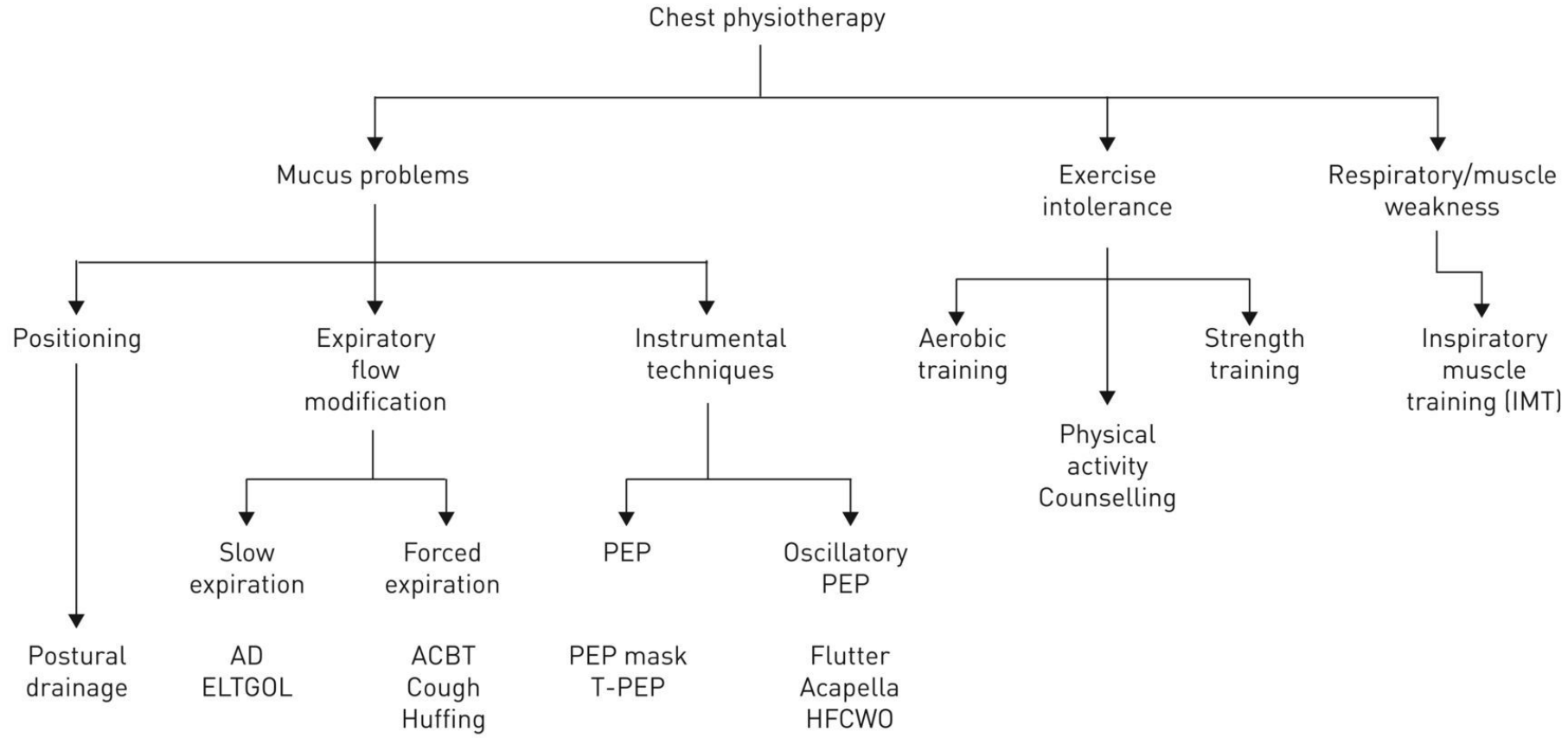


European Respiratory Society guidelines for the management of adult bronchiectasis
Eva Polverino, Pieter C. Goeminne, Melissa J. McDonnell, Stefano Aliberti, Sara E. Marshall, Michael R. Loebinger, European Respiratory Journal 2017 50: 1700629;

- Offer annual influenza immunisation to all patients with bronchiectasis. (D)
- Offer polysaccharide pneumococcal vaccination to all patients with bronchiectasis.

T Hill A, L Sullivan A, D Chalmers J*, et al*
British Thoracic Society Guideline for bronchiectasis in adults
Thorax 2019;**74:**1-69.

Physiotherapy – drainage promotion



AD: autogenic drainage; ELTGOL: total slow expiration with open glottis and infralateral position; ACBT: active cycle of breathing techniques; PEP: positive expiratory pressure; T-PEP: temporary positive expiratory pressure; HFCWO: high frequency chest wall oscillation.

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Airway clearance techniques

- should be taught by a respiratory physiotherapist.
- Patients admitted with an exacerbation of bronchiectasis should be seen daily by a respiratory physiotherapist until their airway clearance is optimised.
- CT imaging should be reviewed to complement the physiotherapy assessment. Where indicated, this information could be used in order to teach the patient the appropriate postural drainage position(s) for their affected bronchopulmonary segment(s).

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- Consider autogenic drainage, positive expiratory pressure, high frequency chest wall oscillation and intrapulmonary percussive ventilation as an alternative airway clearance technique if other techniques are not effective or acceptable to the patient.
- Patients should be encouraged to perform regular physical exercise (plus the forced expiration technique/huff) to promote airway clearance.
- If there is ongoing haemoptysis, refer back to the respiratory physiotherapist to determine the optimum airways clearance technique.
- Advise individuals to perform their airway clearance technique for a minimum of 10 minutes (up to a maximum of 30 minutes). After this time they should continue until two clear huffs or coughs are completed, or until the patient is starting to become fatigued.

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Airway clearance techniques during an acute exacerbation

- Manual techniques may be offered to enhance sputum clearance when the patient is fatigued or undergoing an exacerbation.
- Consider intermittent positive pressure breathing or non-invasive ventilation during an acute exacerbation to offload the work of breathing so fatigued and/or breathless patients can tolerate a longer treatment session and can adopt postural drainage positions.

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Mucoactives in bronchiectasis

- Do not routinely use recombinant human DNase in adults with bronchiectasis.
- Consider the use of humidification with sterile water or normal saline to facilitate airway clearance.
- Consider a trial of mucoactive treatment in patients with bronchiectasis who have difficulty in sputum expectoration.
- Perform an airway reactivity challenge test when inhaled mucoactive treatment is first administered.
- Consider pre-treatment with a bronchodilator prior to inhaled or nebulised mucoactive treatments especially in individuals where bronchoconstriction is likely (patients with asthma or bronchial hyper-reactivity and those with severe airflow obstruction FEV_1<1 litre).
- If carbocysteine is prescribed, a 6 month trial should be given and continued if there is ongoing clinical benefit.

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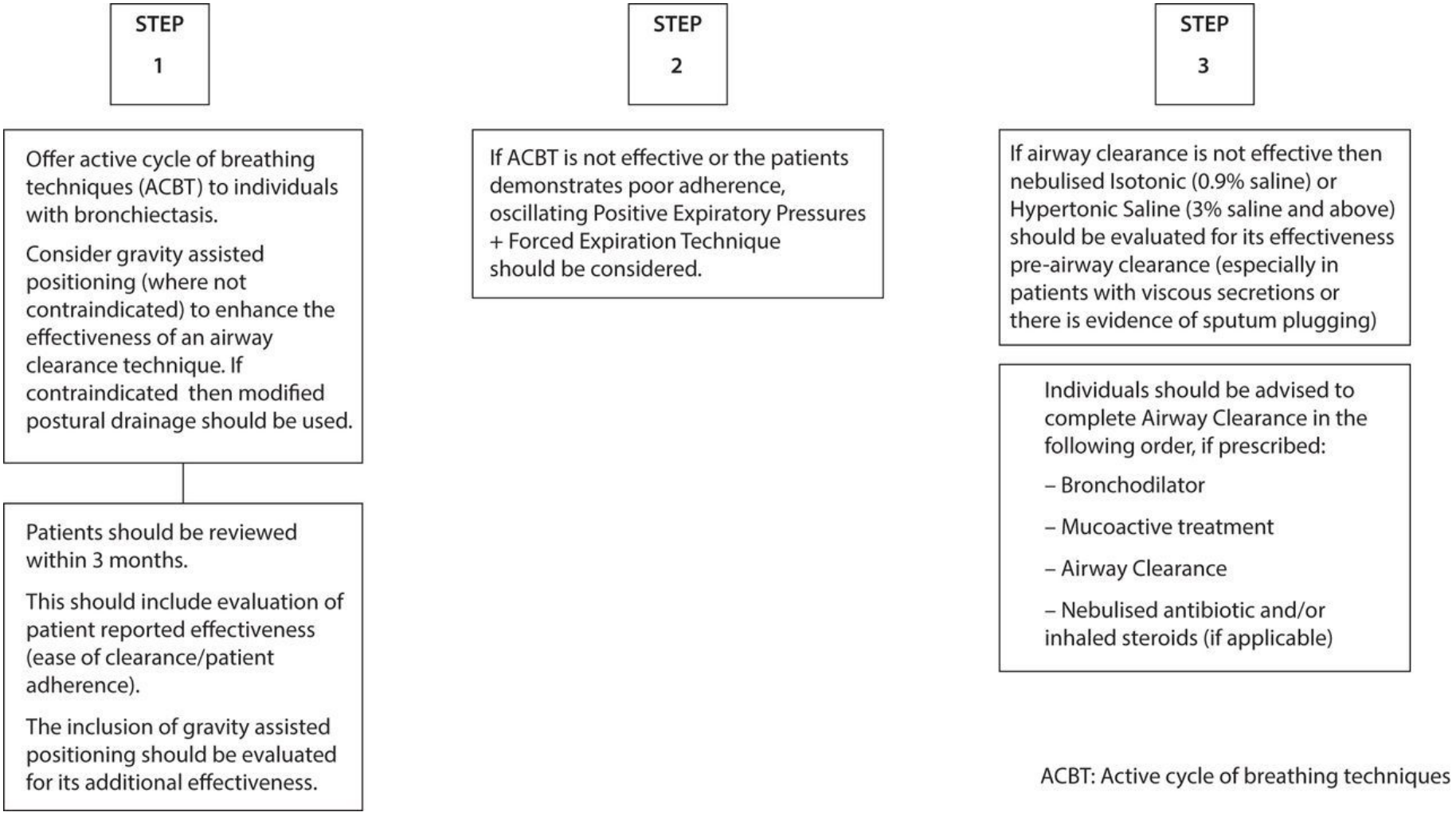

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STEP 1

Increase airway clearance frequency. E.g.: from twice daily to three/four times daily.

STEP 2

Commence the use of mPD or PD if tolerated.

For individuals with radiological changes, PD or mPD should be targeted appropriately.

STEP 3

Individuals with ongoing difficulty with airway clearance may benefit from the addition of other techniques. It is recommended that these should be commenced and evaluated in the following order (unless contraindicated)

1. Enhanced humidification / hydration of airways if secretions viscous (isotonic (0.9% saline) or hypertonic saline (3% saline and above)/humidification/increased fluid intake)
2. Manual Techniques
3. Positive pressure devices including Intermittent Positive Pressure Breathing (IPPB) or Non Invasive Ventilation (NIV) to be used during Airway Clearance

PD=postural drainage; mPD= modified postural drainage

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Inhaled GCS:

- Do not offer long-term oral corticosteroids for patients with bronchiectasis without other indications (such as ABPA, chronic asthma, COPD, inflammatory bowel disease). (D)
- Inhaled corticosteroids have an established role in the management of asthma and in a proportion of patients with COPD which are common co-morbid conditions in bronchiectasis.

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PDE inhibitors, CXCR2 antagonists, statins etc

- Do not routinely offer phosphodiesterase type 4 (PDE4) inhibitors, methylxanthines or leukotriene receptor antagonists for bronchiectasis treatment. (D)
- Do not routinely offer CXCR2 antagonists, neutrophil elastase inhibitors or statins for bronchiectasis treatment. (B)

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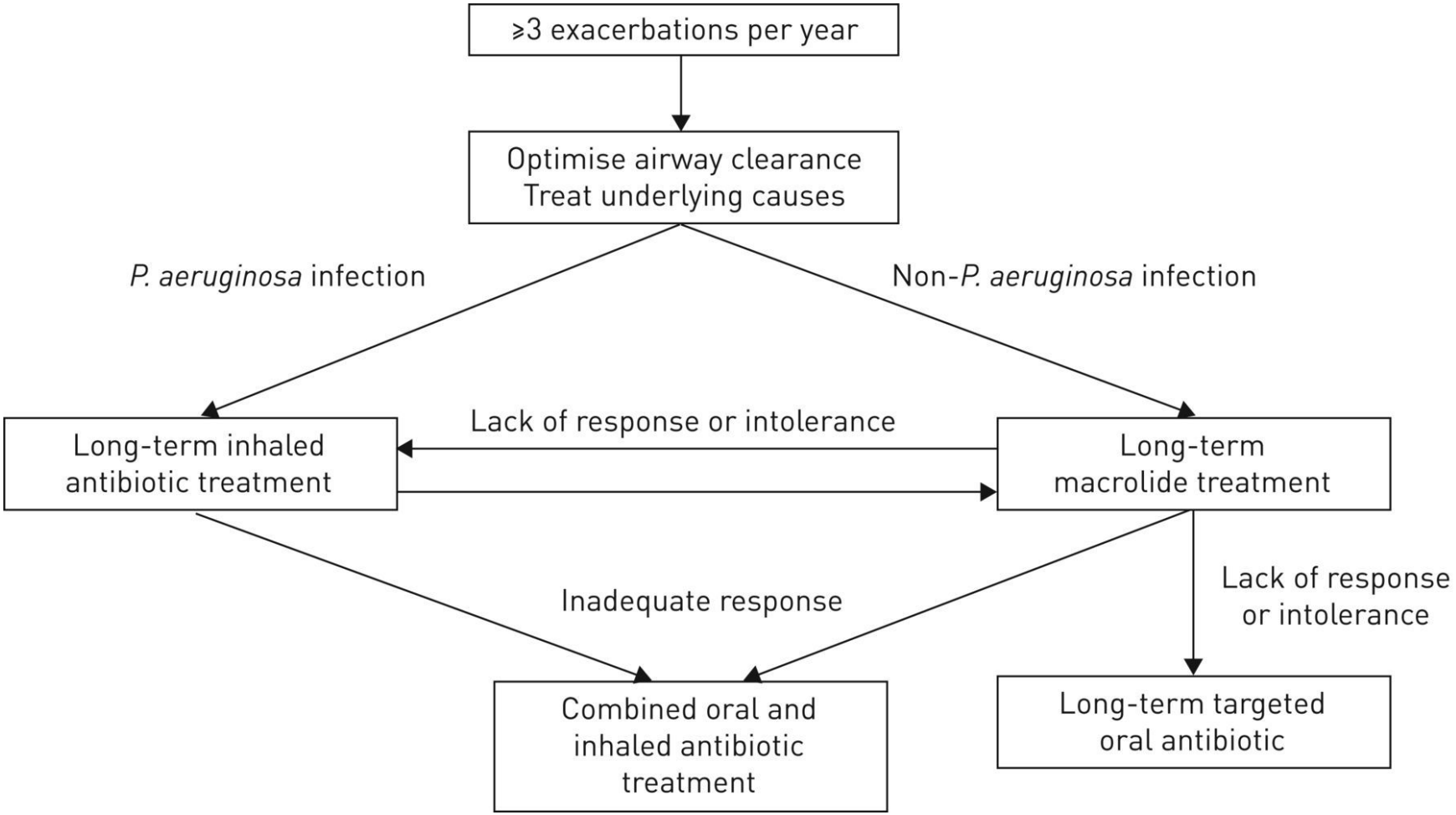
Antibiotics



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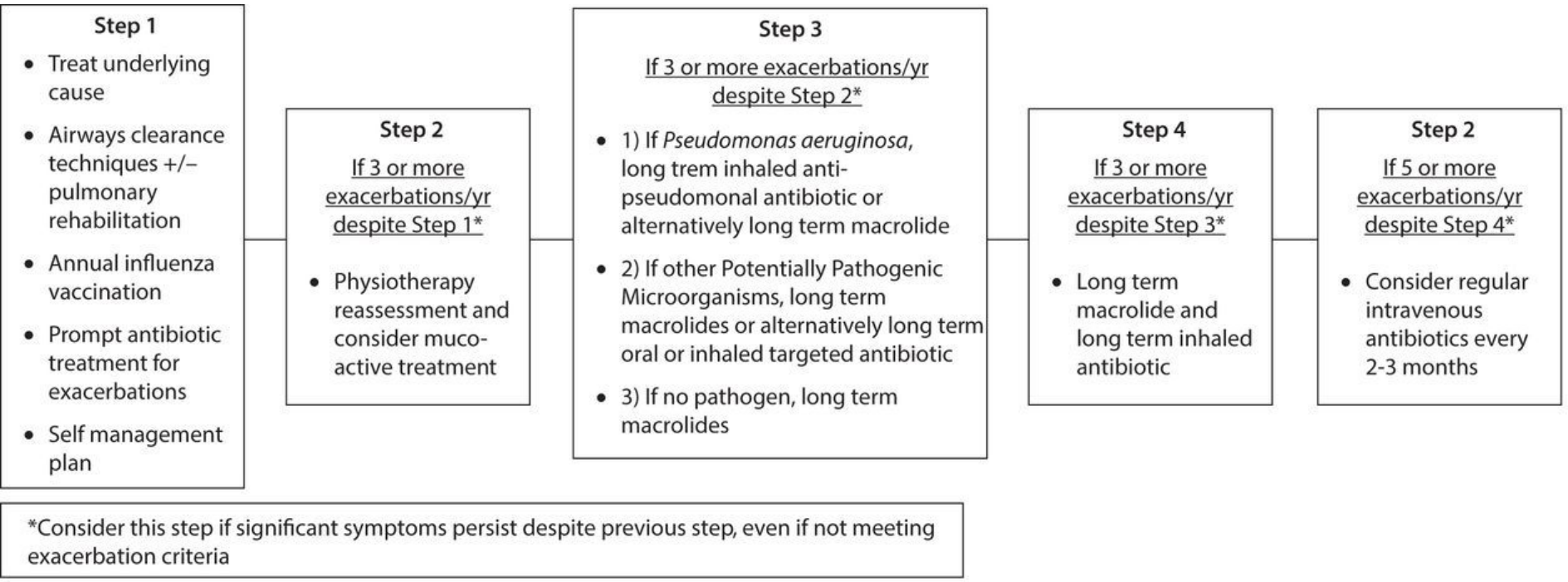

Antibiotics are used to treat exacerbations that present with an acute deterioration (usually over several days) with worsening local symtoms (cough, increased sputum volume or change of viscosity, increased sputum purulence with or without increasing wheeze, breathlessness, haemoptysis) and/or systemic upset. The flow diagram refers to three or more annual exacerbations.

Consider long term antibiotics in patients with bronchiectasis who experience 3 or more exacerbations per year. (A)

Non- P. aeruginosa colonised patients

e. Use azithromycin or erythromycin for patient with bronchiectasis. (A)

f. Consider inhaled gentamicin as a second line alternative to azithromycin or erythromycin.

g. Consider doxycycline as an alternative in patients intolerant of macrolides or in whom they are ineffective. (C)

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Safety

- Prior to starting long term macrolides, for safety reasons:
- (1) ensure no active NTM infection with at least one negative respiratory NTM culture;
- (2) use with caution if the patient has significant hearing loss needing hearing aids or significant balance issues.
- Prior to starting long term inhaled aminoglycosides, for safety reasons:
- (1) avoid using if creatinine clearance <30ml/min;
- (2) use with caution if the patient has significant hearing loss needing hearing aids or significant balance issues;
- (3) avoid concomitant nephrotoxic medications.

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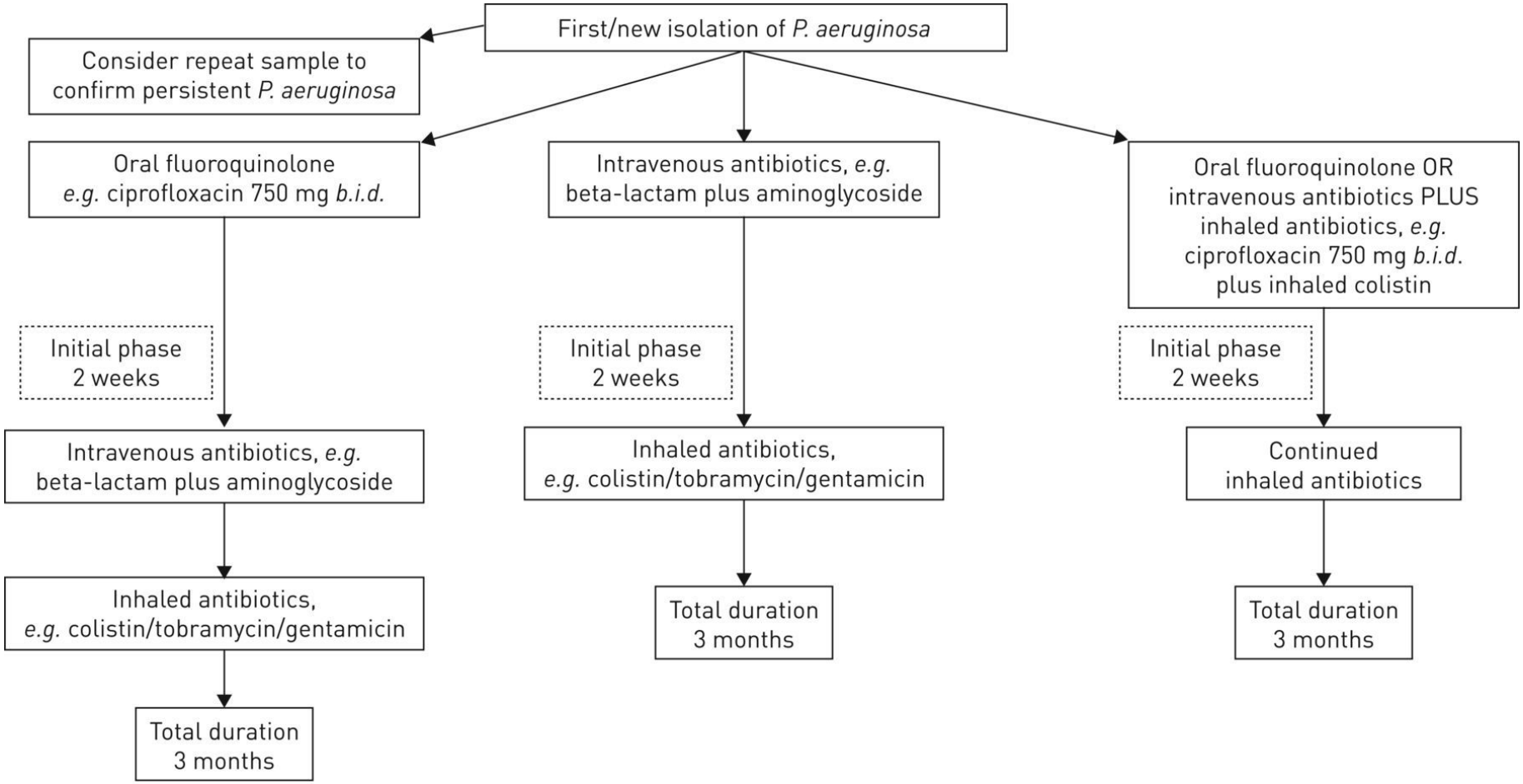
Ps.aeruginosa



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- Offer patients with bronchiectasis associated with clinical deterioration and a new growth of *P. aeruginosa* (1st isolation or regrowth in the context of intermittently positive cultures) eradication antibiotic treatment.
- first line treatment: ciprofloxacin 500–750 mg bd for 2 weeks;
- second line treatment: iv antipseudomonal beta-lactam ± an iv aminoglycoside for 2 weeks, followed by a 3 month course of nebulised colistin, gentamicin or tobramycin).

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- Offer patients with bronchiectasis associated with clinical deterioration and a new growth of methicillin-resistant *S. aureus* (MRSA) (1st isolation or regrowth in the context of intermittently positive cultures) eradication. This should be attempted especially in view of infection control issue

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- Consider long term oxygen therapy for patients with bronchiectasis and respiratory failure, using the same eligibility criteria as for COPD. (D)
- Consider domiciliary non-invasive ventilation with humidification for patients with bronchiectasis and respiratory failure associated with hypercapnia, especially where this is associated with symptoms or recurrent hospitalisation.

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- Consider lung resection in patients with localised disease whose symptoms are not controlled by medical treatment optimised by a bronchiectasis specialist. (D)
- Consider transplant referral in bronchiectasis patients aged 65 years or less if the FEV_1 is <30% with significant clinical instability or if there is a rapid progressive respiratory deterioration despite optimal medical management. (D)
- Consider earlier transplant referral in bronchiectasis patients with poor lung function and the following additional factors: massive haemoptysis, severe secondary pulmonary hypertension, ICU admissions or respiratory failure (particularly if requiring NIV).(D

Hill AT, Sullivan AL, Chalmers JD, *et al*. British Thoracic Society Guideline for bronchiectasis in adults. *Thorax* 2019;**74**:1-69.

allergic broncho-pulmonary aspergillosis

- Offer oral corticosteroid to patients with active ABPA. An initial dose of 0.5 mg/kg/d, for 2 weeks is recommended. Wean steroids according to clinical response and serum IgE levels. (D)
- Consider itraconazole as a steroid sparing agent for patients dependent on oral corticosteroids where difficulty in weaning is experienced. (B)
- Monitor patients with active ABPA with total IgE level to assess treatment response

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