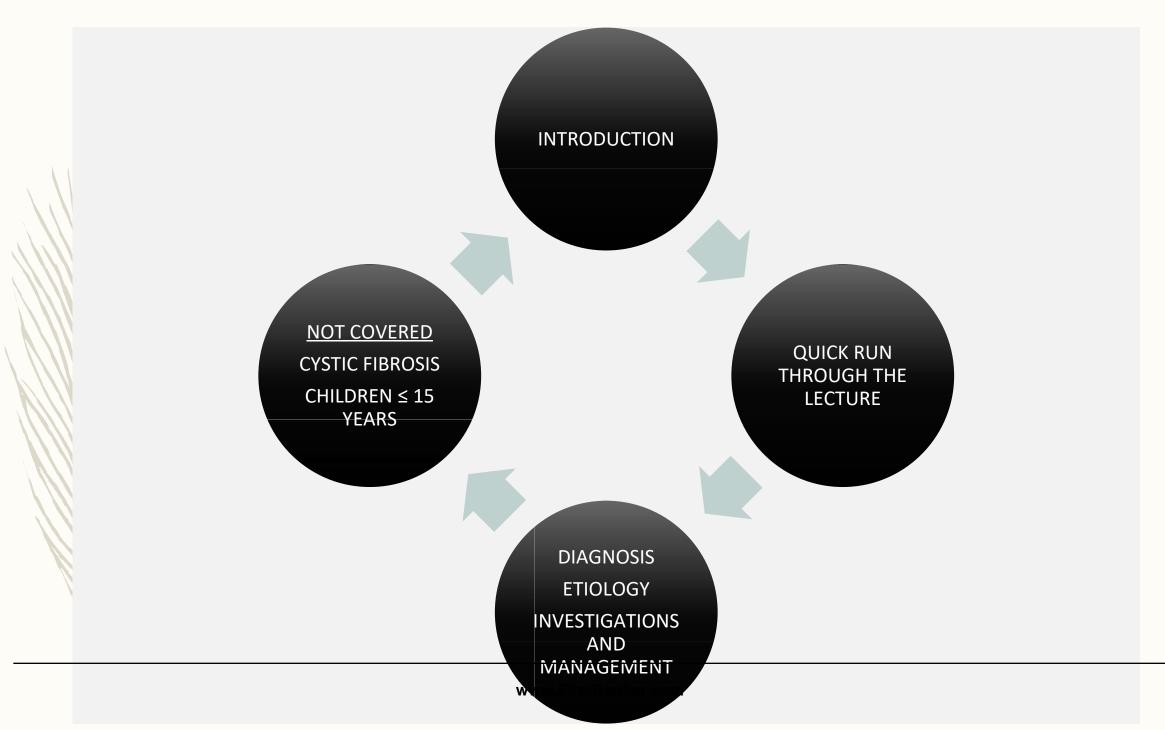




APPROACH TO BRONCHIECTASIS & LUNG ABSCESS







LEARNING OBJECTIVES

- Definition
- Etiology
- Pathogenesis
- Clinical manifestations
- Diagnosis
- Treatment

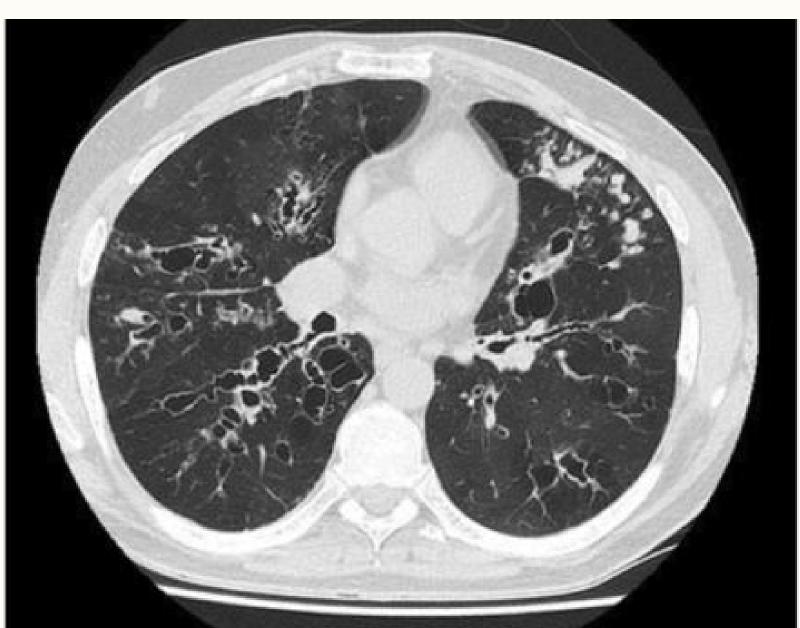
CASE 1



- Long history of respiratory problems starting in early childhood.
- Previously diagnosed as asthma.
- Frequent absence from work due to "recurrent chest infections".
- Unaware of any neonatal issues but believes that he was born at home without complications and is unsure of any previous tests he has had as he is now estranged from his parents.
- Has a cousin with a "lung disease".
- Married but has "no kids"







INVESTIGATIONS

- Sputum culture: P. aeruginosa
- Sweat chloride = 73 meq/liter
- Cystic fibrosis genetics: genotype was F508del/R117H
- CYSTIC FIBROSIS: Multisystem disorder caused by mutations in the gene that encodes the CF transmembrane conductance regulator (CFTR) protein, a chloride channel expressed in epithelial cells.
- More than 2000 CFTR mutations have been identified to date, but only the functional importance of a small number is known to cause the disease







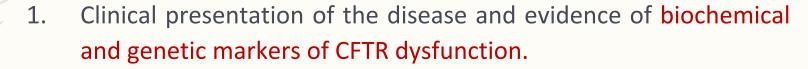
HRCT THORAX

- An upper lobe predominant distribution of cylindrical, cystic and varicose bronchiectasis associated with airway wall thickening, mucus plugging and parenchymal opacities on a HRCT scan should raise the suspicion of CF disease.
- The presence of nasal polyposis and/or chronic rhinosinusitis, recurrent pancreatitis, malabsorption, diabetes, osteoporosis and male infertility are other typical features of CF

DIAGNOSIS

Guidelines published by the Cystic Fibrosis Foundation in the

USA allows diagnosis if:



- 2. Clinical features of the disease with concentration of chloride >60 mmol·L⁻¹ at the sweat test or a concentration in the intermediate range (30–59 mmol·L⁻¹) but two disease-causing CFTR mutations.
- 3. CFTR genotype is undefined: CFTR physiologic tests, such as nasal potential difference and intestinal current measurement, should be performed.





MANAGEMENT

- 1. CFTR modulator therapies
- 2. Airway clearing techniques
- 3. Chest physical therapy
- 4. Humidification with sterile water or normal saline to facilitate airway clearance
- 5. Antibiotics
- 6. Mucus thinners
- 7. Lung transplantation

CASE 2



- Complaints: Decline in his exercise tolerance and an increase in cough which has become productive of purulent sputum with occasional thick/solid components.
- Respiratory exacerbations not responding well to standard steroid and antibiotic treatment.
- He was noted to have variable pulmonary infiltrates on chest radiographs during these episodes







INVSETIGATIONS

- Marked peripheral blood eosinophilia
- Total IgE > 1000 IU/ ml
- Aspergillus specific IgE > 0.35

ABPA: ABPA is an inflammatory disease caused by hypersensitivity to the ubiquitous fungus Aspergillus fumigatus

- ABPA occurs most commonly in patients with asthma and CF
- ABPA is the cause of 1–10% of cases of bronchiectasis
- Most ABPA cases occur in the third and fourth decade without a sex predilection.





DIAGNOSIS

- Long standing uncontrolled asthma/ Cystic fibrosis
- Brownish sputum
- Peripheral eosinophilia > 500/ mm3
- Total IgE > 1000 IU/ ml
- Specific IgE for A. fumigatus > 0.35

HRCT thorax:

- > Central bronchiectasis
- ➤ High attenuation mucus
- ➤ Finger in glove/ TIB
- > Tram track
- Mosaic attenuation

MANAGEMENT



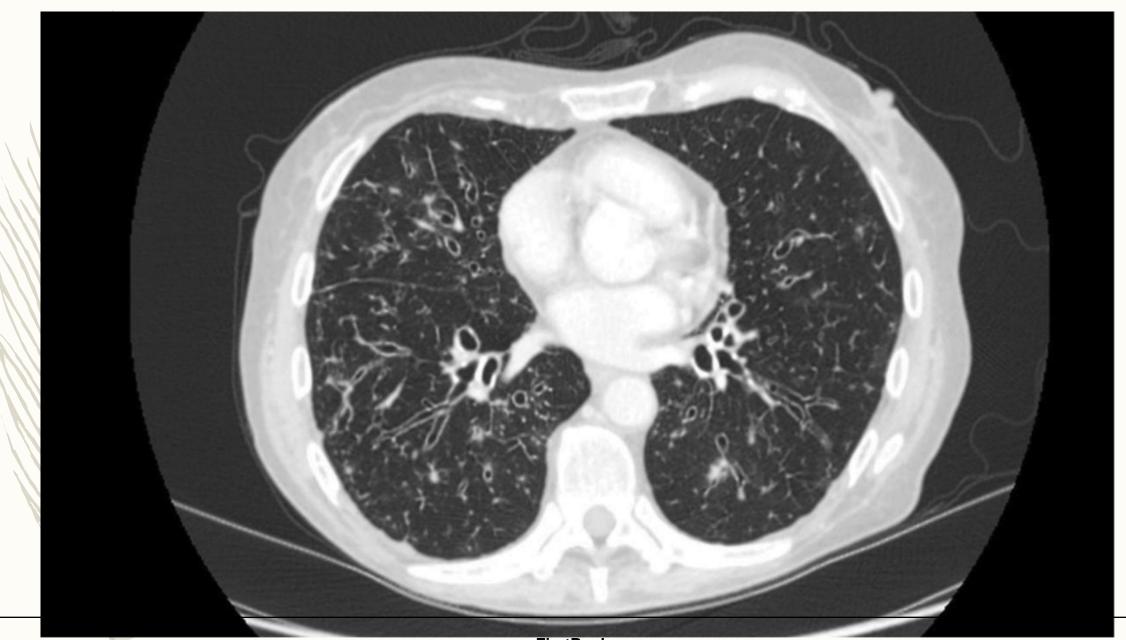
- 1. Corticosteroids
- 2. Antifungals
- 3. Airway clearing techniques
- 4. Chest physical therapy
- 5. Mucus thinners





CASE 3

- 77-year-old retired librarian.
- Cough for many years with new symptoms of fatigue, weight loss and fever.
- A chest CT scan was performed looking for a possible occult malignancy and bronchiectasis was found.



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DIAGNOSIS

- HRCT thorax: cylindrical bronchiectasis and tree-in-bud pattern in middle and lower lobes
- Sputum for M. Tuberculosis: negative
- MGIT culture: MAC growth at 4 weeks
- Repeat MGIT: Positive for MAC
- Tests for immunodeficiency and ABPA: Negative

MANAGEMENT



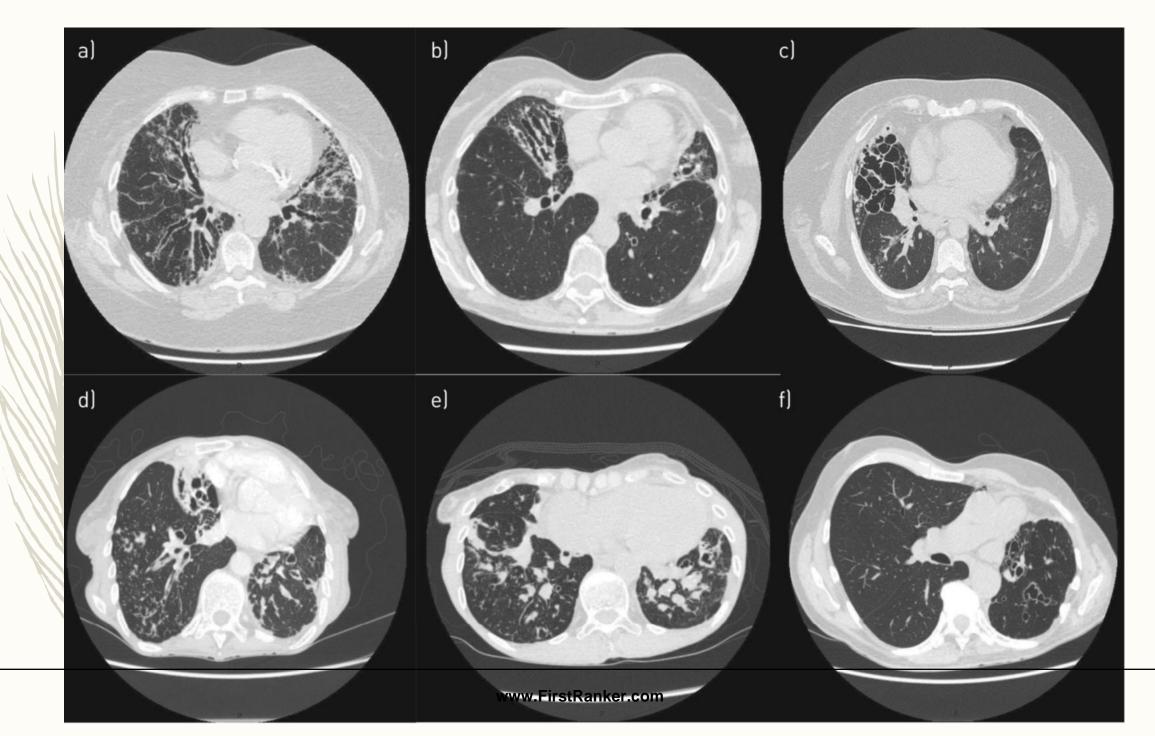
- 1. Management of NTM as per the organism and clinical picture
- 2. Airway clearing techniques
- 3. Chest physical therapy
- 4. Mucus thinners



CASE 4



- A 66-year-old woman with established idiopathic bronchiectasis has had three to four exacerbations per year for the past 3 years despite performing daily chest physiotherapy.
- Produces large volumes of sputum daily despite performing the active cycle of breathing technique.
- Testing for NTM, ABPA and other complications were negative, but sputum shows persistent infection with P. aeruginosa.





- One of the most common presentations of bronchiectasis
- Exacerbations are one of the most important manifestations of bronchiectasis and *P. aeruginosa* is the most frequent organism in severe bronchiectasis worldwide
- Cylindrical bronchiectasis is the most common morphological pattern identified on CT scans





- 1. Review current airway clearance regime.
- 2. Repeat sputum microbiology and repeat testing for NTM, ABPA and ensuring the all possible treatable causes and comorbidities have been identified.
- 3. First-line recommendation for *P. aeruginosa* with frequent exacerbations is an inhaled antibiotic.





ETIOLOGY

Category	Cause/notes	Clinical phenotype	Specific treatment
Post-infection	Viral, bacterial, fungal, mycobacteria (usually classified separately)	Past history of severe infection; classically unilobar bronchiectasis	No specific treatment
NTM	M. avium and M. abscessus most frequent	Middle-aged or elderly; females with low BMI; middle lobe and lingual nodular bronchiectasis; cavitation; tree-in- bud	Antibiotic treatment
Post-TB	M. tuberculosis	Upper lobe most frequently	No specific therapy
АВРА	Hypersensitivity to A. fumigatus	History of asthma (not universal); thick sputum; S. aureus in sputum; central bronchiectasis; fleeting infiltrates	Steroids±antifungals
COPD	Smoking, biomass exposure	Fixed airflow obstruction; smoking history; bilateral lower lobe; tubular bronchiectasis	No specific therapy
Asthma	Not universally accepted as a cause of bronchiectasis	Long history of asthma; frequent	Inhaled corticosteroids, biologics e.g. anti-IgE and anti-IL5
	www.FirstR	exacerbations; ankertcomilic airway inflammation	



Aspiration/inhalation	Foreign body aspiration, gastric contents aspiration, inhalation of corrosive substances	Lower lobe bronchiectasis	Speech and language therapy, fundoplication, removal of exacerbating drugs
Obstruction	Benign tumours, enlarged lymph nodes	Single lobe bronchiectasis	Removal of obstruction via bronchoscopy or thoracic surgery
Congenital defects of large airways	Marfan syndrome, Mounier- Kuhn syndrome (tracheobronchomegaly), Williams-Campbell syndrome (bronchial cartilage deficiency)	Specific features depending on the congenital defect	Dependant on the underlying disorder
AATD	Unopposed protease activity	Combined emphysema and bronchiectasis	Augmentation therapy is available in some countries
Yellow nail syndrome	Lymphatic obstruction	Dystrophic nails, pleural effusions, rhinosinusitis	Local treatment for nails e.g. vitamin-E, management of lymphoedema
Immunological defects	Primary: common variable immune deficiency, agammaglobulinemia, hyper-IgE syndrome; secondary: chemotherapy, immunosuppressant therapy, malignancy, HIV/AIDS	Varied clinical pattern depending on the underlying cause; patient may give a history of non-respiratory infections	lg replacement, prophylactic antibiotics, treatment of underlying disorder, removal of iatrogenic immunosuppression
Young's syndrome	Cause not known	Bronchiectasis, rhinosinusitis and reduced fertility	See ciliary disorders below

PCD	Genetic	Middle lobe and lower lobe bronchiectasis; rhinosinusitis; middle ear infections; situs inversus in some cases	Recognition and treatment of associated problems (including rhinosinusitis, middle ear disease, infertility, ectopic pregnancy), genetic counselling, intensive airway clearance
Systemic inflammatory disease	Rheumatoid arthritis, sarcoidosis, systemic lupus erythematosus, Sjögren syndrome	Varied clinical pattern, often rapidly progressive	No specific treatment
Inflammatory bowel disease	Ulcerative colitis, Crohn's syndrome, coeliac disease	Varied clinical pattern often high sputum volumes and steroid responsive	Inhaled and systematic corticosteroids, treatment of the underlying condition
Adult CF	CFTR mutations	Upper lobe bronchiectasis; P. aeruginosa or S. aureus in sputum; non-respiratory manifestations	Specialist multidisciplinary care in adult CF centres, recognition and treatment of non-respiratory manifestations, CFTR modulator/corrector therapy
Diffuse	Idiopathic inflammatory disease	Mostly patients of	Macrolide antibiotics
panbronchiolitis	www.Firstl	Far Eastern ethnic Ranker com	



INVESTIGATIONS FOR CAUSE

IN ALL

- COMORBIDITIES AND RELEVANT PAST HISTORY
- FULL BLOOD COUNT/ SERUM TOTAL IGE/ SKIN PRICK TEST TO A. FUMIGATUS
- SERUM Ig G/ IgA/ IgM
- BASELINE SPECIFIC ANTIBODY LEVELS AGAINST CAPSULAR POLYSACCHRIDES OF STREPTOCOCCUS PNEUMONIAE

CLINICALLY STABLE • SPUTUM CULTURE: ROUTINE AND MYCOBACTERIAL

CLINICALLY SUSPECT

- HIV
- TEST FOR CYSTIC FIBROSIS/ PCD/ GERD
- RA, ANTI CCP , ANCA, ANA
- ALPHA 1 AT
- BRONCHIAL ASPIRATION OR WASH

Hill A, Welham S, Sullivan A, Loebinger M. Updated BTS Adult Bronchiectasis Guideline 2018: a multidisciplinary approach to comprehensive care. Thorax. 2018;74(1):1-3.



STEPWISE MANAGEMENT



Step 1

- Treat underlying cause
- Airways clearance techniques +/pulmonary rehabilitation
- Annual influenza
 vaccination
- Prompt antibiotic treatment for exacerbations
- Self management plan

Step 2

If 3 or more exacerbations/yr despite Step 1*

 Physiotherapy reassessment and consider mucoactive treatment Step 3

If 3 or more exacerbations/yr despite Step 2*

- 1) If Pseudomonas aeruginosa,
 long trem inhaled antipseudomonal antibiotic or alternatively long term macrolide
- 2) If other Potentially Pathogenic
 Microorganisms, long term macrolides or alternatively long term oral or inhaled targeted antibiotic
- 3) If no pathogen, long term macrolides

Step 4

If 3 or more exacerbations/yr despite Step 3*

 Long term macrolide and long term inhaled antibiotic Step 2

If 5 or more exacerbations/yr despite Step 4*

Consider regular intravenous antibiotics every 2-3 months

*Consider this step if significant symptoms persist despite previous step, even if not meeting exacerbation criteria

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.





AIRWAY CLEARANCE



Physiotherapy management-stepwise airway clearance.

STEP

1

Offer active cycle of breathing techniques (ACBT) to individuals with bronchiectasis.

Consider gravity assisted positioning (where not contraindicated) to enhance the effectiveness of an airway clearance technique. If

contraindicated then modified postural drainage should be used.

Patients should be reviewed within 3 months.

This should include evaluation of patient reported effectiveness (ease of clearance/patient adherence).

The inclusion of gravity assisted positioning should be evaluated for its additional effectiveness.

STEP

2

If ACBT is not effective or the patients demonstrates poor adherence.

should be considered.

oscillating Positive Expiratory Pressures + Forced Expiration Technique

STEP

3

If airway clearance is not effective then nebulised Isotonic (0.9% saline) or Hypertonic Saline (3% saline and above)

should be evaluated for its effectiveness pre-airway clearance (especially in patients with viscous secretions or there is evidence of sputum plugging)

Individuals should be advised to complete Airway Clearance in the following order, if prescribed:

- Bronchodilator
- Mucoactive treatment
- Airway Clearance
- Nebulised antibiotic and/or inhaled steroids (if applicable)

ACBT: Active cycle of breathing techniques

THORAX

ACBT

Breathing control 20/30 seconds

Huffing followed by cough if needed

3-4 deep breaths

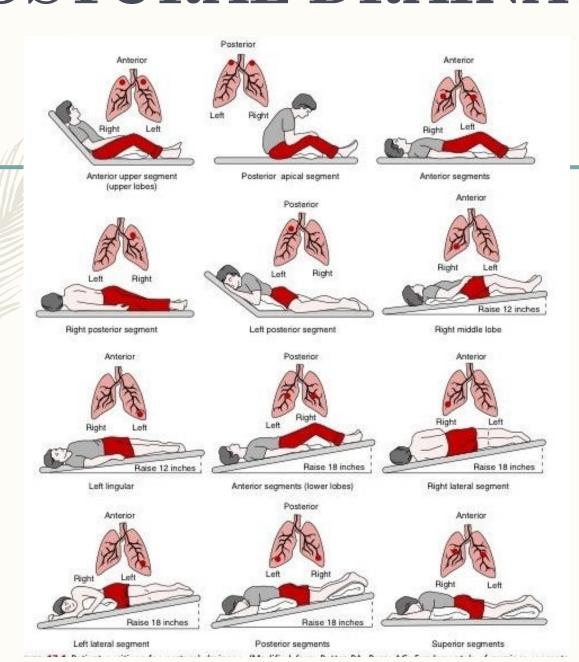
Breathing control

Breathing control

3 - 4 deep breaths



POSTURAL DRAINAGE



Airway clearance - exacerbations.

STEP

STEP

STEP

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

Commence the use of mPD or PD if tolerated.

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

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)
- Manual Techniques
- Positive pressure devices including Intermittent Positive Pressure Breathing (IPPB) or Non Invasive Ventilation

(NIV) to be used during Airway Clearance





ANTIBIOTIC TREAMENT FOR EXACERBATION

Organism	Recommended first line treatment	Length of treatment	Recommended second line treatment	Length of treatmen
Streptococcus pneumoniae	Amoxicillin 500 mg Three times a day	14 days	Doxycycline 100 mg BD	14 days
Haemophilus influenzae- beta lactamase negative	Amoxicillin 500 mg Three times a day Or Amoxicillin 1G Three times a day Or Amoxicillin 3G BD	14 days	Doxycycline 100 mg BD Or Ciprofloxacin 500 mg or 750 mg BD Or Ceftriaxone 2G OD (IV)	14 days
Haemophilus influenzae- beta lactamase positive	Amoxicillin with clavulanic acid 625 one tablet Three times a day	14 days	Doxycycline 100 mg bd Or Ciprofloxacin 500 mg or 750 mg BD Or Ceftriaxone 2G OD (IV)	14 days
Moraxella catarrhalis	Amoxicillin with clavulanic acid 625 one tablet Three times a day	14 days	Clarithromycin 500 mg BD Or Doxycycline 100 mg BD Or Ciprofloxacin 500 mg or 750 mg BD	14 days
Staphylococcus aureus (MSSA)	Flucloxacillin 500 mg Four times a day	14 days	Clarithromycin 500 mg BD Or Doxycycline 100 mg BD	14 days

a day



	Doxycycline 100 mg BD			
	Rifampicin (<50 Kg)			
Staphylococcus	450 mg OD		Third line	
aureus (MRSA)	Rifampicin (>50 Kg)	14 days	Linezolid	14 days
Oral preparations	600 mg OD		600 mg BD	
	Trimethoprim			
	200 mg BD			
	200 1118 22			
Staphylococcus	Vancomycin 1 gm BD* (monitor			
aureus (MRSA)	serum levels and adjust dose		Linezolid	
Intravenous	accordingly) or Teicoplanin 400 mg	14 days	600 mg BD	14 days
preparations	OD			
Coliforms for			Intravenous	
example,	Oral Ciprofloxacin	14 days	Ceftriaxone	14 days
Klebsiella,	500 mg or 750 mg BD	1 Tudys	2G OD	Trudys
enterobacter			2005	
			Monotherapy:	
			Intravenous	
			Ceftazidime	
			2G TDS	
			or	
			Piperacillin with tazobactam	
			4.5G TDS	
			or	
			Aztreonam	
			2G TDS	
	Oral Ciprofloxacin		OF	
Pseudomonas	500 mg bd		Meropenem	2.2
aeruginosa	(750 mg bd in more severe	14 days	2G TDS	14 days
Same of the Control o	infections)		Combination therapy	
			The above can be combined with gentamicin or	
			tobramycin or	
			Colistin 2MU TDS (under 60 kg, 50 000-75 000 Units/kg	
			daily in 3 divided doses)	
			Patients can have an in vivo response despite in vitro	
			resistance. Caution with aminoglycosides as highlighted	
			below but also if previous adverse events, particularly	
			previous	
			ototoxicity/acute kidney injury due to aminoglycosides	

WHAT IS THE ROLE OF SURGERY IN MANAGING BRONCHIECTASIS?

RECOMMENDATIONS

- Consider lung resection in patients with localized disease whose symptoms are not controlled by medical treatment optimized by a bronchiectasis specialist. (D)
- Offer multidisciplinary assessment, including a bronchiectasis physician, a thoracic surgeon and an experienced anesthetist, of suitability for surgery and pre-operative assessment of cardiopulmonary reserve post resection. (D)



LUNG TRANSPLANTATION FOR BRONCHIECTASIS

Recommendations

- 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)

LUNG ABSCESS

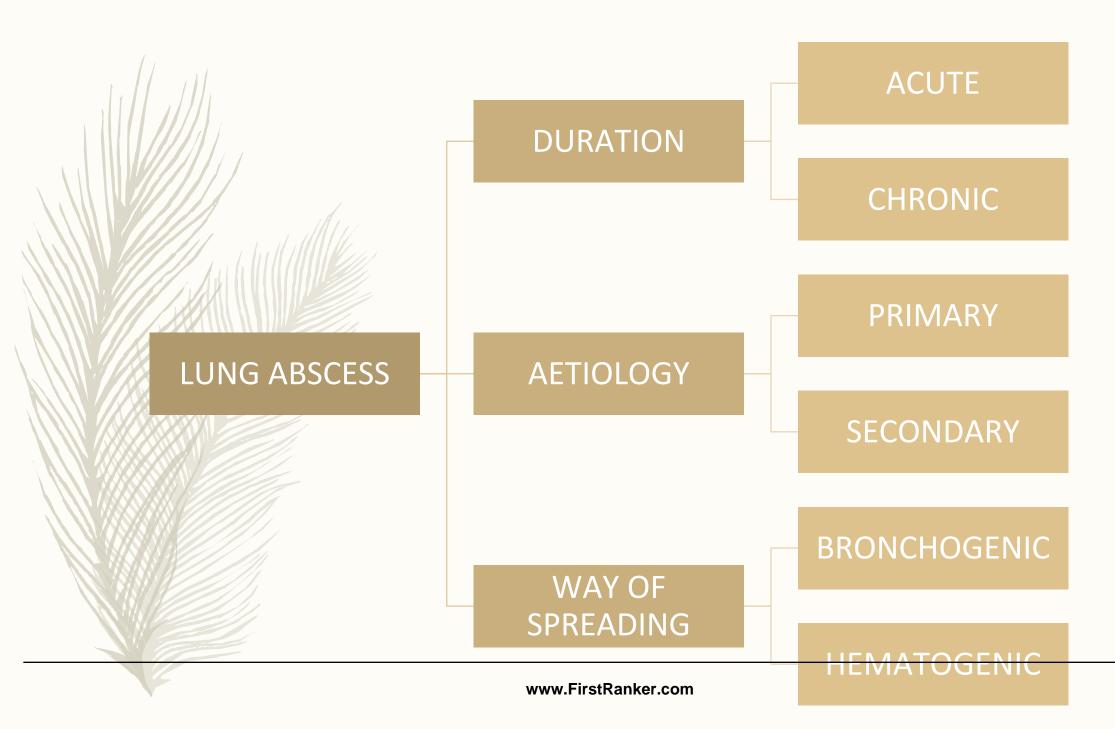




DEFINITION

Localized area of lung suppuration, leading to necrosis of the lung parenchyma with or without cavity formation.

Type of liquefactive necrosis of the lung tissue and formation of cavities (more than 2 cm) containing necrotic debris or fluid caused by microbial infection.







CLASSIFICATION (CONTD.)

• * ACCORDING TO THE DURATION:

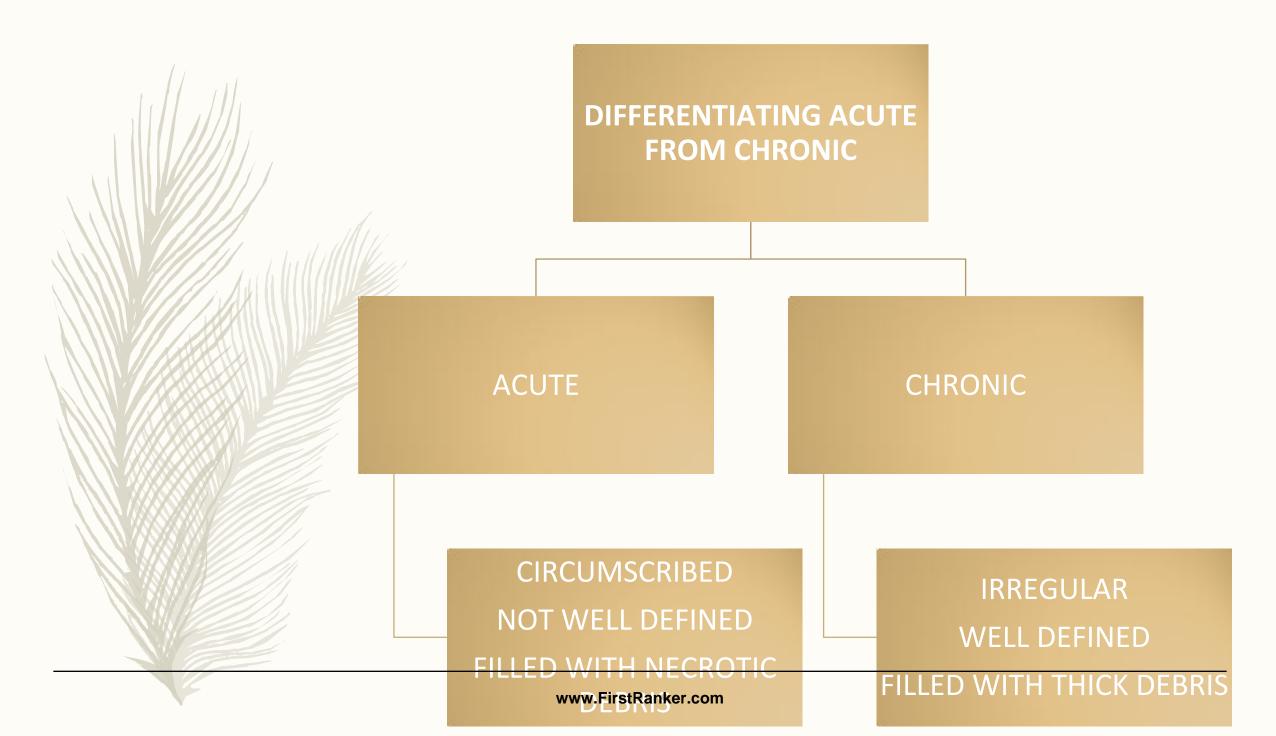
- Acute (less than 6 weeks);
- Chronic (more than 6 weeks)

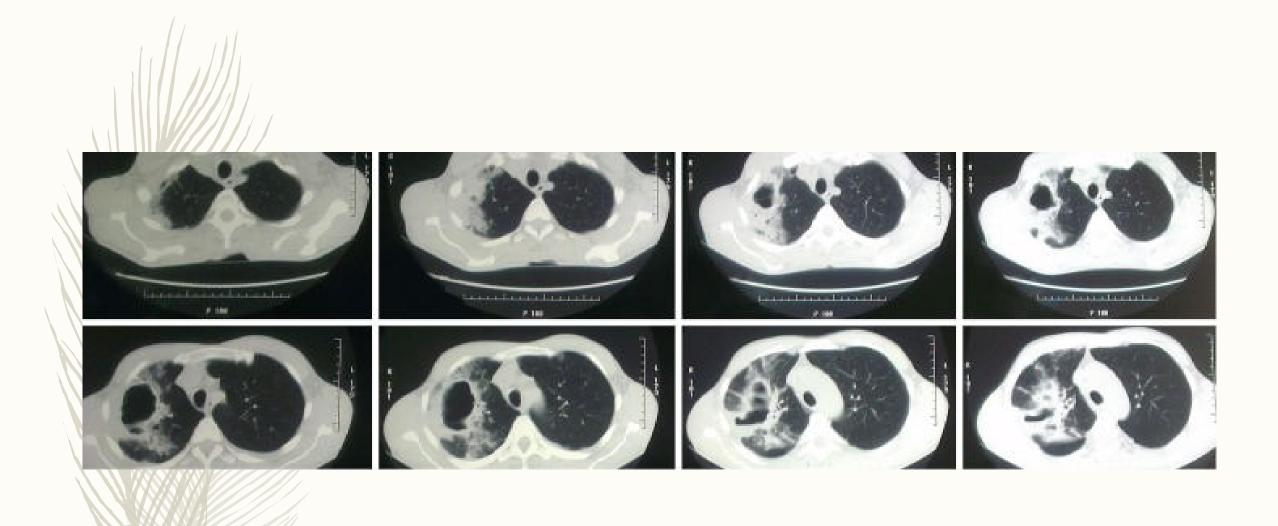
• * BY ETIOLOGY:

- Primary (aspiration of oropharyngeal secretions, necrotizing pneumonia, immunodeficiency);
- Secondary (bronchial obstructions, haematogenic dissemination, direct spreading from mediastinal infection, from sub phrenic space, coexisting lung diseases)

• * WAY OF SPREADING:

- Bronchogenic (aspiration of oropharyngeal secretions, bronchial obstruction by tumour, foreign body, enlarged lymph nodes, congenital malformation);
- Haematogenic (abdominal sepsis, infective endocarditis, septic thromboembolisms)





DIFFERENTIAL DIAGNOSIS

- Excavating bronchial carcinoma (squamo-cellular or microcellular)
- Excavating tuberculosis
- Localized pleural empyema
- Infected emphysematous bullae
- Cavitary pneumoconiosis
- Hiatus hernia
- Pulmonary hematoma
- Hydatid cyst of lung
- Cavitary infarcts of lung
- Wegener's granulomatosis...



DIAGNOSIS

- Diagnostic bronchoscopy is a part of diagnostic protocol for taking the material for microbiological examination and to confirm intrabronchial cause of abscess-tumor or foreign body.
- Sputum examination is useful for identification of microbiological agents or confirmation of bronchial carcinoma

MANAGEMENT

STANDARD CONSERVATIVE THERAPY: MEDICAL MANAGEMENT

- It is recommended to treat lung abscess with broad spectrum antibiotics, due to poly microbial flora, such as Clindamycin (600 mg IV on 8 h) and then 300 mg PO on 8 h or combination ampicilin/sulbactam (1.5-3 gr IV on 6 h).
- Alternative therapy is piperacilin/tazobactam 3.375 gr IV on 6 h or Meropenem 1 gr IV on 8 h.
- For MRSA it is recommended to use linezolid 600 mg IV on 12 h or vancomycin 15 mg/kg BM on 12 h.





MANAGEMENT

SURGICAL

- Endoscopic drainage of lung abscesses is described as an alternative to chest tube drainage and is performed during the bronchoscopy with usage of laser.
- Per cutaneous trans thoracic tube drainage
- Surgical resection of lung abscess is the therapy of choice for about 10% of patients.
- Lobectomy is the resection of choice for large or central position of abscess. Atypical resection or segmentectomy are satisfactory procedures, if it is possible to remove complete abscess and if necessary surrounding lung tissue with necrotizing pneumonia

THANK YOU





BRONCHIECTASIS

DEFINITION

- Bronchiectasis (broncos, airways; ectasia, dilatation) is a morphologic term used to describe abnormal, irreversibly dilated and thick walled bronchi.
- This is an anatomic definition that evolved from Laennec's original description in 1819 of ectatic bronchi in pathologic specimens.



PREVALENCE

US¹

Prevalence increased every year from 2000 to 2007 by an annual percentage change of 8.74%.

- Increased with age (peak= 80-84 years)
- Higher in women

UK²

- Prevalence in women 566/lakh; men= 486/lakh
- Women and age more than 60 years associated with higher rate of hospitalization

INDIA

- There is no good data on bronchiectasis in India
- EMBARC INDIA REGISTRY (European Multi Centre Bronchiectasis Audit and Research Collaboration)
- 1. McShane P, Naureckas E, Tino G, Strek M. Non–Cystic Fibrosis Bronchiectasis. American Journal of Respiratory and Critical Care Medicine. 2013;188(6):647-656.
- 2. Hill A, Welham S, Sullivan A, Loebinger M. Updated BTS Adult Bronchiectasis Guideline 2018: a multidisciplinary approach to comprehensive care. Thorax. 2018;74(1):1-3.

BURDEN

LONGER HOSPITAL STAY

FREQUENT OPD VISITS

INCREASED EXPENDITURE ON MEDICINES

MORTALITY RATE= 10-16%

RISK FACTORS FOR MORTALITY

LOW FEV1

MALE

INCREASED DYSPNOEA GRADE ADVANCED AGE

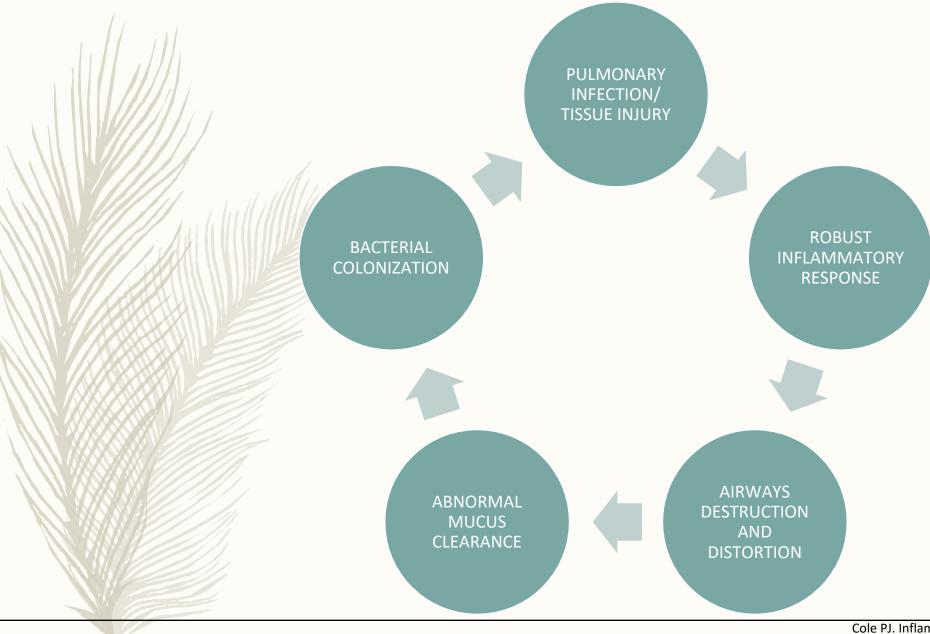
COPD

P. aeruginosa SPUTUM POSITIVITY

- Goeminne PC, Scheers H, Decraene A, Seys S, Dupont LJ. Risk factors for morbidity and death in non–cystic fibrosis bronchiectasis: a retrospective cross-sectional analysis of CT diagnosed bronchiectatic patients. Respir Res 2012;13:21.
- Weycker D, Edelsberg J, Oster G, Tino G. Prevalence and economic burden of bronchiectasis. Clin Pulm Med 2005;12:205–209 •

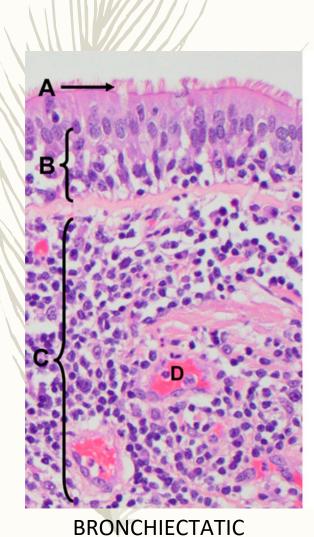


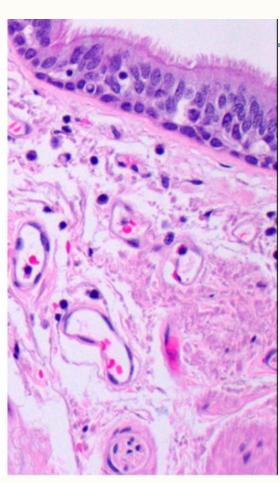












NORMAL

Hematoxylin and eosin stain of the bronchial wall

in a patient with bronchiectasis (left) versus a normal subject (right).

A: Pseudostratified columnar, ciliated epithelium

B: thickened epithelium with intraepithelial lymphocytes

C: submucosa with dense infiltrate of lymphocytes and plasma cells

D: blood vessel with reactive endothelial cells.

TYPES

REIDS CLASSIFICATION

CYLINDRICAL

VARICOSE

SACULAR/CYSTIC



TYPES OF BRONCHIECTASIS

Cylindrical bronchiectasis	Varicose bronchiectasis	Cystic bronchiectasis
Mild	Moderate	Sever
Tram track appearance	String of beads	Cluster of grapes
		SCIENCEPHOTOLIBRARY



ETIOLOGY



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Category	Cause/notes	Clinical phenotype	Specific treatment
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Aspiration/inhalation	Foreign body aspiration, gastric contents aspiration, inhalation of corrosive substances	Lower lobe bronchiectasis	Speech and language therapy, fundoplication, removal of exacerbating drugs
Obstruction	Benign tumours, enlarged lymph nodes	Single lobe bronchiectasis	Removal of obstruction via bronchoscopy or thoracic surgery
Congenital defects of large airways	Marfan syndrome, Mounier- Kuhn syndrome (tracheobronchomegaly), Williams-Campbell syndrome (bronchial cartilage deficiency)	Specific features depending on the congenital defect	Dependant on the underlying disorder
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Immunological defects	Primary: common variable immune deficiency, agammaglobulinemia, hyper-lgE syndrome; secondary: chemotherapy, immunosuppressant therapy, malignancy, HIV/AIDS	Varied clinical pattern depending on the underlying cause; patient may give a history of non-respiratory infections	lg replacement, prophylactic antibiotics, treatment of underlying disorder, removal of iatrogenic immunosuppression
Young's syndrome	Cause not known	Bronchiectasis,	See ciliary disorders below
	www.FirstR	rhinosinusitis and ankeucomertility	

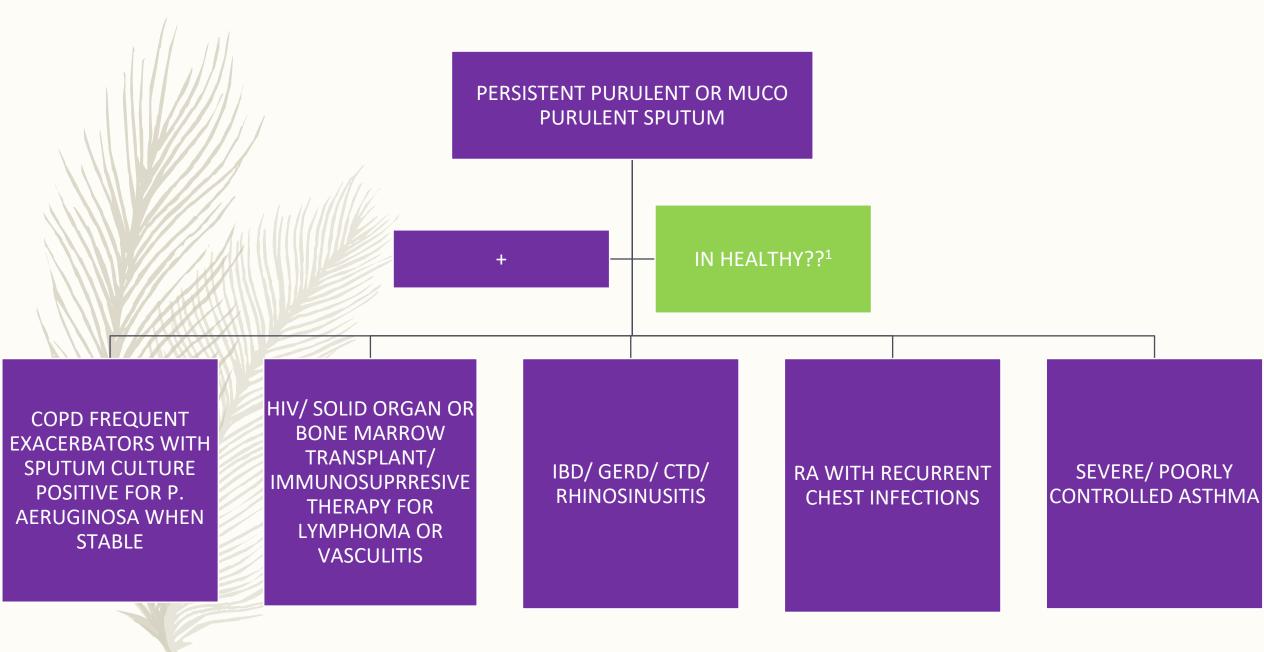


PCD	Genetic	Middle lobe and lower lobe bronchiectasis; rhinosinusitis; middle ear infections; situs inversus in some cases	Recognition and treatment of associated problems (including rhinosinusitis, middle ear disease, infertility, ectopic pregnancy), genetic counselling, intensive airway clearance
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Adult CF	CFTR mutations	Upper lobe bronchiectasis; P. aeruginosa or S. aureus in sputum; non-respiratory manifestations	Specialist multidisciplinary care in adult CF centres, recognition and treatment of non-respiratory manifestations, CFTR modulator/corrector therapy
Diffuse panbronchiolitis	Idiopathic inflammatory disease	Mostly patients of Far Eastern ethnic origin	Macrolide antibiotics



IN WHOM TO SUSPECT?

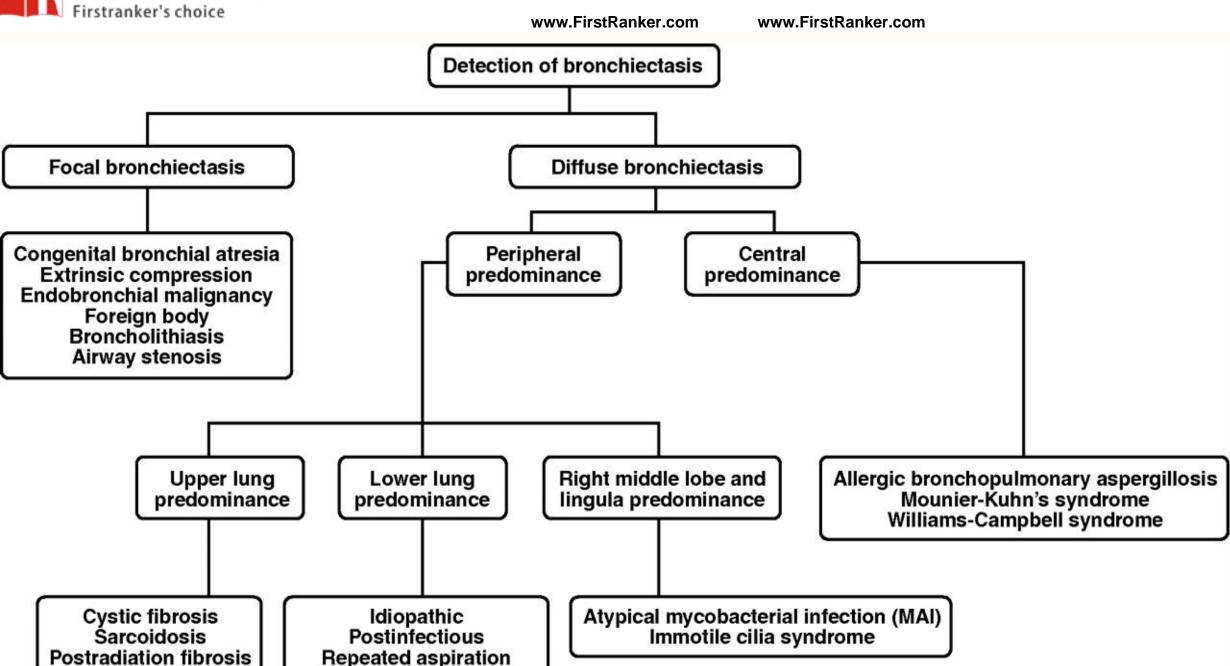




Hill A, Welham S, Sullivan A, Loebinger M. Updated BTS Adult Bronchiectasis Guideline 2018: a multidisciplinary approach to comprehensive care. Thorax. 2018;74(1):1-3.



ALGORITHM FOR EVALUATION OF BRONCHIECTASIS



INVESTIGATION: RADIOLOGY

Cantin L, Bankier A, Eisenberg R. Bronchiectasis. American Journal of Roentgenology.

2009;193(3):W158-W171.

- BASELINE CHEST RADIOGRAPH

- THIN SECTION CT [HRCT THORAX]

CT FEATURES OF BRONCHIECTASIS

***BRONCHIAL DILATATION SUGGESTED BY**

- 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.

❖ INDIRECT SIGNS

- BRONCHIAL WALL THICKENING
- MUCUS IMPACTION
- MOSAIC PERFUSION / AIR TRAPPING ON EXPIRATORY CT

Fibrotic lung disease Posttransplant rejection

Hypogammaglobulinemia



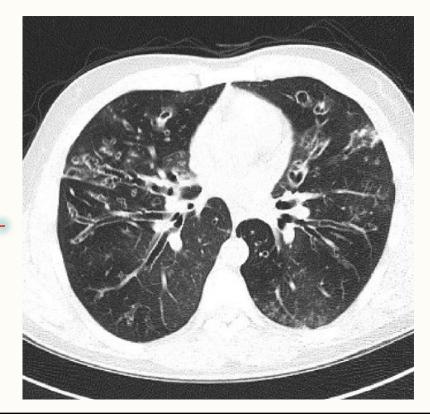
CHEST RADIOGRAPH



CYLINDRICAL BRONCHIECTASIS

WITHIN I CM OF PLEURA

TRAM TRACK SIGN

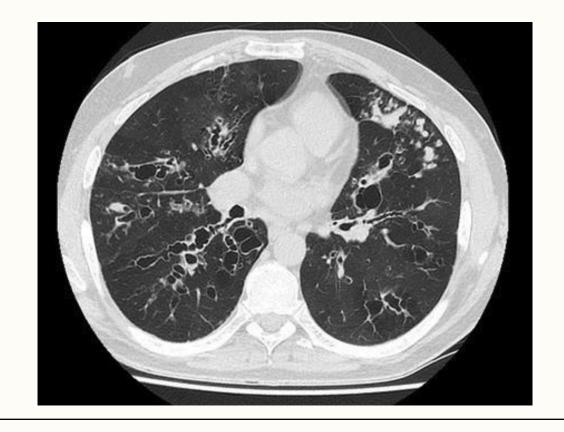




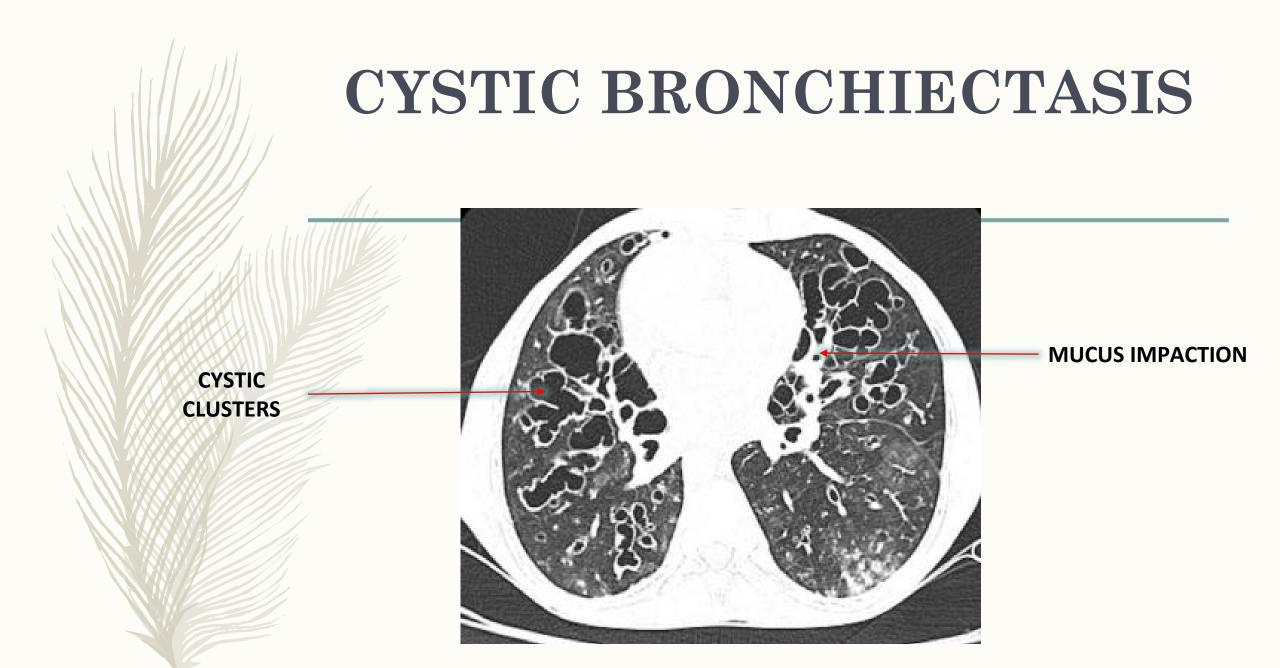
VARICOSE



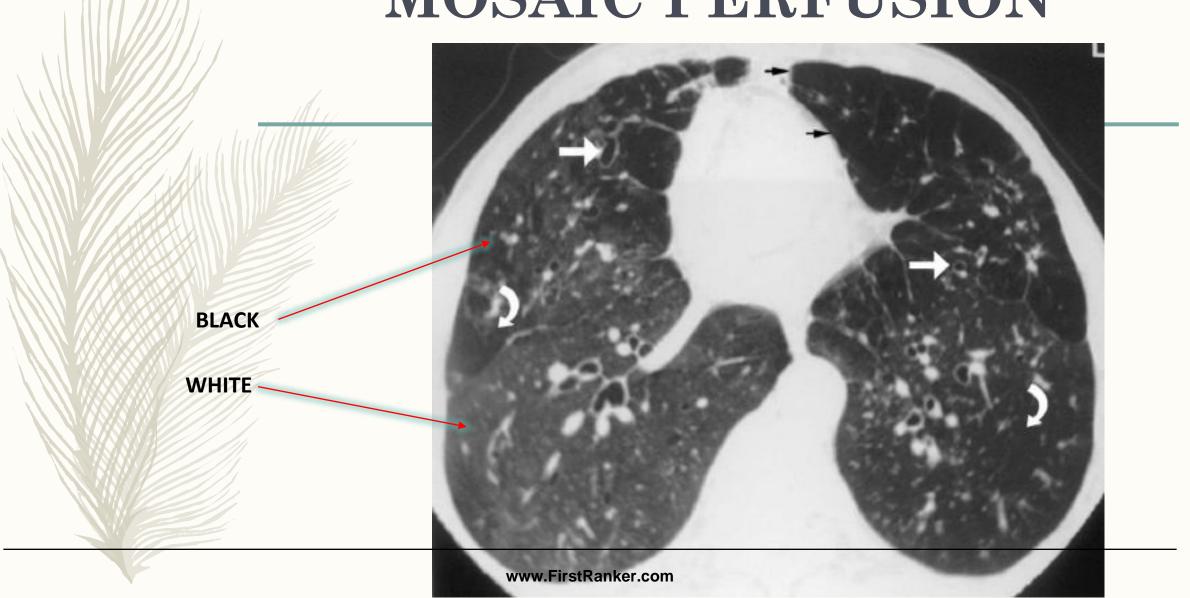
VARICOSE BRONCHIECTASIS







MOSAIC PERFUSION





INVESTIGATIONS FOR CAUSE

IN ALL

- COMORBIDITIES AND RELEVANT PAST HISTORY
- FULL BLOOD COUNT/ SERUM TOTAL IGE/ SKIN PRICK TEST TO A. FUMIGATUS
- SERUM Ig G/ IgA/ IgM
- BASELINE SPECIFIC ANTIBODY LEVELS AGAINST CAPSULAR POLYSACCHRIDES OF STREPTOCOCCUS PNEUMONIAE

CLINICALLY STABLE • SPUTUM CULTURE: ROUTINE AND MYCOBACTERIAL

CLINICALLY SUSPECT

- HIV
- TEST FOR CYSTIC FIBROSIS/ PCD/ GERD
- RA, ANTI CCP , ANCA, ANA
- ALPHA 1 AT
- BRONCHIAL ASPIRATION OR WASH

Hill A, Welham S, Sullivan A, Loebinger M. Updated BTS Adult Bronchiectasis Guideline 2018: a multidisciplinary approach to comprehensive care. Thorax. 2018;74(1):1-3.



STEPWISE MANAGEMENT



Step 1

- Treat underlying cause
- Airways clearance techniques +/pulmonary rehabilitation
- Annual influenza
 vaccination
- Prompt antibiotic treatment for exacerbations
- Self management plan

Step 2

If 3 or more exacerbations/yr despite Step 1*

 Physiotherapy reassessment and consider mucoactive treatment Step 3

If 3 or more exacerbations/yr despite Step 2*

- 1) If Pseudomonas aeruginosa,
 long trem inhaled antipseudomonal antibiotic or alternatively long term macrolide
- 2) If other Potentially Pathogenic
 Microorganisms, long term macrolides or alternatively long term oral or inhaled targeted antibiotic
- 3) If no pathogen, long term macrolides

Step 4

If 3 or more exacerbations/yr despite Step 3*

 Long term macrolide and long term inhaled antibiotic Step 2

If 5 or more exacerbations/yr despite Step 4*

Consider regular intravenous antibiotics every 2-3 months

*Consider this step if significant symptoms persist despite previous step, even if not meeting exacerbation criteria

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.





AIRWAY CLEARANCE



Physiotherapy management-stepwise airway clearance.

STEP

1

Offer active cycle of breathing techniques (ACBT) to individuals with bronchiectasis.

Consider gravity assisted positioning (where not contraindicated) to enhance the effectiveness of an airway clearance technique. If

contraindicated then modified postural drainage should be used.

Patients should be reviewed within 3 months.

This should include evaluation of patient reported effectiveness (ease of clearance/patient adherence).

The inclusion of gravity assisted positioning should be evaluated for its additional effectiveness.

STEP

2

If ACBT is not effective or the patients demonstrates poor adherence.

should be considered.

oscillating Positive Expiratory Pressures + Forced Expiration Technique

STEP

3

If airway clearance is not effective then nebulised Isotonic (0.9% saline) or Hypertonic Saline (3% saline and above)

should be evaluated for its effectiveness pre-airway clearance (especially in patients with viscous secretions or there is evidence of sputum plugging)

Individuals should be advised to complete Airway Clearance in the following order, if prescribed:

- Bronchodilator
- Mucoactive treatment
- Airway Clearance
- Nebulised antibiotic and/or inhaled steroids (if applicable)

ACBT: Active cycle of breathing techniques

THORAX

ACBT

Breathing control 20/30 seconds

Huffing followed by cough if needed

3-4 deep breaths

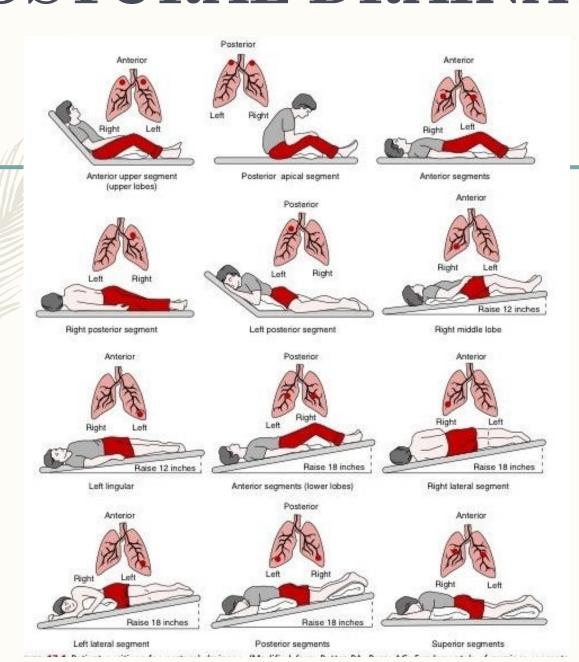
Breathing control

Breathing control

3 - 4 deep breaths



POSTURAL DRAINAGE



Airway clearance - exacerbations.

STEP

STEP

STEP

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

Commence the use of mPD or PD if tolerated.

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

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)
- Manual Techniques
- Positive pressure devices including Intermittent Positive Pressure Breathing (IPPB) or Non Invasive Ventilation

(NIV) to be used during Airway Clearance





ANTIBIOTIC TREAMENT FOR EXACERBATION

Organism	Recommended first line treatment	Length of treatment	Recommended second line treatment	Length of treatment
Streptococcus pneumoniae	Amoxicillin 500 mg Three times a day	14 days	Doxycycline 100 mg BD	14 days
Haemophilus influenzae- beta lactamase negative	Amoxicillin 500 mg Three times a day Or Amoxicillin 1G Three times a day Or Amoxicillin 3G BD	14 days	Doxycycline 100 mg BD Or Ciprofloxacin 500 mg or 750 mg BD Or Ceftriaxone 2G OD (IV)	14 days
Haemophilus influenzae- beta lactamase positive	Amoxicillin with clavulanic acid 625 one tablet Three times a day	14 days	Doxycycline 100 mg bd Or Ciprofloxacin 500 mg or 750 mg BD Or Ceftriaxone 2G OD (IV)	14 days
Moraxella catarrhalis	Amoxicillin with clavulanic acid 625 one tablet Three times a day	14 days	Clarithromycin 500 mg BD Or Doxycycline 100 mg BD Or Ciprofloxacin 500 mg or 750 mg BD	14 days
Staphylococcus aureus (MSSA)	Flucloxacillin 500 mg Four times a day	14 days	Clarithromycin 500 mg BD Or Doxycycline 100 mg BD	14 days

a day



		***************************************	outamonoon www.noutamonoon	
	Doxycycline 100 mg BD			
	Rifampicin (<50 Kg)			
Staphylococcus	450 mg OD		Third line	
aureus (MRSA)	Rifampicin (>50 Kg)	14 days	Linezolid	14 days
Oral preparations	600 mg OD		600 mg BD	
	Trimethoprim			
	200 mg BD			
	200 111g 55			
Staphylococcus	Vancomycin 1 gm BD* (monitor			
aureus (MRSA)	serum levels and adjust dose		Linezolid	
Intravenous	accordingly) or Teicoplanin 400 mg	14 days	600 mg BD	14 days
preparations	OD			
Coliforms for			Intravenous	
example,	Oral Ciprofloxacin	14 days	Ceftriaxone	14 days
Klebsiella,	500 mg or 750 mg BD	1 1 days	2G OD	11 days
enterobacter			2500	
			Monotherapy:	
			Intravenous	
			Ceftazidime	
			2G TDS	
			or or	
			Piperacillin with tazobactam	
			4.5G TDS	
			or	
			Aztreonam	
			2G TDS	
	Oral Ciprofloxacin		or	
Pseudomonas	500 mg bd		Meropenem	
aeruginosa	(750 mg bd in more severe	14 days	2G TDS	14 days
	infections)			
			Combination therapy	
			The above can be combined with gentamicin or	
			tobramycin or	
			Colistin 2MU TDS (under 60 kg, 50 000-75 000 Units/kg	
			daily in 3 divided doses)	
			Patients can have an in vivo response despite in vitro	
			resistance. Caution with aminoglycosides as highlighted	
			below but also if previous adverse events, particularly	
			previous	

WHAT IS THE ROLE OF SURGERY IN MANAGING BRONCHIECTASIS?

RECOMMENDATIONS

- Consider lung resection in patients with localized disease whose symptoms are not controlled by medical treatment optimized by a bronchiectasis specialist. (D)
- Offer multidisciplinary assessment, including a bronchiectasis physician, a thoracic surgeon and an experienced anesthetist, of suitability for surgery and pre-operative assessment of cardiopulmonary reserve post resection. (D)



LUNG TRANSPLANTATION FOR BRONCHIECTASIS

Recommendations

- 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)

LUNG ABSCESS

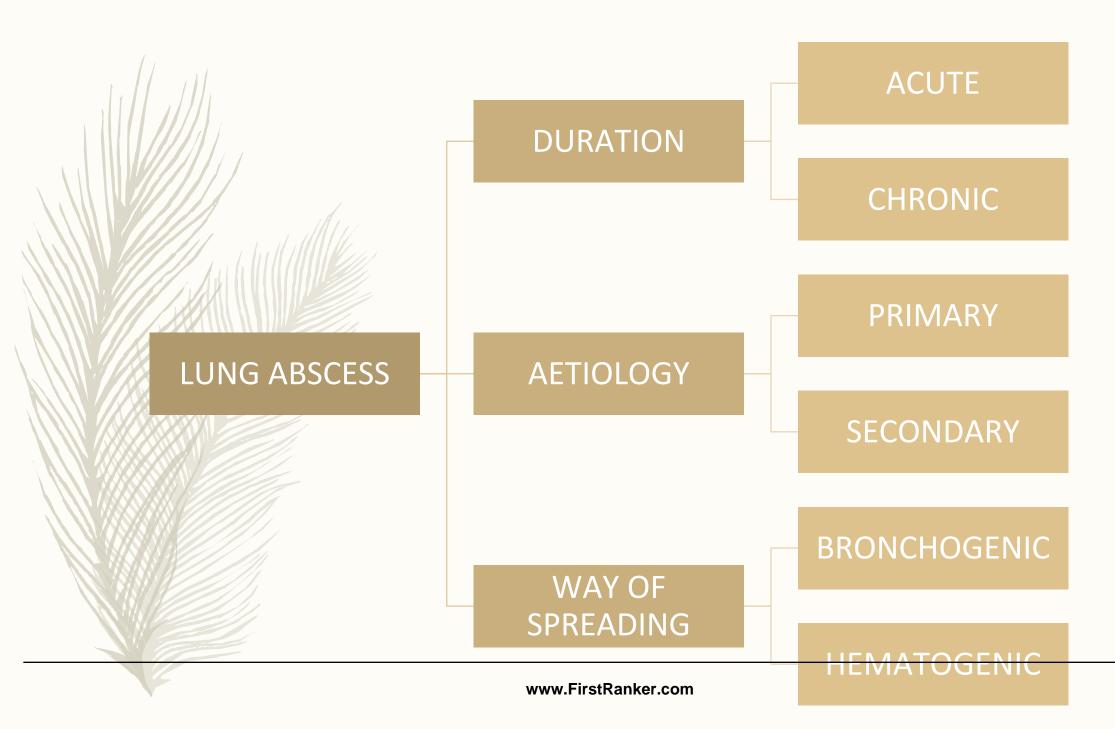




DEFINITION

Localized area of lung suppuration, leading to necrosis of the lung parenchyma with or without cavity formation.

Type of liquefactive necrosis of the lung tissue and formation of cavities (more than 2 cm) containing necrotic debris or fluid caused by microbial infection.







CLASSIFICATION (CONTD.)

• * ACCORDING TO THE DURATION:

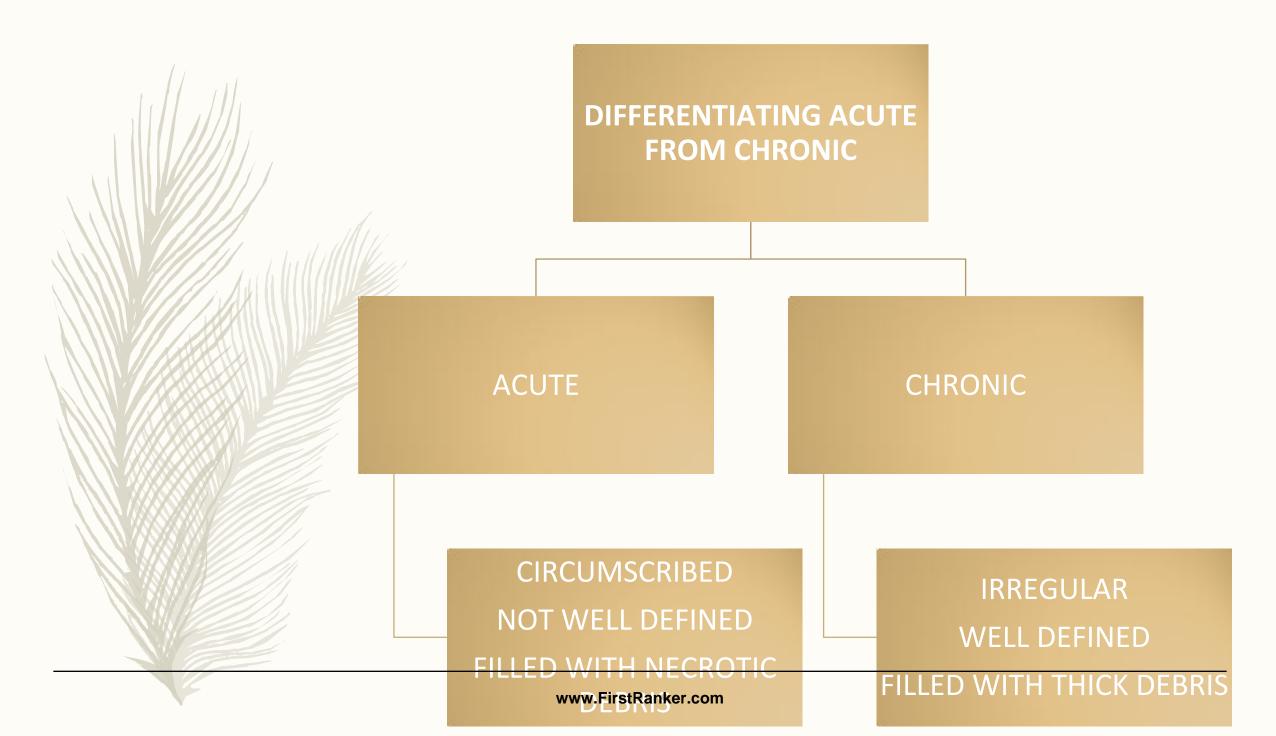
- Acute (less than 6 weeks);
- Chronic (more than 6 weeks)

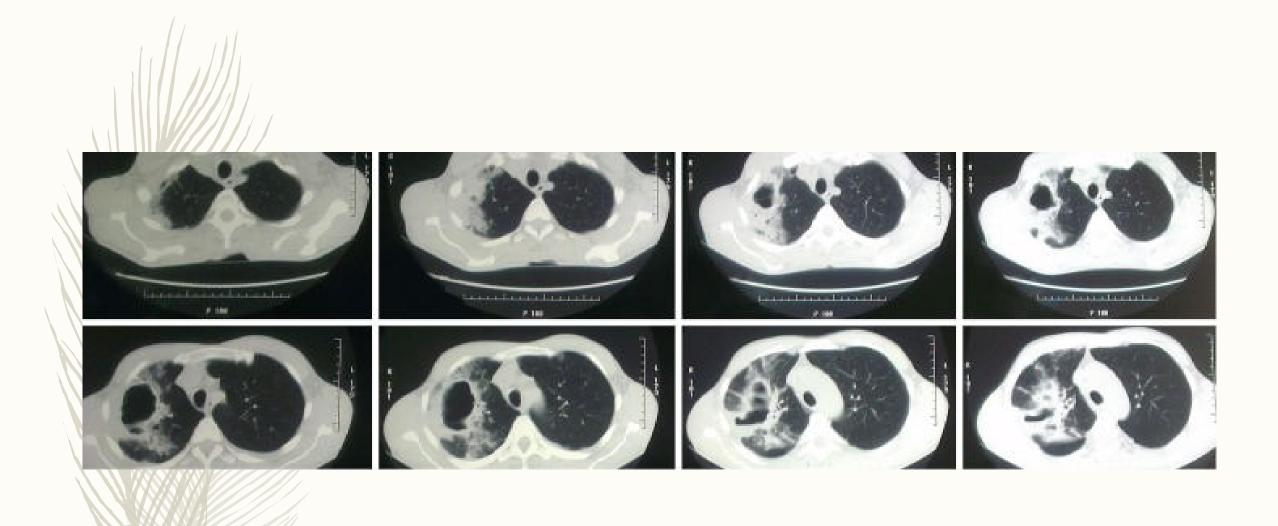
• * BY ETIOLOGY:

- Primary (aspiration of oropharyngeal secretions, necrotizing pneumonia, immunodeficiency);
- Secondary (bronchial obstructions, haematogenic dissemination, direct spreading from mediastinal infection, from sub phrenic space, coexisting lung diseases)

• * WAY OF SPREADING:

- Bronchogenic (aspiration of oropharyngeal secretions, bronchial obstruction by tumour, foreign body, enlarged lymph nodes, congenital malformation);
- Haematogenic (abdominal sepsis, infective endocarditis, septic thromboembolisms)





DIFFERENTIAL DIAGNOSIS

- Excavating bronchial carcinoma (squamo-cellular or microcellular)
- Excavating tuberculosis
- Localized pleural empyema
- Infected emphysematous bullae
- Cavitary pneumoconiosis
- Hiatus hernia
- Pulmonary hematoma
- Hydatid cyst of lung
- Cavitary infarcts of lung
- Wegener's granulomatosis...



DIAGNOSIS

- Diagnostic bronchoscopy is a part of diagnostic protocol for taking the material for microbiological examination and to confirm intrabronchial cause of abscess-tumor or foreign body.
- Sputum examination is useful for identification of microbiological agents or confirmation of bronchial carcinoma

MANAGEMENT

STANDARD CONSERVATIVE THERAPY: MEDICAL MANAGEMENT

- It is recommended to treat lung abscess with broad spectrum antibiotics, due to poly microbial flora, such as Clindamycin (600 mg IV on 8 h) and then 300 mg PO on 8 h or combination ampicilin/sulbactam (1.5-3 gr IV on 6 h).
- Alternative therapy is piperacilin/tazobactam 3.375 gr IV on 6 h or Meropenem 1 gr IV on 8 h.
- For MRSA it is recommended to use linezolid 600 mg IV on 12 h or vancomycin 15 mg/kg BM on 12 h.





MANAGEMENT

SURGICAL

- Endoscopic drainage of lung abscesses is described as an alternative to chest tube drainage and is performed during the bronchoscopy with usage of laser.
- Per cutaneous trans thoracic tube drainage
- Surgical resection of lung abscess is the therapy of choice for about 10% of patients.
- Lobectomy is the resection of choice for large or central position of abscess.
 Atypical resection or segmentectomy are satisfactory procedures, if it is possible to remove complete abscess and if necessary surrounding lung tissue with necrotizing pneumonia

THANK YOU



CASE 1



- A 42-year-old man, gardener
- Long history of respiratory problems starting in early childhood.
- Previously diagnosed as asthma.
- Frequent absence from work due to "recurrent chest infections".
- Unaware of any neonatal issues but believes that he was born at home without complications and is unsure of any previous tests he has had as he is now estranged from his parents.
- Has a cousin with a "lung disease".
- Married but has "no kids"





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INVESTIGATIONS



- Sweat chloride = 73 meq/liter
- Cystic fibrosis genetics: genotype was F508del/R117H
- CYSTIC FIBROSIS: Multisystem disorder caused by mutations in the gene that encodes the CF transmembrane conductance regulator (CFTR) protein, a chloride channel expressed in epithelial cells.
- More than 2000 CFTR mutations have been identified to date, but only the functional importance of a small number is known to cause the disease

HRCT THORAX

- An upper lobe predominant distribution of cylindrical, cystic and varicose bronchiectasis associated with airway wall thickening, mucus plugging and parenchymal opacities on a HRCT scan should raise the suspicion of CF disease.
- The presence of nasal polyposis and/or chronic rhinosinusitis, recurrent pancreatitis, malabsorption, diabetes, osteoporosis and male infertility are other typical features of CF



DIAGNOSIS

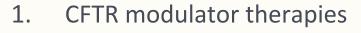


USA allows diagnosis if:



- 2. Clinical features of the disease with concentration of chloride >60 mmol·L⁻¹ at the sweat test or a concentration in the intermediate range (30–59 mmol·L⁻¹) but two disease-causing CFTR mutations.
- 3. CFTR genotype is undefined: CFTR physiologic tests, such as nasal potential difference and intestinal current measurement, should be performed.

MANAGEMENT



- 2. Airway clearing techniques
- 3. Chest physical therapy
- 4. Humidification with sterile water or normal saline to facilitate airway clearance
- 5. Antibiotics
- 6. Mucus thinners
- 7. Lung transplantation



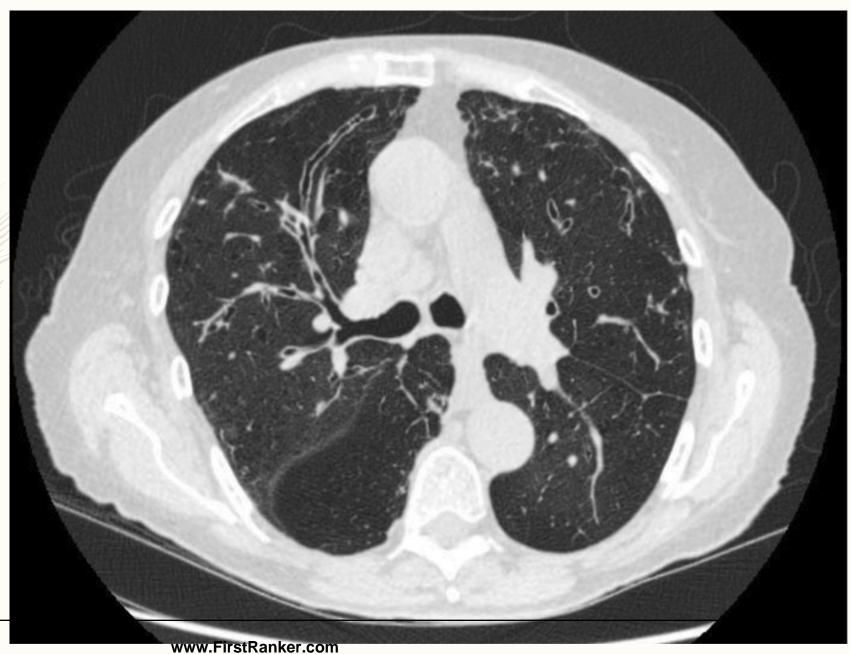


CASE 2



- 45-year-old farmer with asthma since childhood.
- Complaints: Decline in his exercise tolerance and an increase in cough which has become productive of purulent sputum with occasional thick/solid components.
- Respiratory exacerbations not responding well to standard steroid and antibiotic treatment.
- He was noted to have variable pulmonary infiltrates on chest radiographs during these episodes







INVSETIGATIONS



- Total IgE > 1000 IU/ ml
- Aspergillus specific IgE > 0.35

ABPA: ABPA is an inflammatory disease caused by hypersensitivity to the ubiquitous fungus Aspergillus fumigatus

- ABPA occurs most commonly in patients with asthma and CF
- ABPA is the cause of 1–10% of cases of bronchiectasis
- Most ABPA cases occur in the third and fourth decade without a sex predilection.

DIAGNOSIS

- Long standing uncontrolled asthma/ Cystic fibrosis
- Brownish sputum
- Peripheral eosinophilia > 500/ mm3
- Total IgE > 1000 IU/ ml
- Specific IgE for A. fumigatus > 0.35

HRCT thorax:

- > Central bronchiectasis
- ➤ High attenuation mucus
- > Finger in glove/ TIB
- > Tram track
- Mosaic werkthe First Banker.com





MANAGEMENT



- 1. Corticosteroids
- 2. Antifungals
- 3. Airway clearing techniques
- 4. Chest physical therapy
- 5. Mucus thinners

CASE 3



- Cough for many years with new symptoms of fatigue, weight loss and fever.
- A chest CT scan was performed looking for a possible occult malignancy and bronchiectasis was found.





DIAGNOSIS



- HRCT thorax: cylindrical bronchiectasis and tree-in-bud pattern in middle and lower lobes
- Sputum for M. Tuberculosis: negative
- MGIT culture: MAC growth at 4 weeks
- Repeat MGIT: Positive for MAC
- Tests for immunodeficiency and ABPA: Negative



MANAGEMENT



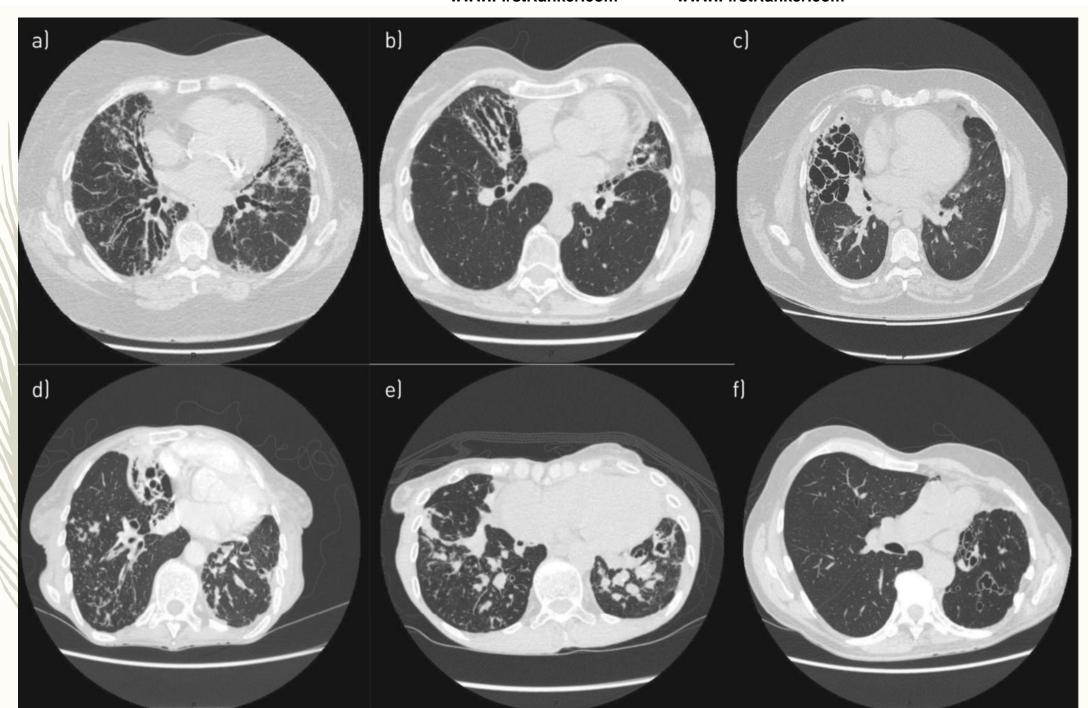
- 1. Management of NTM as per the organism and clinical picture
- 2. Airway clearing techniques
- 3. Chest physical therapy
- 4. Mucus thinners

CASE 4



- A 66-year-old woman with established idiopathic bronchiectasis has had three to four exacerbations per year for the past 3 years despite performing daily chest physiotherapy.
- Produces large volumes of sputum daily despite performing the active cycle of breathing technique.
- Testing for NTM, ABPA and other complications were negative, but sputum shows persistent infection with P. aeruginosa.





- One of the most common presentations of bronchiectasis
- Exacerbations are one of the most important manifestations of bronchiectasis and *P. aeruginosa* is the most frequent organism in severe bronchiectasis worldwide
- Cylindrical bronchiectasis is the most common morphological pattern identified on CT scans



MANAGEMENT



- 1. Review current airway clearance regime.
- 2. Repeat sputum microbiology and repeat testing for NTM, ABPA and ensuring the all possible treatable causes and comorbidities have been identified.
- 3. First-line recommendation for *P. aeruginosa* with frequent exacerbations is an inhaled antibiotic.

THANK YOU