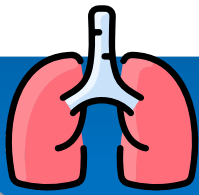




Round : Internal medicine

Group no : A4

Done By : Batool ALzubaidi



BRONCHIECTASIS

I. PATHOPHYSIOLOGY

A. BRONCHIECTASIS

II. CLASSIC FINDINGS OF BRONCHIECTASIS

- A. PRODUCTIVE AND FOUL SMELLING COUGH

III. COMPLICATIONS OF BRONCHIECTASIS

- A. HEMOPTYSIS
- B. RESPIRATORY FAILURE
- C. COR PULMONALE

IV. DIAGNOSTIC APPROACH TO BRONCHIECTASIS

- A. ASSESS FOR RADIOGRAPHIC EVIDENCE OF BRONCHIECTASIS
- B. ASSESS FOR EVIDENCE OF OBSTRUCTIVE LUNG DISEASE
- C. ASSESS FOR CAUSES OF BRONCHIECTASIS

V. TREATMENT OF BRONCHIECTASIS

- A. SUPPRESS CHRONIC BRONCHIOLE INFLAMMATION
- B. IMPROVE MUCUS CLEARANCE
- C. ANTIBIOTIC THERAPY
- D. MANAGEMENT OF HEMOPTYSIS

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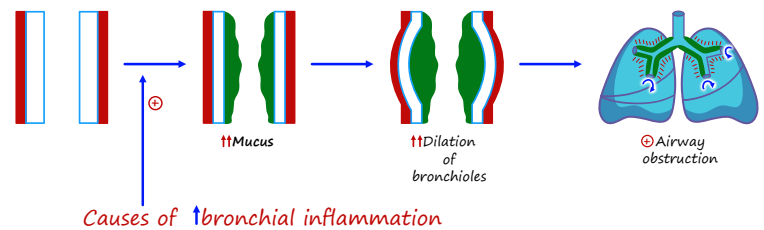
I. PATHOPHYSIOLOGY

A. BRONCHIECTASIS

1. Pathophysiology:

- Mucociliary dysfunction or Chronic Bronchiole Inflammation → ↑ Mucus production by goblet cells → Trapping of pathogens in the mucus which propagates more inflammation → The inflammation leads to bronchiole dilation along, bronchospasm and more mucus production → Bronchiole luminal obstruction occurs secondary to mucus plugging and bronchospasm → Airway obstruction ensues

Massive bronchial dilatation secondary to a lot of inflammation in the airways, super thick layer of mucus that smells very bad



2. Causes of Bronchiectasis

a) Mucociliary Dysfunction

i) Cystic Fibrosis

- CFTR Mutation →
 - ↓ Chloride secretion in bronchioles →
 - ↓ Water drawn into mucus →
 - Thick mucus in bronchioles →
 - Inability to move the mucus through the bronchioles
 - This disease leads to the trapping of pathogens (e.g. Pseudomonas)
- Think about frequent pulmonary infections & extra pancreatic insufficiency

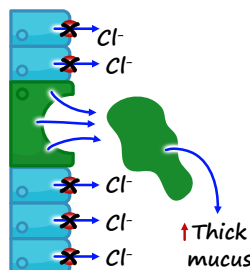
ii) Primary Ciliary Dyskinesia

- Ciliary Dysfunction →
 - Inability to move the mucus through the bronchioles
 - This disease leads to situs inversus and chronic sinusitis
- Function of cilia is to move the mucus upward so you can spit it out » their dysfunction results with build up of mucus

iii) Bronchiole Obstruction

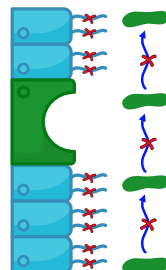
- Tumor or Foreign Bodies
 - Physically obstructs the bronchioles →
 - Leads to the inability to move the mucus through the bronchioles
- COPD
 - Leads to Bronchospasm →
 - This obstructs the bronchioles →
 - leads to the inability to move the mucus through the bronchioles
 - This disease leads to the trapping of pathogens (e.g. H.flu)

The cystic fibrosis transmembrane receptor protein is unable to produce chloride » these transporters will be defected and chloride won't be secreted » mucus can not be thinned by chloride which will make it massively thick



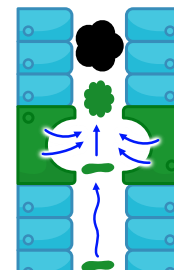
Thick mucus

→ Cystic fibrosis



↓ Ciliary function

→ Primary Ciliary Dyskinesia



Airway obstruction

→ Tumor
→ COPD
→ Foreign body



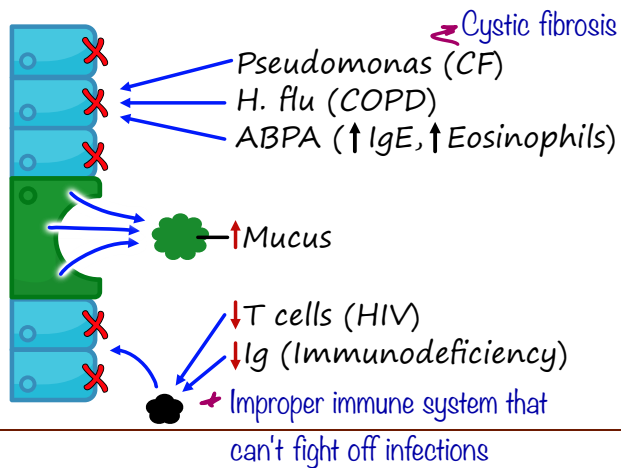
b) Recurrent Infections

i) Allergic Bronchopulmonary Aspergillus (ABPA)

- **Aspergillus** → Chronic airway inflammation
 - This disease presents with Asthma exacerbations, brown mucus production, and ↑IgE and Eosinophils

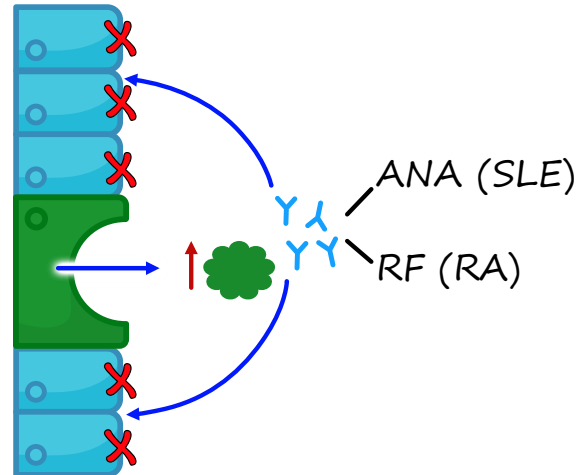
ii) Immunodeficiency

- **HIV**
 - ↓CD4 count → ↑Risk of **opportunistic infections**, such as Pneumocystis Jiroveci Pneumonia (PJP) → Resulting in **Chronic Airway Inflammation**
- Immunoglobulin Deficiency:
 - ↓IgA, IgG → ↓immune response → ↑Risk of infections → Resulting in **Chronic Airway Inflammation**



c) Chronic Inflammation

- **SLE/RA** → Chronic systemic inflammation → **Chronic Airway Inflammation** (Result as an adverse effect)
- **Assess for Antibody Involvement:**
 - **SLE** → ANA and Anti-dsDNA antibodies
 - **RA** → RF and Anti-CCP antibodies



00:36

II. CLASSIC FINDINGS OF BRONCHIECTASIS

A. PRODUCTIVE AND FOUL SMELLING COUGH

- This is secondary to **massive mucus production** in the bronchioles that traps pathogens and propagates more mucus production → Inflammation of the bronchioles triggers a cough reflex → Production of a cough filled with **"heavy mucus"** and pathogens leading to a **"foul smell"**



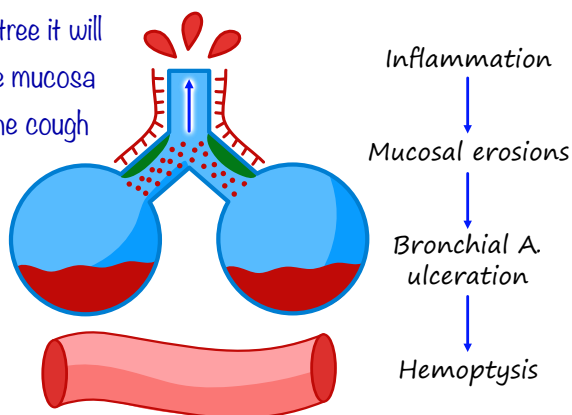
III. COMPLICATIONS OF BRONCHIECTASIS

A. HEMOPTYSIS

14:58

- **Pathophysiology:**
 - Chronic bronchiole inflammation → Erosion and destruction of bronchiole walls → Erosion of bronchiole arteries, which reside in the bronchiole wall → **Blood leaks into the bronchiole lumen**
- **Presentation:**
 - **Hemoptysis**
 - Most common cause of massive hemoptysis
 - **Acute Respiratory Failure in the setting of hemoptysis**
 - This may be due to blood clogging up the alveoli, **impairing gas exchange**
 - Assess for **↑RR** and **↑WOB**
 - Assess for **Hypoxia**

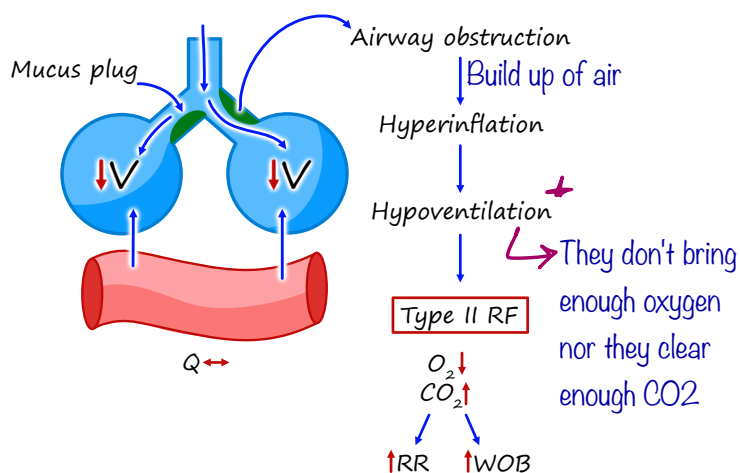
When blood enters the bronchial tree it will irritate the mucosa causing the cough



B. RESPIRATORY FAILURE

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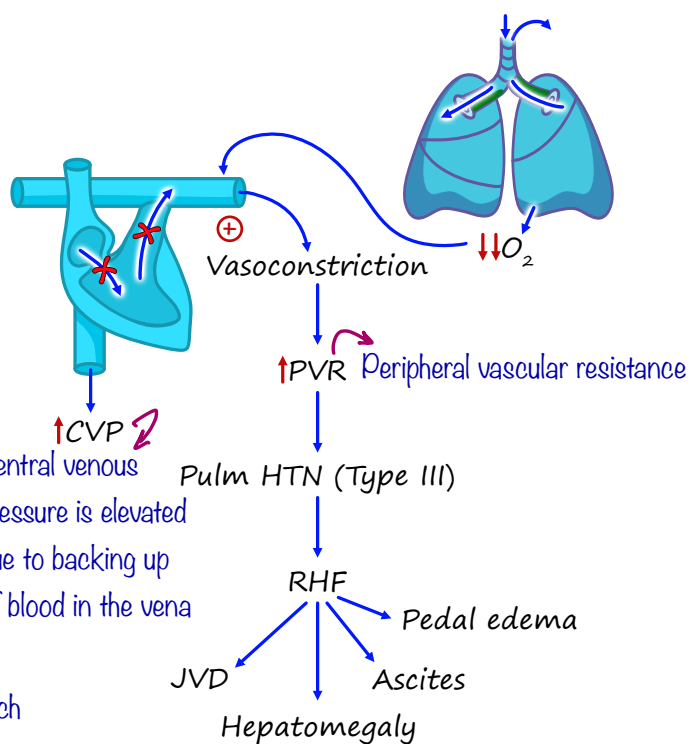
- **Pathophysiology:**
 - Chronic bronchiole inflammation → Erosion and destruction of bronchiole walls → ↑Mucus production and bronchospasm → **Airway obstruction** → **Alveolar hypoventilation** → **Impaired gas exchange**
- **Presentation:**
 - **↑RR** and **↑WOB**
 - **Hypoxia**
 - **Wheezing and Rhonchi** upon auscultation



C. COR PULMONALE

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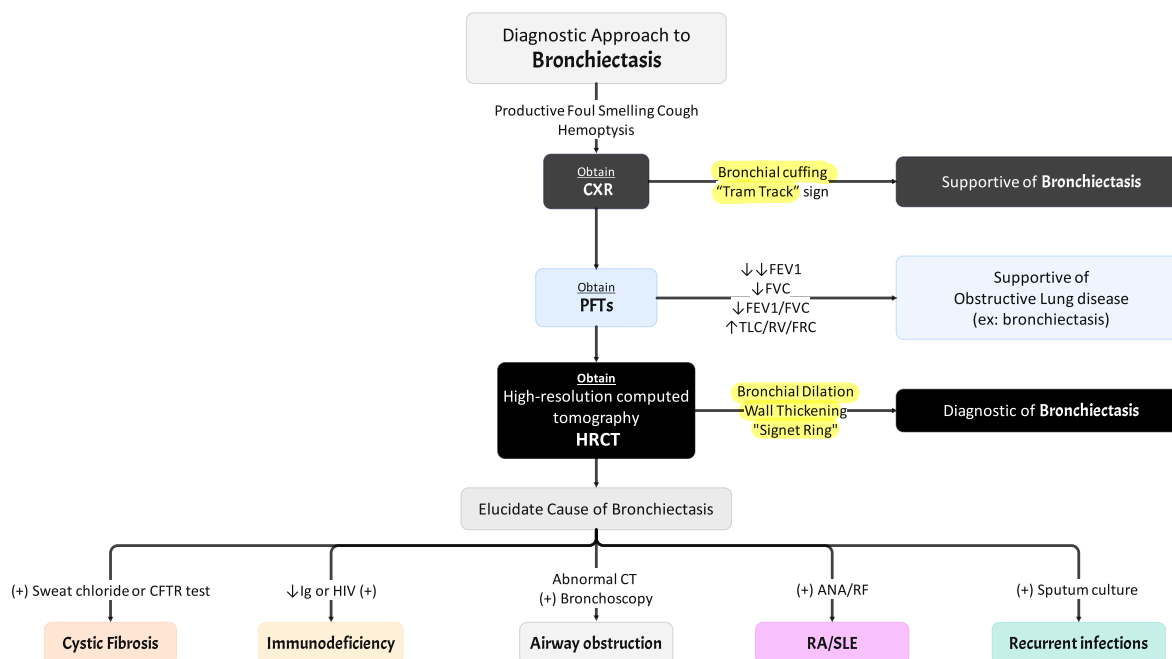
- **Pathophysiology:**
 - Chronic bronchiole inflammation and mucus plugging → **Chronic respiratory failure** → **Chronic hypoxic pulmonary vasoconstriction** → ↑Pulmonary vascular resistance → ↑Pulmonary artery pressures → **Right heart strain** arises (e.g. RVH) → **Right heart failure develops from chronic right heart strain**
- **Presentation:**
 - **JVD** (related to SVC backflow)
 - **Hepatomegaly** (related to IVC backflow)
 - **Ascites** (related to IVC backflow)
 - **Lower extremity edema** (related to IVC backflow)



Generally speaking the patients with bronchiectasis the common cause is recurrent infections » they build up mucus which they can't clear from their airway » perfect media for the bacteria to colonize and stay » they have high amounts of bacteria in their airways that can cause infections » they can develop very frequent pulmonary infections



IV. DIAGNOSTIC APPROACH TO BRONCHIECTASIS



A. ASSESS FOR RADIOGRAPHIC EVIDENCE OF BRONCHIECTASIS

1. Obtain CXR

- **Purpose:**
 - Assess for findings of bronchiectasis in a patient presenting with a **foul and productive cough** or **hemoptysis**
 - Rule out other lung pathologies (e.g. PNA, Lung Mass)
- **Abnormal Findings:**
 - **Bronchial Cuffing:**
 - This is representative of bronchiole wall thickening
 - **"Tram Track" Sign:**
 - This is representative of bronchiole wall thickening and dilation

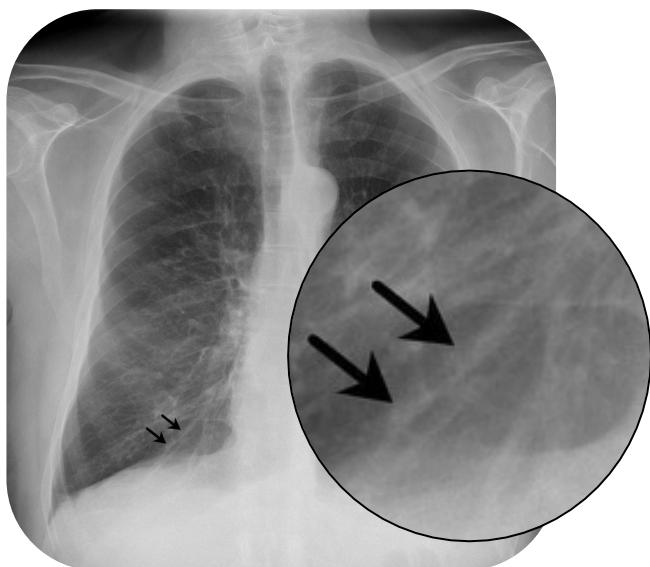


FIGURE 1. XRAY SHOWING "TRAM TRACK" SIGN

2. Obtain High-Resolution CT (HRCT)

- **Purpose:**
 - Definitively diagnose bronchiectasis based on CXR and history (e.g. Foul/productive cough and hemoptysis) being highly suggestive of **Bronchiectasis**
- **Abnormal Findings:**
 - **Bronchial dilation**
 - **Wall thickening of bronchioles**
 - **"Signet Ring" sign**
 - Dilated bronchus with an associated bronchial artery next to it
 - If the ratio between (e.g. Bronchus and Artery) is > 1 → Supports Bronchiectasis

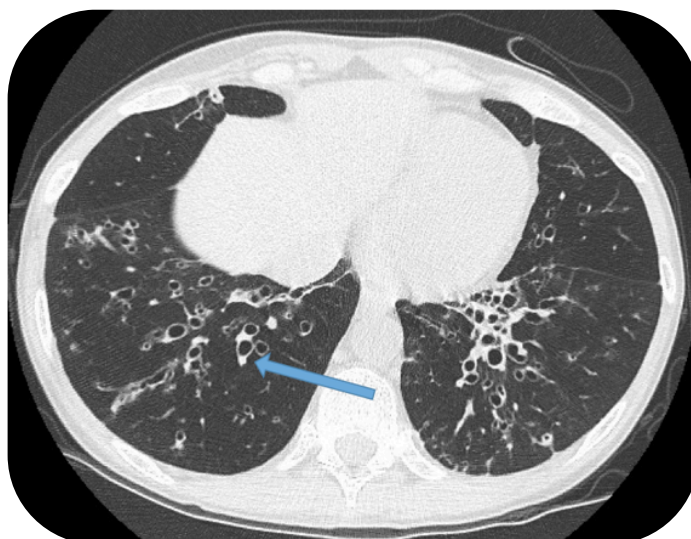


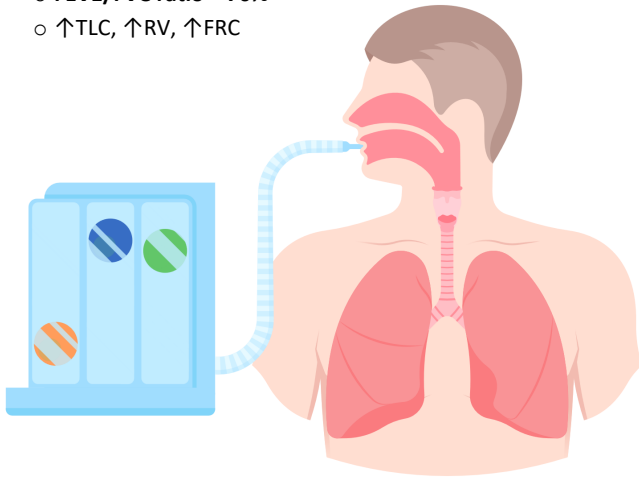
FIGURE 2. HRCT SHOWING THE "SIGNET RING" SIGN



B. ASSESS FOR EVIDENCE OF OBSTRUCTIVE LUNG DISEASE

1. Obtain PFTs:

- **Purpose:**
 - **Identify Obstructive Lung Disease Findings:**
 - If the history (foul/productive cough, hemoptysis) and Imaging (CXR, HRCT) suggest bronchiectasis → The use of PFTs will aid in the diagnosis (Not mandatory to perform)
- **Abnormal Findings:**
 - ↓↓ FEV1
 - ↓ FVC
 - FEV1/FVC ratio < 70%
 - ↑ TLC, ↑ RV, ↑ FRC



C. ASSESS FOR CAUSES OF BRONCHIECTASIS

1. Obtain Sweat Chloride Test:

- **Indications:**
 - **Bronchiectasis** in childhood or early adulthood → Support **Cystic Fibrosis**
- **Abnormal findings:**
 - ↑ Sweat chloride suggests cystic fibrosis
 - In individuals with CFTR dysfunction, the sweat glands are unable to effectively transport chloride from the sweat into the gland cells. As a result, chloride is not pumped into the cells and instead accumulates on the skin surface

2. Obtain Serum Immunoglobulins and HIV/CD4 Count Testing:

- **Indications:**
 - Recurrent pulmonary infections and poor response to vaccines → **Ig Deficiency**
 - Recurrent Pulmonary infections with a low CD4 count → **Opportunistic infection in the setting of HIV**
- **Abnormal Findings:**
 - ↓↓ IgA, IgG, and/or IgM levels → Ig Deficiency-related Bronchiectasis
 - **HIV (+) test and CD4 < 200** → HIV/AIDS-related Bronchiectasis

3. Obtain ANA, Anti-dsDNA, RF, and Anti-CCP Antibodies:

- **Indications:**
 - Assess for the possibility of RA/SLE-related bronchiectasis given joint pain, rash, and other systemic involvement
- **Abnormal Findings:**
 - **SLE** → (+) ANA and Anti-dsDNA antibodies
 - **RA** → (+) RF and Anti-CCP antibodies

4. Obtain IgE & IgG for Aspergillus, Eosinophil Count:

- **Indications:**
 - Assess for ABPA in patients with recurrent asthma, brown mucus production
- **Abnormal Findings:**
 - ↑ IgE and IgG against Aspergillus
 - ↑ Eosinophil count

5. Obtain Sputum Culture

- **Indications:**
 - Assess for the type of pathogens in Bronchiectasis
- **Abnormal Findings:**
 - Pseudomonas (seen in CF)
 - H.Flu (seen in COPD)
 - Aspergillus (seen in ABPA)



A. SUPPRESS CHRONIC BRONCHIOLE INFLAMMATION

- **Treatment:**
 - Treatment of the underlying cause
- **Purpose:**
 - Treating the underlying cause leads to a reduction in further airway inflammation, mucus production, and bronchiole inflammation and/or destruction

B. IMPROVE MUCUS CLEARANCE

- **Treatment:**
 - **Chest Physiotherapy** (Vest Therapy, Postural Drainage)
 - **Mucolytic Nebulizers** (n-acetylcysteine, 3% hypertonic nebulizers)
- **Indications:**
 - Frequent and copious thick secretions
- **Purpose:**
 - **Chest Physiotherapy**
 - Percussion helps to aid in the facilitation of **mobilizing the secretions**
 - **Mucolytics**
 - **Thin the secretions** aiding in easier mobilization of secretions
 - Common to be performed for Cystic Fibrosis
- **Monitoring:**
 - Improvement in sputum clearance
 - This will reduce the risk of bacterial infections → The improved clearance of mucus leads to less trapping of pathogens

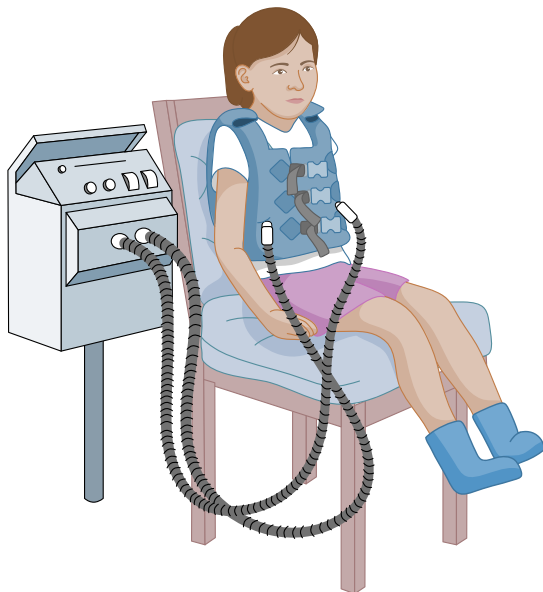


FIGURE 3. VEST THERAPY

C. ANTIBIOTIC THERAPY

- **Treatment:**
 - **Azithromycin** or **Doxycycline**
 - This is preferred for non-pseudomonas due to its anti-inflammatory properties
 - Acute Exacerbation, or Chronic suppressive therapy
 - **Fluoroquinolones** (Ciprofloxacin, Levofloxacin)
 - This is preferred for pseudomonas infection
 - Acute Exacerbation, or Chronic suppressive therapy
- **Indications:**
 - **Acute Exacerbations:**
 - ↑ Productive cough and Respiratory distress
 - **Chronic Suppressive Therapy:**
 - ≥ 3 exacerbations/year
- **Purpose:**
 - ↓↓ Bacterial growth → ↓↓ Inflammation → ↓↓ Further lung damage
- **Monitoring:**
 - Improvement in sputum production and reduced respiratory distress

≥ 3 Exacerbations/year

Antibiotic therapy

D. MANAGEMENT OF HEMOPTYSIS

- **Treatment:**
 - **Bronchial Artery Embolization**
 - If this fails this patient may need surgical resection of the diseased lung segment
- **Indications:**
 - **Severe Hemoptysis**
 - Hemodynamic instability
 - Acute respiratory failure
- **Purpose:**
 - Embolize the bleeding bronchiole artery to reduce hemoptysis
- **Monitoring:**
 - Improvement in hemoptysis
 - Improvement in HD status
 - Improvement in respiratory failure

Mechanism of Treatment	Treatment
↓↓ Chronic Inflammation	- Treat cause
↑↑ Mucus Clearance	- Chest Physiotherapy
↓↓ Bacterial Growth → ↓↓ Exacerbations	<u>Antibiotic Therapy (tailored towards pathogen):</u> - Fluoroquinolones for Pseudomonas coverage - Azithromycin if not pseudomonas
↓↓ Hemoptysis	- Bronchial Artery Embolization - Surgical Resection

