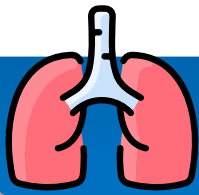




Round : Internal medicine

Group no : A4

Done By : Batool ALzubaidi



RESTRICTIVE LUNG DISEASE

I. PATHOPHYSIOLOGY

- A. INTRINSIC LUNG DISEASES
- B. EXTRINSIC LUNG DISEASES

II. CAUSES

- A. CAUSES OF INTRINSIC LUNG DISEASES
- B. CAUSES OF EXTRINSIC LUNG DISEASES

III. CLASSIC FINDINGS OF RESTRICTIVE LUNG DISEASES

- A. DRY COUGH
- B. BIBASILAR CRACKLES
- C. CLUBBING

IV. COMPLICATIONS OF RESTRICTIVE LUNG DISEASES

- A. RESPIRATORY FAILURE
- B. PULMONARY HYPERTENSION
- C. RIGHT HEART FAILURE (E.G. COR PULMONALE)
- D. LUNG CANCER

V. DIAGNOSTIC APPROACH TO RESTRICTIVE LUNG DISEASES

- A. ASSESS FOR RADIOGRAPHIC EVIDENCE OF INTRINSIC LUNG DISEASE
- B. ASSESS FOR EVIDENCE OF RESTRICTIVE VS OBSTRUCTIVE LUNG DISEASE
- C. ASSESS FOR THE CAUSE OF INTRINSIC LUNG DISEASE

VI. TREATMENT OF RESTRICTIVE LUNG DISEASES

- A. TREAT THE CAUSE OF ILD

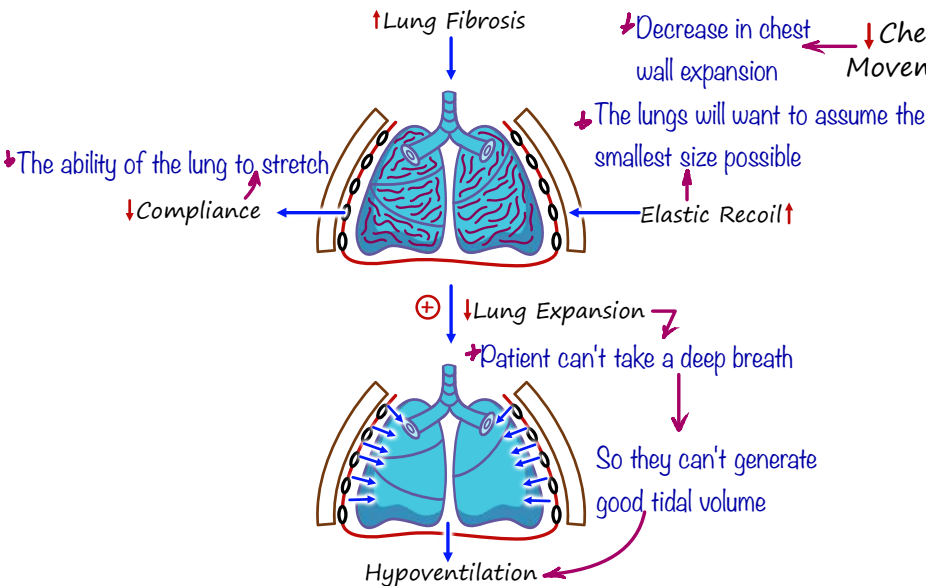
I. PATHOPHYSIOLOGY

A. INTRINSIC LUNG DISEASES

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- Lung fibrosis → ↓ Lung compliance and ↑ Elastic recoil → ↓ Lung expansion → Hypoventilation

• Lungs are the problem » lung fibrosis or injury

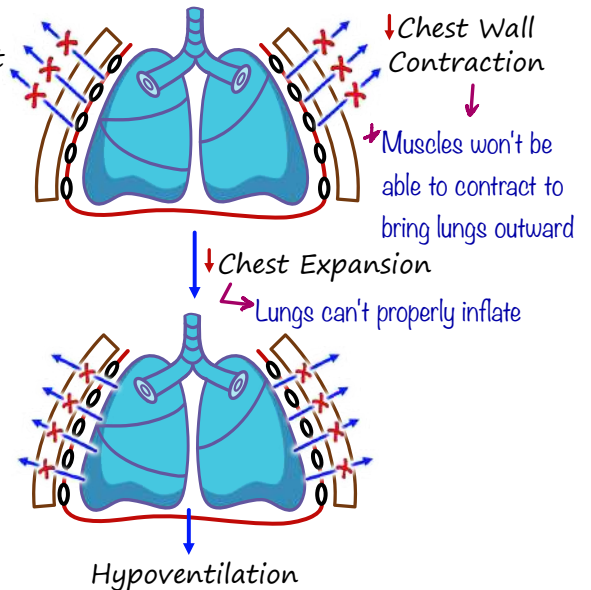


B. EXTRINSIC LUNG DISEASES

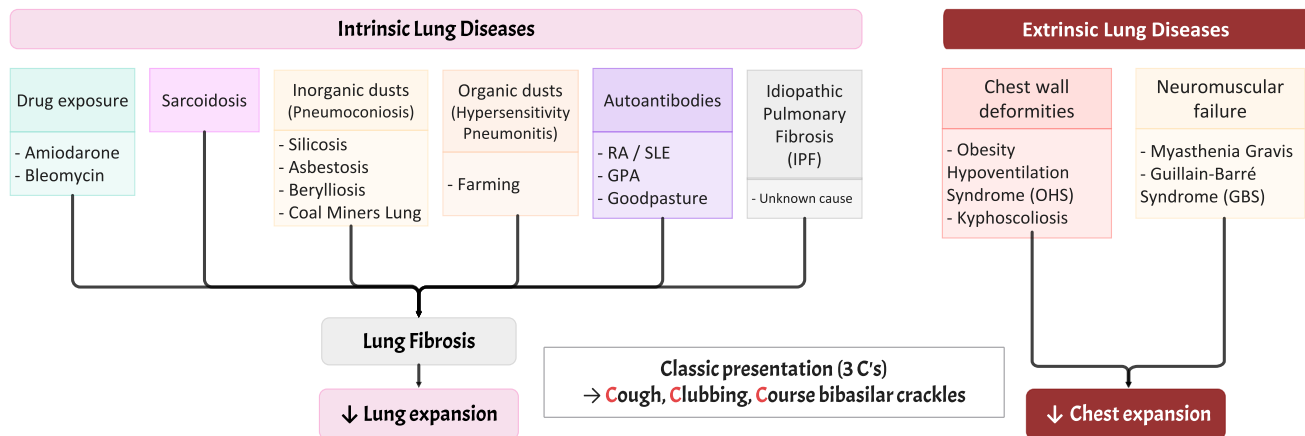
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- ↓ Chest wall movement or contraction → ↓ Chest wall expansion → ↓ Lung expansion → Hypoventilation

• Lungs are healthy there's no fibrosis or injury » chest wall muscles are the problem



II. CAUSES



A. CAUSES OF INTRINSIC LUNG DISEASES

1. Granulomatous Diseases

a) Sarcoidosis

- **Noncaseating granuloma formation** in lung tissue → Lung inflammation and fibrosis
 - Suspected in **young African-American** patients with **bilateral hilar lymphadenopathy** and **erythema nodosum**

b) Hypersensitivity Pneumonitis

- **Noncaseating granuloma formation** in lung tissue → Lung inflammation and fibrosis
 - Suspected in **farmers or bird handlers** → Symptoms improve when **avoiding exposure**

2. Pneumoconiosis

a) Asbestosis

- Asbestos inhalation → Lung fibrosis
 - Suspected in patients who worked in **shipbuilding, insulation, or construction**

b) Silicosis

- Silica inhalation → Lung fibrosis
 - Suspected in patients who worked in **sandblasting and stone cutters**
 - ↑Risk of TB infection

c) Berylliosis

- Beryllium exposure → Lung fibrosis
 - Suspected in patients who work in **aerospace jobs, dental equipment industries**

d) Coal Miners Lung

- Coal inhalation → Lung fibrosis
 - Suspected in **coal miners**

3. Autoimmune Diseases

a) Rheumatoid Arthritis

- **RF and Anti-CCP antibodies** → Lung injury and inflammation → Lung fibrosis
 - Assess for Joint disease

b) Systemic Lupus Erythematosus

- **ANA and Anti-dsDNA antibodies** → Lung injury and inflammation → Lung fibrosis
 - Assess for Joint and skin disease

c) Systemic Sclerosis

- **Anti-Scl-70** → Lung injury and inflammation → Lung fibrosis
 - Assess for **CREST syndrome** findings

4. Drug-Induced Lung Injury

a) Amiodarone

- Direct toxic effect on lung tissue → Lung fibrosis
 - Assess patients with **atrial fibrillation or ventricular arrhythmias** who are on chronic amiodarone

b) Bleomycin + Anti-neoplastic drug

- Direct toxic effect on lung tissue → Lung fibrosis
 - Assess patients with a **history of cancer** who had been on bleomycin

5. Idiopathic Pulmonary Fibrosis (IPF)

- Progressive lung fibrosis without an identifiable trigger
 - More common in **older adult males without any identifiable cause**



B. CAUSES OF EXTRINSIC LUNG DISEASES

1. Chest Wall Deformities

a) Obesity Hypoventilation Syndrome

- Obesity impedes abdomen and chest wall movement

b) Kyphoscoliosis

- Abnormal rib cage anatomy → ↓ Chest wall movement

2. Neuromuscular Dysfunction

a) Guillain Barré Syndrome

- ↓ Amplitude of action potentials propagate down the intercostal and phrenic nerves → ↓ Ability to take a deep breath and expand lungs → Impaired ventilation of multiple alveoli →

Hypercapnia ± Hypoxemia

- Suspect this in a patient with ascending paralysis and areflexia

b) Myasthenia Gravis

- Weak contraction of intercostals and diaphragm → ↓ Ability to take a deep breath and expand lungs → impaired ventilation of multiple alveoli → hypercapnia +/- hypoxemia

- Suspect this in a patient with ptosis, diplopia, dysphagia, and dysarthria

c) Spinal Cord Injury

- ↓ Amplitude of action potentials propagate down the intercostal and phrenic nerves → ↓ Ability to take a deep breath and expand lungs → impaired ventilation of multiple alveoli → Hypercapnia +/- hypoxemia

- Suspect this in a patient with paralysis

III. CLASSIC FINDINGS OF RESTRICTIVE LUNG DISEASES

A. DRY COUGH

- Lung fibrosis → Airway irritation → Dry cough

B. BIBASILAR CRACKLES

- Lung fibrosis → ↑ Elastic recoil and ↓ Lung compliance → Bronchiole and alveolar collapse → During inspiration the airways "pop open" creating the crackling sound at the bases

C. CLUBBING

- Lung fibrosis → ↑ Elastic recoil and ↓ Lung compliance → Hypoventilation → Chronic hypoxia → Poor nail bed oxygenation → Vascular changes of digits → Spoon appearance or clubbing of digits result



IV. COMPLICATIONS OF RESTRICTIVE LUNG DISEASES

A. RESPIRATORY FAILURE

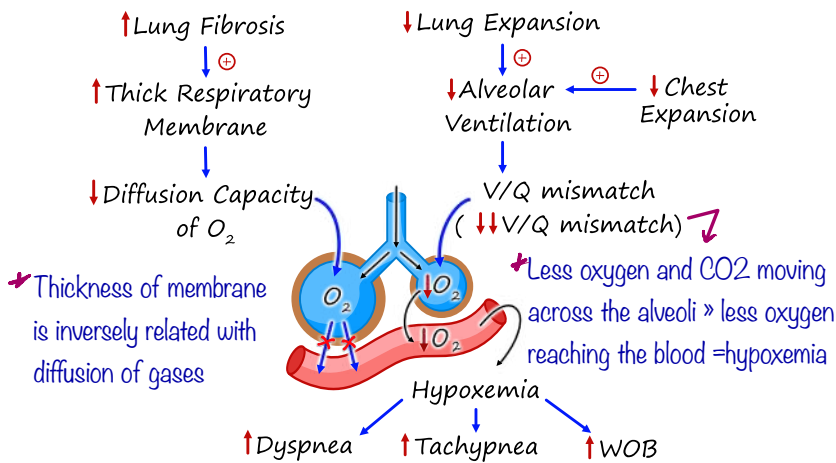
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● Pathophysiology:

- Chronic lung fibrosis → ↓ Lung expansion → Hypoventilation → Impaired gas exchange → Hypoxia
- Chronic lung fibrosis → Thick respiratory membrane → Impaired gas exchange → Hypoxia

● Presentation:

- Dyspnea, ↑RR, and ↑WOB
- Hypoxia (SpO₂ < 90%)
- ABG with Type I Respiratory Failure
 - ↓PaO₂ (hypoxemia)
 - Normal or ↓PaCO₂



B. PULMONARY HYPERTENSION

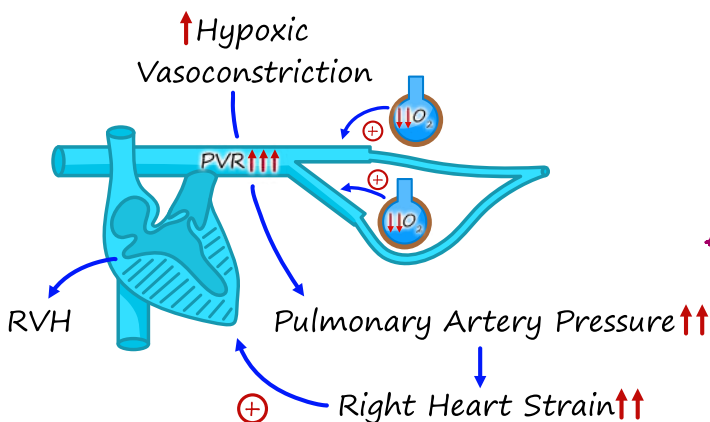
● Pathophysiology:

- **Chronic hypoxia** → ↑ Hypoxic Pulmonary Vasoconstriction → ↑ PVR → ↑ Pulmonary BP

● Presentation:

- Dyspnea on exertion
- ↑ Risk of Right Heart Failure (e.g. Cor Pulmonale)

Pulmonary Hypertension (Type III PAH)



C. RIGHT HEART FAILURE (COR PULMONALE)

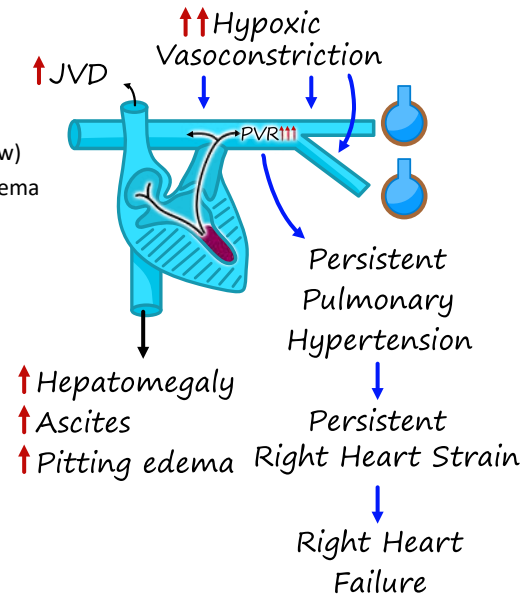
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● Pathophysiology:

- **Chronic respiratory failure** → Chronic hypoxic pulmonary vasoconstriction → ↑ Pulmonary vascular resistance → ↑ Pulmonary artery pressures → Right heart strain arises (e.g. RVH) → **Development of Right Heart Failure from Chronic Right Heart Strain**

● Presentation:

- JVD (SVC backflow)
- Hepatomegaly (IVC backflow)
- Ascites (IVC backflow)
- Lower extremity edema (IVC backflow)



D. LUNG CANCER

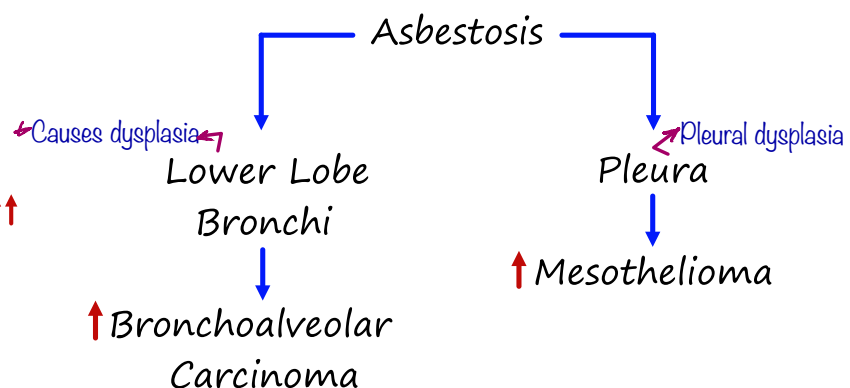
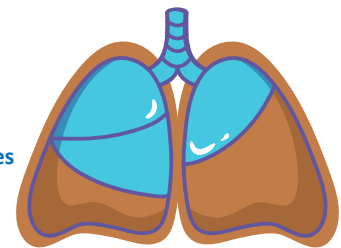
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● Pathophysiology:

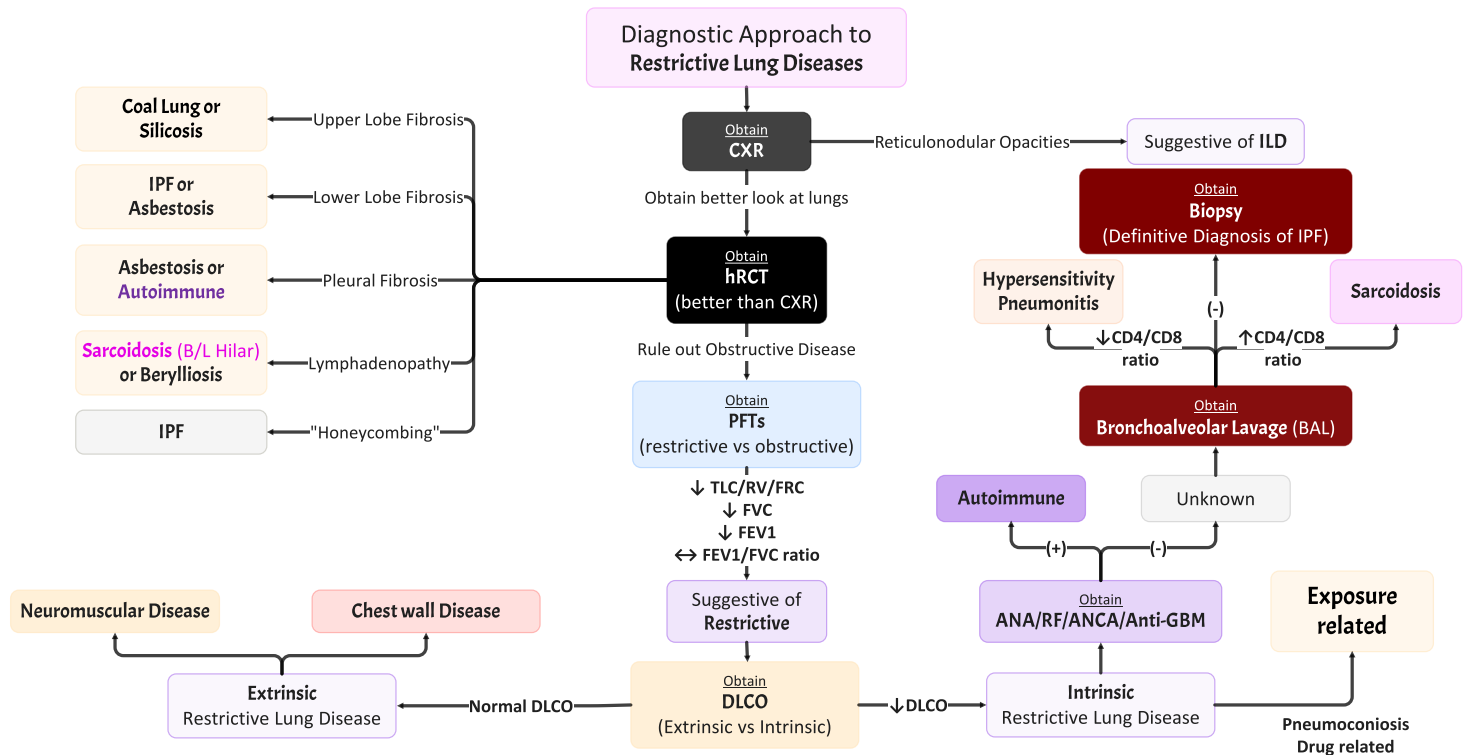
- **Asbestosis** → Chronic lung inflammation → ↑ Risk of lung dysplasia → ↑ Risk of Bronchogenic Carcinoma or Mesothelioma

● Presentation:

- **Bronchogenic Carcinoma:**
 - Hemoptysis
 - Weight loss
 - ↑ Risk of Paraneoplastic Syndromes
- **Mesothelioma:**
 - Weight loss
 - ↑ Risk of recurrent Pleural Effusions



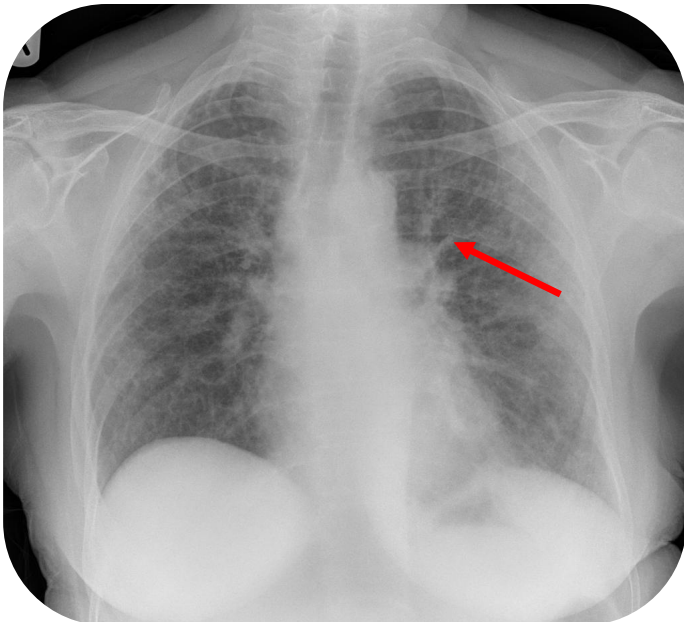
V. DIAGNOSTIC APPROACH TO RESTRICTIVE LUNG DISEASES



A. ASSESS FOR RADIOGRAPHIC EVIDENCE OF INTRINSIC LUNG DISEASE

1. Obtain CXR

- **Purpose:**
 - Assess for alternative causes of dyspnea
- **Abnormal Findings:**
 - **Reticulonodular Opacities**
 - This indicates **Interstitial Fibrosis**



2. Obtain an HRCT

- **Purpose:**
 - CXR suggests ILD and requires a definitive evaluation of the lungs
- **Abnormal Findings:**
 - Upper Lobe Fibrosis → “Coal Miner’s” Lung or Silicosis
 - Lower lobe Fibrosis → IPF or Asbestosis
 - Pleural Fibrosis → Asbestosis or Autoimmune
 - Lymphadenopathy → Sarcoidosis
 - Honeycombing and ground glass Opacities → IPF

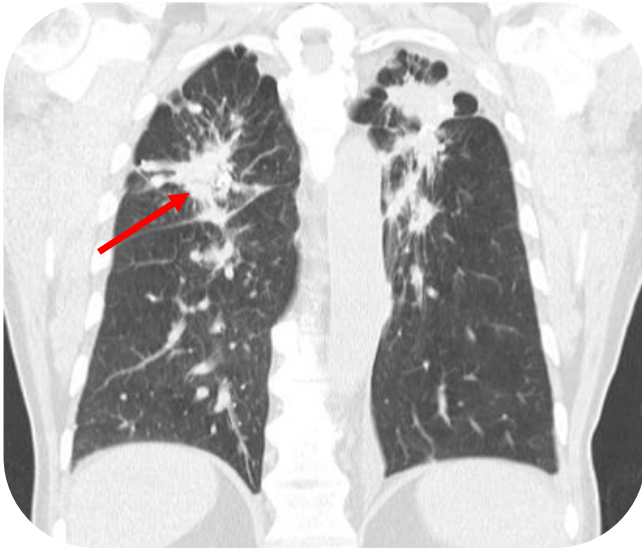


FIGURE 1. HRCT OF SILICOSIS



FIGURE 2. HRCT OF SARCOIDOSIS

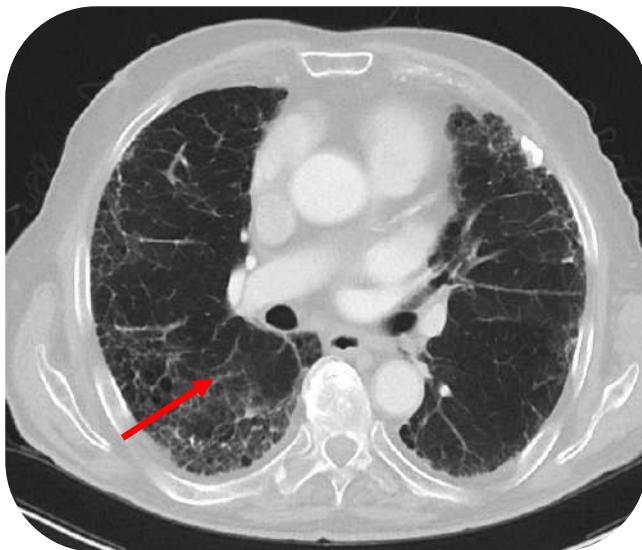


FIGURE 4. HRCT OF ASBESTOSIS



FIGURE 3. CT SHOWING HONEYCOMBING SIGN



B. ASSESS FOR EVIDENCE OF RESTRICTIVE VS OBSTRUCTIVE LUNG DISEASE

1. Obtain PFTs

- **Parameters of PFTs:**

- FEV₁/FVC ratio
- Diffusion limitation of carbon monoxide (DLCO)
- Lung Volumes (e.g. TLC, RV, and FRC)

- **Purpose:**

- Determine if the patient has an obstructive vs restrictive lung disease

- **Findings Supportive of Restrictive Lung Disease:**

- Obstructive Lung Disease pattern that is reversible:
 - **FEV₁/FVC ratio > 70% suggests Restrictive Lung Disease**

- **DLCO:**

- **ILD exhibits a ↓DLCO**

- ILD increases the thickness of the respiratory membrane's alveolar surface area → Decreasing the diffusion of gases

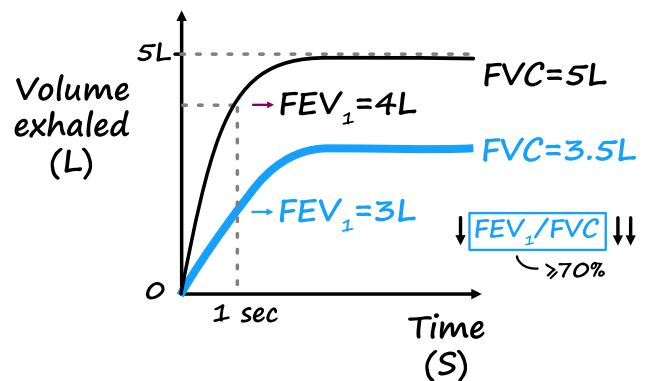
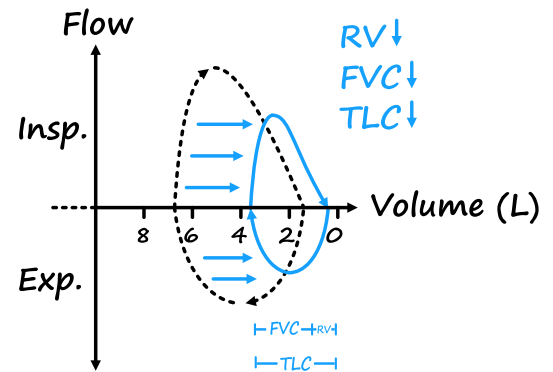
- **Extrinsic lung diseases exhibit a normal DLCO**

- Extrinsic Lung Diseases do not affect the surface area or thickness of the respiratory membrane

- **Lung Volumes:**

- **↓↓TLC, RV, and FRC**

- Due to poor lung expansion from lung fibrosis



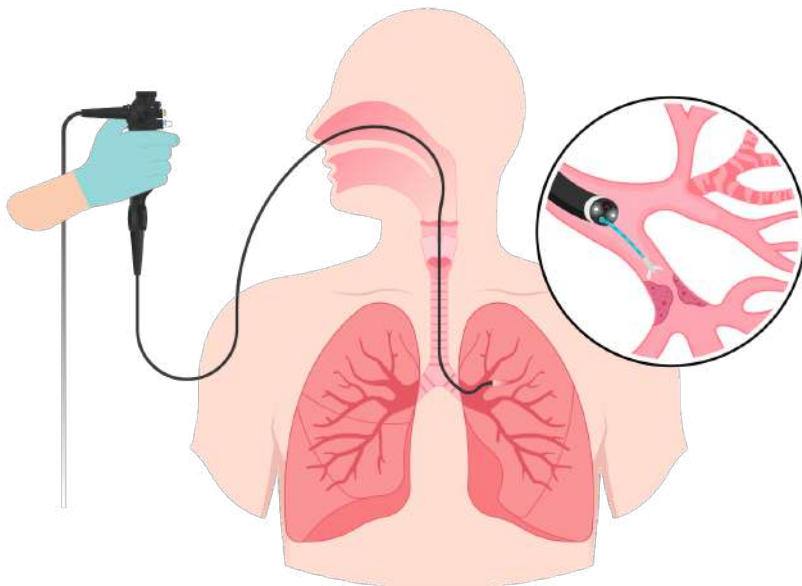
C. ASSESS FOR THE CAUSE OF INTRINSIC LUNG DISEASE

1. Obtain **Abstinence from Exposure to Trigger**

- **Indications:**
 - CXR/HRCT and PFTs suggest ILD, and history suggests exposure-related (e.g. Drugs or Pneumoconiosis)
- **Abnormal Findings suggestive of exposure-related ILD**
 - **Improvement in ILD with avoidance of occupational exposure or drug**
 - **Asbestosis:**
 - Pleural plaques and lower lobe fibrosis in the setting of avoiding construction, insulation, or shipbuilding
 - **Silicosis:**
 - Upper lobe fibrosis in the setting of avoiding stone cutting and sandblasting
 - **Coal Miner's Lung:**
 - Upper lobe fibrosis in the setting of avoiding coal mines

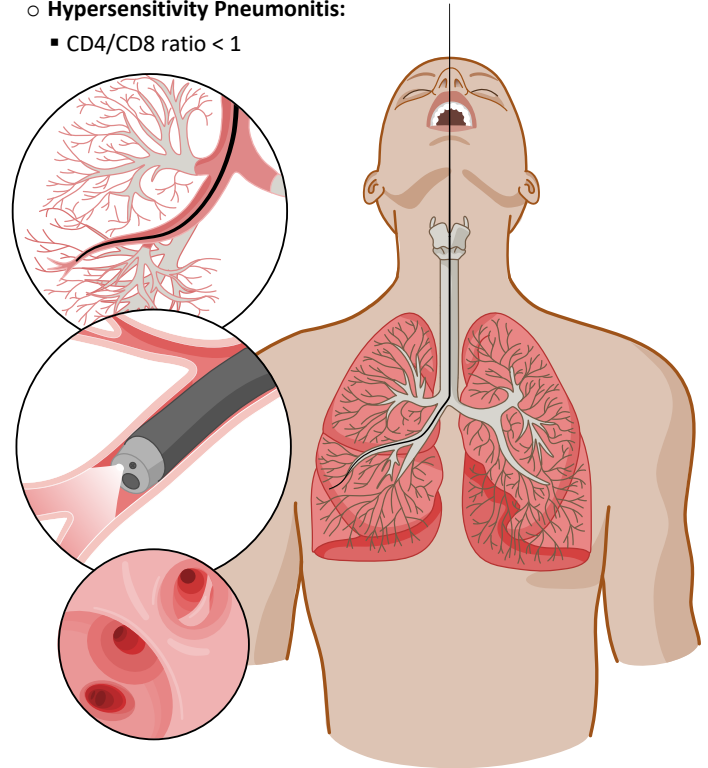
2. Obtain **Autoantibody panel** (e.g. ANA, RF, Anti-scl-70)

- **Purpose:**
 - Assess for autoimmune-related causes of ILD such as **SLE**, **RA**, or **systemic sclerosis**
- **Abnormal Findings:**
 - RA → (+) RF and Anti-CCP antibodies
 - SLE → (+) ANA and Anti-dsDNA antibodies
 - Systemic sclerosis (+) Anti-Scl-70 antibodies



3. Obtain **Bronchoalveolar Lavage (BAL)**

- **Indications:**
 - CXR/HRCT and PFTs suggest ILD and antibody panel negative and no improvement with avoidance of exposure and need to assess for CD4/CD8 ratio
- **Abnormal Findings:**
 - **Sarcoidosis:**
 - CD4/CD8 ratio > 1
 - **Hypersensitivity Pneumonitis:**
 - CD4/CD8 ratio < 1



4. Obtain **Lung Biopsy**

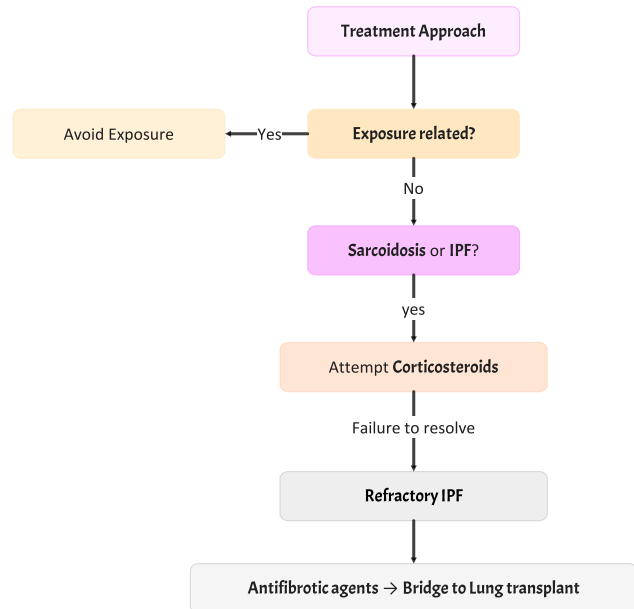
- **Indications:**
 - CXR/HRCT and PFTs suggest ILD, antibody panel, and BAL negative and no improvement with avoidance of exposure
- **Abnormal Findings:**
 - **IPF**
 - Definitively diagnose IPF as the cause of ILD
 - Not always required for diagnosis due to the invasive nature of obtaining a biopsy



VI. TREATMENT OF RESTRICTIVE LUNG DISEASES

Treatment of Restrictive Lung Diseases

Restrictive Lung Disease	Treatment
Drug Induced - Amiodarone - Bleomycin	- Discontinue drugs
Pneumoconiosis - Silicosis - Asbestosis - Berylliosis - Coal Miners Lung	- Avoid exposure
Hypersensitivity Pneumonitis - Farming	- Avoid exposure - Corticosteroids
Granulomatous - Sarcoidosis	- Corticosteroids
Autoimmune - RA / SLE - GPA - Goodpasture	- Corticosteroids
IPF	- Pirfenidone - Nintedanib - Lung transplant
Extrinsic Disease - OHS	- Weight loss



A. TREAT THE CAUSE OF ILD

1. Exposure-Related ILD

- **Indications:**
 - ILD with occupational and drug exposure
- **Purpose:**
 - ↓ Progressive lung fibrosis
- **Monitoring:**
 - Improvement in dyspnea, cough, crackles
 - Improvement in PFTs
 - Improvement in CXR/HRCT

2. Sarcoidosis and Autoimmune ILD

- **Types of Therapy:**
 - Corticosteroids
- **Indications:**
 - Sarcoidosis or autoimmune ILD
 - IPF
- **Purpose:**
 - ↓ Progressive lung fibrosis
- **Monitoring:**
 - Improvement in dyspnea, cough, crackles
 - Improvement in PFTs
 - Improvement in CXR/HRCT

3. Idiopathic Pulmonary Fibrosis (IPF)

- **Types of Therapy:**
 - Corticosteroids
 - Anti-fibrotic agents (e.g. Pirfenidone or Nintedanib)
 - Lung transplantation
- **Indications:**
 - **Corticosteroids**
 - Acute exacerbation of IPF
 - Not commonly used
 - **Anti-Fibrotic Agents** (e.g. Pirfenidone or Nintedanib)
 - Mild to moderate IPF
 - **Lung Transplantation**
 - Severe IPF refractory to anti-fibrotic agents with high mortality risk
- **Purpose:**
 - **Anti-Fibrotic Agents** (e.g. Pirfenidone or Nintedanib)
 - ↓ Progressive lung fibrosis
- **Monitoring:**
 - Improvement in dyspnea, cough, crackles
 - Improvement in PFTs
 - Improvement in CXR/HRCT

