

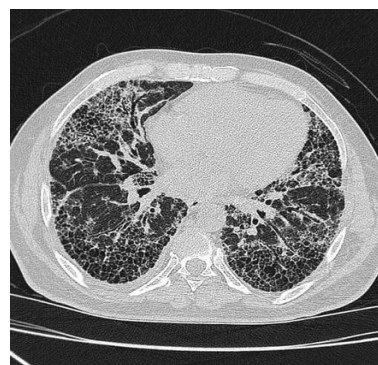
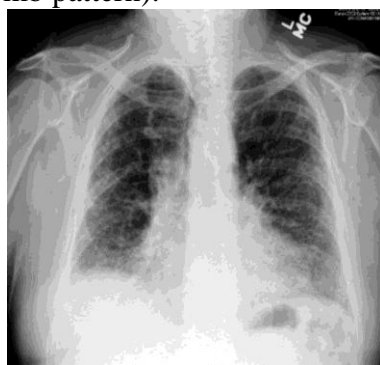


- **Interstitial lung diseases (introduction):**

- **Definition:** it is chronic inflammation and fibrosis of lung interstitium and parenchyma. This will result in thickened alveolar-capillary membrane thus impairing gas exchange.
- **Etiology:**
 - ✓ Idiopathic pulmonary fibrosis.
 - ✓ Sarcoidosis.
 - ✓ Pneumoconiosis and occupational lung diseases.
- **Clinical manifestation:**
 - ✓ Most common: exertional dyspnea.
 - ✓ Non-productive cough.
 - ✓ Physical examination: coarse crackles on auscultation; clubbing of fingers (not always).
- **Diagnosis:**
 - ✓ Chest x-ray and CT-scan: ground-glass appearance.
 - ✓ PFT: restrictive pattern ($FEV_1 < 80\%$; $FEV_1/FVC \geq 80\%$) and decreased DLco.
 - ✓ Lung biopsy has to be done to differentiate between these diseases.

- **Idiopathic pulmonary fibrosis:**

- **Definition:** it is an inflammatory lung disease of unknown origin resulting in lung fibrosis and restrictive lung disease.
- **Epidemiology:** 5th decade of life. Males = females.
- **Clinical manifestations:**
 - ✓ Exertional dyspnea.
 - ✓ Physical examination: coarse crackles and clubbing of the fingers.
- **Diagnosis:**
 - ✓ Chest x-ray: reticular or reticulonodular disease.
 - ✓ CT-scan: ground-glass appearance (with advanced disease you will see honeycomb pattern).



- ✓ PFT: restrictive lung pattern.
- ✓ Biopsy is done to exclude other possible differentials.
- **Treatment:**
 - ✓ Pharmacological: pirfenidone (has anti-fibrotic effect).
 - ✓ Non-pharmacological: lung transplantation.

- **Sarcoidosis:**

- **Definition:** it is a systemic disease of unknown etiology resulting in non-specific, non-caseating granulomas which involve the lung and any other organs of the body.
- **Epidemiology:** age (20-40); more among blacks.
- **Clinical manifestations:**
 - ✓ Lung involvement in 90% of patients: represented by hilar adenopathy on chest x-ray.



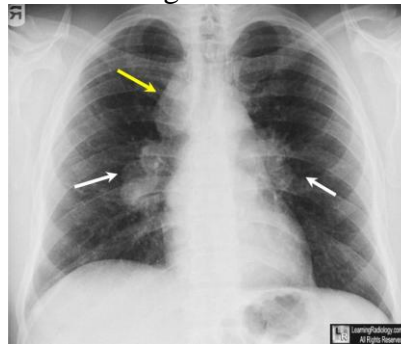
- ✓ Skin manifestations in 25% of patients: lupus pernio (image), erythema nodosum, papules, non-scarring alopecia.



- ✓ Uveitis/conjunctivitis in > 25% of patients.

- **Diagnosis:**

- ✓ Laboratory investigations: hypercalcemia/hypercalciuria; ↑ACE in 60% of patients; abnormal LFTs in 30% of patients.
- ✓ Chest x-ray will show 4 stages of the disease: bilateral hilar adenopathy, hilar adenopathy with reticulonodular parenchyma; reticulonodular parenchyma only; honeycombing of bilateral lung fields with fibrosis.



- ✓ PFT: normal or showing a restrictive pattern.
- ✓ Biopsy (definitive diagnosis): non-caseating granulomas.

- **Treatment**: there is no evidence that any therapy alters the course of the disease.
- **Prognosis**: 80% of patients will remain stable or resolve spontaneously.

- **Pneumoconioses:**

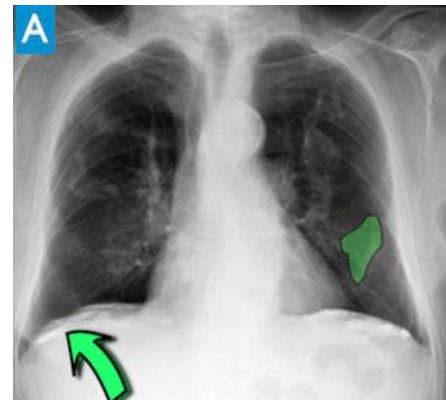
- **Introduction:**

- ✓ Definition: also known as occupational lung diseases in which inhalation of certain dusts/fibers results in inflammation and fibrosis of lung parenchyma (this occurs 20-30 years after chronic exposure to these substances).
- ✓ Pathology: alveolar macrophages will engulf those fibers and initiate inflammatory response which eventually will result in fibrosis of lung parenchyma.

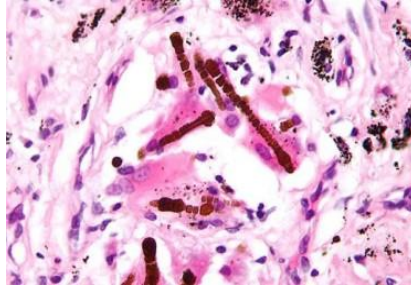
- **Asbestosis:**

- ✓ Definition: it is an occupational lung disease in which chronic exposure to asbestose dust will result in lung fibrosis and restrictive lung disease. Asbestose dust is found in mining, foundry work, shipyards and pipes.
- ✓ Clinical manifestations: exertional dyspnea, cough and wheezing (especially in smokers).
- ✓ Diagnosis:

- ❖ Chest x-ray: diffuse or local pleural thickening; pleural plaques and calcifications at the level of the diaphragm. It commonly involves lower lung fields.



- ❖ *Biopsy (definitive diagnosis):* showing barbell-shaped asbestose fiber.



- ✓ Complications:
 - ❖ Most common cancer associated with asbestosis is bronchogenic carcinoma (adenocarcinoma and squamous cell carcinoma).
 - ❖ Mesothelioma is also associated with asbestosis but not common.
- ✓ Treatment: no specific treatment is offered.

- **Silicosis:**

- ✓ Definition: it is an occupational lung disease which is caused by chronic inhalation of silica dust thus resulting in lung fibrosis. Silica dust is found in mining and glass/pottery making.
- ✓ Clinical manifestations: exertional dyspnea, cough and wheezing (especially among smokers).
- ✓ Diagnosis:
 - ❖ *Chest x-ray:* 1-10 mm hyaline nodules throughout the lung especially in upper lung fields.



- ❖ *Biopsy (definitive diagnosis).*

- ✓ Complications:
 - ❖ Silicosis is associated with tuberculosis (thus patient must undergo regular screening with PPD test).
- ✓ Treatment: there is no effective therapy for silicosis.

- **Coal Worker's Pneumoconiosis (CWP):**

- ✓ Definition: it is an occupational lung disease caused by prolonged inhalation of coal dust. It is seen in 12% of all miners.
- ✓ Clinical manifestations: dyspnea and cough.
- ✓ Diagnosis:
 - ❖ *Chest x-ray:* small round densities in lung parenchyma involving upper lung fields.
 - ❖ *Associated immunologic abnormalities:* elevated IgA, IgG, C3 and presence of ANA or RF.

