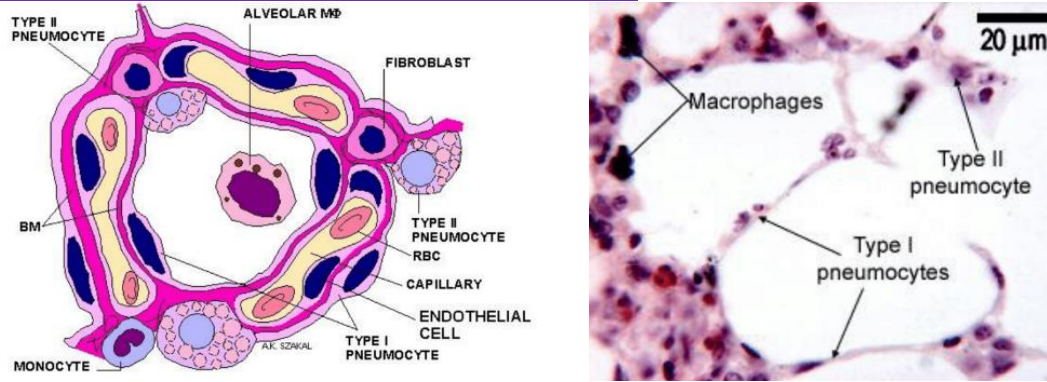




- The figure below demonstrates pulmonary alveoli:



- Restrictive disease: inability to fill the lung due to the following causes

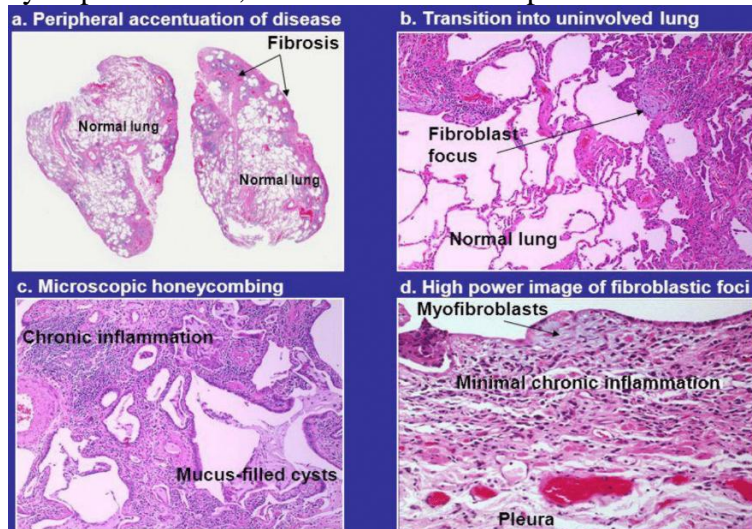
- **Interstitial lung disease:** a disorder in which there is fibrosis in interstitium resulting in thickening of barrier of gas exchange. Pulmonary interstitium is composed of:
  - ✓ Basement membrane.
  - ✓ Collagen fibers.
  - ✓ Elastic tissue.
  - ✓ Fibroblasts.
  - ✓ Few mast cells.
  - ✓ Occasionally mononuclear cells.
- **Chest wall abnormalities (such as obesity):** in which there is a force in the chest wall that is inhibiting the lung from being opened and filled.

- Restrictive disease spirometry changes:

- ↓ TLC
- ↓↓ FVC
- ↓ FEV<sub>1</sub>
- ↑  $\frac{FEV_1}{FVC}$  (> 80%).

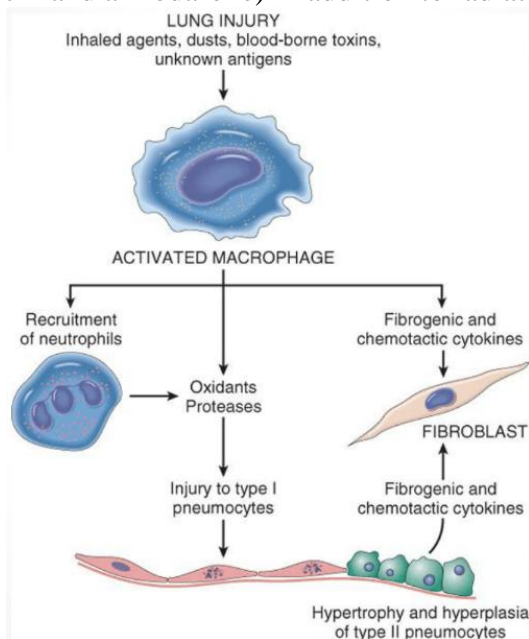
- Idiopathic pulmonary fibrosis:

- **Definition:** fibrosis of lung interstitium which is more common among males ≥ 60 years old.
- **Histology:**
  - ✓ Simply: thickened air sacs walls.
  - ✓ Dense fibrosis → collapse of alveolar walls + formation of cystic spaces lined by hypertrophic type-II pneumocytes of bronchiolar epithelium (honeycomb fibrosis).
  - ✓ The interstitial inflammation is composed mostly of lymphocytes and occasionally of plasma cells, mast cells and eosinophils.





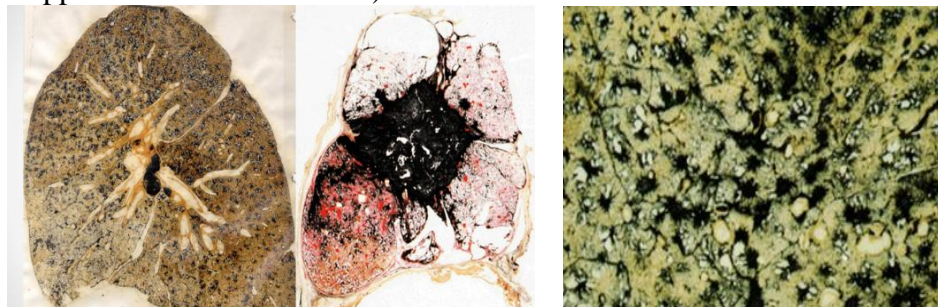
- **Cause:** cyclical lung injury (alveolitis) with cyclical lung healing in which there is generation of TGF- $\beta$  that induces fibrosis. Secondary causes for this disease include drugs (such as bleomycin and amiodarone) in addition to radiation therapy.



- **Clinical features:**
  - ✓ Progressive dyspnea (as fibrosis is getting worse with time) and cough.
  - ✓ Fibrosis is seen on lung CT-scan (initially in subpleural region and then progressing to involve the entire lung).
- **Treatment:** lung transplantation.

#### - Pneumoconioses:

- **Definition:** interstitial fibrosis due to occupational exposure to small particles (that are fibrogenic) and going to activate macrophages which will mediate fibrosis.
  - ✓ Coal workers pneumoconioses: carbon gets into macrophages which will induce massive fibrosis of the lung. This will result in black shrunken lung (coal macules 1-2 mm and coal nodules which are more noticed in upper lobes and upper zones of lower lobes).



- ✓ Silicosis: there is exposure to silica which impairs phagolysosomes to be formed in macrophages. This is the only pneumoconioses causing  $\uparrow$  risk for tuberculosis.
- ✓ Berylliosis: seen in miners and workers in aerospace industry. It is resulting in non-caseating granuloma in lungs, hilar lymph nodes and systemic organs. It increases the risk for lung cancer.
- ✓ Asbestosis: seen in construction workers, plumbers and shipyard workers. It creates fibrosis in lung/pleura or cancer in lung/pleura. Asbestosis bodies are long rod-shaped particles with iron deposited on them. Fibrosis begins in lower lobes and subpleurally (while coal workers pneumoconioses and silicosis affect upper lobes).

