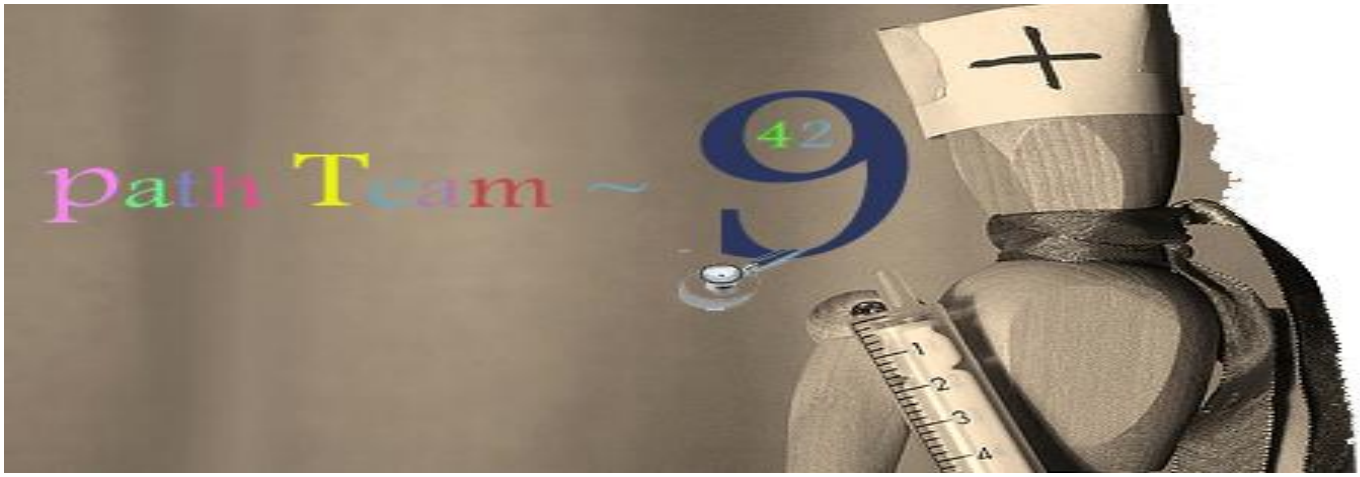


RESTRICTIVE PULMONARY DISEASES

هذه المذكرة عبارة عن هاند أوت الدكتور **عمار الركابي** بعد تنسيقها.
شاملة لكل ما هو مطلوب في هذه الجزئية

وأيضا:
توجد بعض الجداول والصور التوضيحية مضافة من محاضرات الدكتورة **مها عرفة**.

Path. Team - 429
Abdullah Ak-Faris
Muhammad Al-Harbi
Dona Barakah

GENERAL CONSIDERATIONS, DEFINITIONS AND CAUSES:

A. ***Restrictive Pulmonary Disease*** is a group of disorders characterized by **reduced** expansion of the lung and reduction in total lung capacity.

B. ***Examples*** include abnormalities of the chest wall from *Bony Abnormalities* or *Neuromuscular Disease* that restrict lung expansion.

C. ***The Interstitial Lung Disease*** a heterogeneous-group of disorders, characterized by interstitial accumulations of cells or non cellular material within the alveolar walls ((that restrict expansion and often interfere with gaseous exchange.))

Prominent examples are acute conditions

such as

- *The Adult and Neonatal Respiratory Distress Syndromes*
- *Pneumoconioses (e.g.* ***coal worker's pneumoconiosis***, ***silicosis*** and ***asbestosis***)
- *Diseases Of Unknown Etiology* (*e.g.* ***sarcoidosis*** and ***idiopathic pulmonary fibrosis)***
- *Various Other Conditions* (*e.g.* ***eosinophilic granuloma***, ***hypersensitivity pneumonitis***)
- *Chemical Or Drug Associated Disorders* (*e.g.* ***berylliosis*** or ***the pulmonary fibrosis*** *associated with* ***bleomyxin toxicity***)
- *Immune Disorders* (*e.g.* ***systemic lupus erythematosus***, ***systemic sclerosis (scleroderma)***, ***Wegener granulomatosis*** and ***Goodpasture syndrome.***)

ADULT RESPIRATORY DISTRESS SYNDROME (ARDS):

1) **Produced By**
Diffuse alveolar damage WITH resultant *increase* in Alveolar capillary permeability,
→ *causing* leakage of protein-rich fluid into alveoli.

2) **Characteristics** include:
the formation of an ***Intra-Alveolar Hyaline Membrane***
Composed of fibrin and cellular debris.

3) **The result** severe impairment of respiratory gas exchange with consequent severe hypoxia.

4) **Causes** include:
a wide variety of mechanisms and toxic agents, including

- Shock - sepsis - trauma - uremia - aspiration of gastric contents - acute Pancreatitis
- Inhalation of chemical irritants (e.g. chlorine, oxygen)
- toxicity or overdose with street drugs (e.g. heroin) or therapeutic drugs (e.g. bleomycin.)

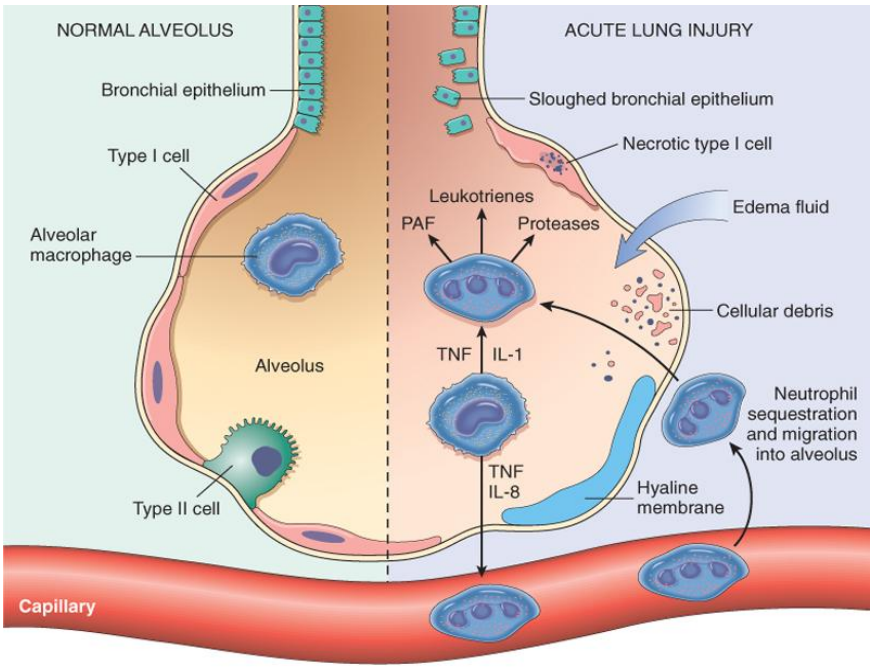
DIRECT LUNG INJURY	INDIRECT LUNG INJURY
COMMON CAUSES	
Pneumonia	Sepsis
Aspiration of gastric contents	Severe trauma with shock
UNCOMMON CAUSES	
Pulmonary contusion	Cardiopulmonary bypass
Fat embolism	Acute pancreatitis
Near-drowning	Drug: - ***Bleomycin*** - overdose ***Heroin***
Inhalational injury	Transfusion of blood products
Reperfusion injury after lung transplantation	Uremia

5) ARDS can be a manifestation of ***The Severe Acute Respiratory Syndrome*** **(SARS).**

The SARS virus is a corona virus that destroys the type II pneumocytes
causes diffuse alveolar damage.

6) ***ARDS* is initiated by**
damage to alveolar capillary endothelium and alveolar epithelium
influenced by the following pathogenic factors:

(a) Neutrophils release substances toxic to the alveolar wall.
(b) Activation of the coagulation cascade is suggested by the presence of **microemboli.**
(c) Oxygen toxicity is mediated by the formation of oxygen-derived free radicals.



<< With previous page (ARDS)

NEONATAL RESPIRATORY DISTRESS SYNDROME (HYALINE 'MEMBRANE DISEASE)

General Considerations

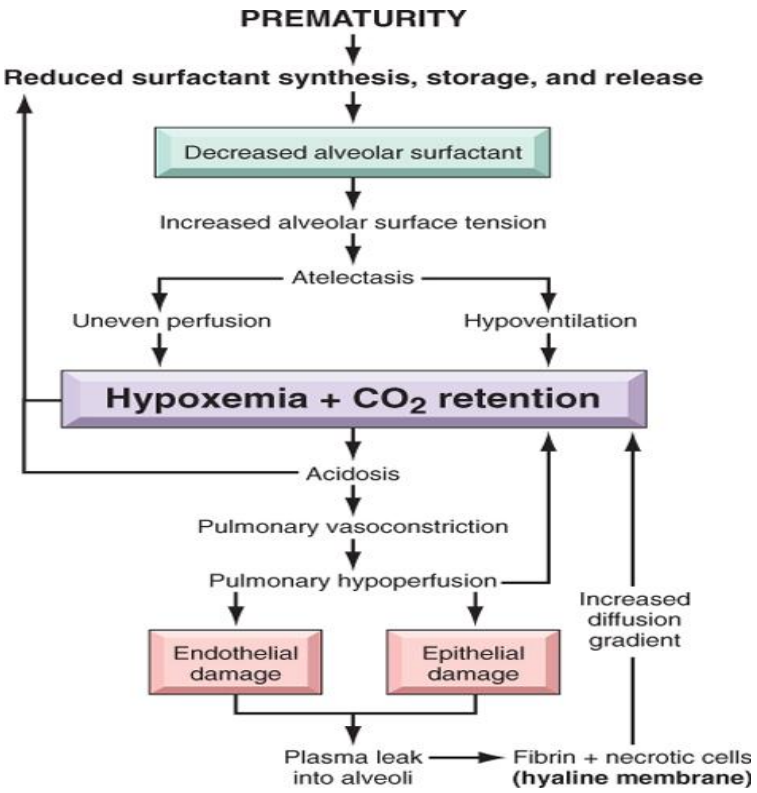
- It is the most common cause of ***respiratory failure*** in the newborn
- It is the most common cause of ***death*** in premature infants.

It is ***marked by:*** dyspnea - cyanosis - tachypnea (high breath rate) →WHEN? Shortly after birth**.**

It ***results from:*** a deficiency of surfactant, most often as a result of immaturity.

Predisposing factors:

- Prematurity.
- Maternal diabetes mellitus and delivery by cesarean section

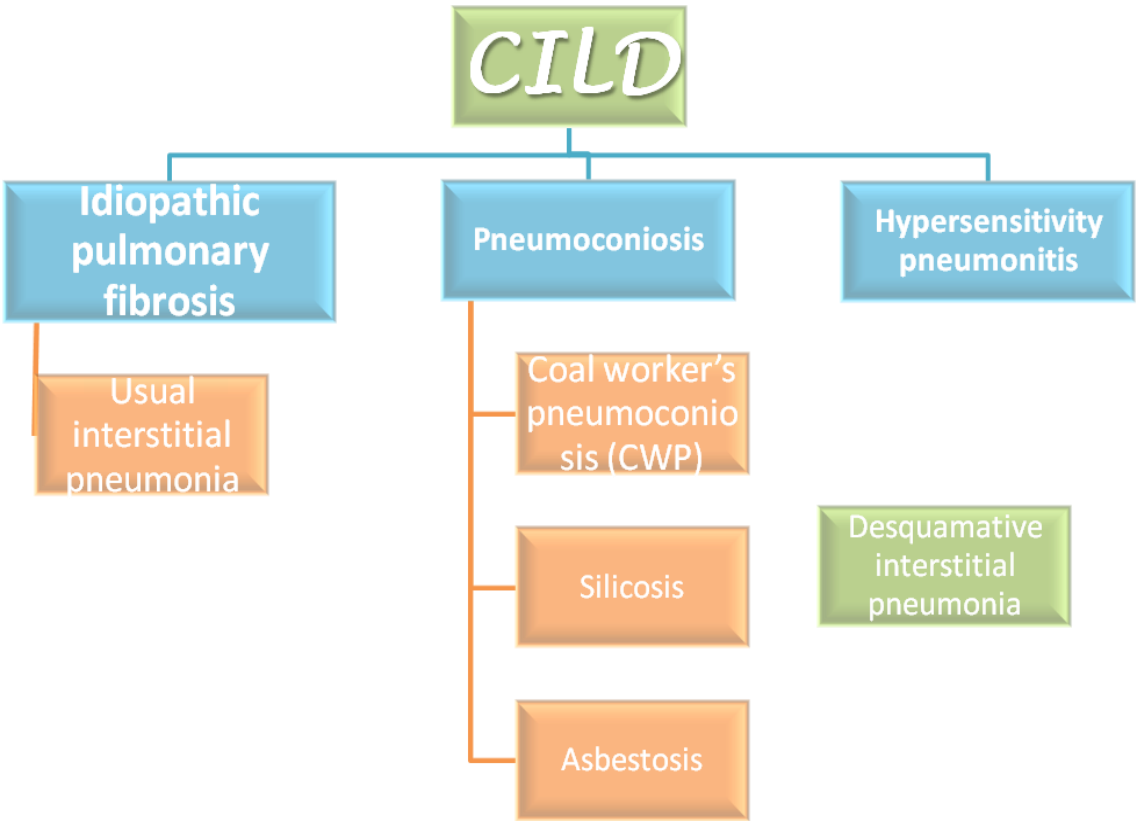


INTERSTITIAL LUNG DISEASE

Selected Examples of Interstitial Lung Disease

Disorder	Description
Hypersensitivity Pneumonitis (Extrinsic Allergic Alveolitis)	Interstitial Pneumonia caused by inhalation of various antigenic substances *Exemplified By* inhalation of spores of ***Thermophili Actinomycetes*** from *moldy hay* → **causing "farmer's lung"**
Goodpastures Dynrome	Hemorrhagic Pneumonitis and Glomerulonephritis caused by antibodies directed against *glomerular basement membranes* .
Eosinophilic Granuloma	Proliferation of histocytic cells ((Related to *Langerhan's cells* of the skin.))
Idiopathic Pulmonary Fibrosis	Immune complex disease with progressive fibrosis of the alveolar wall.
Sarcoidosis	***Granulomatous*** disorder of unknown etiology.

CHRONIC INTERSTITIAL LUNG DISEASE



PNEUMOCONIOSES:

- ➔ **Environmental diseases, caused by inhalation of inorganic duct particles.**
- ➔ **exemplified by (illustrated by) the following conditions:**

Acanthracosis

Caused By
inhalation of carbon dust --- it is endemic (spread) in urban areas and causes no harm.

Characterized by
carbon-carrying macrophages → it results in irregular black patches visible on gross inspection.

COAL WORKER'S PNEUMOCONIOSIS

Caused By inhalation of **coal dust** ((which contains both carbon and silica)).
Types:
(a) Simple coal worker's pneumoconiosis:

- **Marked by** coal macules around the bronchioles,
  How are those formed ? by ingestion of coal dust particles by macrophages.
-
- In most cases, it is inconsequential and produces no disability.

(b) Progressive massive fibrosis:

- **Marked by** fibrotic nodules filled with necrotic black fluid.
- **It can result in (it is a complication of)**
  bronchiectasis, pulmonary hypertension,
  or death from respiratory failure or right-sided heart failure.

SILICOSIS

A chronic occupational lung disease **caused by** exposure to ***free silica dust***
Seen in miners (men who work at mines), glass manufacturers and stone cutters.

➢ In the **Gulf region** and **in"desert climate",** it could be due to inhalation of sand.

(a) **Initiated By** ingestion of silica dust by alveolar macrophages
→ damage to macrophages → initiates an inflammatory response
mediated by lysosomal enzymes and various chemical mediators.

(b) Silicotic nodules that enlarge and eventually obstruct the airways and blood vessels are characteristic.

(c) **Associated With** increased susceptibility to **tuberculosis**;
the frequent concurrence ((meeting of both diseases)) is referred to as **silicotuberculosis**.

ASBESTOSIS

Caused By inhalation of asbestos fibers.

(a) **Initiated By** uptake or asbestos fibers by alveolar macrophages
→ A fibroblastic response occurs
((probably from release of fibroblast-stimulation growth factors by macrophages))
→ leads to diffuse interstitial fibrosis mainly in the lower lobes.

(b) **Characterized By** the presence of
- ***Ferruginous Bodies:*** are yellow-brown, rod-shaped bodies with clubbed ends
  stain positively with **Prussian blue**
  arising from iron and protein coating on asbestos fibers.

- ***Dense hyalinized fibrocalcific plaques*** of the parietal pleura.

(c) **Results In**
marked predisposition **TO**
*Bronchogenic Carcinoma **AND** Malignant Mesothelioma of The Pleura Or Peritoneum.*

((Cigarette smoking further increases the risk of bronchogenic carcinoma.))