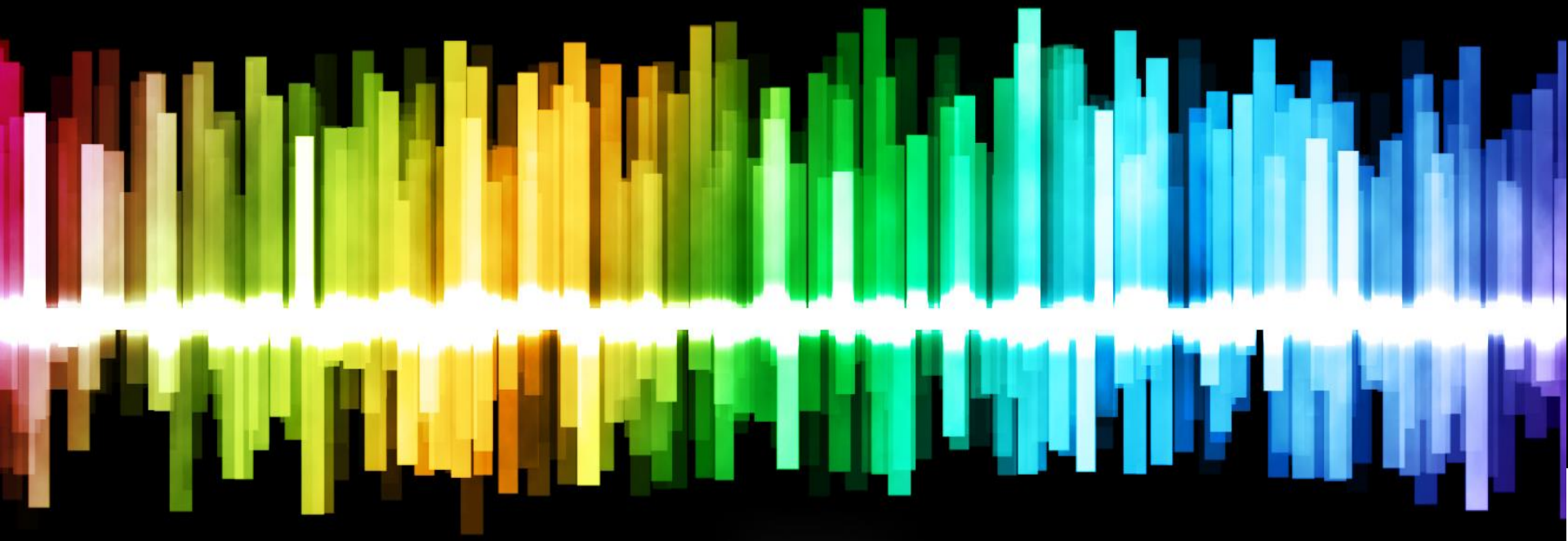
PATHOLOGY
TEAM 433

Lecture3:
Pathology of RESTRICTIVE LUNG DISEASES



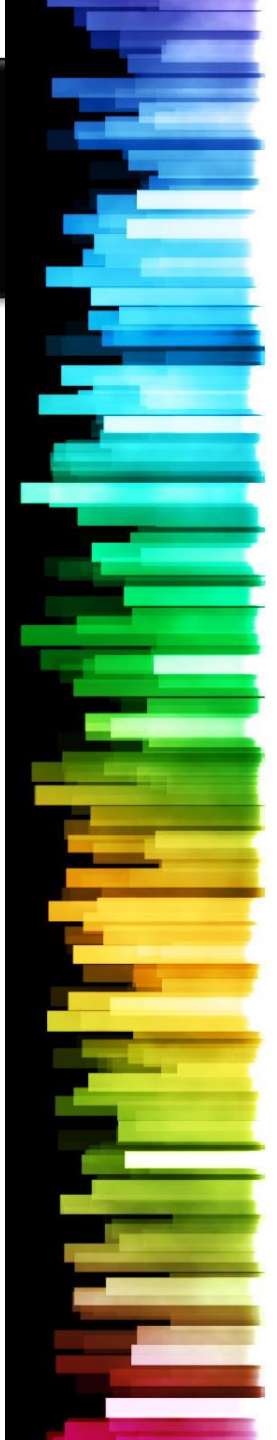
Objectives

- **at the end of this lecture , the student should able to:**

A) Understand the structure and constituents of the lung interstitial as well as the restrictive changes which occur in these diseases and lead to the development of symptoms of progressive breathlessness and cough in affected patients .

B) Appreciate the pathogenesis of interstitial lung diseases regardless of their type. this pathogenesis include the influx of inflammatory cells into the alveoli and alveolar walls, distortion of the normal structure of alveoli, release of chemical mediators and promotion of fibrosis (honey-comb lung).

C) Become aware of the classification of interstitial lung diseases.



Definition of RESTRICTIVE PULMONARY DISEASES *(interstitial) :*

- Are a category of diseases that restrict lung expansion, resulting in a decreased total lung capacity (lung volume).

The signs and symptoms for RESTRICTIVE PULMONARY DISEASES:

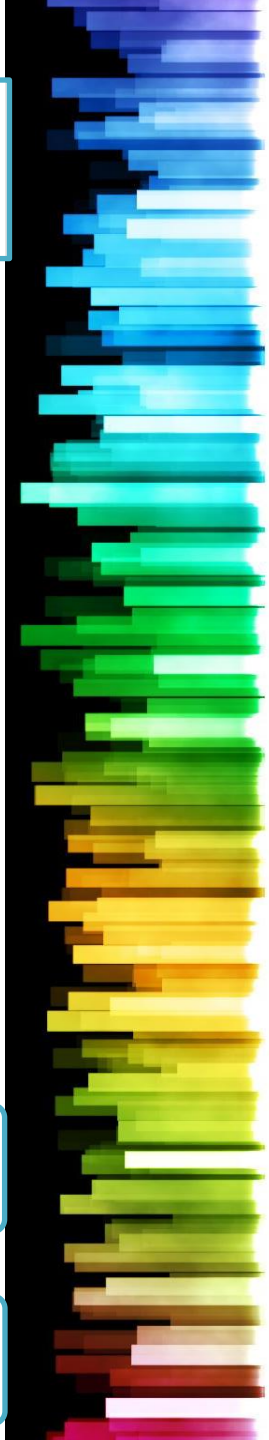
1-Dyspnea at rest and exertion

2- Persistent dry cough

3-Hypoxia and cyanosis (not sever as in chronic bronchitis and emphysema)

4- On auscultation, very sharp crackles sound usually in the **bases of lower lobes** can be heard. (respiratory crackles)

5- On spirometry : FEV1 is reduced, FVC is more reduced , Ratio is normal higher than 80%



Extrinsic type of restrictive pulmonary diseases :

Caused by :

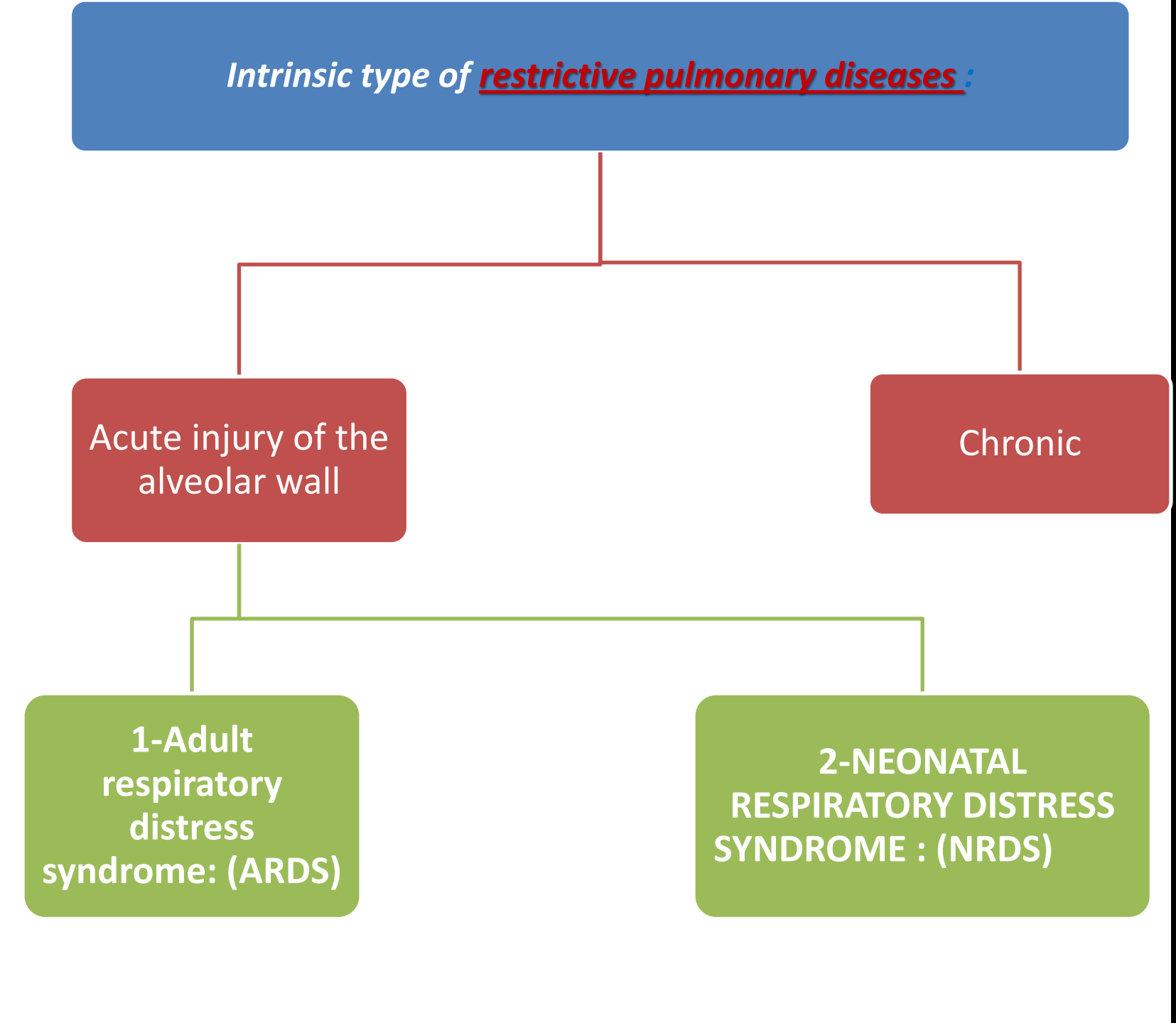
2.Neuromuscular disease:

- Neurological diseases such as: Guillain–Barré syndrome*
- Weakness in the intercostal muscles or paralysis of diaphragm

1.Chest deformity:

- e.g: kyphosciliosis.

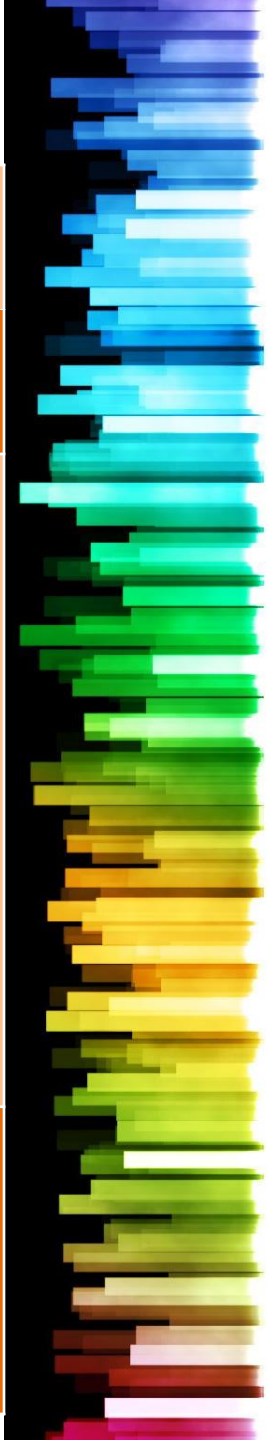
* viral infection leading to neuropathy affecting limbs muscles, in advanced cases the neuromuscular muscles may be affected leading to Restrictive lung disease.


Intrinsic type of restrictive pulmonary diseases :
Acute injury of the alveolar wall
Chronic
1-Adult respiratory distress syndrome: (ARDS)
2-NEONATAL RESPIRATORY DISTRESS SYNDROME : (NRDS)

1) ADULT RESPIRATORY DISTRESS SYNDROME (ARDS)

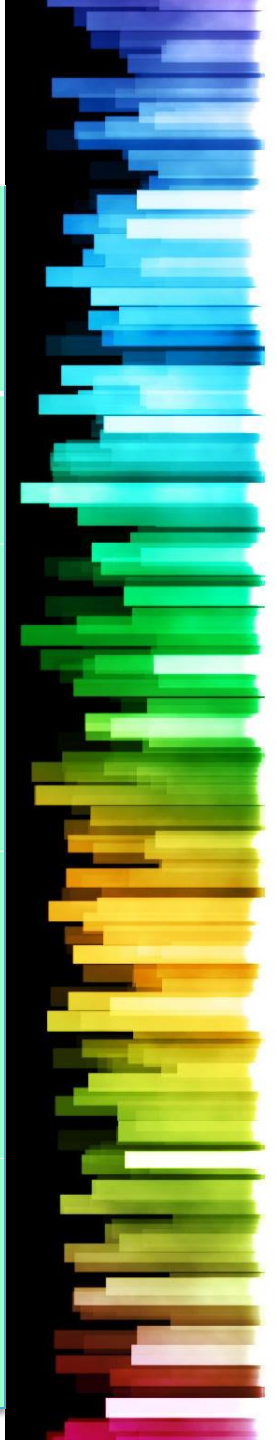
Or diffused alveolar damage(DAD)

Characteristics	**hyaline membrane** composed of fibrin and cellular debris due to injury to endothelial cells
produced by	diffuse **alveolar damage** with resultant increase in alveolar capillary permeability **(edema)**
Causes	Direct injury to lung: - Post anesthesia (aspiration of **gastric contents**) - inhalation of **chemical irritants** such as chlorine -**Treatment with O2:** Because O2 can lead to formation of free radicals which form superoxide (O-) **- Indirect injury to lung:** severe trauma, Sepsis, Shock, acute pancreatitis, uremia overdose with street drugs such as heroin, therapeutic drugs such as bleomycin.
Pathogenesis	1- Injury to lung. 2-Damage to alveolar lining cell specially type I pneumocytes + damage to alveolar capillary endothelium 3-Inflammatory reactions which lead to interstitial edema. 4-hyaline membrane lining alveoli.



2) NEONATAL RESPIRATORY DISTRESS SYNDROME : (NRDS) (HYALINE MEMBRANE DISEASE)

General considerations	most common cause of **respiratory failure in the newborn** and is the most common **cause of death in premature infants.**
marked by	dyspnea, cyanosis and tachypnea shortly after birth
Causes	✓**deficiency of** pulmonary **surfactants** in new borns, most often as a result of immaturity of type II pneumocyte . ✓**Stroke or brain hemorrhage** of the infant or if the infant **inhaled the amniotic fluid**
Predisposing factors:	✓Prematurity (less than 36 weeks). ✓Maternal diabetes mellitus ✓delivery by cesarean section ✓Amniotic fluid aspiration ✓multiple birth
Pathogenesis	results in diffuse alveolar damage with the development of a hyaline membrane lining the alveoli. (It is composed of fibrin and necrosis and interfere with gas exchange and inflammation), Immature type ll pneumocytes



Chronic restrictive lung disease:

Are a heterogeneous group of diseases, they have similar clinical signs, symptoms, radiographic alterations and pathophysiologic changes.

1- «تغبر الرئة»Pneumoconiosis: Occupational

Pneumoconiosis is a group of pulmonary diseases caused by chronic exposure to inorganic mineral dust inhalation and this leads to lung damage. They include **carbon dust, silica, asbestos**, beryllium etc.

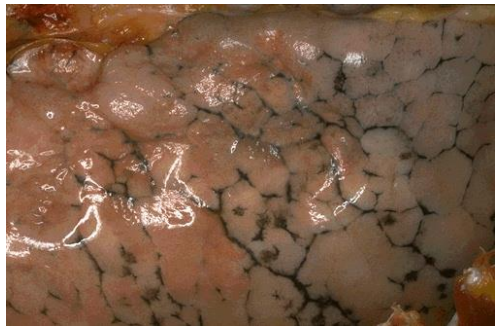
N.B: Dust particles can be either: organic or inorganic (metals).

The dust particles "between" 5-10 µm don't reach the alveoli because of the action of the cilia, and the dust that is less than 0.5 µm is very tiny dust cann't cause a disease, so the dust has to be from 1-5 µm to cause an interstitial lung disease.

- Anthracosis:

is the asymptomatic accumulation of coal pigment/carbon particles without consequent cellular reaction.

Inhaled coal dust enters the terminal bronchioles, and the carbon pigment is engulfed by alveolar and interstitial macrophages.



1.Coal worker's pneumoconiosis (CWP) :

Can be defined as the accumulation of coal dust in the lungs and the tissue's reaction to its presence.

The disease is divided into 2 categories:

	1.Simple coal «الفحم» worker's pneumoconiosis	2. complicated coal worker's pneumoconiosis (CCWP), or Progressive massive fibrosis (fiberogenic)
General notes	✓Mostly in the upper zones of the lower and upper lobes of the lungs. ✓ Patients have slight cough and blackish sputum	✓Occurs after many years of underground mine work. ✓Miners who have rheumatoid arthritis and PMF are called Caplan's syndrome. ✓ Patients have cough, dyspnea, and lung function impairment.
Morphology	✓Focal aggregations of black coal dust-laden macrophages (coal molecules)1 to 5 µmare scattered through the lung. ✓Little or no fibrosis	✓marked by fibrotic nodules filled with necrotic black fluid ✓ Black scars exceed 2 cm in diameter some times up to 10 cm ✓ It consists of dense collagen and carbon pigments.
Picture		

2.Silicosis :

Silicosis is a fibro-nodular lung disease caused by long term exposure to inhalation of dust containing crystalline silica particles.

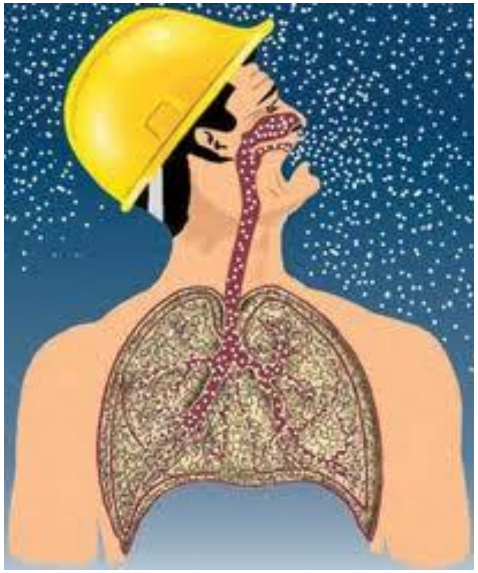
Encountered in industries: mining of gold, tin, copper and coal, sandblasting, metal grinding, ceramic manufacturing.

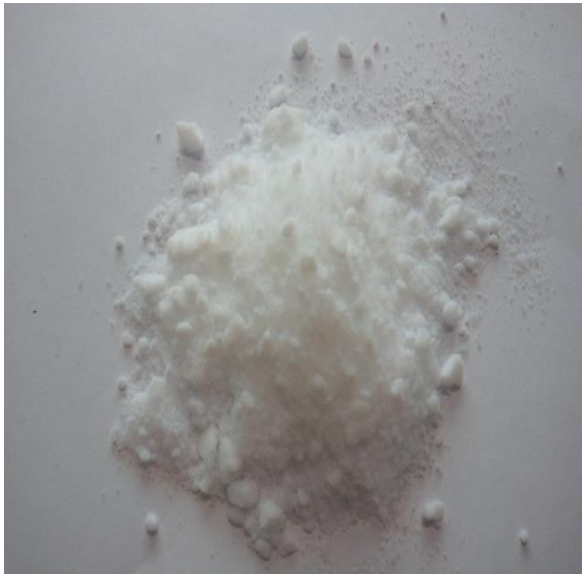
-Pathogenesis :

✓**A lot of carbon and calcified Silicotic nodules** (eggshell calcification)

✓**increased susceptibility to tuberculosis (silicotuberculosis)**

✓Chronic silicosis can lead to complicated progressive massive fibrosis or lung cancer.





3.Asbestosis:

is caused by inhalation of asbestos fibers (needle-shaped) are found in the insulation materials, workers who are exposed to these materials are susceptible to this disease.

Pathogenesis :

presence of **ferruginous bodies which are yellow-brown**

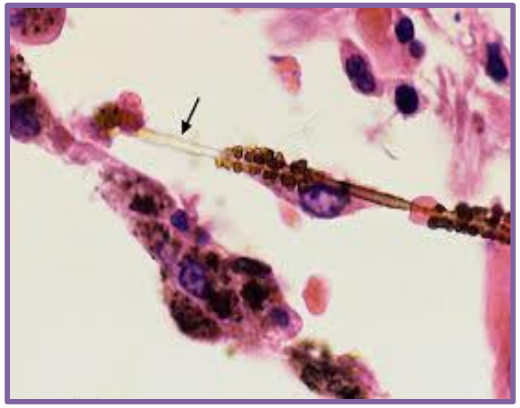
Asbestos may cause other disease:

1-Fibrous pleural block :

It prefers the periphery (pleura) causing area of fibrosis in the pleura and it is usually without symptoms.

2-Asbestosis (restrictive lung disease)

3-Mesothelioma (Lethal) Smokers have increased incidence of developing mesothelioma and bronchogenic carcinoma.



- Hypersensitivity pneumonitis:

An immunologically mediated inflammatory lung disease primarily affects the alveoli and is therefore often called **allergic alveolitis**.

1.Farmers lung :

Thermophilic actinomycetes in hay " يتحسس من عفن القش"

2.(Pigeon-breeder's lung Disease)

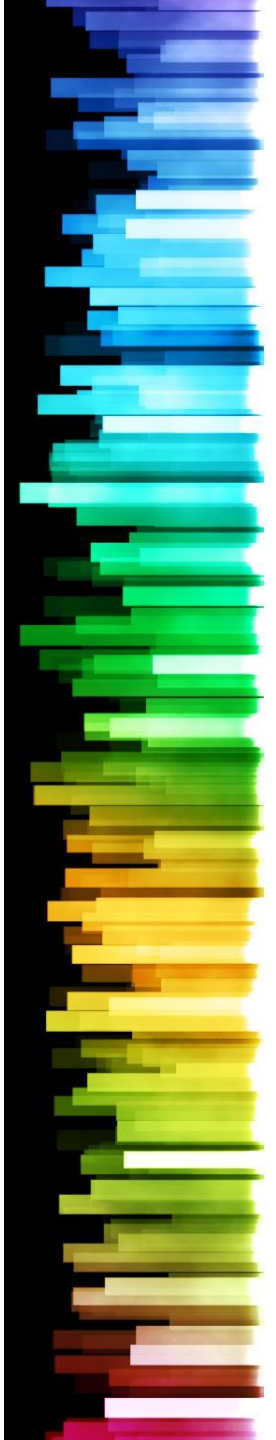
For the people who are dealing with birds and pigeon

The feces of these animals cause hypersensitivity pneumonitis

Idiopathic restrictive pulmonary disease:

1.Idiopathic Pulmonary Fibrosis (IPF), or usual interstitial pneumonia (UIP):

- ✓ Idiopathic pulmonary fibrosis (IPF) is progressive fibrosis disorder of unknown cause. It is an idiopathic interstitial pneumonia with diffuse interstitial fibrosis and inflammation.
- ✓ Live about 6 months to 2 years
- ✓ It has a poor prognosis. Respiratory and heart failure may develop within few years. No effective therapy is available for the treatment of idiopathic pulmonary fibrosis. Lung transplant is the only solution.

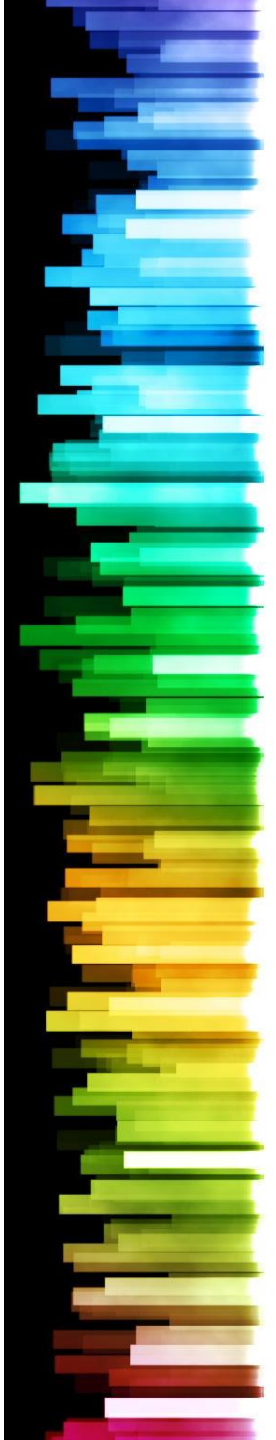


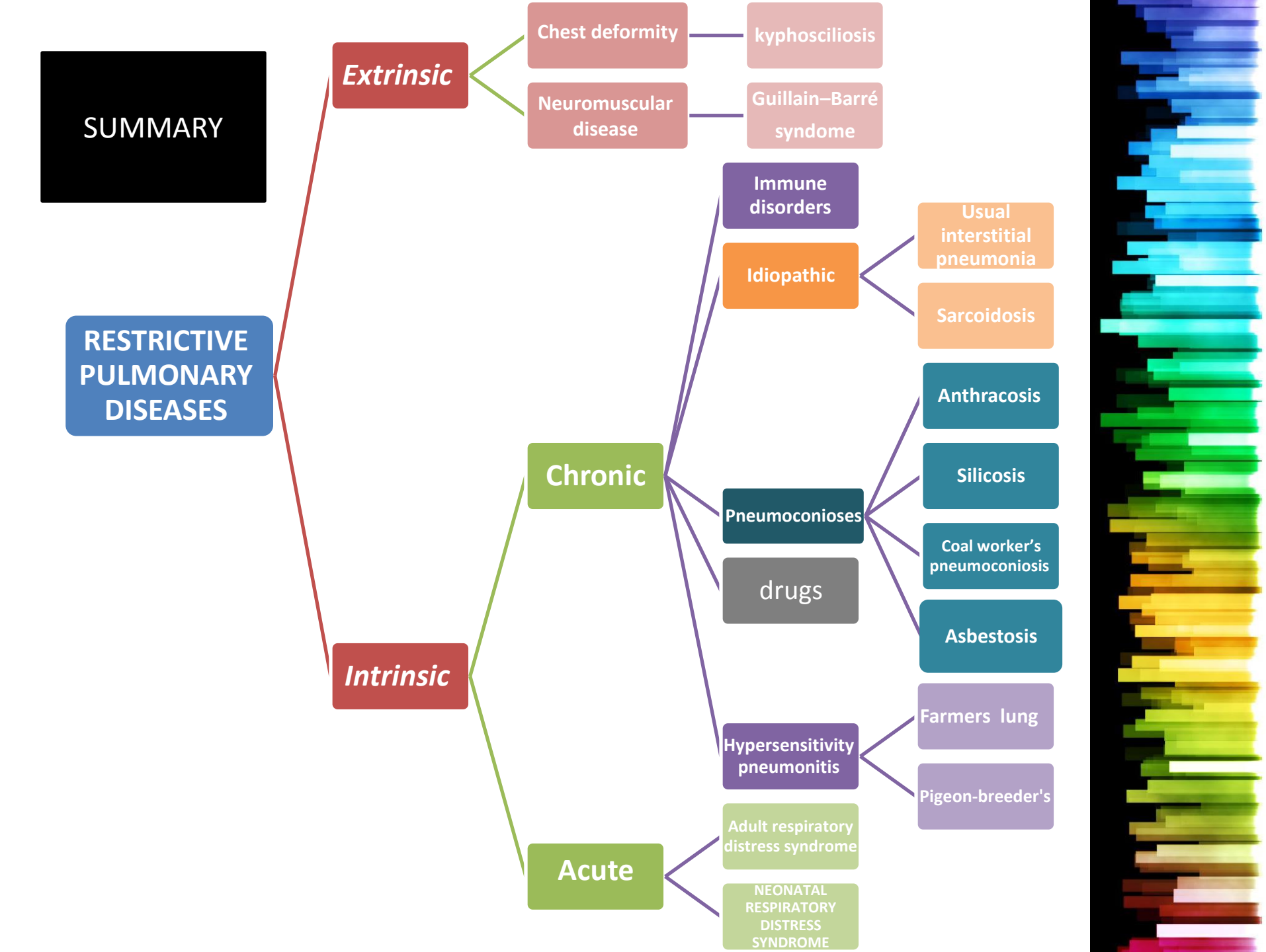
2.Sarcoidosis:

Sarcoidosis is a multisystem inflammatory disease of unknown cause/etiology that predominantly affects the lungs and intrathoracic lymph nodes.

- ✓ **Drugs associated disorders:**
- ✓ Such as berylliosis or Pulmonary fibrosis associated **Bleomyxin toxicity**

- ✓ **Immune disorders:**
- ✓ **SLE**
- ✓ ***goodpasture syndrome:*** anti-glomerular basement antibody disease attack the lungs and kidneys, leading to bleeding from the lungs and to kidney failure.
- ✓ **Eosinophilic granuloma** is proliferation of histiocytosis cells related to Langerhan's cells of the skin.




SUMMARY
RESTRICTIVE PULMONARY DISEASES
Extrinsic
Chest deformity
kyphosciliosis
Neuromuscular disease
Guillain–Barré syndome
Intrinsic
Chronic
Immune disorders
Idiopathic
Usual interstitial pneumonia
Sarcoidosis
Pneumoconioses
Anthracosis
Silicosis
Coal worker's pneumoconiosis
Asbestosis
drugs
Hypersensitivity pneumonitis
Farmers lung
Pigeon-breeder's
Acute
Adult respiratory distress syndrome
NEONATAL RESPIRATORY DISTRESS SYNDROME

Questions

1- How can we diagnose the severity of **RESTRICTIVE PULMONARY DISEASE**?
By asking him when did he get dyspnea 'at rest = very sever , after exercise = mild'

2- What are the results of fibrosis in **RESTRICTIVE PULMONARY DISEASE?**

A. Thickened of interstitium of alveoli.
B. interrupted of alveolar duct and alveoli
C. Shrinkage of lung (lung volume)

3- Hyaline membrane composed of:
(Fibrin + dead cells 'necrotic cells' +inflammatory cells)

4- Long form of "DAD" and other name for it:
Diffuse Alveolar Damage =ADULT RESPIRATORY DISTRESS SYNDROME (ARDS)

5- What is the cause of Edema in **RESTRICTIVE PULMONARY DISEASE?**
Inflammation ↑ membrane permeability cause edema

6- What are the disease under this category "**ACUTE INJURY OF ALVEOLAR WALL**"?

A. ADULT RESPIRATORY DISTRESS SYNDROME
B. NEONATAL RESPIRATORY DISTRESS SYNDROME

MCQs

1. Most common symptoms of RESTRICTIVE PULMONARY DISEASE:

a) Dyspnea
b) Productive cough
c) Dry cough
d) Both A+B
e) Both A+C

2. Honeycomb is the best describe of :

a) COPD.
b) Restrictive pulmonary disease
c) Both A+B
d) None of them

3. The size of dust that can cause disease:

a) <1µm
b) Between 1 to 5 µm
c) >5 µm
d) None of them

MCQs

4. Which one of these restrictive diseases has unknown etiology:

a) Silicosis

b) Asbestosis

c) Sarcoidosis

5. Which one of these diseases is an acute restrictive lung disease:

a) Adult respiratory distress syndrome

b) Farmers lung

c) Coal worker's pneumoconiosis

6. Which one of these diseases is found in the insulation workers:

a) Asbestosis

b) Anthracosis

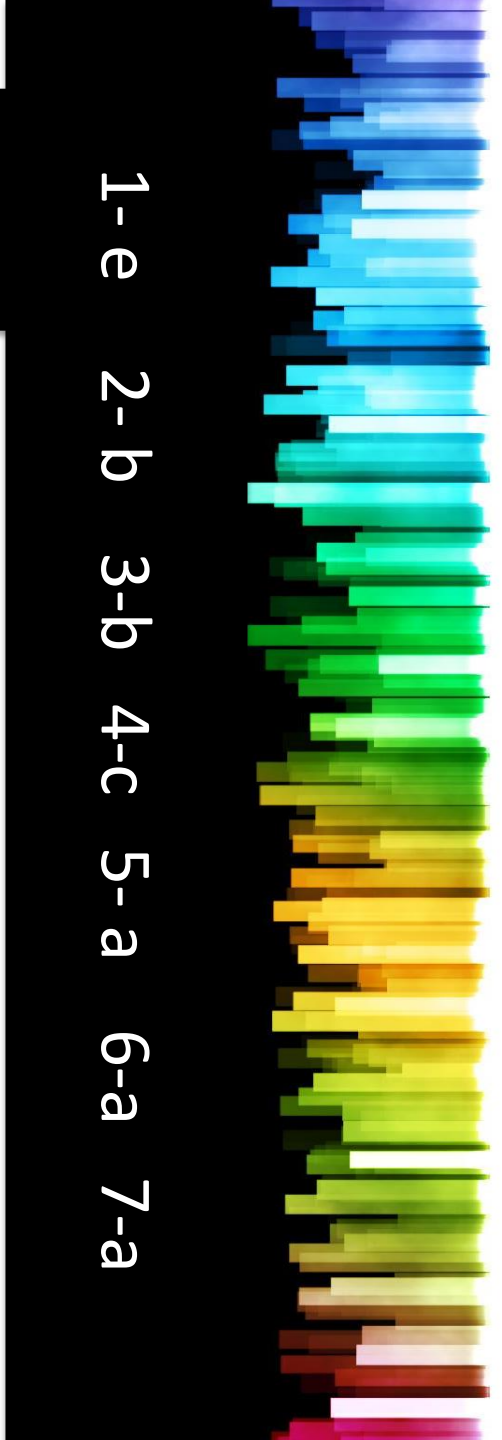
c) Silicosis

7. Which one of the following could be found in a normal person:

a) Anthracosis

b) Farmers lung

1- e 2- b 3-b 4-c 5- a 6-a 7-a



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https://www.youtube.com/watch?v=ASQMDrg2fNg&feature=youtube_gdata_player

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