

Interstitial lung disease

(Infiltrative or restrictive lung disease)

Definition

- Heterogeneous group of disorder
- Characterized by diffuse & chronic involvement of pulmonary connective tissue
- Involving the interstitium in the alveolar wall - (BM, endothelium, epithelium, collagen, fiber, elastic tissue, proteoglycan, fibroblast, mast cell & lymphocyte)
- Restrictive lung disease

Interstitial lung diseases

Fibrosing -

- Interstitial pneumonia (interstitial pulmonary fibrosis)
- Cryptogenic organizing pneumonia
- Associated with collagen vascular pneumonia
- ***Pneumoconiosis***
- Drug reaction
- Radiation pneumonitis

Granulomatous –

- Sarcoidosis
- Hypersensitive pneumonia

Eosinophilic

Smoking related –

- Desquamative pneumonia
- Respiratory bronchitis associated ILD

Pathogenesis

- Alveolitis – collection of inflammatory & immune cells in alveolar space.
- Leukocytes accumulation – distort alveolar structure and release mediators - parenchymal injury & stimulate fibrosis
- End stage fibrotic lung disease – air space is replaced by cystic space separated by thick fibrotic band and inflammatory cells (honey comb)

Clinical and radiological findings

- Dyspnea, tachypnea, end inspiratory crackles, cyanosis,
- Reduction in CO diffusing capacity, lung vol & compliance
- CXR – diffuse infiltration by small nodules, irregular lines or ground glass shadow
- Secondary pul. HT, RHF – cor – pulmonale
- Scarring of lung - end stage lung or honey comb lung

Pneumoconiosis

(An entity of ILD)

Definition

- Group of non-neoplastic lung disorders
- Result from the inhalation of
 - Inorganic dust
 - Mineral
 - Organic dusts
 - Chemical fumes
 - Vapours

Classification

- Pneumoconiosis is usually divided into three groups:
 - Major pneumoconiosis
 - Minor pneumoconiosis
 - Benign pneumoconiosis

Major Pneumoconiosis

- Inhalation of some dusts results in “major fibrosis” of the lungs, which results in interference of lung architecture or lung function tests.

- Silica → silicosis
- Asbestos → asbestosis
- Talc → talcosis
- Coal → coal workers
pneumoconiosis
(anthracosis)

- Cane fibers – Bagassosis
- Cotton dust - Byssinosis (In Textile industries)
- Grain dust – farmer's lung

Minor Pneumoconiosis

- Inhalation of some dusts results in “minor fibrosis” of the lungs
- There is minimal fibrosis of the lungs without interference of lung architecture or lung function tests.
- These dusts include:
 - Clay
 - Koalin
 - Mica
 - Feldspar (non-fibrous silicates)

- Major pneumoconiosis
and
- Minor pneumoconiosis
are called

“ Fibrotic Pneumoconiosis ”

Benign Pneumoconiosis

- No reaction in the lungs
- Inhalation of dust
- Deposition casts a shadow in x-ray of the lung.
- There is no fibrosis
- No disturbance of lung functions.

Examples of Benign Pneumoconiosis

- Iron dust → siderosis
 - Tin dust → stannosis
 - Calcium dust → chalicosis
-
- Characterized by the presence of small rounded dense opacities on a chest film due to perivascular collections of dusts.
 - Hilar lymph nodes may be prominent.
 - Deposits in the lung disappear when exposure is discontinued.

Etiology

Etiologic Determinants

- Size of inhaled particle
 - ***0.3 and 0.5 μm reach the alveoli (Imp to note)***
- Chemical nature of the particle
- Concentration of the particle
- Length of exposure
- Effectiveness of clearing mechanism
- Cigarette smoking
- The individual's susceptibility

Deposition of particles in the body

- Larger particles are trapped in the mucous of the airways and moved out of the lung by the action of cilia, the “mucociliary escalator”
 - **>10 μm particles are deposited in the upper respiratory tract (nose and throat)**
 - **5-10 μm particles are deposited in the airways of the lung**
 - **<5 μm particles are deposited in the respiratory bronchioles and alveoli**
 - **Removed by macrophages (white blood cells)**

Etiology

Asbestosis

- Acoustic products
- Automobile undercoating
- Brake lining
- Cements
- Clutch casings
- Floor tiles
- Fire-fighting suits
- Fireproof paints
- Insulation
- Roofing materials
- Ropes
- Steam pipe material

Etiology

Coal Worker's Pneumoconiosis

- The deposition and accumulation of large amounts of coal dust cause what is known as coal worker's pneumoconiosis (CWP)
- Also called:
 - Coal miner's lung
 - Black lung
 - Black phthisis
 - Miner's phthisis

Etiology Silicosis

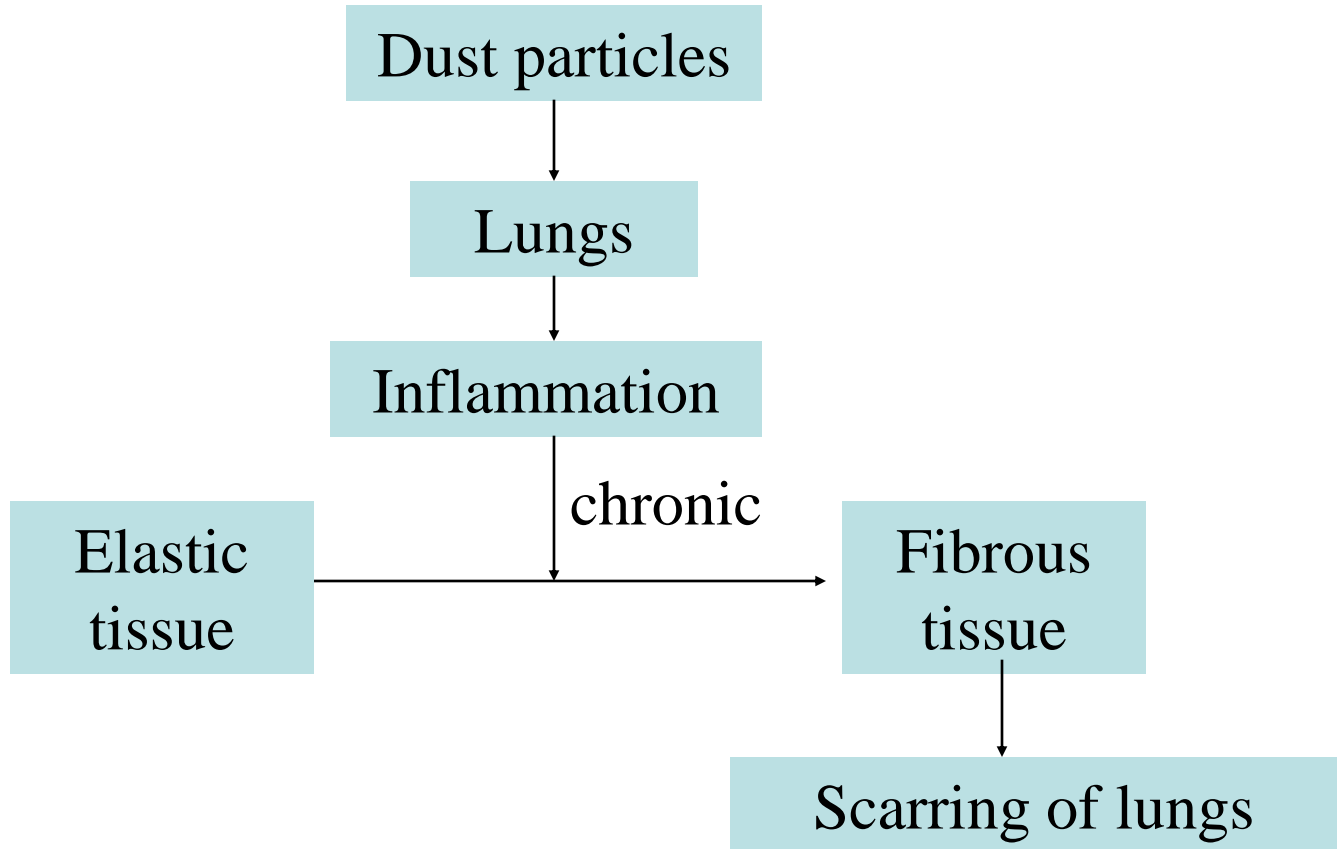
- Tunneling
- Hard-rock mining
- Sandblasting
- Quarrying
- Stonecutting
- Foundry work
- Ceramics work
- Abrasive work
- Brick making
- Paint making
- Polishing
- Stone drilling
- Well drilling

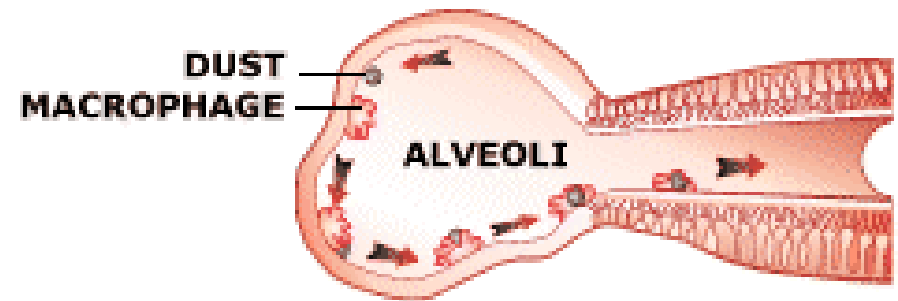
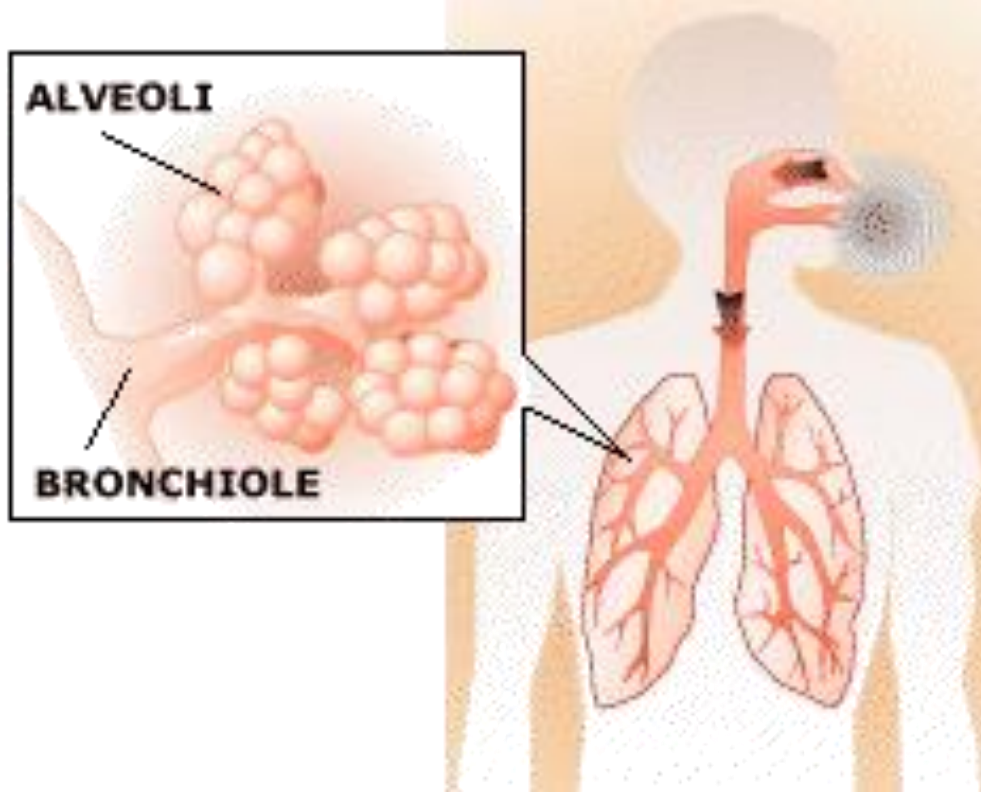
Etiology

Berylliosis

- Beryllium is a steel-gray, lightweight metal found in:
 - Certain plastics and ceramics
 - Rocket fuels
 - X-ray

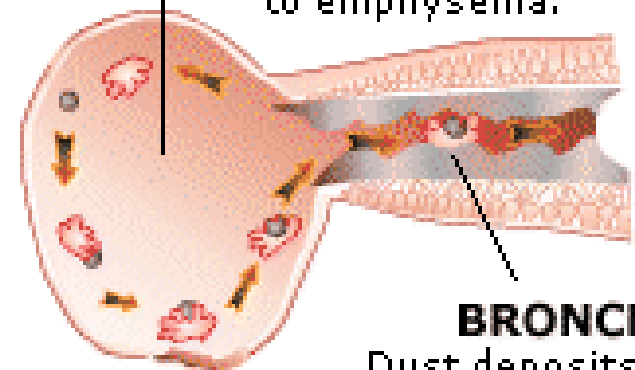
Pathogenesis





ALVEOLI

Years of cleaning out dust deposits cause the alveoli walls to become weaker and less elastic. This leads to emphysema.



Dust deposits lead to scarring and inflammation, which clogs passageways, obstructing airflow and causing chronic bronchitis.

Pneumoconiosis

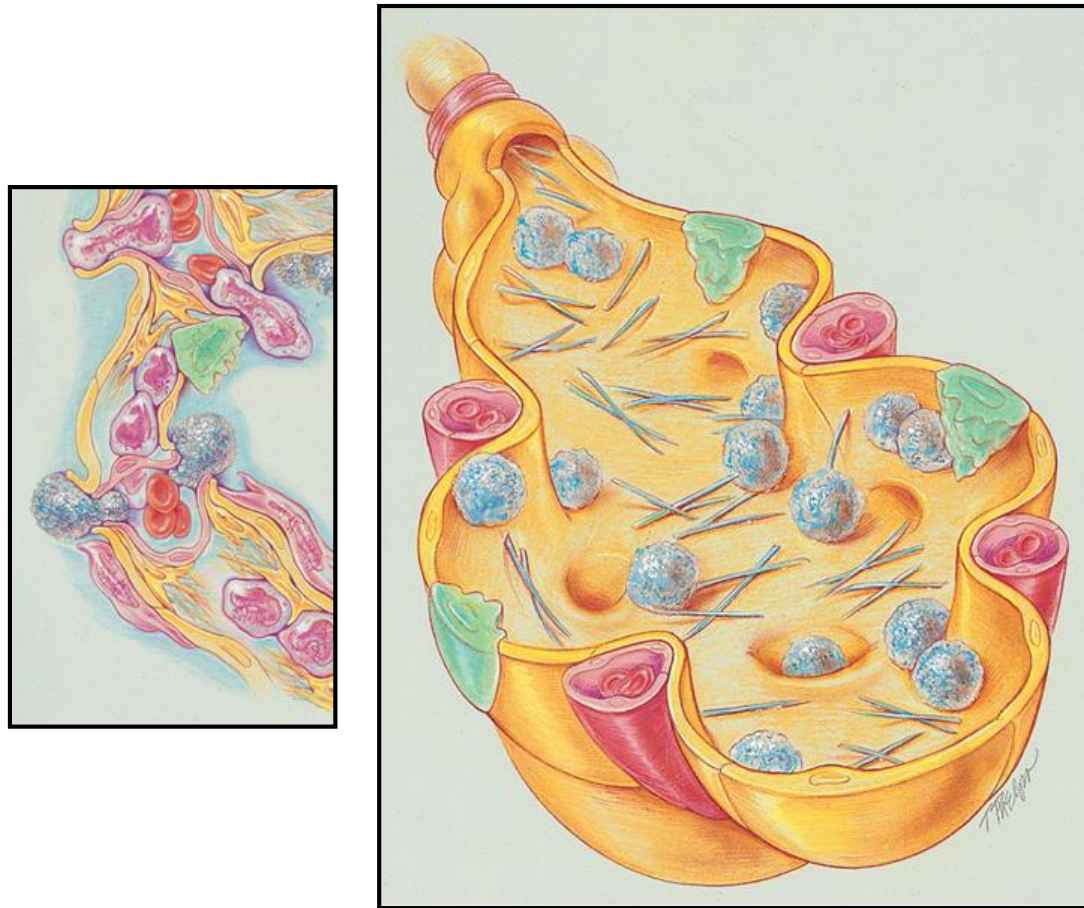


Figure 25-1. Pneumoconiosis, illustrated here in a case of asbestosis (close-up of one alveolar unit). *AF*, Asbestos fiber; *FIB*, fibrosis; *M*, macrophage. *Inset*, Cross-section showing fibrotic thickening of the alveolus, a common secondary anatomic alteration of the lungs.

Anatomic Alterations of the Lungs

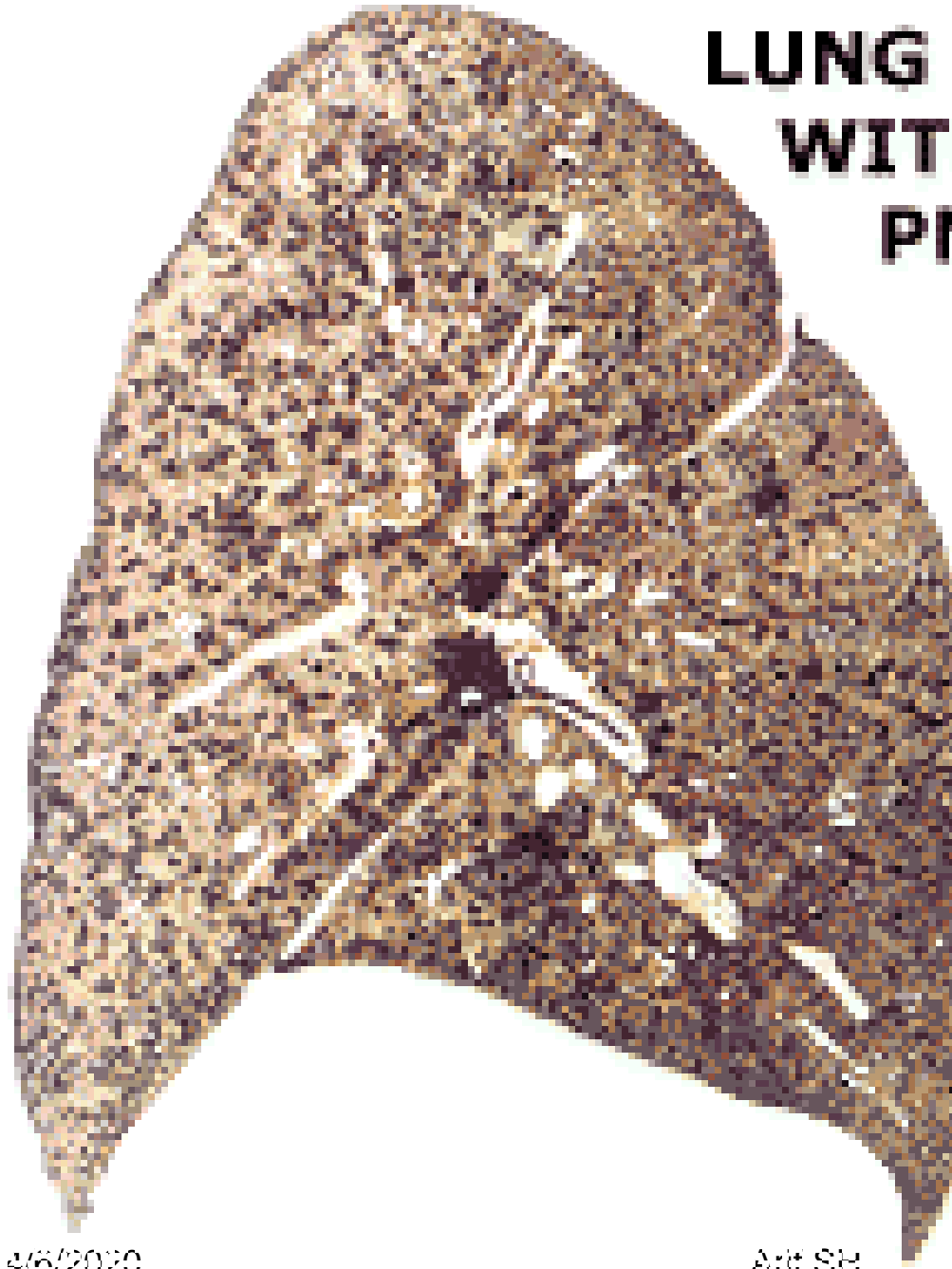
- Destruction of the alveoli and adjacent pulmonary capillaries
- Fibrotic thickening of the respiratory bronchioles, alveolar ducts, and alveoli
- Cystlike structures (honeycomb appearance)
- Fibrocalcific pleural plaques (e.g., asbestosis)
- Airway obstruction caused by inflammation and excessive bronchial secretions
- Bronchospasm
- Bronchogenic carcinoma
- Mesothelioma (in asbestosis)

Mechanism of injury

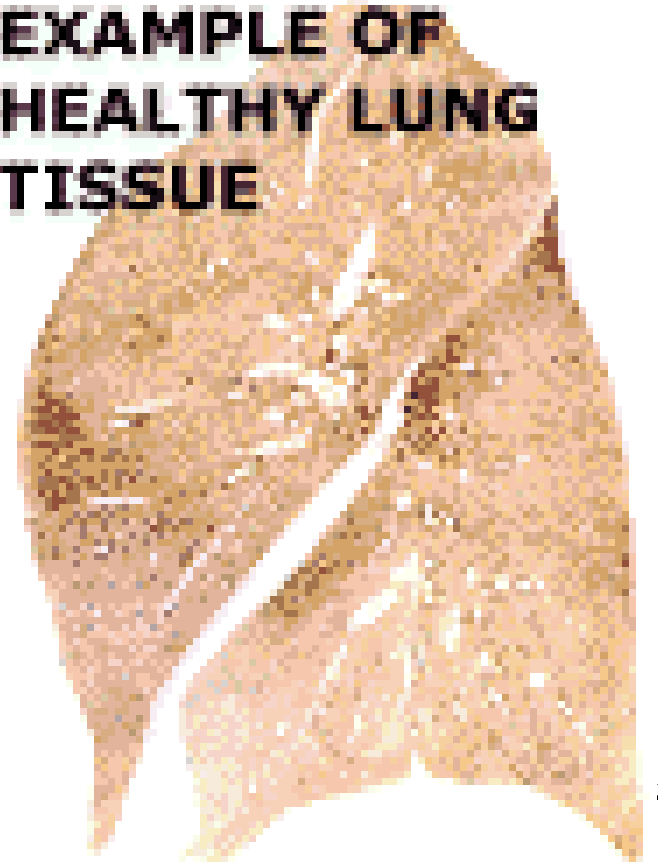
- Direct – injury by free radicals present on the particle – membrane damage causing cell death. Trigger macrophages to release mediators causing inflammatory response – fibroblast proliferation collagen deposition (silica & asbestos)

- Indirect – may be taken by epithelial cell or cross epithelial cells lining and reach to fibroblast and macrophages – local reaction – inflammatory response – fibroblast proliferation – interstitial fibrosis (asbestos)
- Cigarette smoke worsen the situation

LUNG TISSUE WITH TYPICAL PNEUMOCONIOSIS



**EXAMPLE OF
HEALTHY LUNG
TISSUE**



4/6/2020

ART SH

<http://www.courier-journal.com/dust/>

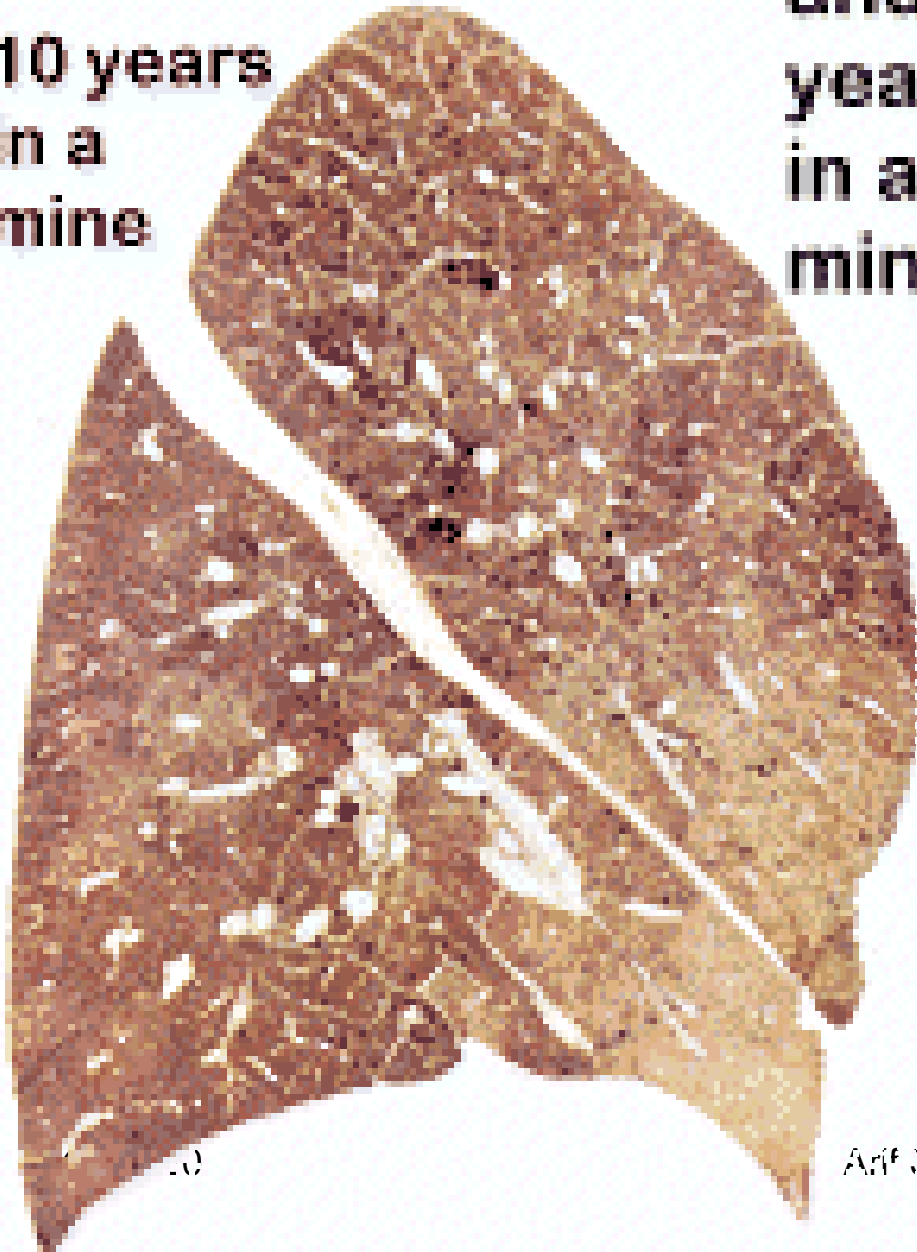
- CWP
- Asbestos related disease
- silicosis

Coal worker pneumoconiosis (CWP)

- Occurs in 5% of coal workers
- **The CXR in these patients is characterized by the presence of Reticulonodular pattern**
- Nodules up to 1 cm (**could progressively get larger in size, reaching up to 1 cm in diameter**).
- Progressive massive fibrosis (**eventually, there will be fibrosis and formation of large nodules and this stage is called progressive massive fibrosis**).

Black Lung

10 years
in a
mine



Between 20
and 30
years
in a
mine





4/6/2020

Simple CWP
Arif, SM

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Gross specimen demonstrating

progressive massive fibrosis in a coal miner

Anthracosis

- A benign deposition of coal dust within the lungs from inhalation of dirty air.



Coal workers' pneumoconiosis

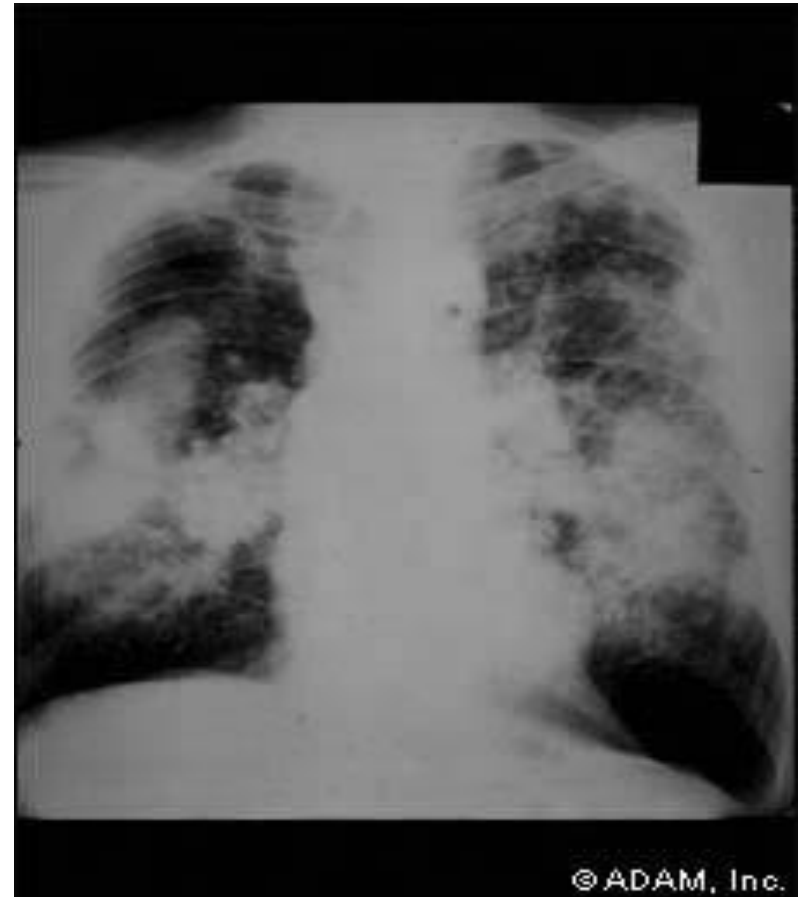
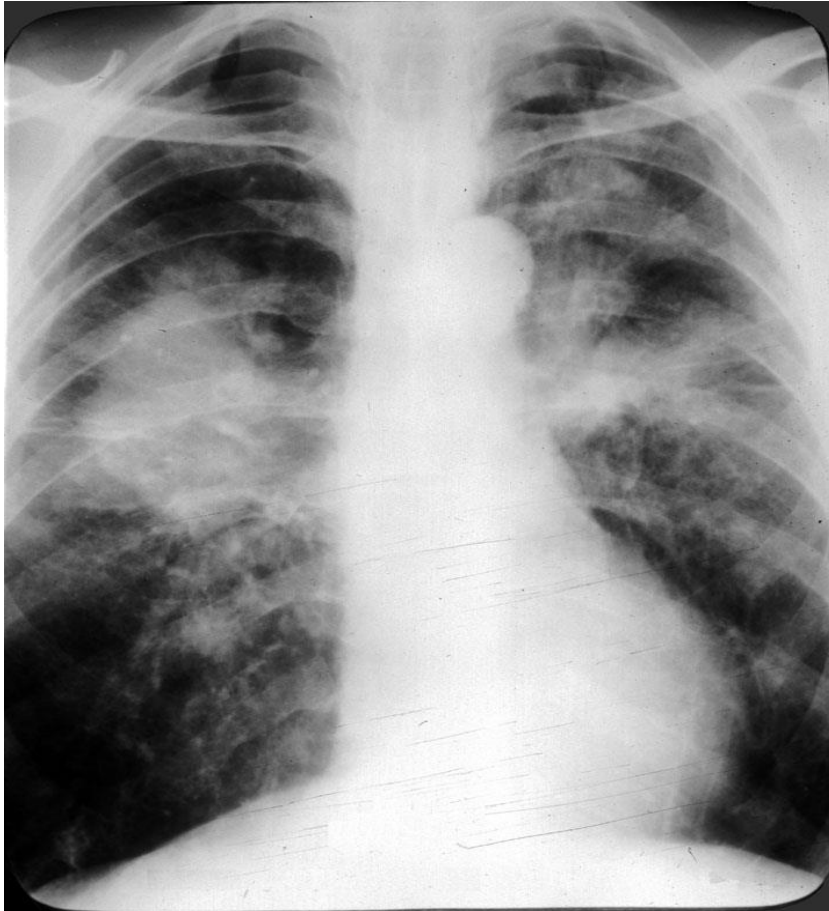
- Spectrum of findings in coal workers
- Asymptomatic anthracosis
- Simple CWP (little pulmonary dysfunction) - slight fibrosis, non-progressive

- **Progressive massive fibrosis**
(lung function compromised) -
fibrous nodules (up to 2 cm) -
sometimes coalesce - "black
lung", central necrosis

- clinically - breathlessness, cough
- <10% of CWP progress to PMF
- sometimes associated with rheumatoid arthritis - **Caplan's syndrome** (rapidly developing nodular pulmonary lesions)

CXR - CWP

Reticulo-nodular pattern on Xray



Morphology - Anthracosis

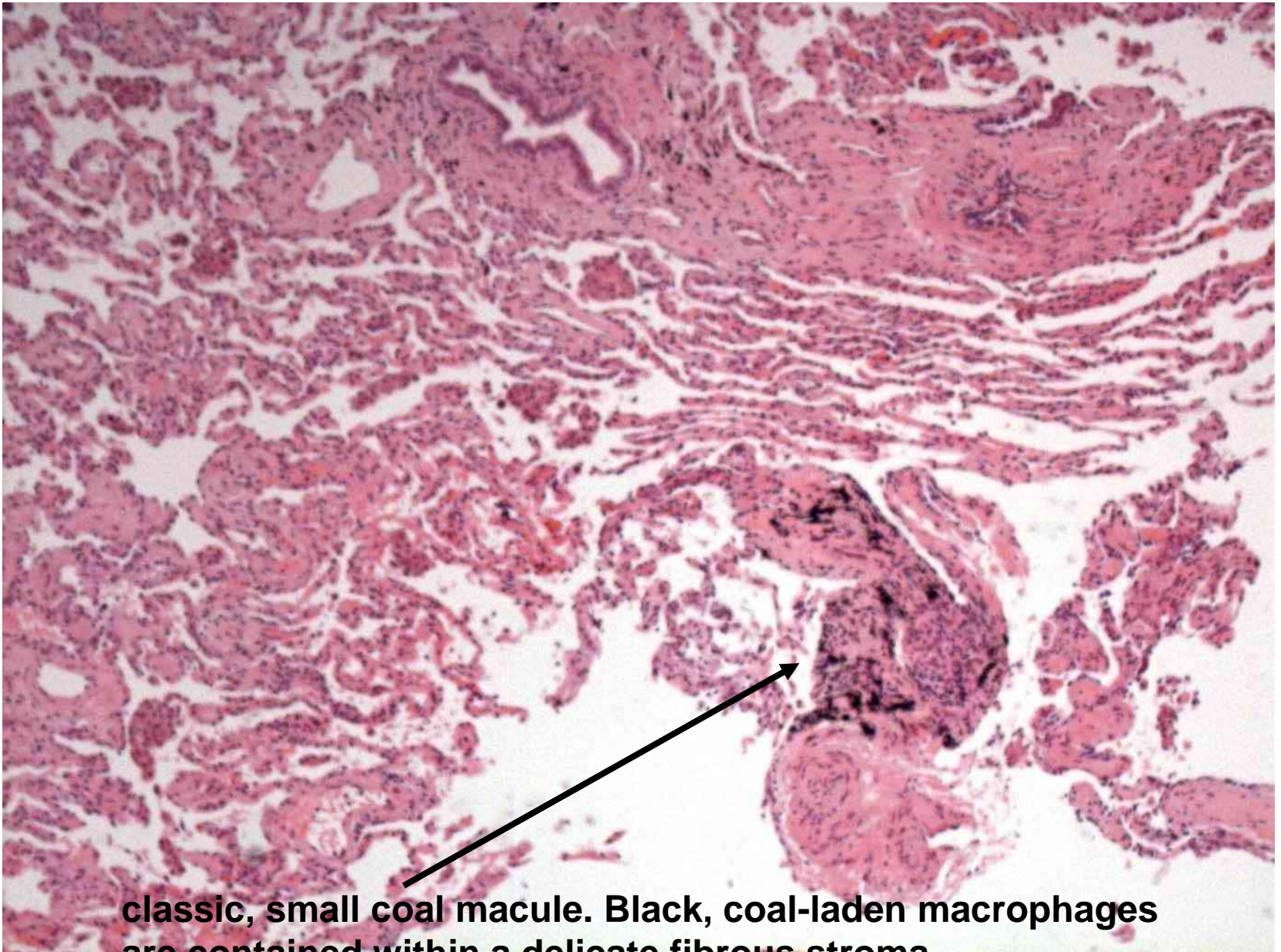
- Most innocuous lesion
- Commonly seen in urban population other than coal worker and cigarette smoker
- Carbon particles are engulfed by macrophages
- Accumulated in the connective tissue along the lymphatics

Morphology – simple CWP

- Characterized by coal macules 1 – 2 mm in size or coal nodules (1 cm)
- Macules – carbon laden macrophage
- Nodules – carbon laden macrophages and collagen fibres
- Upper lobe or upper zone of lower lobe
- Primarily adjacent to respiratory bronchioles

Morphology – complicated CWP (PMF)

- Intensely black scar - 2 - 10 cm in size
- Usually multiple
- M/E – dense collagen and pigments, centre is necrotic due to local ischemia
- Only 10% - develops pul dysfunction, pul HT, cor pulmonale
- Once develop – it become progressive even dust ceases



classic, small coal macule. Black, coal-laden macrophages are contained within a delicate fibrous stroma.

4/6/2020

There is surrounding emphysema, consistent with the history of smoking.

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