

Interstitial Lung Disease

The Pleura and Chest Wall

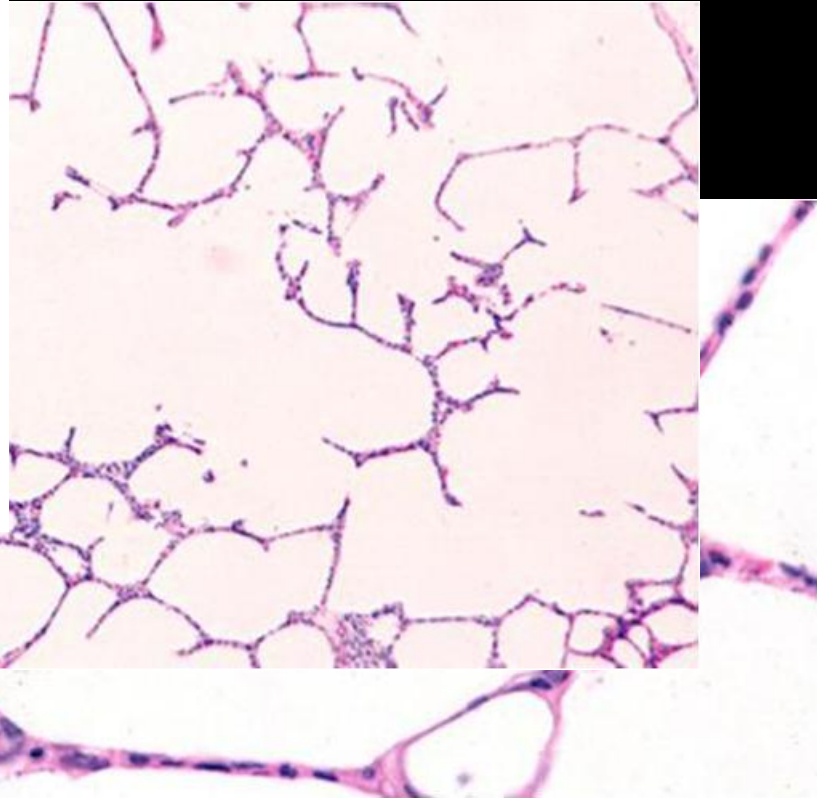
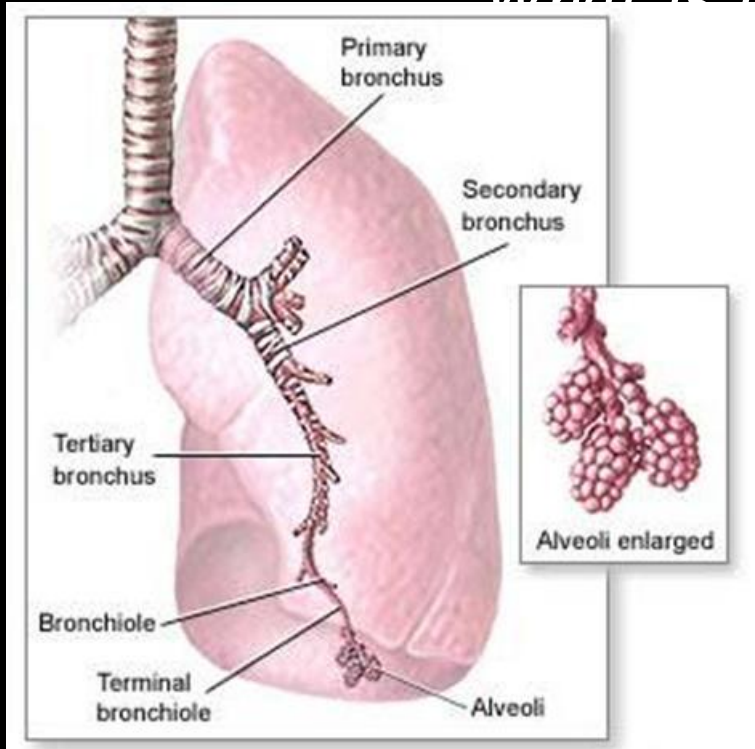
Objectives

- Interstitium
- Pleural disease
- Chest wall disease

Interstitial disease

- What is the interstitium?
- What does the interstitium do?
- What are the pathophysiological effects of interstitial disease?
- What are the clinical manifestations?

What is the interstitium?



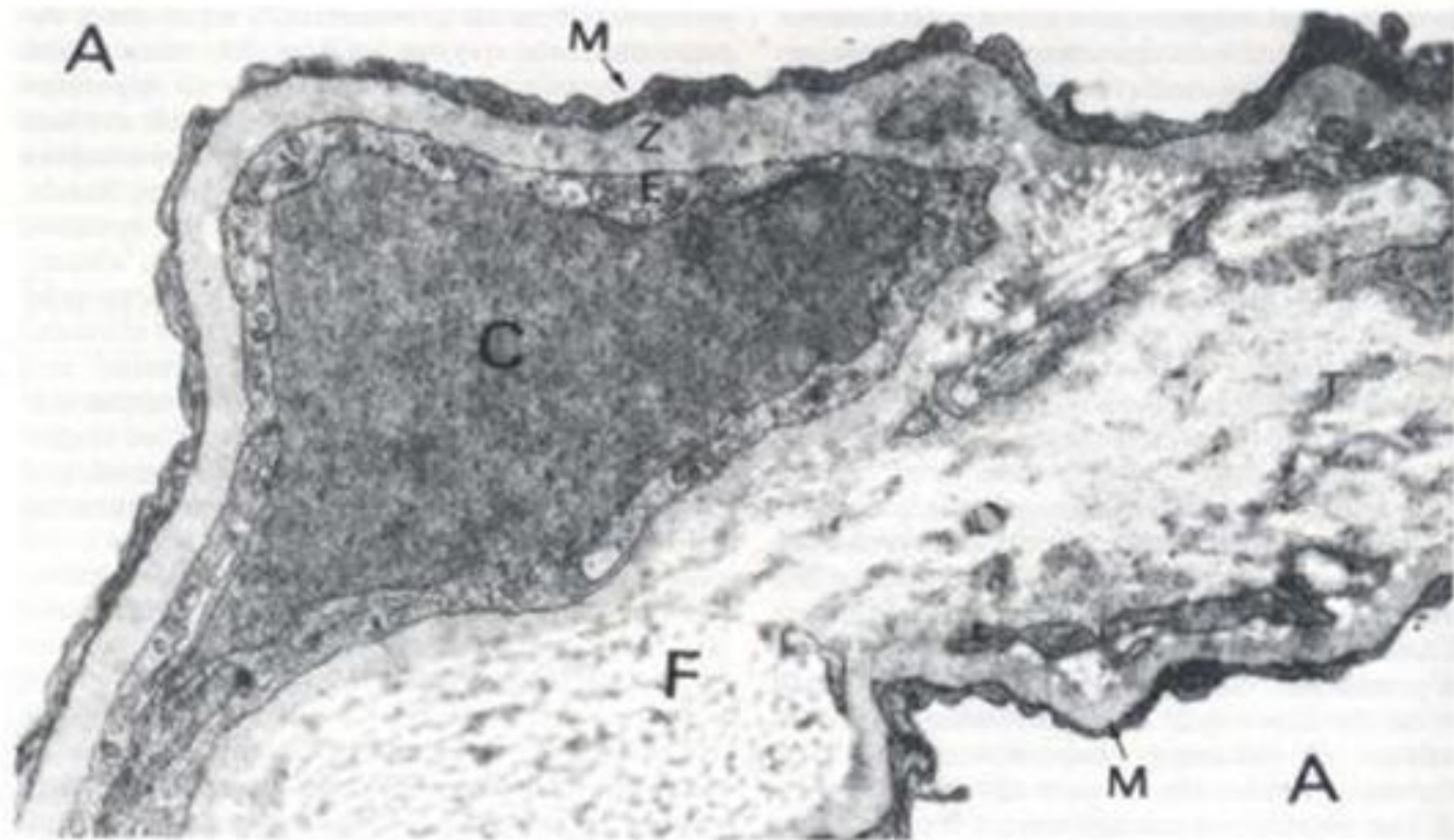
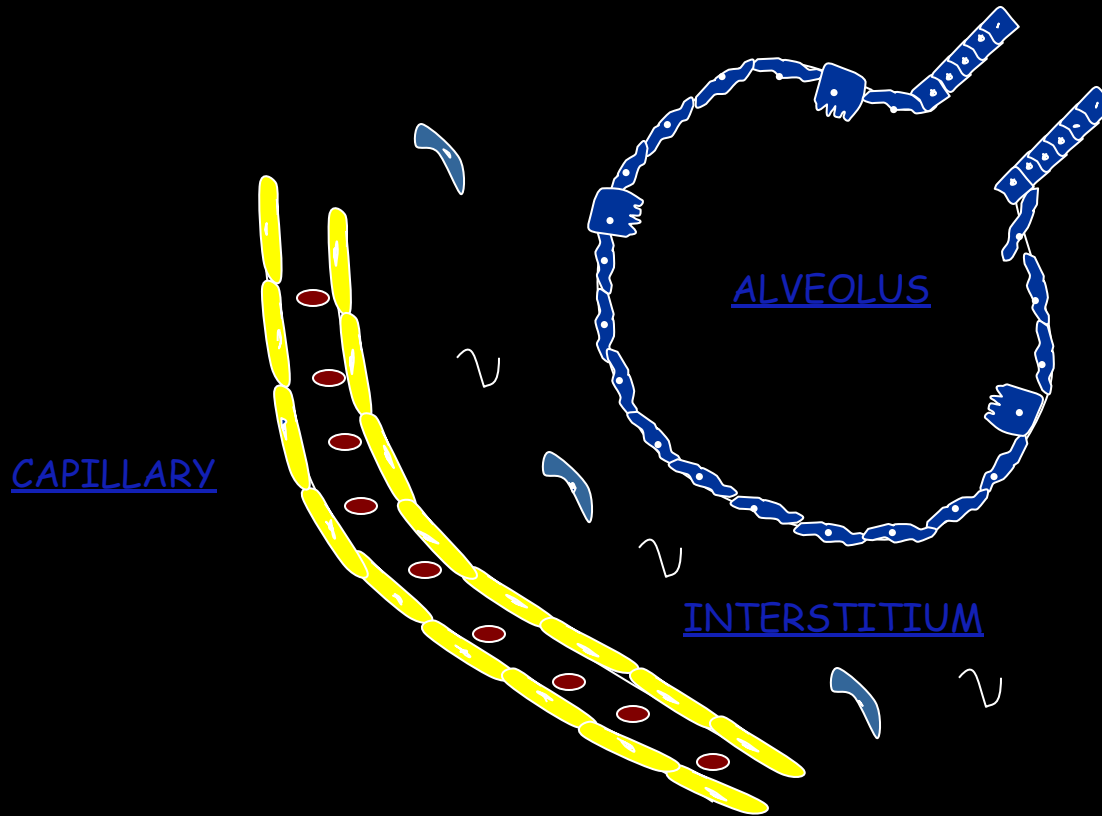


Fig. 16.11 Alveolar wall in normal human lung. The capillary lumen (C) is lined by endothelial cells (E). The alveolar spaces (A) are lined by membranous pneumocytes (M). In the thinnest portion of the blood-air pathway the endothelial cell is separated from the membranous pneumocyte by a granular amorphous zone (Z) consisting of their fused basement membranes. Elsewhere, the endothelial and epithelial cells are separated by an interstitial space containing collagen (F) and elastic fibres (T). Electron micrograph. $\times 25\,000$.

What is the interstitium and what does it do?

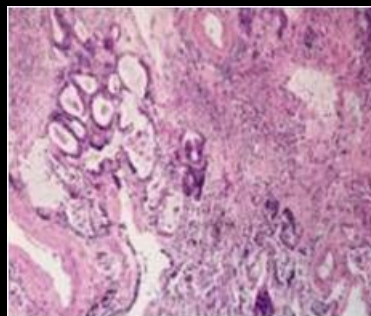
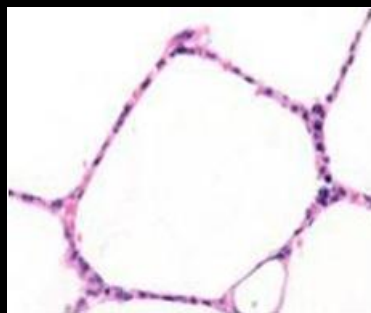


Does interstitial disease effect just the interstitium?

NO !

Structures affected:

Acini
Alveoli lumen
Bronchiolar lumen
Bronchioles

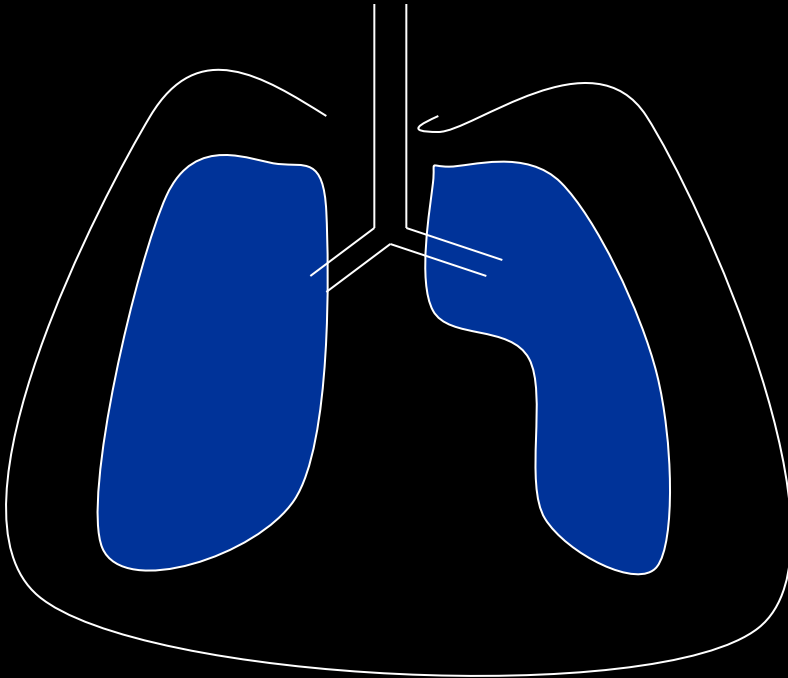


Cells involved:

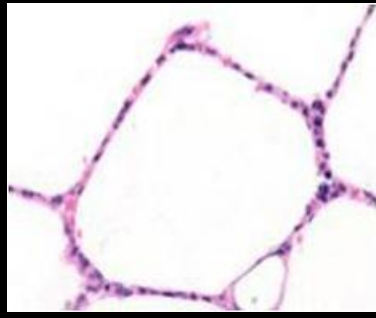
Epithelial
Endothelial
Mesenchymal
Macrophages
Recruited inflammatory cells

Chronic Diffuse parenchymal lung disease' ...

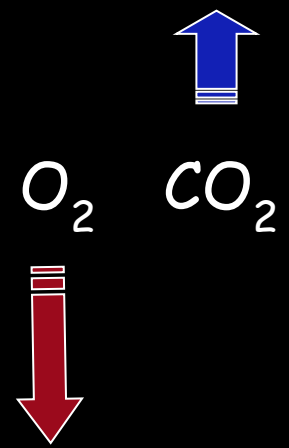
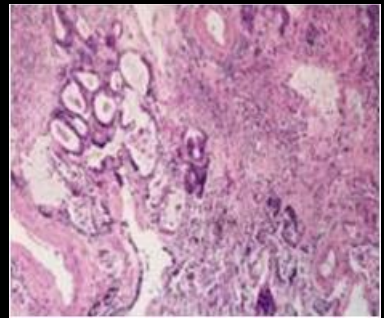
Ventilation



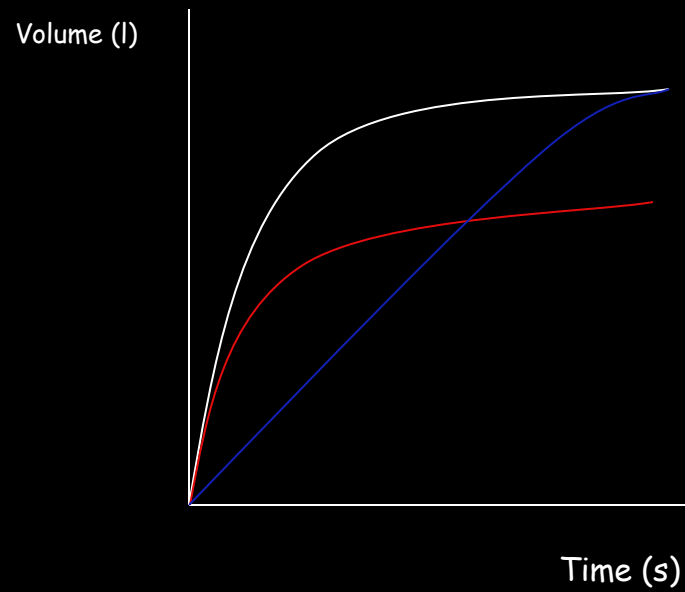
Diffusion



Perfusion



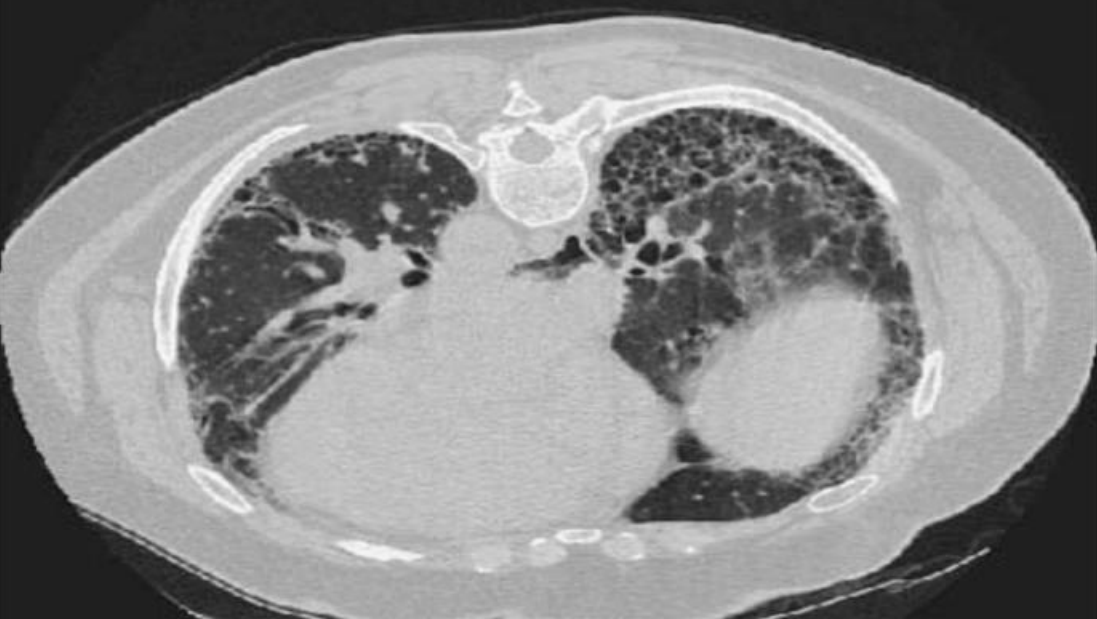
Pulmonary function tests



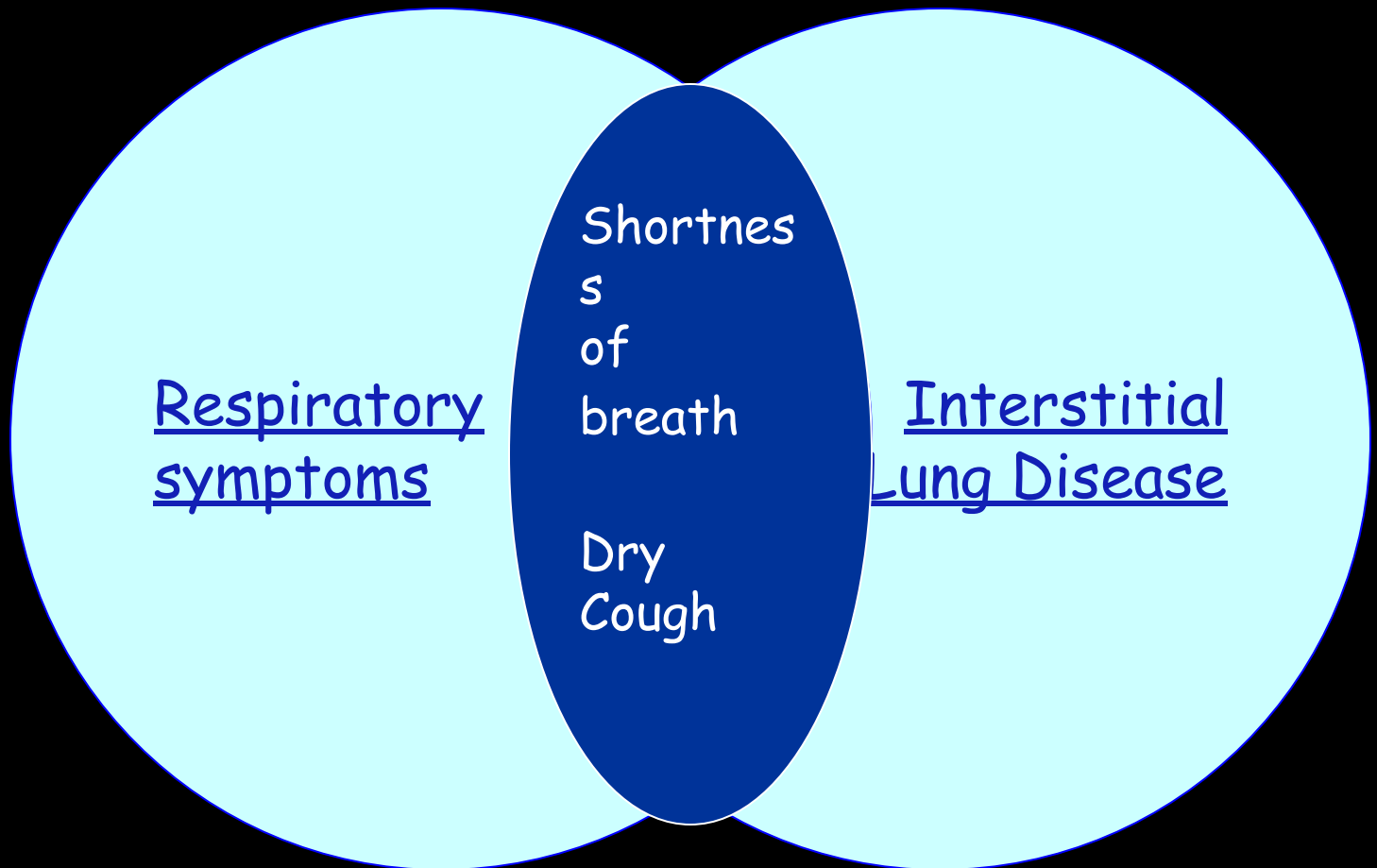
Patient 1

- 59 year old male
- Shortness of breath & dry cough, increasing 1 year - breathless with dressing
- Rheumatoid arthritis (on methotrexate) x 15 years
- Current smoker 40 years.
- Pigeon fancier
- Respiratory rate 24/min, HR 106, Oxygen saturations 87%
- Chest examination - diffuse bilateral crackles, reduced air entry
- Bilateral pitting ankle oedema





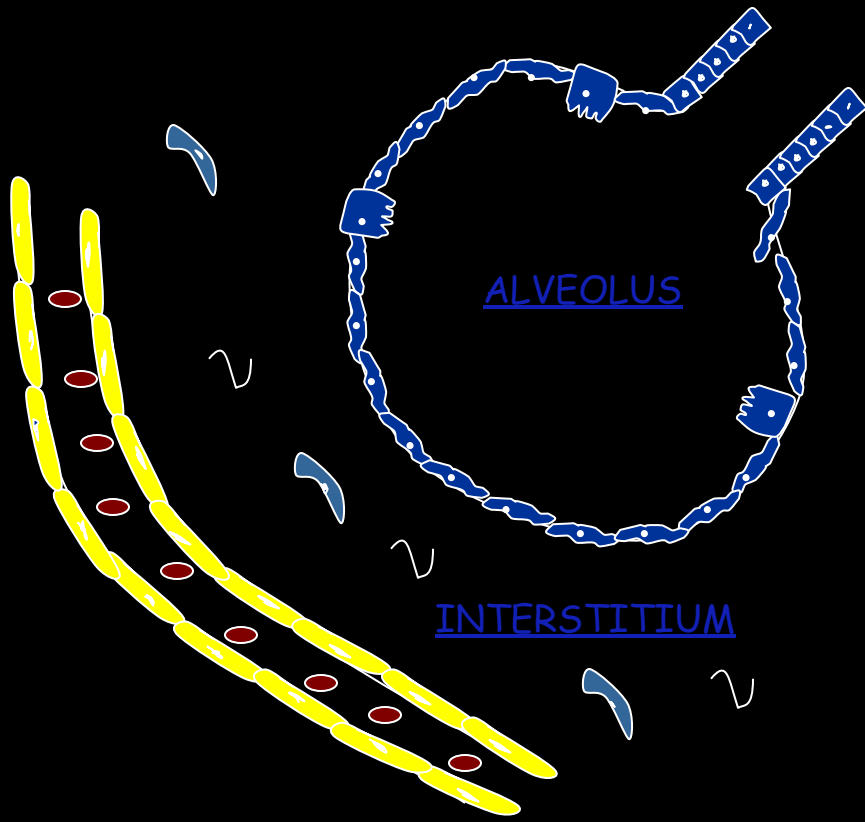
Symptoms & history taking



CAPILLARY

ALVEOLUS

INTERSTITIUM



- **Common clinical features**

- **Symptoms 1-Chronic dry cough**

- **2-Exertional dyspnea**

- **Signs 1-Clubbing**

- **2-Basal inspiratory crepitations**

- **Laboratory 1-High ESR**

- **2-Pulmonary infeltrate and reduced lung size**

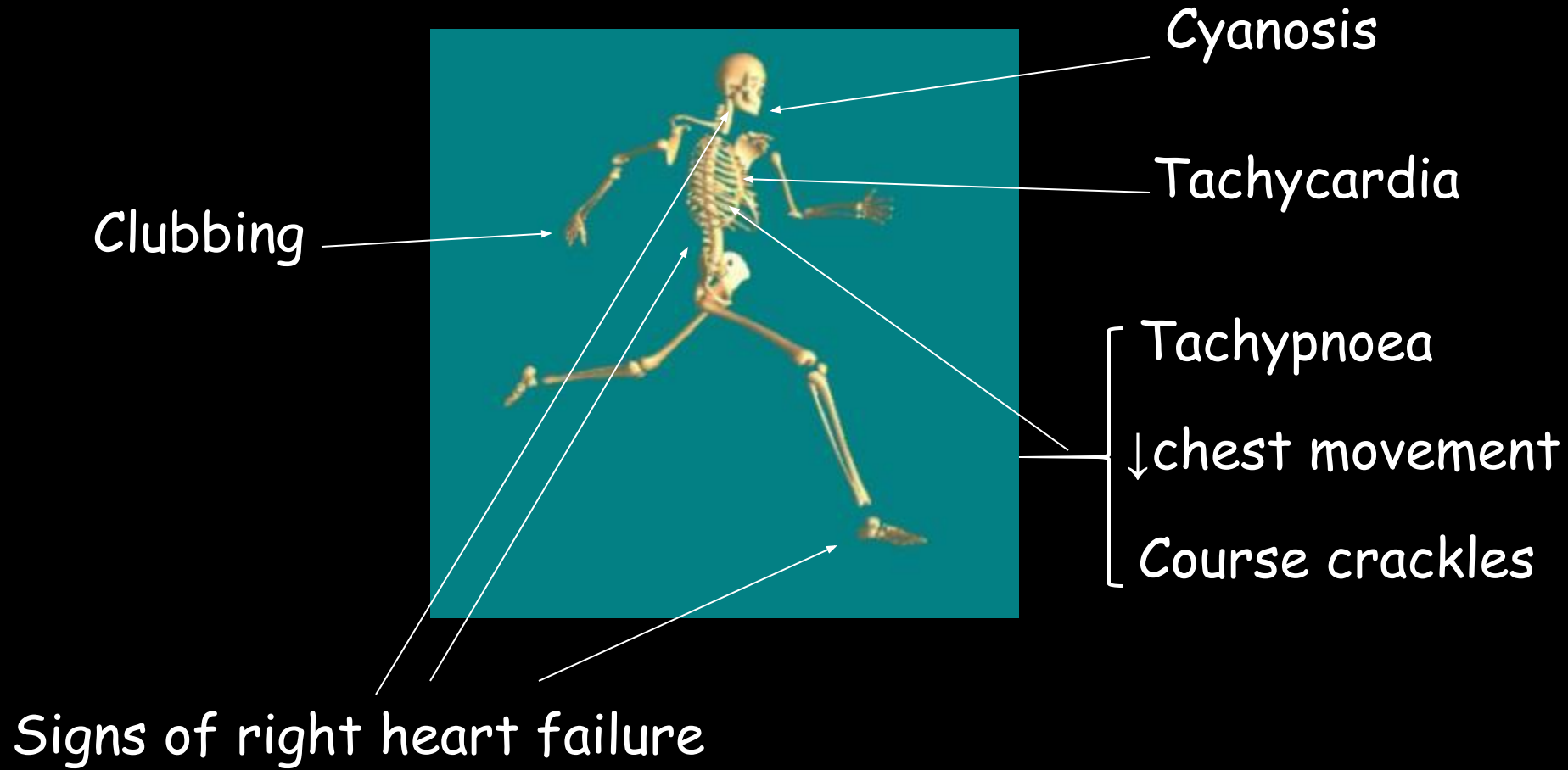
- **3-Restrictive pattern of pulmonary function tests**

Pulmonary function tests

- **Spirometry 1-Decreased FEV₁,FEV₁**
 - **(Normal FEV₁/FVC)**
 - **2-Decreased TLC**
 - **3-Mildly Decreased PEF**
 - **4-Markedly Decreased DLCO**
- **Blood gasses 1-Hypoxia**
 - **2-Hypocapnea**
- **(Type 1 respiratory failure)**

Examination

Signs of underlying disease



Interstitial Lung Disease

Occupational

- Asbestosis
- Silicosis
- Coal Workers pneumoconiosis

Treatment related

- Radiation
- Methotrexate
- Nitrofurantoin
- Amiodarone
- Chemotherapy

Connective tissue disease

- Rh. Arthritis
- SLE
- Polymyositis
- Schleroderma
- Sjogren's

Immunological

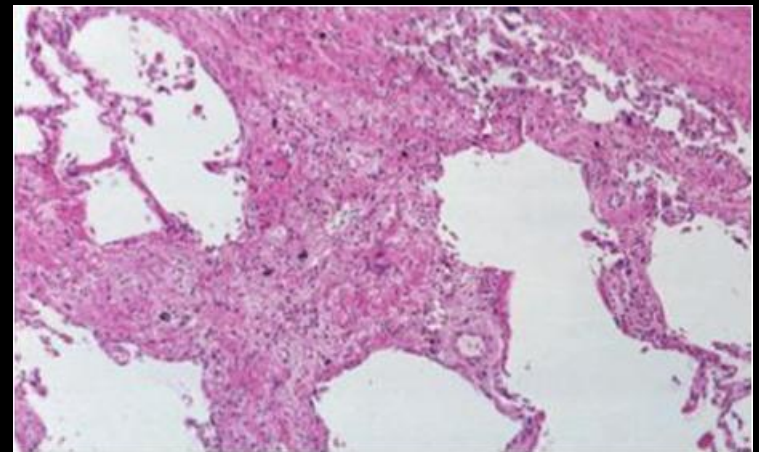
- Sarcoidosis
- Hypersensitivity pneumonitis

Idiopathic

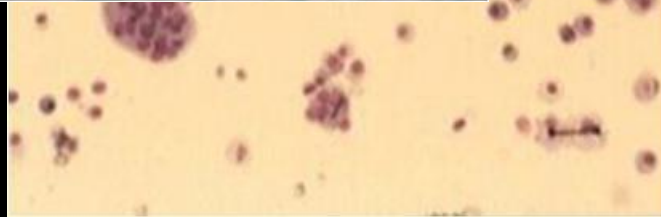
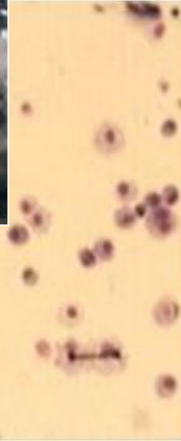
- CFA/IPF
- UIP/NSIP
- DIP
- LIP
- RB-ILD
- BOOP

Idiopathic interstitial pneumonitis (IIP)

- A variety of histological descriptions (UIP, NSIP, DIP, RB-ILD, BOOP)
- Histological descriptions - high inter and intra observer variability
- Often poor correlation with CT chest & clinician
- Biopsy may not help with management
- **More cellular - more steroid responsive**
- Presents 60-70 years old
- Cough/ Breathlessness
- CXR/Chest - basilar, bilateral, subpleural fibrosis +/- ground glass
- Restrictive PFT's
- Biopsy - variable findings
- Treatment- observe/steroids
- Prognosis - depends on cause



Asbestos



Asbestos



- Asbestos plaques
- Diffuse pleural thickening
- Benign asbestos pleural effusions (BAPE)
- **ASBESTOSIS**
- Mesothelioma
- Bronchogenic lung cancer
- Rounded atelectasis

- **Asbestosis**

Exposure history

Interstitial fibrosis - CXR/CT scan

Restriction - PFT's

Fibrosis - Biopsy

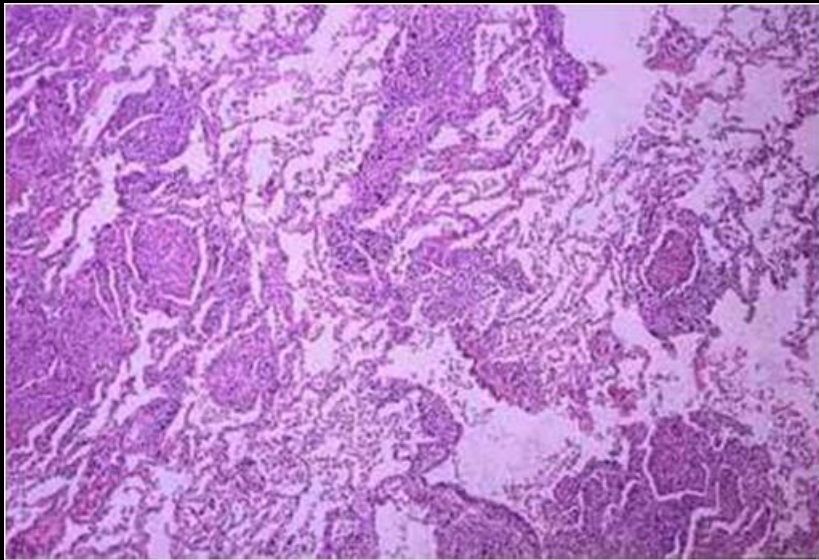
Variable progression - Prognosis

Limited treatment options

Compensation issues - Occupational lung disease

Drug induced ILD

- Methotrexate
- Bleomycin
- Amiodarone
- Nitrofurantoin



- **Methotrexate**

Treatment & cause of lung disease

Dose & duration important

Variable CXR/CT findings

PFT's Restriction

Biopsy - variable

Treatment - withdrawal/steroids

Prognosis - variable according to dose and duration

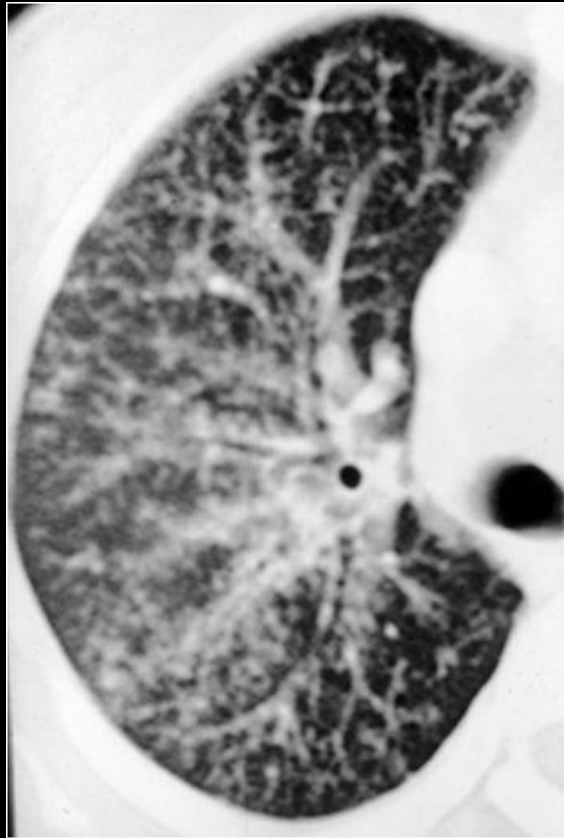
Rheumatoid lung disease

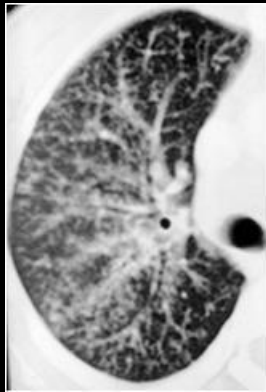


Connective tissue disease

- Dermatomyositis/ Polymyositis
 - Sjogren's Syndrome
 - Systemic Lupus erythematosus
 - Scleroderma
 - Rheumatoid arthritis
 - Rheumatoid lung disease
- May predate arthritic symptoms
Disease or treatment may be cause
Male > female
Variable CXR/CT findings
PFT's Restriction/normal
Biopsy- variable findings
Treatment - rheumatoid
drugs/observation
Prognosis - variable

Sarcoidosis





Sarcoidosis

- Often asymptomatic
- Genetic predisposition
- Cough & breathlessness
- Normal chest examination
- May get better, remain static, worsen...unpredictable
- Grading system 0-4
- CXR/ CT -specific features
- Restriction/mixed PFT's
- Biopsy - transbronchial, non-caseating granuloma
- Differential diagnosis - lymphoma & TB
- Treatment - Observation vs. prednisolone

Interstitial disease

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- What are the clinical manifestations?

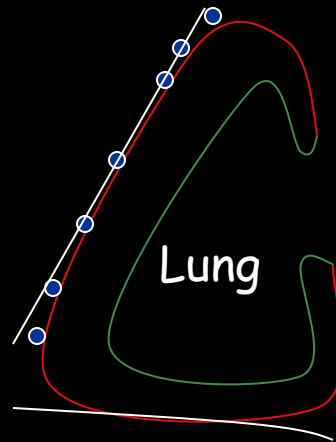
Objectives

- Interstitium
- Pleural disease
- Chest wall disease

Pleural Disease

- Anatomy
- Effusions
- Malignancy

Pleura

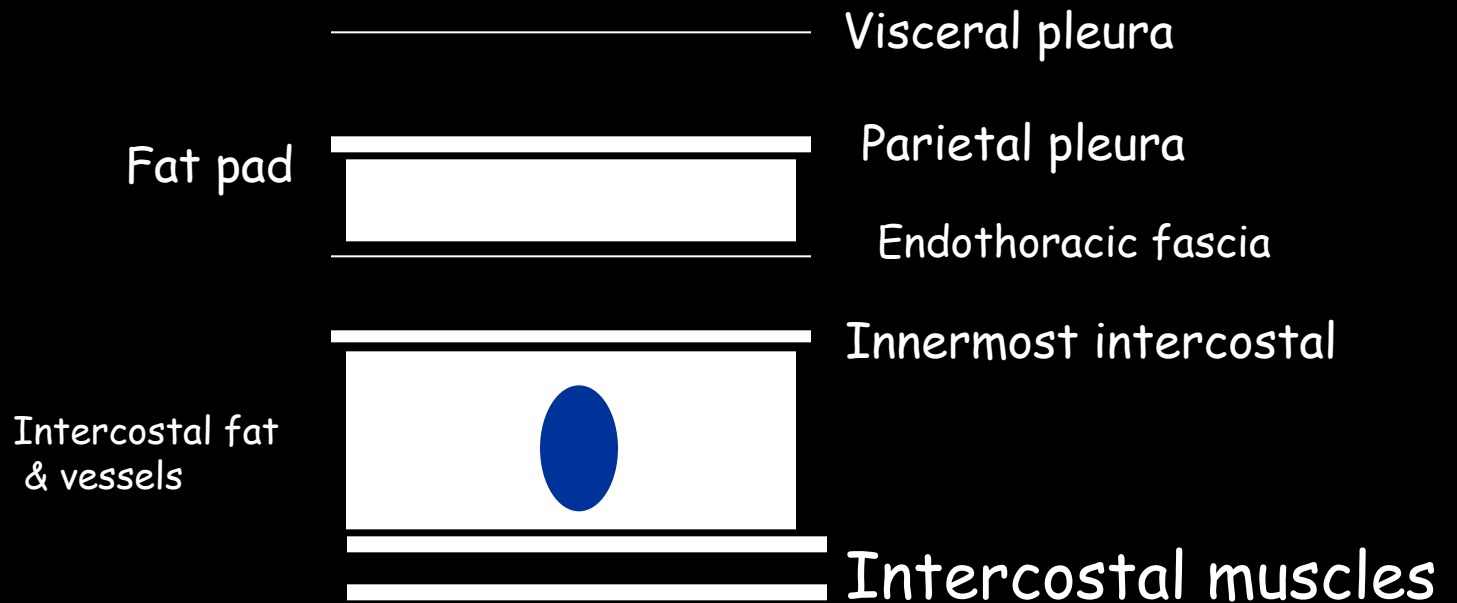


Parietal pleura

Visceral pleura

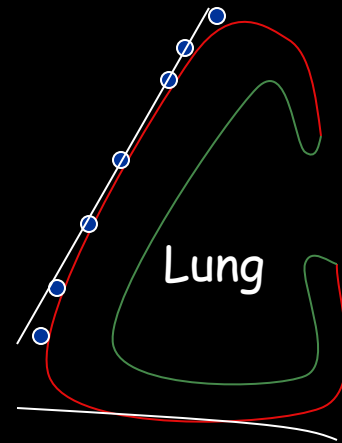
Pleural Space

LUNG



Functions of the pleural space

- Allow movement of lung and chest wall
- Coupling of chest wall and lung - inward lung recoil, outward chest wall recoil
- Pleural fluid circulation



Blood supply

- Parietal pleura

Intercostals & IMA

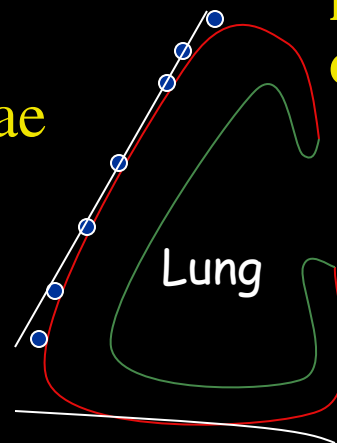
- subclavian artery

Venous drainage -
peribronchial and venae
cavae

- Visceral pleura

Bronchial arteries and
pulmonary circulation

Venous drainage -
pulmonary venous
circulation



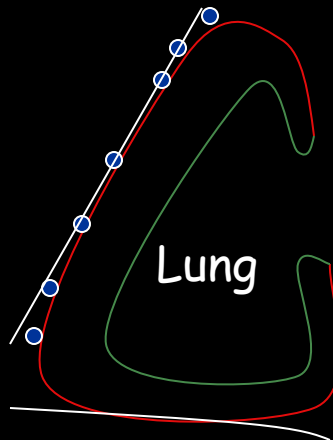
Lymphatic drainage

- Parietal

Intercostal and internal mammary
lymph vessels

- Visceral

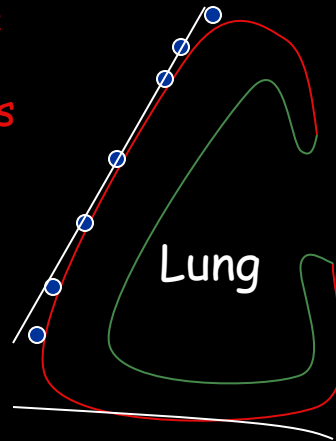
Pulmonary lymphatics



Pleura - innervation

Parietal pleura - somatic,
sympathetic & parasympathetic

Phrenic & intercostal nerves



Visceral pleura - devoid of
somatic innervation

Pleural fluid turnover

- 15ml per day (can increase to 300 ml/day)
- **Production** - Capillary filtration(Starling forces) - Parietal pleura only
- **Absorption** - Lymphatic drainage, parietal pleural lymphatics - via stomata on parietal pleural surface (mainly mediastinal, diaphragmatic regions)

Pathogenesis of pleural fluid accumulation

- **Increased production**

Lung interstitial fluid increase

Hydrostatic pressure increase

Permeability increase

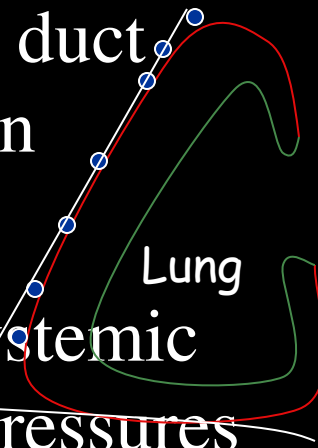
Oncotic pressure decrease

- **Decreased absorption**

Lymphatic blockage

Thoracic duct disruption

Elevated systemic venous pressures



Pleural effusions

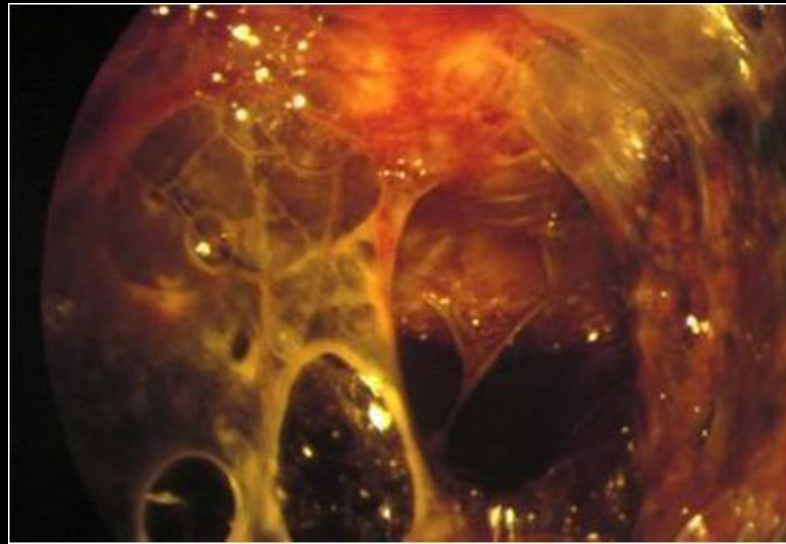
Transudate

- Hydrothorax
- Haemothorax
- Chylothora
- **Empyema**

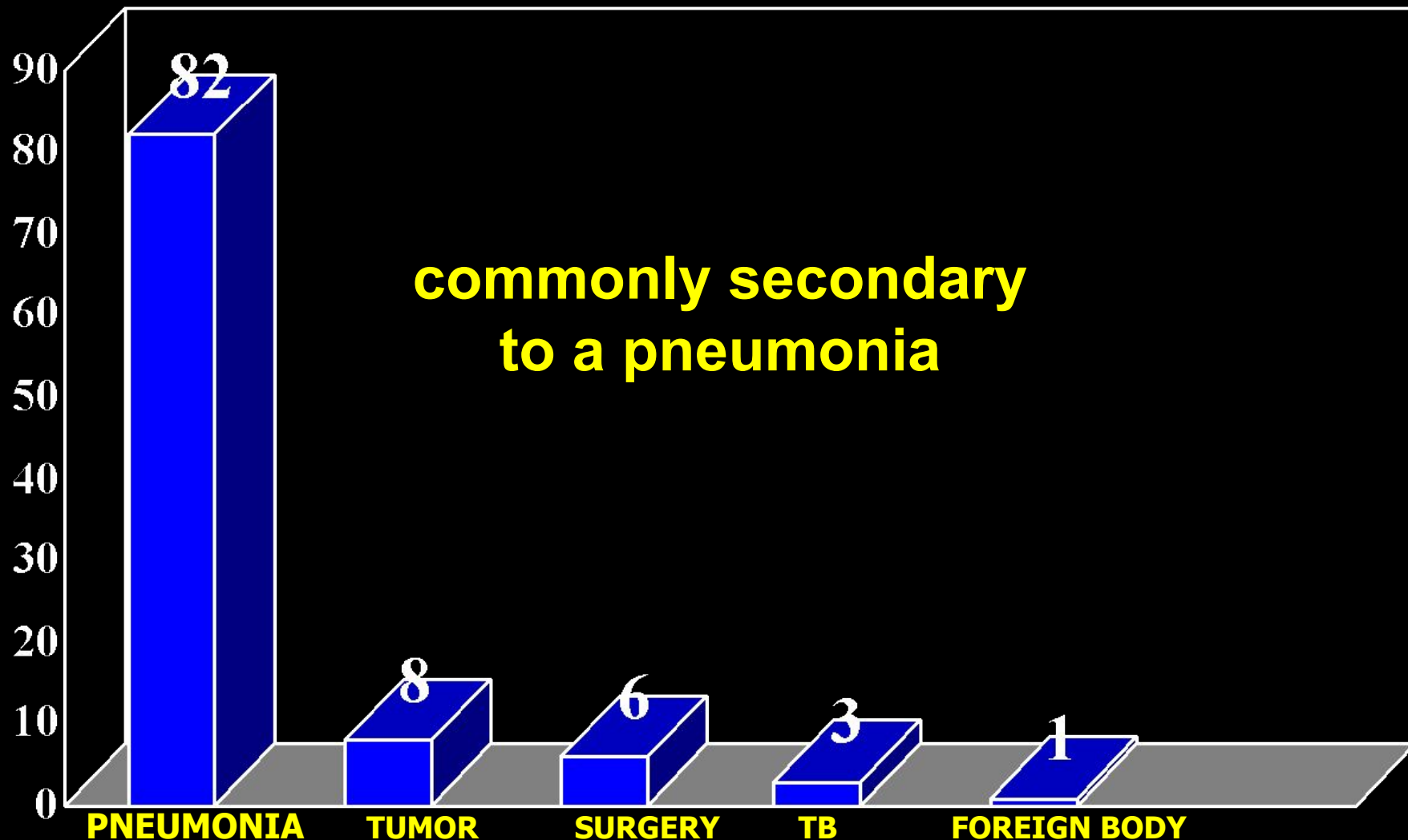
Exudate

Thoracocentesis

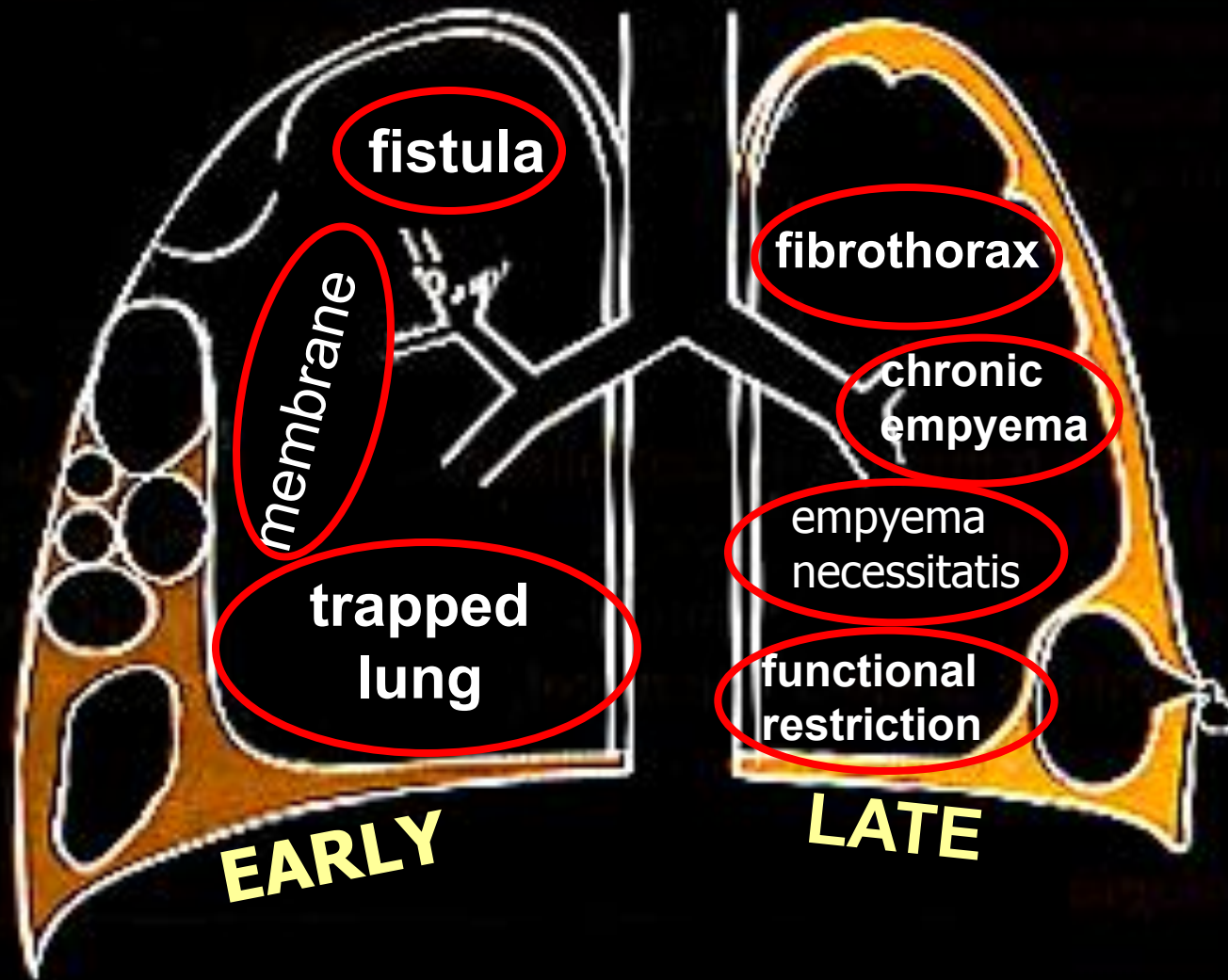
PLEURAL EMPYEMA



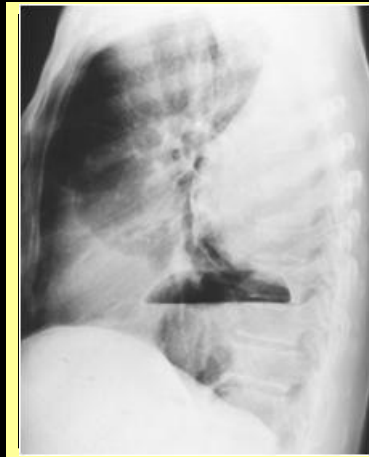
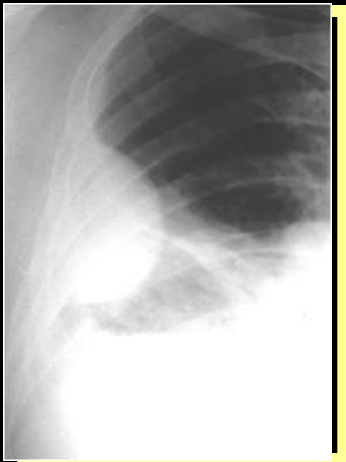
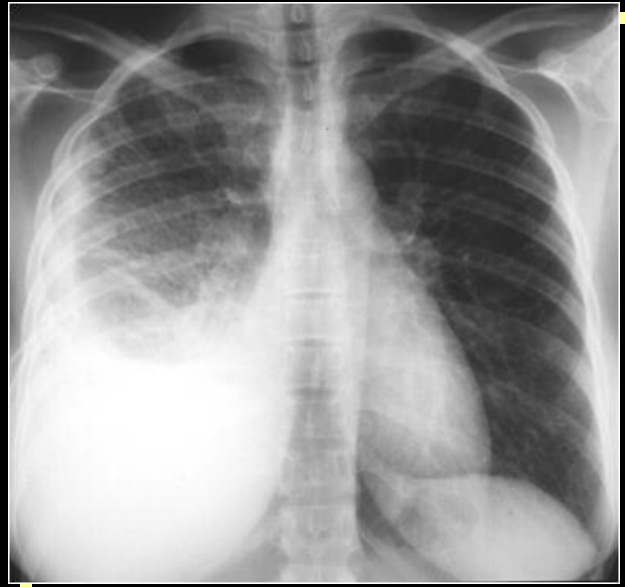
PLEURAL EMPYEMA



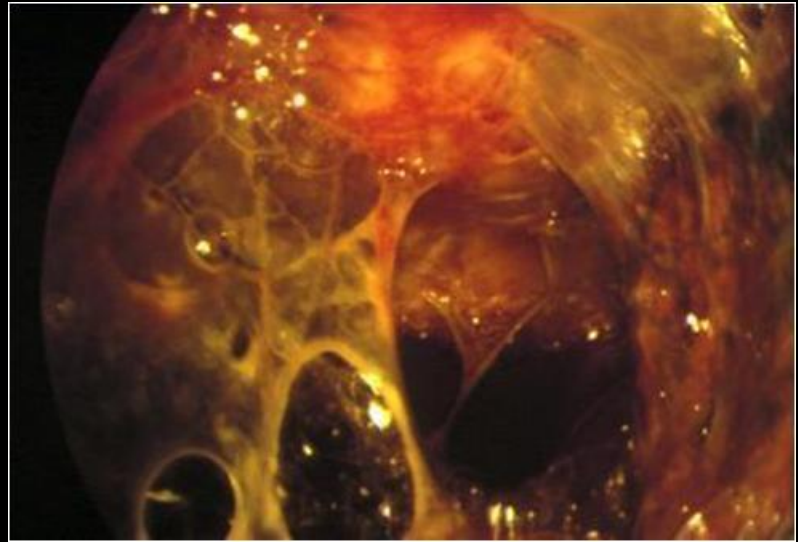
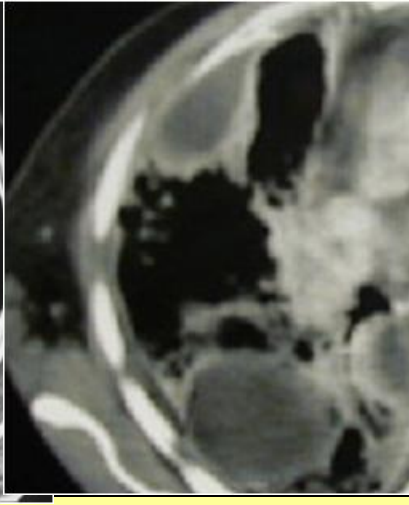
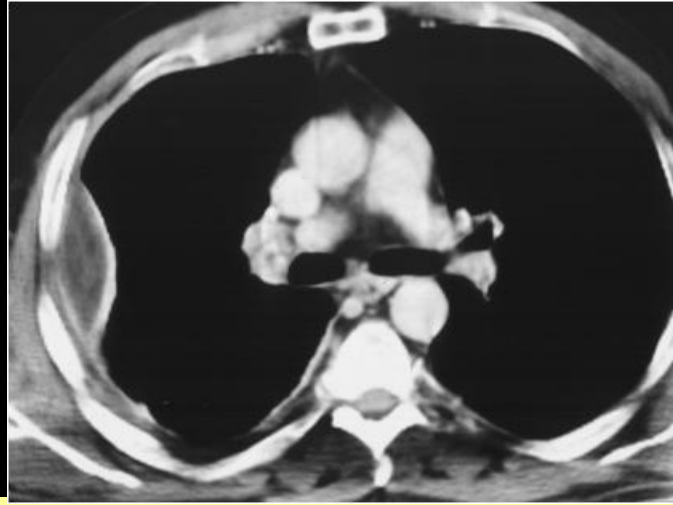
EMPHYEMA: complications



PLEURAL EMPYEMA



PLEURAL EMPYEMA



Pleural malignancy

- Metastatic

- Primary - mesothelioma

- Mesothelioma

Asbestos exposure

Pain, breathlessness

Effusion, mediastinal pleural enhancement

Chemotherapy, palliative & radical surgery

Poor prognosis

Pleural Disease

- Anatomy
- Effusions
- Malignancy

Objectives

- Interstitium
- Pleural disease
- Chest wall disease

Chest wall disease

- Congenital

Pectus deformities

Scoliosis

Kyphosis

Muscular dystrophy

- Acquired

Trauma

Iatrogenic

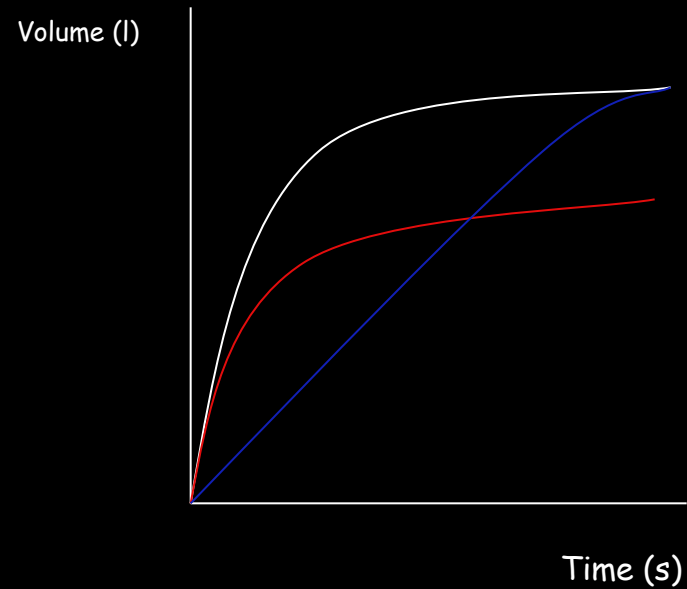
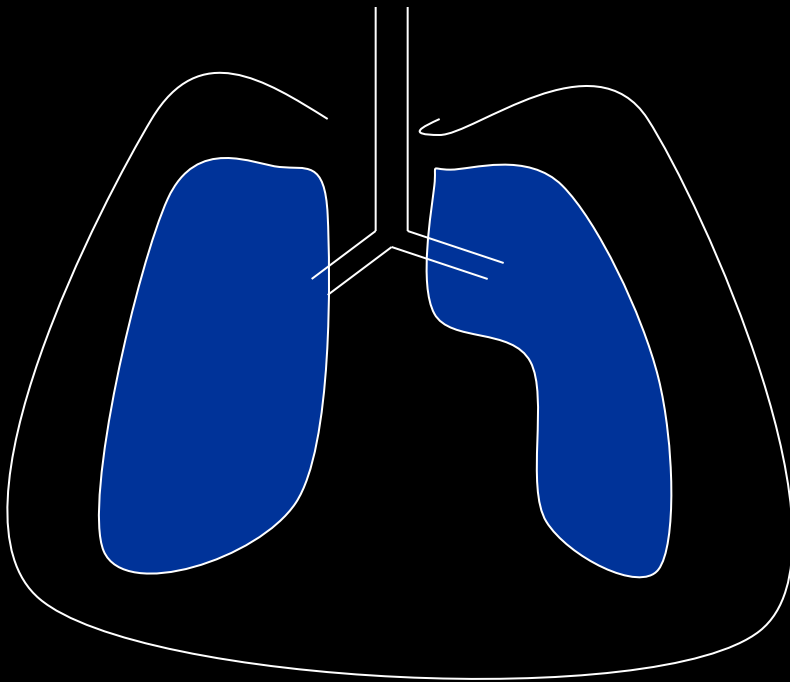
Ankylosing spondylitis

Motor neurone disease



Chest wall disease

Ventilation



Chest wall disease

- Ventilation
- Sleep disordered breathing
- Poor clearance of secretions
- Atelectasis
- Pneumonia

Clubbing

