Idiopathic pulmonary fibrosis

PRESENTED BY:

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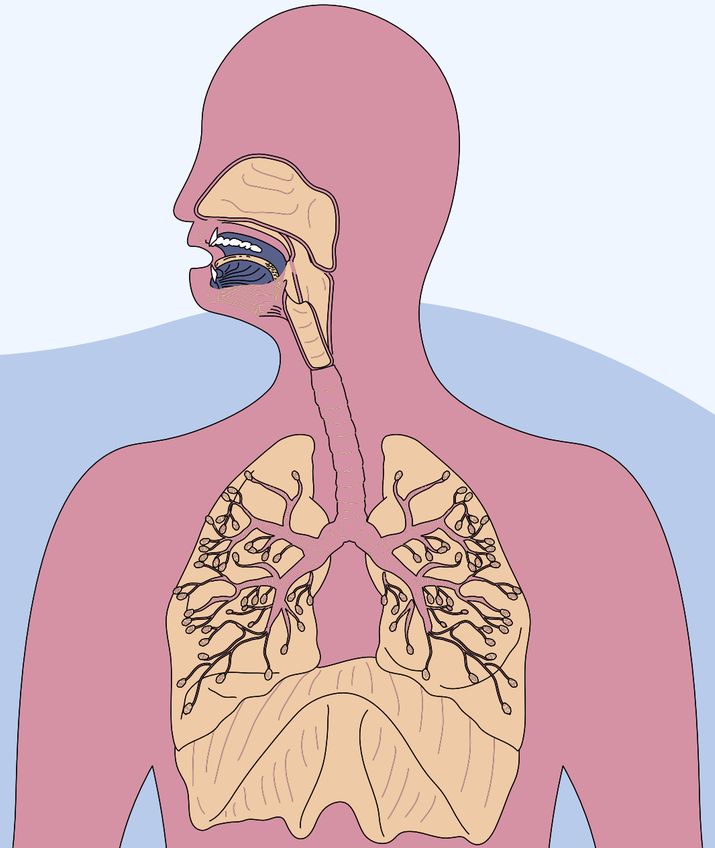
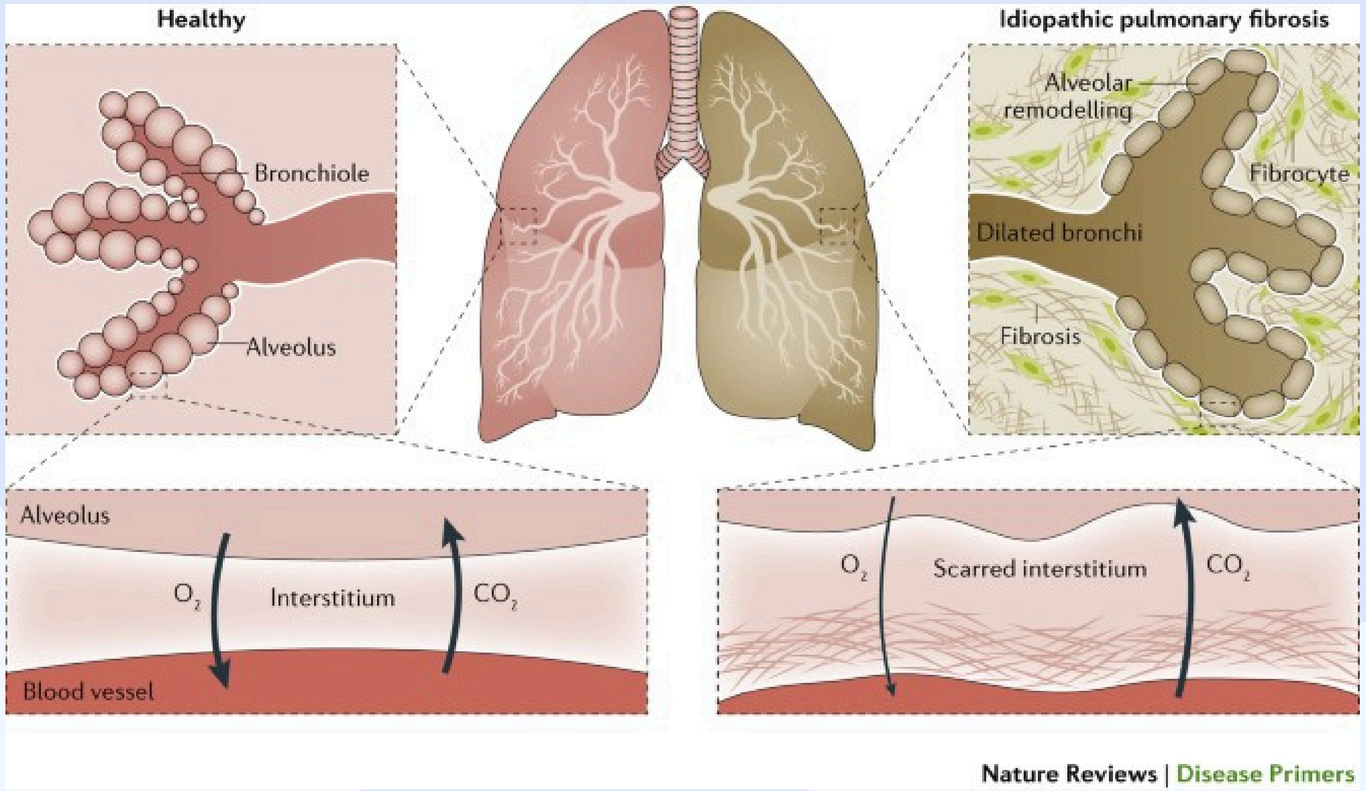


Definition :

Refers to a pulmonary disorder of unknown etiology that is characterized by patchy , progressive bilateral interstitial fibrosis.

it is the most common type of ILD.



- Males are affected more often than females .
- It is a disease of aging, affect mostly men 50-70 years of age.
- More common in smokers.
- IPF is one of chronic interstitial lung disease that show restrictive pulmonary function.

Prognosis:

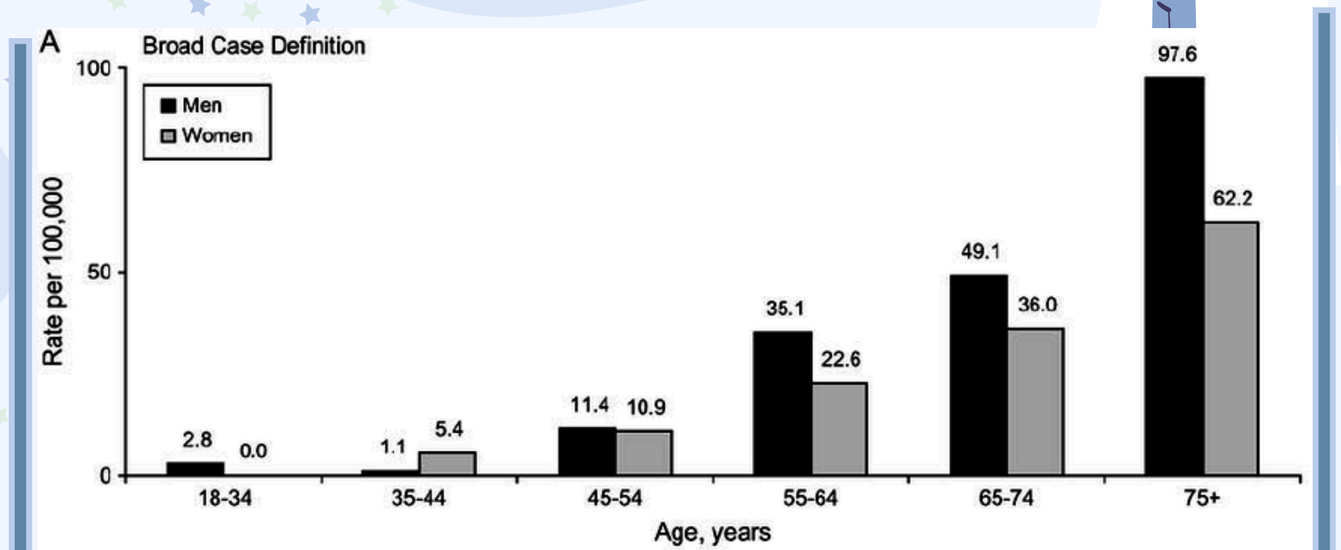
poor , the mean survival is only 3 to 7 years after first diagnosis due to respiratory failure.

Epidemiology:

Estimated to affect 5 million people worldwide.

Most cases are sporadic but rare cases of familial IPF have been described.

The most common (and deadly)interstitial lung disease.



Pathophysiology:

1: Injury to Alveolar Epithelium:

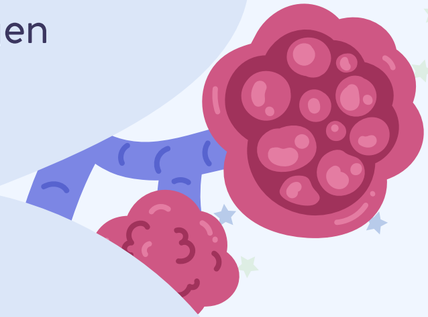
The initial trigger is often unknown This injury may be caused by environmental factors (as smoking , fume ...etc), genetic predisposition, or other unknown agents.

2: Inflammatory Response:

leads to the activation of various immune cells, including macrophages and lymphocytes.

3: Fibroblast Activation:

The persistent inflammation stimulates fibroblasts to proliferate and migrate to the site of injury. This results in excess collagen deposition in the lung interstitium



4: Fibrosis:

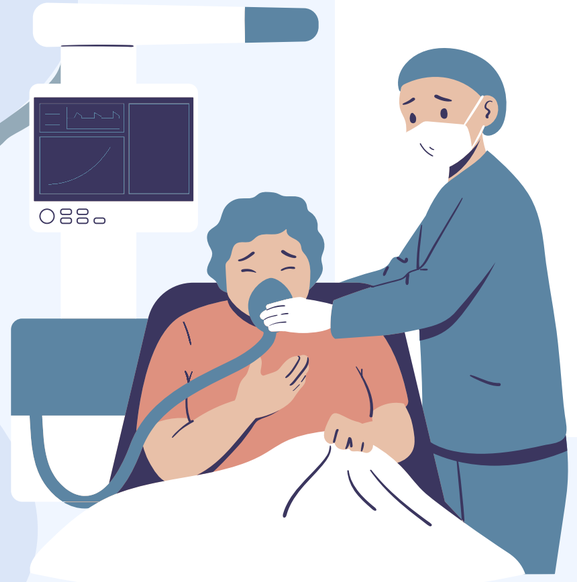
Over time, the accumulation of collagen leads to fibrosis. This disrupts the normal architecture of the lungs and leads to impaired lung function.

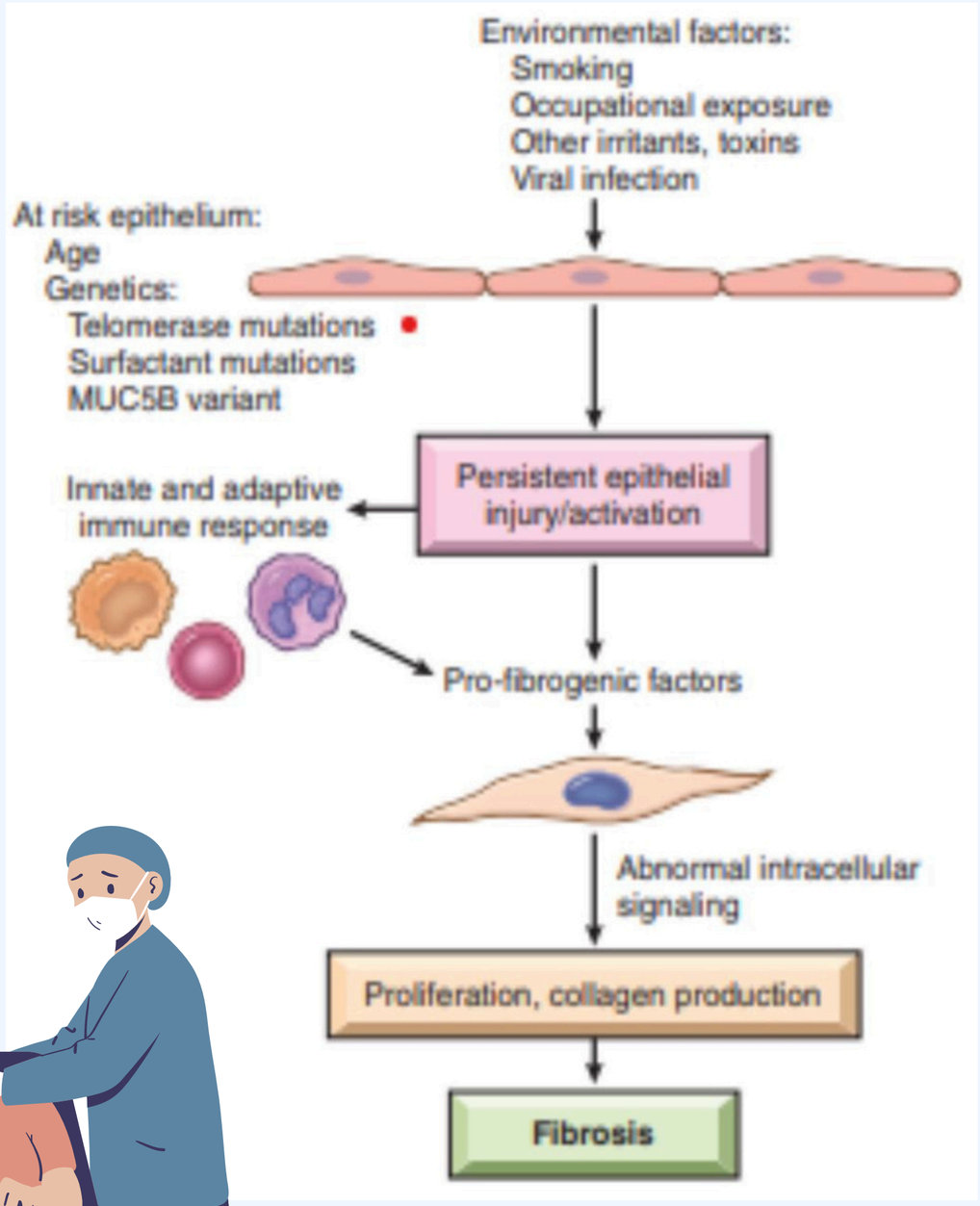
5: Impaired Gas Exchange

The fibrosis reduces lung compliance and impairs gas exchange, resulting in symptoms like shortness of breath, dry cough, and decreased exercise tolerance.

6: Progression:

IPF is progressive, meaning that the fibrosis worsens over time, leading to respiratory failure and increased mortality.



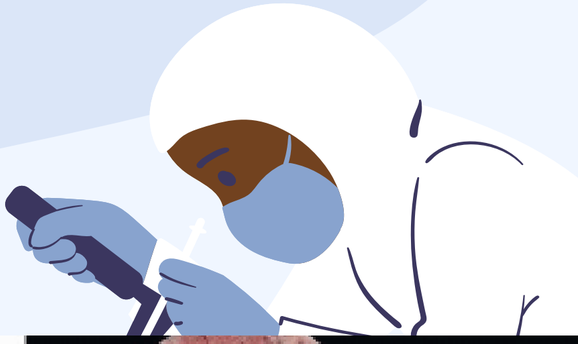


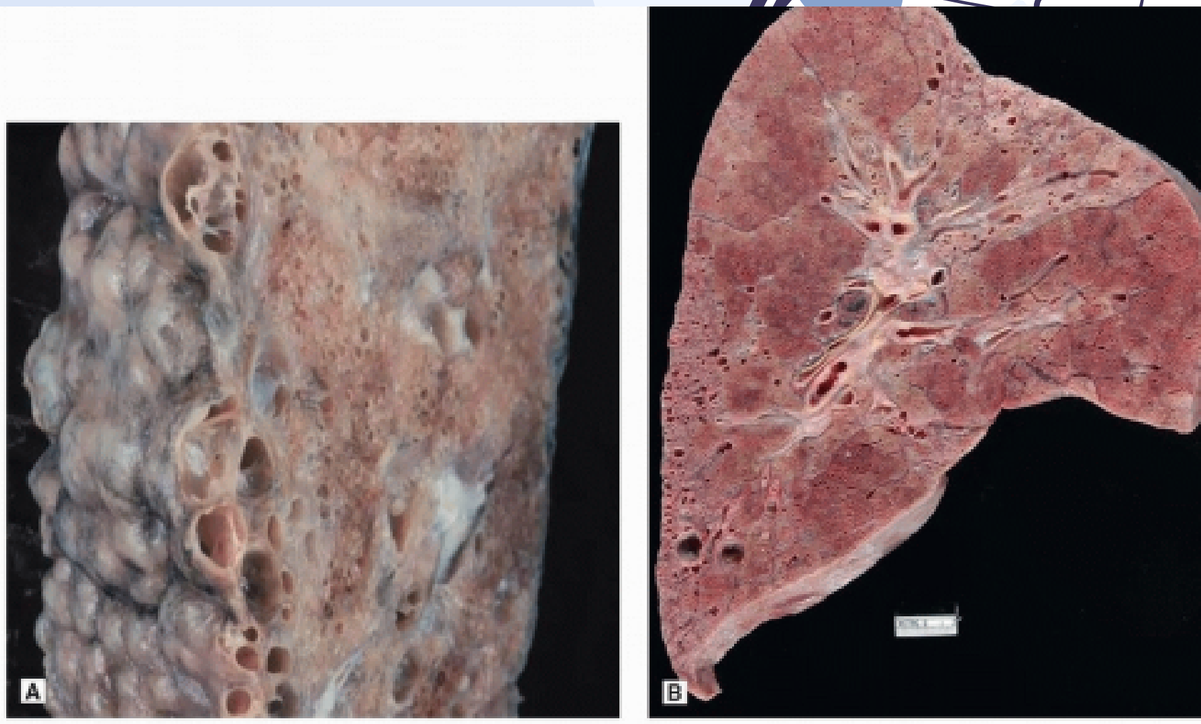
★ **So it is Repeated cycles of tissue injury in the lung parenchyma with aberrant wound healing → collagenous fibrosis → remodeling of the pulmonary interstitium**

Grossly:

the pleural surfaces of the lung are cobblestoned due to retraction of scars along the interlobular septa.

The cut surface shows firm, rubbery white areas of fibrosis, which occurs preferentially within the lower lobe, the subpleural regions, and along the interlobular septa





Histologically:

the hallmark is patchy interstitial fibrosis, which varies in intensity and worsens with time .

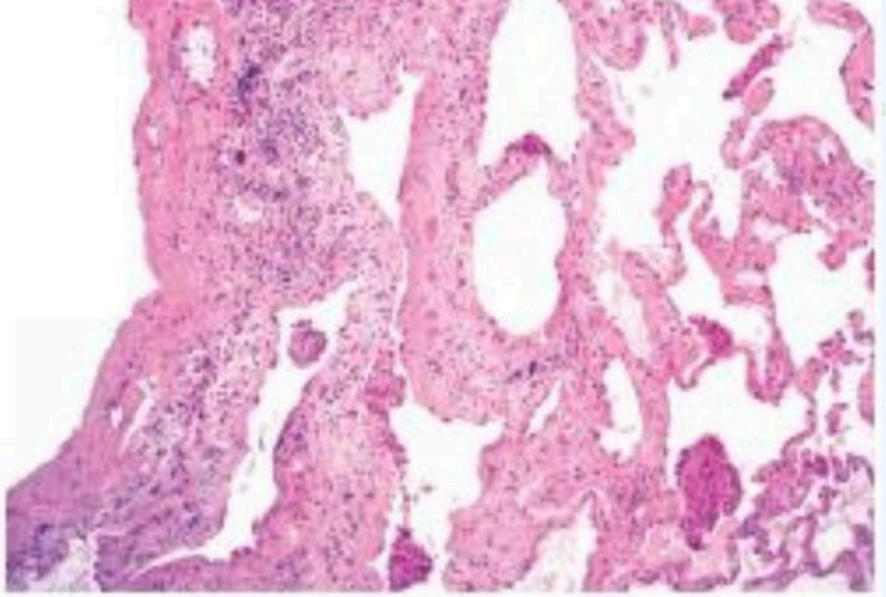
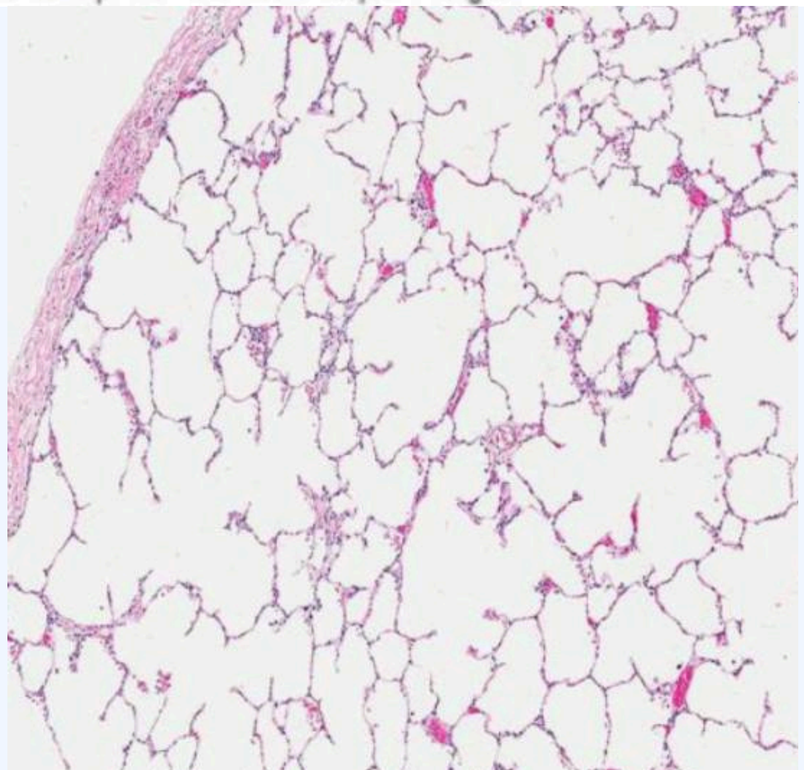


Fig. 13.14 Usual interstitial pneumonia. The fibrosis, which varies in intensity, is more pronounced in the subpleural region.

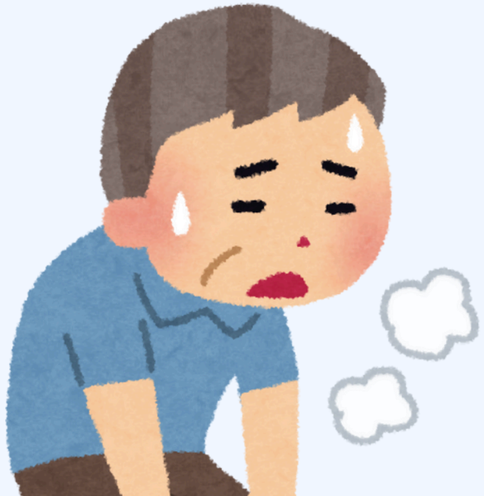


Normal

Symptoms:

Shortness of breath:

Onset: Over YEARS (3-5 years)!
Timing: Exertion later during rest.
Course: continuous & progressive.
Triggers: No trigger.
Severity: variable.

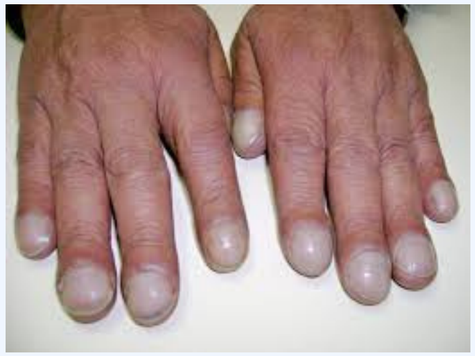


Cough:

Dry cough (unlike obstructive lung disease).

Signs Of IPf

FINGER CLUBBING



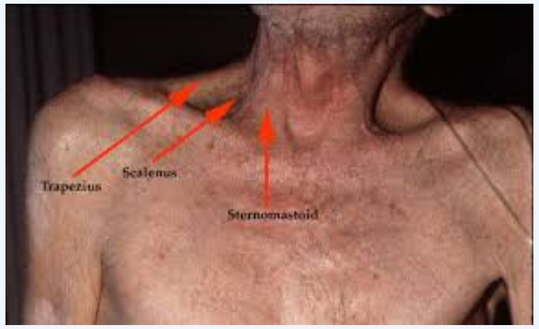
Finger clubbing is a significant physical finding often associated with idiopathic pulmonary fibrosis (IPF). It presents as a bulbous enlargement of the tips of the fingers and toes, accompanied by changes in the angle between the nail bed and the nail itself.

Signs of respiratory distress

Vital signs:

A. Respiratory rate >20 times/ min

B. O2 Saturation<94%

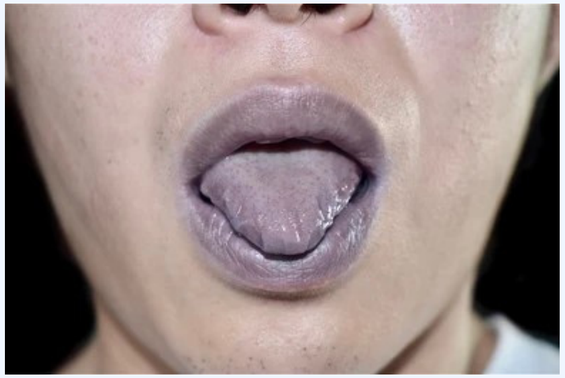


General examination:

A. Level of consciousness (hypoxia and hypercarbia)

B. Asterixis

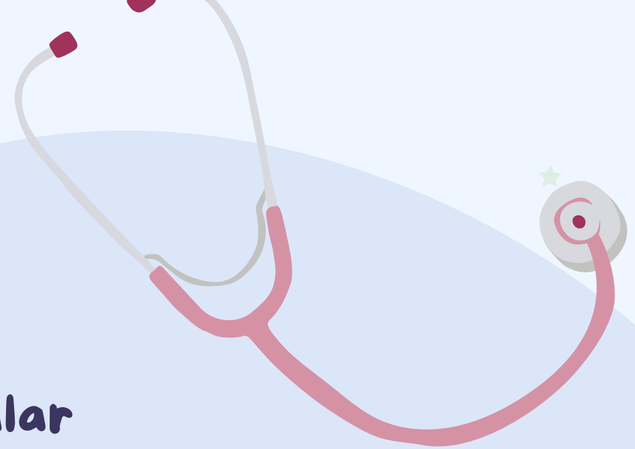
C. Central cyanosis



D. Signs of using of accessory respiratory muscles (sternocleidomastoid, abdominal muscles, subcostal and suprasternal retraction)

E. Nasal flaring

Chest examination:



1. Inspection

2. Palpation: Parasternal heave (right ventricular hypertrophy in cor pulmonale)

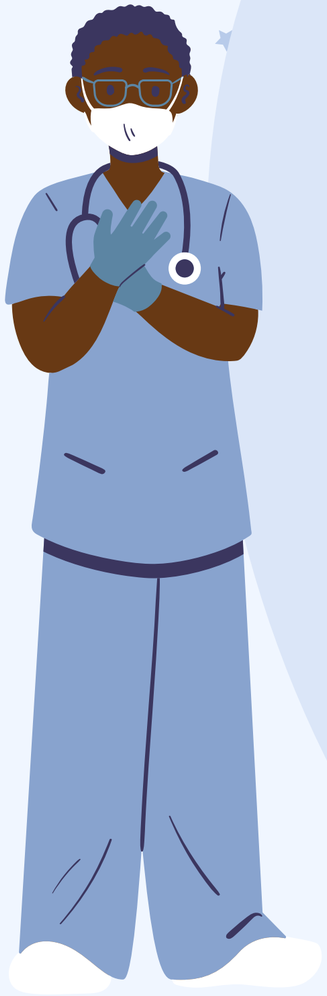
3. Percussion: resonant bilaterally

4. Auscultation:

A. Added sounds: End-inspiratory fine crackles (Dry "velcro-like" rales)

B. Heart sounds (in cor pulmonale) : Accentuated S2 (pulmonary HTN) + holosystolic murmur best heard left sternal border (functional tricuspid regurgitation).

Diagnosis:

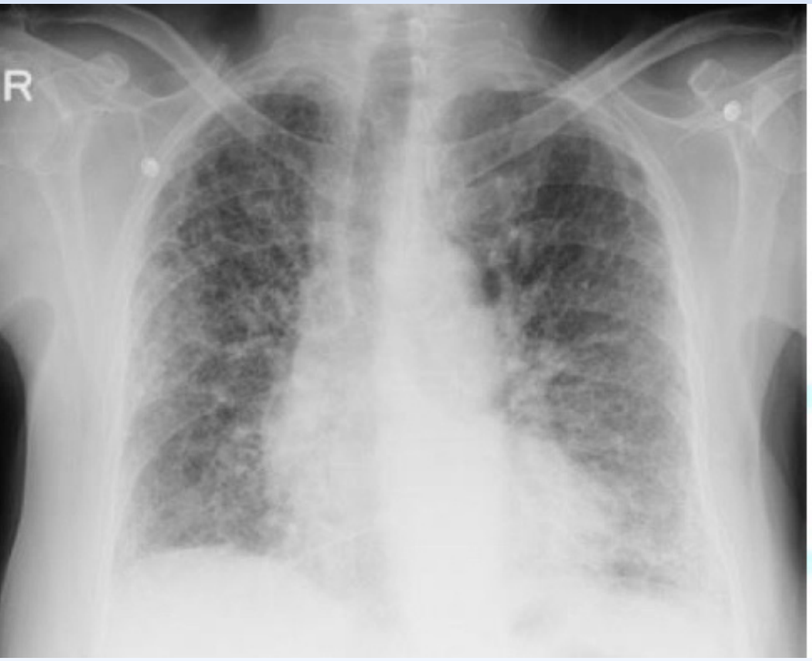


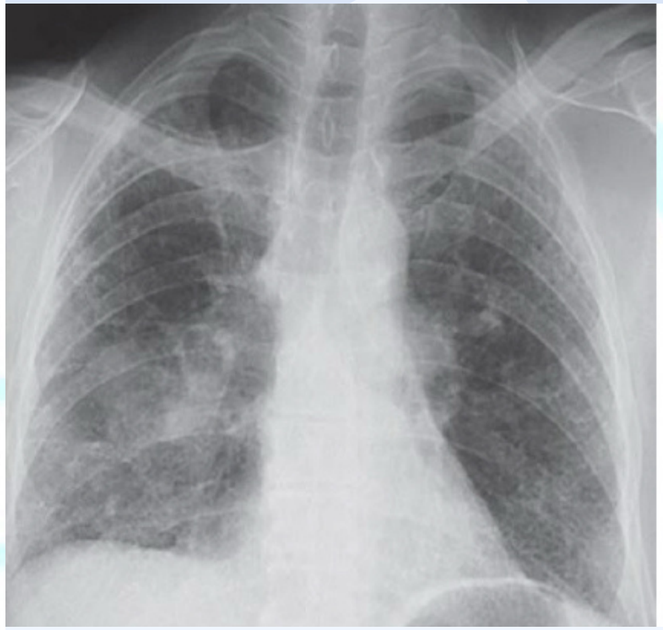
Combination of clinical history & examination + classic HRCT findings and the absence of other identifiable causes for fibrosis (e.g., environmental exposures, connective tissue disease, drug toxicity).

Diagnosis:

CHEST X-Ray

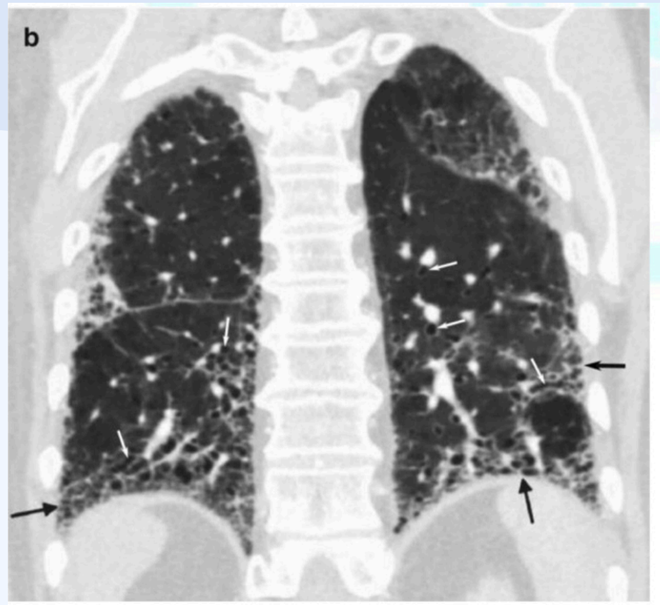
- basilar, peripheral, bilateral, asymmetric, reticular opacities
- ground-glass or a honeycombed appearance





high resolution CT

- HRCT typically demonstrates a patchy, predominantly peripheral, subpleural and basal reticular pattern and, in more advanced disease, the presence of honeycombing cysts and traction bronchiectasis



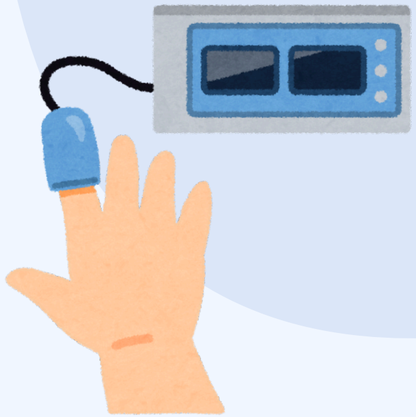
Diagnosis:

Pulmonary function tests (PFTs)

Restrictive disease with decreased forced vital capacity (FVC),
total lung capacity (TLC),
FEV1
and diffusing capacity for carbon monoxide (DLCO)
Normal or increased FEV1/FVC ratio

ABG:

Increased A-a gradient
Decreased PaO2



the 6-minute-walk test

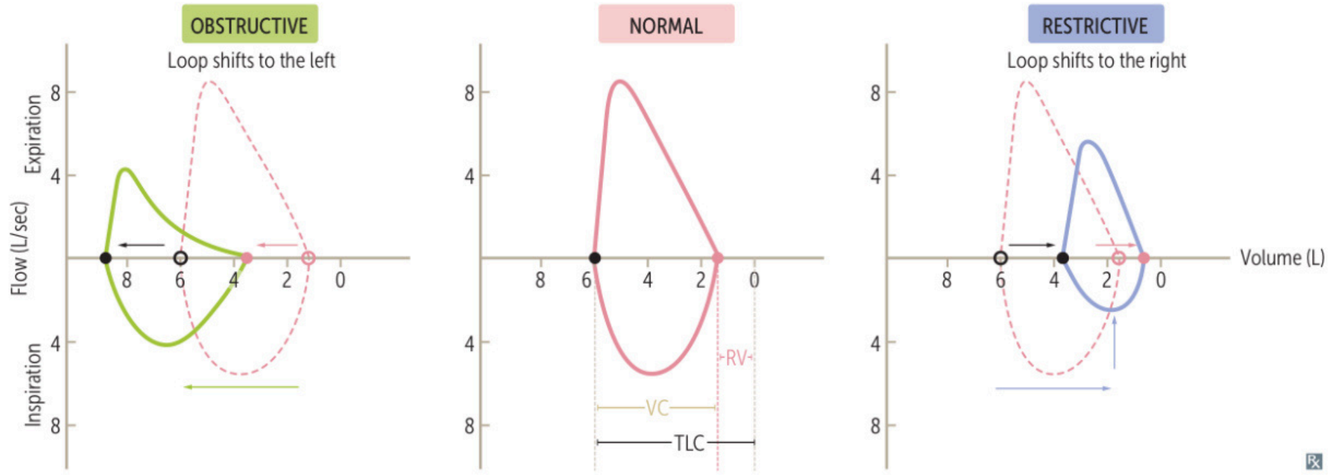
Assess Oxygen requirement during Exercise
but as IPF advances, arterial hypoxaemia present at rest

Diagnosis:

Flow Volume Loop

Shift to right and smaller in size

Flow volume loops Obstructive lung volumes > normal (↑ TLC, ↑ FRC, ↑ RV); restrictive lung volumes < normal. In obstructive, FEV_1 is more dramatically reduced compared with FVC → decreased FEV_1/FVC ratio. In restrictive, FVC is more reduced or close to same compared with FEV_1 → increased or normal FEV_1/FVC ratio.



Lung biopsy

Surgical lung biopsy is only needed if there is a disconnect between the clinical picture and HRCT findings. Biopsy pathology shows a usual interstitial pneumonitis (UIP) pattern.

Treatment:

No cure or treatment to reverse fibrosis

Pirfenidone (a tumor growth factor β-antagonist) and **nintedanib** (a tyrosine kinase inhibitor) have been shown to slow loss of lung function.

Lung transplantation remains the only effective therapy for patients with progressive or end-stage disease

Case scenario:

A 68-year-old man presents to the emergency department for worsening shortness of breath. His dyspnea was most notable with exertion; however, over the course of the past several months, it presented at rest. He reports that he has a non-productive cough and denies have any sinus pain, fever, malaise, chills, or night sweats. He has a past medical history of hypertension and hyperlipidemia. He is a retired aircraft mechanic and has smoked 1 pack of cigarettes daily for the past 45 years. His temperature is 98.6°F (37°C), blood pressure is 134/90 mmHg, pulse is 106/min, respirations are 23/min, and oxygen saturation is 88% on room air. On physical exam, the patient is alert to person but not place or time. There are rales on pulmonary auscultation and digital clubbing. A chest radiograph demonstrates a reticular pattern and honeycombing.

Thank you for your attention