- 50 year old Police man
- Presents with cough and SOB for 2 yr
- Moving to different hospitals, doctors and hakeems.
- Treated for asthma with inhaled β2 stimulant and steroides plus oral theophylines.
- ▲?

▲, DD

- Asthma
- COPD
- VHD/CHD/Heart failure
- Pulmonary fibrosis
- Pulmonary hypertension
- Bronchiectasis

Details of presenting complaint

- SOB
- Cough

- Other information required?

Relevant and systemic enquiry

- Orthopnea, syncope, palpitations, wheezing, cough, sputam, hemoptysis, rash, joint problem, allergy

- Other information required?

Other details

- Father of two kids
- Non-smoker
- No other medication except for described earlier. Some improvement with hakeem Rx
- Well off
- No pets

- ▲?

▲, DD

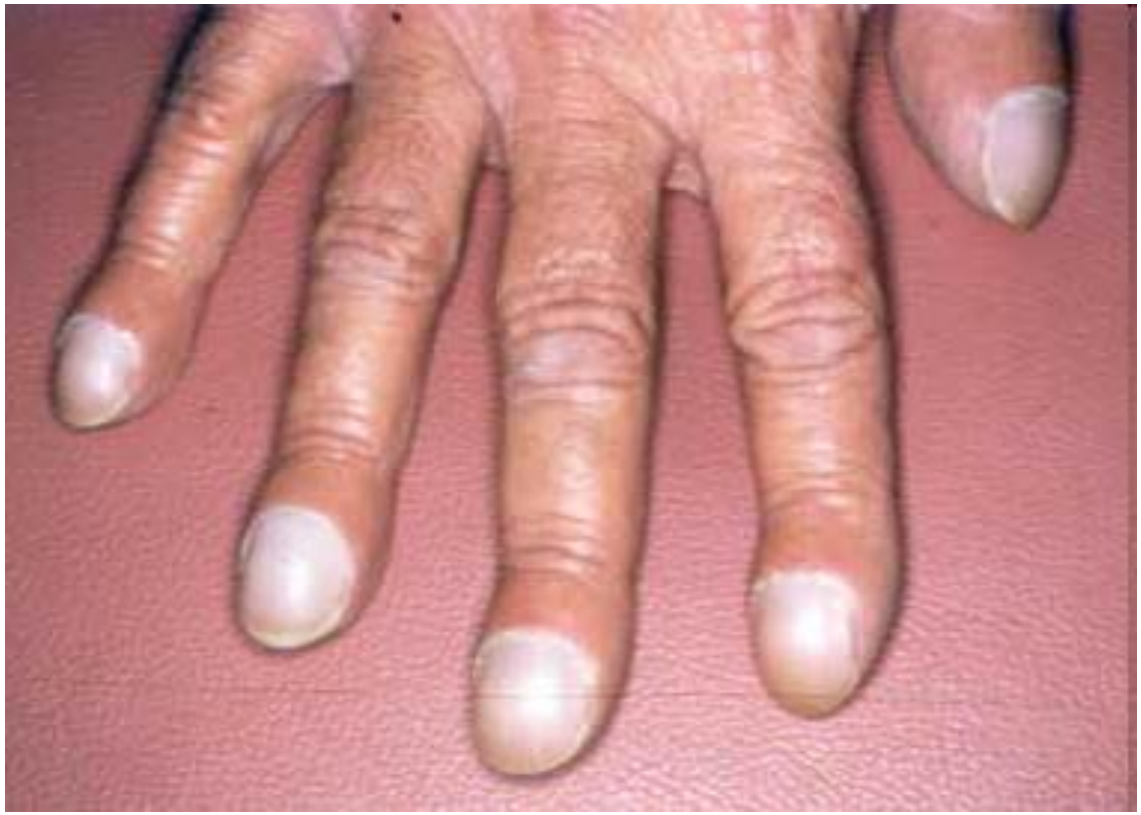
- Asthma
- COPD
- VHD/CHD/Heart failure
- Pulmonary fibrosis
- Pulmonary hypertension
- Bronchiectasis

▲, DD

- Pulmonary fibrosis
- Pulmonary hypertension
- Asthma
- COPD
- VHD/CHD

- What next?

Clinical evaluation



Clinical evaluation

- Tachycardia, raised JVP, edema
- RVH, palpable & loud P2, systolic murmur.

- Chest fine crackles at both bases

▲, DD

▲, DD

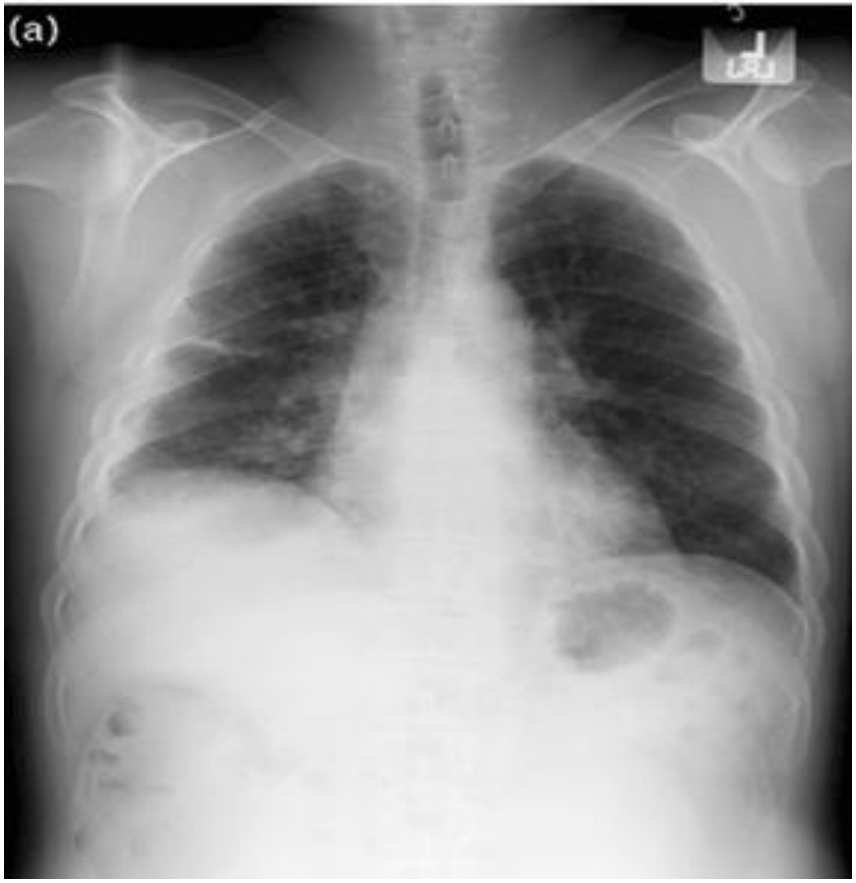
- Pulmonary fibrosis
- Pulmonary hypertension
- Asthma
- COPD
- VHD/CHD

- What next?

How to investigate

Investigations

- Routine
  - Blood CP
  - RFTs
  - LFTs
  - Coagulation profile
  - CXR
  - ECG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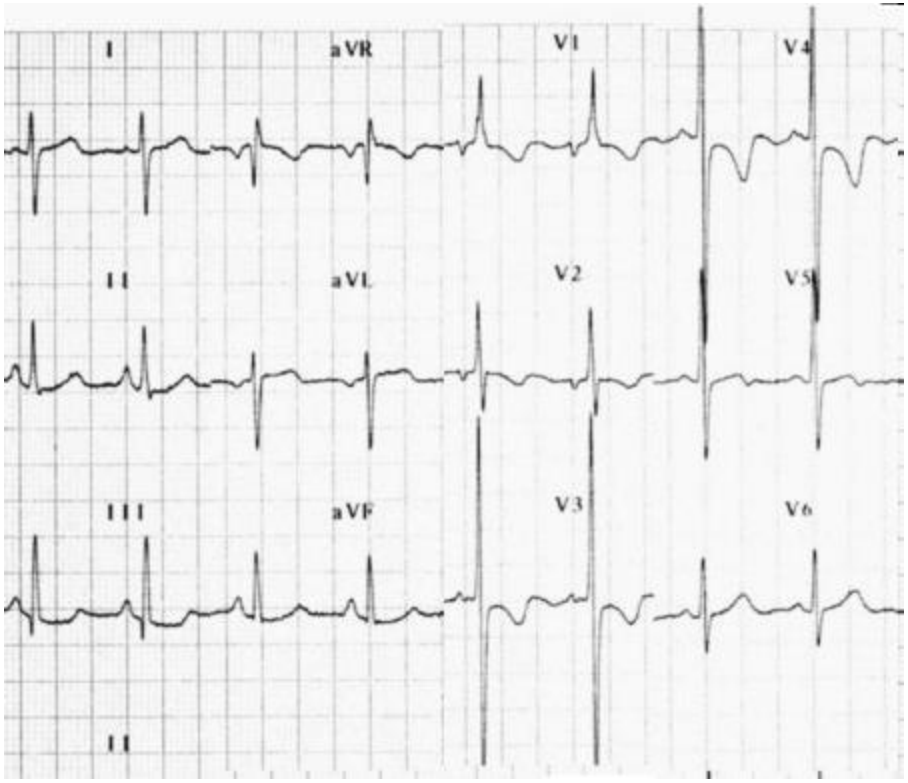
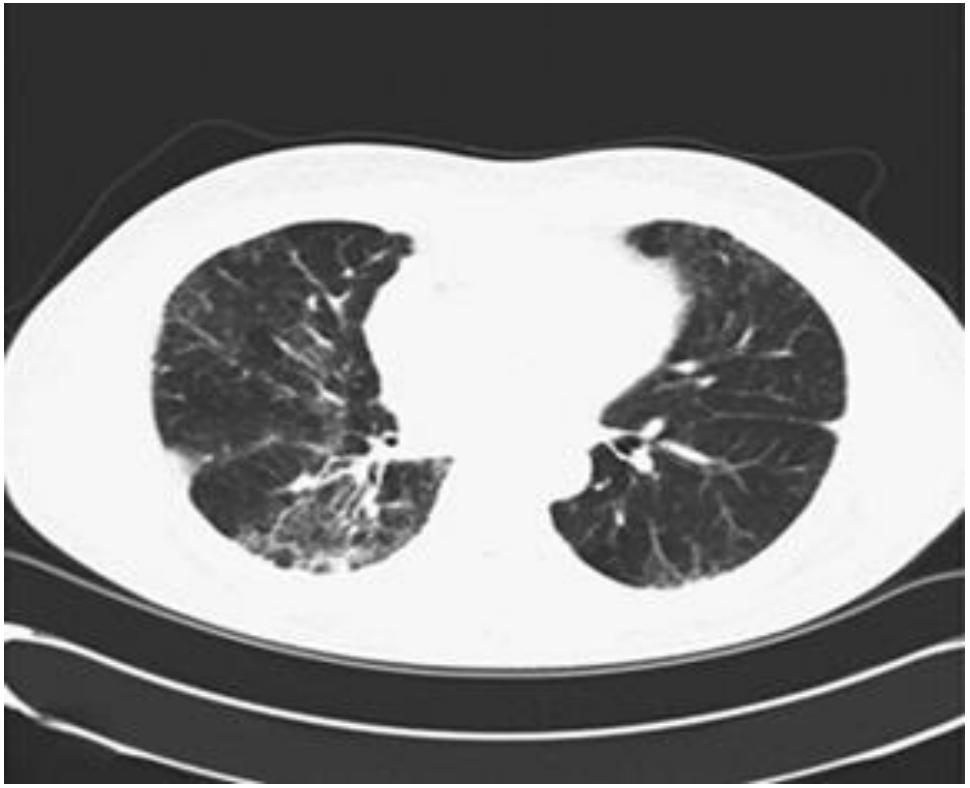

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II

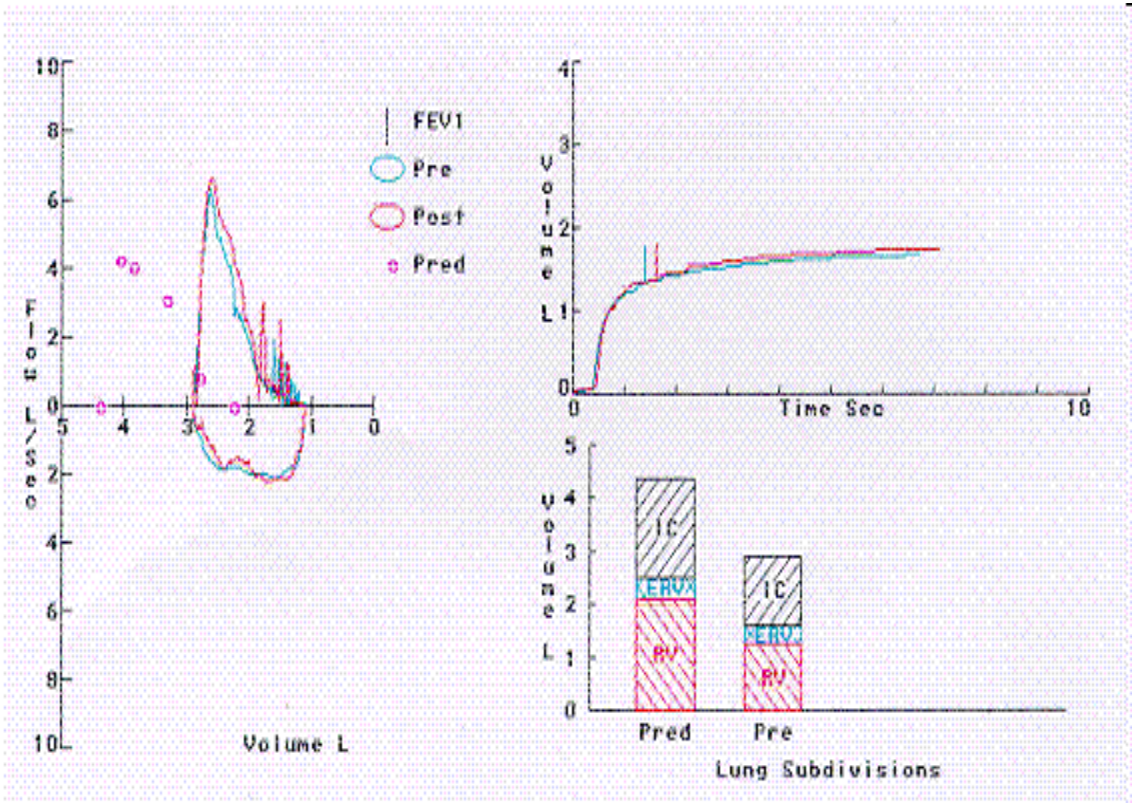
Other specific investigations

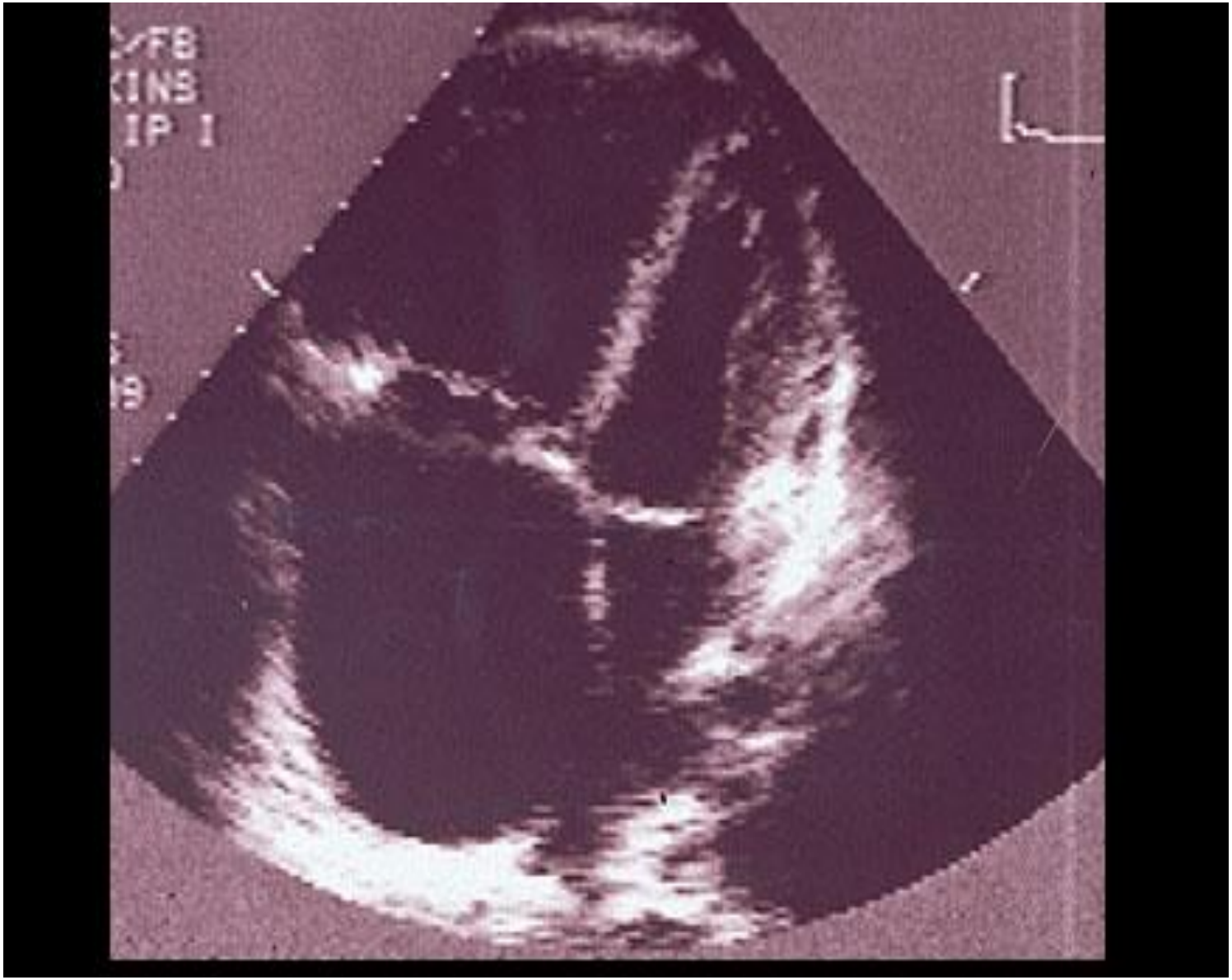
- ??

Other specific investigations

- CT scan
- CTD screening
- PFTs
- Echo




FEV1
Pre
Post
Pred
Flow L/Sec
Volume L
Volume L
Time Sec
Volume L
IC
ERV
RV
Pred
Pre
Lung Subdivisions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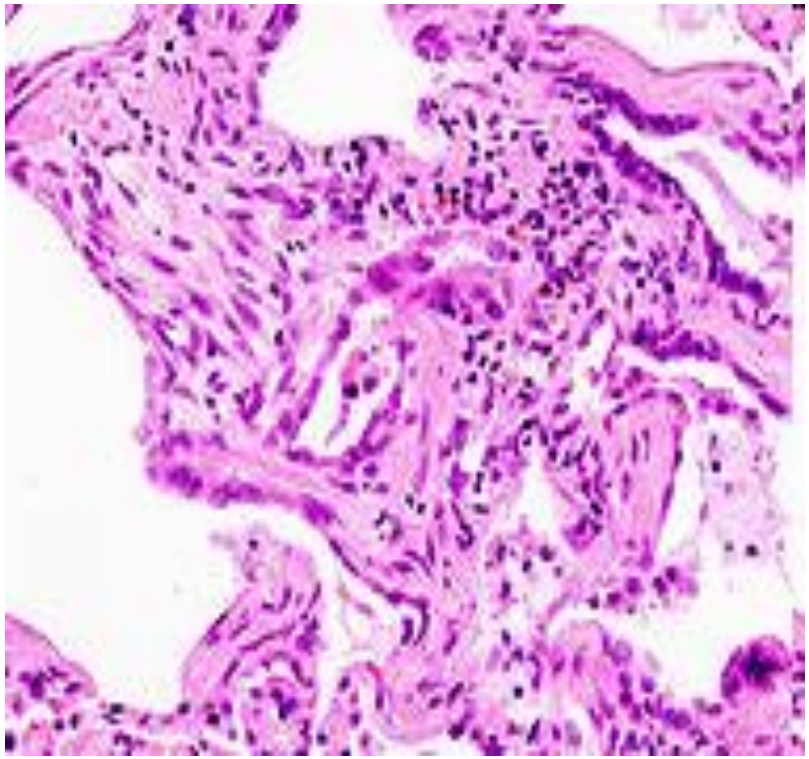
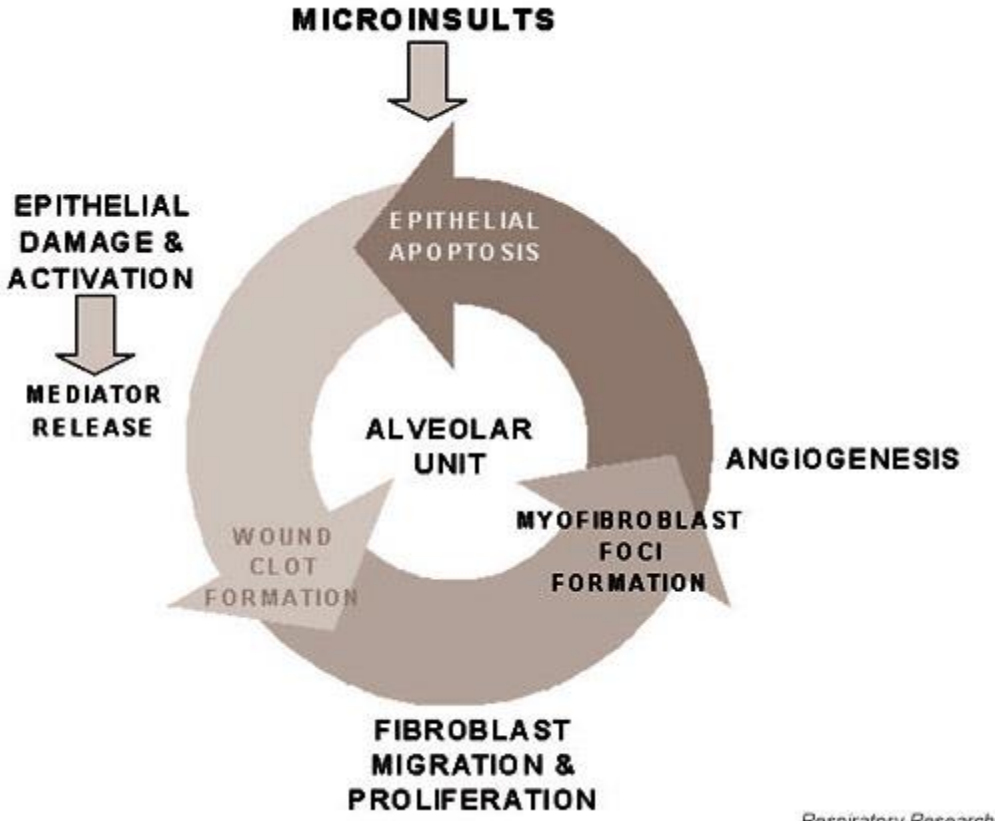
KINS
IP I



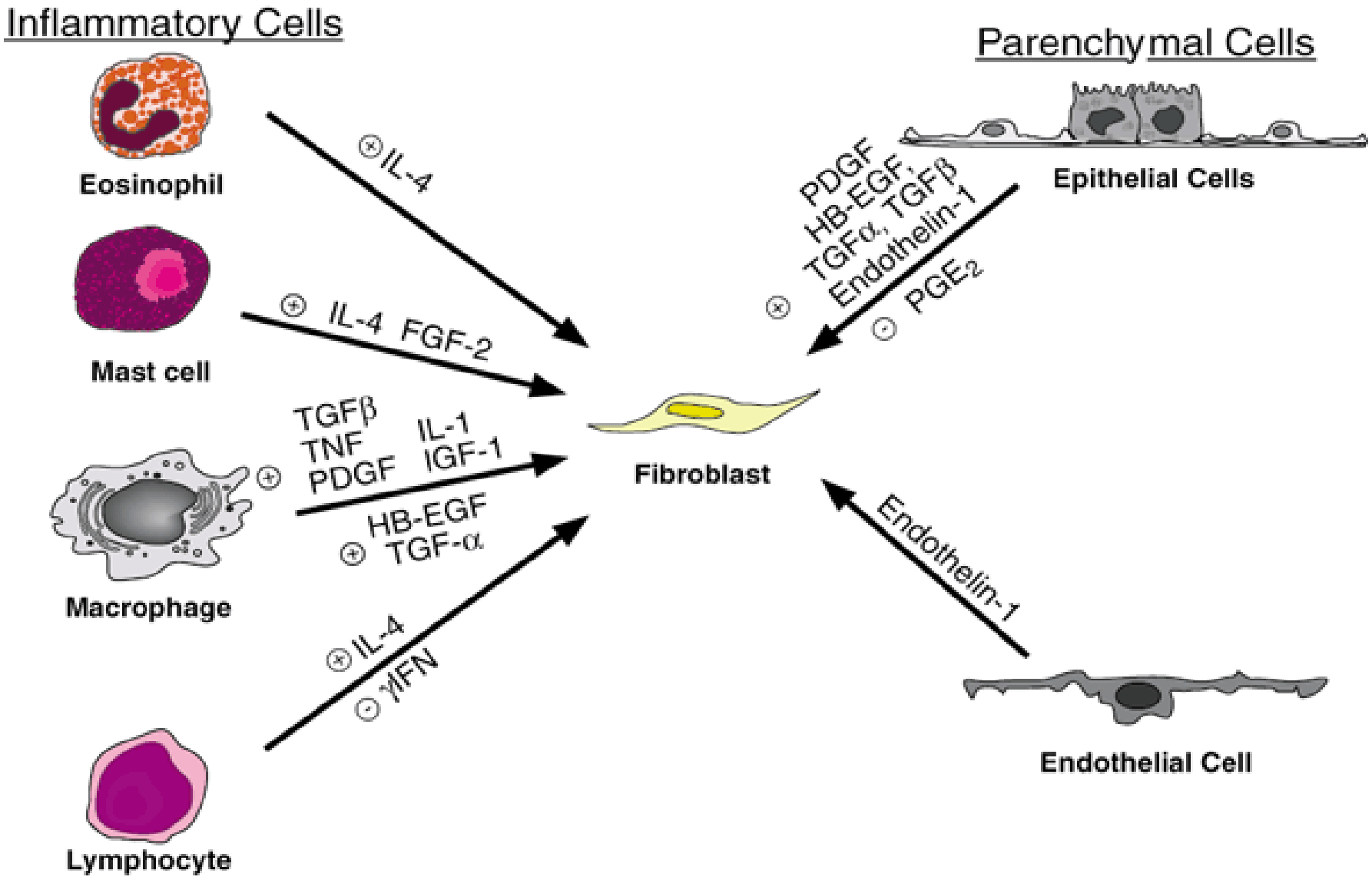
Diagnosis

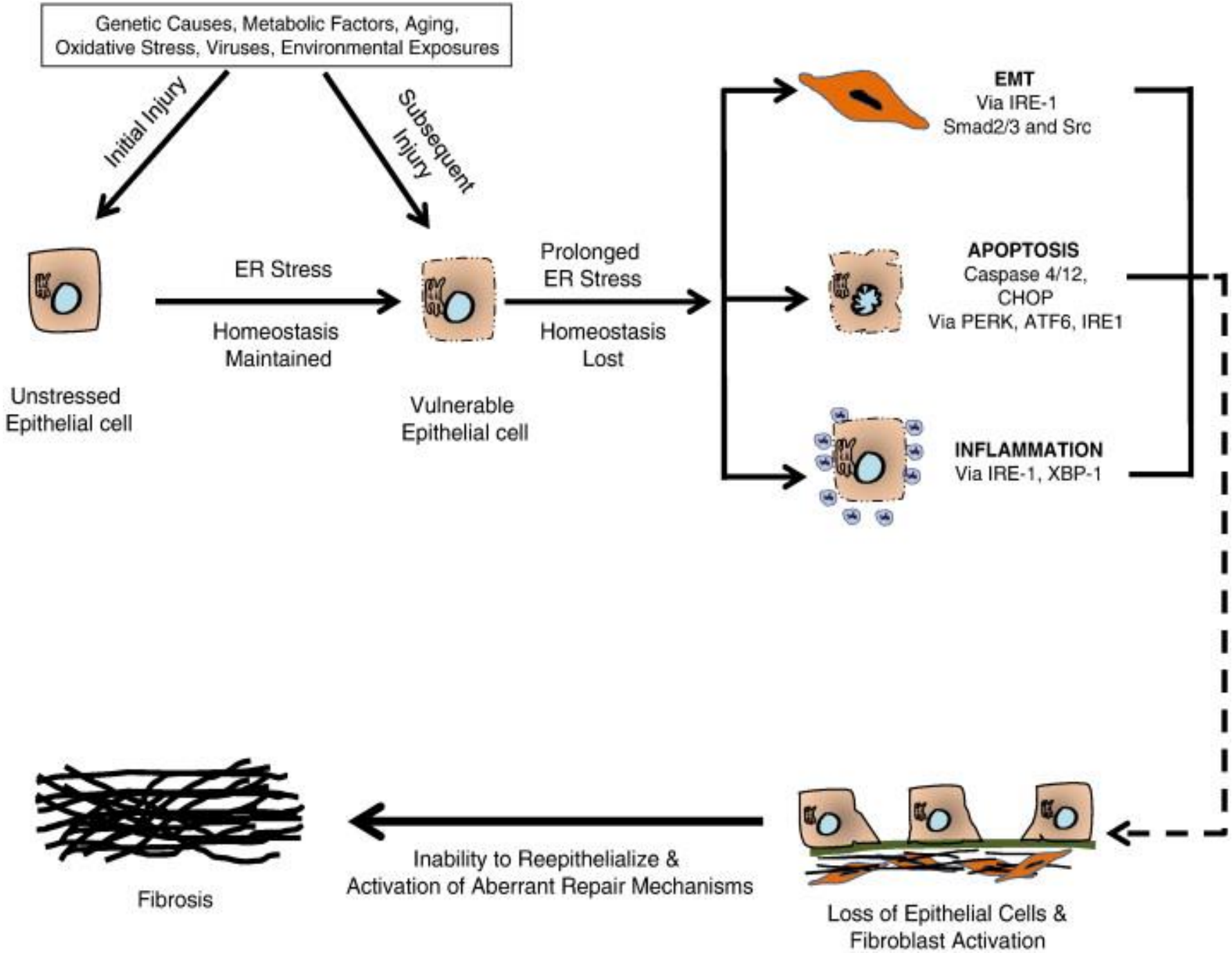
IPF



Respiratory Research

Pulmonary Fibrosis




Genetic Causes, Metabolic Factors, Aging,
Oxidative Stress, Viruses, Environmental Exposures
Initial Injury
Subsequent Injury
ER Stress
Homeostasis Maintained
Unstressed Epithelial cell
Vulnerable Epithelial cell
Prolonged ER Stress
Homeostasis Lost
EMT
Via IRE-1
Smad2/3 and Src
APOPTOSIS
Caspase 4/12,
CHOP
Via PERK, ATF6, IRE1
INFLAMMATION
Via IRE-1, XBP-1
Loss of Epithelial Cells &
Fibroblast Activation
Inability to Reepithelialize &
Activation of Aberrant Repair Mechanisms
Fibrosis

When to suspect?

- Age over 45 years
- Persistent breathlessness on exertion
- Persistent cough
- Bilateral inspiratory crackles
- Clubbing of the fingers
- Normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern.

Magnitude

- 10-20 and 7-10 cases per 100,000 persons, respectively

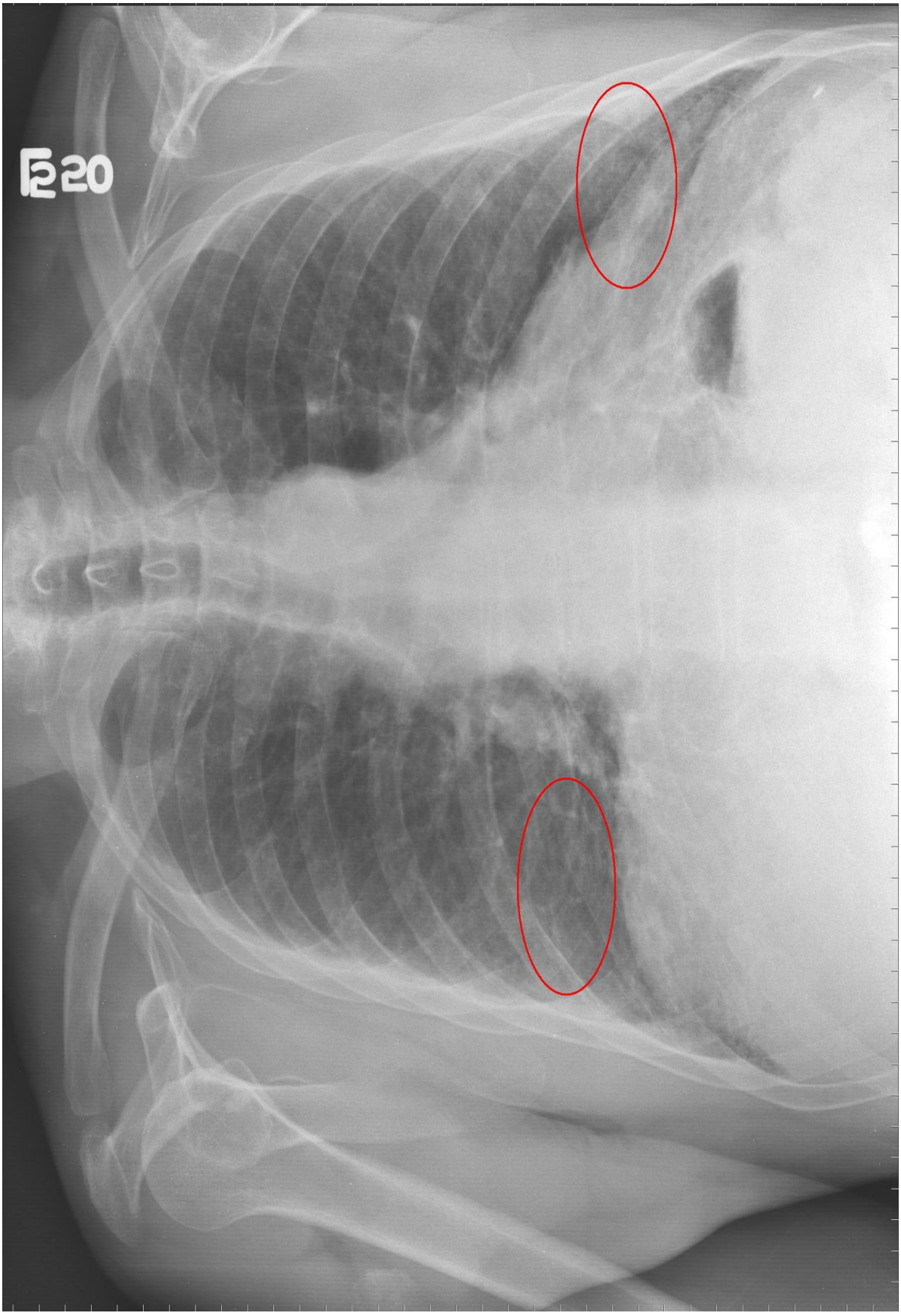
Age and gender

- Older
- Prevalence: 20 cases per 100,000 persons for males and 13 cases per 100,000 persons for females

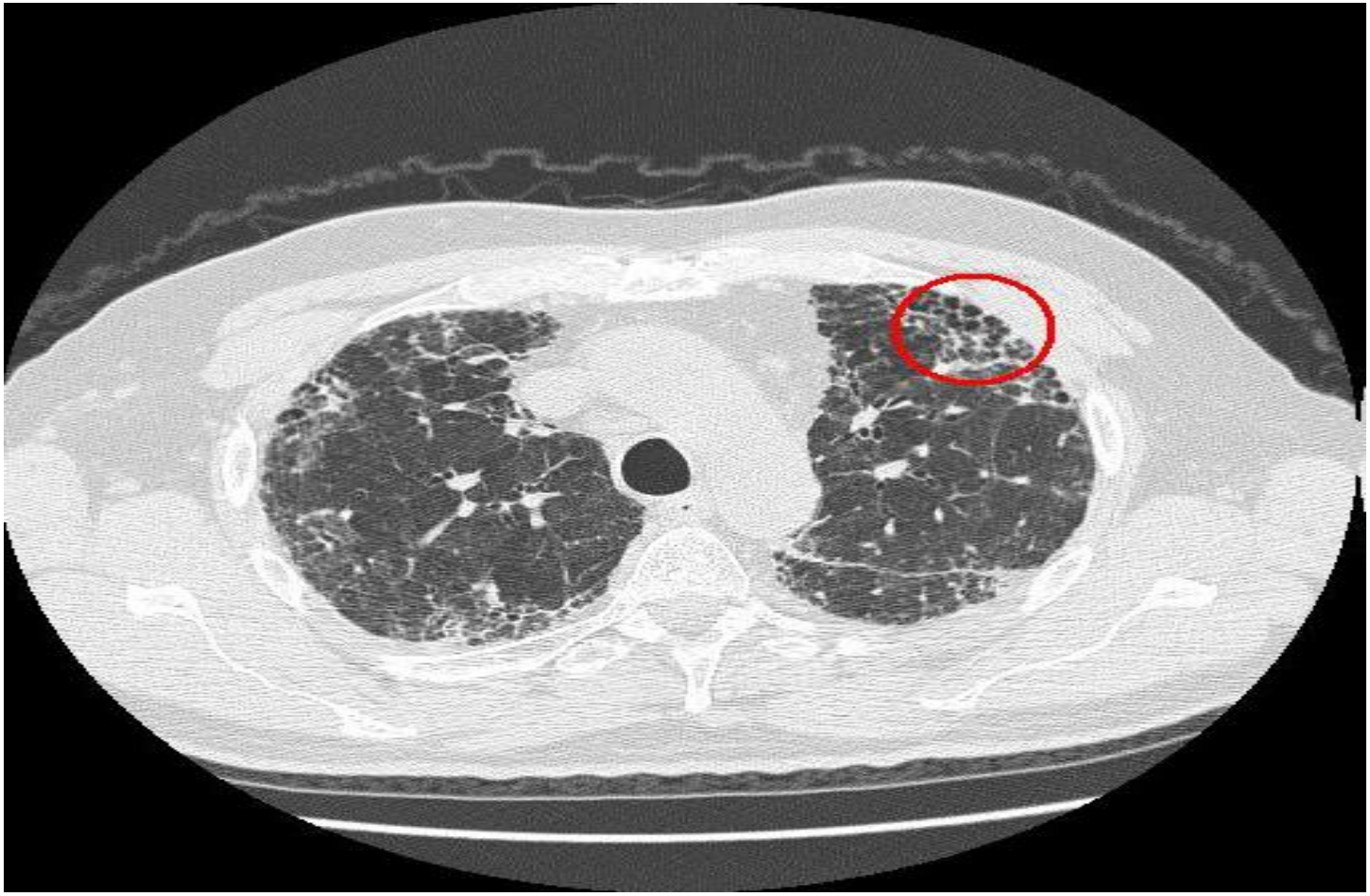
Clinical features

Investigations

Xrays


F220

CT scan



Treatment

- Steroides
- Immunospressive agents
- Anti Fibrotics
  - Pirfenidone
  - Nintedanib
- Antioxidants
- Lung transplantation

Prognosis

The following factors are associated with worse prognosis:

- Older age
- Male sex
- Cigarette smoking
- Higher predominance of honeycombing on the lung HRCT scan
- Lower diffusing capacity on pulmonary function test results at the time of diagnosis
- Higher rate of acute exacerbations of IPF
- Presence of pulmonary hypertension

Thank u