Interstitial Lung Disease

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Disclosures

No relevant financial disclosures

Most of the therapies we will discuss are off-label

Objectives

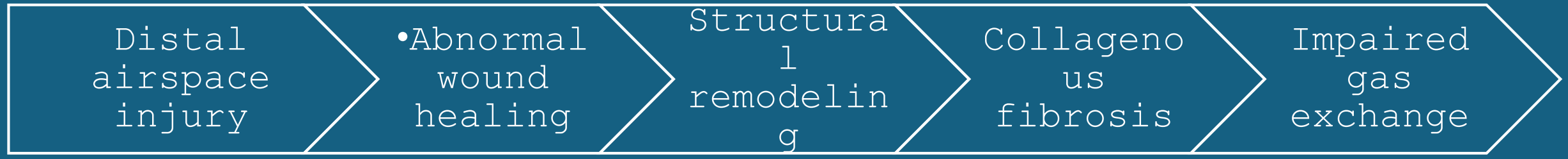
- **Understand** the definition, classification, and common etiologies of ILD.

- **Recognize** clinical presentations suggestive of ILD, **differentiate** between various types of ILDs, and **order** appropriate diagnostic tests.

- **Discuss** the general principles of managing interstitial lung disease.

Pathophysiology of ILD

- Inflammatory process



Exam and Work-up

- HPI: shortness of breath, dry cough
- PE: dry inspiratory crackles, digital clubbing, cyanosis
- Imaging: CXR, HRCT
- Other studies: PFT, biopsy
- Most important clues to etiology will be in history: age, gender, medical history, medications, smoking, occupation, hobbies, pets, family history, duration of symptoms

CXR

Volume loss

Reticular changes

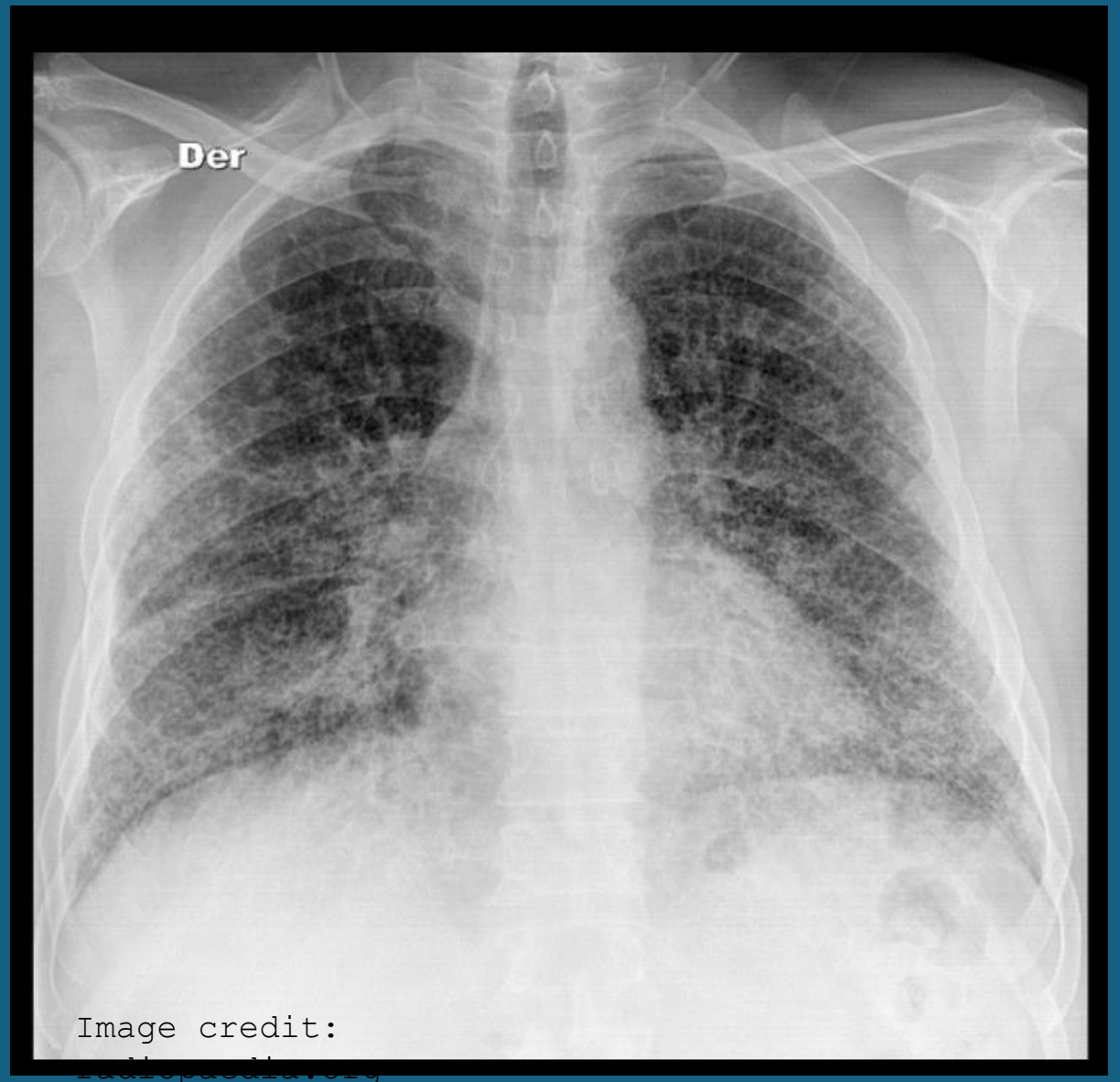


Image credit:

High resolution CT

UIP pattern

- Asymmetric, subpleural basilar predominance, honeycombing, traction bronchiectasis, architectural distortion/volume loss
- No groundglass opacities or micronodules

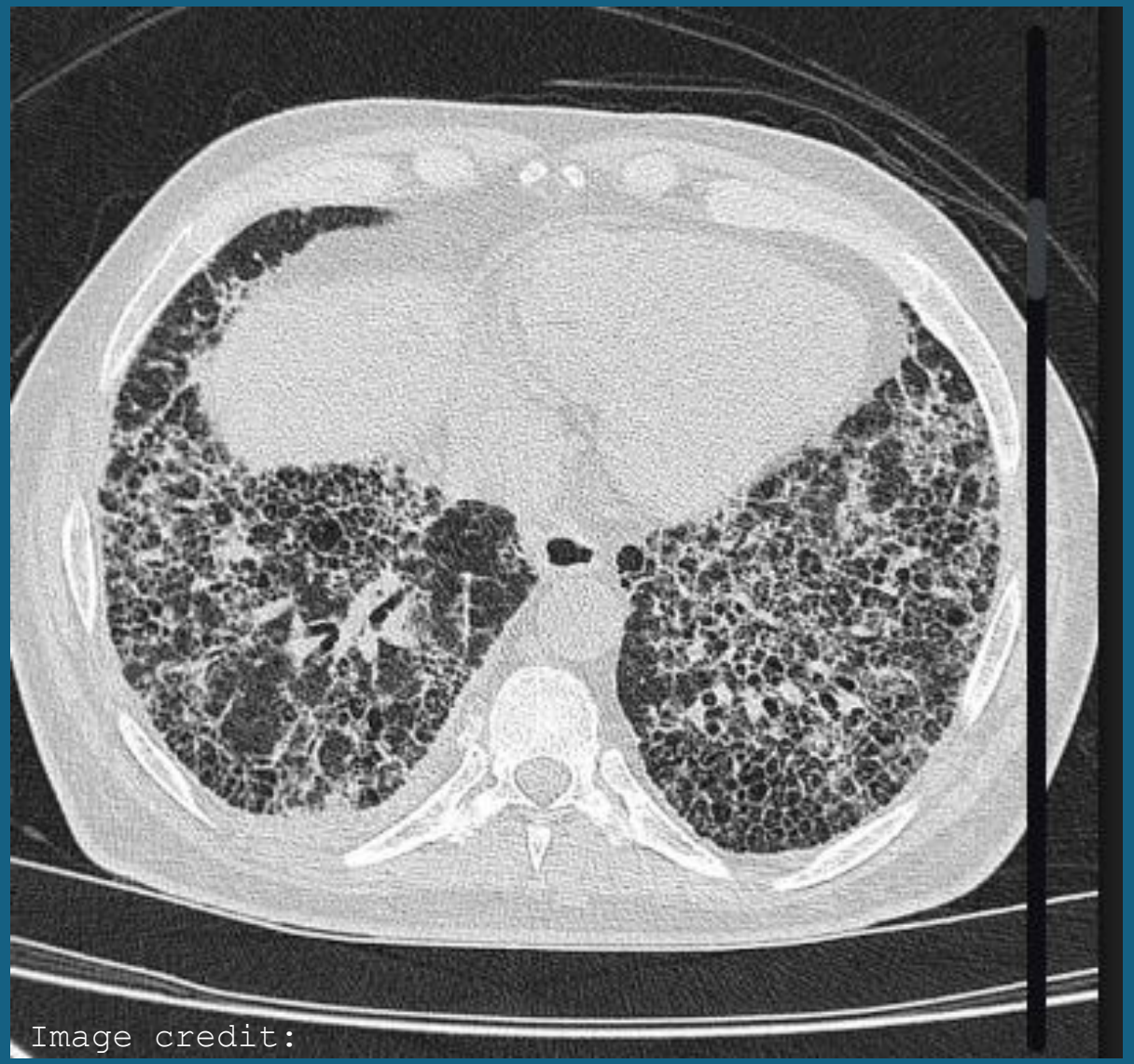


Image credit: radiopaedia.org

High resolution CT

NSIP pattern

- Fibrotic or cellular
- Groundglass opacities, reticular opacities, typically symmetric, subpleural sparing possible

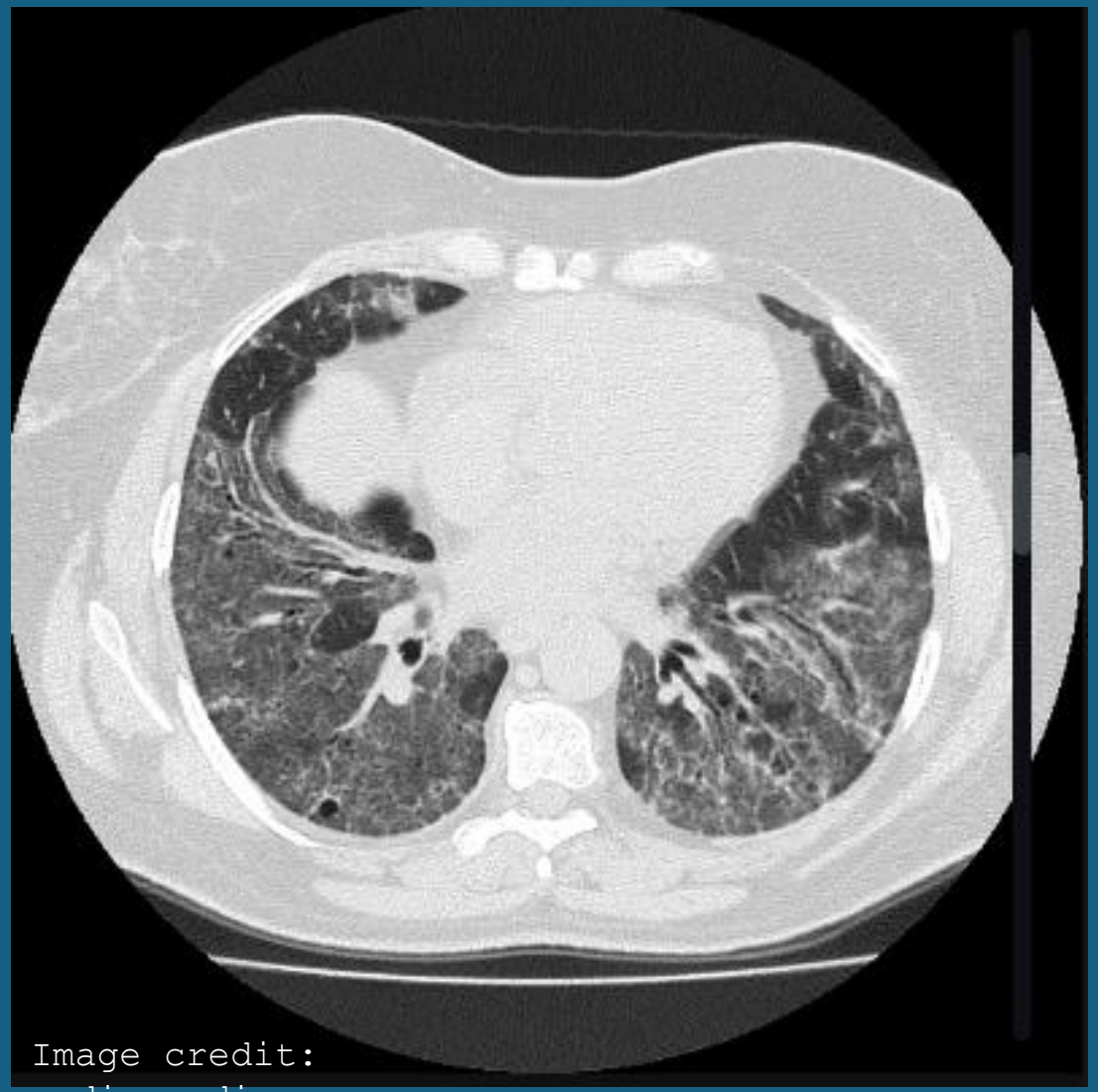
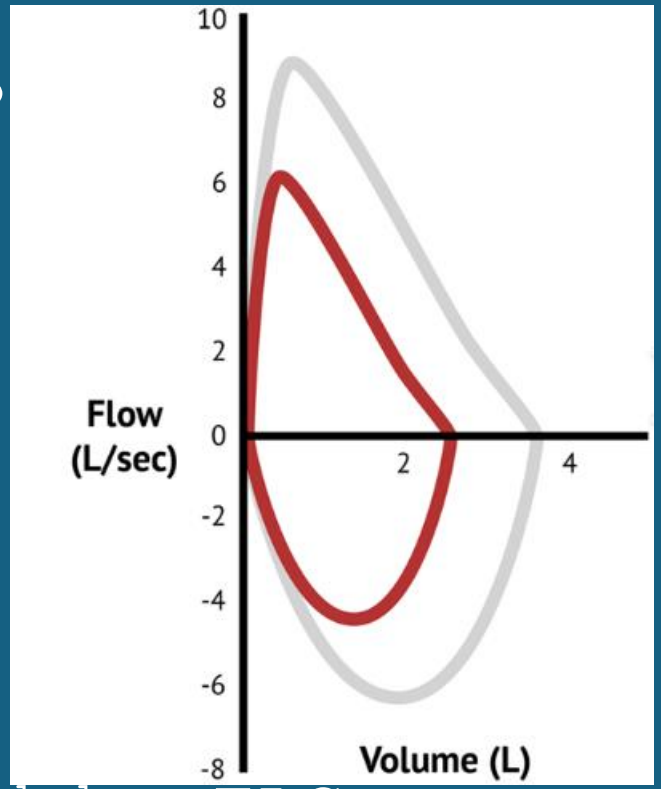


Image credit: radiopaedia.org

Pulmonary Function Tes

- Restrictive pattern
  - FEV1/FVC ratio normal to high
  - FEV1 and FVC low
  - Lung volumes reduced: TLC, RV
- Severity of restriction determined by TLC
- Reduced diffusion capacity



“Common” Types of ILD

Idiopathic Pulmonary Fibrosis (IPF)

Connective Tissue Disease-associated ILD (CTD-ILD)

Hypersensitivity Pneumonitis (extrinsic allergic alveolitis)

Pneumoconioses (asbestos, coal, silica, beryllium)

Smoking related ILD

Radiation-induced

Drug-induced

“Common” Types of ILD

Idiopathic Pulmonary Fibrosis (IPF)

- UIP pattern
- GGO during exacerbations
- Diagnosis of exclusion
- Typically males 50-80yo
- Median survival 3 years

“Common” Types of ILD

Hypersensitivity Pneumonitis (extrinsic allergic alveolitis)

- Numerous antigen triggers (moldy hay, bird feces, grain dust, HVAC)
- Presents as recurrent “pneumonia”
- Symptoms temporal to exposures
- Can progress to fibrotic ILD
- Tx: Antigen avoidance

"Common" Types of ILD

Pneumoconiosis

- Asbestos – shipyards, roofing, mechanic
  - Basilar bronchiectasis, interstitial thickening, 20-40y latent period
- Coal dust/mines
- Silica - sandblasting, brickyards, tile, glass
  - Upper lobe predominant, micronodules, hilar eggshell calcifications
  - Progressive massive fibrosis
- Beryllium – electronics, ceramics, dental fillings
  - Sarcoid mimic with hilar adenopathy

“Common” Types of ILD

Smoking related ILD

- Respiratory bronchiolitis associated (RB-ILD)
  - Patchy GGO, vague nodules
- Desquamative interstitial pneumonia (DIP)
  - GGO, reticular opacities, cysts, honeycombing
- Mixed fibrosis and emphysema
  - Upper lobe emphysema, basilar fibrosis, preserved spirometry/lung volumes
  - High incidence of PH

“Common” Types of ILD

Radiation-induced

- Pneumonitis (4-12 weeks)
- Fibrosis (6-12 months)
- Organizing pneumonia
  - Can present outside radiation fields
  - Most common in breast cancer

“Common” Types of ILD

Drug-induced

- Amiodarone
- Nitrofurantoin
- Methotrexate
- Chemotherapy

ILD Complications

- Pulmonary fibrosis—reversible vs irreversible
- Respiratory failure
- Prognosis variable and depends on underlying cause

Case 1

- 74yo M cough and worsening SOB over 9 months.
- PMH Afib, HTN, DLD. On amiodarone, apixaban, lisinopril, amlodipine, simvastatin, current smoker with 40ppd history. Retired mechanic.
- AF, 145/79, HR 83, 16 RR, 86% on RA. Mild digital clubbing, inspiratory crackles bilaterally.
- CXR with reticular opacities
- What's your suspicion? What

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Case 2

- 60yo M with shortness of breath and dry cough for several weeks
- PMH HTN, DLD. Farmer. Non-smoker.
- Meds: lisinopril/HCTZ, atorvastatin
- AF, BPs 130s/90s, HR 84, RR 18, O2 94% on RA.
- PE: inspiratory crackles, tachypnea, no clubbing
- What's your suspicion? What do you do next?

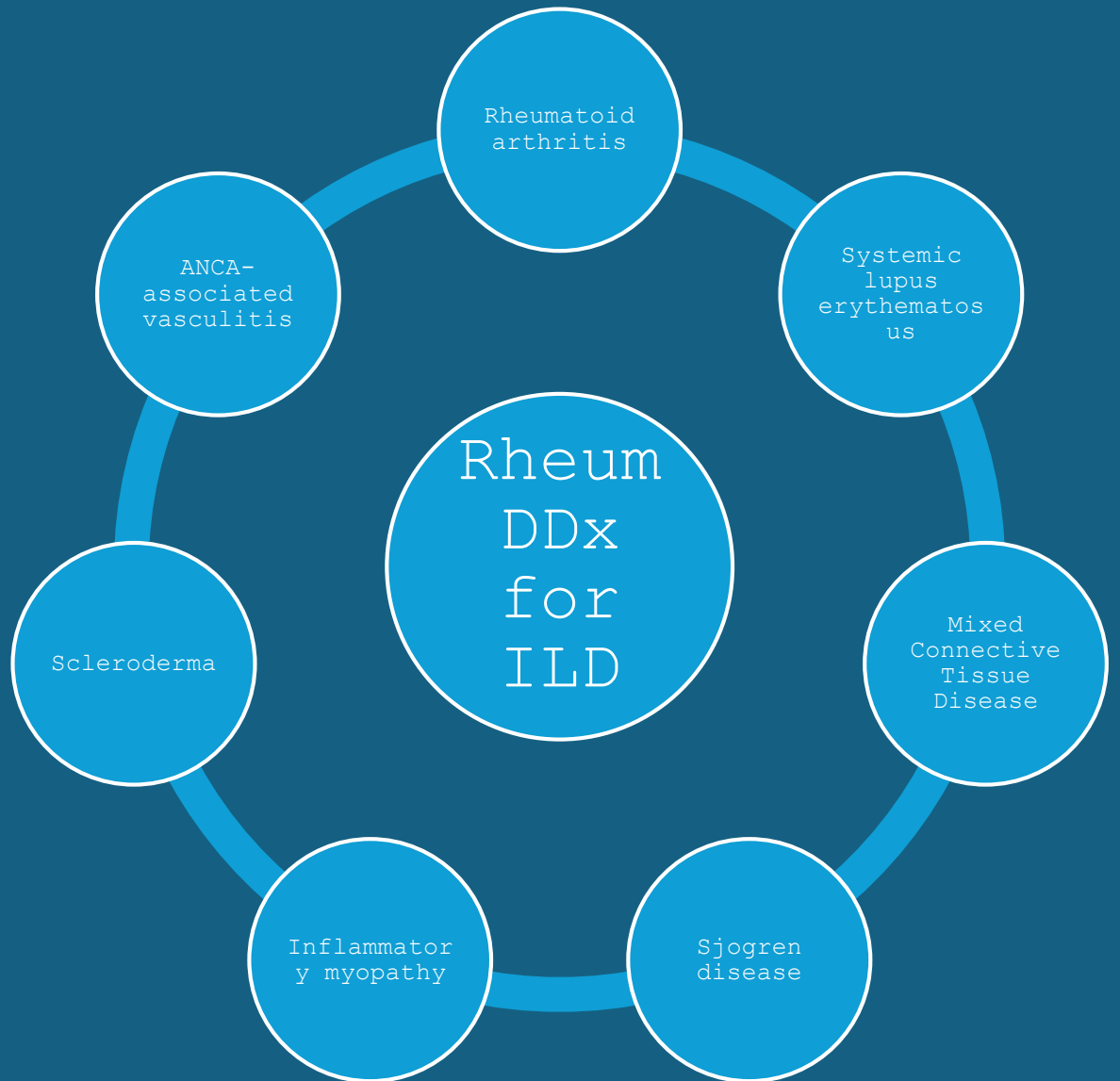
Case 3

- 67yo m SOB for 1 year
- PMH asthma, constipation. Albuterol and sodium docusate. Retired navy officer. 52pack year smoking history, quit 2years ago.
- PFTs with restriction.

- What's your suspicion? What do you do next?

Image credit: open

A Crucial Part of the Work-up: Identifying an Underlying Cause



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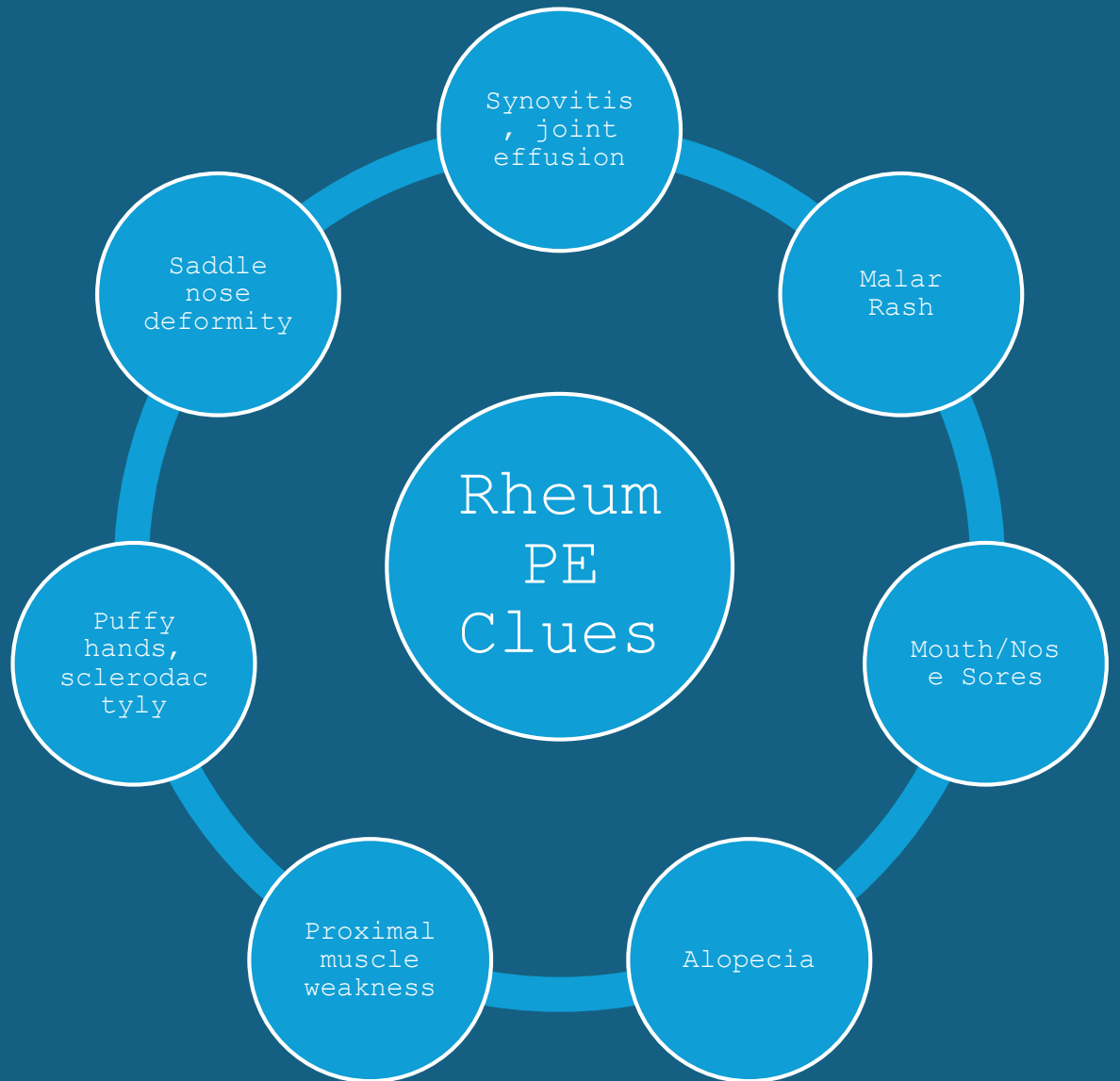

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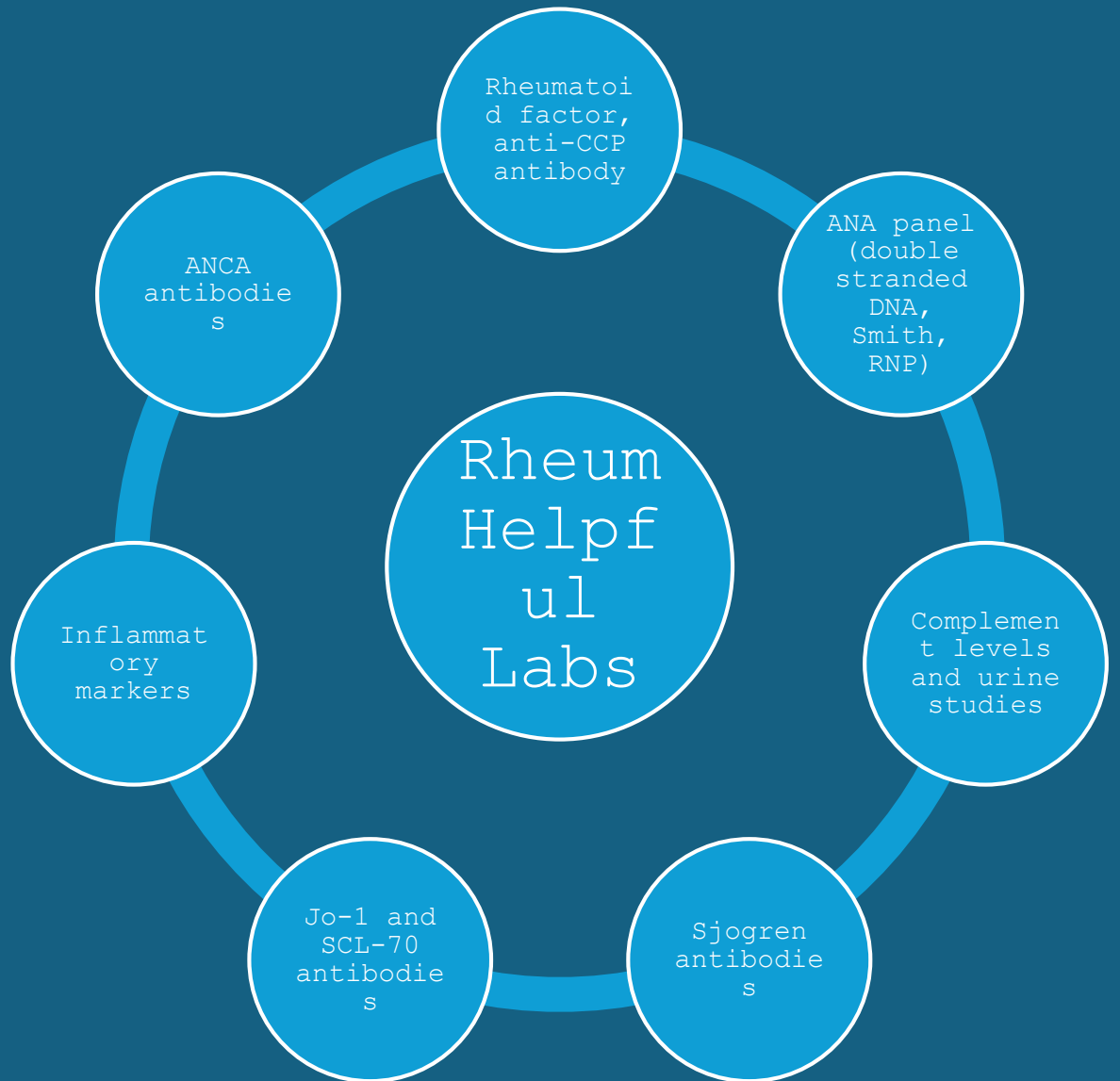


Case 4

- 39yo F SOB x1year, now with rest.
- No occupational exposures, works in an office.
- AF, BP 125/82, HR 92, O2 90% on RA
- Exam with puffy hands, skin thickening on hands, forearms, decreased oral aperture
- What's your suspicion? What do you do next?

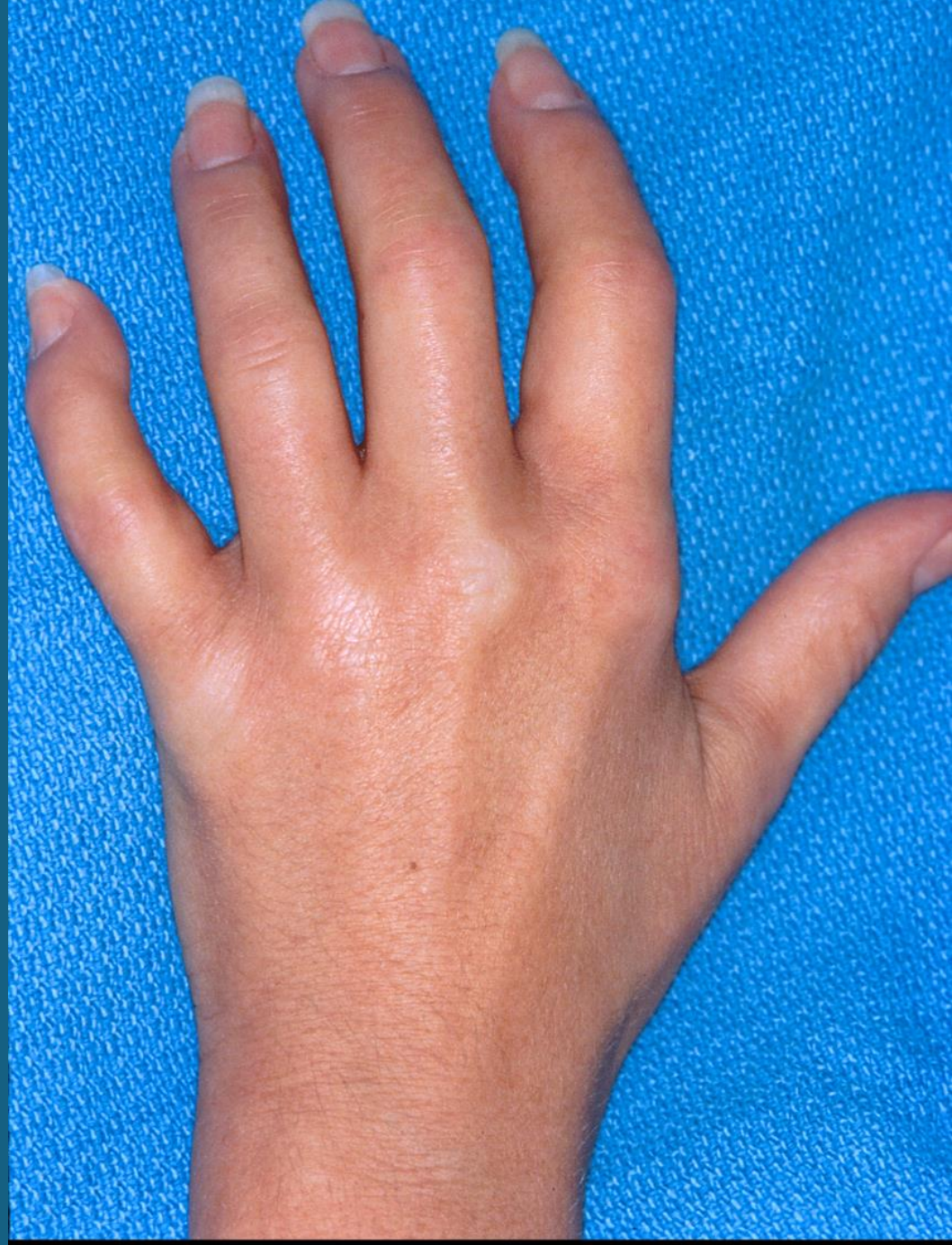
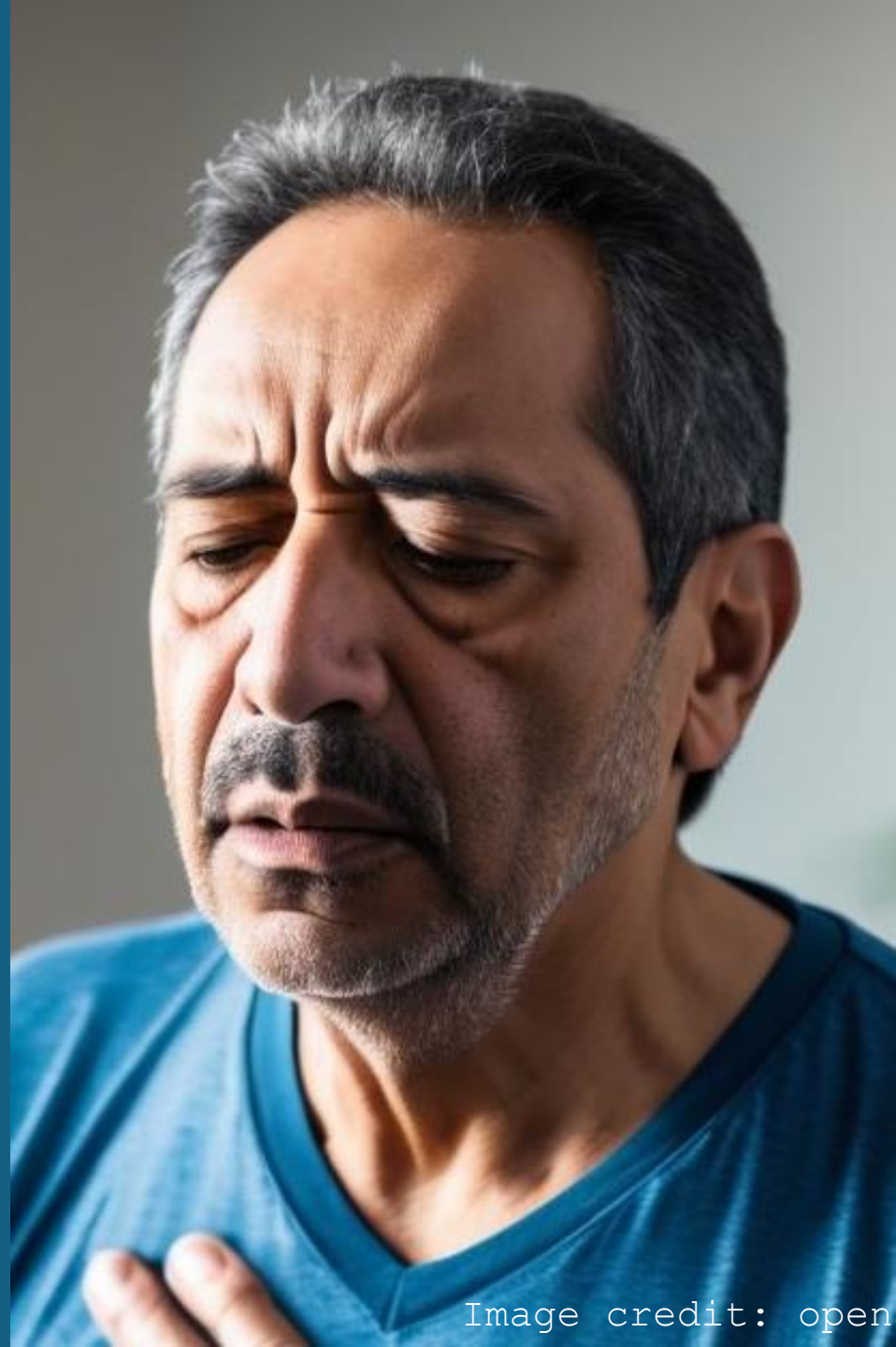


Image credit: ACR Image

Case 5

- A 59yo M with a 20year history of RA (on methotrexate, intermittently adherent) reports a 1year history of SOB with exertion but progressive
- 15ppd history but quit 5years ago.
- Exam with crackles on auscultation, active synovitis of MCPs and PIPs bilaterally.
- What's your suspicion? What do you do next?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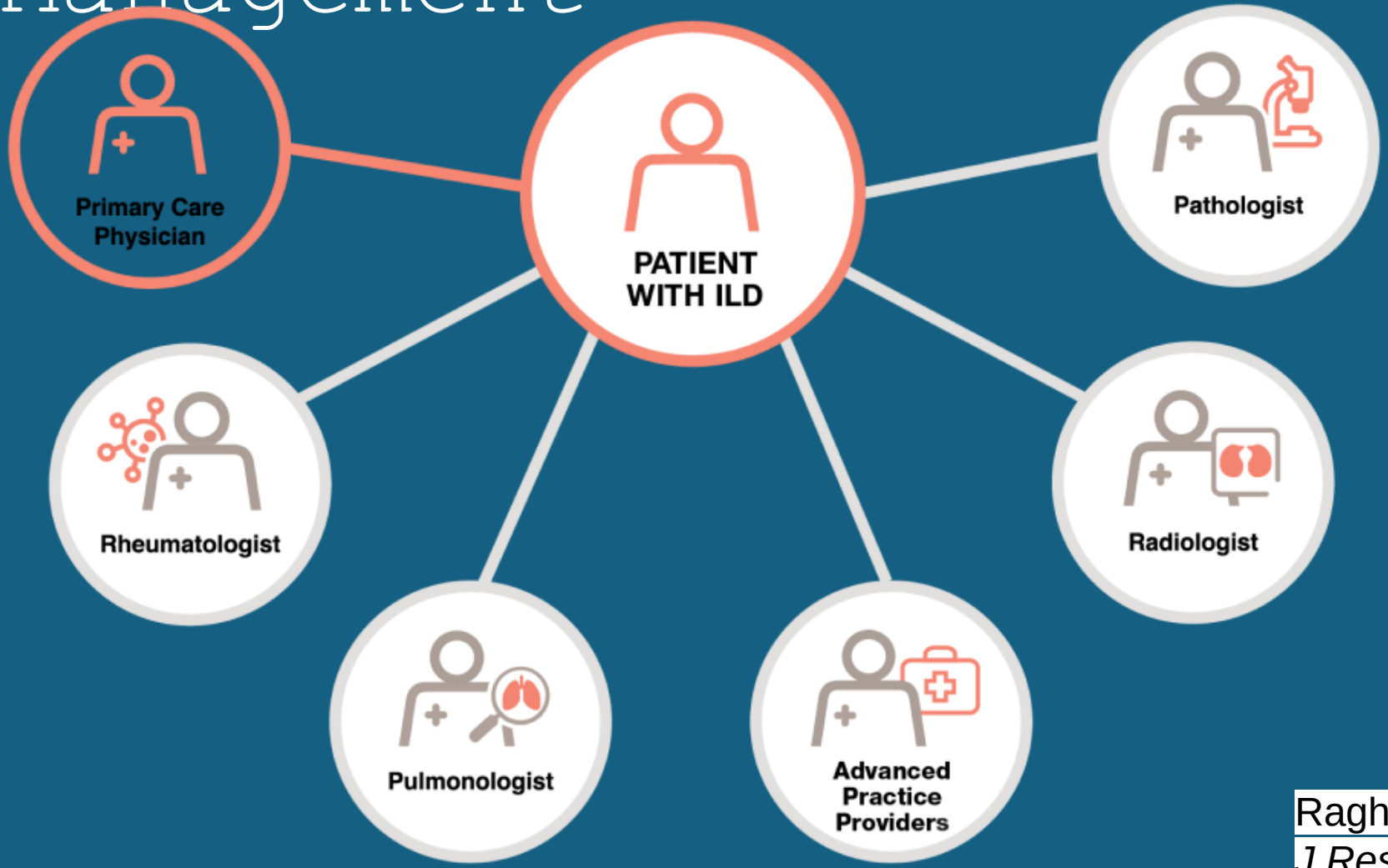
Multidisciplinary Diagnosis and Management



Raghu G et al; *Am J Respir Crit Care Med*. 2018.

Emerging Therapies: Anti-Fibrotic Agents

- IPF vs non-IPF
- Nintedanib – tyrosine kinase inhibitor
  - FDA-approved to treat IPF and scleroderma-related lung disease
  - Oral
  - Common GI adverse effects
- Pirfenidone
  - FDA-approved to treat IPF

Other Therapies: CTD-Targeted Agents

- Glucocorticoids
- Azathioprine
- Mycophenolate
- Methotrexate
- Rituximab
- Cyclophosphamide

Other Therapeutic Considerations

- PJP prophylaxis
- GI prophylaxis
- Pulmonary rehabilitation
- Oxygen
- Palliative care

Key Takeaways

- Dry inspiratory crackles on exam are unique to ILD
- High-resolution CT is the diagnostic test of choice for ILD
- Screening for serologic evidence of CTD in the setting of ILD is worthwhile.
- CTD-related ILD has a variety of treatment options
- Newer agents are becoming available for fibrotic ILD

Epilogue

- Known ILD with acute pulmonary decompensation:
  - Infection, infection, infection
  - Advanced fibrotic disease
  - If CTD-related ILD, consider immunosuppression

Yioe et al. Clin Med (Lond). 2021.

Questions

Photo credit: Dr. Allison Rogers