UCSF Health

The Journey to a Diagnosis: A Multidisciplinary Approach

Seminar for Patients, Caregivers, and Families

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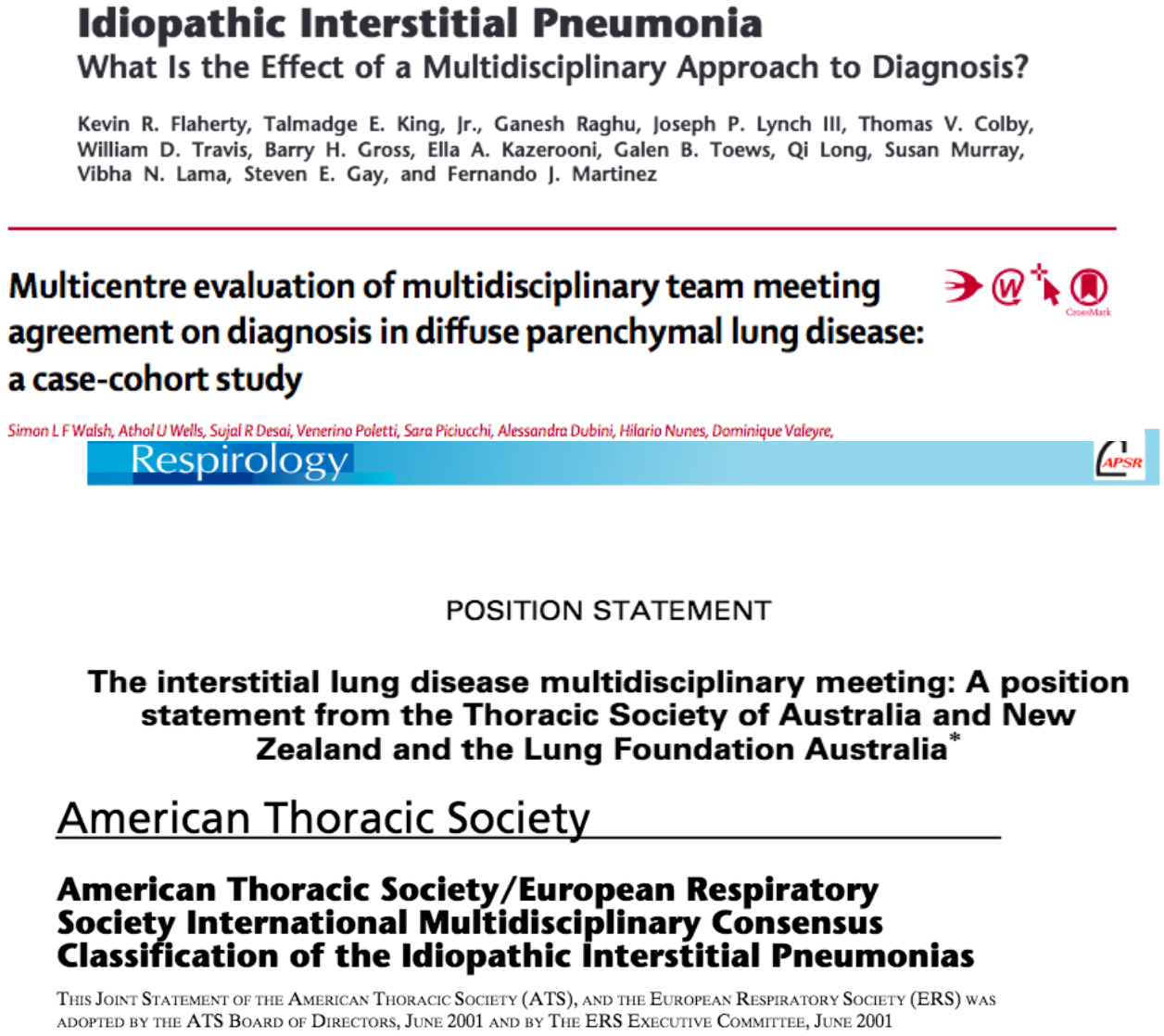
Disclosures

- I have nothing to disclose

Why multidisciplinary teams?

- Making an accurate diagnosis of a specific form of interstitial lung disease is challenging
- Pulmonary clinicians, radiologist and pathologists are traditionally separated in a hospital setting
- Agreement between providers and diagnostic confidence improve as more data is available
- Dynamic discussions lead to changes in diagnosis and improved confidence



Idiopathic Interstitial Pneumonia

What Is the Effect of a Multidisciplinary Approach to Diagnosis?

Kevin R. Flaherty, Talmadge E. King, Jr., Ganesh Raghu, Joseph P. Lynch III, Thomas V. Colby, William D. Travis, Barry H. Gross, Ella A. Kazerooni, Galen B. Toews, Qi Long, Susan Murray, Vibha N. Lama, Steven E. Gay, and Fernando J. Martinez

Multicentre evaluation of multidisciplinary team meeting agreement on diagnosis in diffuse parenchymal lung disease: a case-cohort study

Simon L F Walsh, Athol U Wells, Sujal R Desai, Venerino Poletti, Sara Piciucchi, Alessandra Dubini, Hilario Nunes, Dominique Valeyre,

Respirology

POSITION STATEMENT

The interstitial lung disease multidisciplinary meeting: A position statement from the Thoracic Society of Australia and New Zealand and the Lung Foundation Australia*

American Thoracic Society

American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias

THIS JOINT STATEMENT OF THE AMERICAN THORACIC SOCIETY (ATS), AND THE EUROPEAN RESPIRATORY SOCIETY (ERS) WAS ADOPTED BY THE ATS BOARD OF DIRECTORS, JUNE 2001 AND BY THE ERS EXECUTIVE COMMITTEE, JUNE 2001

What is a multidisciplinary discussion?

- Expert ILD clinicians, radiologists, and pathologists integrate all available clinical data, laboratory results, high-resolution computed tomography [HRCT] findings, and lung biopsy [when performed].
- Goals:
  - Diagnosis
  - Therapeutic decisions
  - Discussion of prognosis

Walsh et al. The Lancet Respiratory Medicine. 2016

Case Presentation

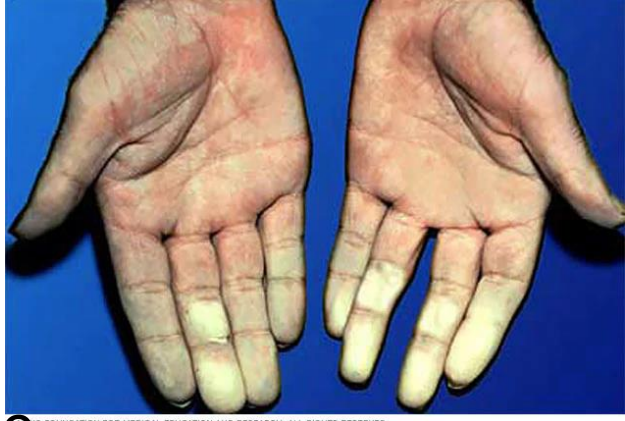
History

- 67 year old man with slow development of dry cough and shortness of breath. Recently diagnosed with IPF
- Medical History: atrial fibrillation, diabetes mellitus
- Medications: apixaban, metformin, pirfenidone
- Allergies: None
- Family history:
  - No family history of ILD, cryptogenic cirrhosis, aplastic anemia, myelodysplastic syndrome, leukemia
  - No family history of early graying

History: Exposures

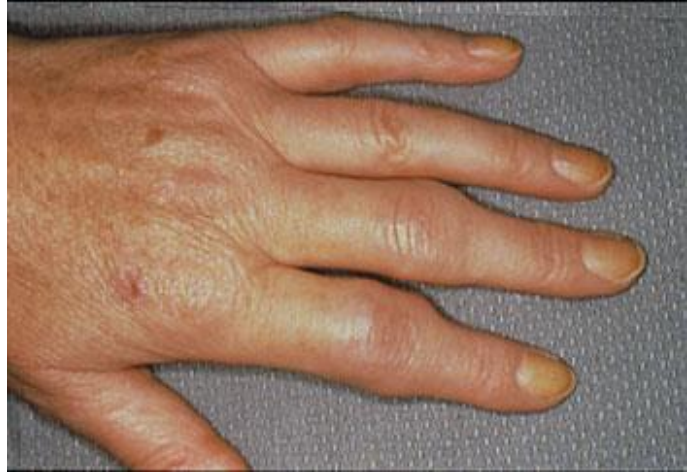
- Smoking:
  - Former smoker – smoked 1 pack per day for 30 years.
  - Quit 20 years ago
- Environmental:
  - Down comforter
  - Nightly humidifier use
  - Possible mold in work environment
- Occupational:
  - Retired general contractor

Review of Systems



- Constitutional: no weight loss, fevers, chills, night sweats, fatigue
- Head, Eyes, Ears, Nose, Throat: No dry eyes, dry mouth
- Respiratory: cough, shortness of breath; No pain with deep breaths, hemoptysis, or wheezing
- Cardiovascular: palpitations, no chest pain
- Gastrointestinal: no heartburn, no difficulty swallowing, no nausea, vomiting, diarrhea or constipation
- Musculoskeletal: back pain, joint pain in hands and fingers, chronic knee pain
- Skin: No rashes, Raynaud's

Physical Exam

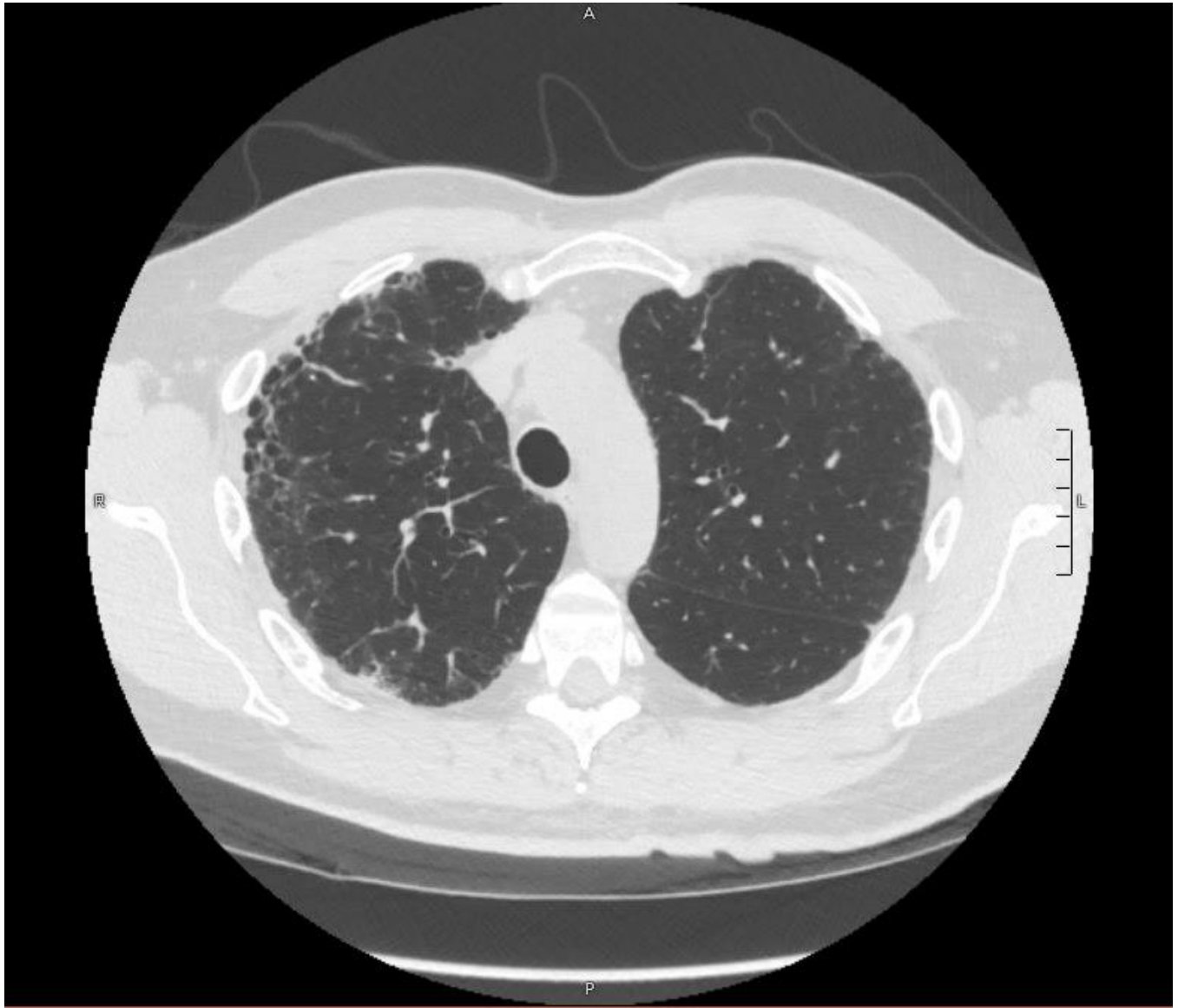


- Eyes: no redness/inflammation, normal vision
- Ears, Nose and Throat: normal salivary pool, good dentition
- Cardiovascular: Irregular heart rhythm, normal heart rate and sounds.

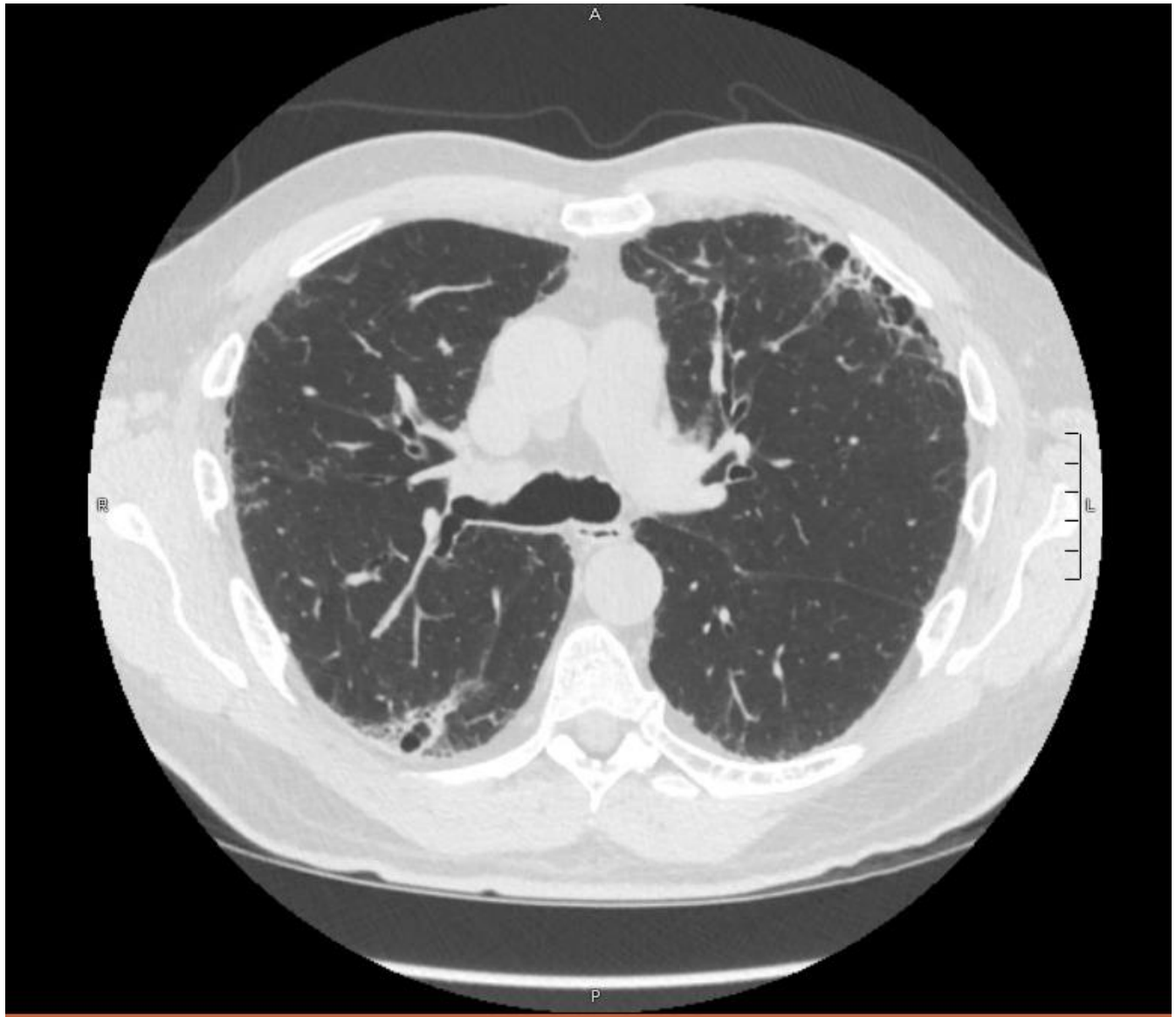
No leg swelling

- Respiratory: normal effort, no wheezes, rales, or rhonchi, crackles at the bases of the lungs
- Gastrointestinal: abdomen soft, nontender, no masses
- Skin: no rashes, skin dryness or cracking, no changes to the nail bed, thickening or tightening of the skin. No nodules. No clubbing
- Neuro: normal muscle strength, normal reflexes
- Psych: normal mood and affect
- Musculoskeletal: Swelling of the finger joints, right hand and right knee. Tenderness of both wrists and right knee, No warmth or redness.

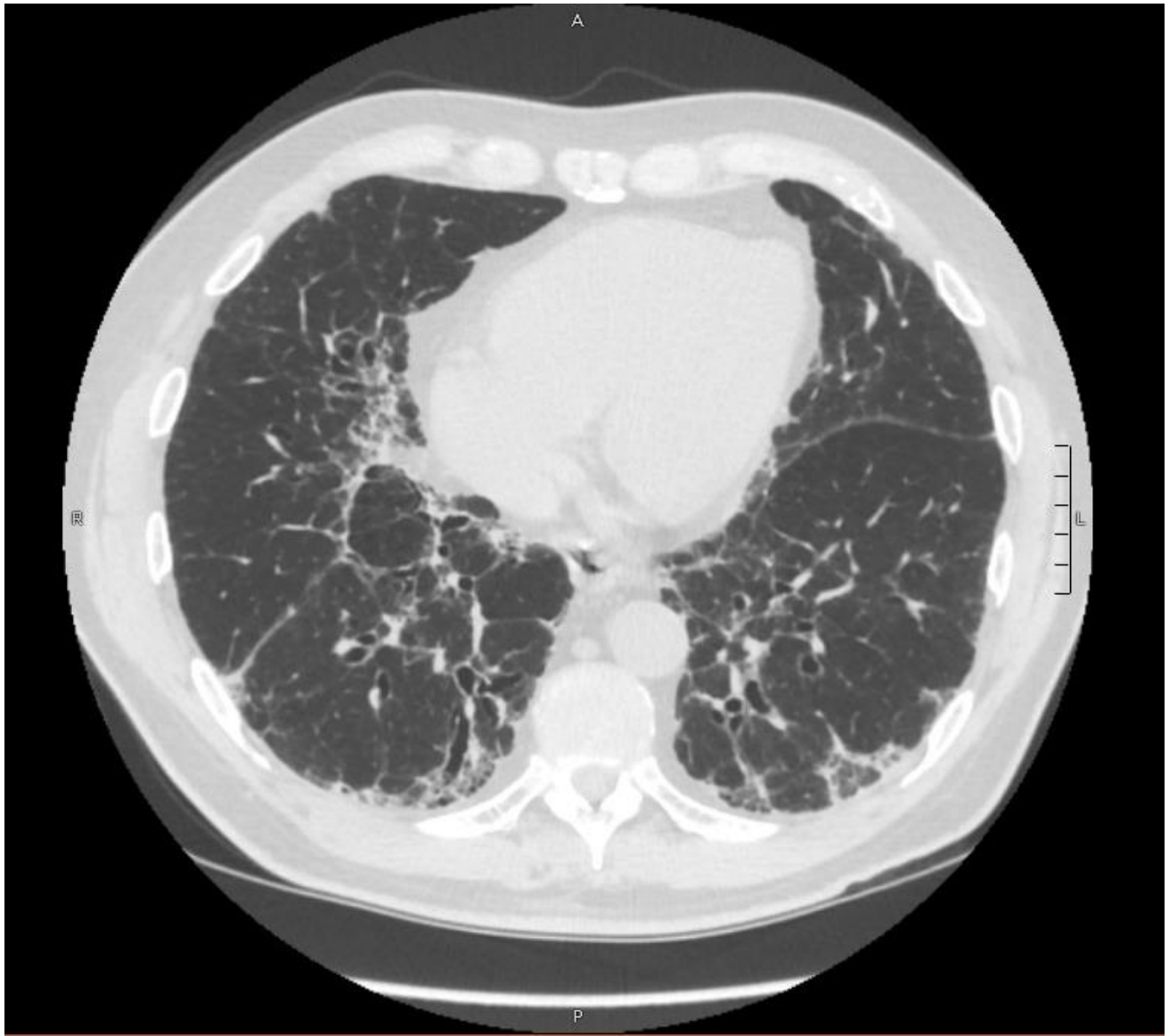
Diagnostic studies



Diagnostic studies



Diagnostic studies



Diagnostic studies

Information recap

- History:
  - 67 year old male, former smoker
  - Respiratory symptoms + joint pain
  - Down comforter use
  - No clear medication exposures
  - No family history or findings to suggest genetic predisposition
- Exam: crackles, joint tenderness and swelling
- PFTs: Normal lung volumes, reduced diffusion capacity
- HRCT: Definite UIP pattern
- TTE: Normal

Question for Multidisciplinary Conference

- What is the diagnosis?
- What are the next steps? Does he need a surgical lung biopsy?
- What is the appropriate therapeutic options? Should he continue pirfenidone?

What is the diagnosis?

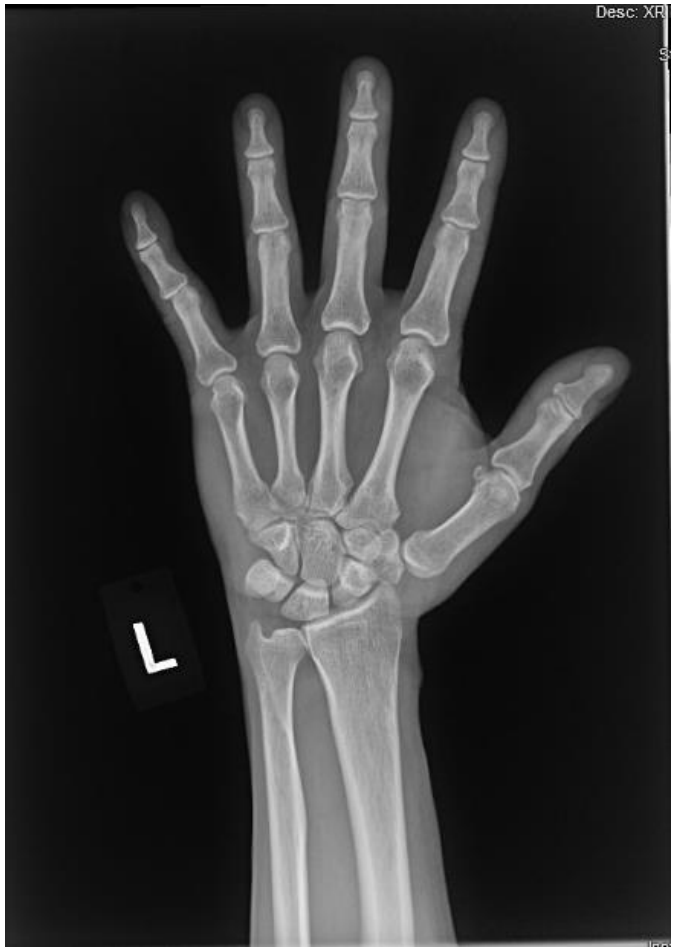
- Idiopathic Pulmonary Fibrosis
- Connective tissue-interstitial lung disease
- *Chronic Hypersensitivity Pneumonitis*

Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
   ANA - negative
   RF - positive
   Anti-CCP - elevated
   C-Reactive Protein - elevated

Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
2. Hand films



Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
2. Hand films
3. Rheumatology referral – inflammatory arthritis

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Rheumatoid Arthritis

Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
2. Hand films
3. Rheumatology referral – inflammatory arthritis

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Rheumatoid Arthritis

4. Surgical lung biopsy???

Rheumatoid Arthritis - Interstitial Lung Disease

- Rheumatoid arthritis is an inflammatory and autoimmune disease ~1% of the population
- Up to 60% of patients with RA have interstitial lung disease, but a minority of those patients will develop clinically significant disease
- Diagnosis:
  - Various histologic patterns
  - Various pathologic patterns
- Variable response to treatment and clinical course

Olson et al. Am J Respir Crit Care Med. 2011
Hyldgaard et al. Ann Rheum Dis. 2017.

Therapeutic Options: Anti-inflammatory

- Pretreatment:
  - Baseline blood work
  - Evaluate for infectious risks: hepatitis B and C, latent tuberculosis

Therapeutic Options: Anti-inflammatory

- Pretreatment:
- Types of Therapies
  - Rapid acting: NSAIDs and glucocorticoids
  - Corticosteroid sparing agents: mycophenolate mofetil, azathioprine, cyclophosphamide
  - Disease-modifying anti-rheumatic drug (DMARD): hydroxychloroquine, leflunomide*, methotrexate *
  - Biologics:
    - Anti-TNF therapy: etanercept*, infliximab*, adalimumab*
    - Abatacept
    - Rituximab

Therapeutic Options: Additional Therapies

- Pirfenidone and nintedanib?
  - Under investigation as a therapeutic option for patients with RA-ILD (ClinicalTrials.gov identifier: NCT02808871)

- Smoking cessation
- Supplemental oxygen
- Pulmonary rehabilitation
- Lung transplantation

Take Home Points

- Interstitial lung diseases are challenging to diagnose
- Increased information (clinical, physical exam, blood work, diagnostic testing) improves agreement amongst providers
- Multidisciplinary approach improves the accuracy and confidence of a diagnosis

Thank you for involving us in your care

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