

# Non-Specific Interstitial Pneumonia

Maria L. Padilla, MD

Professor of Medicine

Director Advanced Lung Disease Program

Icahn Sch. of Medicine at Mount Sinai



**Mount  
Sinai**

# COI

1. Consultant : Boehringer Ingelheim
2. Speaker: Genentech

# Non-Specific Pneumonia

## Objectives

**Upon Completion of this learning activity,  
participants should be able to:**

1. Recognize the clinical and radiographic presentation of NSIP
2. Distinguish NSIP from other Idiopathic Interstitial Pneumonias
3. Develop treatment strategies for management of NSIP

# NSIP- Chronology



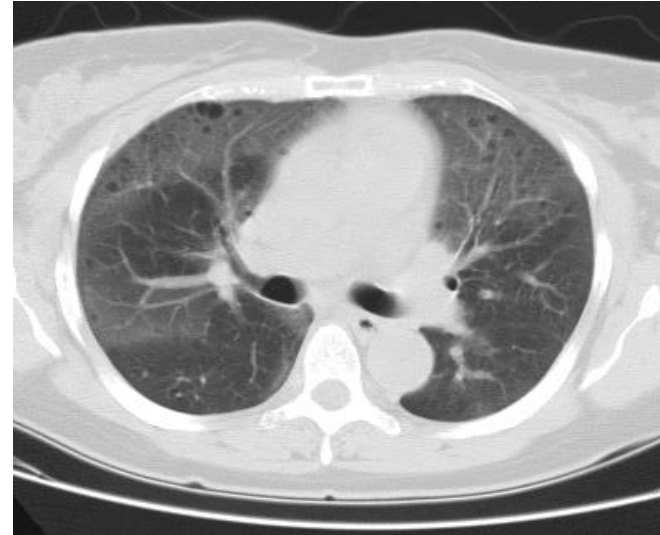
# IIP Radiographic Patterns

Which radiographic pattern is most consistent with NSIP?

A



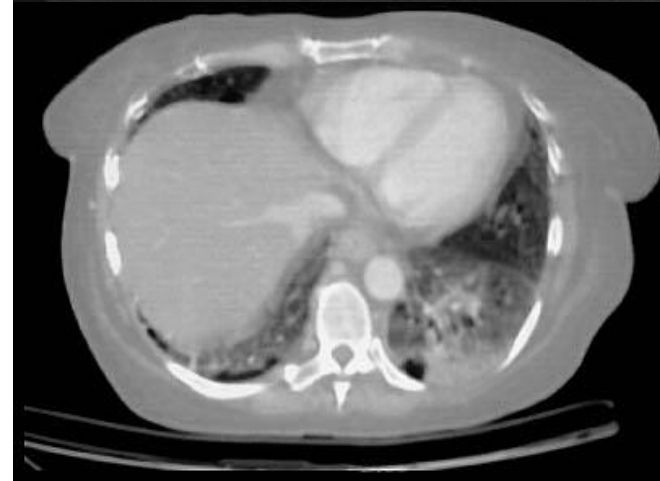
B



C



D



# Non-Specific Interstitial Pneumonia

1. Definition : NSIP is a chronic interstitial pneumonia that is characterized by a homogeneous appearance of with mild to moderate interstitial inflammation and varying degree of dense or loose interstitial fibrosis
2. Idiopathic
3. Associated with other diseases:
  1. CTD
  2. HIV
  3. Drug Toxicity
  4. HSP
  5. IgG4 Disease
  6. Familial IIP
  7. GVH disease

# Epidemiology of NSIP

1. 1-9/100.000; 14-36 % of IIP
2. Idiopathic: middle to later age female >>M, non- smoker
3. Associated NSIP no gender predominance
4. Usually responsive to Rx
5. Good Prognosis
  - A. Cellular iNSIP 100% at 5 y and 10 y
  - B. Fibrotic
    1. 5 y 86-92% (vs UIP 43%)
    2. 10 y 26-40% (vs UIP 15 %)

# NSIP Clinical Presentation

1. Subacute to chronic course
2. Cough and Dyspnea
3. Systemic symptoms: F, Flu-like illness, fatigue, weight loss
4. Physical exam
  1. Crackles
  2. Clubbing is infrequent (less than with UIP)
  3. Signs and symptoms related to associated disease : skin findings, fissures of fingers, Raynauds, Nail bed abnormalities, muscle weakness



# NSIP

## Evaluation

- HX
- PE
- Serology
- PFT's , 6MWT
- Echocardiogram
- Cxr, HRCT
- MDD
- Biopsy

# ILD PE: Rales and Clubbing but Connective Tissue Disease ...

TALKS TO THE  
HAND ....



AND WE SHOULD LISTEN!

# Gottron's Papules in Dermatomyositis



# Mechanic's Hands in Anti-Synthetase Syndrome



*Your PAH Resource*

# Raynaud's Phenomenon



*Your PAH Resource*



# Serological Evaluation

- ▶ ANA, RF, CCP
- ▶ Based on history & physical exam, consider:
  - Sjogren AB, Myositis Panel, Jo1 AB
  - Immuno-electrophoresis
  - Sclero 70 and Anti-centromere antibody, RNA polym
  - ESR & CRP, Creatine kinase, aldolase
  - MPO/PR3 (ANCA) antibodies
  - Anti-cardiolipin antibodies, lupus anticoagulant
  - +/-Hypersensitivity pneumonitis panel
- ▶ Should be performed before a biopsy

# Non Specific Interstitial Pneumonia

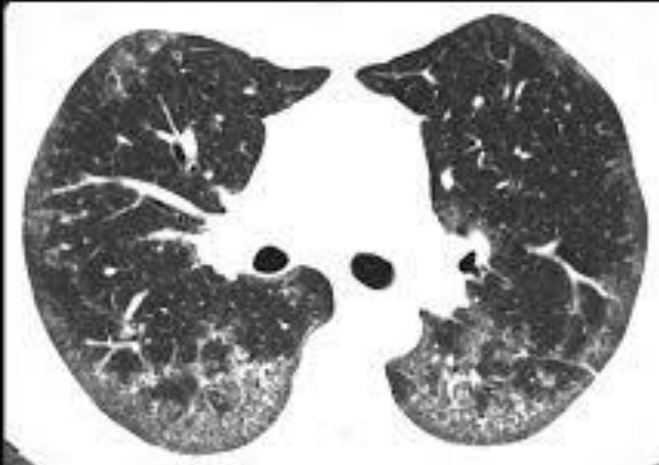
## ► Imaging

Antisynthetase syndrome  
3/5/17

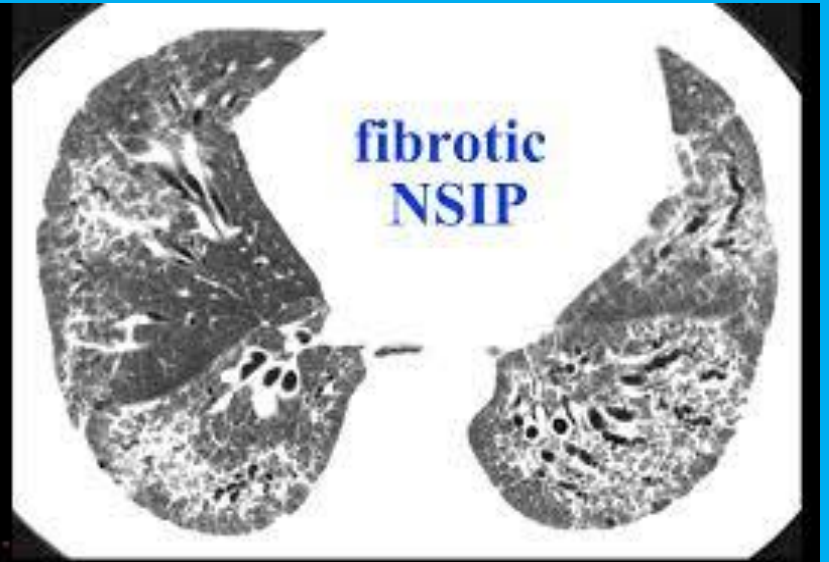


# NSIP Imaging

**Cellular NSIP with infiltration**

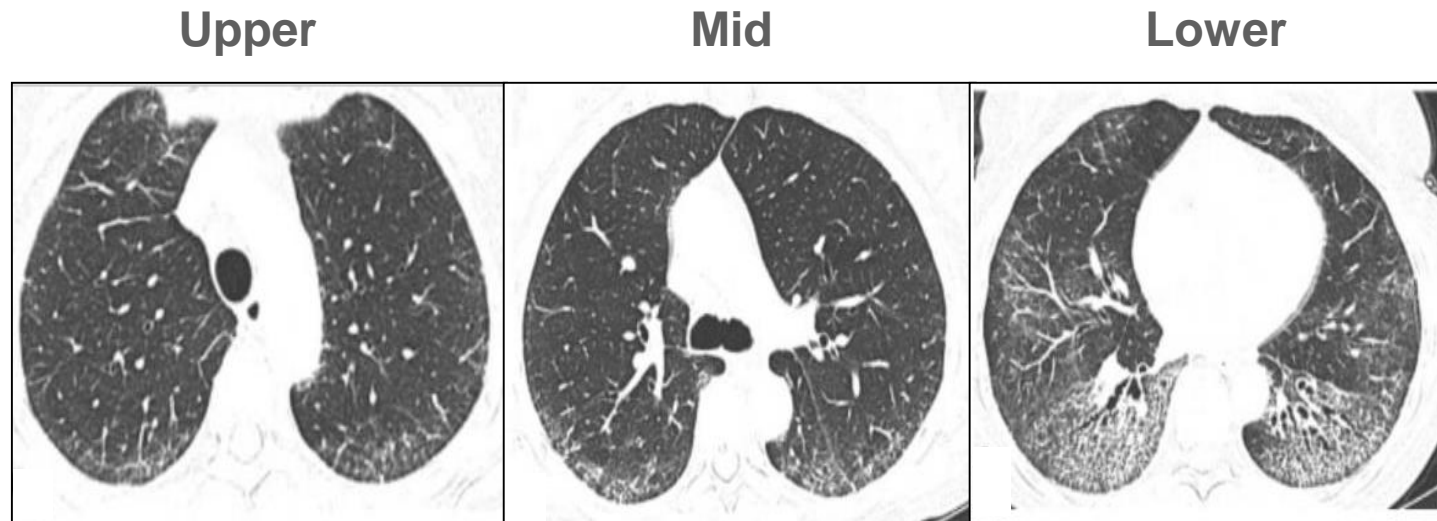


**fibrotic  
NSIP**



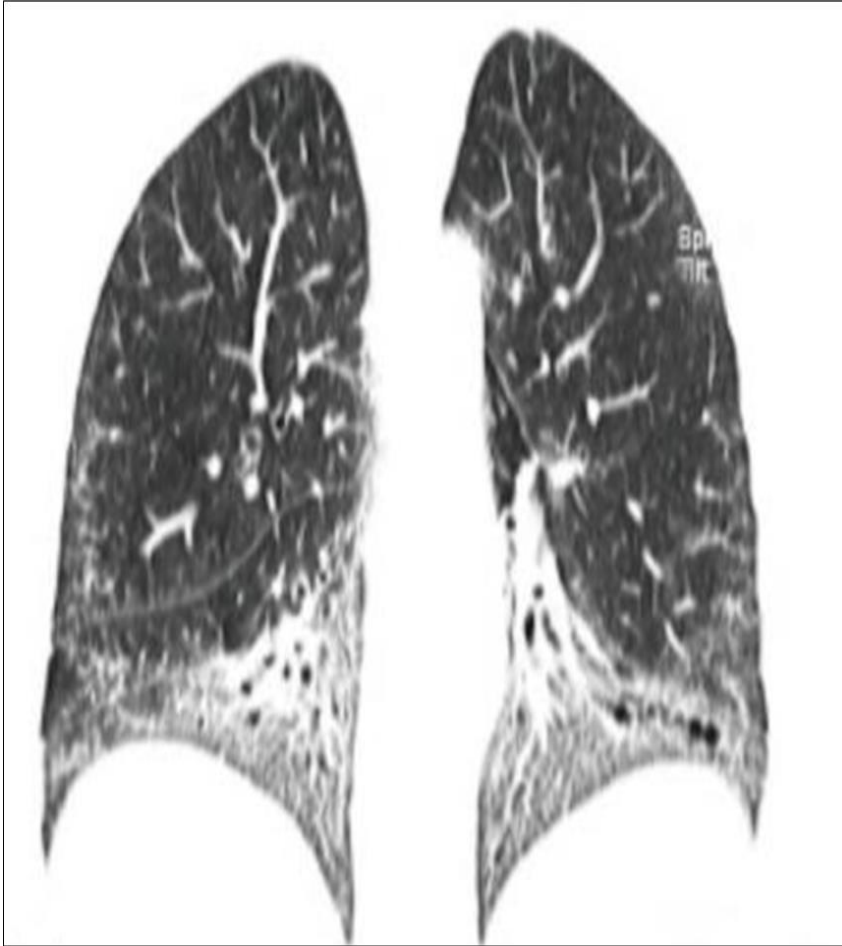


## *Nonspecific Interstitial Pneumonia (NSIP)*



- ▶ HRCT images show a typical pattern of fibrotic NSIP:
  - Subpleural and basilar predominance of abnormalities
  - Irregular reticulation
  - Traction bronchiectasis
  - Honeycombing is rare (,5%)

# *Non Specific Interstitial Pneumonia*



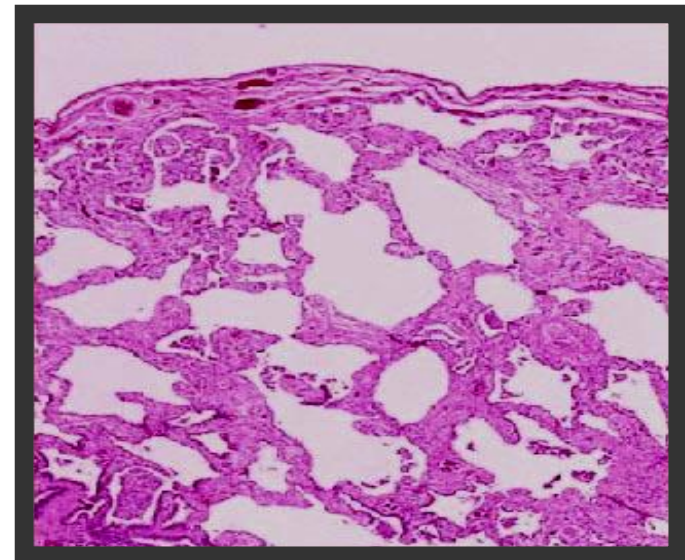
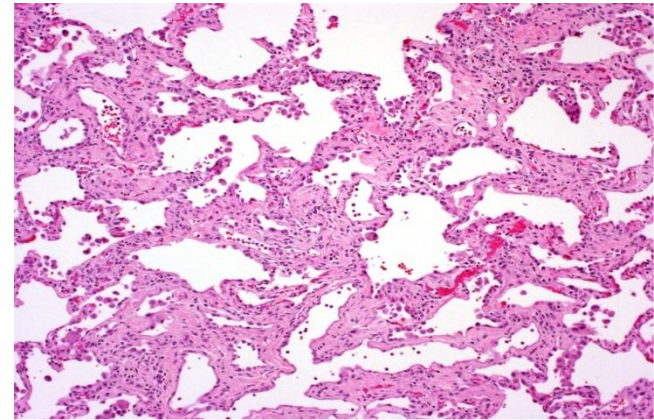
**Coronal**

- Coronal HRCT image confirms:
  - Basilar distribution of irregular reticulation
  - Traction bronchiectasis
  - Subpleural sparing
  - Peribronchovascular extension

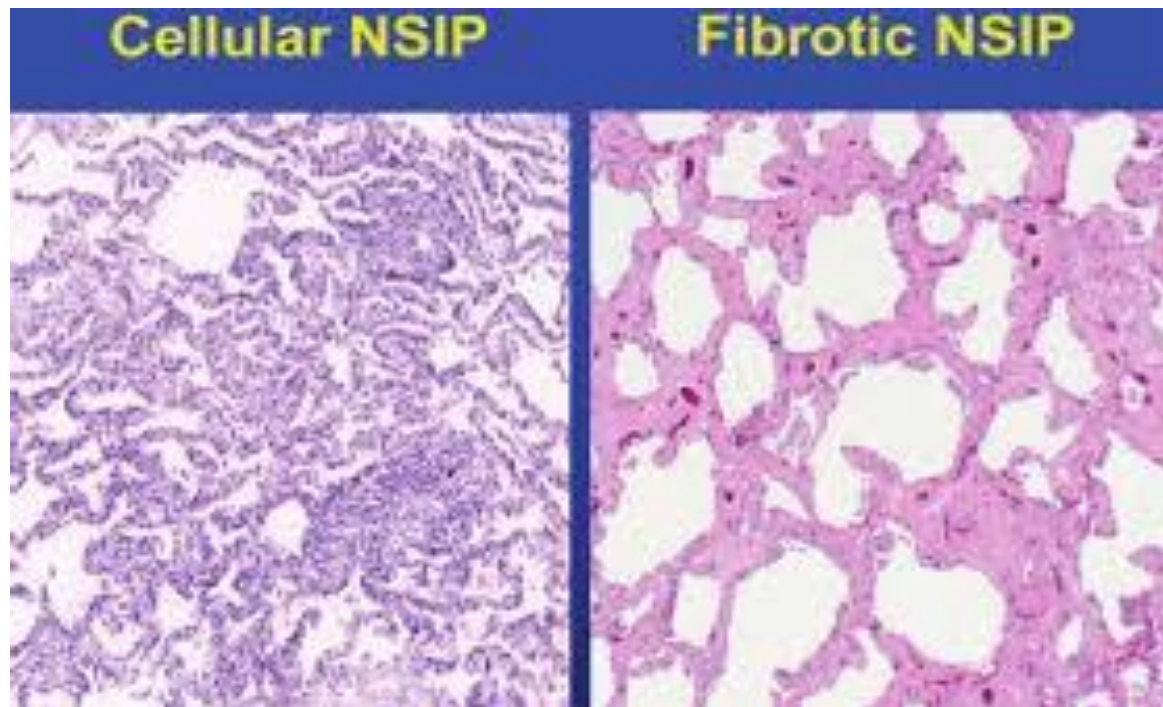
# Non-Specific Interstitial Pneumonia

## Histology

- ▶ Temporal uniformity
- ▶ Inflammation and Fibrosis (cellular, Fibrotic, mixed)
- ▶ No architectural disturbance
- ▶ Rare FBF
- ▶ OP may be present (<10%)



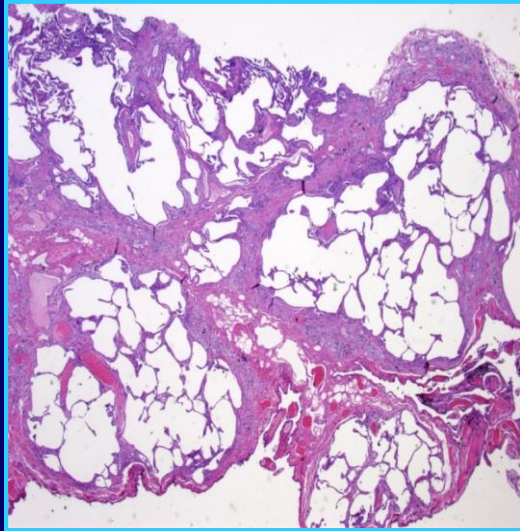
► Histology



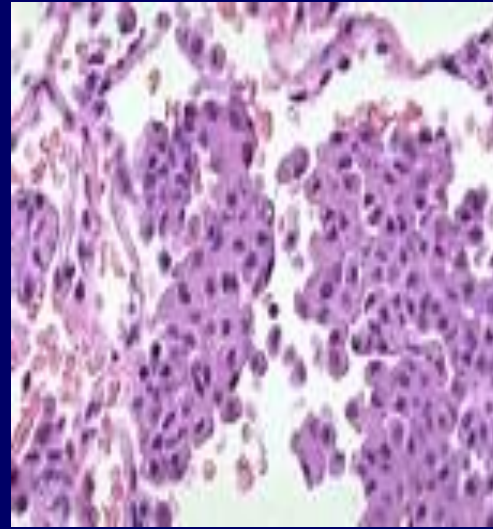


# IIP Histologic Patterns

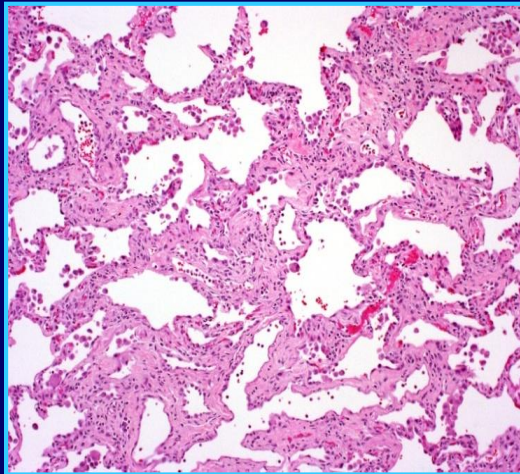
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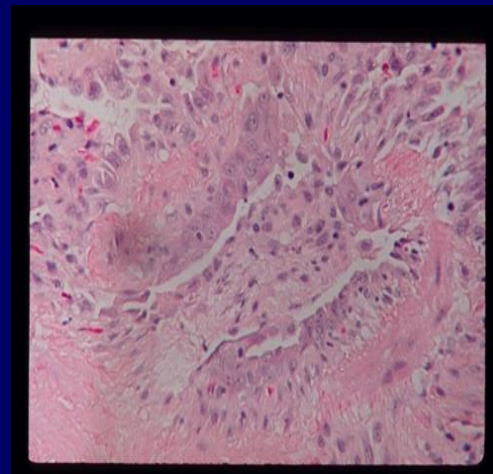
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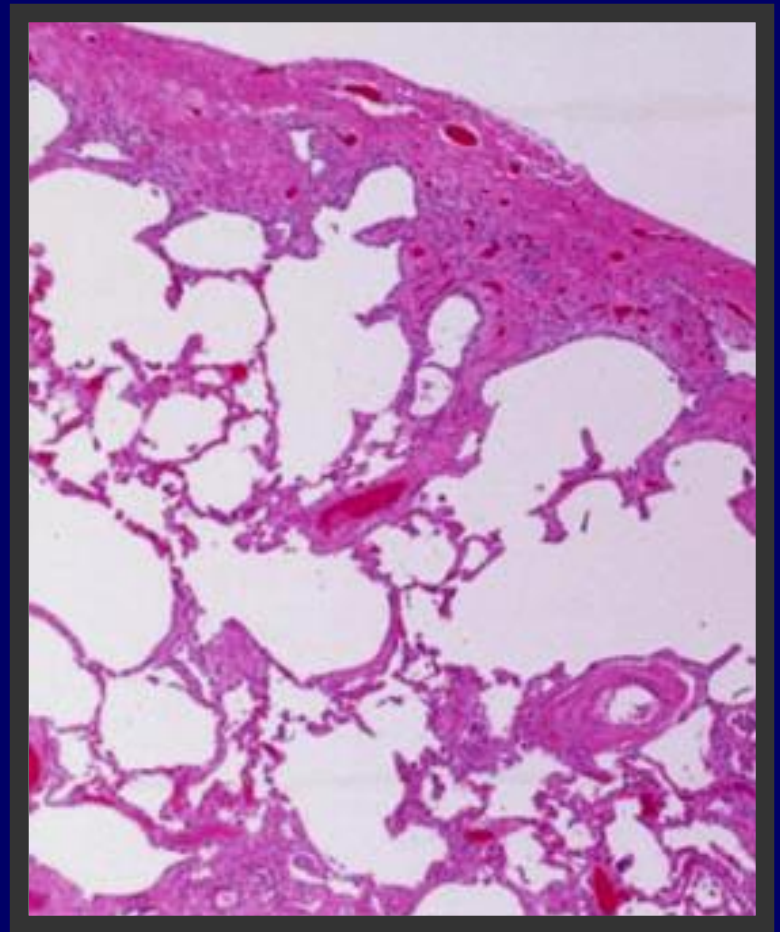
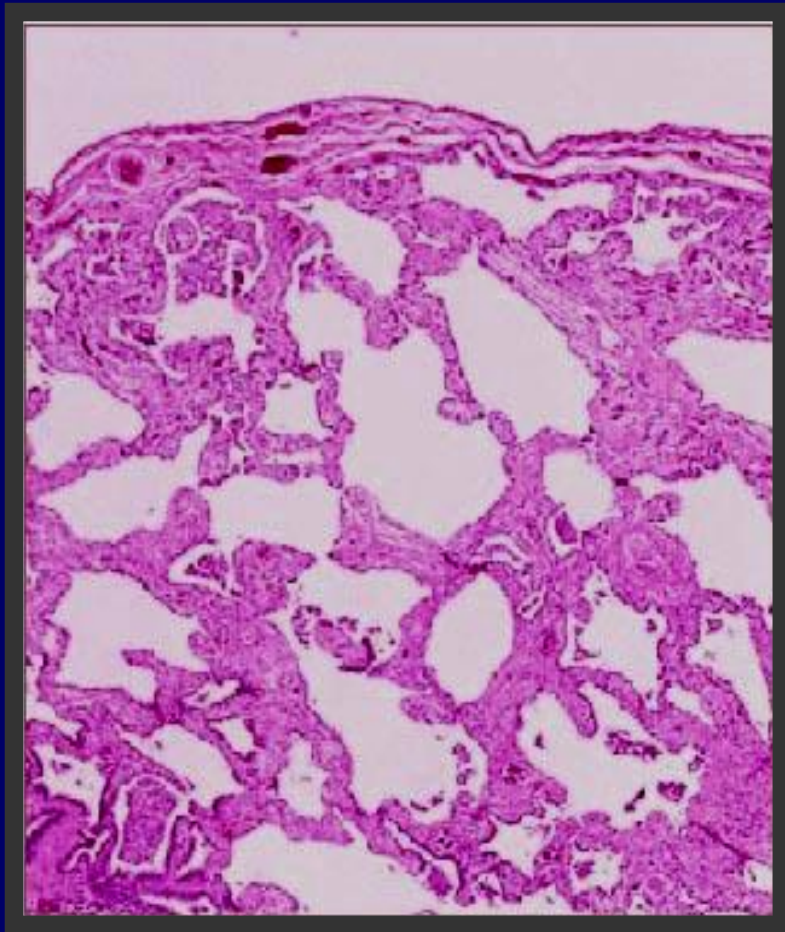
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# Different Patterns of Fibrosis

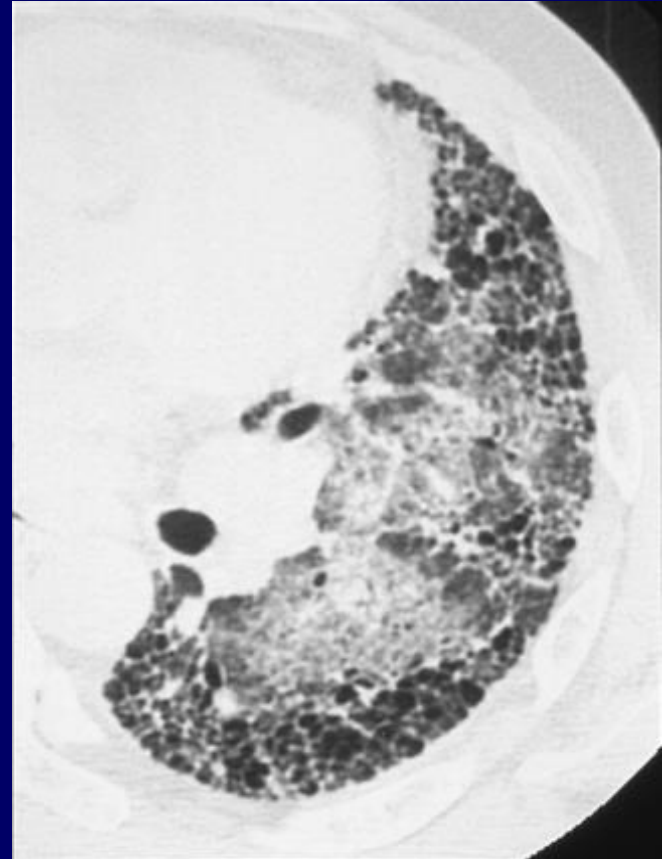
**NSIP/F**

**UIP**



# Acute Exacerbations

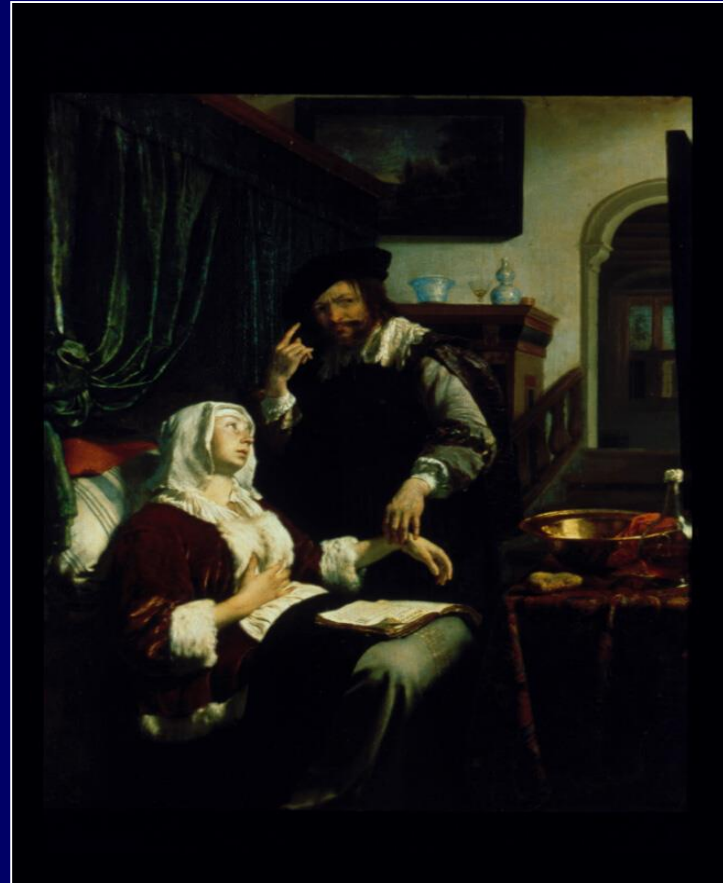
- Acute exacerbations IIP
  - Previous or concurrent dx of IIP
  - Worsening dyspnea, < 30 days
  - HRCT with new ground glass and/or consolidation on IIP pattern
  - No evidence of infection
  - Exclude alternative causes





# NSIP Treatment

Treatment



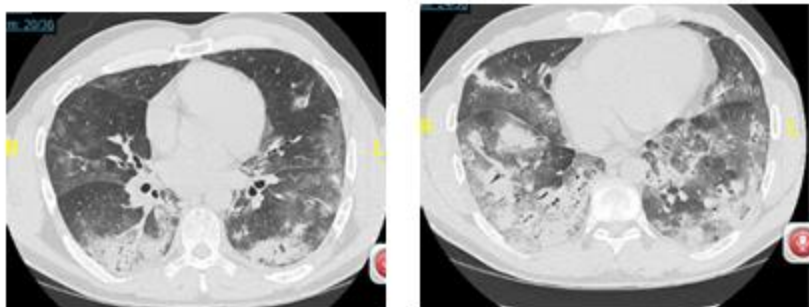


# Therapy

- Glucocorticoids (1 mg/kg)
- Other:
  - Azathioprine
  - Mycophenolate mofetil (MMF)
  - Cyclosporine, tacrolimus
  - Rituximab
  - Cyclophosphamide
  - Intravenous immunoglobulin (IVIG)
- Anti-fibrotic therapy????
- Lung Transplantation

## Antisynthetase Syndrome

46 yo WM admitted in ARD. Rx for pneumonia



3/6/17

Jo1+, Ro 52+ subseq: muscle weakness and fissuring of hands

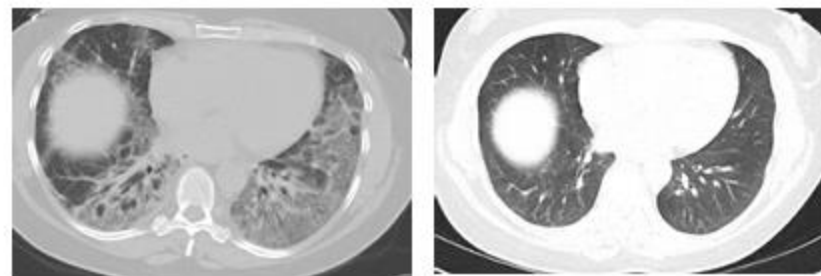
8/7/18

Rx:  
medrol  
Imuran  
CC +Rtx  
Tacro



## Anti-Synthetase Syndrome

52 yo WF with hand changes and progressive SOB, cough and desaturation , RF



December 28, 2016

January 26, 2018

PM/Scl+

Rx: Pred + CC +Ritux

# Non Specific Interstitial Pneumonia



THANK YOU

# Advanced Lung Disease/ILD Program

## Icahn School of Medicine at Mount Sinai

### ILD Team Members (MDD)

Maria Padilla, MD

Aditi Mathur, MD

Sakshi Dua, MD

Mary Beth Beasley MD

Michael Chung, MD

Adam Bernheim, MD

Ioannis Tassiulas, MD

Margrit Wiesendanger, MD

Timothy Harkin, MD

Andrew Kaufman, MD

Jennifer Hastings, PA



**Thank You**