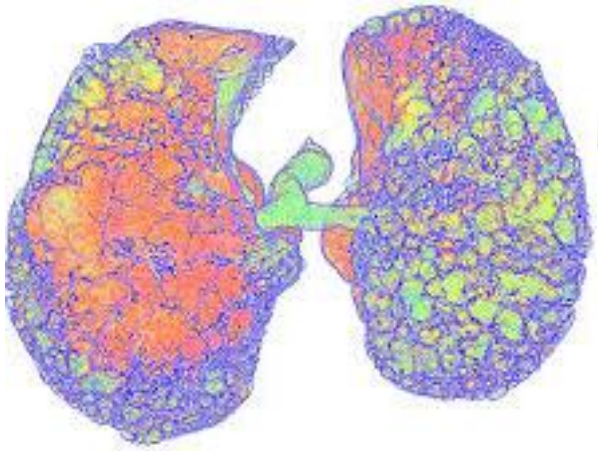


Approach to Interstitial Lung Disease

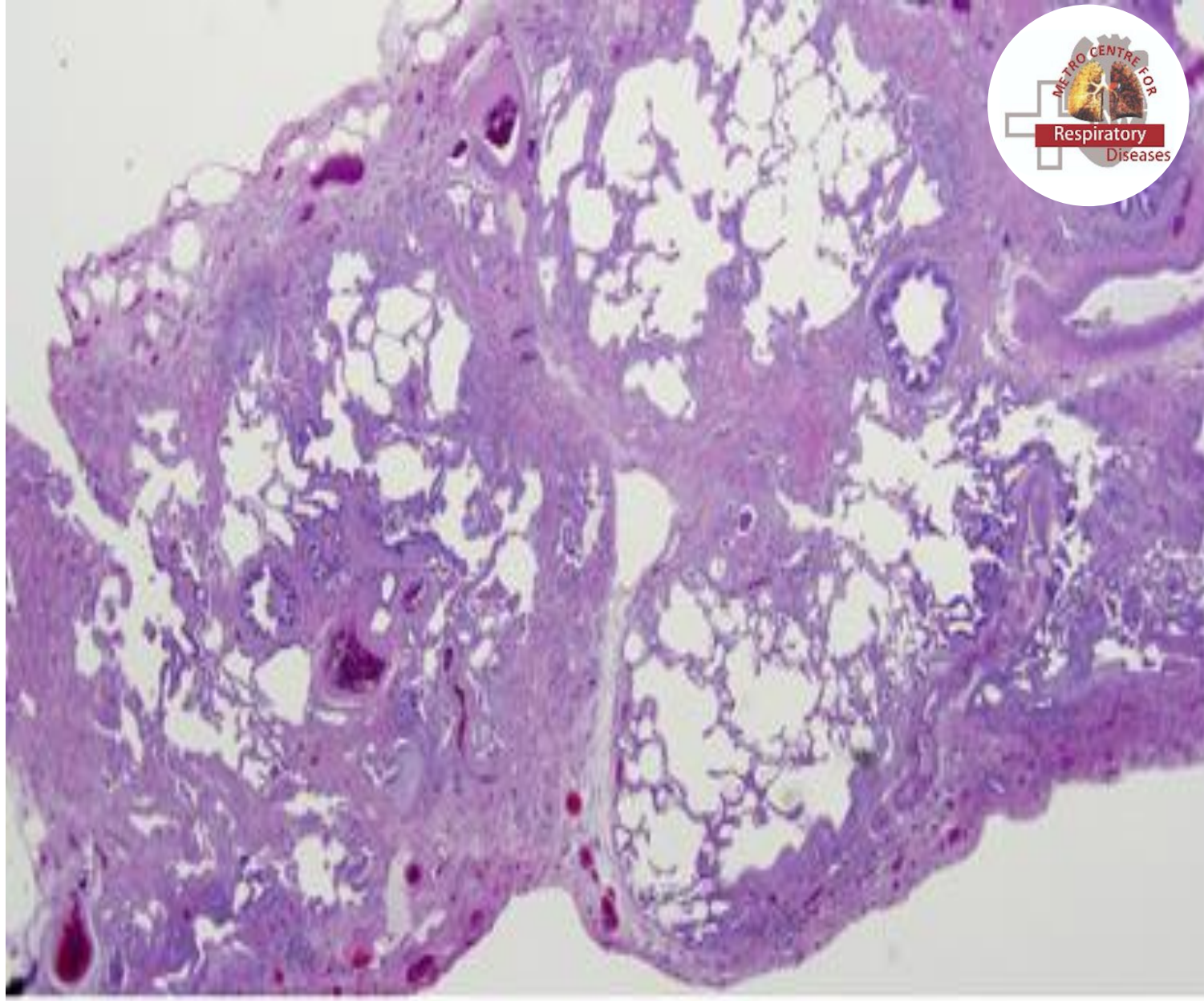
Deepak Talwar

MD, DTCD, DNB, DM (Pulmonary & Critical Medicine) FISDA, FCCP (USA), FNCCP

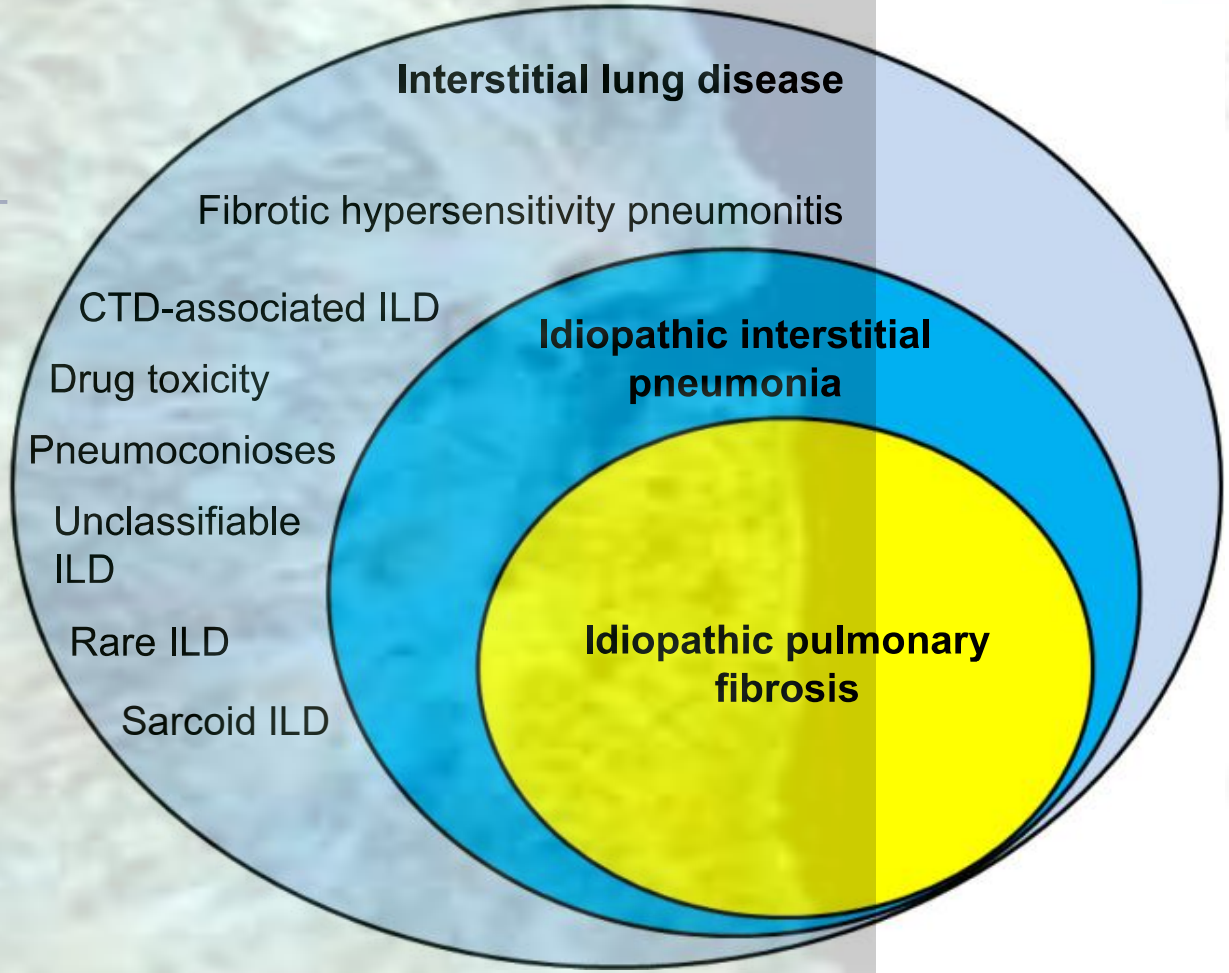
Director & Chair

Pulmonary, Sleep, Allergy & Critical Care Medicine

Metro Group of Hospitals, INDIA



ILD is …



- *> 1000 diseases*
- *> 200 entities*
- *2 / year new added*

ILD Diagnosis is Delayed Worldwide !

Diagnoses 2 years prior Diagnosis of ILD	IPF (%)	Controls (%)
COPD	43	11
Pneumonia	40	7
Asthma	21	6

of patients with ILD were misdiagnosed at least once

Sometimes requiring 3 or more different doctors

Appropriate diagnosis can be delayed by up to 1-2 years

9.9 % - 17.6% ILD cases get diagnosis of Tuberculosis and get ATT

Cosgrove GP et al. *BMC Pulm Med*. 2018;18(1):9.

Late ILD Diagnosis – More Mortality & Morbidity

< 50% Survive beyond 4 years after Diagnosis

It takes 4.5 years to diagnose ILD in India twice the time taken in the developed nations !

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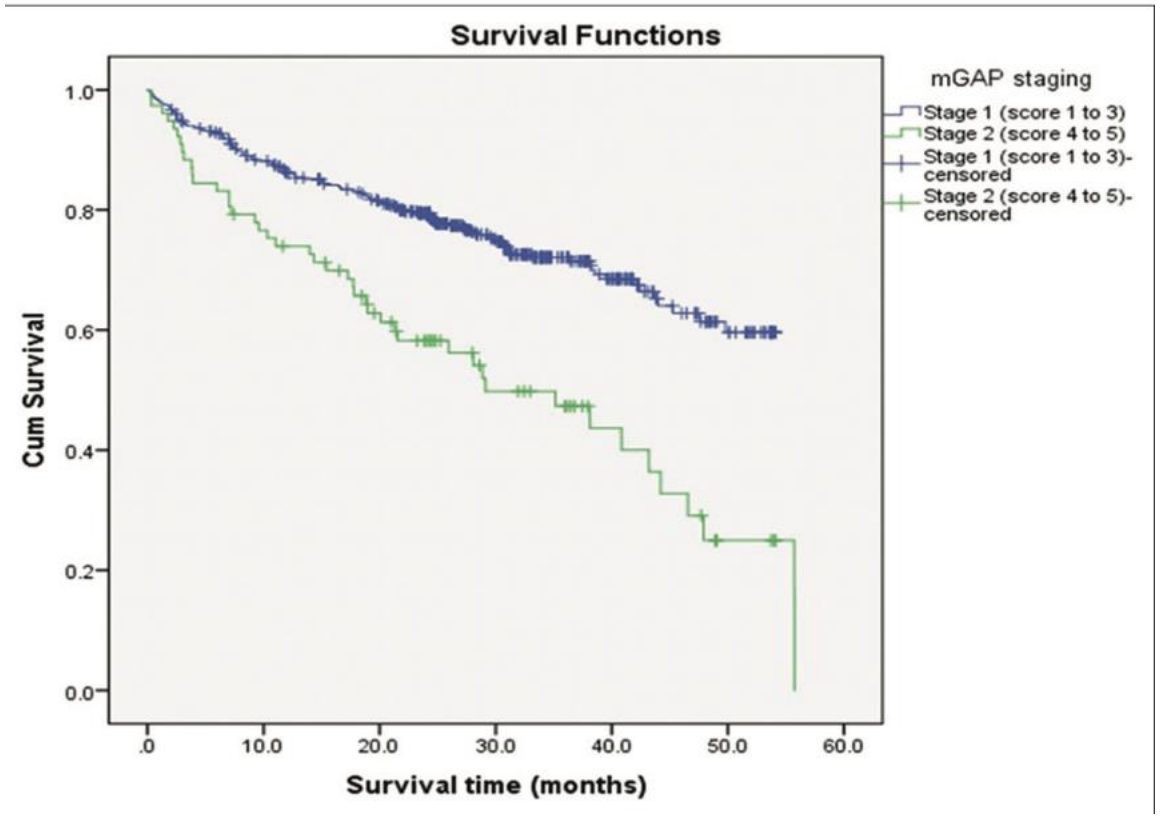
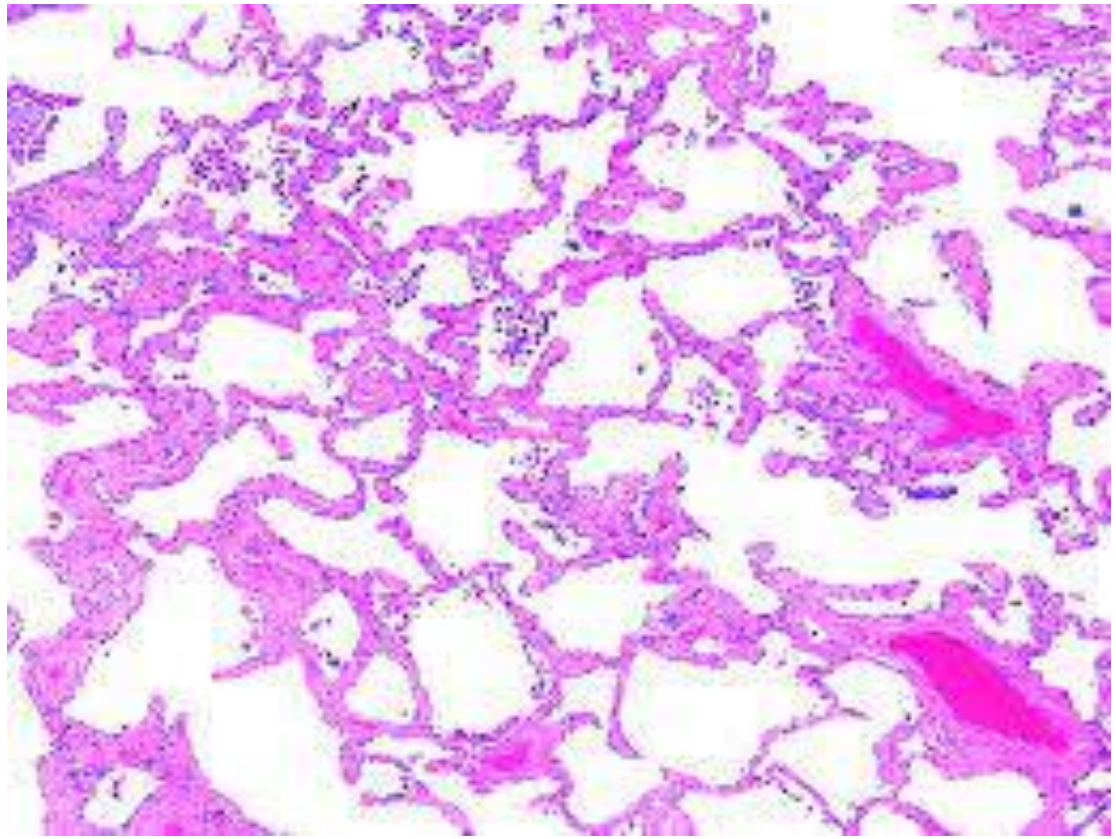
Survival predictors of interstitial lung disease in India: Follow-up of Interstitial Lung Disease India registry

Sheetu Singh[1], Mohan Bairwa[2], Bridget F. Collins[3], Bharat Bhushan Sharma[4], Jyotsana M Joshi[5], Deepak Talwar[6],



Lung India. 2021 Jan-Feb;38(1):5-11

Step 1 : Suspect Diagnosis of ILD



Disease of interstitium but involves air spaces, terminal airways as well as pulmonary vasculature

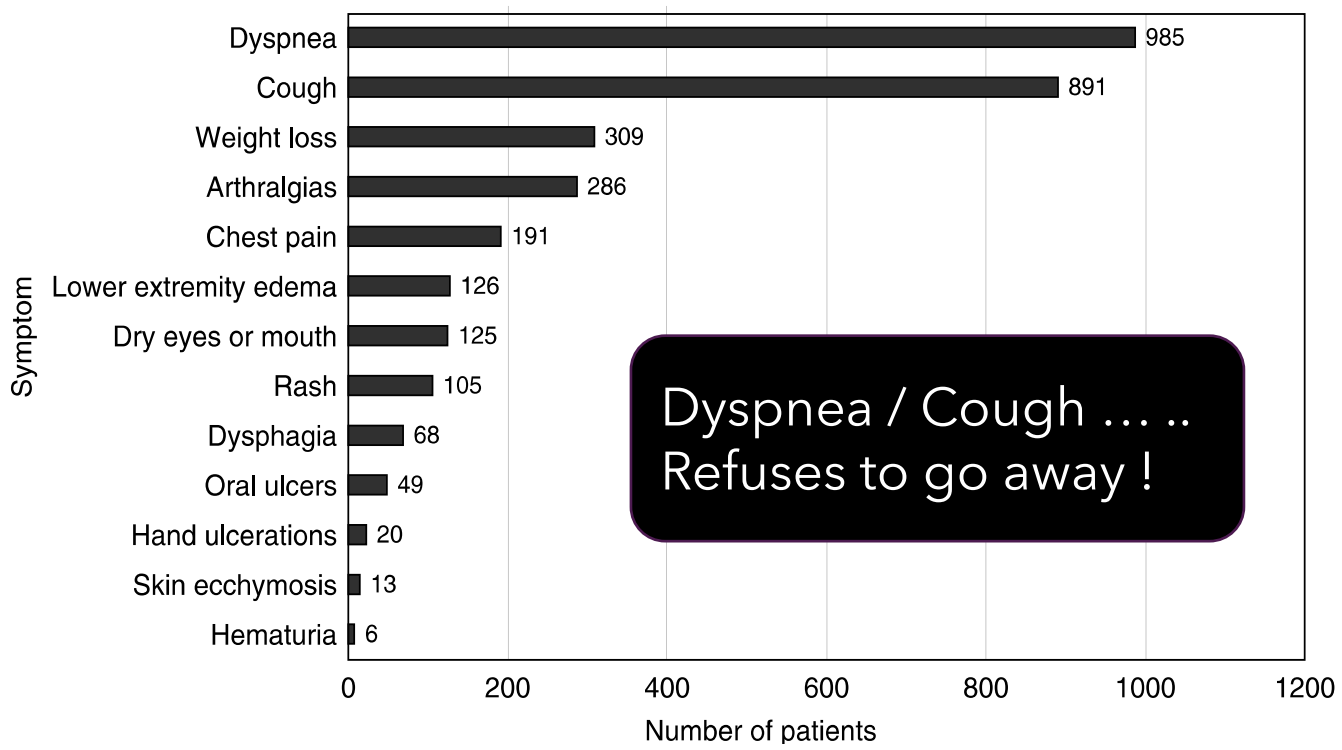
Yu et al. ATS 2014

Step 1 : *Suspect ILD – Clinically : History*

ORIGINAL ARTICLE

Interstitial Lung Disease in India
Results of a Prospective Registry

Sheetu Singh[1], Bridget F. Collins[2], Bharat B. Sharma[3], Jyotsna M. Joshi[4], Deepak Talwar[5],



Dyspnea / Cough … ..
Refuses to go away !

Am J Respir Crit Care Med Vol 195, Iss 6, pp 801–813, Mar 15, 2017

Step 1 : *Suspect ILD – Clinically - Chest Exam*

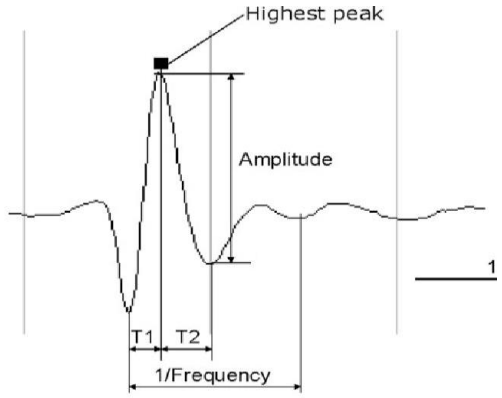
EDITORIAL

Velcro crackles: the key for early diagnosis of idiopathic pulmonary fibrosis?

Vincent Cottin and Jean-François Cordier

Sensitivity : 50%

Specificity : 85%



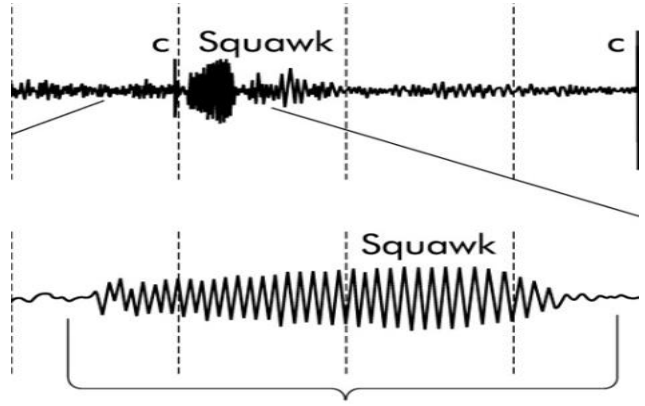
Cottin V et al. *Eur Respir J* 2012;40:519

EUROPEAN RESPIRATORY *journal*

FLAGSHIP SCIENTIFIC JOURNAL OF ERS

Squeaks in hypersensitivity pneumonitis: prevalence and clinical correlates

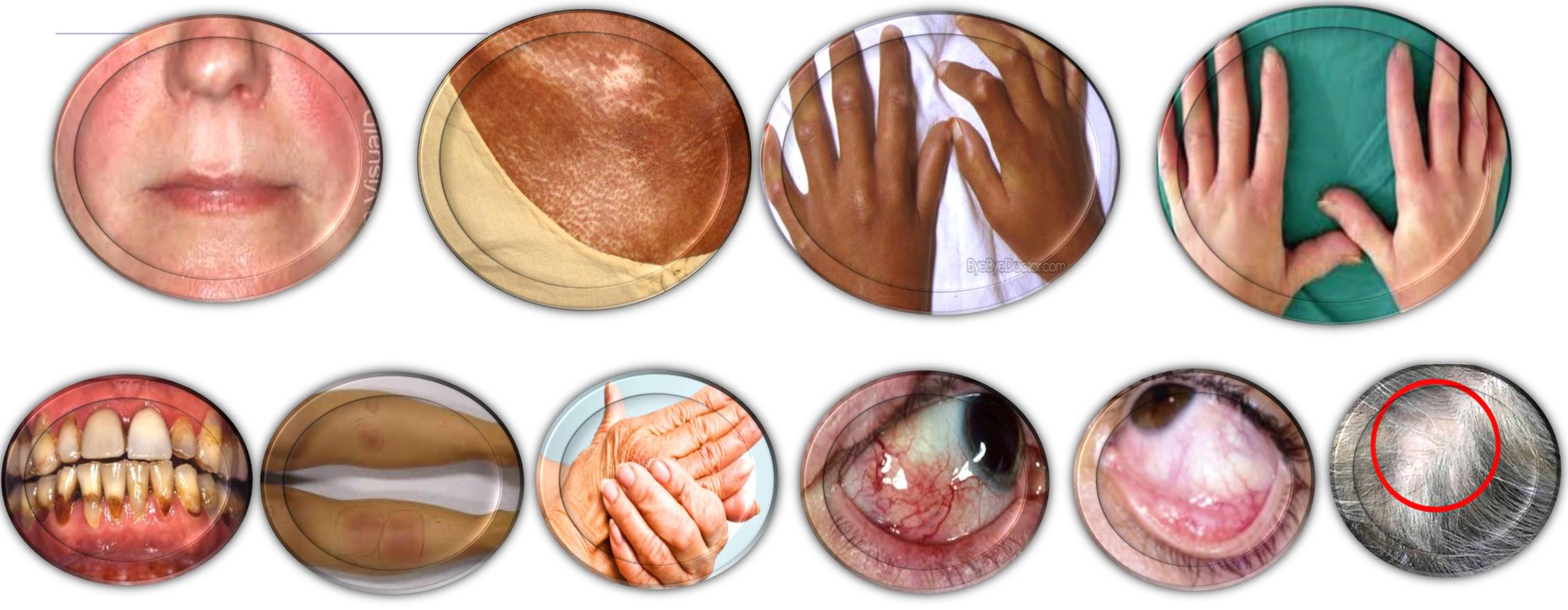
Eleni Diamanti, Cecile Daccord, Selma Ahmetovic, Nathalie Bacco, Romain Lazor



~ 40% of patients with HP, more in females & may suggest the diagnosis of HP in the appropriate context

ERJ 2019, 54 (suppl

Step 1 : *Suspect ILD : Clinically –Look Beyond Lungs*



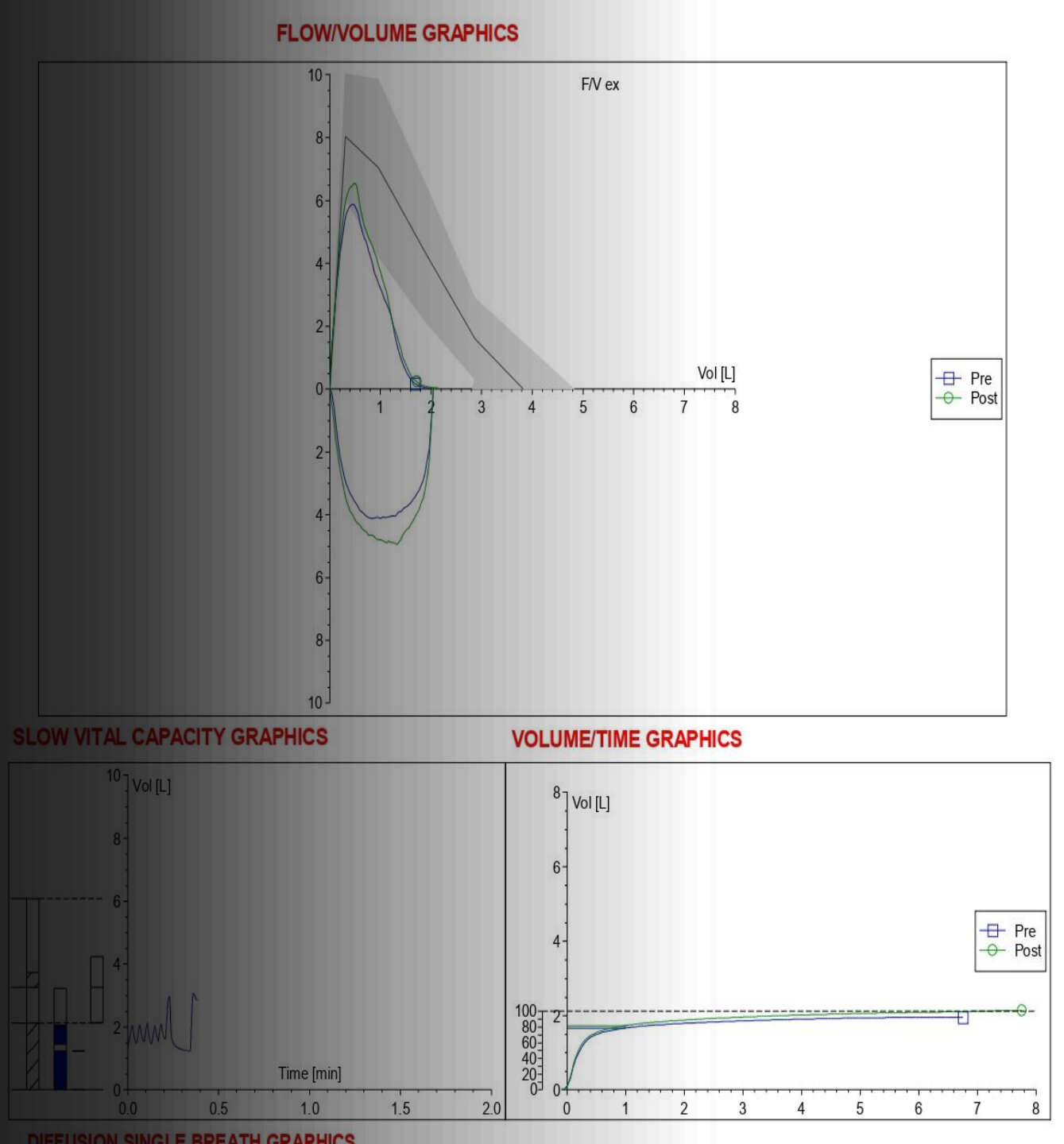
Step 1 : *Suspect ILD : Lung Functions*

Restrictive Lung Functions

- Spirometry
- TLC reduced

Reduced Diffusion Capacity

Desaturation on 6 MWT



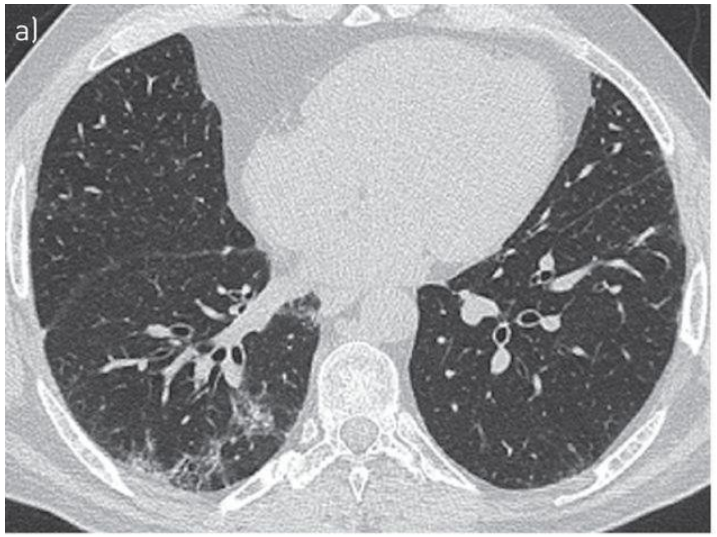
Step 1 : *Suspect ILD – Chest X Ray*

Normal in 10%

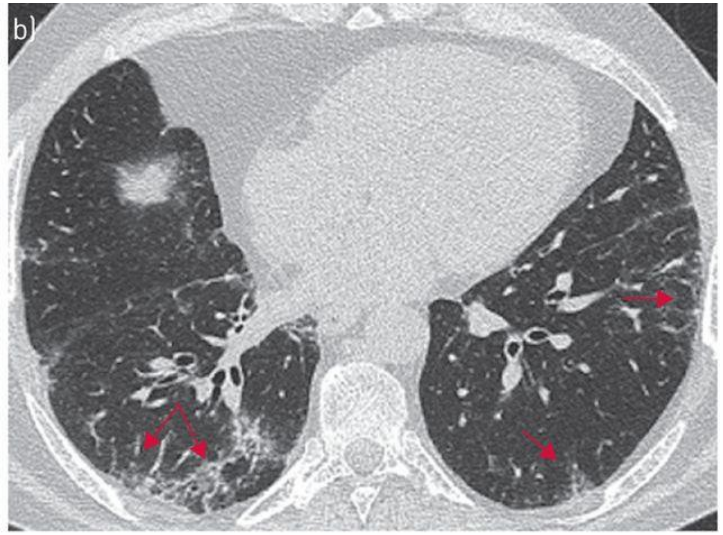
Frequently Normal :

- HP
- CPFE
- RB-ILD

HRCT Scan to Diagnose ILD



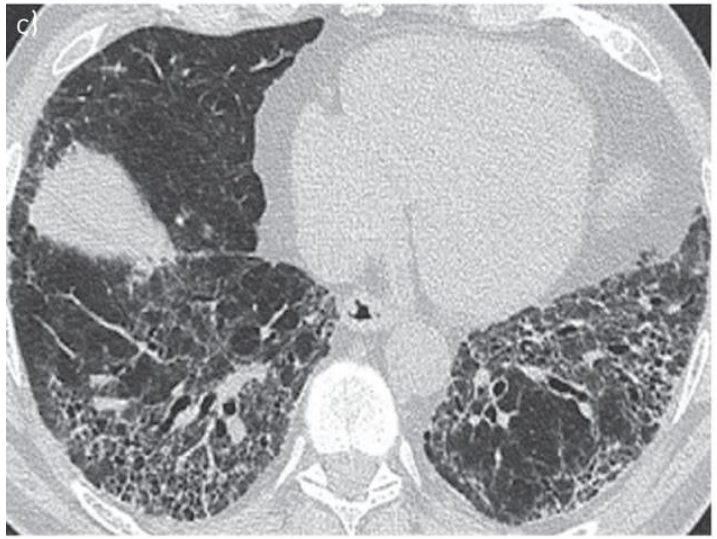
0 Year



2 years

?

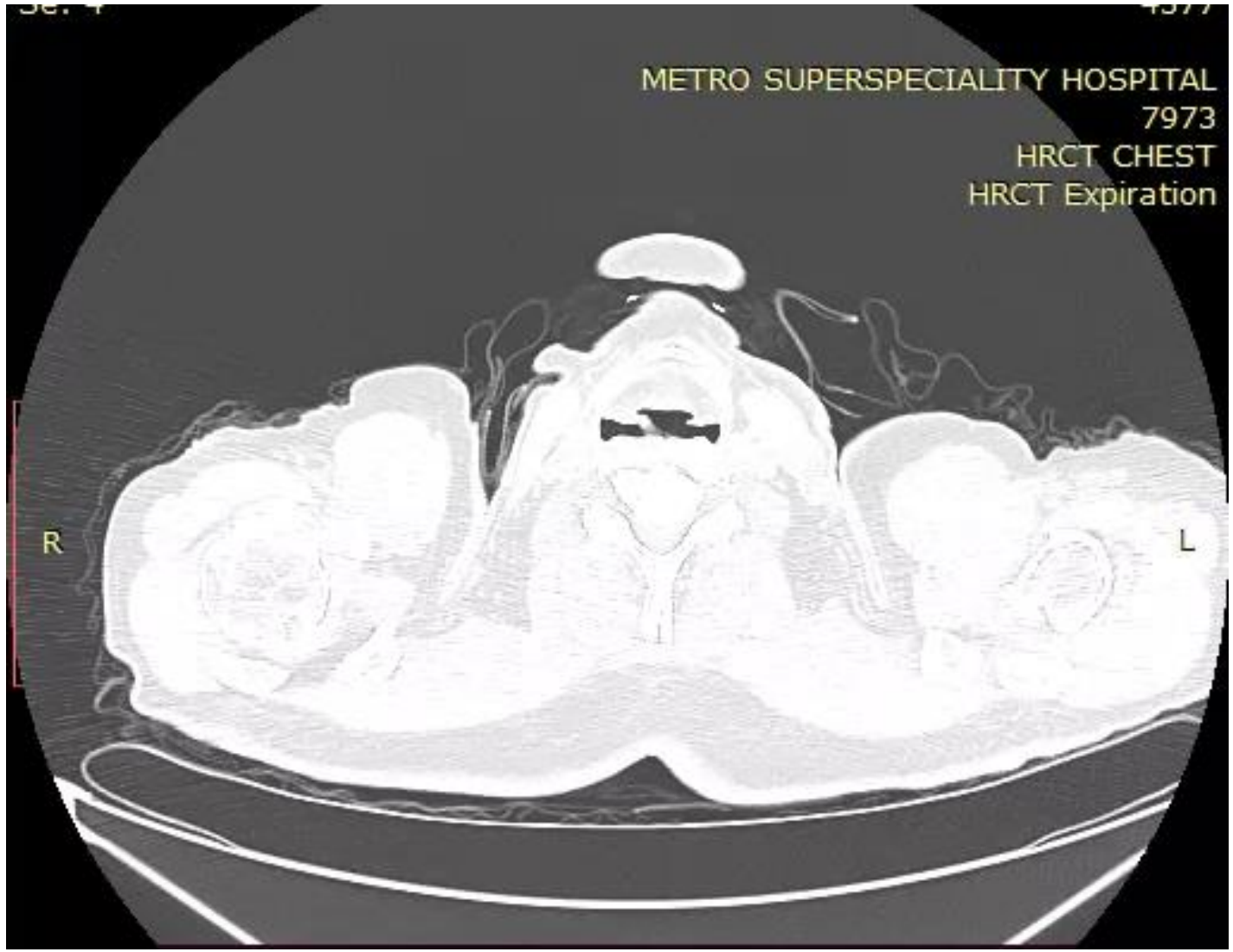
To change the disease trajectory, intervene timely



4 years

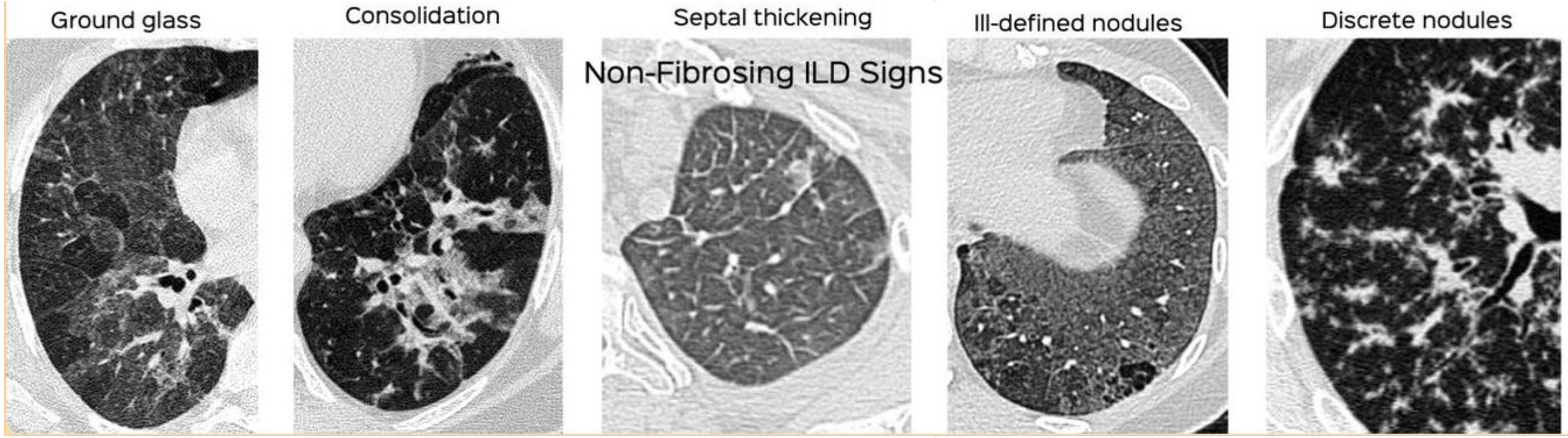
Step 2 : Confirm ILD : *Typical CT Scan*

- NON-Contrast HRCT scan (minimum 1 mm sections)
- Paired - Full inspiration & Expiratory images
- Volumetric images on soft copy



Step 3 : Non-Fibrotic vs Fibrotic ILD vs Progressively Fibrotic ILD's

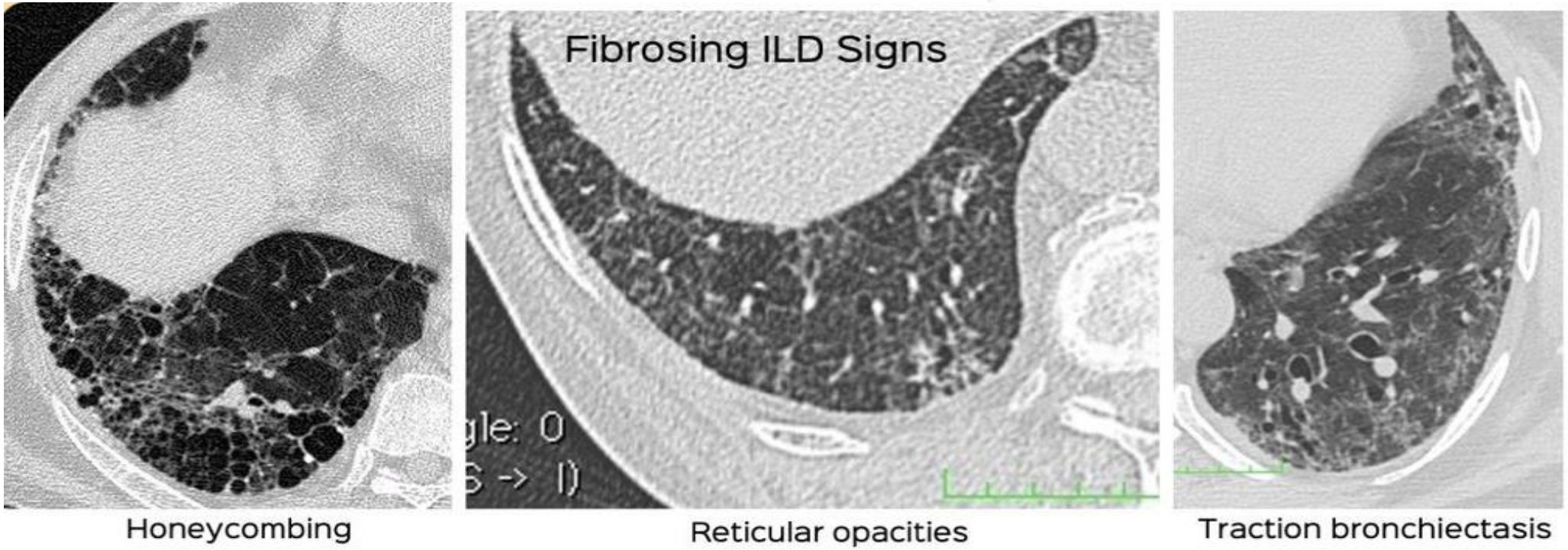
Non-Fibrotic ILD's survive : ~ 10 years



J Clin Med. 2020;9(4):1082.

Step 3 : Non-Fibrotic vs *Fibrotic ILD* vs Progressively Fibrotic ILD's

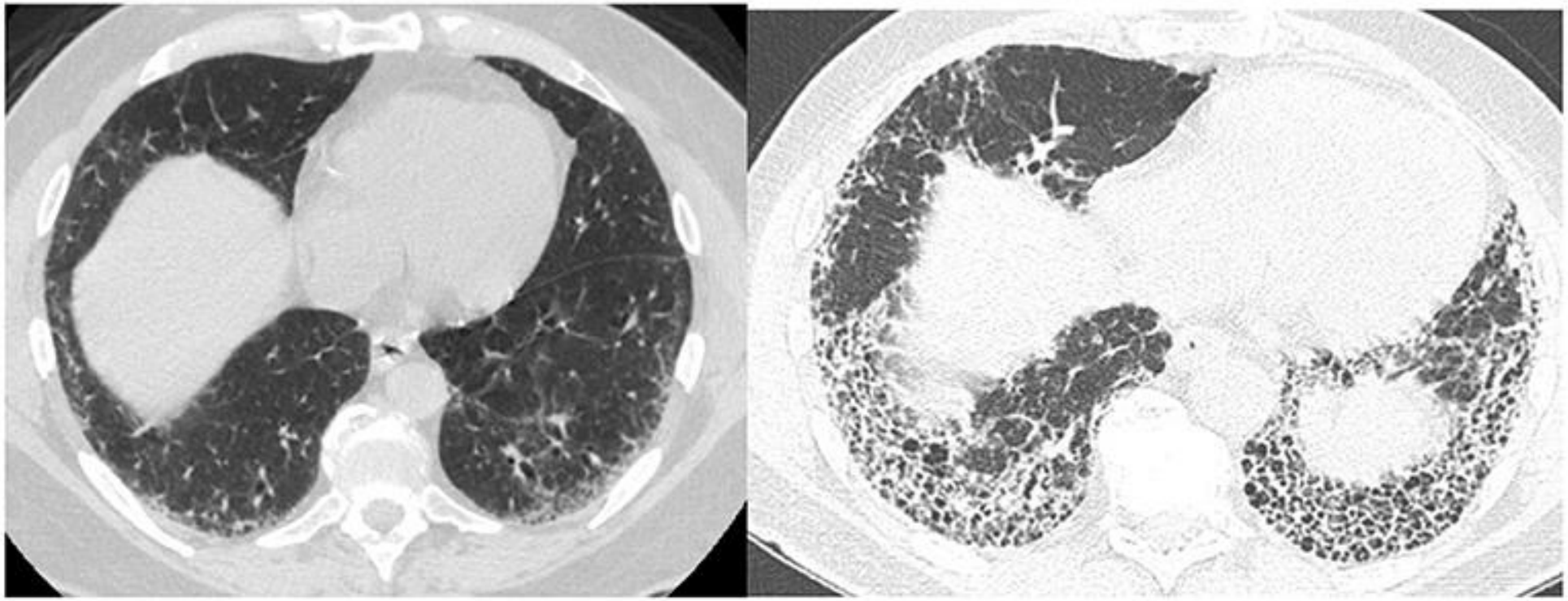
Fibrotic ILD's survive :
~ 5 years



J Clin Med. 2020;9(4):1082.

Step 3: Non-Fibrotic vs Fibrotic ILD vs *Progressively Fibrotic ILD*'s

Progressive Fibrotic ILD's
Survive : ~ 3 years



1 Year later

J Clin Med. 2020;9(4):1082.

ALL ILD's are *NOT* *Equally* Deadly …

Sarcoid ILD's survive much longer than CHP or CTD-ILD's

Its heterogenous disease with least survival in IPF and best in sarcoid ILD

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Survival predictors of interstitial lung disease in India: Follow-up of Interstitial Lung Disease India registry

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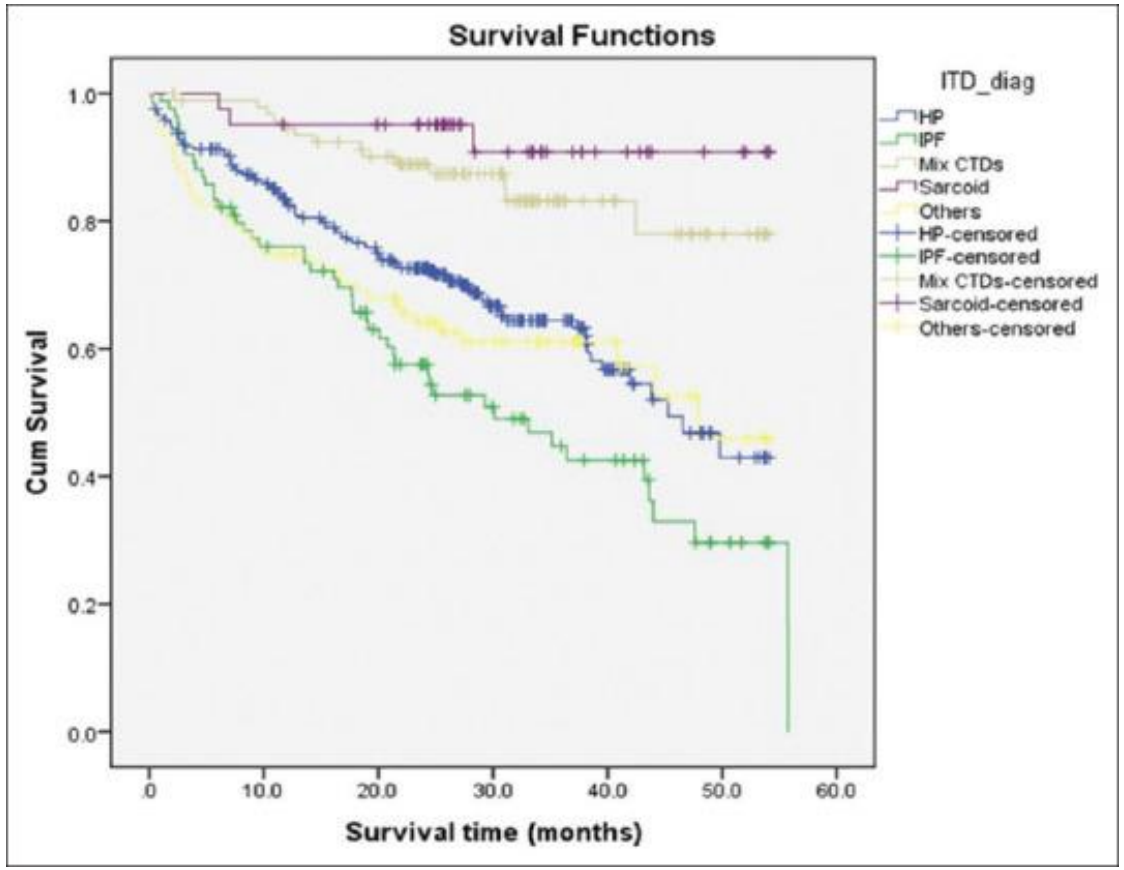
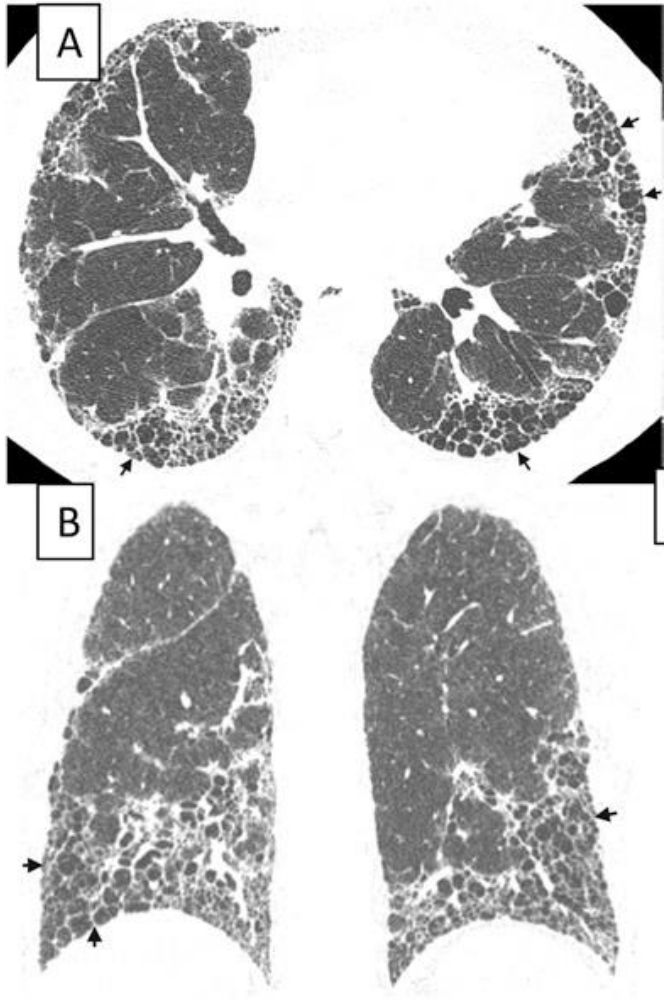


Lung India. 2021 Jan-Feb;38(1):5-11

Step 4 : IPF Vs Non IPF- ILD



Honeycombing # IPF but presence indicates poor survival

Fibrotic HP

- CTD – ILD
- Sarcoid ILD

25% of ILD cases in India have Honeycombing at the diagnosis while IPF is only 15% of newly seen ILD's

ATS/ERS Am J Respir Crit Care. 2018 ; 165 ; 277-304

IPF Label : Death Sentence

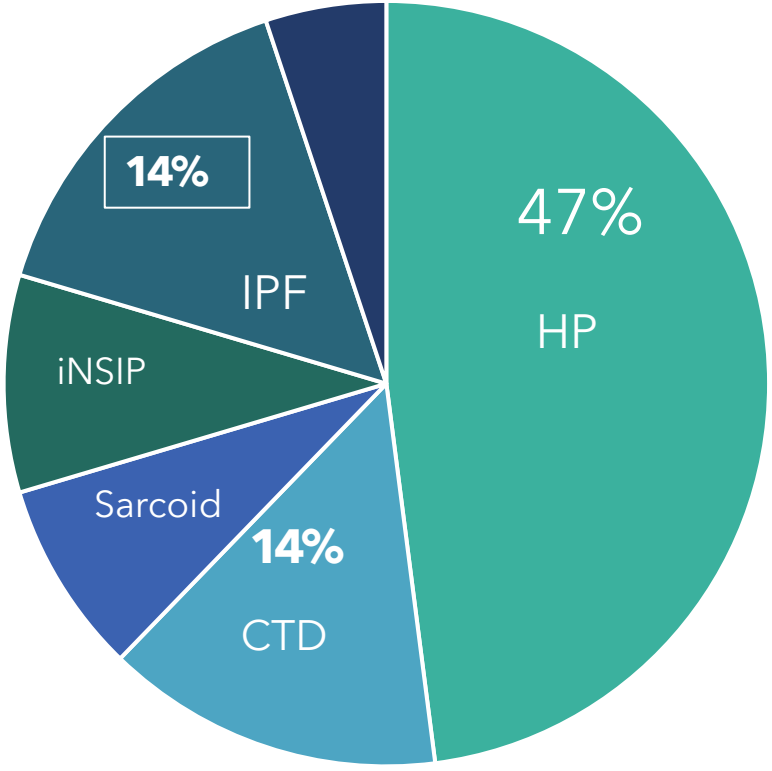
IPF is aging Disease

What's Typical IPF ?

- > 65 years, Male, Ex-smoker
- Dyspnea on exertion
- Chronic dry cough
- Exercise desaturation
- Basilar "Velcro" crackles
- Definite/Probable UIP on HRCT
- PFTs: Mixed defect with ↓ DL_{CO}
- Often previously diagnosed with different lung disease
- Concomitant emphysema (30%)

Brody J The New York Times , 11 Jan 2016

Step 5 : Which Non IPF ILD ?



Type of ILD	Number of patients (%)
Hypersensitivity pneumonitis	513 (47.3)
Connective tissue associated ILD	151 (13·9)
Idiopathic pulmonary fibrosis	148 (13·7)
Idiopathic Nonspecific interstitial pneumonia	92 (8·5)
Sarcoidosis	85 (7.8)
Pneumoconiosis	33 (3.0)

HP CTD-ILD Sarcoidosis NSIP IPF Others

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Diagnosis of ILD : *Next Step – CTD Etiology !*

AMERICAN THORACIC SOCIETY DOCUMENTS

Diagnosis of Idiopathic Pulmonary Fibrosis
An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Jeffrey L. Myers, Luca Richeldi, Christopher J. Ryerson, David J. Lederer, Juergen Behr, Vincent Cottin, Sonye K. Danoff, Ferran Morell, Kevin R. Flaherty, Athol Wells, Fernando J. Martinez, Arata Azuma, Thomas J. Bice, Demosthenes Bouros, Kevin K. Brown, Harold R. Collard, Abhijit Duggal, Liam Galvin, Yoshikazu Inoue, R. Gisli Jenkins, Takeshi Johkoh, Ella A. Kazerooni, Masanori Kitaichi, Shandra L. Knight, George Mansour, Andrew G. Nicholson, Sudhakar N. J. Pipavath, Ivette Buendía-Roldán, Moisés Selman, William D. Travis, Simon Walsh, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society

This official clinical practice guideline of the American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society (JRS), and Latin American Thoracic Society (ALAT) was approved by the ATS, JRS, and ALAT May 2018, and the ERS June 2018

Question 2: Should Patients with Newly Detected ILD of Unknown Cause Who Are Clinically Suspected of Having IPF Undergo Serological Testing to Exclude CTDs as Potential Causes of the ILD?

Panelists routinely recommend :

- CRP (C-reactive protein)
- ESR
- ANA (by immunofluorescence)
- RF(rheumatoid factor) & Anti CCP
- Myositis panel ??

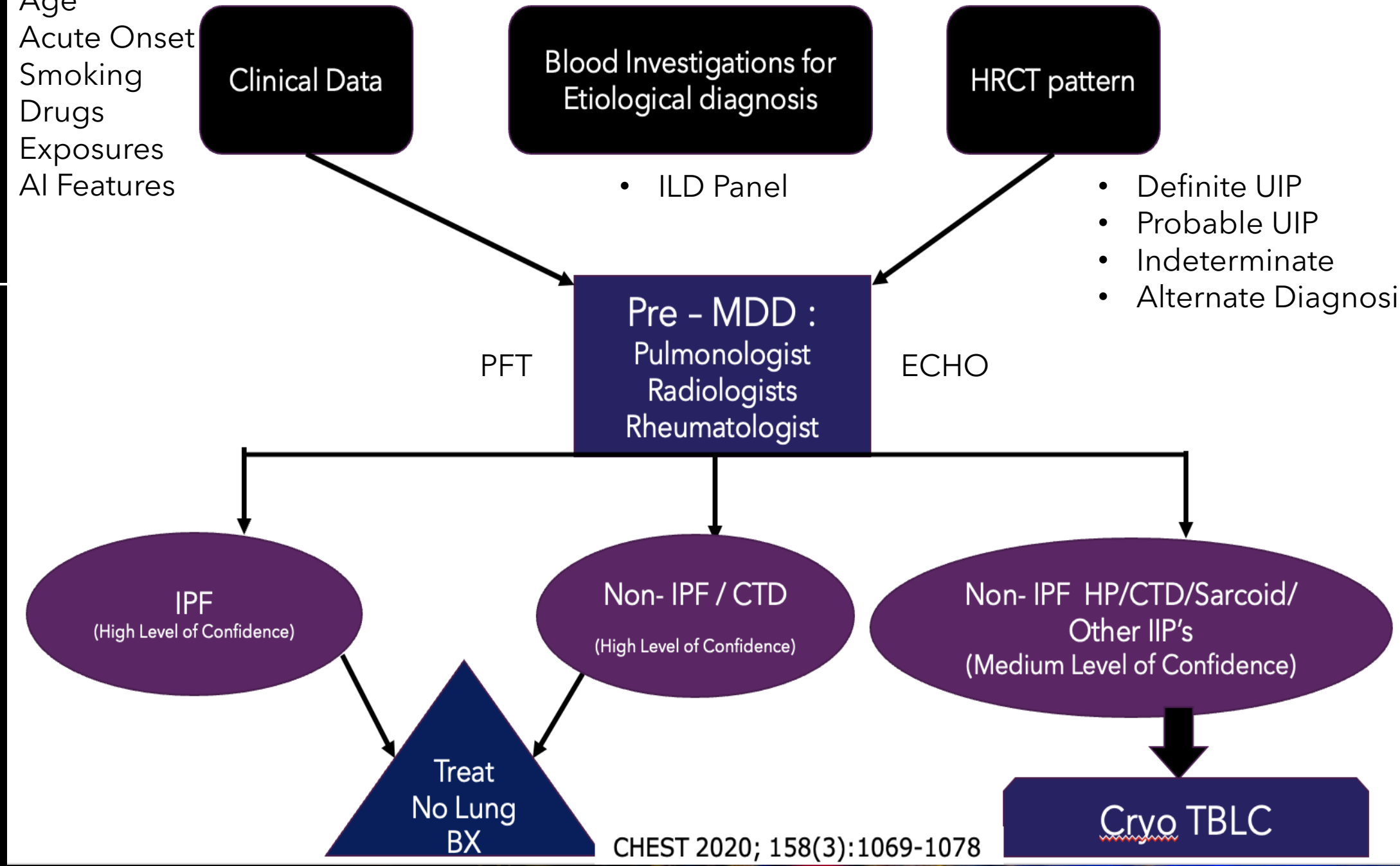
ATS/ERS/JRS/ALAT recommendation.

- **For patients with newly detected ILD of apparently unknown cause who are clinically suspected of having IPF, we recommend serological testing to aid in the exclusion of CTDs as a potential cause of the ILD** (*motherhood statement*).

Am J Respir Crit Care Med Vol 198, Iss 5, pp e44–e68, Sep 1, 2018

Step 6 MDD

- Age
- Acute Onset
- Smoking
- Drugs
- Exposures
- AI Features



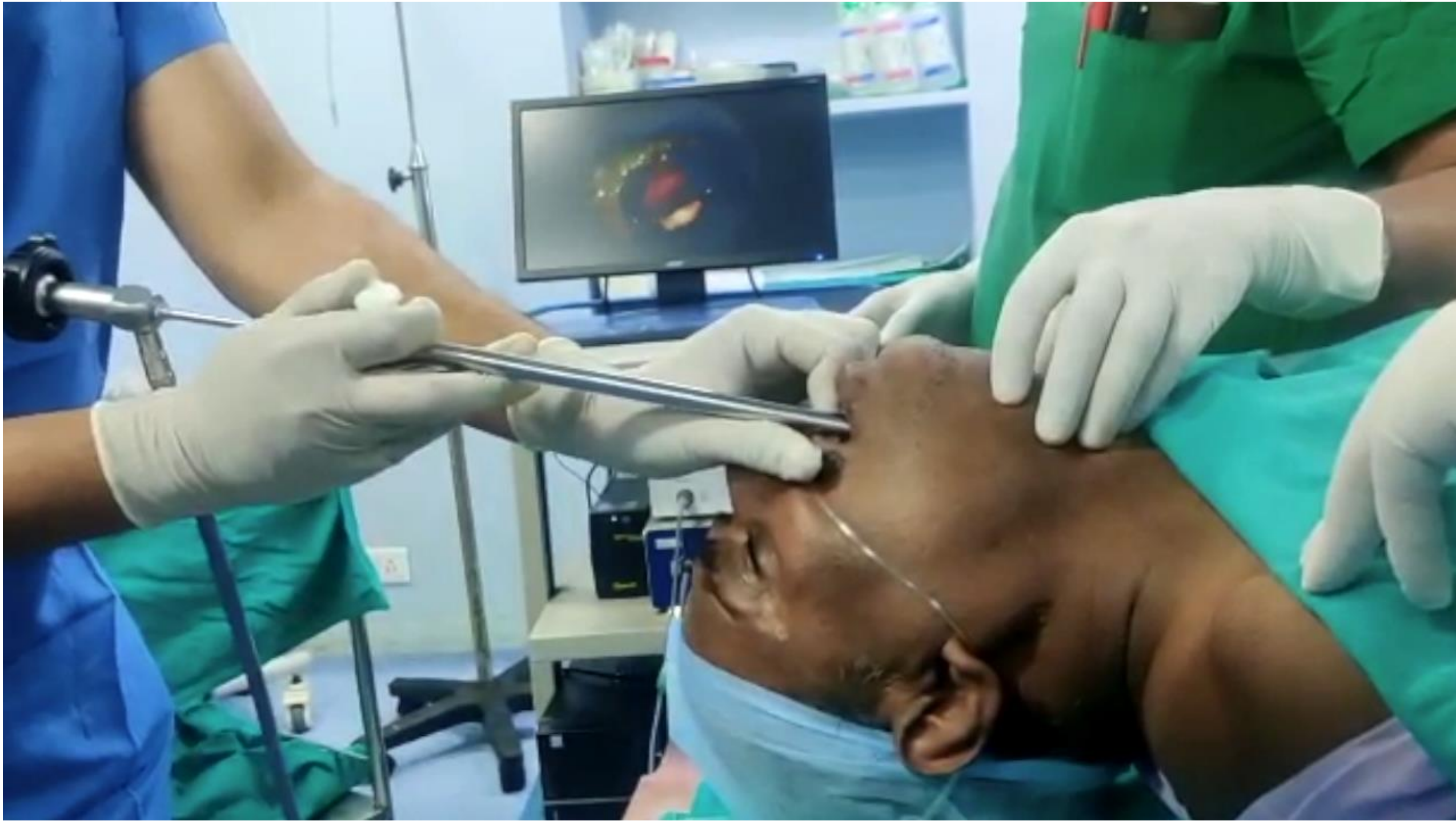
Grey Zone in ILD's : *HP/ NSIP/Sarcoid / IPF*

When clinical-radiological findings are not Typical of Any ILD

Pretest probability of these is around 50%-90%

Cryo TBLB (TBLC)

MDD



Conclusions:

- ILD should be clinically suspected & HRCT with ILD protocol to confirm the diagnos is the beginning of ILD Journey
- Sub type of ILD determines treatment, prognosis and future
- MDD is the way forward to get high probability ILD sub type
- Clinical, HRCT and Labs achieve specific diagnosis in < 40 %
- Lung Biopsy needed when MDD not confident in subtype ILD
- Cryobiopsy – OPD procedure subtype ILDs with > 90% success
- MDT with with Pulmo-Radiologist & Pathologist is *MUST* @ ILD care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AUGUST 2025
Radisson Blu, Kaushambi, Ghaziabad
Dr. Deepak Talwar
Organizing Chair
Dr. Deepak Prajapat
Organizing Secretary
Dr Kanishka Kumar Singh
Organizing Secretary

Thank You