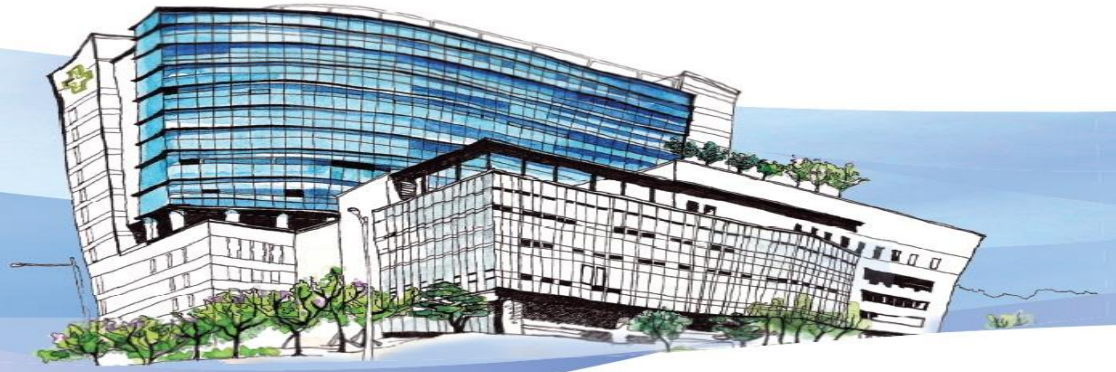


간질성 폐이상(ILA): 경과 관찰 가능한가? 악화 징후 포착하기

해운대백병원

이재하

CASE

▶ M / 76

▶ C.C : Dyspnea (onset: 3 months ago)

▶ PI : 상환 3개월 전 부터 지속되는 기침과 호흡곤란으로 내원하였다. 빨리 걷거나, 계단을 오르면 숨이차서 쉬어야 된다고 하고, 점점 나빠진다고 한다.

▶ Arthritis (-) Raynaud (-) Sicca symptom (-)

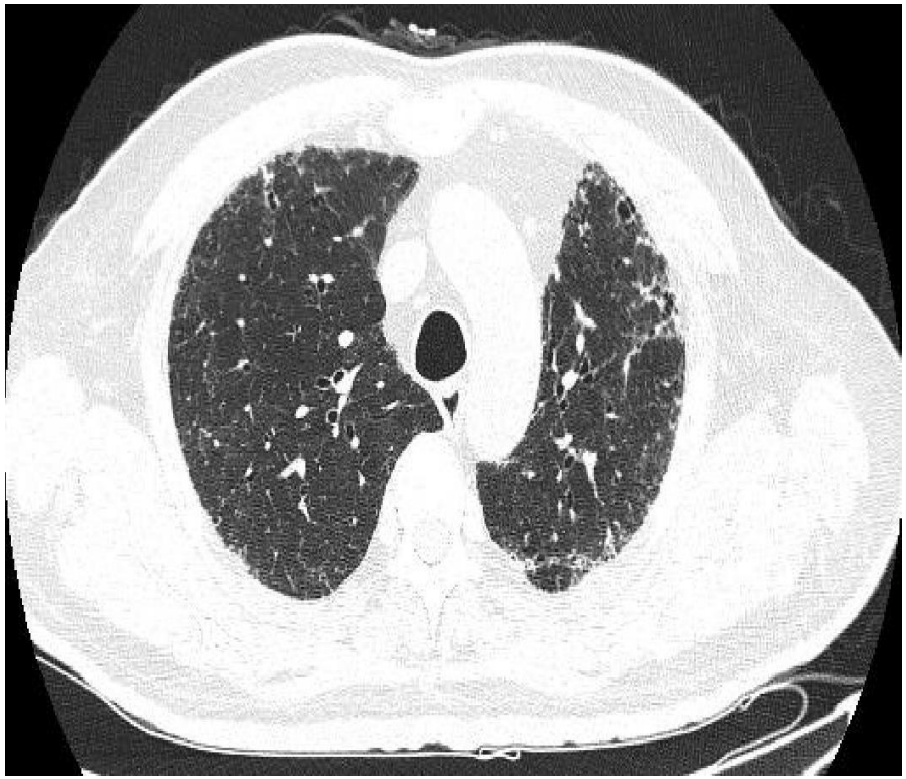
▶ Ex-smoker (40 갑년, 10년 전 중단)

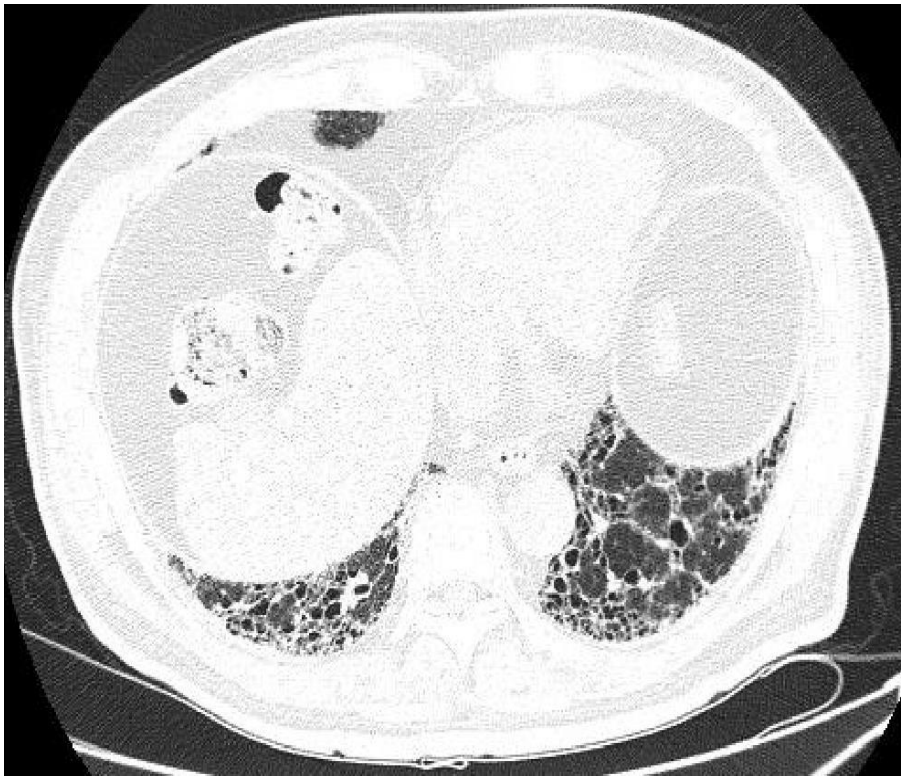
▶ Exposure history (-), Family history (-)

▶ DM (40년 전), Hypothyroidism(20년 전), HTN, Dyslipidemia

Chest CT

2022.10.13





PFT and 6MWT

• PFT

	% predicted
FEV1/FVC	75
FVC	50
DLco (mL/min/mmHg)	32

▶ **2022.10**

6MWT	Initial SpO_2(%)	Lowest SpO_2(%)	Distance (m)
	92	71	340

증상 : Cough, Dyspnea / mMRC G2

• Auto-Ab screening

ANA	Negative (1:40) Cytoplasmic Ab
ANCA-MPO(Qn)	Negative(0.12)
ANCA-PR3(Qn)	Negative(0.77)
ANCA-MPO(QI)	Negative
ANCA-PR3(QI)	Negative
Rheumatoid Factor	244.4
SS-A(Ro) Ab	Negative(0.20)
SS-B(La) Ab	Negative(0.20)
KL-6(U/mL)	1430

CASE

- 특발성폐섬유증으로 진단 되었습니다.
- 환자의 현재 폐기능은 FVC 50% 와 DLco 32%로 중증의 상태이고, 많이 진행되었습니다.
- 항섬유화제 및 가정용산소 치료가 필요합니다.

IPF 진단 후 외래

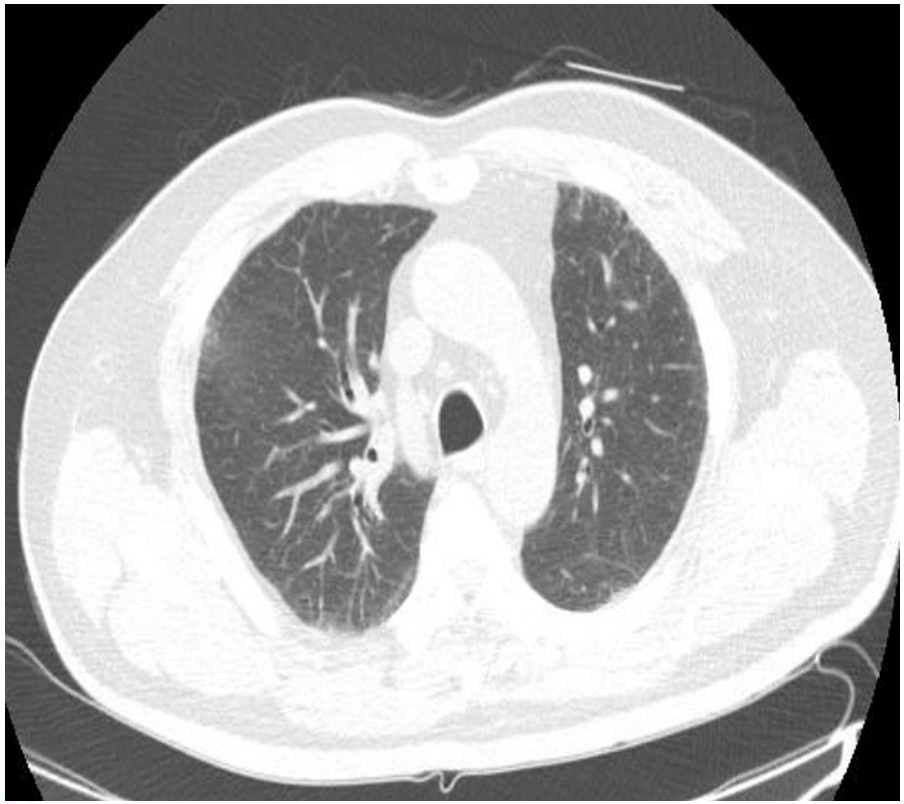
- 증상이 생긴 지는 얼마 안되었는데, 벌써 중증이라고요?
- 별로 불편하지도 않았습니다. 원래 초기에는 이상 증상이 없는 것인가요?
- 건강 검진도 꾸준히 해왔고, 이 병원에서 몇 년 전에 검진으로 CT 검사도 했습니다.
- 그때는 이상이 없었나요?

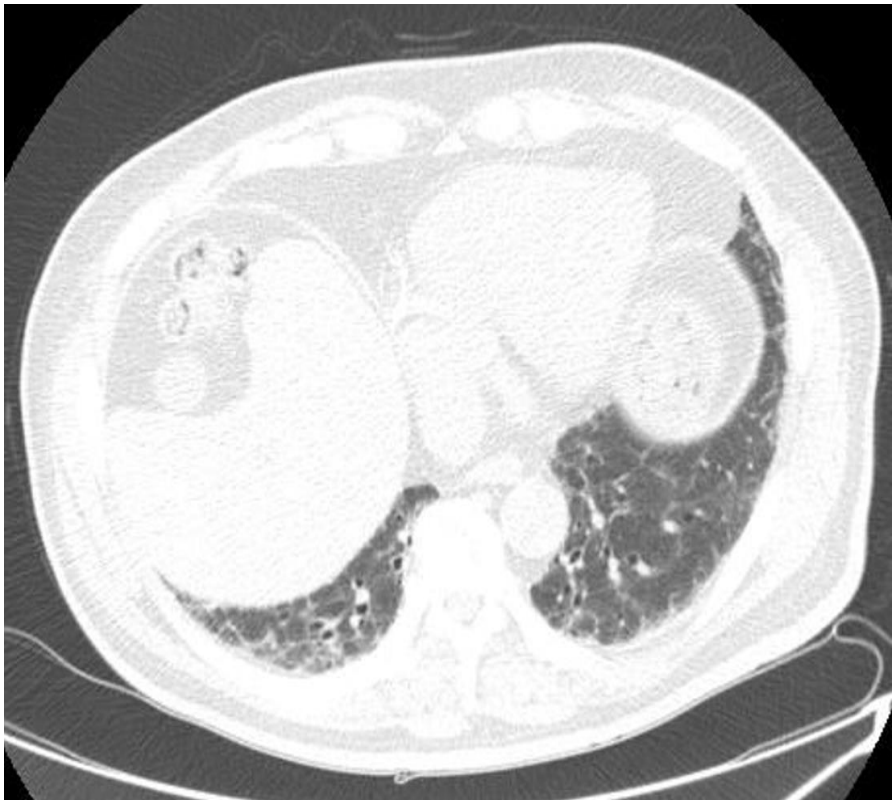
5년 전 검진 결과

2017.07.20

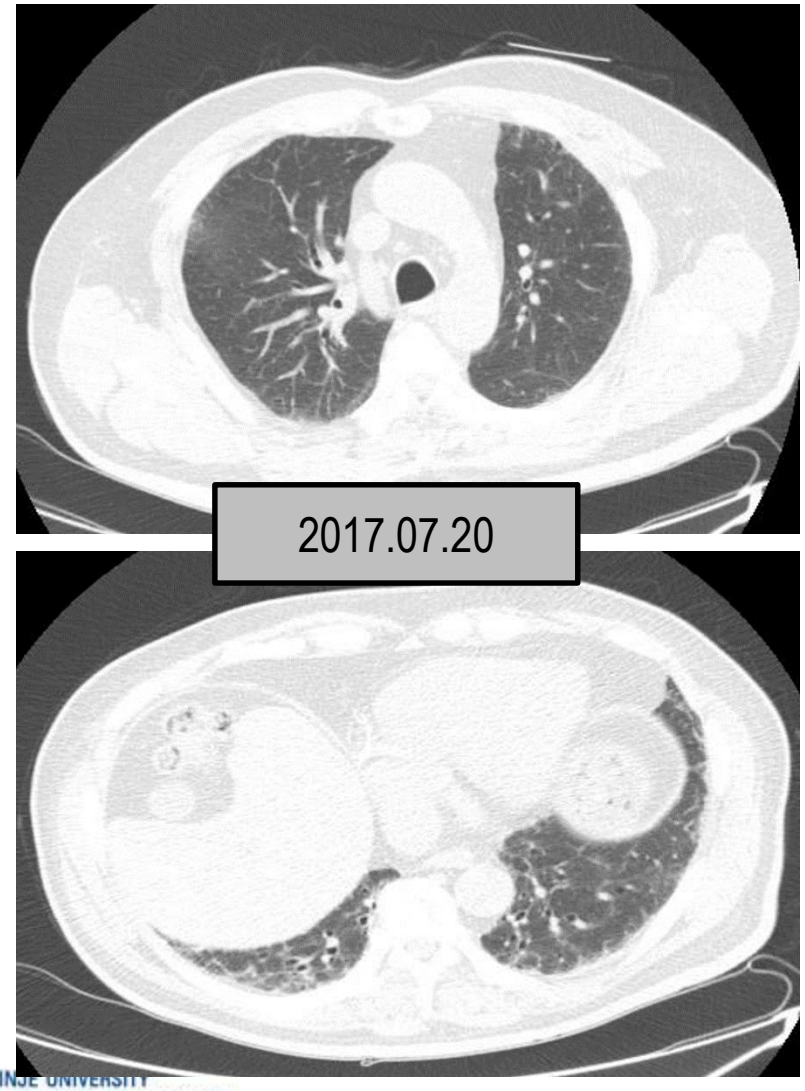
2017년 건강검진으로 LDCT 및 PFT 시행함

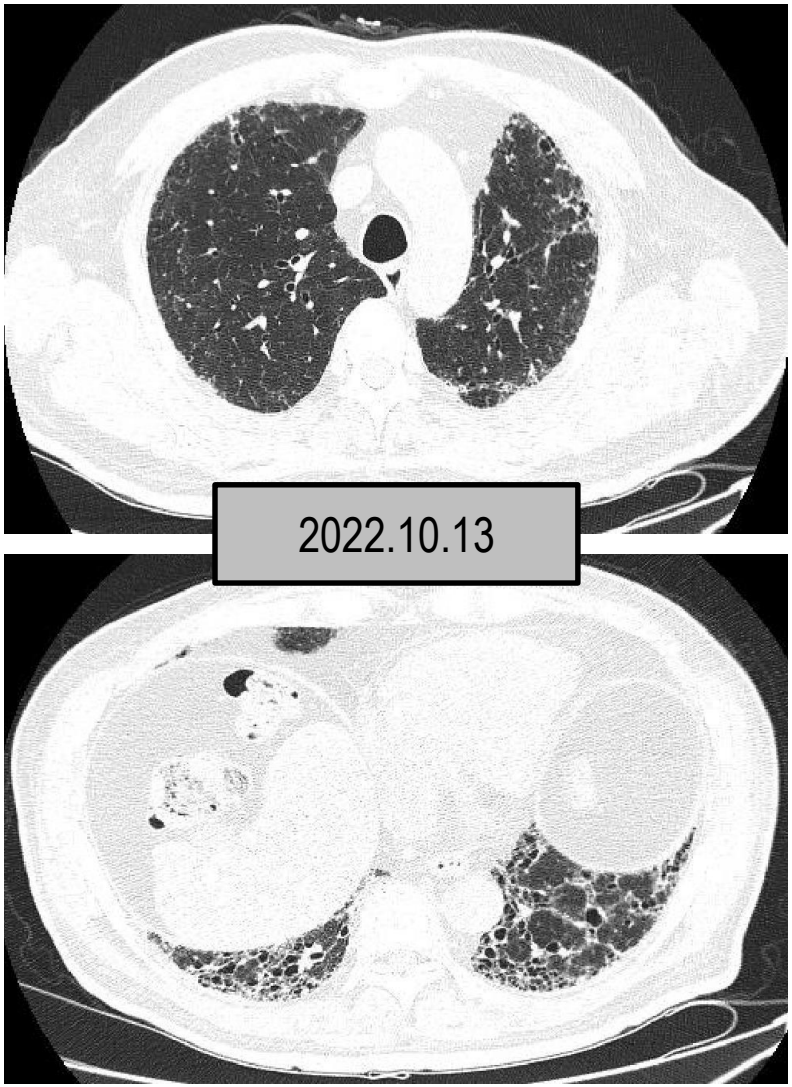
- 호흡기 증상 없었음
- PFT – ratio 76%, FVC 85%
- 정상 폐기능 및 CT 설명 없었음. 추적 검사없이 f/u loss





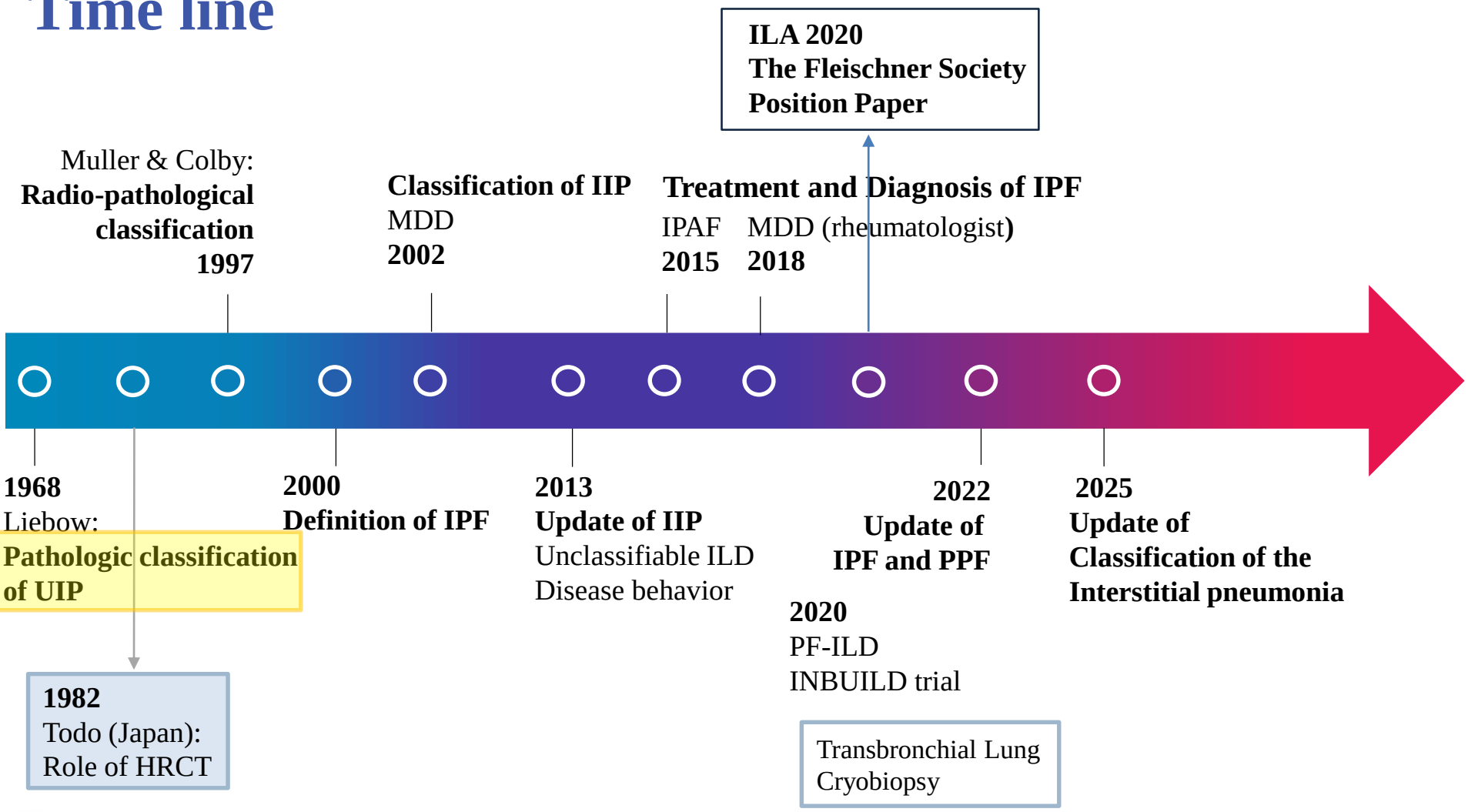
CT progression from ILA to IPF





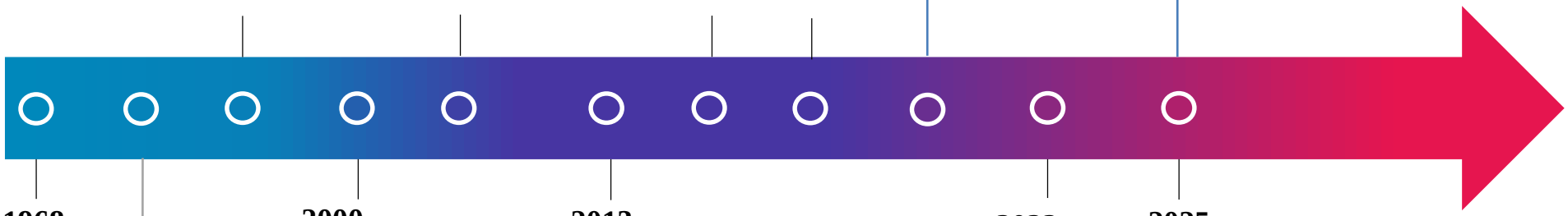
Milestones of ILD classification

Time line



Milestones of ILD classification

Time line



1968
Liebow:
Pathologic classification of UIP

1982
Todo (Japan):
Role of HRCT

Muller & Colby:
Radio-pathological classification
1997

2000
Definition of IPF

Classification of IIP
MDD
2002

2013
Update of IIP
Unclassifiable ILD
Disease behavior

Treatment and Diagnosis of IPF
IPAF **2015** MDD (rheumatologist) **2018**

2020
PF-ILD
INBUILD trial

2022
Update of IPF and PPF

Transbronchial Lung Cryobiopsy

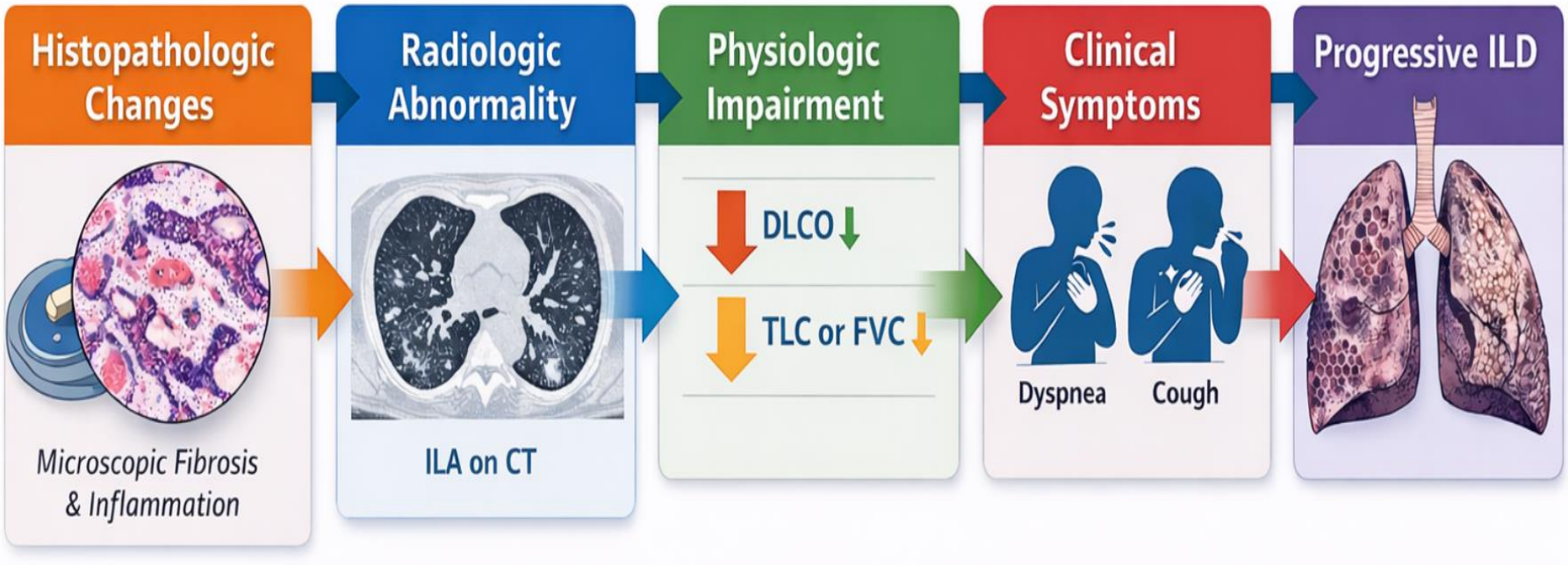
ILA 2020
The Fleischner Society Position Paper

2025 ATS statement
Approach to the Evaluation and Management of ILA

2025
Update of Classification of the Interstitial pneumonia

Trajectory of ILD development

- After pathological change (microscopic fibrosis), Imaging abnormalities may represent the earliest detectable manifestation of interstitial lung disease.
- Pathologic fibrosis can be below the resolution of imaging.



Analysis of the pathologic findings by ILA status

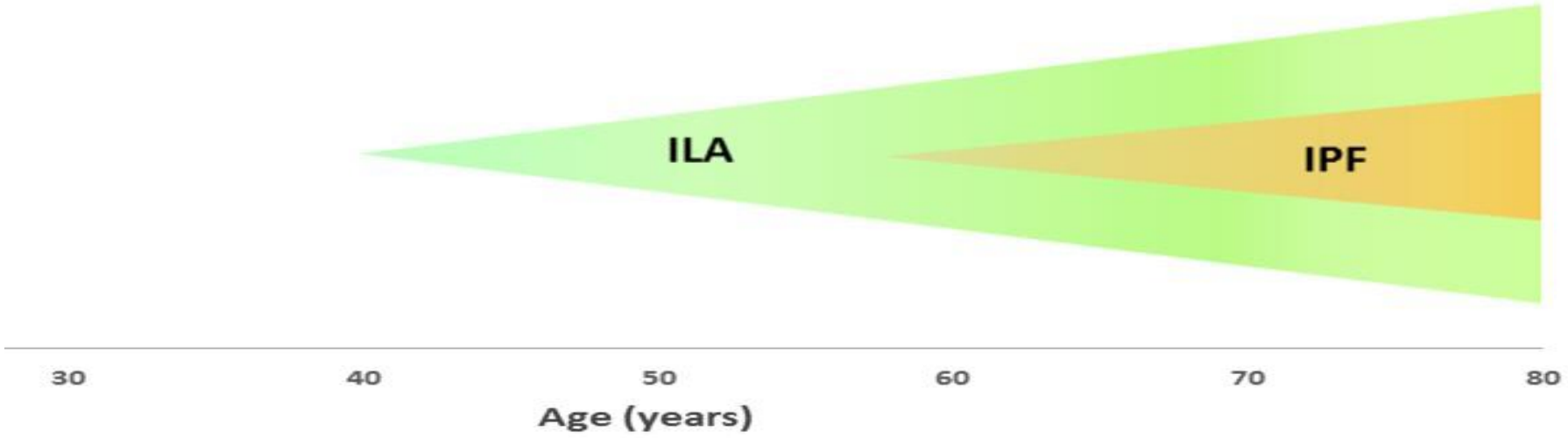
- A retrospective single-cohort study – 424 patients who had undergone lung nodule resection without ILD diagnosis in Brigham and Women's hospital between 2001 and 2015
- ILA on CT and histopathologic findings were compared

	No ILA ($n = 257$; 61%)	Indeterminate ILA ($n = 141$; 33%)	ILA ($n = 26$; 6%)	*P* Value* All Groups	*P* Value* ILA vs. No ILA
Histopathologic features					
Fibrosis					
Any fibrosis present‡, *n* (%)	133 (52%)	74 (52%)	19 (73%)	0.11	0.04
Subpleural fibrosis, *n* (%)	43 (17%)	24 (17%)	12 (46%)	0.003	0.001
Peribronchiolar fibrosis, *n* (%)	62 (24%)	32 (23%)	9 (35%)	0.41	0.24
Interstitial fibrosis, *n* (%)	53 (21%)	30 (21%)	9 (35%)	0.27	0.13
Emphysematous fibrosis§, *n* (%)	39 (15%)	24 (17%)	4 (15%)	0.88	1.0
Additional histopathologic features					
Fibroblastic foci, *n* (%)	9 (4%)	4 (3%)	7 (28%)	0.0001	0.0001
Honeycombing, *n* (%)	0 (0%)	0 (0%)	2 (8%)	0.004	0.008
UIP, *n* (%)	0 (0%)	0 (0%)	2 (8%)	0.004	0.008
Respiratory bronchiolitis, *n* (%)	156 (67%)	89 (71%)	17 (71%)	0.70	0.82
Airways disease‖, *n* (%)	126 (51%)	62 (47%)	11 (48%)	0.73	0.83
Smoking-related interstitial fibrosis¶, *n* (%)	21 (8%)	8 (6%)	1 (4%)	0.66	0.70
Pulmonary arterial hypertensive changes**, *n* (%)	213 (83%)	115 (82%)	23 (92%)	0.47	0.39
Atypical adenomatous hyperplasia, *n* (%)	43 (17%)	36 (26%)	9 (35%)	0.02	0.03
Pigment-laden macrophages, *n* (%)	188 (73%)	105 (75%)	20 (80%)	0.82	0.63
Pleural disease††, *n* (%)	18 (7%)	8 (6%)	3 (13%)	0.43	0.41

AJRCCM 2018; 197(7)

ILA and clinical course

	Population	Clinical findings	Diagnostic criteria
ILA	Any (High risk + Incidental)	None	Clinical-radiologic
Early ILD	Any (Pre-clinical – at high risk; Subclinical – Silent ILD)	Asymptomatic with normal PFTs	Clinical-radiologic-pathologic
Mild ILD	Any (established)	Minor symptoms/trivial PFT abnormalities	Clinical-radiologic-pathologic



Definition: Interstitial lung abnormality (2020)

- Incidental identification of non-dependent abnormalities (CT finding, not disease), including
  - Ground glass abnormality
  - Reticular abnormality
  - Lung distortion
  - Traction bronchiectasis
  - Honeycombing
  - Non-emphysematous cysts

- Involving at least 5% of a lung zone
- In individuals in whom interstitial lung disease is not suspected

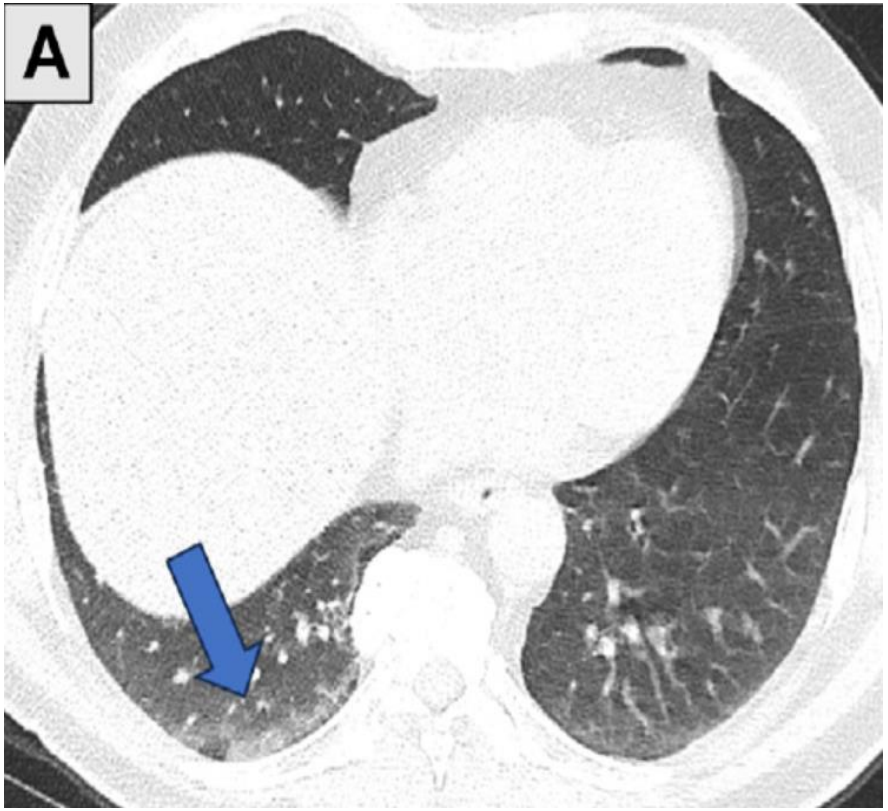
Lancet Respir Med 2020;8:726-37

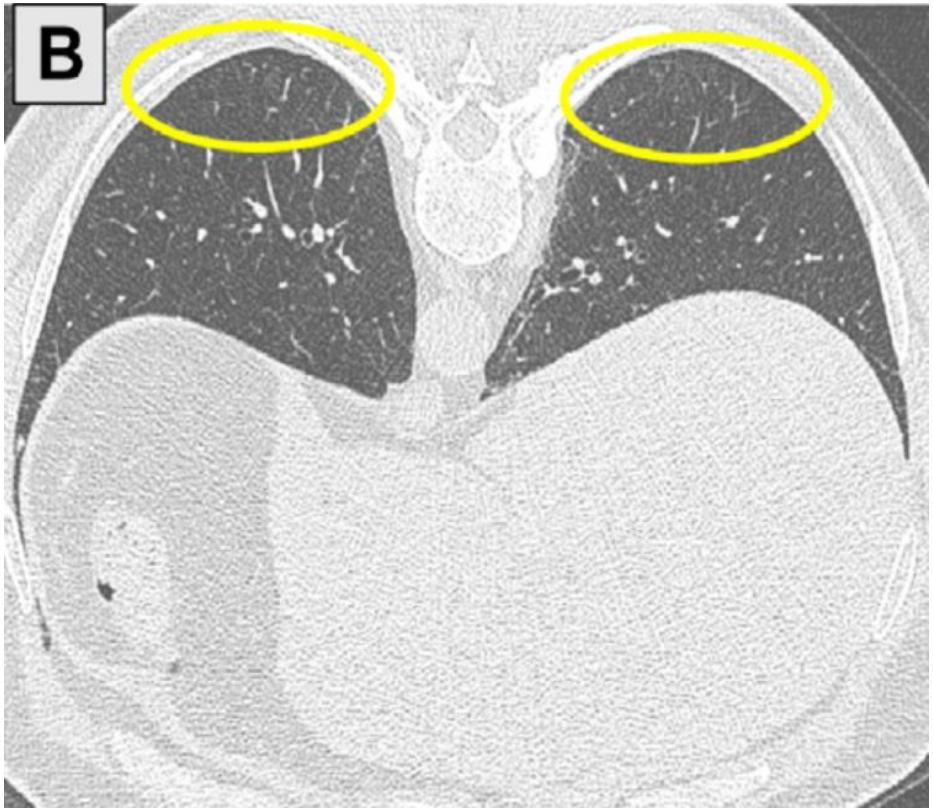
Findings not suggesting ILA (2002)

- Dependent lung atelectasis
- Focal para-spinal fibrosis in contact to spinal osteophytes
- Diffuse (Smoking-related) centrilobular nodularity
- Mild focal or unilateral abnormality
- Interstitial edema
- Findings of aspiration (patch GGO and tree in bud)
- Preclinical interstitial lung abnormalities during screening of high risk subjects (e.q. RA, SSc, occupational exposure, familial PF)

Findings not suggesting ILA

- Dependent lung atelectasis

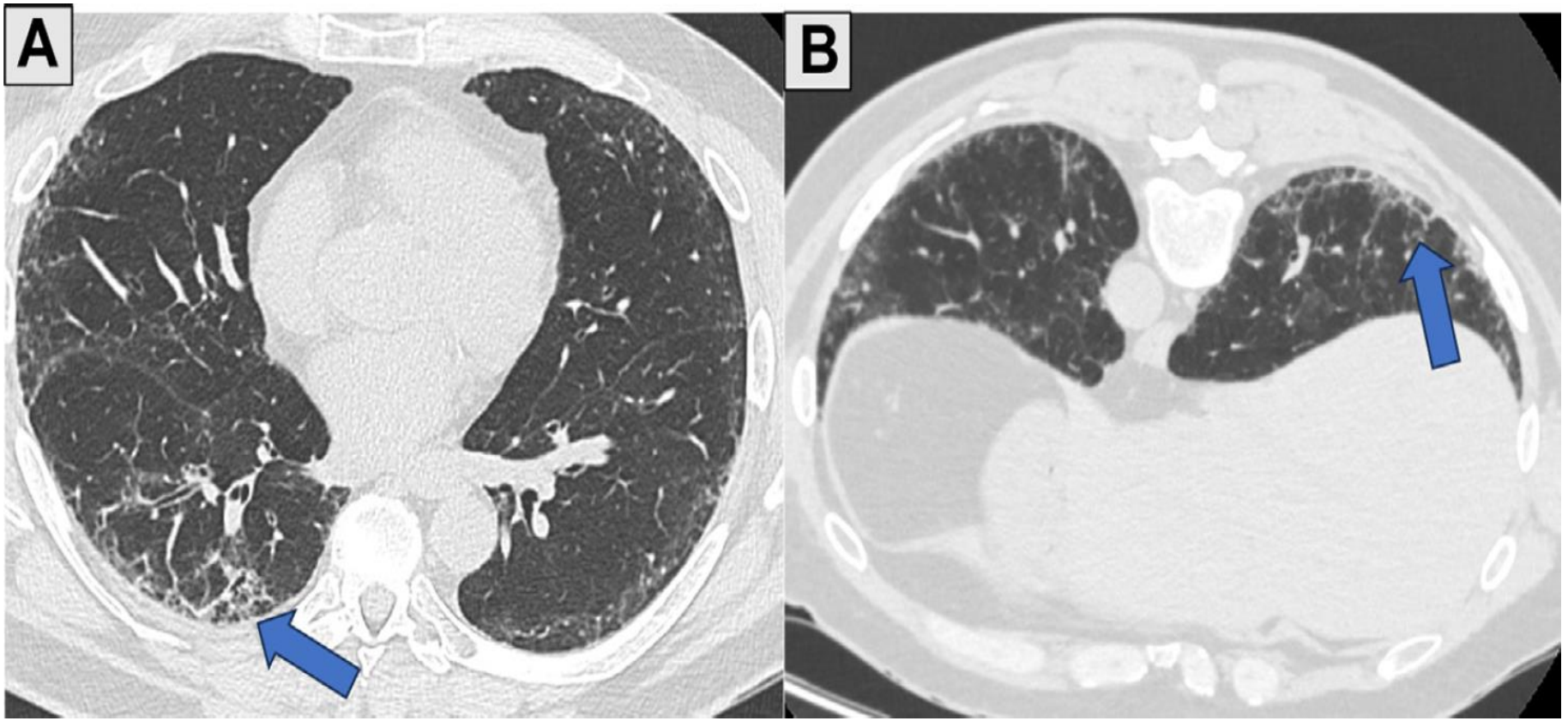




CHEST 2025;167(3):781-799

Findings not suggesting ILA

- True ILA (supine and prone position)

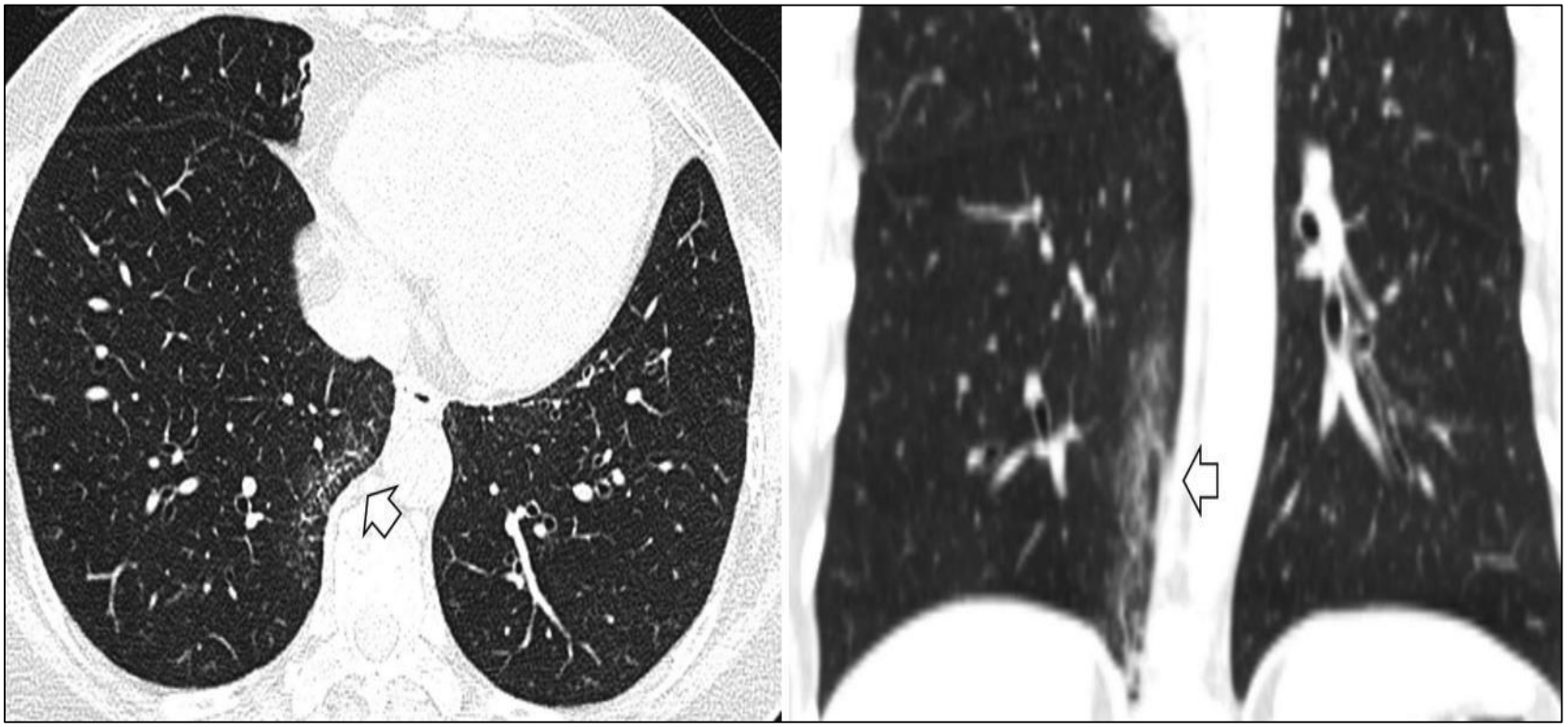


CHEST 2025;167(3):781-799

Findings not suggesting ILA

- Focal para-spinal fibrosis in contact to spinal osteophytes

→ Insignificant focal reactive change

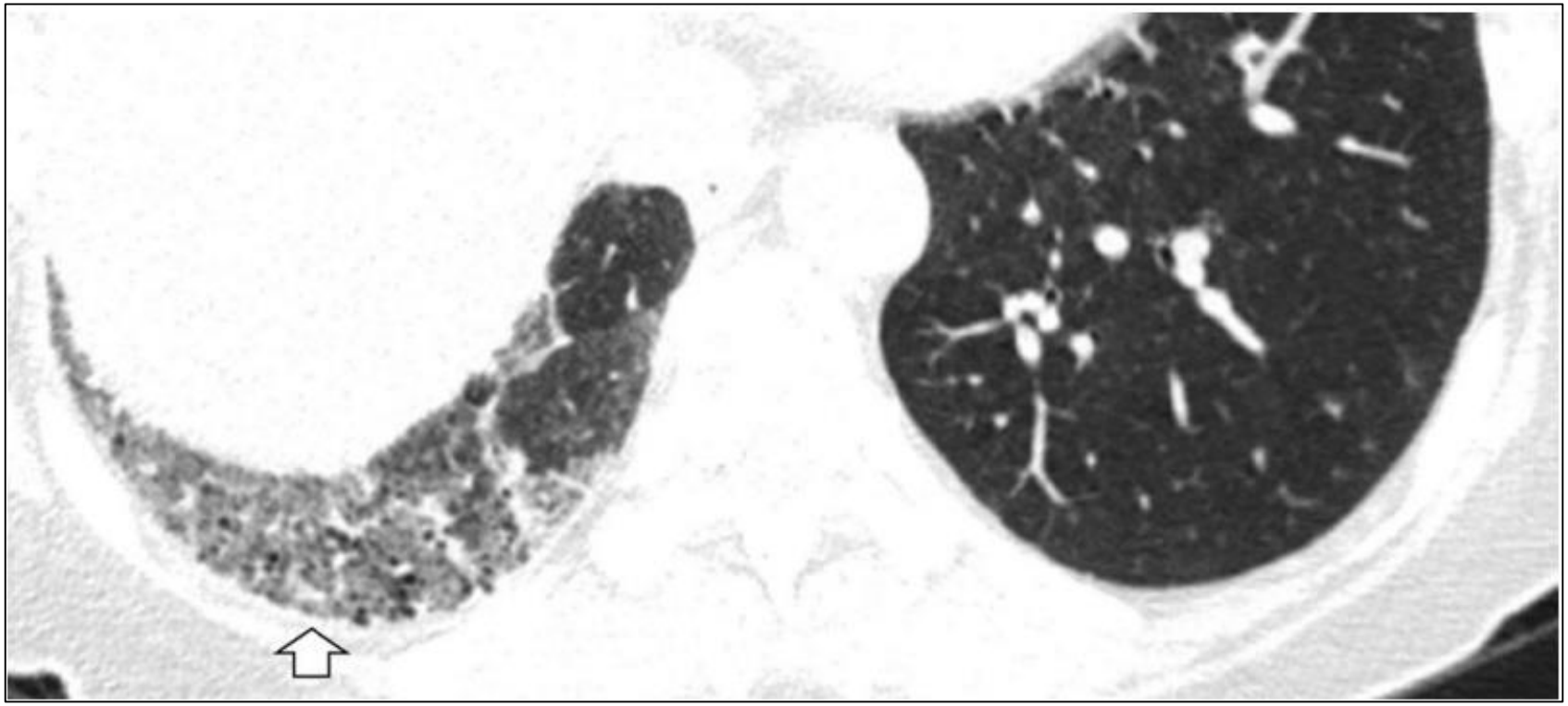


Findings not suggesting ILA

- Mild focal or unilateral abnormality

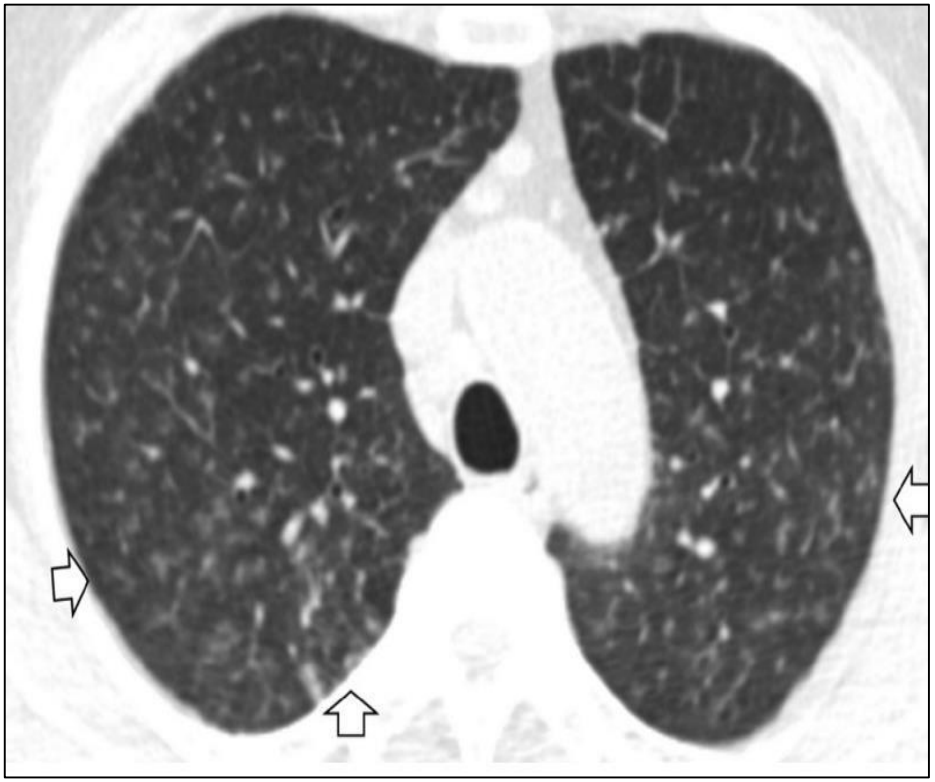
Exceptional considerations

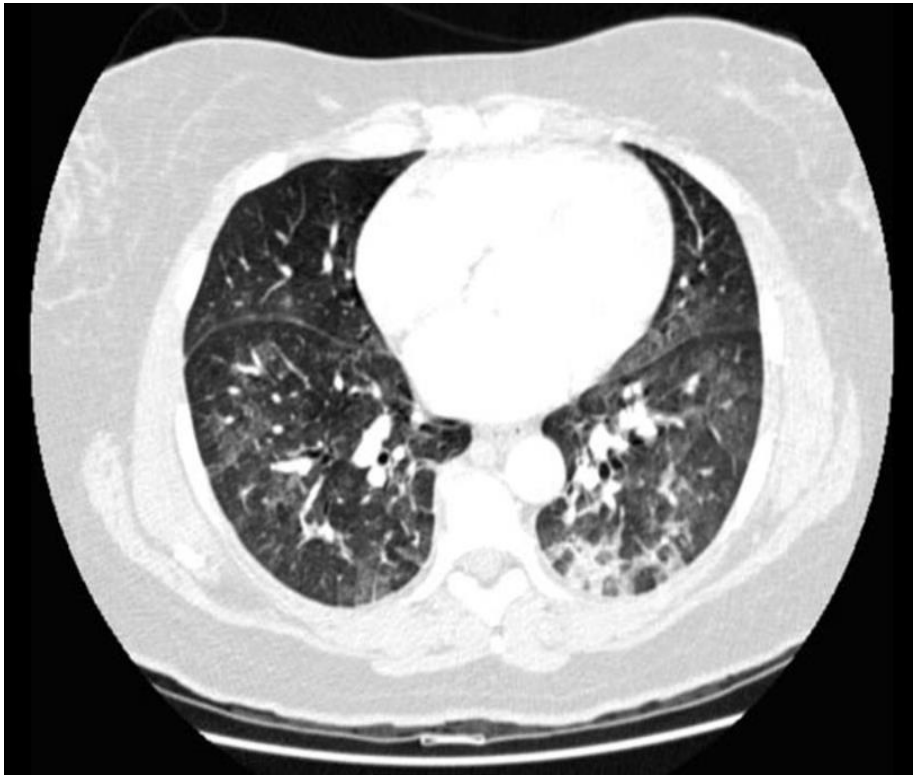
- Early ILA
- In high-risk populations (FPF…)
- Apical fibrosis (PPFE) – not ILA



Findings not suggesting ILA

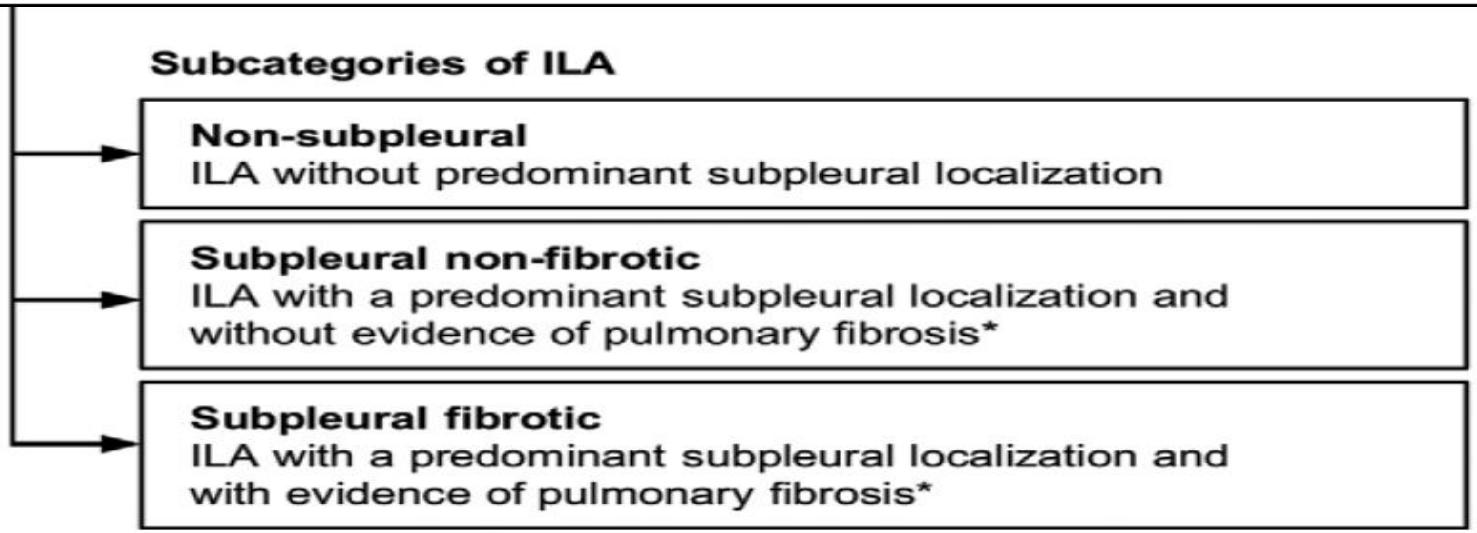
- Findings of centrilobular nodularity (smoking related or aspiration - patch GGO and tree in bud)
- Interstitial edema





Definition and Subtype of ILAs

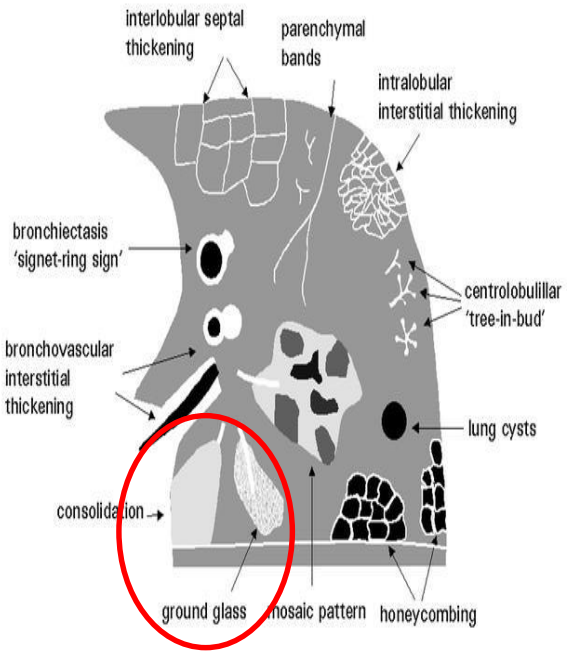
ILA Subtype	Key CT Features	Progression Rates	Key Risk Modifiers
Subpleural fibrotic[1,2,6,15]	Reticulation, traction bronchiectasis, honeycombing	68.8%-89% over 5-8 years; OR-8.4 for progression vs. nonfibrotic	Age, smoking, MUC5B, extent of fibrosis
Subpleural nonfibrotic[1,7]	Ground-glass, mild reticulation, no fibrosis	43.6% over 4.2 years; OR-1.9 if reticulation present	Reticulation, age, smoking
Nonsubpleural[1,8,17]	Nonsubpleural location, no fibrosis	Lower; not well quantified, but <20% over 5 years	Male sex, reduced FVC, age, smoking status, inhalational exposure

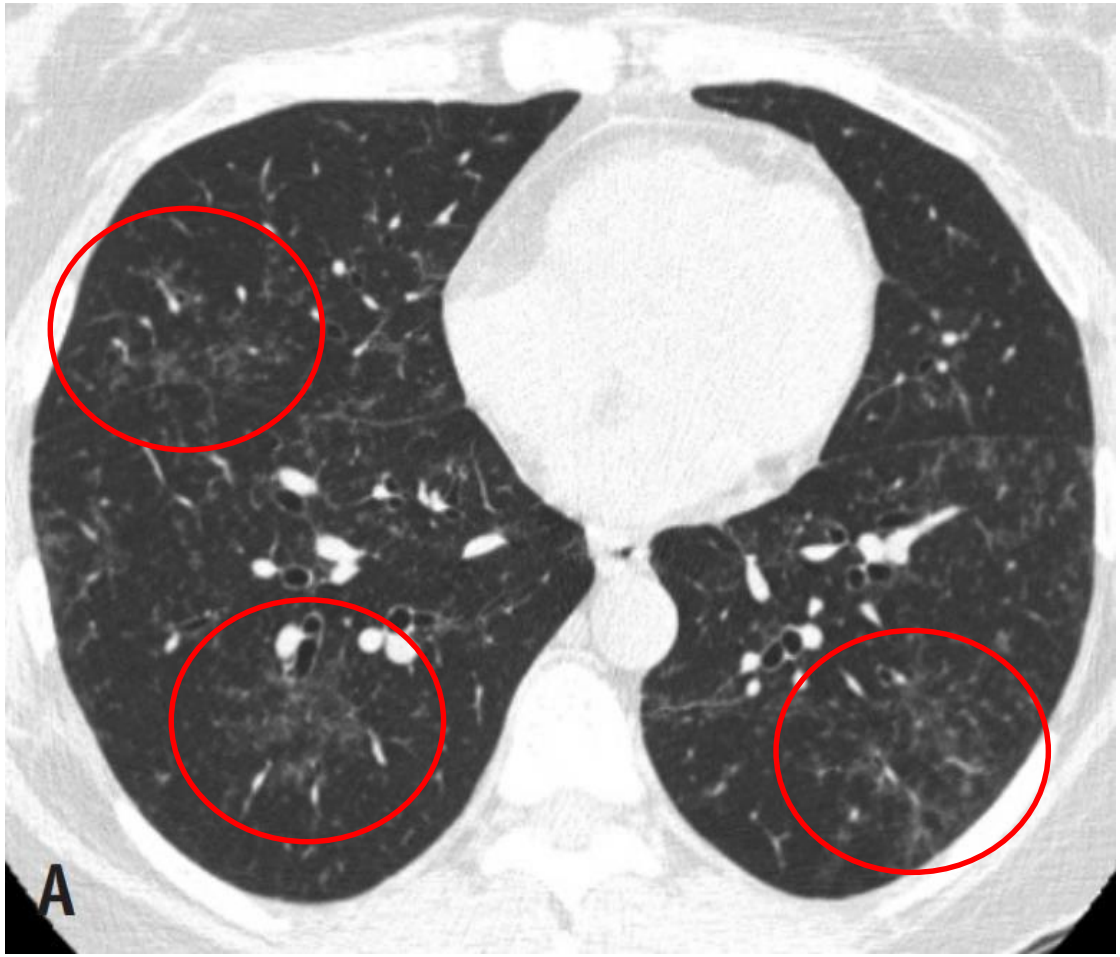


Radiology 2021;301:19-34

Non-subpleural (non-fibrotic) ILA

"Without predominant subpleural localization, Non-fibrotic feature"

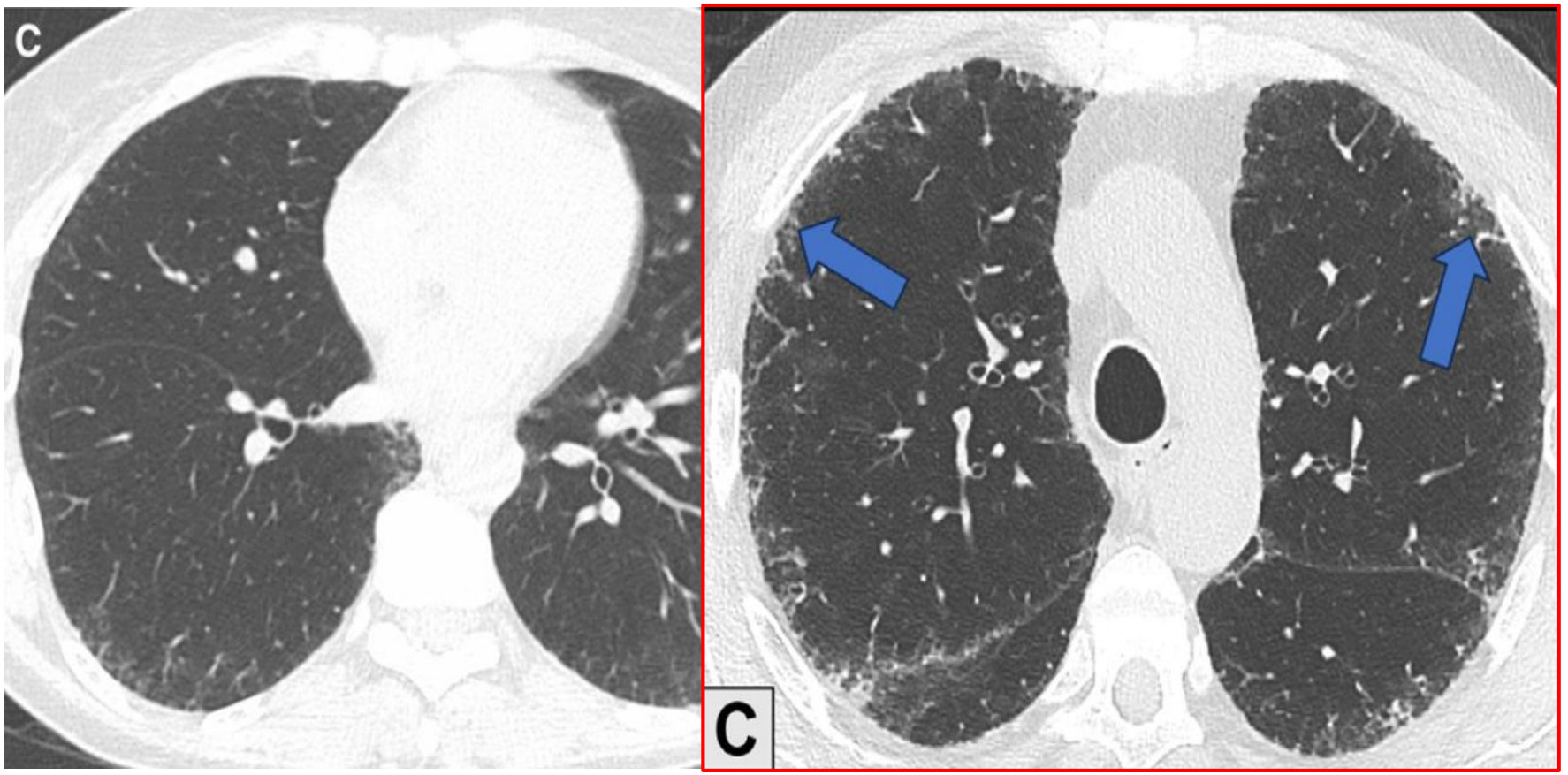




Korean J Radiol 2021;22(5):811-828

Subpleural non-fibrotic ILA

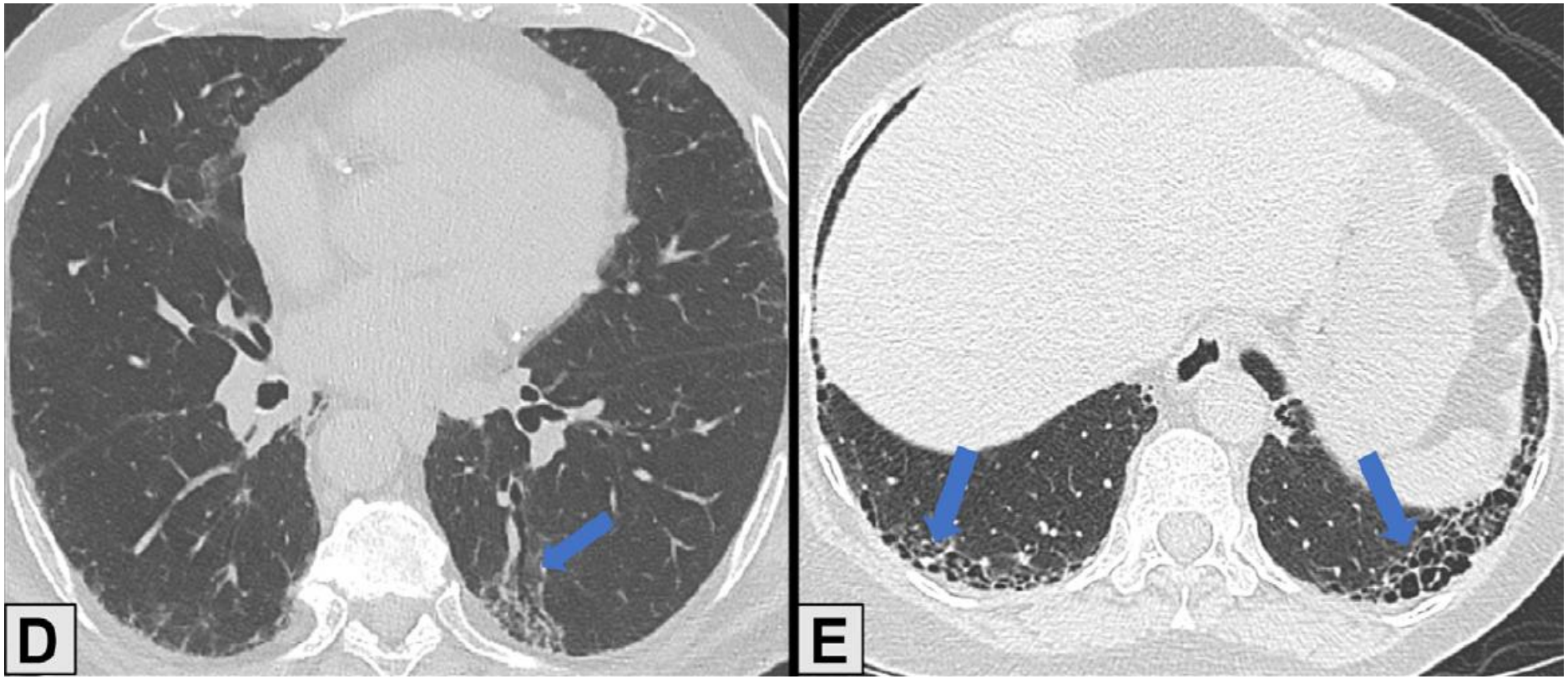
- Predominant subpleural location, without evidence of fibrosis
- <u>GGO & Reticulation</u>



CHEST 2025;167(3):781-799

Subpleural fibrotic ILA

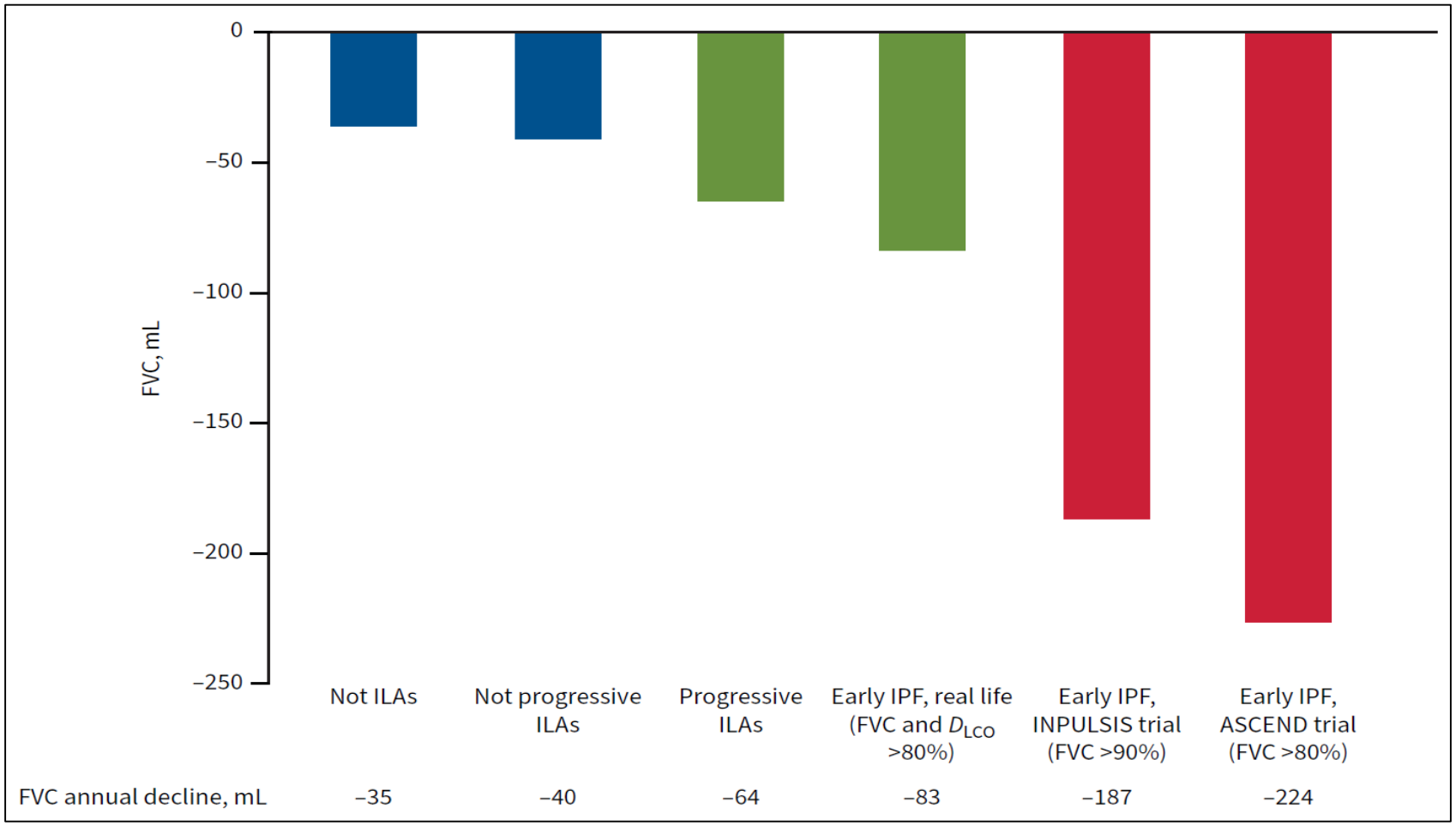
- Predominant subpleural location
- Architectural distortion with traction bronchiectasis or honeycombing



CHEST 2025;167(3):781-799

Clinical Relevance of ILAs

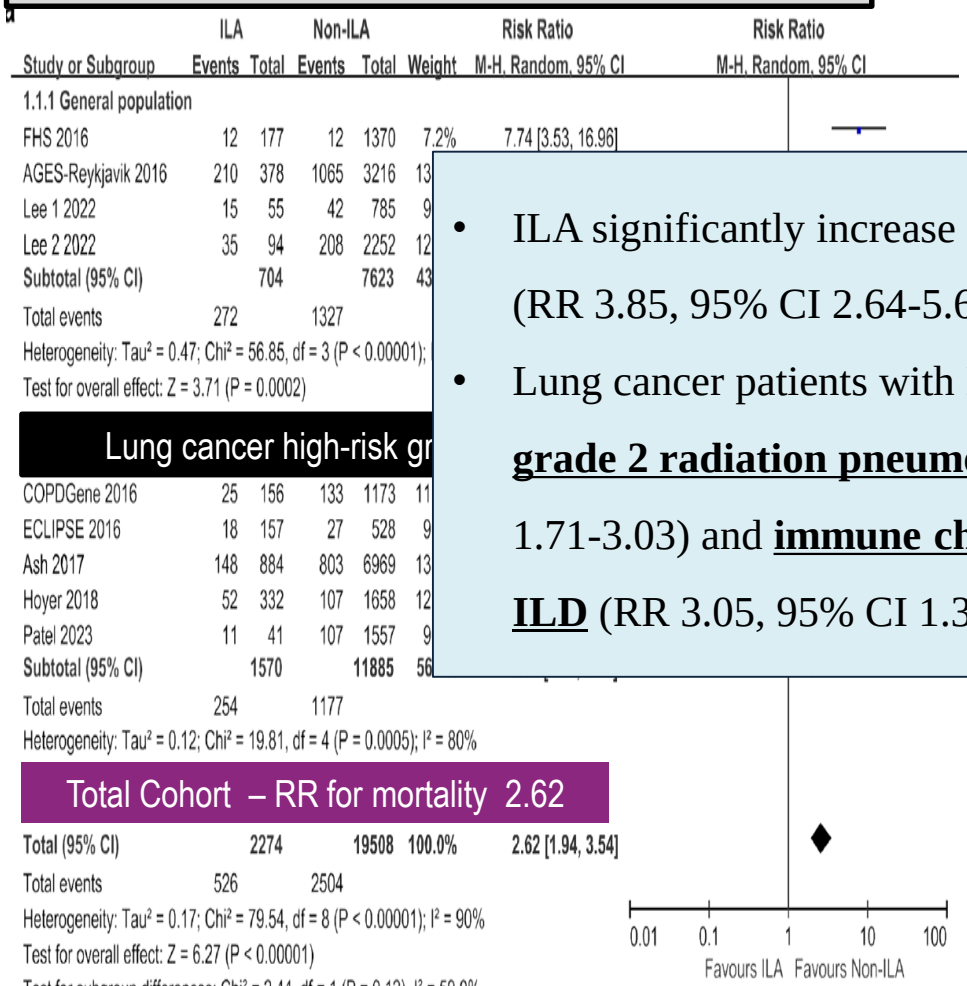
ILAs are associated



AJRCCM 2025;211(7):1132-1155

Clinical Outcomes of ILA
: A Systematic review and Meta-analysis

General Population Cohort – RR for morality 3.83

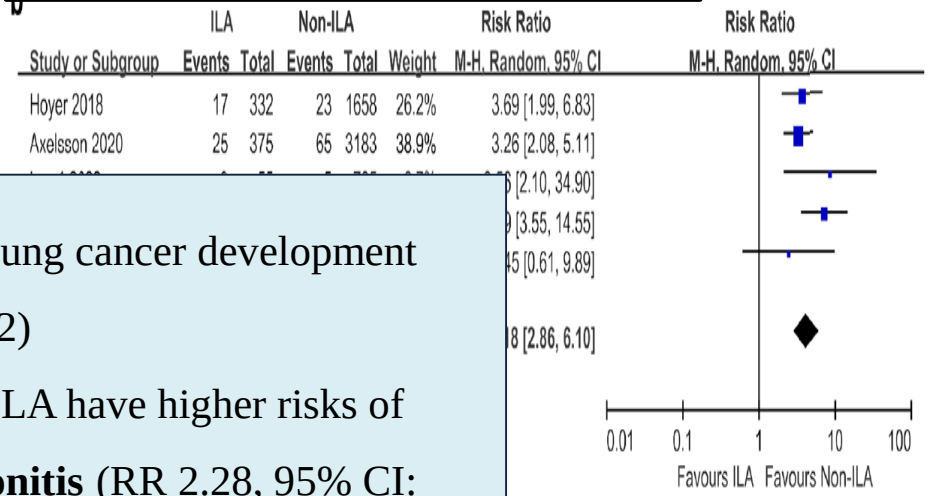


Lung cancer high-risk gr

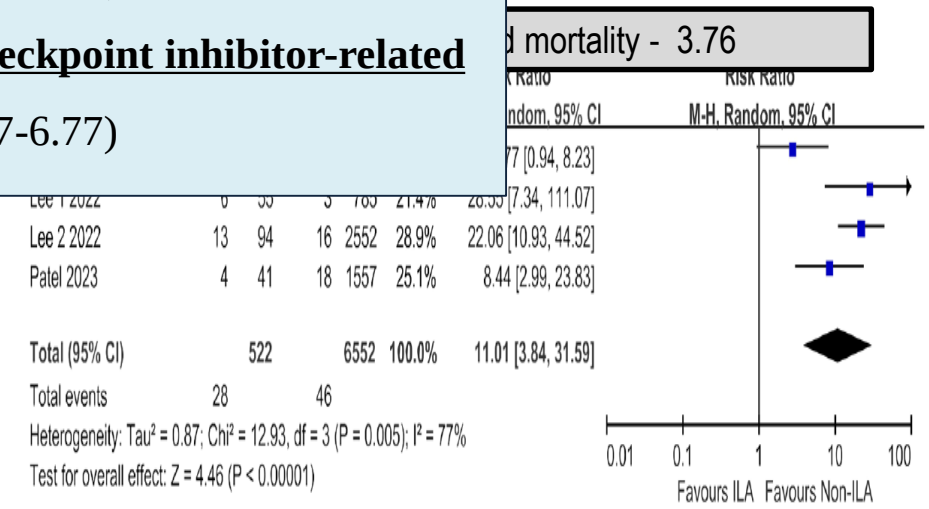
Total Cohort – RR for mortality 2.62

- ILA significantly increase lung cancer development (RR 3.85, 95% CI 2.64-5.62)
- Lung cancer patients with ILA have higher risks of **grade 2 radiation pneumonitis** (RR 2.28, 95% CI: 1.71-3.03) and **immune checkpoint inhibitor-related ILD** (RR 3.05, 95% CI 1.37-6.77)

Lung cancer-related mortality– RR 4.18



d mortality - 3.76



INJE UNIVERSITY
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AJRCCM 2023;208(6):695-708

Approach to the Evaluation and Management of Interstitial Lung Abnormalities

An Official American Thoracic Society Clinical Statement

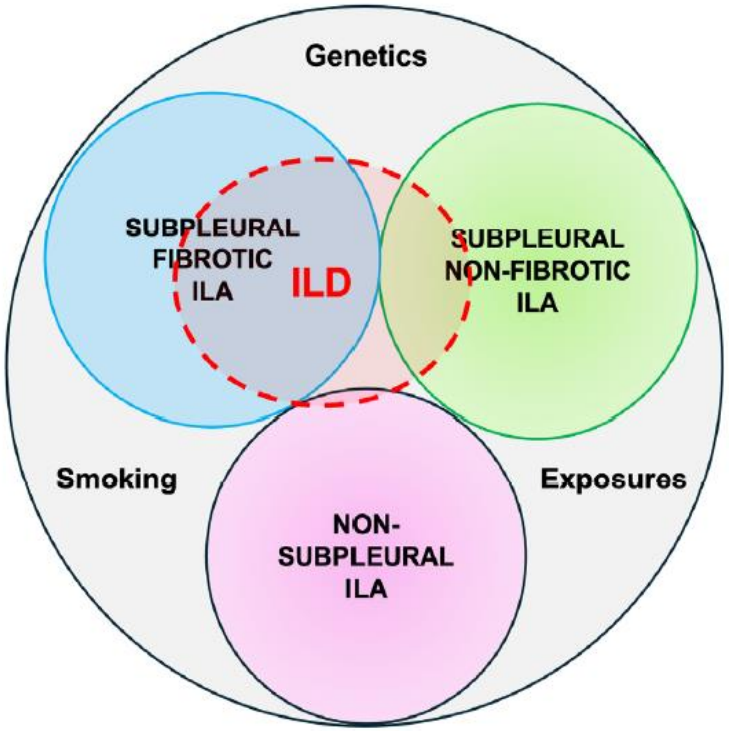
- Definition of ILA: Non-dependent bilateral parenchymal abnormalities detected on CT (ground-glass or reticular abnormalities, lung distortion, traction bronchiectasis, and/or honeycombing) involving ≥ 5% of a lung zone by visual estimate, but without meeting the criteria for ILD.

ILA

involving at least 5% of a lung zone, NOT suspected ILD

Subpleural distribution	**Non-subpleural distribution**
Likely to progress	*Unlikely to progress*

Ground-glass
Reticular abnormalities
Non-emphysematous cysts
Honeycombing
Traction bronchiectasis/bronchiolectasis



AJRCCM 2025;211(7):1132-1155

From Radiologic definition to Clinical Framework : Evolution of ILA (2020→2025)

- The 2020 Fleischner paper – ILA (a radiologic entity) without clinical information
- The 2025 ATS statement – A multidisciplinary (including pulmonology and pathology) clinical framework for evaluation, classification, and management of ILA in real practice

  1) **Removal of incidental and high-risk group restrictions**

  2) Clear distinction between ILA and ILD

  3) Suggestion of screening, baseline assessment, and follow-up strategies

AJRCCM 2025;211(7):1132-1155

Chest CT screening to identify ILAs/ILD in smokers

- The estimated prevalence of ILAs/ILD: 8% (95% CI, 7-10%)
- Advanced age, smoking(OR 1,7-2.2), and male sex – A common features of IPF
- Screening for ILA in all-smokers: not recommended d/t cost burden, anxiety, undesirable anxiety, radiation exposure, and the need for additional test

❖ However, in the context of lung cancer screening, systematic assessment and documentation of ILAs/ILD are recommended.

(The U.S. Preventive Service Task Recommendation of lung cancer screening – aged 50-80 years who have a 20-pack-year smoking history of current smoke or quit within the previous 15 years) → overlapping risk profiles of ILAs/ILD

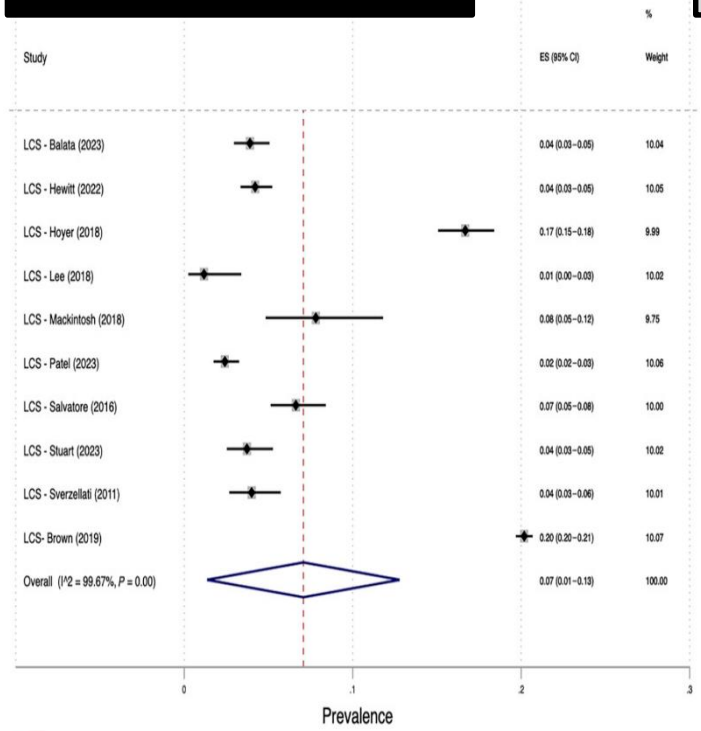
AJRCCM 2025;211(7):1132-1155

Prevalence, Risk Factors, and Outcomes of Adult Interstitial Lung Abnormalities

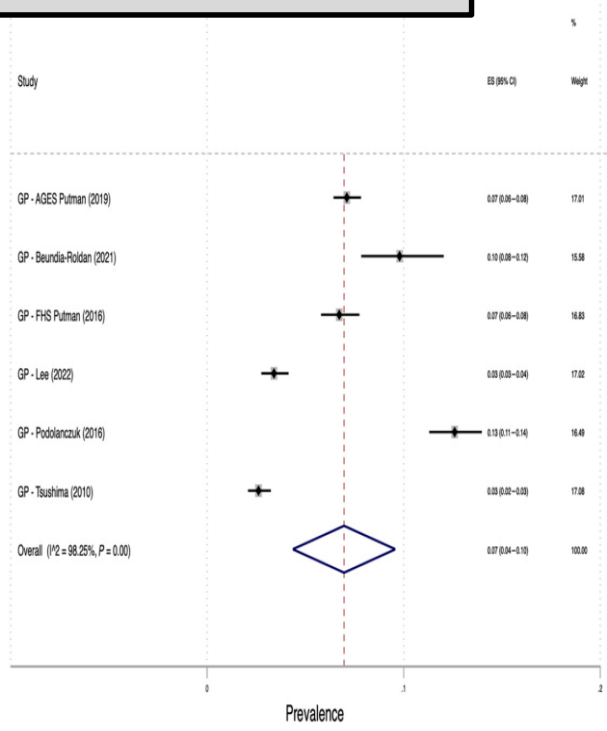
A Systematic Review and Meta-Analysis

- Meta-analyses of observational studies
- Inclusion – 22 studies and 88,325 participants
- Aim to estimate pooled prevalence, risk of ILAs, and ILA-associated mortality risk

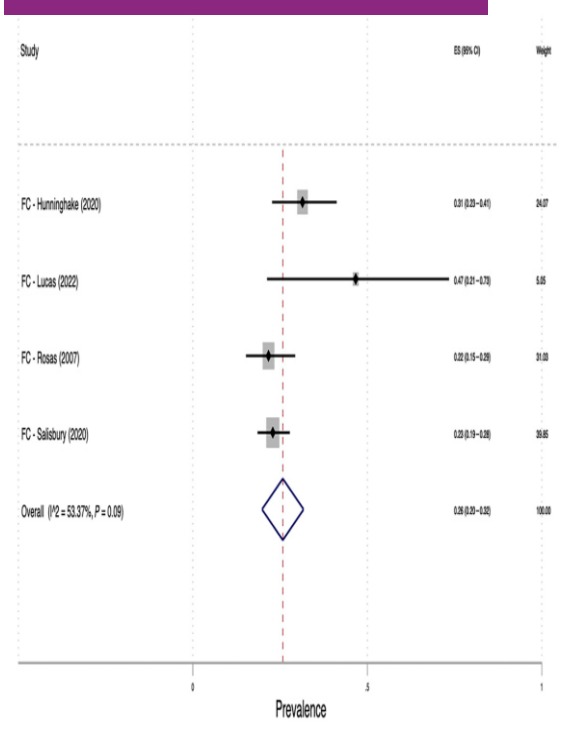
Lung cancer screening – 7%



General Population Cohort – 7%



At-risk Familial Cohort – 26%



INJE UNIVERSITY HAEUNDAE PAIK HOSPITAL

AJRCCM 2023;208(6):695-708

Chest CT screening to identify ILAs/ILD in CTD

- In meta-analysis, the estimated prevalence of ILAs/ILD (RA, systemic sclerosis, polymyositis, dermatomyositis, anti-synthetase syndrome, mixed CTD, Sjogren's disease or overlap syndrome: **40% (95% CI, 37-43%)**
- Rheumatoid arthritis – 23% (95% CI, 17-29%)
- Systemic sclerosis – 45% (95% CI, 42-49%)
- Sjogren's syndrome – 39% (95% CI, 18-59%)
- Dermatomyositis/Polymyositis – 44% (95% CI, 37-52%)

→ Additional benefit – An indication for the initiation or escalation of immunomodulatory therapy targeting the underlying CTD

❖ We suggest a baseline HRCT to screen for ILAs/ILD in adults with CTD

AJRCCM 2025;211(7):1132-1155

Chest CT screening to identify ILAs/ILD in CTD

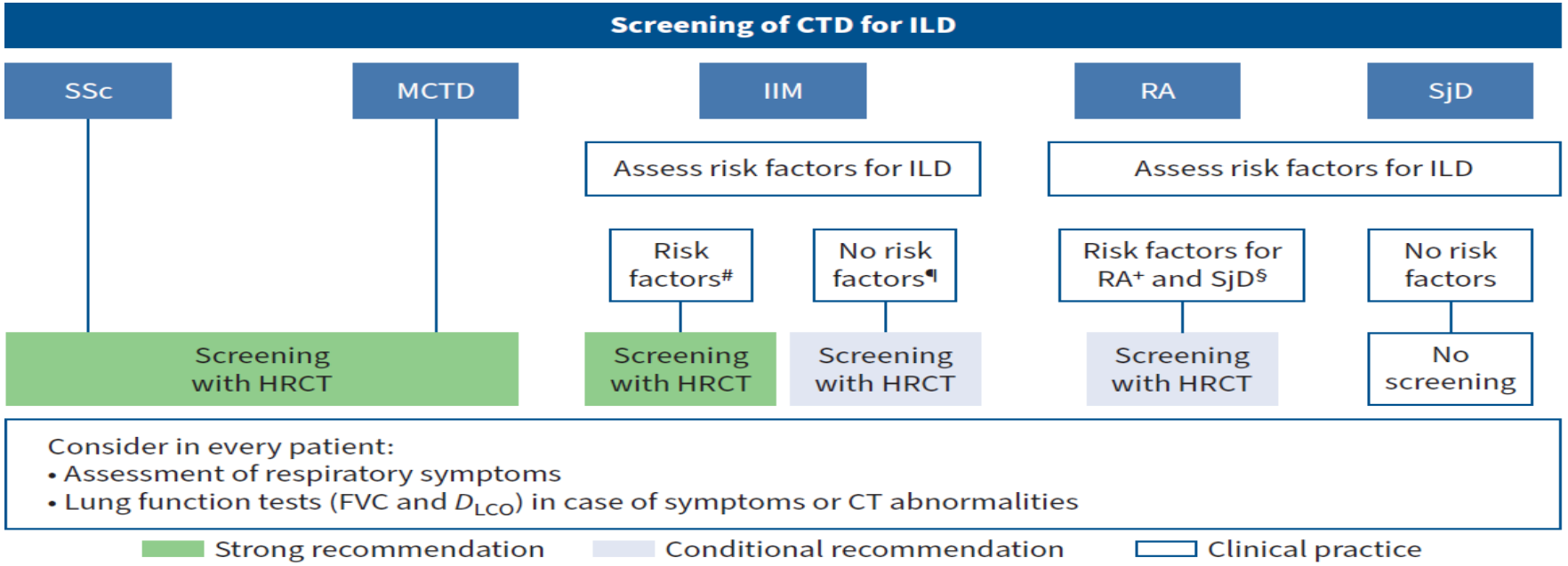
- In meta-analysis, the estimated prevalence of ILAs/ILD (RA, systemic sclerosis, polymyositis, dermatomyositis, anti-synthetase syndrome, mixed CTD, Sjogren's disease or overlap syndrome: **40% (95% CI, 37-43%)**
- Rheumatoid arthritis – 23% (95% CI, 17-29%)
- Systemic sclerosis – 45% (95% CI, 42-49%)
- Sjogren's syndrome – 39% (95% CI, 18-59%)
- Dermatomyositis/Polymyositis – 44% (95% CI, 37-52%)

→ Additional benefit – An indication for the initiation or escalation of immunomodulatory therapy targeting the underlying CTD

❖ We suggest a baseline HRCT to screen for ILAs/ILD in adults with CTD

AJRCCM 2025;211(7):1132-1155

ERS/EULAR clinical practice guidelines for connective tissue disease-associated interstitial lung disease



	SSc	RA	IIM	SjD
Demographics	• Longer disease duration	• Older age • Male sex • Smoking	• Older age	• Older age • Male sex
Circulating markers	• Increased KL-6 • Presence of ATA-I	• Increased ESR • Presence of anti-CCP, RF	• Increased CRP, ESR • Presence of anti-Jo1, anti-MDA-5, anti-Ro52	• Increased CRP • Presence of anti-Ro52
Extrapulmonary involvement	• Diffuse cutaneous SSc • Higher mRSS	• Higher articular disease activity	• Anti-synthetase syndrome • Clinical amyopathic dermatomyositis • Skin involvement# • Arthritis/arthralgia	• Presence of extrapulmonary involvement

Chest CT screening to identify ILAs/ILD in a first-degree relative with pulmonary fibrosis

- Familial Pulmonary Fibrosis – at least two genetically related first or second degree relatives with fibrotic ILD
- In meta-analysis, the estimated prevalence of ILAs/ILD in the first-degree relatives of IPF- **26% (95% CI, 18-34%)**

❖ We suggest a chest CT screening for ILAs/ILD in adults ≥ 50 years who have a first-degree relative with FPF

❖ We recommended neither for nor against chest CT screening for ILAs/ILD in adults ≥ 50 years who have a first-degree relative with IPF and no other known family members with ILD

AJRCCM 2025;211(7):1132-1155

Chest CT screening to identify ILAs/ILD in a first-degree relative with pulmonary fibrosis

- Familial Pulmonary Fibrosis – at least two genetically related first or second degree relatives with fibrotic ILD
- In meta-analysis, the estimated prevalence of ILAs/ILD in the first-degree relatives of IPF- **26% (95% CI, 18-34%)**

❖ We suggest a chest CT screening for ILAs/ILD in adults ≥ **50 years who** have **a first-degree relative with FPF**

❖ We recommended neither for nor against chest CT screening for ILAs/ILD in adults ≥ 50 years who have a first-degree relative with IPF and no other known family members with ILD

AJRCCM 2025;211(7):1132-1155

MUC5B testing and Telomere length measurement screening to identify ILAs/ILD in a first-degree relative with pulmonary fibrosis (including FPF)

- In adults with a first-degree relative with pulmonary fibrosis, the prevalence of ILAs/ILD among those with the *MUC5B* promoter variant was 38% (95% CI, 24-52%) with a sensitivity of 56% and a specificity of 60% for prediction.
- The prevalence of ILAs/ILD among those with a telomere length than the 10th percentile for age was 44% (95% CI, 27-61%) in only one study

❖ We suggest not performing *MUC5B* and telomere length measurement as an alternative test to CT screening or as a preliminary test.

→ The tests should not act as a gatekeeper to obtaining a CT d/t insufficient sensitivity & specificity, small evidence, cost, affordability, and difference between races.

AJRCCM 2025;211(7):1132-1155

From Radiologic definition to Clinical Framework : Evolution of ILA (2020→2025)

- The 2020 Fleischner paper – ILA (a radiologic entity) without clinical information
- The 2025 ATS statement – A multidisciplinary (including pulmonology and pathology) clinical framework for evaluation, classification, and management of ILA in real practice

  1) Removal of incidental and high-risk group restrictions

  2) **Clear distinction between ILA and ILD**

  3) Suggestion of screening, baseline assessment, and follow-up strategies

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Definition of ILD for those with ILAs

The importance of distinguishing between ILA and ILD

1. Simple and Easy application (As defined 2022 international guideline)
2. Progressive abnormalities (not aging)
3. The presence of a major fibrotic ILD pattern

Definition of interstitial lung disease for those with ILAs

In a person with CT features of ILAs, at least one of the following criteria must be present to define ILD*

- Symptoms: Any amount of dyspnea and/or cough that a clinician attributes to ILD

- Physiology (any of)
  - Any abnormality in FVC, TLC, or D_{LCO} that a clinician attributes to ILD (defined as a value or z-score below the lower limit of normal)
  - Satisfies physiologic criteria for progressive pulmonary fibrosis that a clinician attributes to ILD (10)

- Imaging (any of the following on chest CT)
  - Fibrotic abnormalities (honeycombing and/or reticulation with traction bronchiectasis) involving ≥5% of total lung volume by visual estimate
  - Progressive fibrotic abnormality on serial chest CT
  - Presence of a major fibrotic ILD pattern on chest CT (i.e., UIP/probable UIP, fibrotic HP, or fibrotic NSIP)

- Pathology: Presence of a major fibrotic ILD pattern (i.e., UIP/probable UIP, fibrotic HP, or fibrotic NSIP)

AJRCCM 2025;211(7):1132-1155

From Radiologic definition to Clinical Framework : Evolution of ILA (2020→2025)

- The 2020 Fleischner paper – ILA (a radiologic entity) without clinical information
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  1) Removal of incidental and high-risk group restrictions

  2) Clear distinction between ILA and ILD

  3) **Suggestion of screening, baseline assessment, and follow-up strategies**

AJRCCM 2025;211(7):1132-1155

Longitudinal follow-up assessment with serial CT

- The purpose of follow-up imaging is to identify progression to ILD for diagnosis and therapeutic decision
- The prevalence of **radiologic progression** – **46%** (95% CI, 38-55%), and a positive association with **mortality (HR 1.9**; 95% CI, 1.3-2.8)
- In a previous study, **the median time to ILA progression was 3.2 years**

❖ We suggest that patients with ILAs undergo a follow-up chest CT 2-3 years after the baseline chest CT

❖ Earlier follow-up (12 month) in some high risks or some clinical contests

❖ No vote, but most committee members agreed follow-up of annual PFT

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ERS/EULAR clinical practice guidelines for connective tissue disease-associated interstitial lung disease

TABLE 4 Risk factors for poor outcome, defined as disease progression and death, in patients with connective tissue disease (CTD)-associated interstitial lung disease (ILD) and rheumatoid arthritis (RA)-associated ILD

	SSc[#]	RA[#]	IIM[#,¶]
Demographics	• Older age • Male sex • African American ethnicity	• Older age at RA onset • Male sex	
Circulating markers	• Elevated ESR, CRP • ATA-I	• Anti-CCP, RF	• Elevated ferritin • Anti-MDA-5, anti-synthetase
Pulmonary function/markers	• Baseline PFTs (FVC, D_{LCO})	• Baseline PFTs (low FVC and/or D_{LCO})	
Imaging/histology	• Higher extent of ILD on HRCT	• UIP and probable UIP HRCT/histological patterns • Higher extent of ILD on HRCT	• Higher extent of ILD on HRCT and ILD pattern on HRCT
Extrapulmonary involvement	• Recent onset of SSc with rapid skin progression, more extensive skin fibrosis (mRSS)	• Higher articular disease activity	

Approach to the Evaluation and Management of Interstitial Lung Abnormalities
An Official American Thoracic Society Clinical Statement

High-risk ILA features

Demographic and clinical factors

- Family history of pulmonary fibrosis
- Older age
- Smoking history
- Other inhaled exposures (e.g., occupational vapors, gases, dusts, and fumes; air pollution)
- Connective tissue disease

Genetic

- MUC5B promoter variant
- Leukocyte telomere length below age-adjusted 10th percentile

Imaging

- Definite fibrosis on CT (e.g., honeycombing, traction bronchiectasis, distortion)
- Subpleural fibrotic and nonfibrotic subtypes
- Greater extent of abnormalities (multiple lung zones)

Physiologic

- Abnormal or borderline FVC (TLC) and DLco

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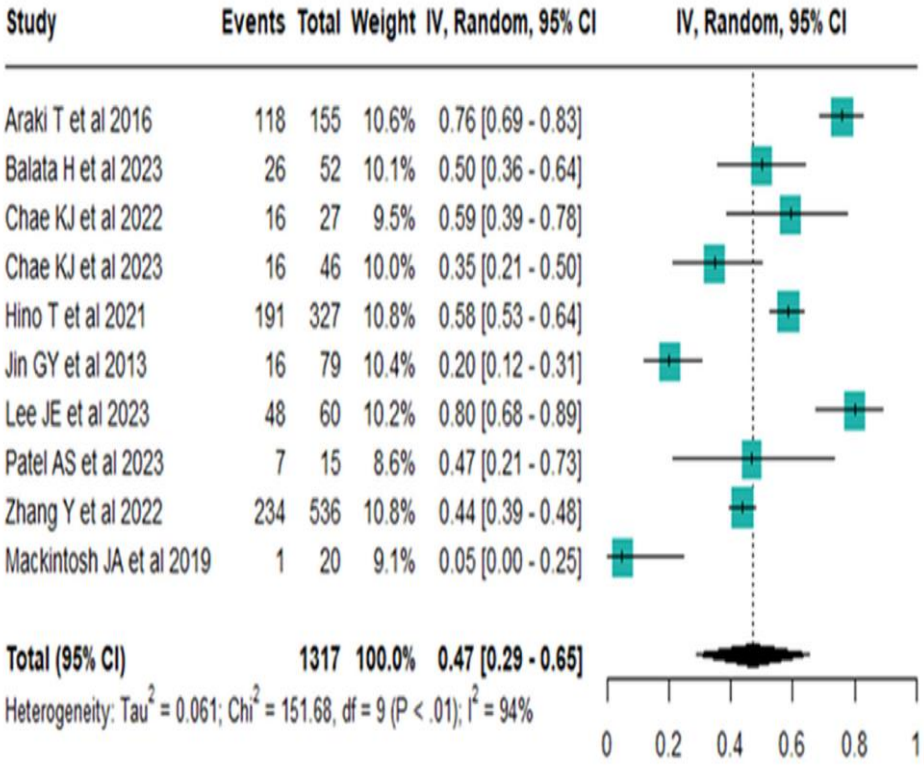
Prevalence and progression rate of interstitial lung abnormalities detected on thoracic CT: a systematic review and meta-analysis

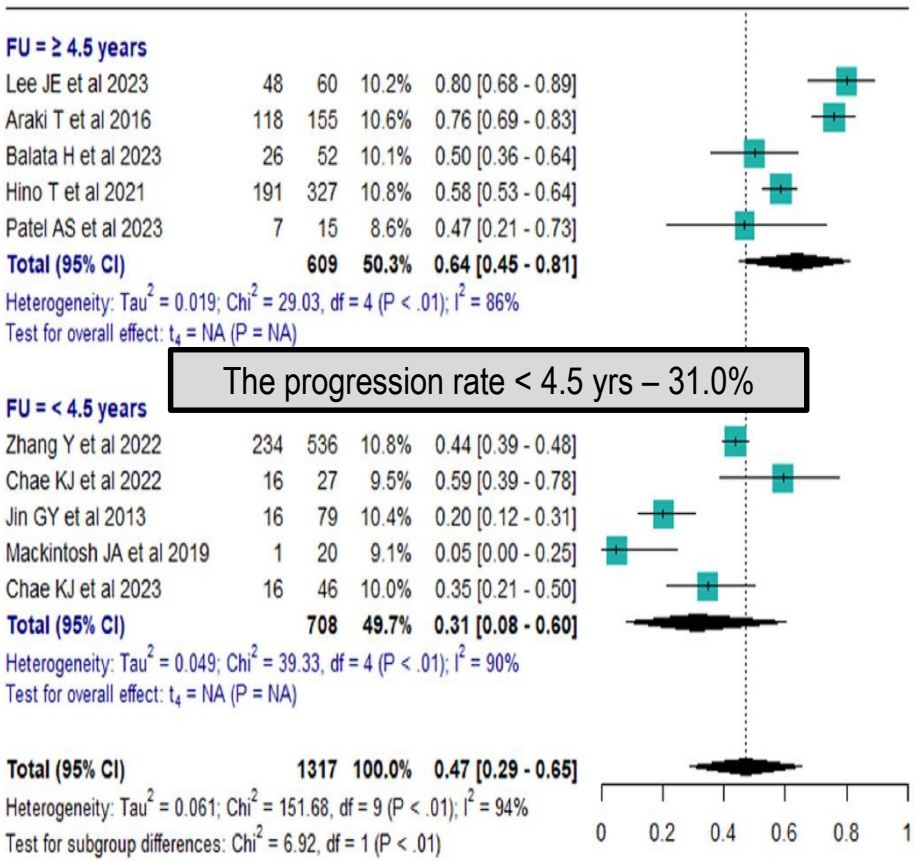
ILA progression – generally as an increase in extent or severity, new abnormalities, or evolution toward fibrotic features on follow-up CT

The pooled progression rate – 47.1%



The progression rate > 4.5 yrs – 64.2%

The progression rate < 4.5 yrs – 31.0%



European Radiology (2025) 35:276-288

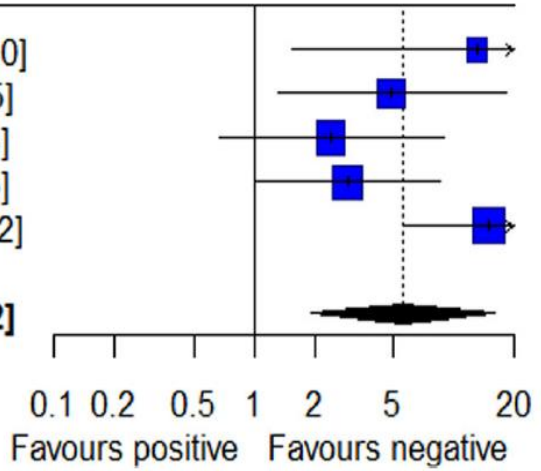
Pooled odds ratio of the progression between fibrotic and non-fibrotic types

Pooled odds ratio Fibrotic vs Non-Fibrotic: OR 5.55 (95%CI: 1.95-15.82)

Study	Positive Events	Positive Total	Negative Events	Negative Total	Weight	Odds Ratio MH, Random, 95% CI
Chae KJ et al 2023	15	31	1	15	10.2%	13.12 [1.53 - 112.40]
Jin GY et al 2013	7	19	5	47	19.7%	4.90 [1.32 - 18.25]
Lee JE et al 2023	34	40	14	20	20.1%	2.43 [0.67 - 8.84]
Zhang Y et al 2022	11	16	223	520	24.2%	2.93 [1.00 - 8.55]
Hino T et al 2021	51	56	47	116	25.9%	14.97 [5.56 - 40.32]
Total (95% CI)		**162**		**718**	**100.0%**	**5.55 [1.95 - 15.82]**

Heterogeneity: $Tau^2 = 0.363$; $Chi^2 = 7.44$, df = 4 (P = .11); $I^2 = 46\%$

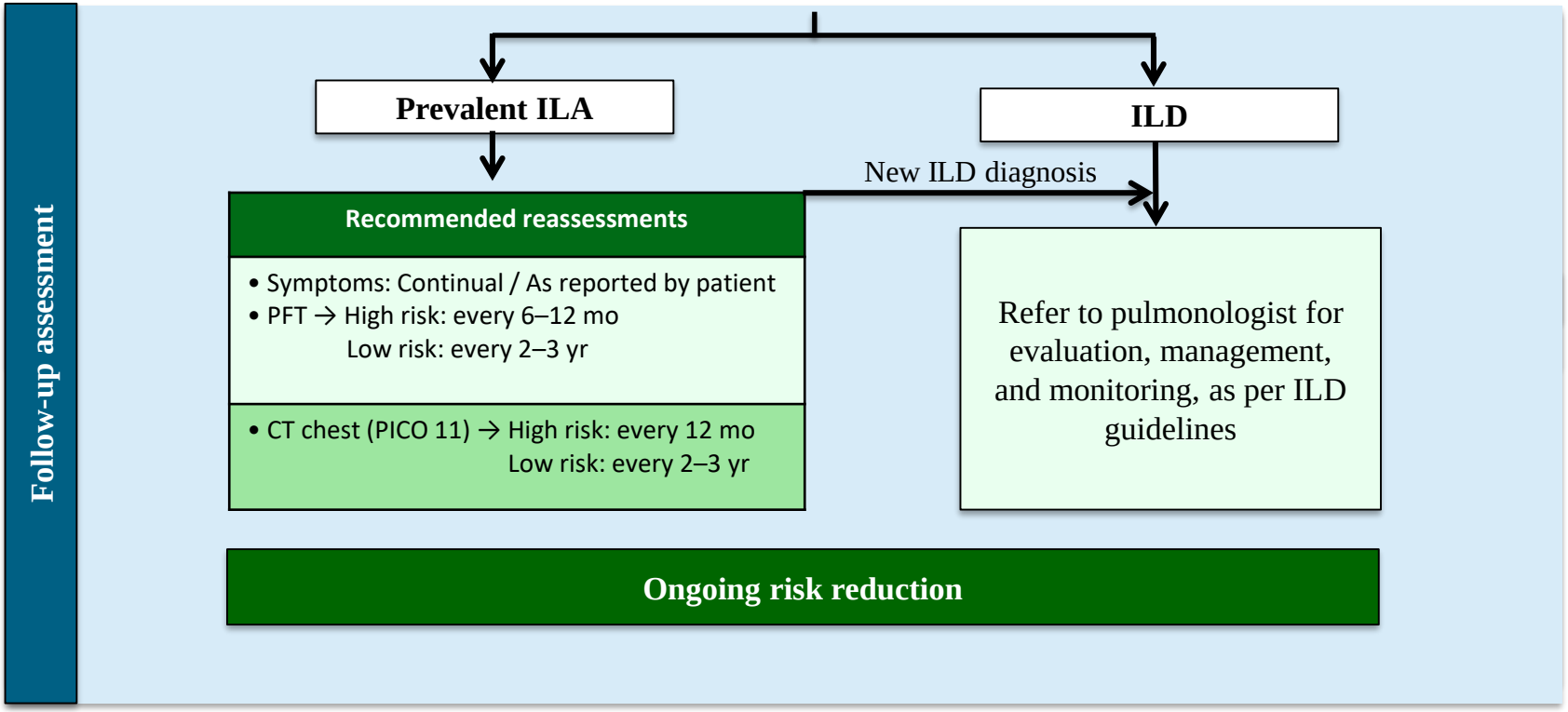
Test for overall effect: $t_4 = 4.54$ (P = .01)



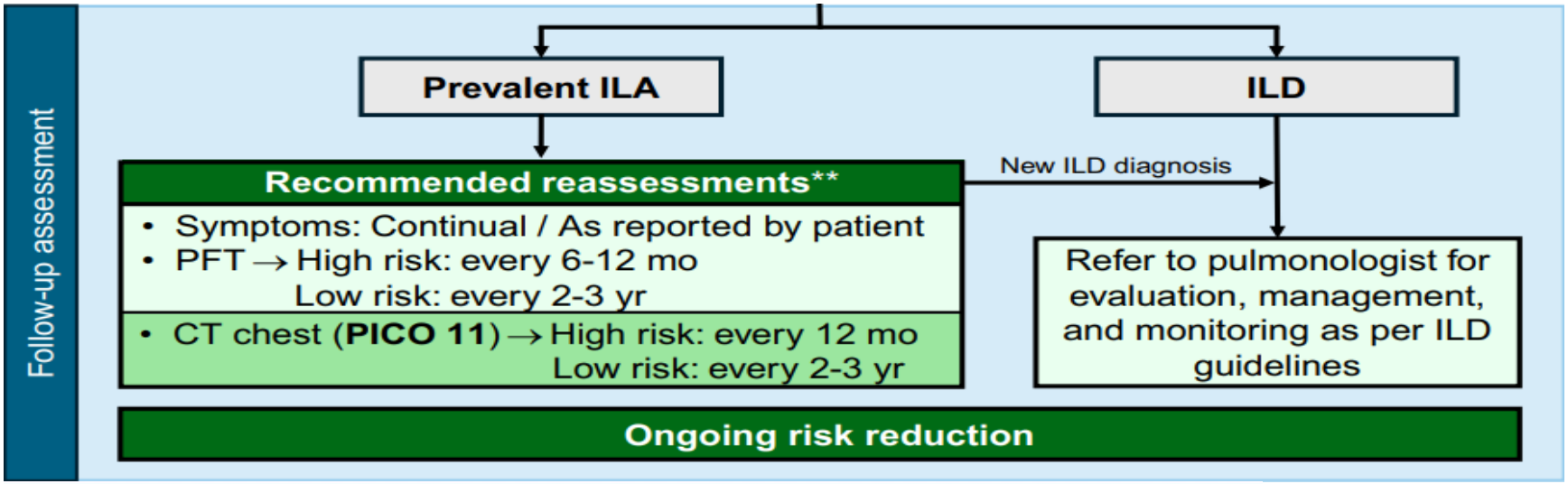
❖ Suggestions

- ILA is not a benign incidental finding – Nearly half progress, and fibrotic ILA (OR 5.55) requires active long-term monitoring
- Risk stratification in ILA – Fibrotic vs Non-fibrotic features
- At least 5 years of follow-up
- Follow-up every 1-2 years

European Radiology (2025) 35:276-288


Follow-up assessment
Prevalent ILA
ILD
New ILD diagnosis
Recommended reassessments
• Symptoms: Continual / As reported by patient
• PFT → High risk: every 6–12 mo
Low risk: every 2–3 yr
• CT chest (PICO 11) → High risk: every 12 mo
Low risk: every 2–3 yr
Refer to pulmonologist for evaluation, management, and monitoring, as per ILD guidelines
Ongoing risk reduction

Management of of ILA



High-risk ILA features

Demographic and clinical factors

- Family history of pulmonary fibrosis
- Older age
- Smoking history
- Other inhaled exposures (e.g., occupational vapors, gases, dusts, and fumes; air pollution)
- Connective tissue disease

Genetic

- MUC5B promoter variant
- Leukocyte telomere length below age-adjusted 10th percentile

Imaging

- Definite fibrosis on CT (e.g., honeycombing, traction bronchiectasis, distortion)
- Subpleural fibrotic and nonfibrotic subtypes
- Greater extent of abnormalities (multiple lung zones)

Physiologic

- Abnormal or borderline FVC (TLC) and DLco

Low risk

- Incidental ILA
- Non-subpleural ILA
- Stable ILA
- No symptoms
- Normal PFT

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CASE

▶ F / 83

▶ C.C : Abnormal chest CT

▶ PI : 상환 2023년 12월 검진으로 시행한 CT 이상 (r/o ILA)으로 호흡기내과 내원하였다.
심부전으로 타과 진료 중이며, 기침 등 호흡기 증상은 없고, never smoker 였다.
2013년 chest CT 검사를 했으나, 이상은 없었다고 한다.

▶ Arthritis (-) Raynaud (-) sicca symptom (-)

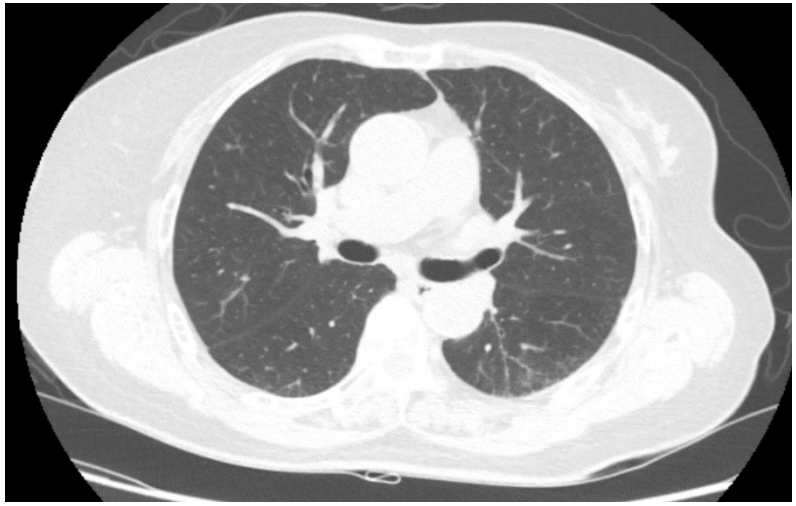
▶ Never-smoker

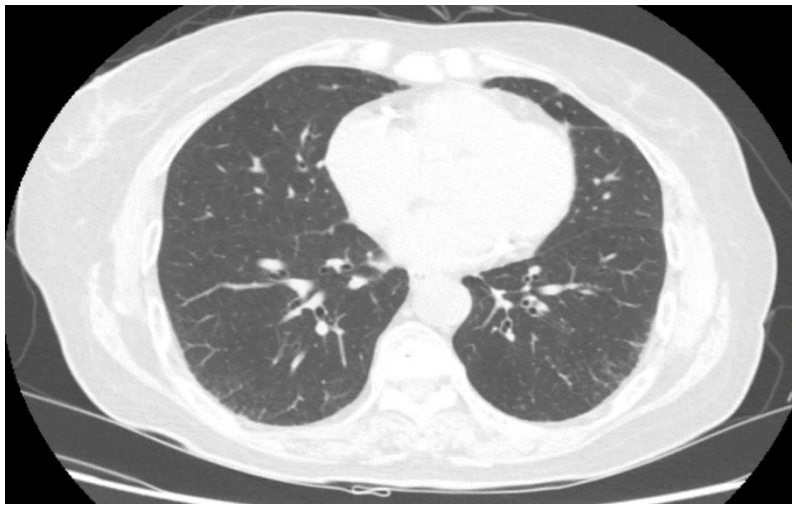
▶ Family-history(-)

▶ CAOD(2005), DM(2007), Rectal cancer(2013), GERD, Osteoporosis(2023)

Case

2013.04





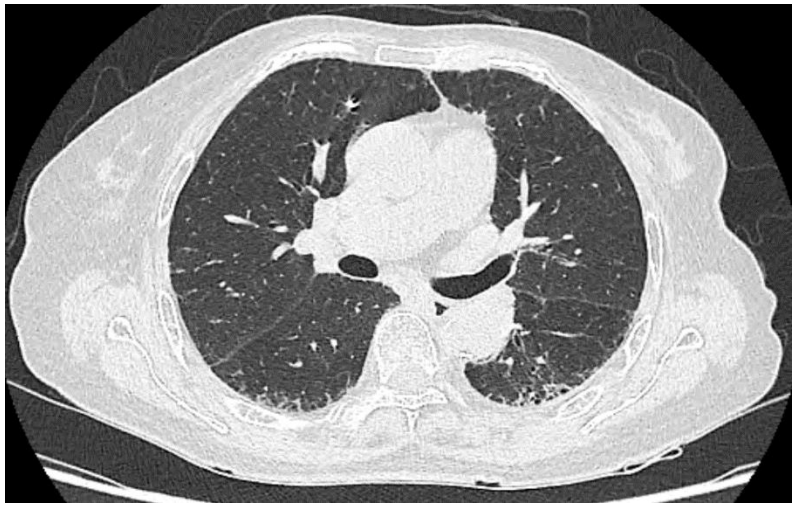
- Symptoms (-)
- CTD(-) Family history(-) Never smoker
- CT – Subpleural non-fibrotic ILA
- **<u>Low risk</u>**

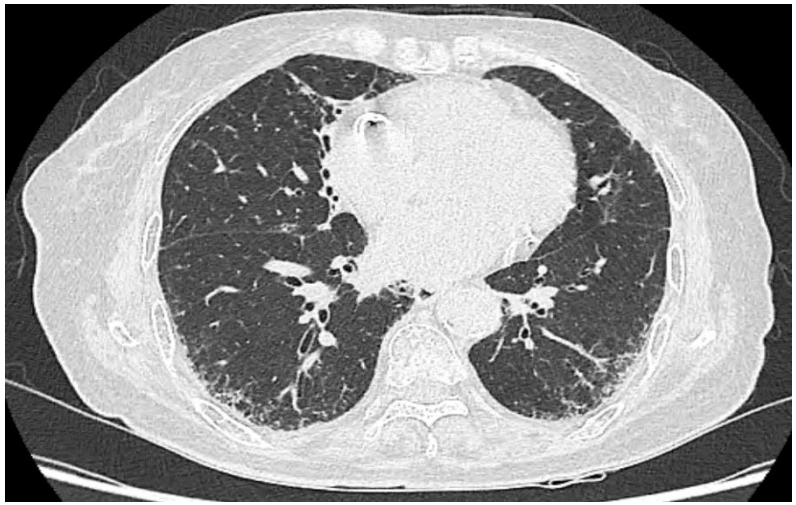
→ CT f/u after 3 years

→ PFT f/u

Case

2023.12





- CTD(-) Family history(-) Never smoker
- CT – **Subpleural fibrotic ILA**

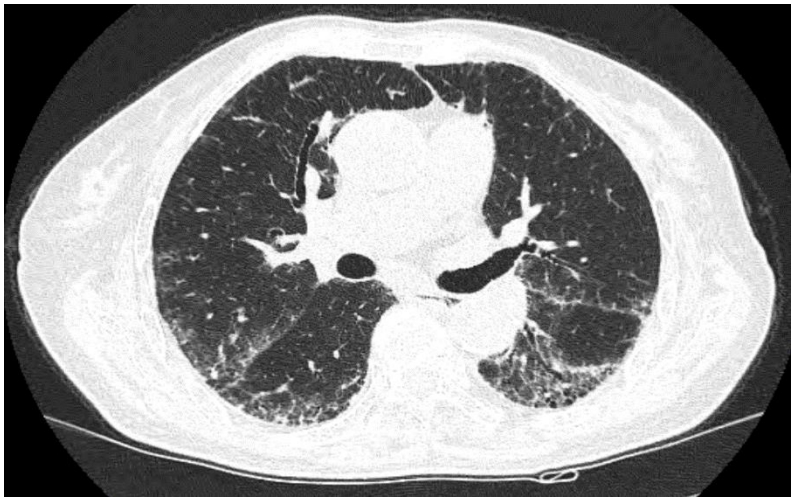
  Progression of ILA (10 years)
- **High risk**
- Symptom (-), HF로 진료 중
- PFT – ratio 83%, FVC 93%, DLco 85%

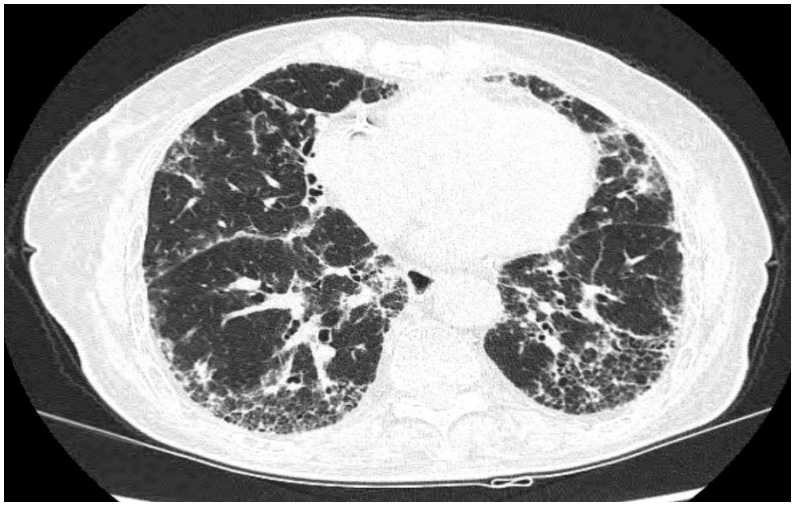
→ CT f/u after 1year

→ PFT after 6 month

Case

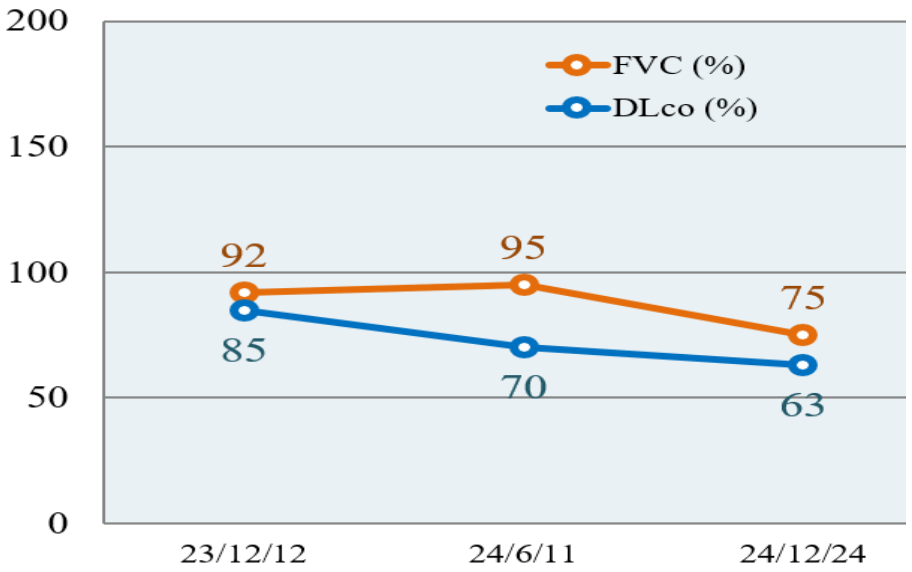
2024.12



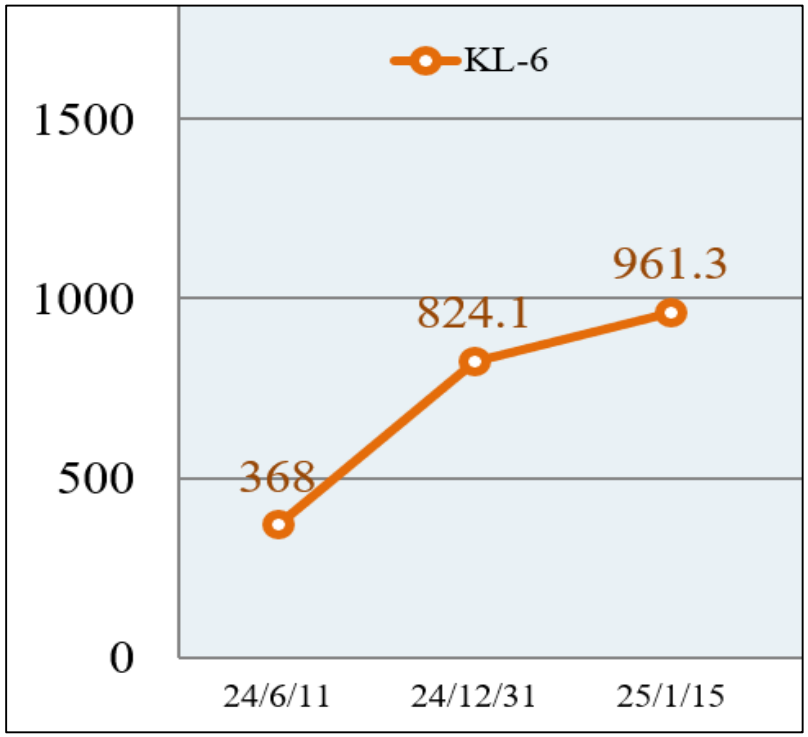


- CTD(-) Family history(-) Never smoker
- CT – **<u>Subpleural fibrotic ILA → NSIP & progression: ILD</u>**
- **<u>High risk</u>**
- Symptom (+) → exertional dyspnea

→ Screening for Auto-Ab & KL-6



Case



ANA	Positive : nuclear envelope(1:1280) & Cytoplasmic Ab
ANCA-MPO(Qn)	Positive(123)
ANCA-PR3(Qn)	Negative(0.68)
ANCA-MPO(QI)	Positive
ANCA-PR3(QI)	Negative
Rheumatoid Factor	▲ 21.1
SS-A(Ro) Ab	Negative(0.66)

- **MDD** – Progressive fibrosing ILD & IPAF
- **Steroid + MMF → add-on antifibrotics**

Case

▶ M / 67

▶ C.C : Abnormal chest CT (r/o ILA)

▶ PI : 상환 건강검진으로 시행한 CT에서 이상 소견으로 내원하였다. 현재 호흡기 증상은 없으며, 30갑년의 흡연력이 있고 2013년 부터 금연 했다고 한다. 폐질환 가족력은 없다고 한다.

▶ Arthritis (-) Raynaud (-) sicca symptom (-)

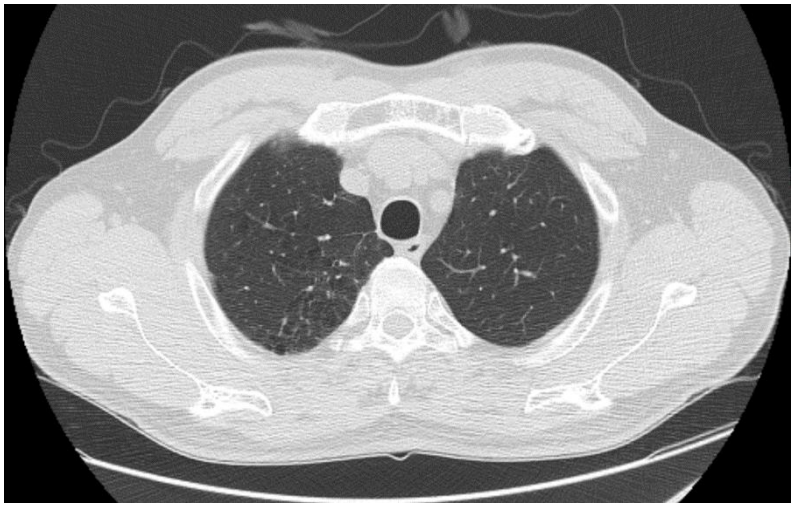
▶ Ex.– smoker 30pyrs (2013년 중단)

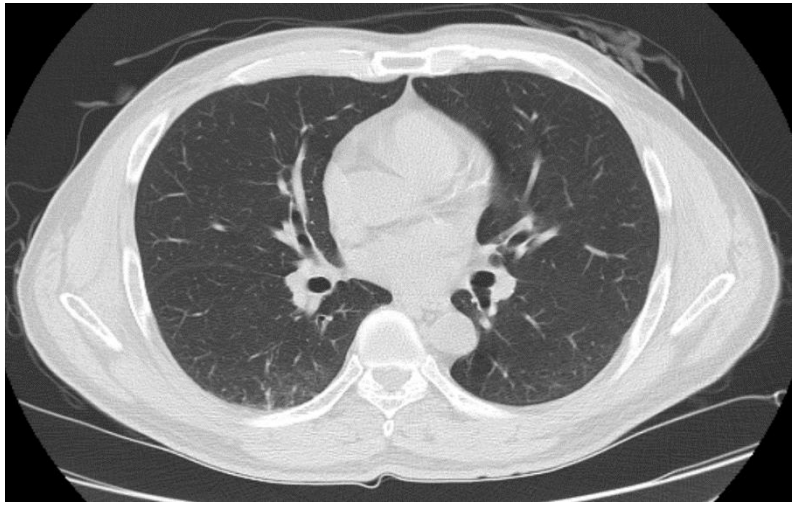
▶ 직업력 : 호텔 (린넨 관련)

▶ Angina(2023), HTN,Hyperlipidemia(2020), DM(2023)

Case

2013.01





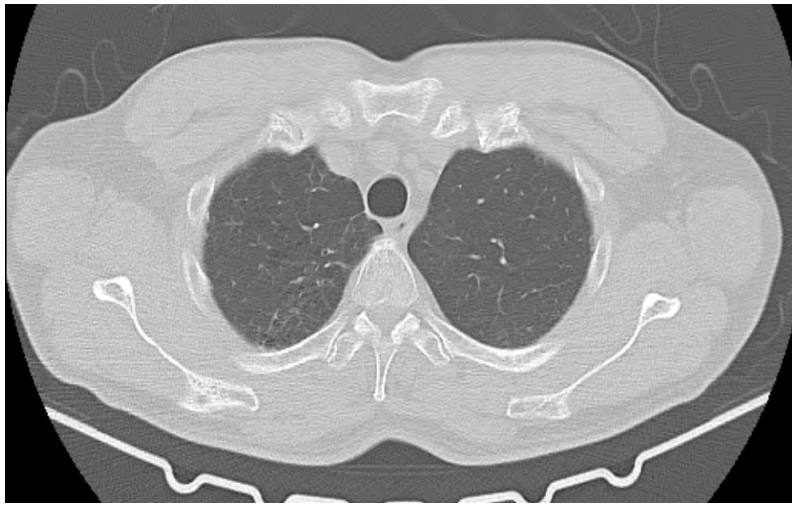
- Symptoms (-)
- CTD(-) Family history(-) **Ex-smoker**
- CT – Subpleural non-fibrotic ILA
- **High risk** – **Lung cancer screening**

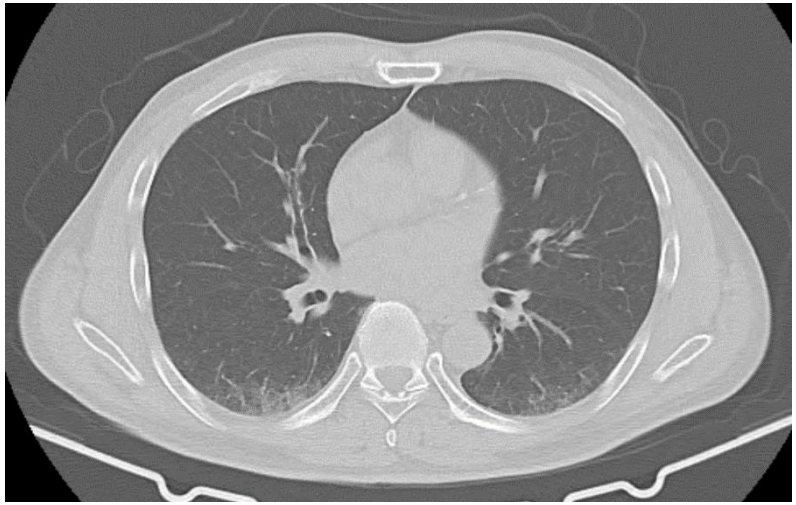
→ CT f/u every year

→ PFT f/u 6-12 months

Case

2025.03





- CTD(-) Family history(-) **Ex-smoker**
- CT – Subpleural non-fibrotic ILA

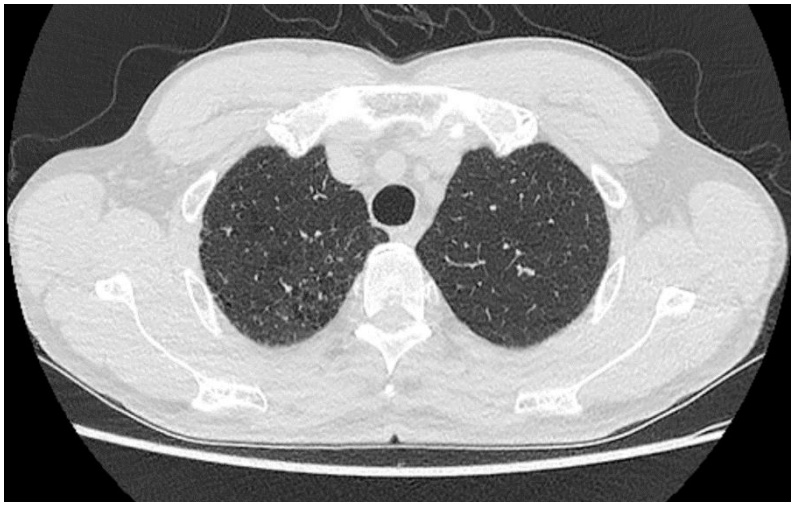
  Progression of ILA (12 years)
- **High risk – Lung cancer screening**
- Symptom (-)
- PFT – ratio 78%, FVC 96%, DLco 103%

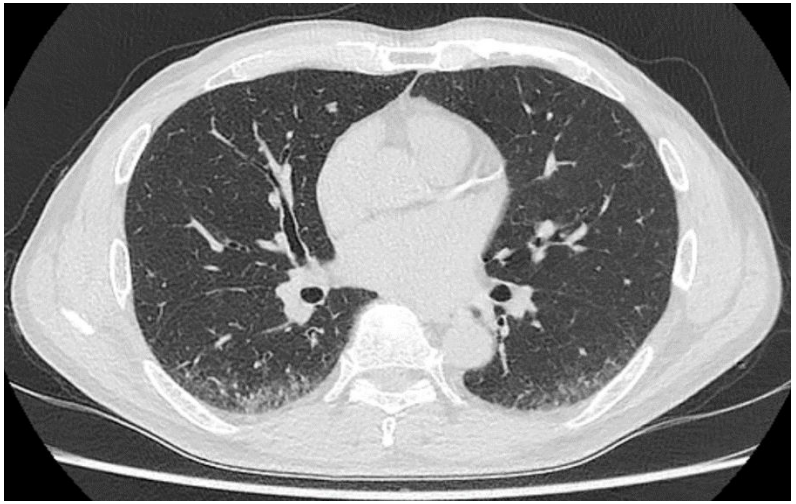
→ CT f/u after 1year

→ PFT after 6 month

Case

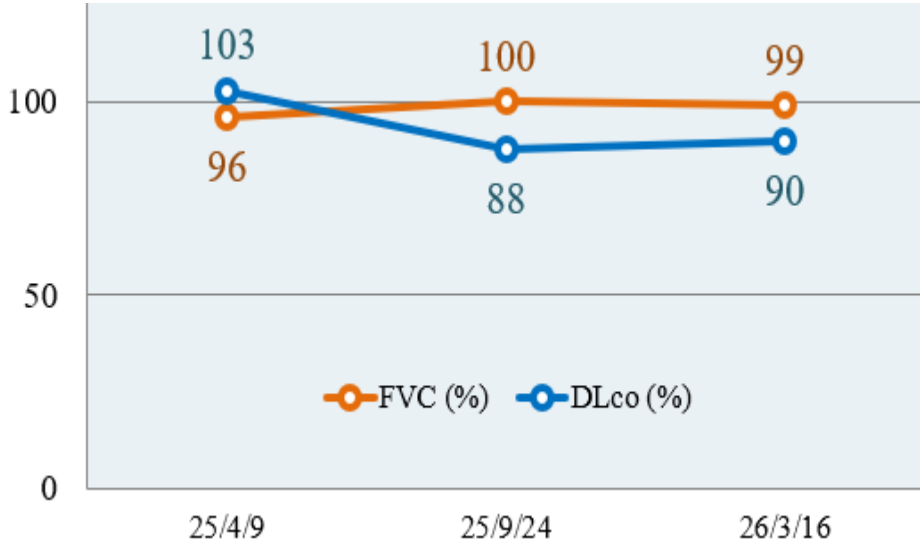
2026.03



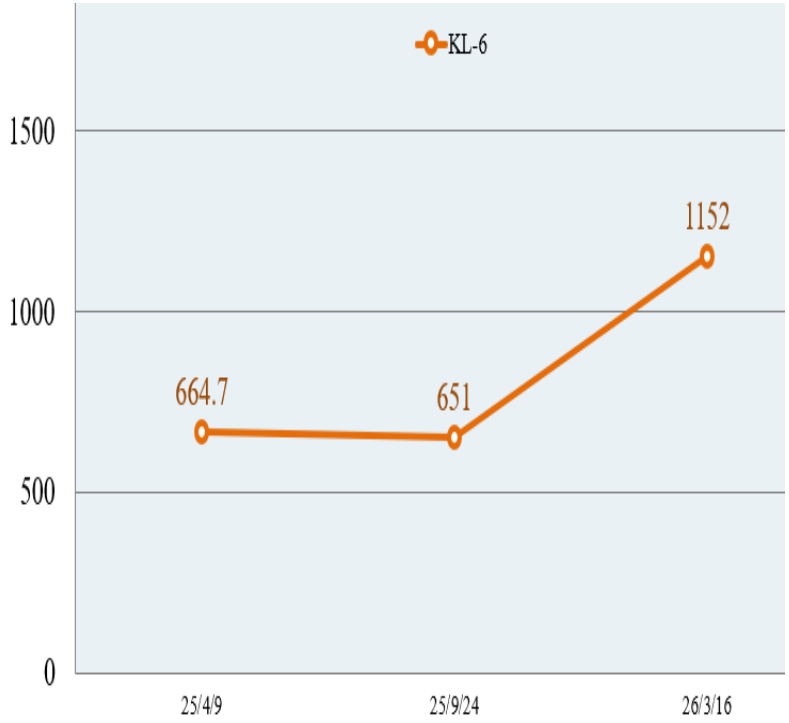


- CTD(-) Family history(-) Never smoker
- CT – **Subpleural non-fibrotic ILA & Progression**
- **High risk – Lung cancer & Progression**
- Symptom (-) exertional dyspnea (-)

→ Screening for Auto-Ab & KL-6



Case

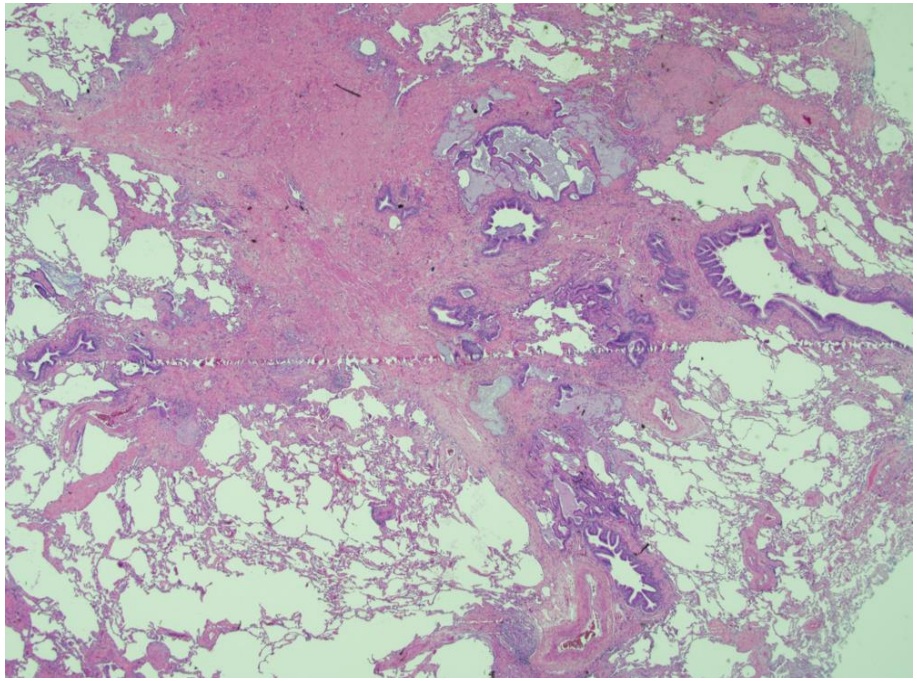


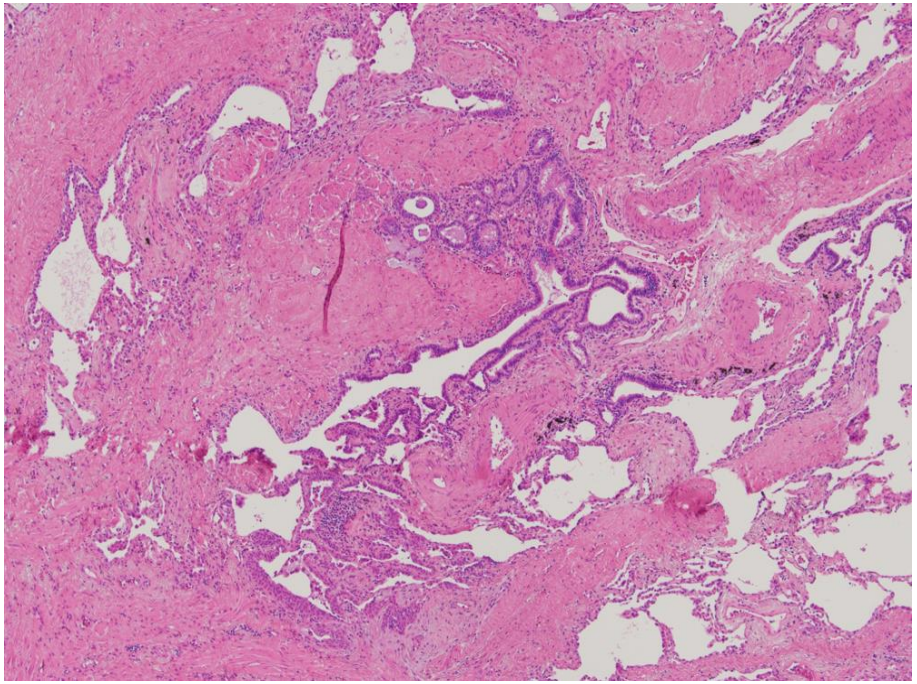
ANA	Positive : dense-fine speckled (1:40)
ANCA-MPO(Qn)	Negative(0.28)
ANCA-PR3(Qn)	Negative(0.62)
ANCA-MPO(QI)	Negative
ANCA-PR3(QI)	Negative
Rheumatoid Factor	< 10
SS-A(Ro) Ab	Negative(0.82)

- **MDD** – Radiology (ILA than ILD)
  Rheumatology – CTD(-)
- Short term-follow up vs VATS

Case

RLL wedge resection





Diffuse fibrosing interstitial pneumonia showing temporal and spatial heterogeneity

1) Scarring and ossifications
2) Positive for fibroblastic foci
3) Frequent peribronchiolar metaplasia
4) No autoimmune features and no granulomas

Summary

Interstitial lung abnormalities

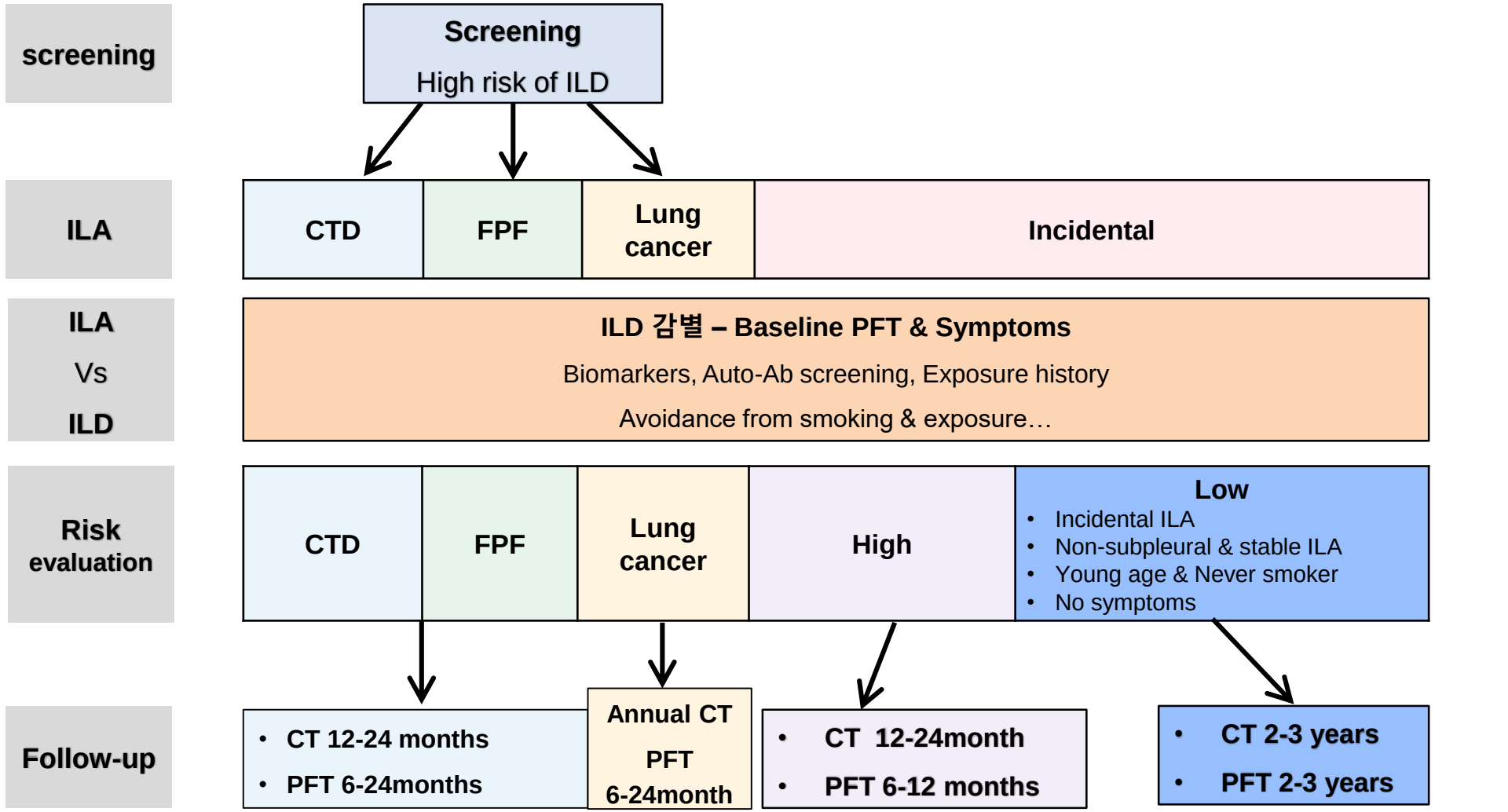
CT findings of the following, which are bilateral, nondependent and involve ≥5% of a lung zone

- Ground-glass opacities
- Reticular abnormalities
- Lung distortion
- Traction bronchiectasis
- Honeycombing

Subpleural distribution	Non-subpleural distribution
Likely to progress	*Unlikely to progress*

- Early detection before PFT abnormality and Symptoms development
- High risk for ILD development → Screening before pre- or sub-clinical ILD
- ILA are associated
  1) Increased respiratory symptoms
  2) Accelerated loss of lung function
  3) Radiologic progression
  4) Increased complications of surgery and drug/radiation induced ILD
  5) Increased all-cause mortality

Summary


screening
Screening
High risk of ILD
ILA
CTD
FPF
Lung cancer
Incidental
ILA
Vs
ILD
ILD 감별 – Baseline PFT & Symptoms
Biomarkers, Auto-Ab screening, Exposure history
Avoidance from smoking & exposure…
Risk evaluation
CTD
FPF
Lung cancer
High
Low
• Incidental ILA
• Non-subpleural & stable ILA
• Young age & Never smoker
• No symptoms
Follow-up
• CT 12-24 months
• PFT 6-24months
Annual CT
PFT
6-24month
• CT 12-24month
• PFT 6-12 months
• CT 2-3 years
• PFT 2-3 years

감사합니다