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Approach to ILD: IPF and PPF

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American T

American Tho
Society Intern
Classification

THIS JOINT STATEMENT OF THE
ADOPTED BY THE ATS BOARD

DPLD of known
cause e.g. drugs
or association e.g.
collagen vascular dise

American Thoraci

An Official American
Respiratory Society S
International Multidi
Idiopathic Interstitia

William D. Travis, Ulrich Costabel, David M
Christopher J. Ryerson, Jay H. Ryu, Moisés
Kevin K. Brown, Thomas V. Colby, Harold
Marjolein Drent, Rosalind F. Dudden, Jim E
Dong Soon Kim, Masanori Kitaichi, James L
Ganesh Raghu, Luca Richeldi, Nicola Sverze
Committee on Idiopathic Interstitial Pneumo

THIS OFFICIAL STATEMENT OF THE AMERICAN T
APPROVED BY THE ATS BOARD OF DIRECTORS, JU

TABLE 2. CATEGORIZATION OF MAJ

Category	Clinical–Ra
Chronic fibrosing IP	Idiopathic pul
	Idiopathic non
Smoking-related IP*	Respiratory br
	Desquamative
Acute/subacute IP	Cryptogenic o
	Acute interstiti

Definition of abbreviation: IP = interstitial pr
*Desquamative interstitial pneumonia can

Update of the international multidisciplinary classification of the interstitial pneumonias: an ERS/ATS statement

Christopher J. Ryerson [1,42], Ayodeji Adegunsoye [2,42], Sara Piciucchi [3,4,42], Lida P. Hariri [5], Yet H. Khor [6,7,8,9], Marlies S. Wijsenbeek[10], Athol U. Wells[11], Amita Sharma [12], Wendy A. Cooper [13], Katerina Antoniou[14], Raphael Borie [15], Aurelie Fabre [16,17], Yoshikazu Inoue [18,19], Kerri Johannson [20], Takeshi Johkoh[21], Leticia Kawano-Dourado [22,23,24], Ella Kazerooni [25], Toby M. Maher [26,27], Philip L. Molyneaux [27,28], Raymond Protti[29], Claudia Ravaglia[3,30], Elisabetta A. Renzoni[27,31], Ryoko Saito-Koyama[32], Nicola Sverzellati[33,34], Simon L.F. Walsh[35,36,37], Paul Wolters [38], Soo-Ryum Yang[39], William Travis[40,42] and Andrew G. Nicholson [27,41,42]

[1]Department of Medicine and Centre for Heart Lung Innovation, University of British Columbia, Vancouver, BC, Canada. [2]Department of Medicine, Section of Pulmonary and Critical Care, University of Chicago, Chicago, IL, USA. [3]Department of Medical and Surgical Sciences (DIMEC), University of Bologna, Bologna, Italy. [4]Department of Radiology, G.B. Morgagni-Pierantoni Hospital, Forlì, Italy. [5]Department of Pathology and Division of Pulmonary and Critical Care Medicine, Massachusetts General Hospital, Harvard Medical School, Boston, MA, USA. [6]Respiratory Research@Alfred, School of Translational Medicine, Monash University, Melbourne, Australia. [7]Department of Respiratory and Sleep Medicine, Austin Health, Heidelberg, Australia. [8]Institute for Breathing and Sleep, Heidelberg, Australia. [9]Faculty of Medicine, University of Melbourne, Melbourne, Australia. [10]Centre of Expertise for Interstitial Lung Diseases and Sarcoidosis, Department of Respiratory Medicine, Erasmus MC, Rotterdam, The Netherlands. [11]Interstitial Lung Disease Unit, Royal Brompton Hospital and Imperial College, London, UK. [12]Department of Radiology, Massachusetts General Hospital, Harvard Medical School, Boston, MA, USA. [13]Tissue Pathology and Diagnostic Oncology, NSW Health Pathology, Royal Prince Alfred Hospital, Camperdown, Australia. [14]Dept of Respiratory Medicine, Laboratory of Molecular and Cellular Pneumonology, School of Medicine, University of Crete, Heraklion, Greece. [15]Université Paris Cité, Inserm, PHERE, Hôpital Bichat, AP-HP, Service de Pneumologie A, Centre Constitutif du Centre de Référence des Maladies Pulmonaires Rares, FHU APOLLO, Paris, France. [16]Histopathology Dept, St Vincent's University Hospital, Dublin, Ireland. [17]University College Dublin School of Medicine, Dublin, Ireland. [18]Clinical Research Center, NHO Kinki Chuo Chest Medical Center, Osaka, Japan. [19]Department of Internal Medicine, Osaka Anti-tuberculosis Association, Osaka Fukujuji Hospital, Osaka, Japan. [20]Department of Medicine and Snyder Institute for Chronic Diseases, University of Calgary, Calgary, AB, Canada. [21]Department of Radiology, Kansai Rosai Hospital, Amagasaki, Japan. [22]Hcor Research Institute, Hcor Hospital, Sao Paulo, Brazil. [23]MAGIC Evidence Ecosystem Foundation, Oslo, Norway. [24]Pulmonary Division, Heart Institute (InCor), University of Sao Paulo, Sao Paulo, Brazil. [25]Departments of Radiology and Internal Medicine, University of Michigan, Ann Arbor, MI, USA. [26]Keck School of Medicine, University of Southern California, Los Angeles, CA, USA. [27]National Heart and Lung Institute, Imperial College London, London, UK. [28]Royal Brompton and Harefield Hospitals, Guy's and St Thomas' NHS Foundation Trust, London, UK. [29]Patient representative. [30]Pulmonology Unit, G.B. Morgagni Hospital, Forlì, Italy. [31]Interstitial Lung Disease Unit, Royal Brompton and Harefield Hospitals, Guy's and St Thomas' NHS Foundation Trust, London, UK. [32]Department of Pathology, National Hospital Organization, Sendai Medical Center, Miyagi, Japan. [33]Department of Medicine and Surgery, University of Parma, Parma, Italy. [34]Scienze Radiologiche, University Hospital of Parma, Parma, Italy. [35]Department of Radiology, Imperial College, London, UK. [36]Department of Radiology, Royal Brompton and Harefield Foundation Trust, London, UK. [37]Qureight Ltd, Cambridge, UK. [38]Department of Medicine, Division of Pulmonary, Critical Care, Allergy and Sleep Medicine, University of California, San Francisco, CA, USA. [39]Department of Pathology and Laboratory Medicine, Memorial Sloan Kettering Cancer Center, New York, NY, USA. [40]Dept of Pathology, Memorial Sloan Kettering Cancer Center, New York, NY, USA. [41]Department of Histopathology, Royal Brompton and Harefield Hospitals, Guy's and St Thomas' NHS Foundation Trust, London, UK. [42]Contributed equally.

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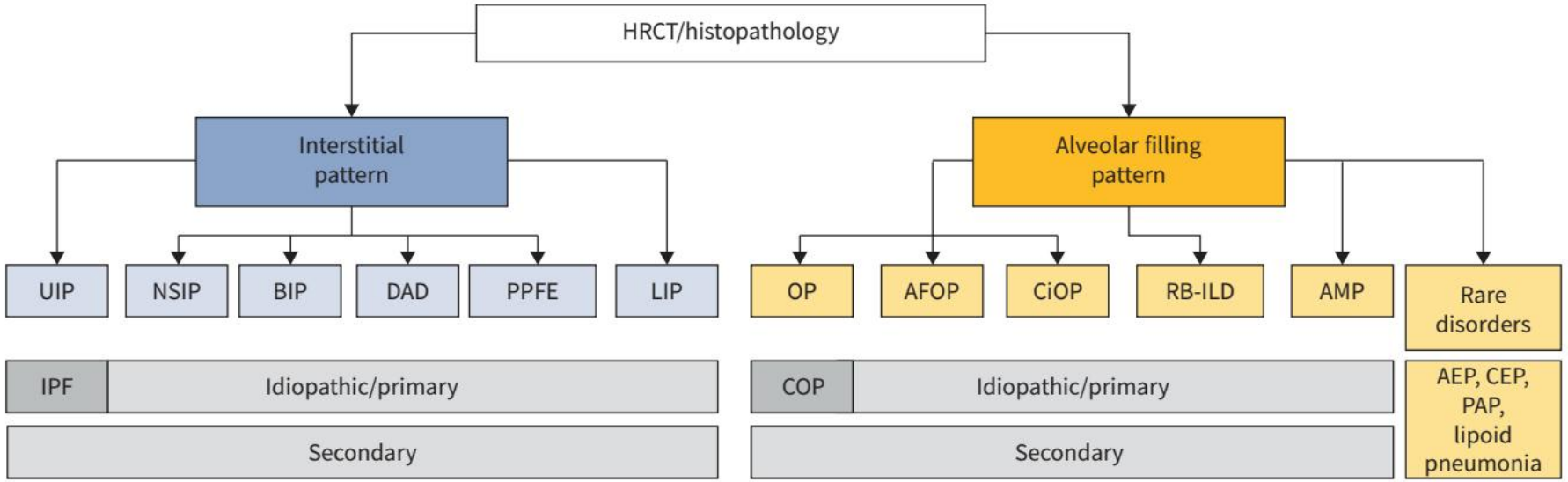
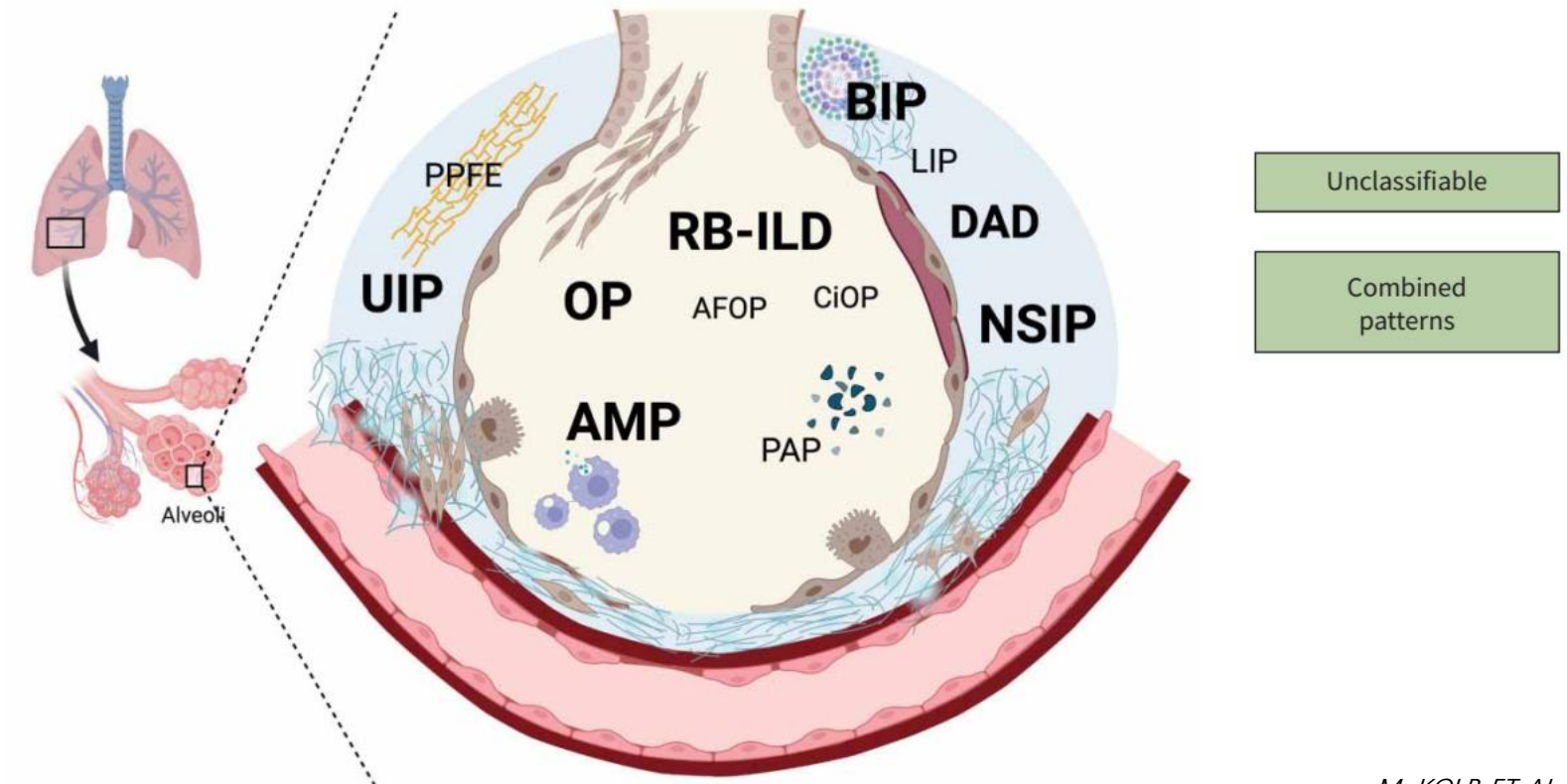
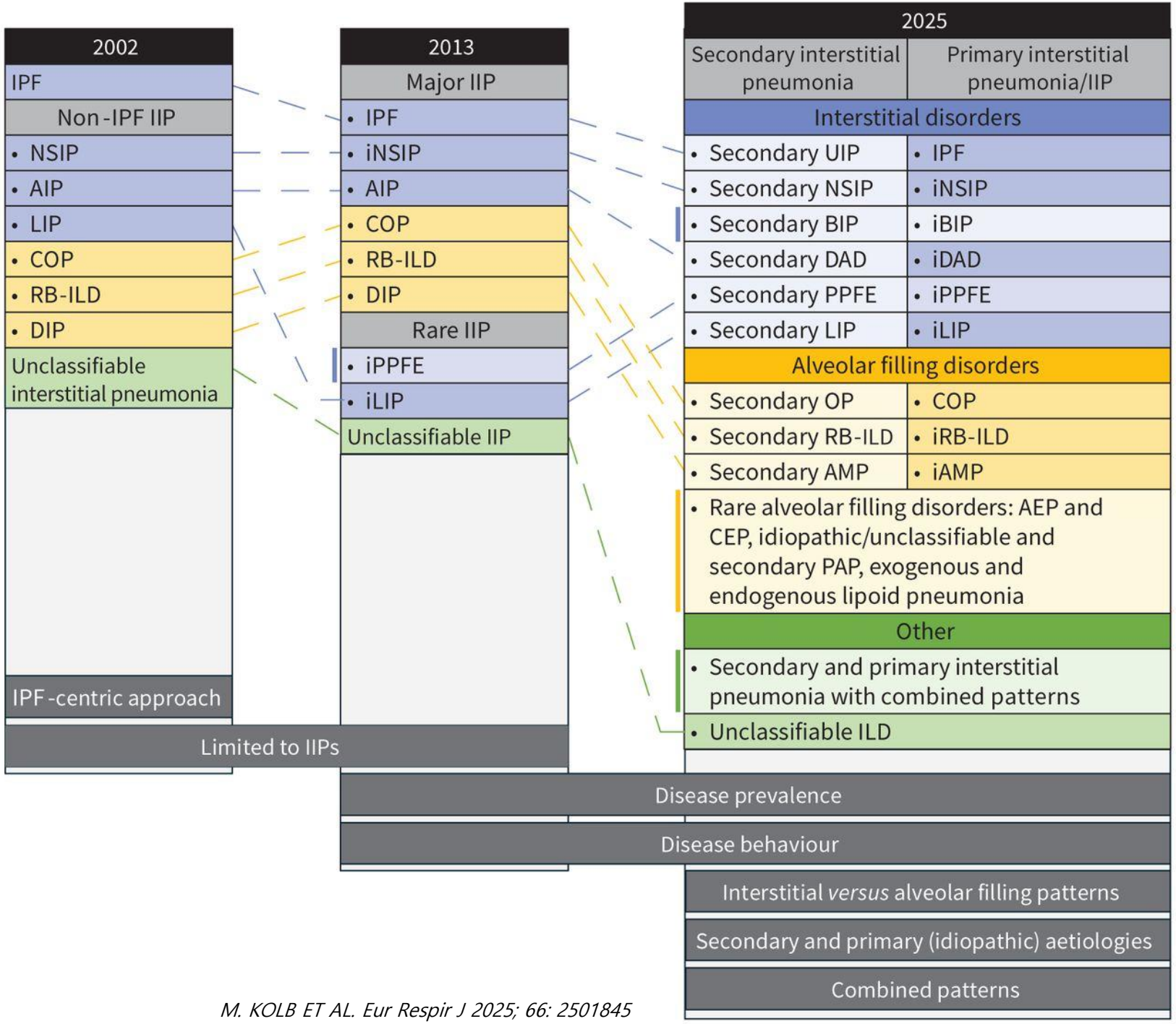
Eur Respir J 2025; 66: 2500158





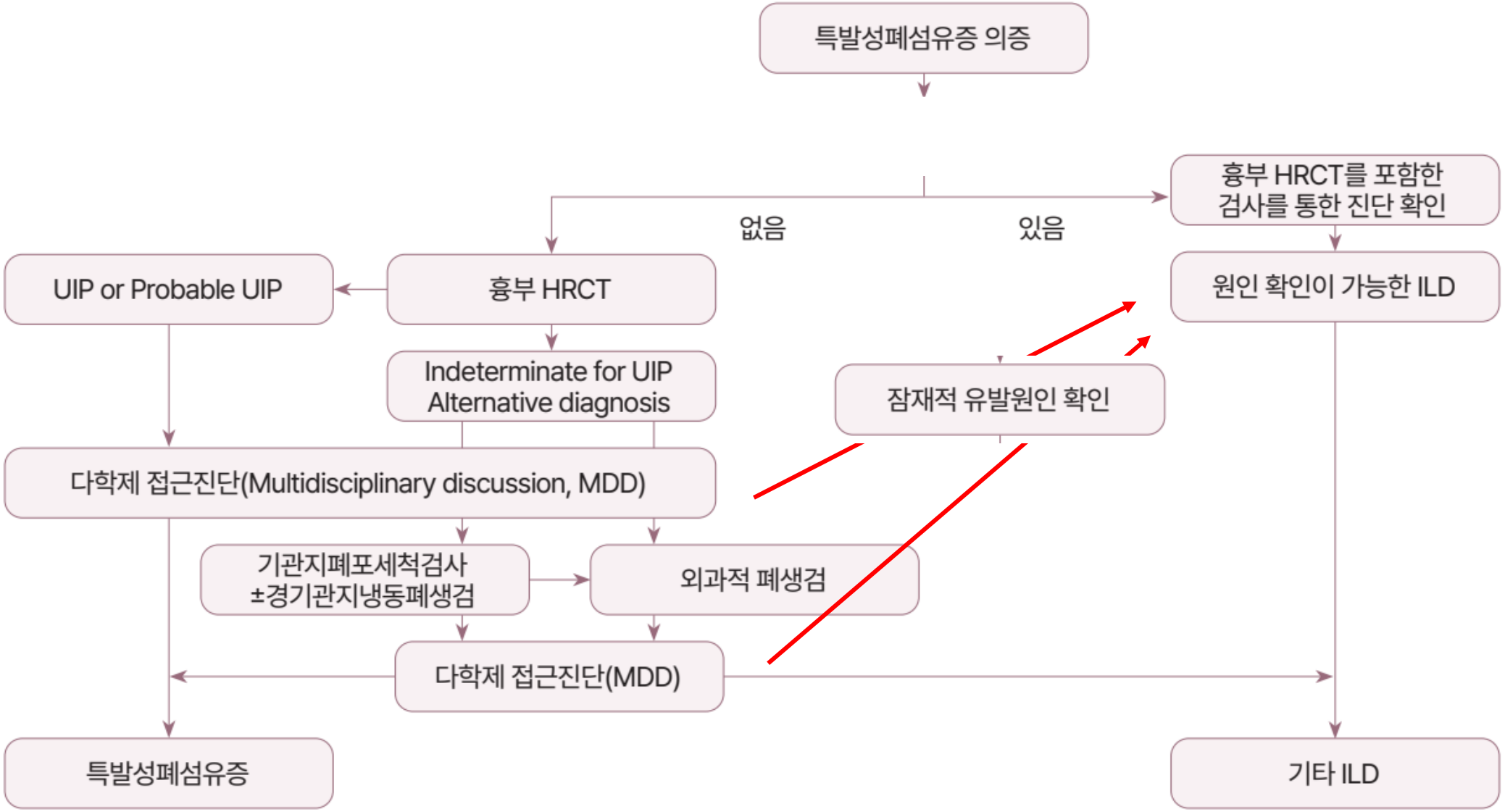
M. KOLB ET AL. Eur Respir J 2025; 66: 2501845

Expansion beyond IIPs



M. KOLB ET AL. Eur Respir J 2025; 66: 2501845

IPF 의 진단과정

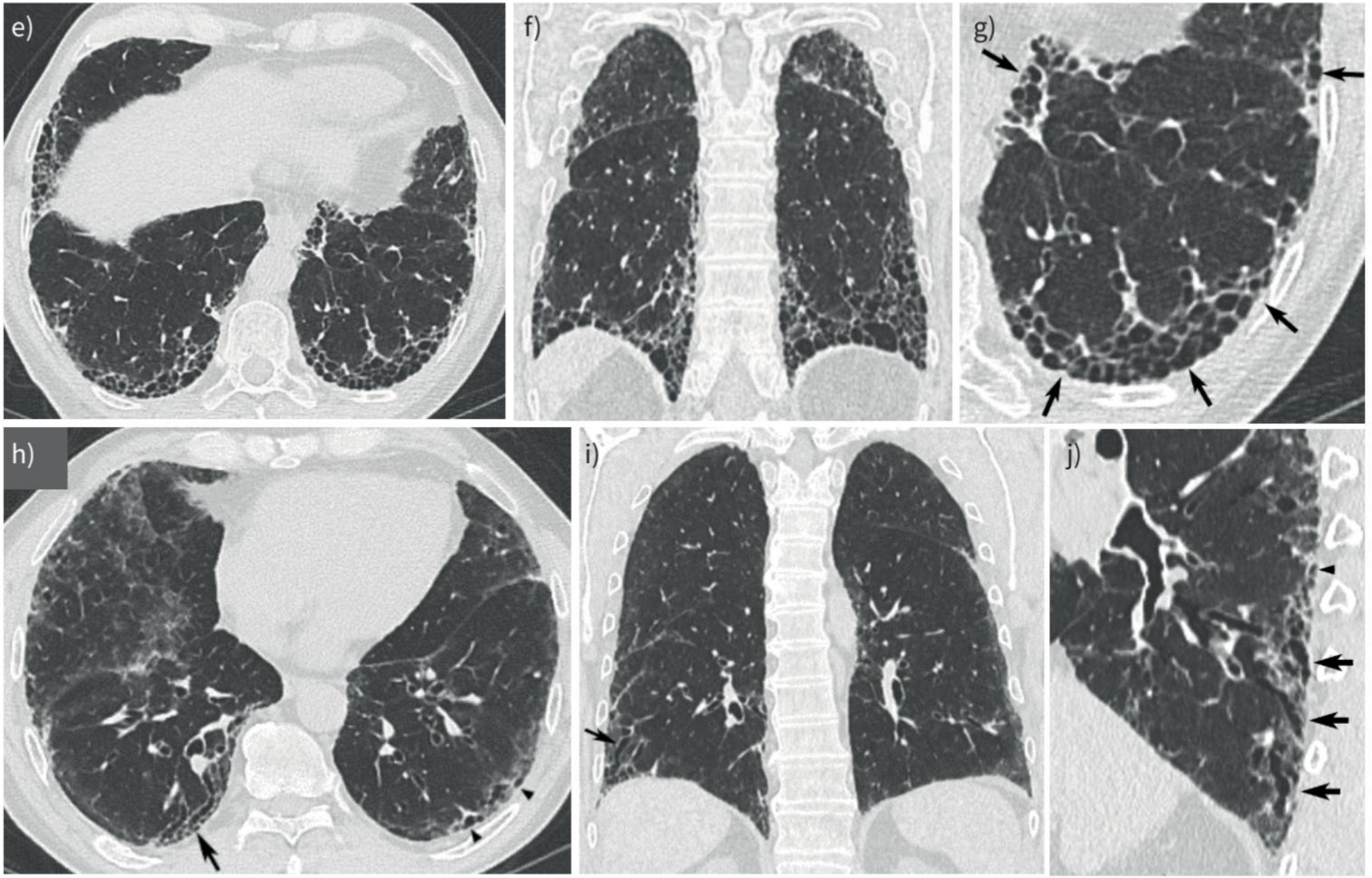


간질성폐질환 임상진료지침 (2023 1차 개정판

UIP 양상의 흉부 HRCT 소견

UIP pattern	Probable UIP	Indeterminate for UIP	Alternative diagnosis
- 흉막하부와 폐하부 우세성 분포 - 불균일한 소견이 자주 보임 - 때로는 미만성 분포가 보일 수 있음 - 비대칭적인 분포를 보일 수도 있음 - 견인성 기관지확장증 또는 세기관지확장증이 동반되거나 동반되지 않은 벌집모양 폐 - 소엽사이막의 불규칙적인 비대가 관찰됨 - 대개 망상 음영, 경도의 간유리 음영이 동반됨 - 폐골화증이 관찰될 수 있음	- 흉막하부와 폐하부 우세성 분포 - 불균일한 소견이 자주 보임 - 견인성 기관지확장증 또는 세기관지확장증이 동반된 망상 음영 - 경도의 간유리 음영이 관찰될 수 있음 - 흉막하부 병변의 존재	- 흉막하의 우세성 침범 없이 미만성 분포를 보이는 경우 - 어떠한 특정 원인을 시사하지 않는 폐섬유화의 CT 소견	- 흉막하부가 보존된 기관지혈관주위 우세성 분포를 보이는 경우(NSIP) - 림프 근처의 분포를 보이는 경우(Sarcoidosis) - 폐상부 또는 중부 우세성 분포(fibrotic HP, CTD-ILD, Sarcoidosis 고려) - 흉막하부 보존(NSIP, smoking-related IP) - 폐낭종(LAM, PLCH, LIP, DIP) - 모자이크 양상의 폐음영 또는 삼상 폐밀도 경향을 보이는 경우(HP) - 망상 음영이 우세한 분포를 보이는 경우(HP, Smoking related disease, drug toxicity, acute exacerbation of fibrosis) - 중심소엽성 결절이 과도하게 관찰되는 경우(HP, Smoking related disease) - 폐결절(Sarcoidosis) - 폐경화(Organizing pneumonia 등) - 흉막석회(asbestosis) - 확장된 식도(CTD)

간질성폐질환 임상진료지침 (2023 1차 개정판



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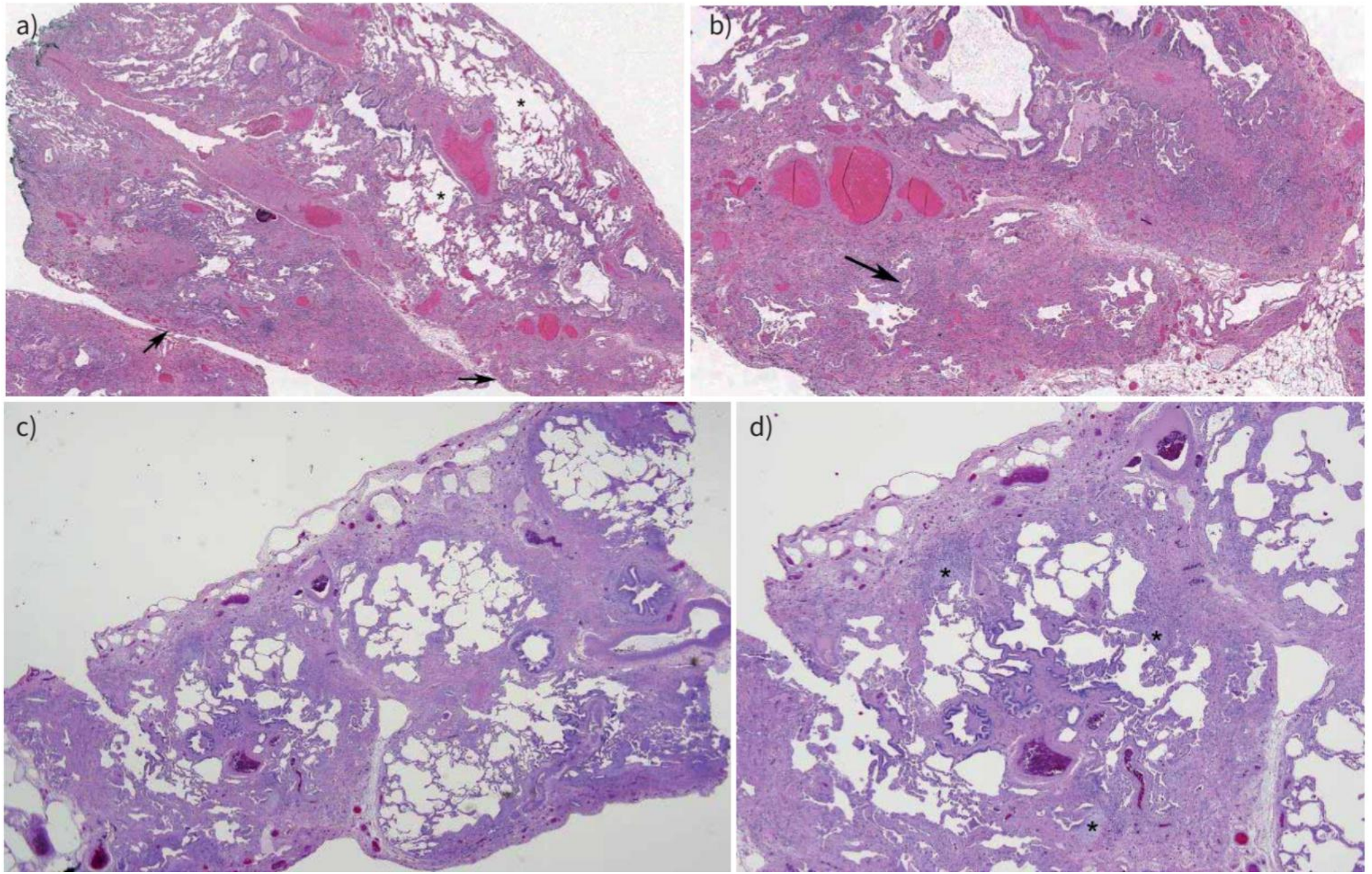
UIP 양상의 병리소견

UIP pattern	Probable UIP	Indeterminate for UIP	Alternative diagnosis
- 산재된 치밀 섬유화와 구조적 왜곡 - 흉막하 및 중격 주위 분포의 우세성 - 섬유화가 반점상으로 분포된 폐실질 - 섬유모세포 병소 - 다른 진단을 시사하는 소견의 부재	- 벌집모양폐만 관찰되는 경우 또는 - UIP의 특성중 몇 가지를 가지고 있으나 UIP/IPF 의 명확한 진단이 제한적인 경우 그리고 - 다른 진단을 시사할 수 있는 특징이 없는 경우	- 구조적 왜곡을 동반하거나 동반하지 않는 섬유화가 있으며, UIP를 시사하는 소견보다는 UIP가 아닌 다른 패턴을 시사하는 소견이 동반된 경우 - UIP의 일부 조직학적 특징이 있지만, 다른 진단을 시사하는 소견이 동반된 경우	- 모든 생검 결과가 다른 IIP의 조직학적 패턴을 보이는 경우 - HP, PLCH, Sarcoidosis, LAM 등의 다른 진단을 시사하는 소견이 보이는 경우

간질성폐질환 임상진료지침 (2023 1차 개정판



Eur Respir J 2025; 66: 2500158

HRCT 와 조직형의 조합에 의한 IPF의 진단

IPF suspected		Histopathology pattern			
		UIP	Probable UIP	Indeterminate for UIP or biopsy not performed	Alternative diagnosis
HRCT pattern	UIP	IPF	IPF	IPF	Non-IPF Dx
	Probable UIP	IPF	IPF	IPF (Likely)[a]	Non-IPF Dx
	Indeterminate	IPF	IPF (Likely)[a]	Indeterminate[b]	Non-IPF Dx
	Alternative diagnosis	IPF (Likely)[a]	Indeterminate[b]	Non-IPF Dx	Non-IPF Dx

[a]IPF (Likely) 다음 중 하나를 만족할 때: 1) 중등도에서 심한 견인성 기관지확장증 및 / 또는 세기관지확장증이 50세 이상의 남성 또는 60세 이상의 여성에서 관찰될 때, 2) HRCT 에서 광범위한(>30%) 망상음영이 관찰되며 70세 이상인 경우, 3) 기관지폐포세척액에서 중성구 증가 및 / 또는 림프구증이 없는 경우, 4) 다학제 접근진단을 통해 IPF에 대한 확신 있는 진단이 나오는 경우.
[b]Indeterminate 1) 충분한 조직이 없는 경우 2) 충분한 조직이 있는 경우에는 다학제 접근진단 또는 추가적인 협진을 통해 구체적인 진단으로 재분류될 수 있음.

간질성폐질환 임상진료지침 (2023 1차 개정판

80/M

20PY exsmoker, 40년 전 금연, 사무직 퇴직

1년 전부터 DOE mMRC 1

Bilateral inspiratory crackles

Arthralgia (-), Raynaud ph (-), Morning stiffness (-)

Dry mouth(-), Dry eye(-)

ANA (-)

Rheumatoid factor (-)

Anti CCP (-)

ANCA (-)

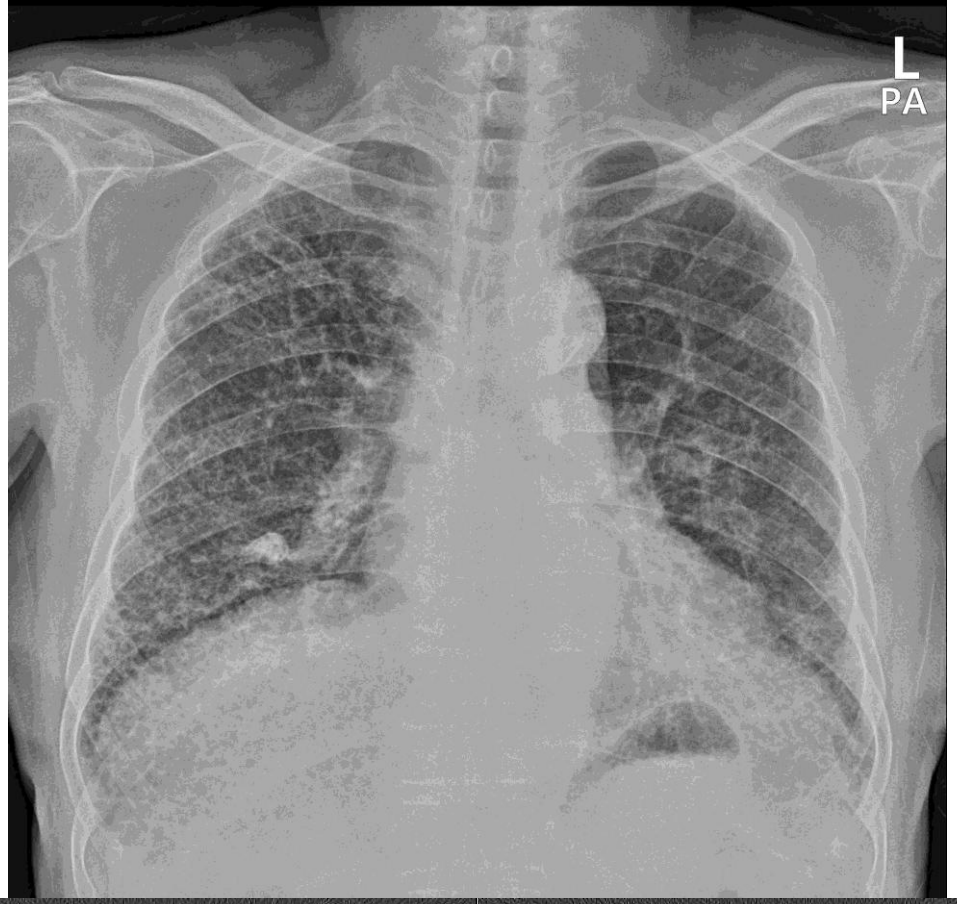
FVC 2.37L (68%)

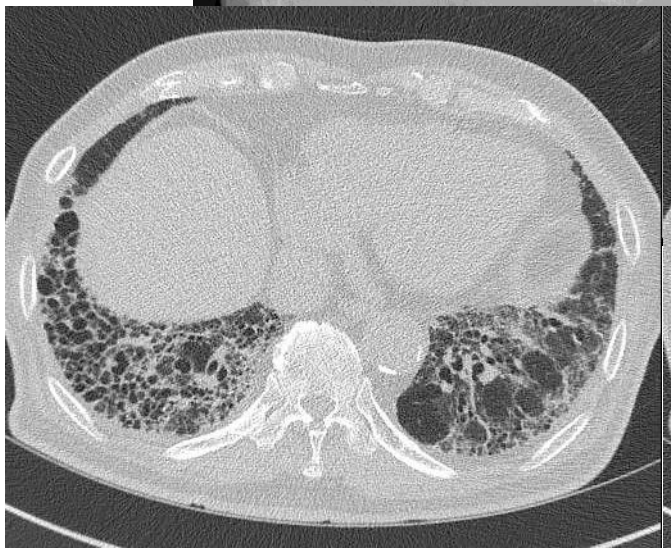
FEV1 1.92L (95%)

FEV1/FVC 81%

DLco 7.52 (55%)

TLC 3.37L (62%)





68/F, Nonsmoker

오빠 IPF 로 2019년 사망, 식당

No respiratory symptoms

보건증위해 찍은 CXR 이상

No definite crackles

Arthralgia (-), Raynaud ph (-),

Morning stiffness (-)

Dry mouth(-), Dry eye(-)

ANA 1: 80, Rheumatoid factor (-)

Anti CCP (-), ANCA (-)

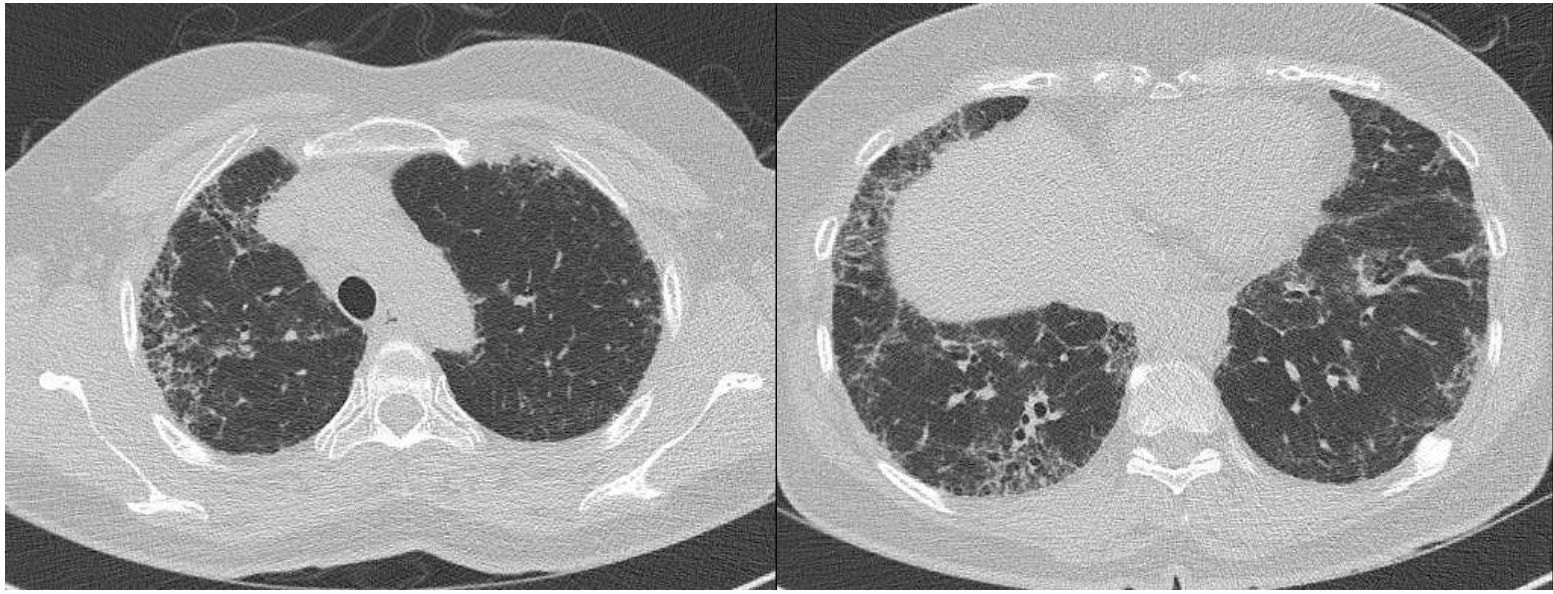
FVC 2.02L (72%)

FEV1 1.72L (81%)

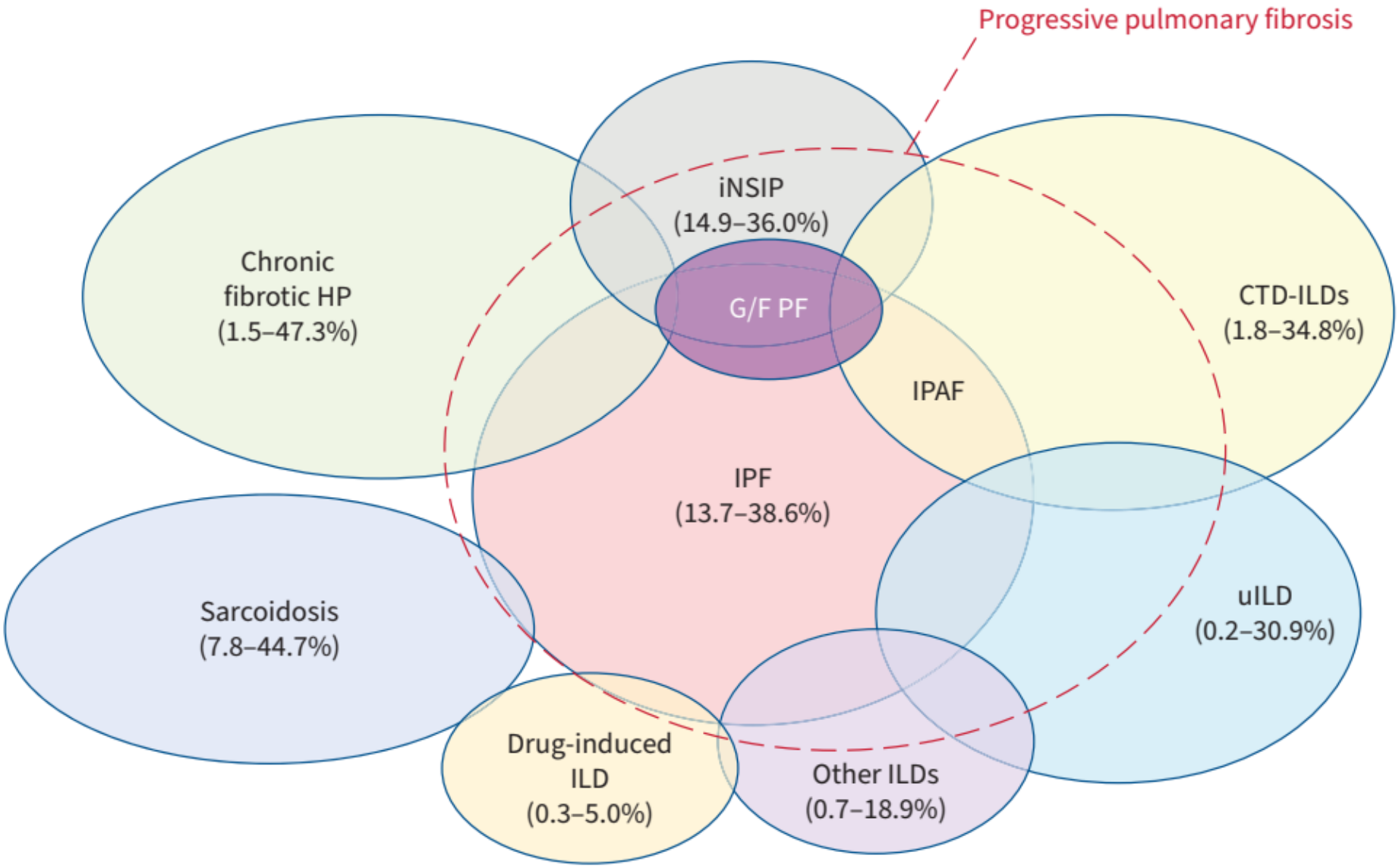
FEV1/FVC 85%

DLco 8.55 (54%)

TLC 3.09L (73%)



ILD a/w Progressive Pulmonary Fibrosis



Rajan et al. Eur Respir J 2023; 61: 2103187

RA 로 치료받던 58세 여자환자

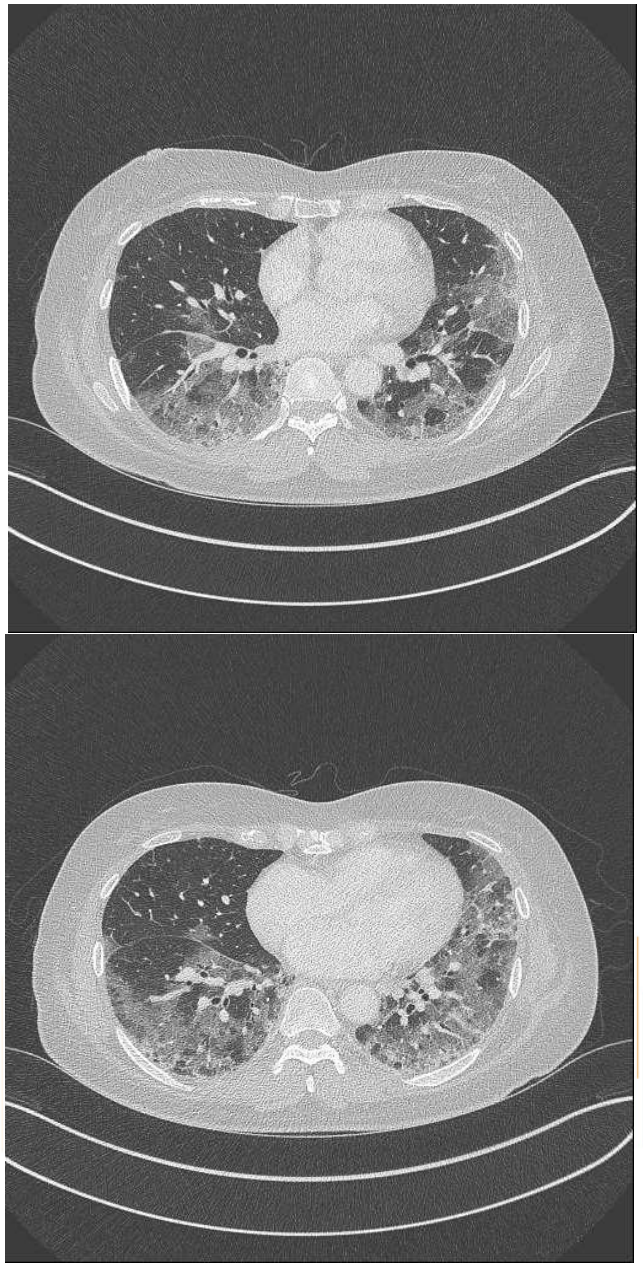
6년 전 호흡곤란을 주소로 내원하여 RA-ILD acute exacerbation 으로 steroid pulse therapy 를 하고 호전

⇒이후 PD tapering, MTX, tacrolimus

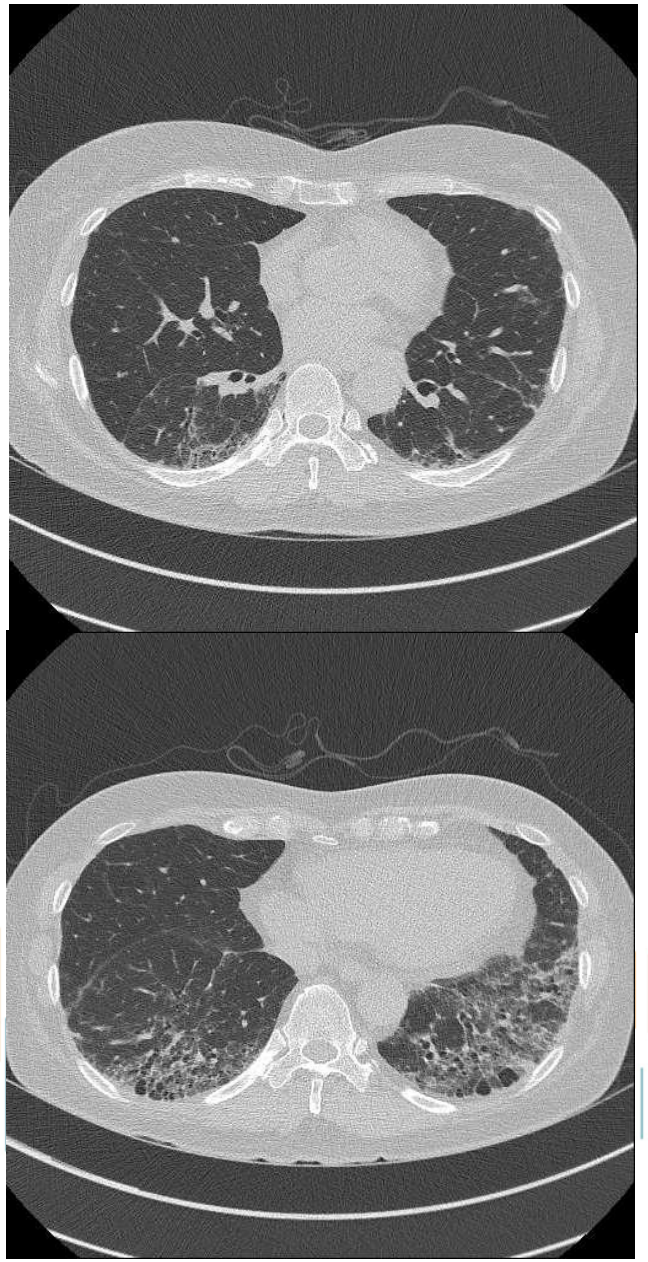
⇒24/11 PFT 악화로 MMF 로 바꿈

⇒약 복용 후 기운이 없고 관절통이 심해져 25/01 자의로 투약 중단

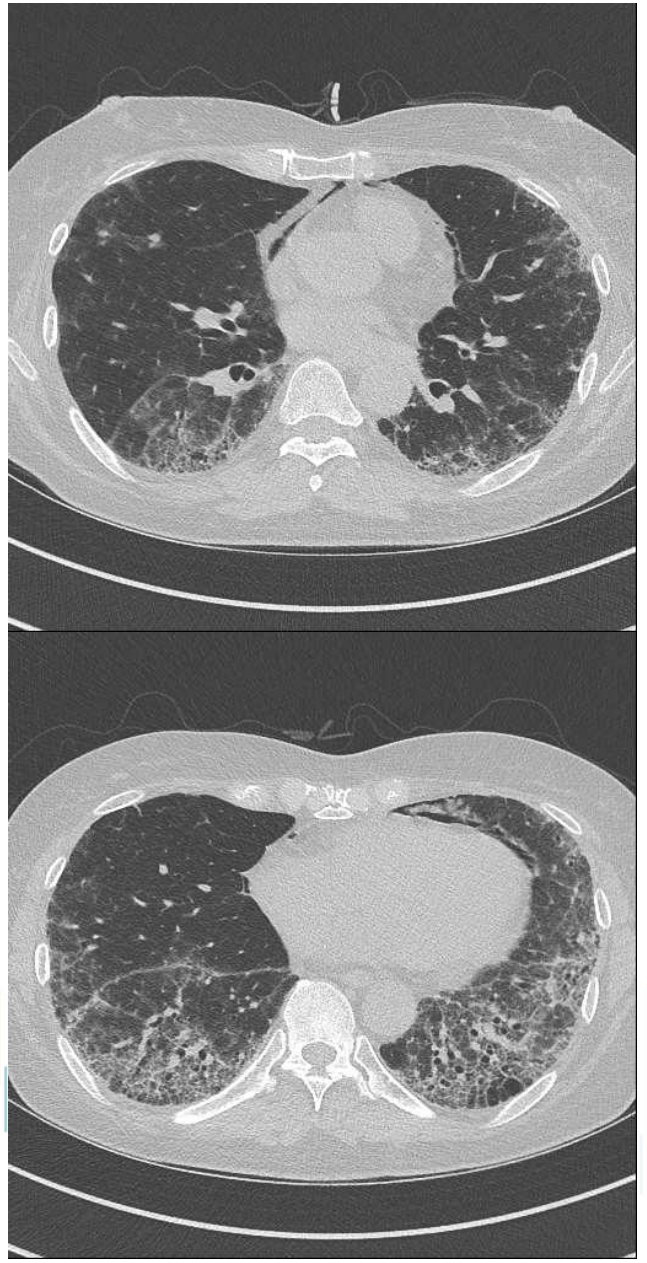
⇒25/2 2주 전부터 시작된 호흡곤란을 주소로 내원



19/04/27



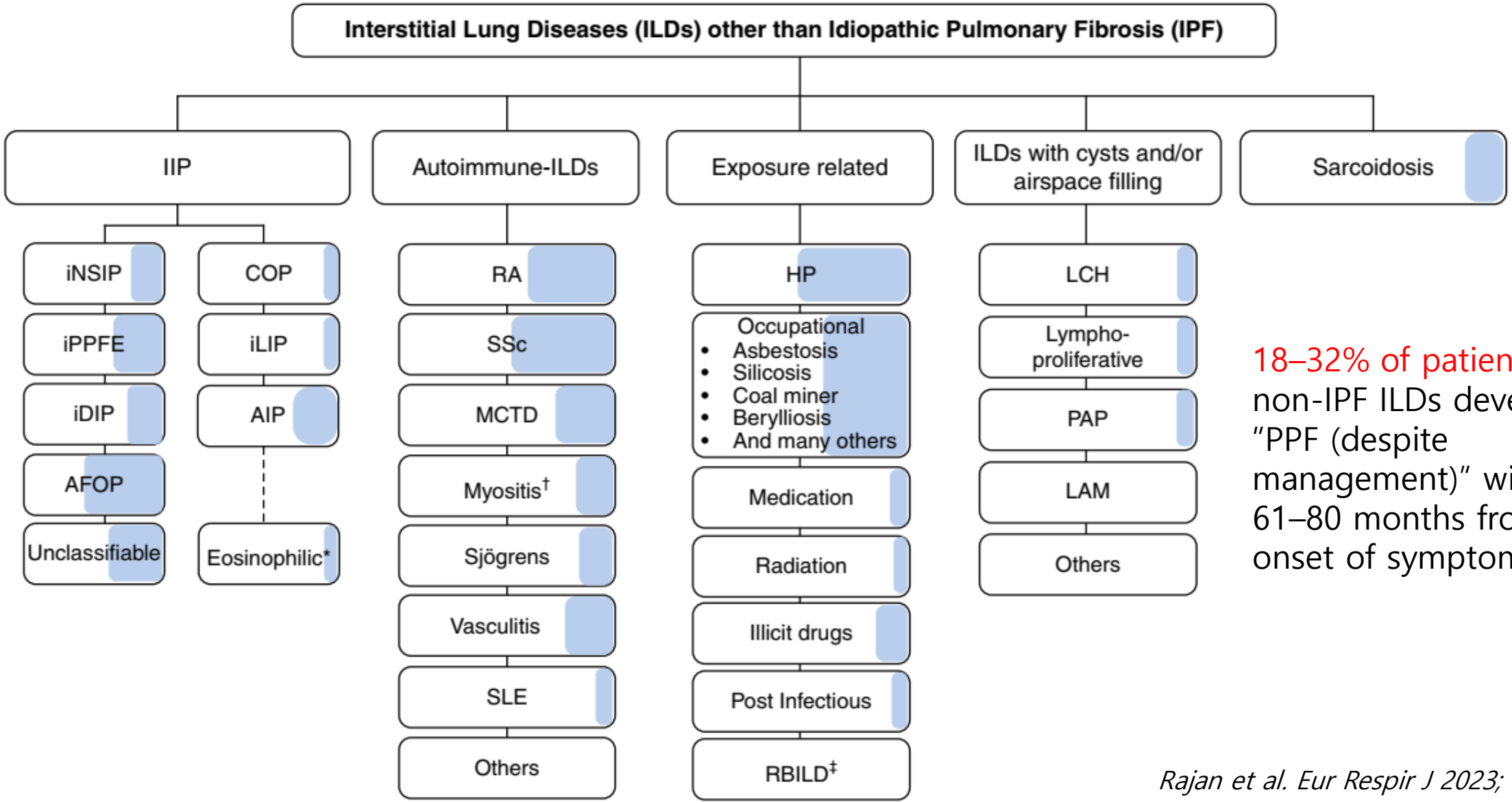
24/10/22



25/02/05

21/03 PD 중단 => 22/08/30시작 =>24/04 다시 중단하고 prn 으로 사용

ILDs manifesting PPF

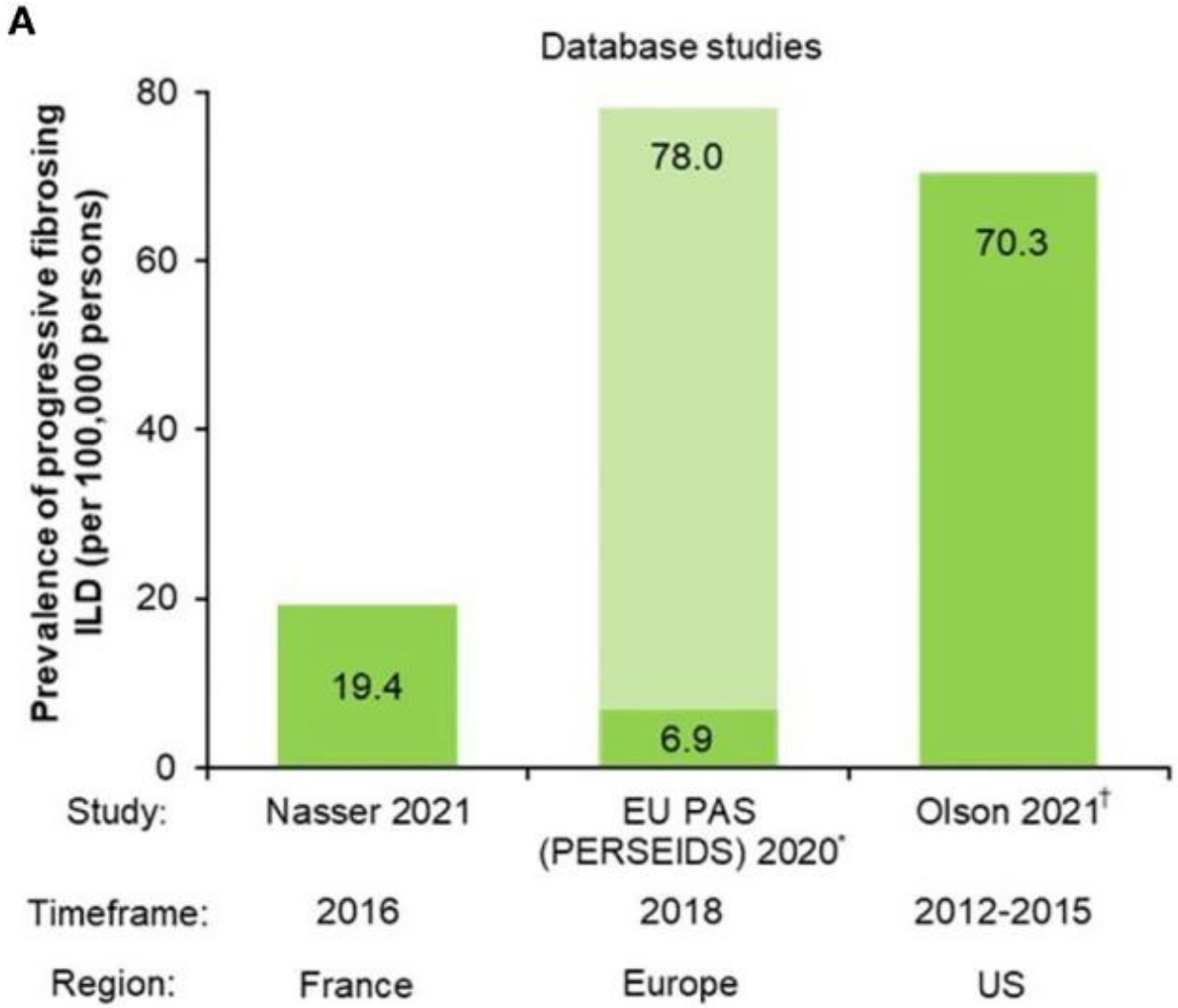


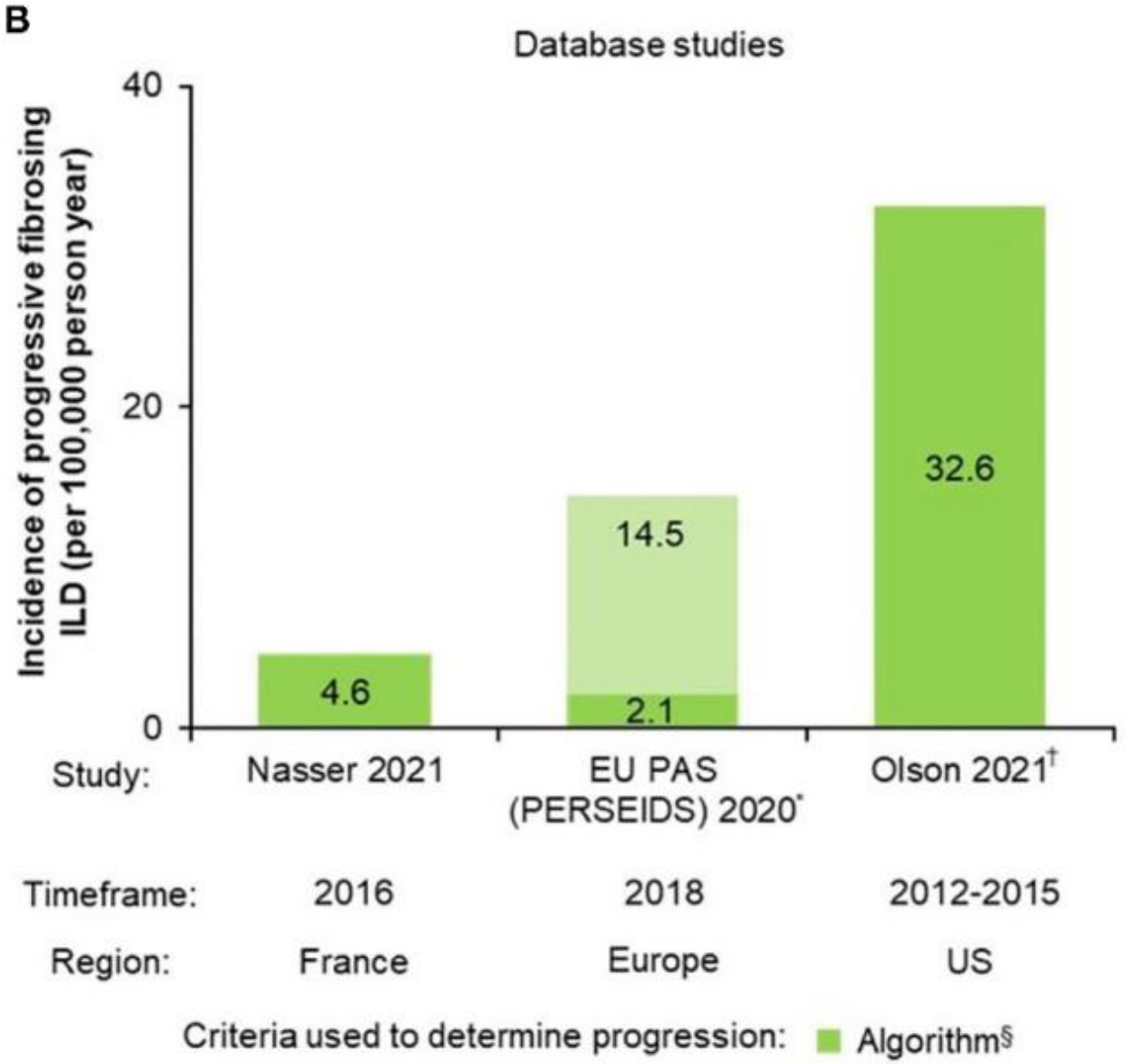
18–32% of patients with non-IPF ILDs develop "PPF (despite management)" within 61–80 months from the onset of symptoms

Rajan et al. Eur Respir J 2023; 61: 2103187

Am J Respir Crit Care Med Vol 205, Iss 9, pp e18–e47, May 1, 2022

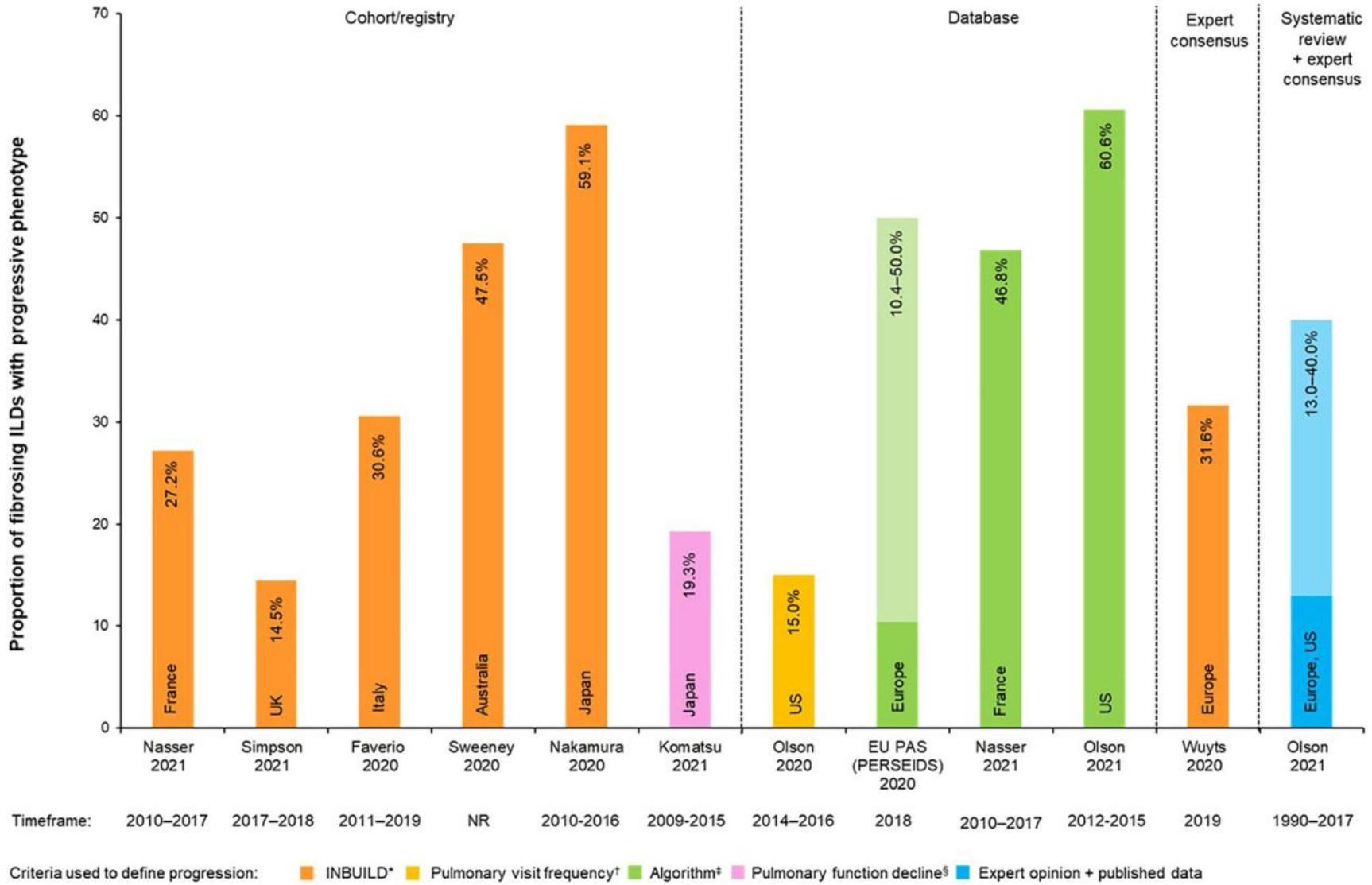
Incidence and Prevalence of PF-ILD





Cottin et al. Front. Med. 9:799912. doi: 10.3389/fmed.2022.799912

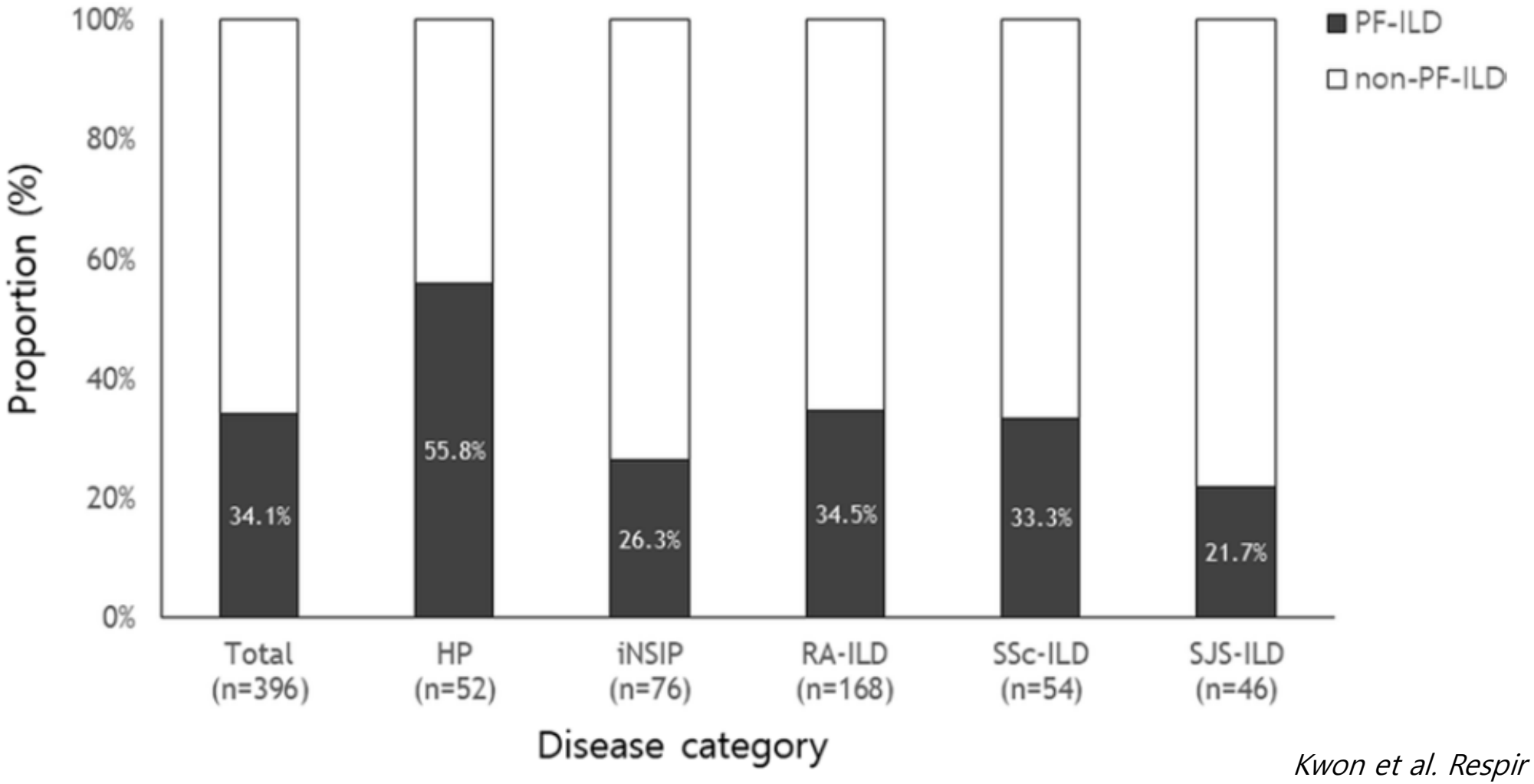
Proportions of non-IPF fibrosing ILDs with progressive phenotype (%)



Cottin et al. Front. Med. 9:799912. doi: 10.3389/fmed.2022.799912

Prevalence of PF-ILD

2005.1-2015.12 Asan Medical center
ILDs other than IPF more than 2 yr FU (n=396)
PF-ILD (n=135)
Non-PF-ILD (n=261)



Kwon et al. Respir Res (2021) 22:282

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Nintedanib in Progressive Fibrosing Interstitial Lung Diseases

K.R. Flaherty, A.U. Wells, V. Cottin, A. Devaraj, S.L.F. Walsh, Y. Inoue, L. Richeldi, M. Kolb, K. Tetzlaff, S. Stowasser, C. Coeck, E. Clerisme-Beaty, B. Rosenstock, M. Quaresma, T. Haeufel, R.-G. Goeldner, R. Schlenker-Herceg, and K.K. Brown, for the INBUILD Trial Investigators*

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Nintedanib for Systemic Sclerosis–Associated Interstitial Lung Disease

Oliver Distler, M.D., Kristin B. Highland, M.D., Martina Gahlemann, M.D., Arata Azuma, M.D., Aryeh Fischer, M.D., Maureen D. Mayes, M.D., Ganesh Raghu, M.D., Wiebke Sauter, Ph.D., Mannaig Girard, M.Sc., Margarida Alves, M.D., Emmanuelle Clerisme-Beaty, M.D., Susanne Stowasser, M.D., Kay Tetzlaff, M.D., Masataka Kuwana, M.D., and Toby M. Maher, M.D., for the SENSCIS Trial Investigators*

Criteria for Progressive Pulmonary Fibrosis

INBUILD		RELIEF	ATS/ERS
Flaherty et al. 2019	George et al. 2020	Behr et al. 2021	Raghu et al. 2022
at least one within 24 months, ● relative decline FVC ≥10% ● relative decline in the 5%≤ FVC <10% +worsening of respiratory symptoms /HRCT ● worsening of respiratory symptoms + an increased extent of fibrosis	at least one within 24 months ● relative decline FVC≥10% ● relative decline FVC ≥ 5% or decline in DLCO ≥15% ● relative decline in FVC ≥5% +worsening of respiratory symptoms /HRCT ● progressive symptoms + increased fibrosis on HRCT,	● CTD-ILD, HP, iNSIP, or asbestos-induced lung fibrosis, FVC 40–90% predicted, DLCO 10–90% predicted ● annual decline of FVC ≥ 5%	Meet at least two of the following three criteria occurring within 1 year ● worsening respiratory symptoms; ● physiological evidence of disease progression (either of the following): a. absolute decline in FVC ≥ 5% b. absolute decline in DLCO (corrected for Hb) ≥10% ● radiological evidence of disease progression

Definition of Progressive Pulmonary Fibrosis

Definition of PPF

FVC 50%=> 40% 로 감소

Absolute decline : 50-40 = 10%

Relative decline : 50-40/0.5 = 20%

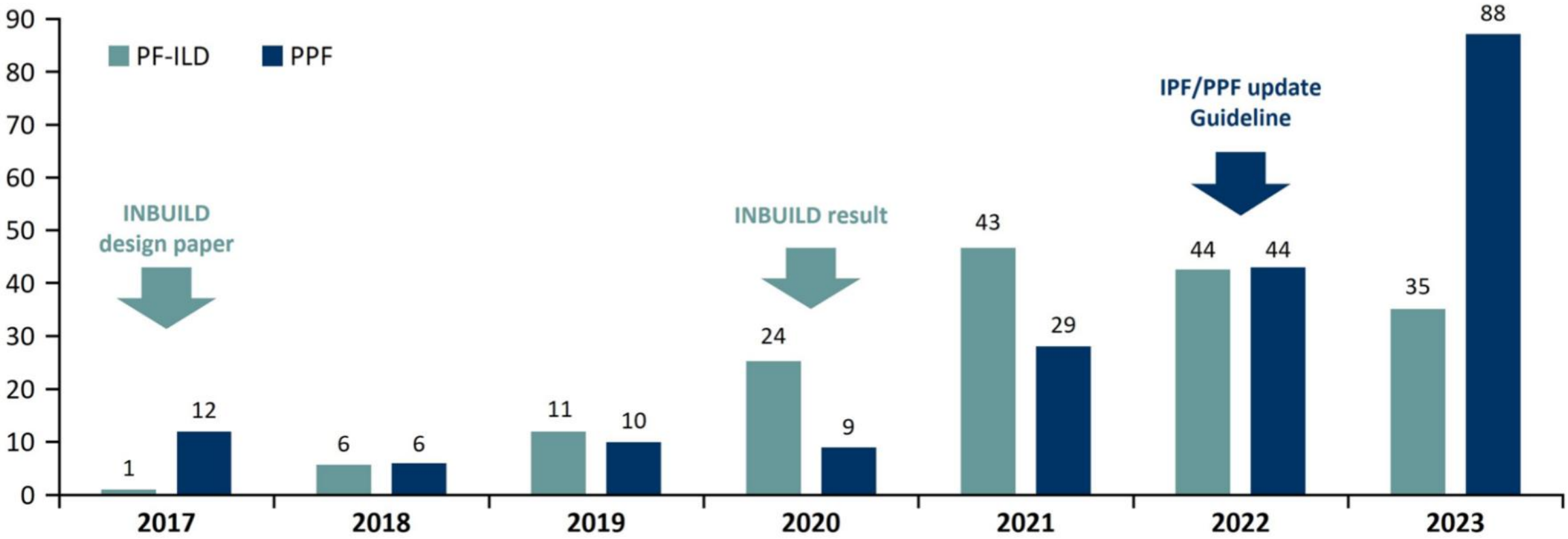
In a patient with ILD of known or unknow
evidence of pulmonary fibrosis, PPF is d
criteria occurring within the past year wit

1 Worsening respiratory symptoms

2 Physiological evidence of disease progression (either of the following):
   a. Absolute decline in FVC ⩾5% predicted within 1 yr of follow-up
   b. Absolute decline in D_{LCO} (corrected for Hb) ⩾10% predicted within 1 yr of follow-up

3 Radiological evidence of disease progression (one or more of the following):
   a. Increased extent or severity of traction bronchiectasis and bronchiolectasis
   b. New ground-glass opacity with traction bronchiectasis
   c. New fine reticulation
   d. Increased extent or increased coarseness of reticular abnormality
   e. New or increased honeycombing
   f. Increased lobar volume loss

Number of Submitted Articles on PF-ILD/PPF



【Survey summary】

Database：PubMed

Search terms：

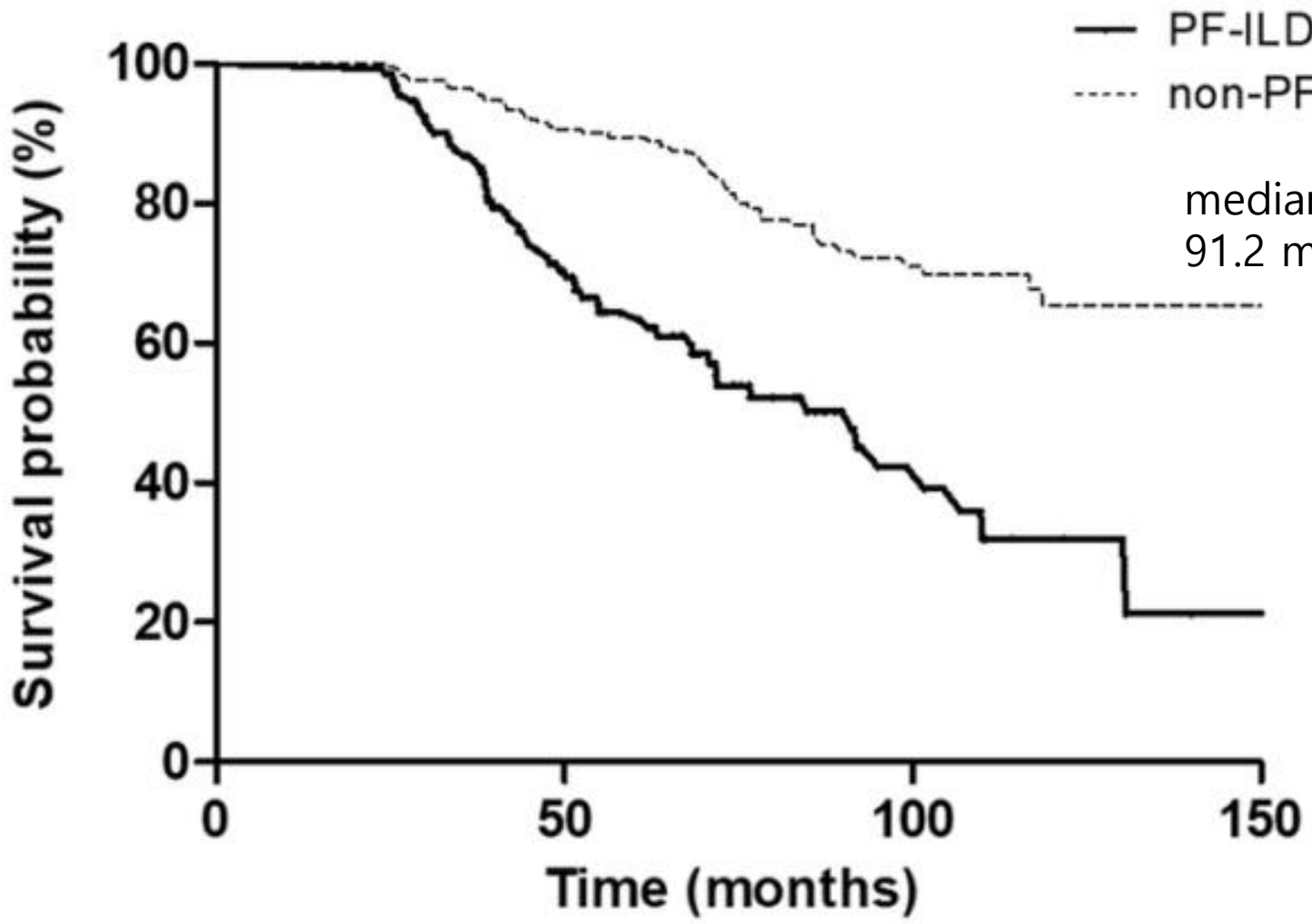
PPF: ["PROGRESSIVE PULMONARY FIBROSIS"[TW] OR PPF[TW] AND PROGRESSIVE PULMONARY FIBROSIS]

PF-ILD: ["PROGRESSIVE FIBROSING INTERSTITIAL LUNG DISEASE*"[TW] OR PFILD[TW] OR PF-ILD[TW] OR PFILDS[TW] OR PF-ILDS[TW]]

As of 31Dec2023

Adv Ther (2025) 42:2988–3001

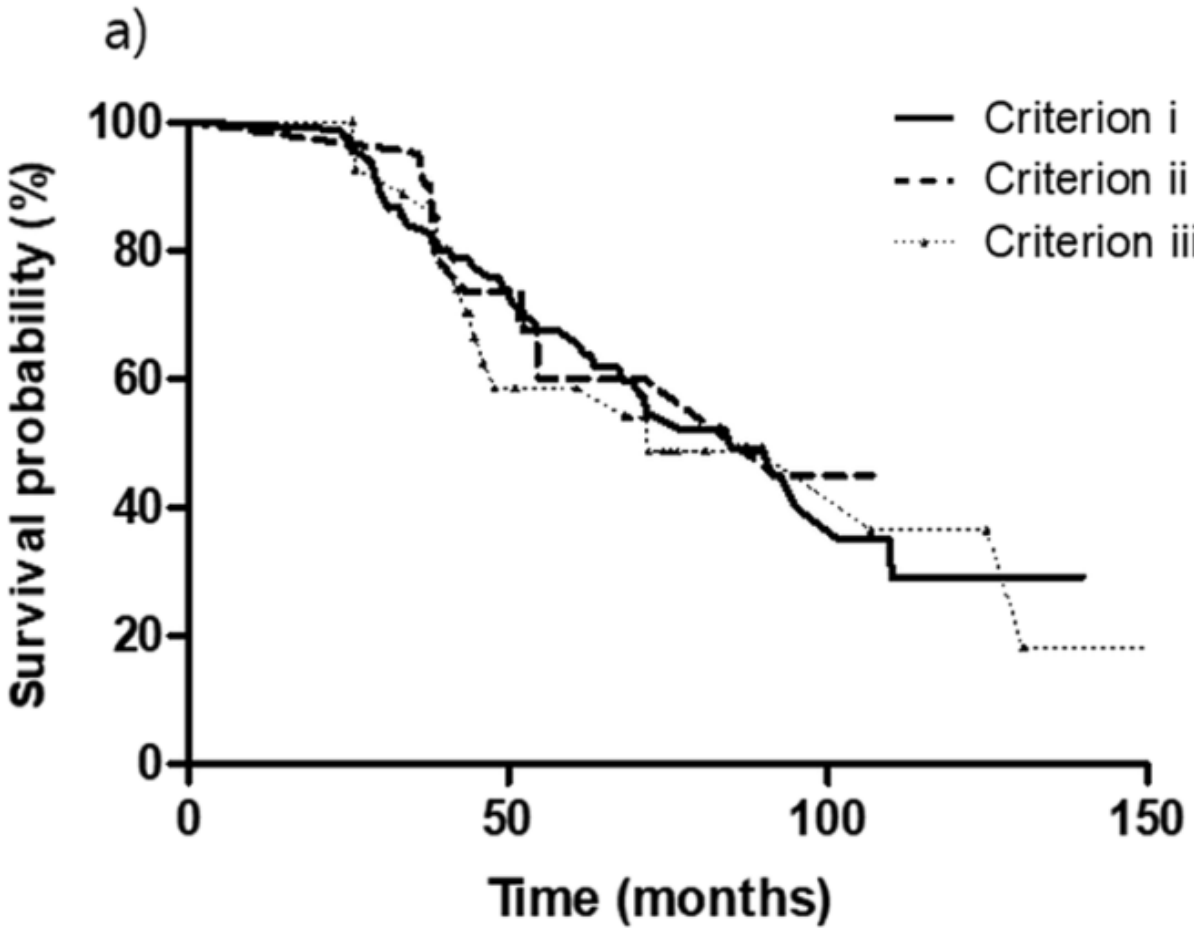
Survival of PF-ILD

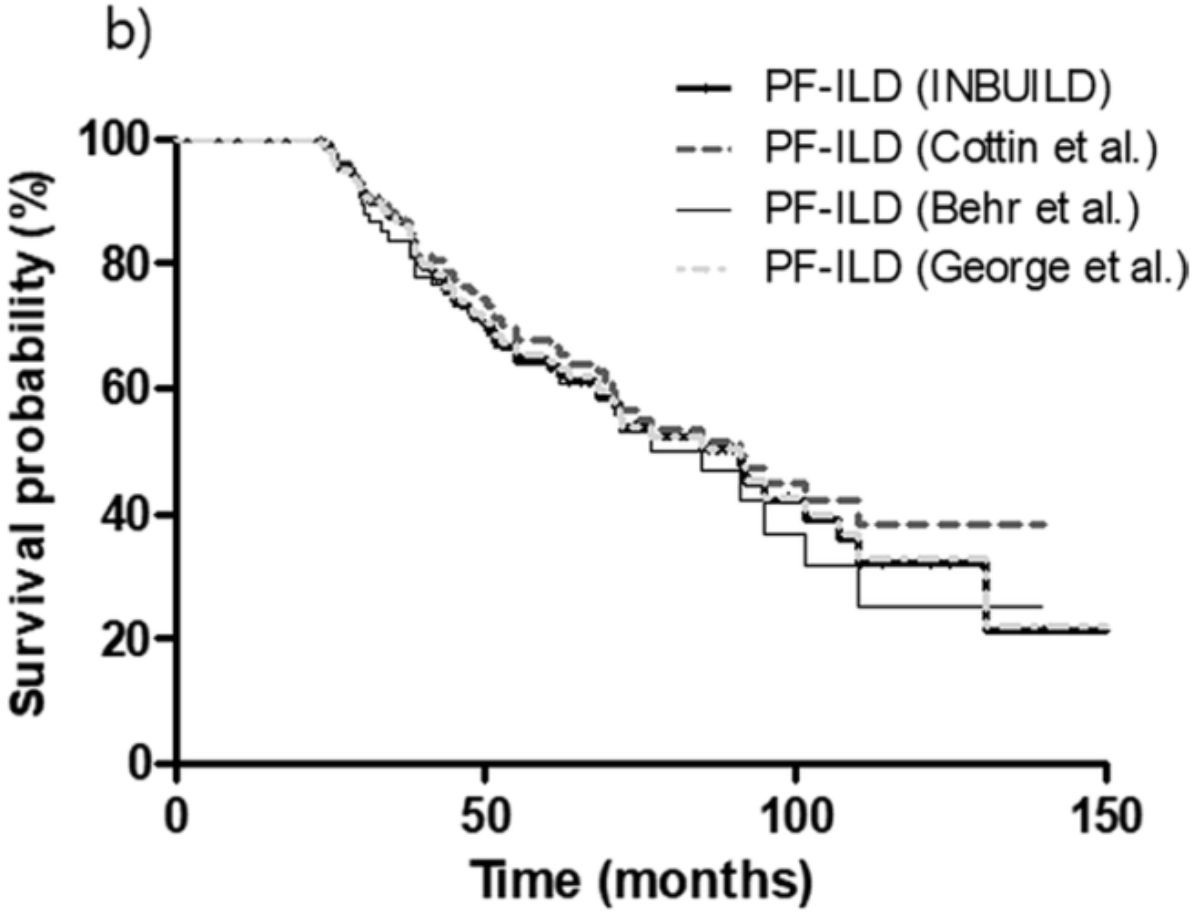


median survival, P < 0.001
91.2 months vs. not reached

Kwon et al. Respir Res (2021) 22:282

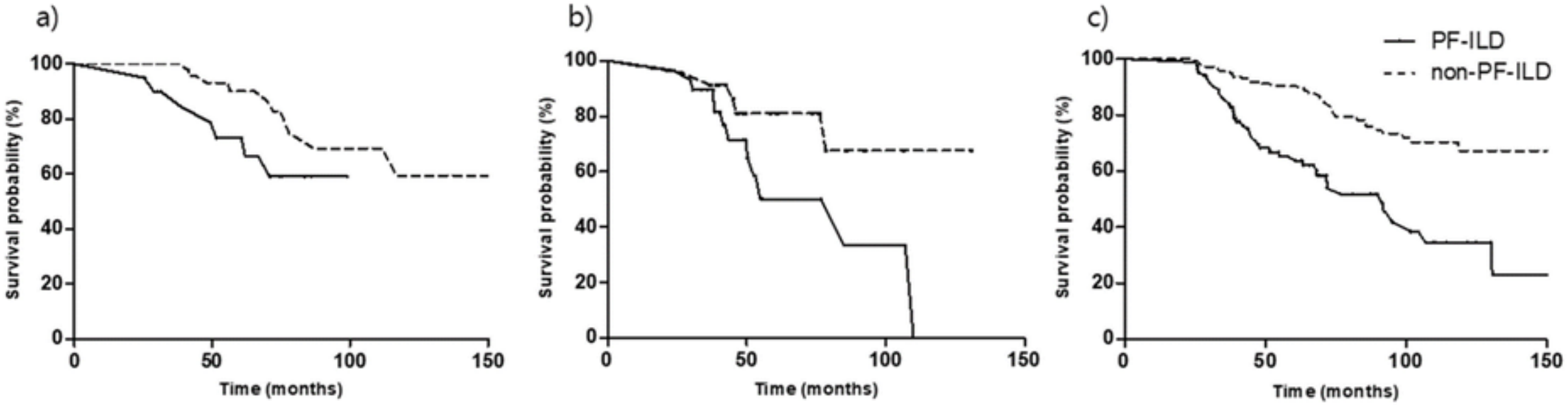
Survival according to diagnostic criteria





Kwon et al. Respir Res (2021) 22:282

Survival of PF-ILD



iNSIP

Fibrotic HP

Autoimmune ILD

Kwon et al. Respir Res (2021) 22:282

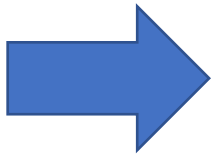
Complications between the PF-ILD and non-PF-ILD

	Total	PF-ILD	Non-PF-ILD	*P*-value
Number of patients	396	135	261	
Follow-up duration, months	68.9 ± 32.5	59.9 ± 28.4	73.5 ± 33.5	< 0.001
Unexpected respiratory hospitalisation	120 (30.3)	62 (45.9)	58 (22.2)	< 0.001
Acute exacerbation[a]	74 (18.7)	38 (28.1)	36 (13.8)	0.001
Time interval from diagnosis (months)	34.6 ± 28.2	29.3 ± 20.5	40.2 ± 33.9	0.037
Pneumonia	35 (8.8)	16 (11.9)	19 (7.3)	0.138
Pneumothorax	7 (1.8)	5 (3.7)	2 (0.8)	0.048
Pulmonary hypertension	89 (22.5)	45 (33.3)	44 (16.9)	< 0.001
Lung cancer	28 (7.1)	14 (10.4)	14 (5.4)	0.065

Kwon et al. Respir Res (2021) 22:282

Prospective registry of PF-ILD

- the Canadian Registry for Pulmonary Fibrosis between 2015-2020
- Definition
  - relative FVC decline ≥10%
  - death, lung transplantation
  - any 2 of: relative FVC decline ≥5 and <10%, worsening respiratory symptoms, or worsening fibrosis on computed tomography
  - within 24 months of diagnosis



PF-ILD1,376/2,746 (50%)

Hambly et al. Eur Respir J 2022; in press

PF-ILD by diagnosis

Diagnosis	Total number of patients†	Proportion meeting PF-ILD criteria‡
IPF	718 (26%)	427 (59%)
Hypersensitivity pneumonitis	216 (8%)	125 (58%)
CTD-ILD	902 (33%)	402 (45%)
Systemic sclerosis	334	163 (49%)
Rheumatoid arthritis	189	87 (46%)
Myositis*	166	69 (42%)
Mixed CTD	65	28 (43%)
Sjogren's	54	20 (37%)
SLE	28	7 (25%)
Undifferentiated	64	26 (41%)
Unclassifiable ILD	550 (20%)	281 (51%)
IPAF	92	51 (55%)
Other fibrotic ILD		
Sarcoidosis		
Idiopathic NSIP		
Occupational ILD		
Drug-induced ILD		
Smoking-related ILD		
Cryptogenic organizing pneumonia		
Vasculitis		
Other°	125	53 (43%)

Table 4: Hazard ratios for progression to PF-ILD by diagnosis

ILD diagnosis	Hazard ratio with 95% confidence intervals
IPF	*Reference*
HP	0.96 (0.79-1.17)
CTD-ILD	0.65 (0.56-0.74)
Unclassifiable	0.82 (0.71-0.96)

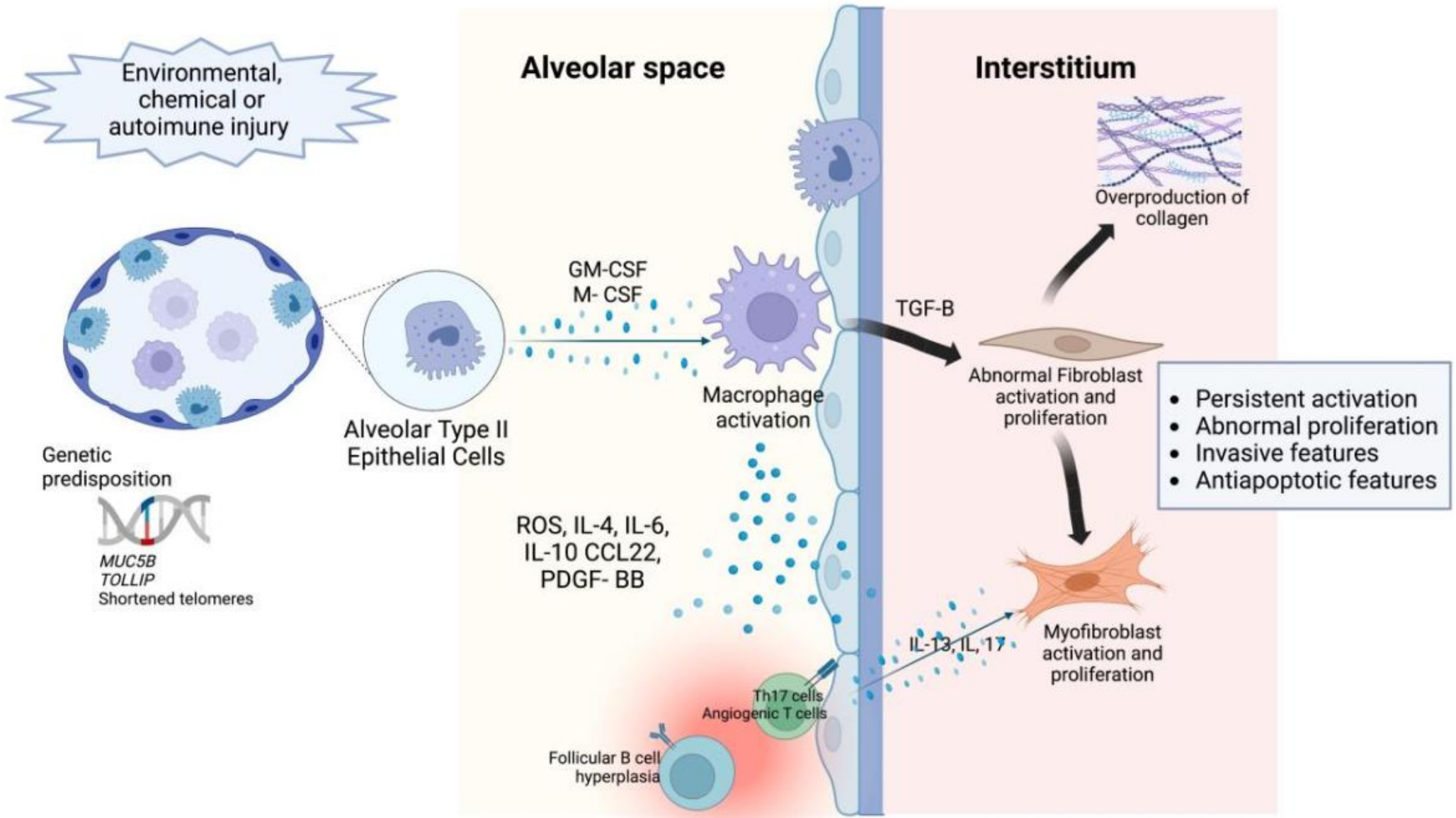
Hambly et al. Eur Respir J 2022;

Risk factors for PF-ILD

Variable	Number (%)	Unadjusted HR	Adjusted HR
Age at diagnosis			
Under 50	403 (15%)	Reference	Reference
50-59	578 (21%)	1.25 (1.04-1.52)*	1.25 (1.02-1.55)*
60-69	910 (33%)	1.28 (1.07-1.52)*	1.29 (1.06-1.57)*
70-79	716 (26%)	1.38 (1.15-1.65)*	1.33 (1.08-1.64)*
Over 80	139 (5%)	1.64 (1.27-2.14)*	1.53 (1.12-2.08)*
Male sex	1,336 (49%)	1.17 (1.06-1.30)*	1.20 (1.06-1.36)*
Ethnicity			
Caucasian	2,196 (80%)	Reference	Reference
Asian	279 (10%)	0.91 (0.76-1.09)	0.91 (0.74-1.12)
Black	52 (2%)	0.81 (0.53-1.23)	0.82 (0.50-1.33)
Other	219 (8%)	0.93 (0.76-1.13)	0.96 (0.74-1.24)
Per-10 pack-year smoking increase	-	1.03 (1.01-1.05)*	1.02 (0.97-1.05)
Family history of pulmonary fibrosis	289 (11%)	0.90 (0.76-1.08)	0.94 (0.78-1.14)
History of COPD	477 (20%)	1.04 (0.90-1.19)	0.93 (0.80-1.08)
History of GERD	558 (23%)	1.20 (1.05-1.36)*	1.22 (1.06-1.40)*
History of surgical lung biopsy	579 (21%)	1.10 (0.97-1.25)	1.12 (0.97-1.29)
Baseline FVC % predicted			
≥90	759 (30%)	Reference	Reference
70-89	938 (37%)	1.30 (1.13-1.50)*	1.13 (0.97-1.31)
<70	818 (33%)	1.51 (1.31-1.74)*	1.23 (1.03-1.43)*
Baseline % predicted DLCO			
>75	611 (24%)	Reference	Reference
60-75	632 (25%)	1.50 (1.26-1.77)*	1.44 (1.21-1.73)*
40-60	897 (36%)	1.55 (1.32-1.82)*	1.42 (1.20-1.69)*
<40	378 (15%)	2.24 (1.87-2.68)*	(1.71-2.56)*

Hambly et al. Eur Respir J 2022

Roles of innate and adaptive immunity in CTD-ILD



Chiang et al. Int. J. Mol. Sci. 2023,24, 2405.

43세 남자

C.C. 3개월 전부터 시작된 다발성 관절통, 한달 전부터 시작된 호흡곤란

P.I. 평소 건강하던 분으로 내원 3개월 전부터 무릎을 포함한 전신 관절통이 발생하여 타원 류마티스내과 내원하여 methylprednisolone 4mg/일로 시작하고 감량 중이었으나 내원 한달 전부터 호흡곤란, 발열 및 관절통 지속되어 응급실 통해 입원

SHx 직업 사무직 음주/흡연 소주 1.5병*주 3회, 흡연자(20 갑년)

P/E 혈압 113/78 mmHg, 심박수 94회/분, 호흡수 20회/분, 체온 37.4℃

양측 폐하부 수포음

안면에는 연보라 발진(Heliotrope rash),

손에는 고트론구진(Gottron's papules),

손톱주위 홍반(Periungual erytherma) 이 보였다

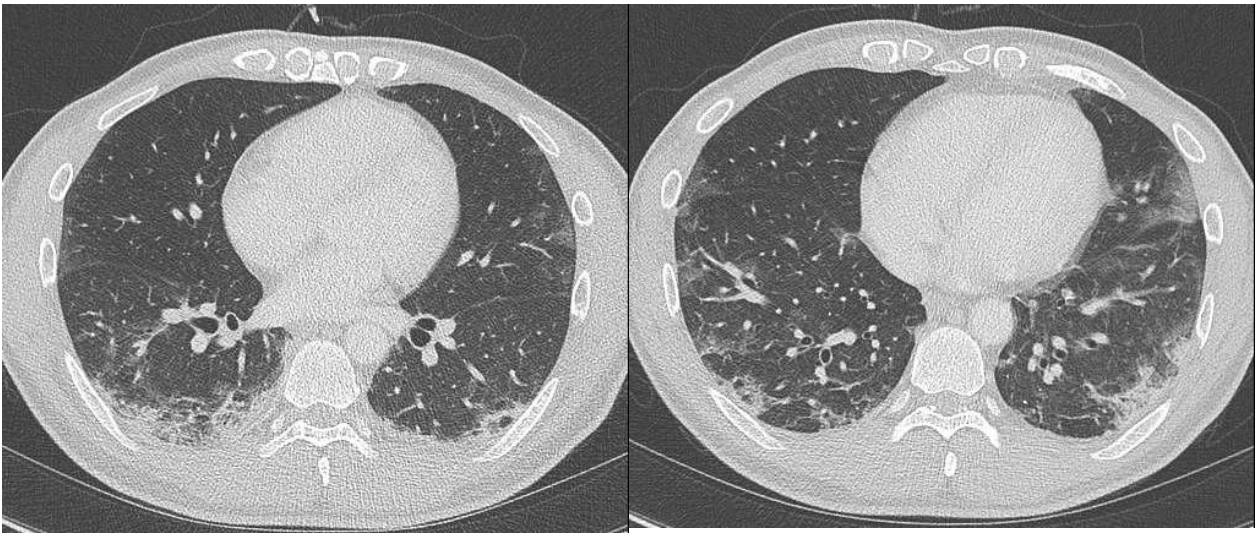
Lab ANA(-), Rheumatoid factor 6.3 IU/ml (0-14),

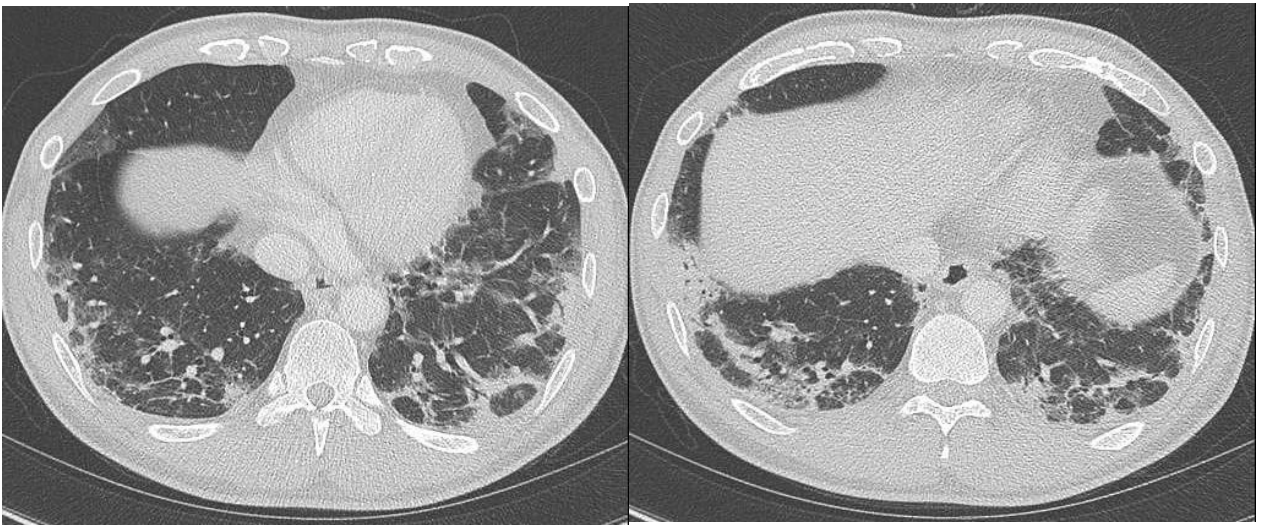
Anti-CCP (-), ANCA (-), Anti Jo-1 Ab (-)

AST/ALT 68/103 IU/L, Aldolase 8.6U/L (WNL)

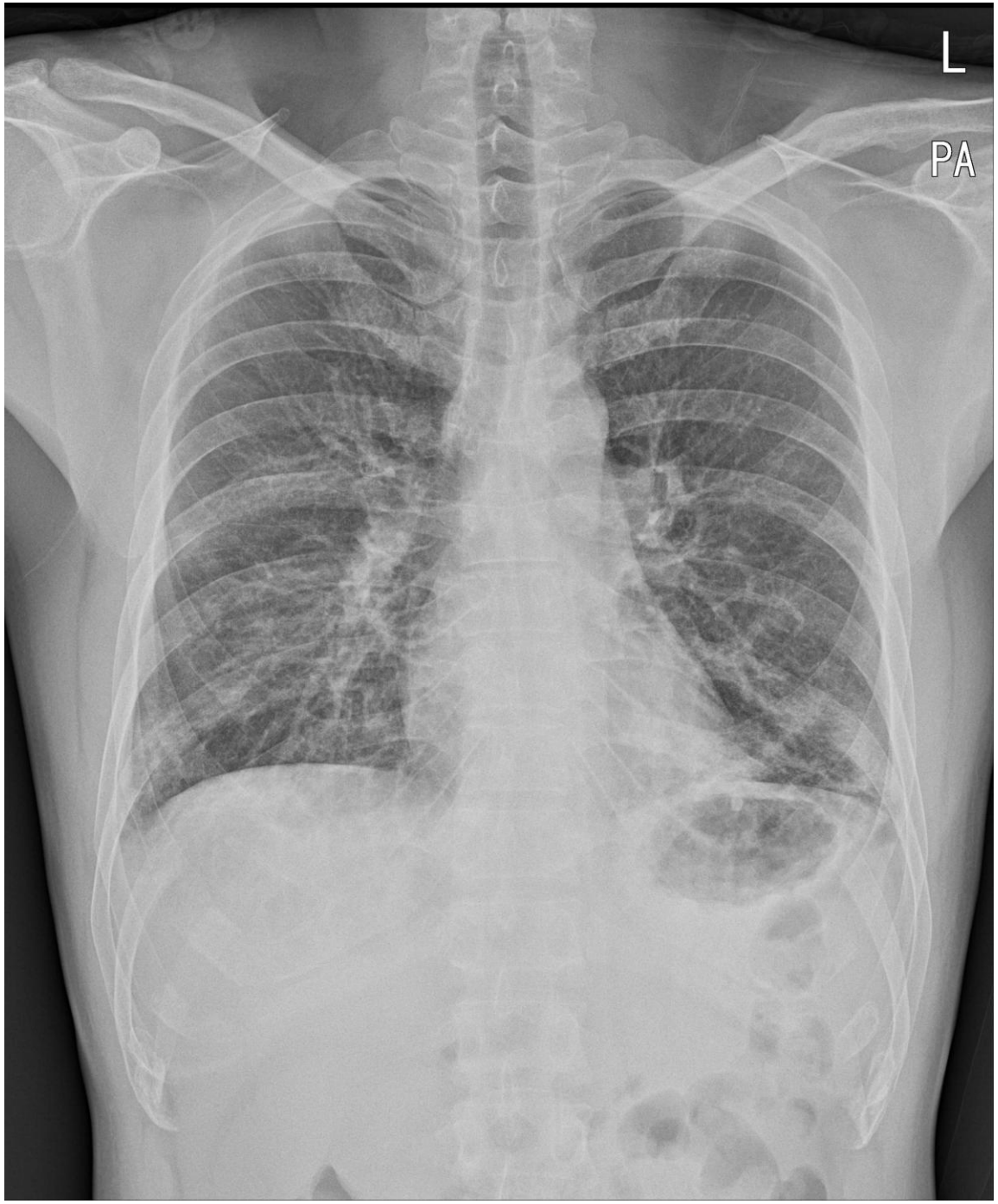
PFT FVC 2.90L (63%), FEV_1 2.21L (57%), FEV_1/FVC 76%

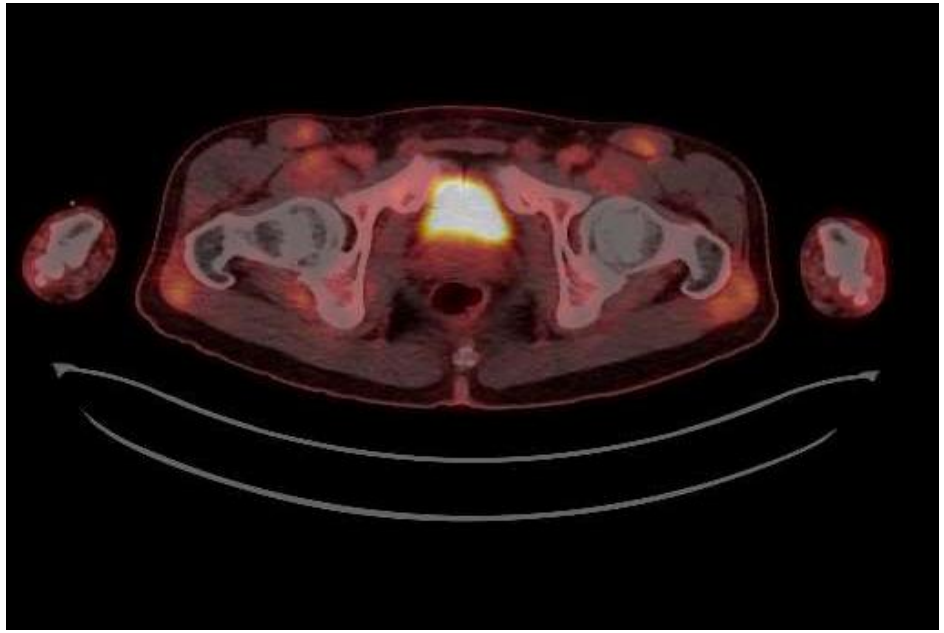
DLco 9.50 ml/min/mmHg (35%)

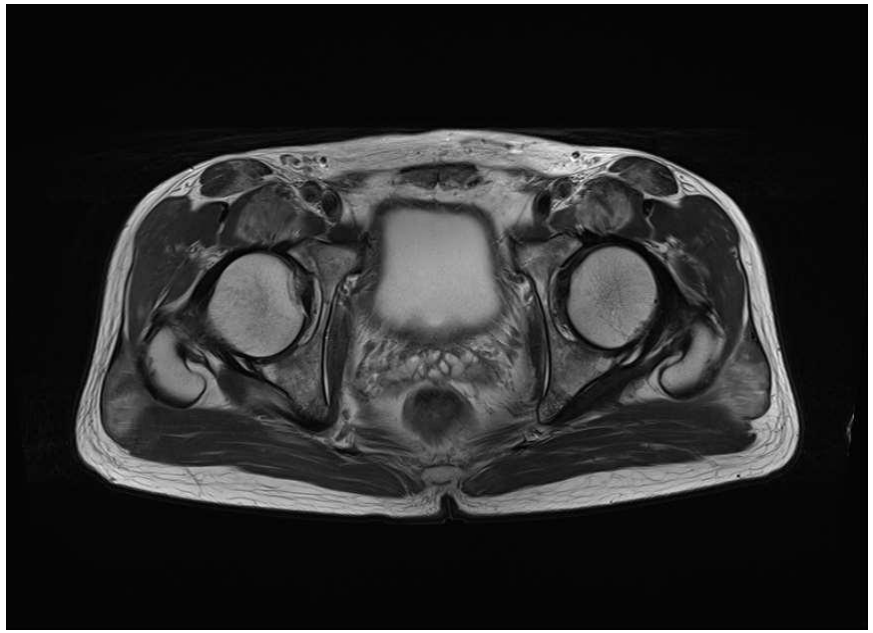


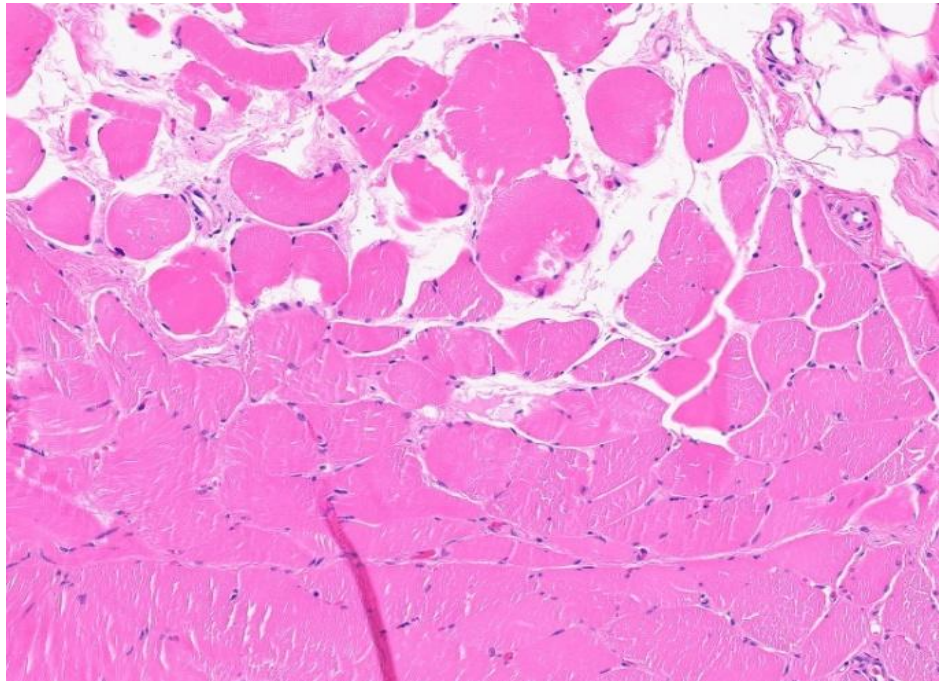


2024-02-19





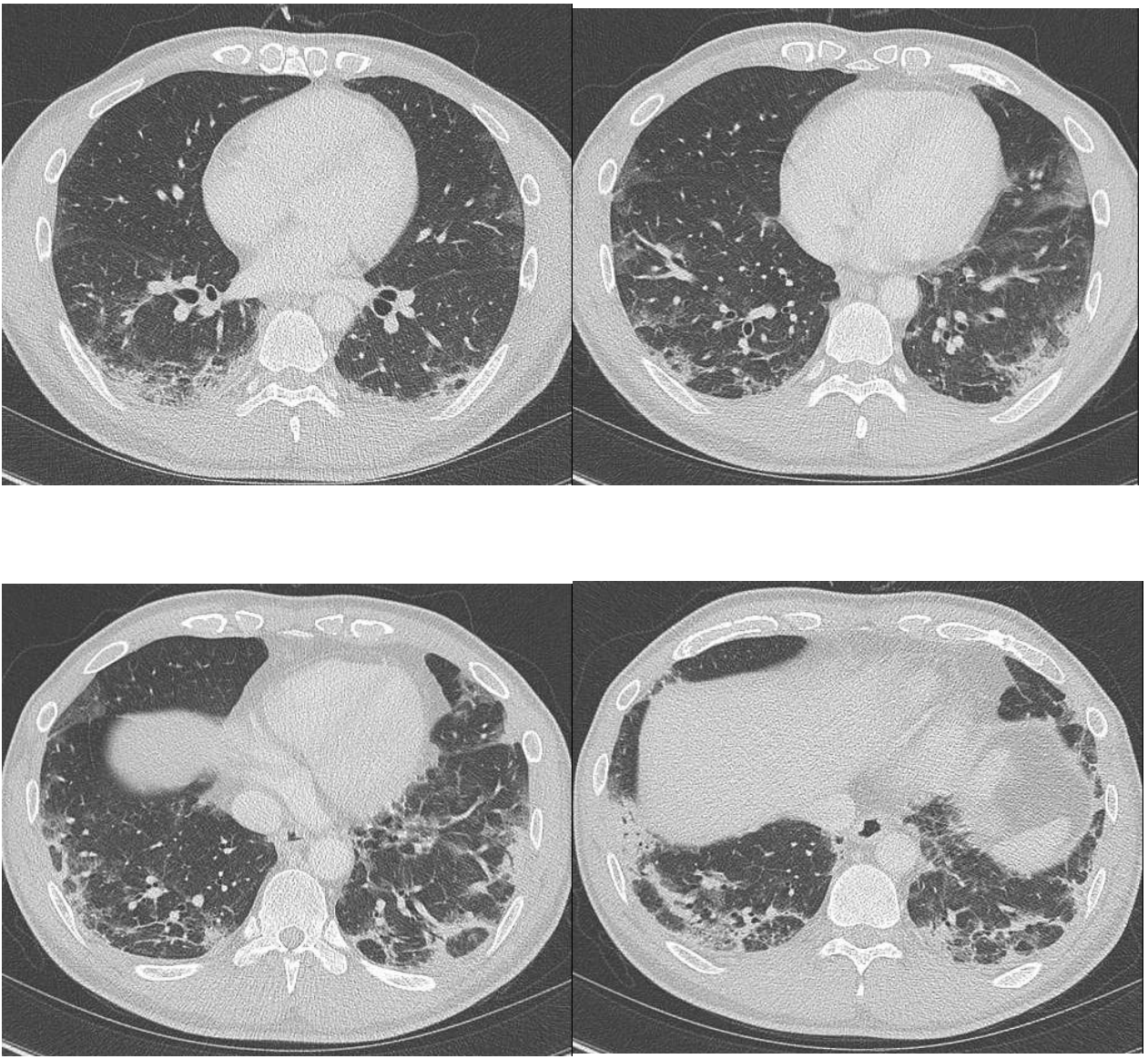




Skeletal muscle, (left thigh), incisional biopsy ;

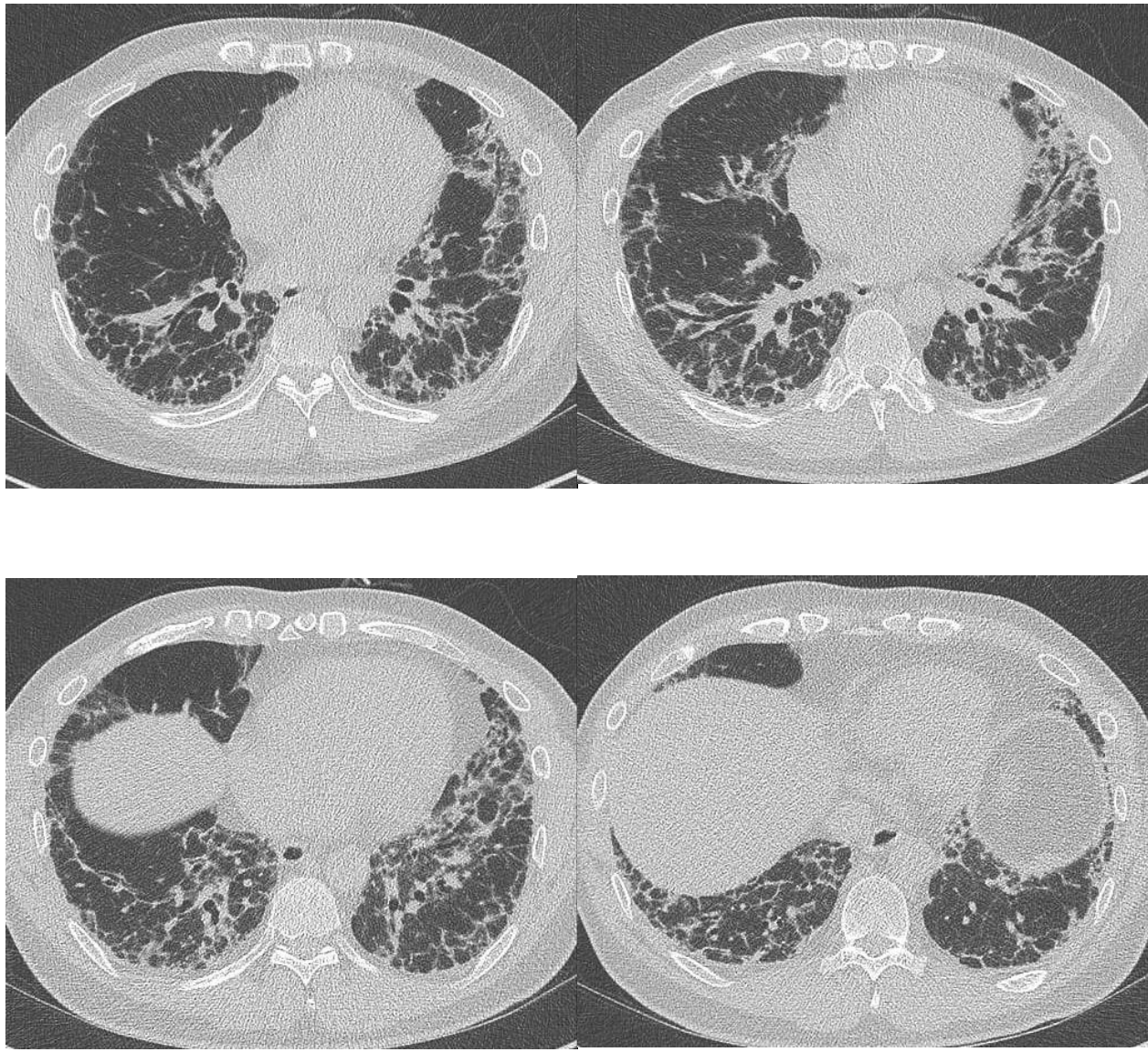
- Group atrophy of muscle fibers
- Minimal endomyseal inflammatory infiltrates
- No evidence of perimyseal vasculitis
- Endomyseal fatty infiltration, mild

suggestive of neurogenic musculopathy.

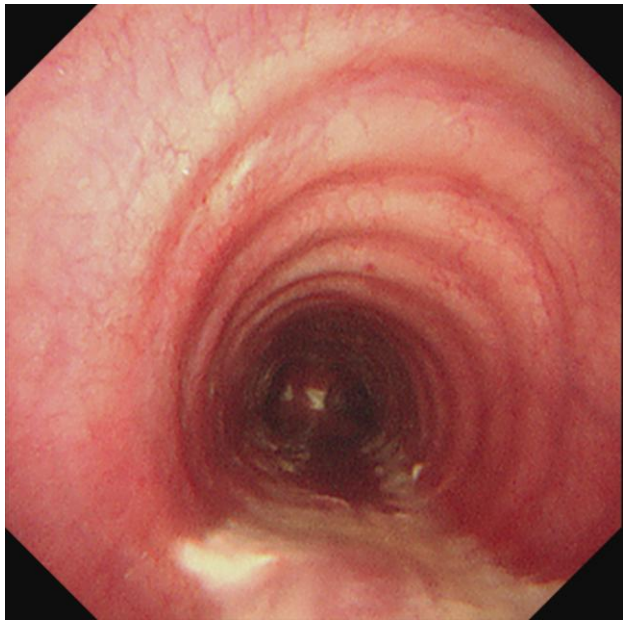


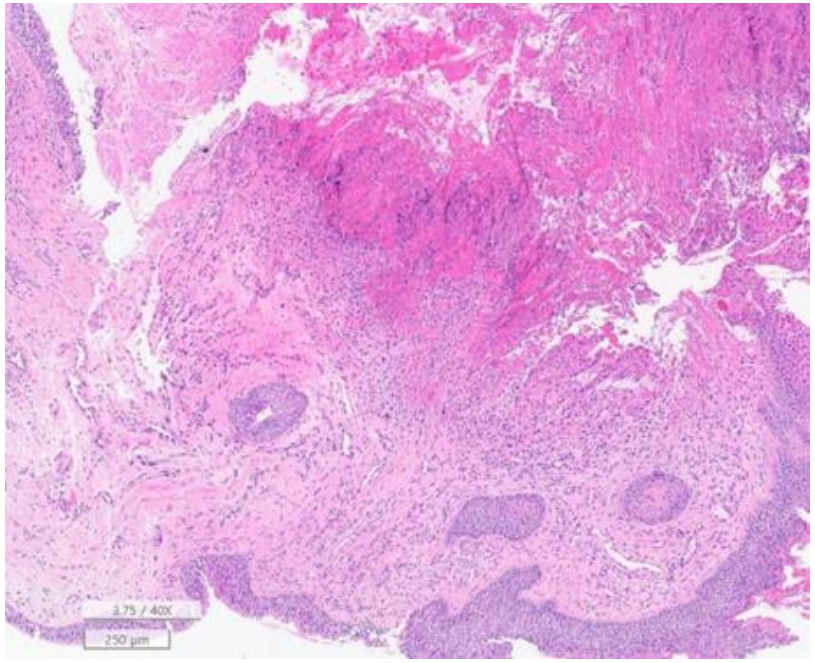
2024-02-19

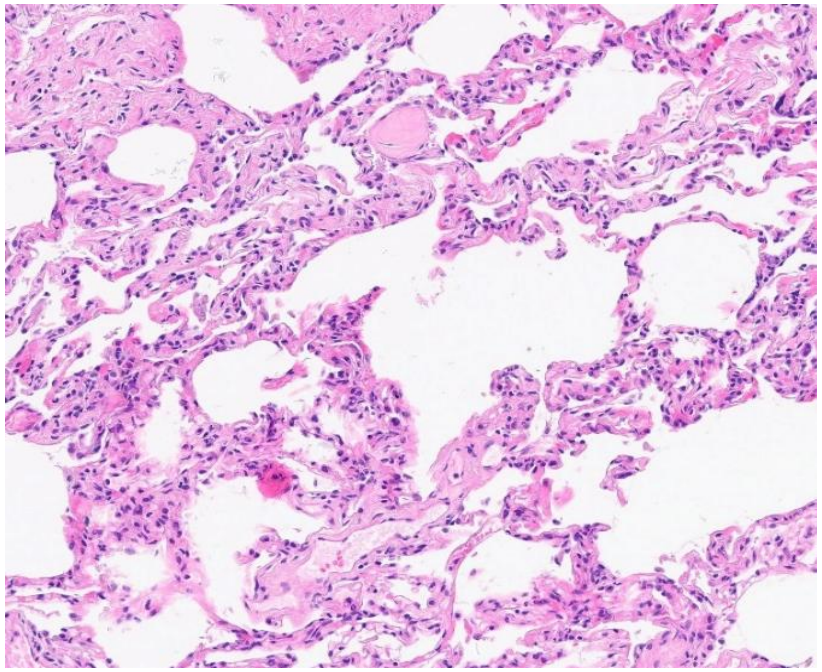
Steroid pulse
PD+ MMF



2024-06-12







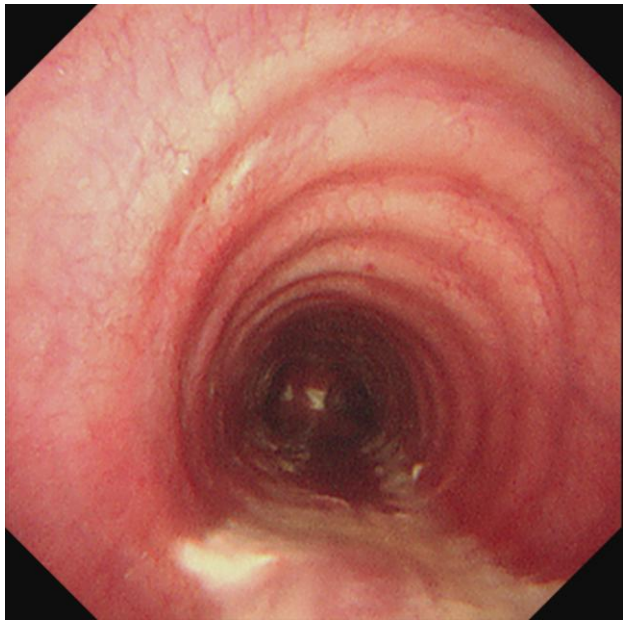
2024-06-19

나이/성별	42/M	의뢰의사	제갈양진	검사일시	2024/07/09 04:06
비 고	24070821824	검체종류	Serum	보고일시	2024/07/24 17:16

검사결과

Dx : Secondary fibrotic NSIP associated with Clinically Amyopathic Dermatomyositis

OJ AB	< 11	< 11 SI	NXP-2 AB	< 11	< 11 SI
SRP AB	< 11	< 11 SI			



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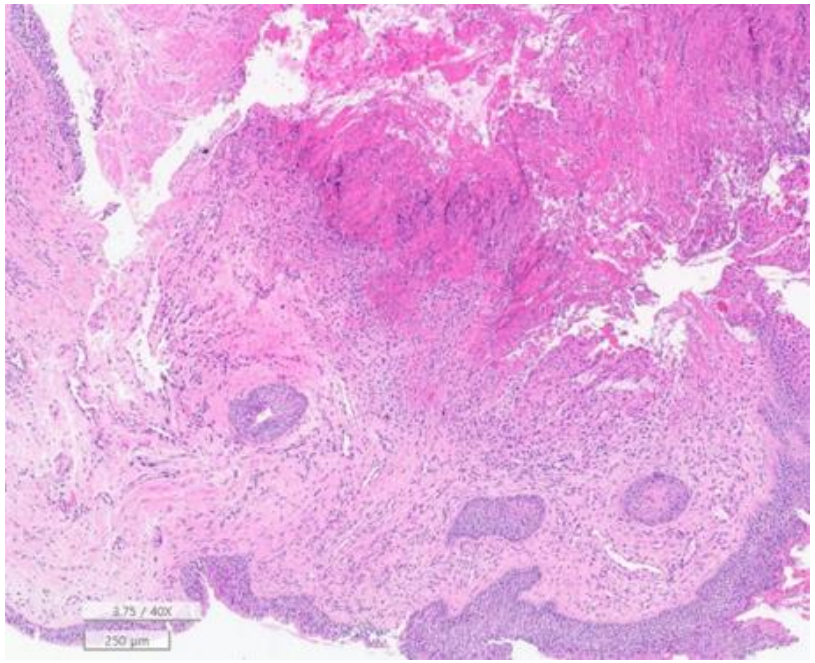
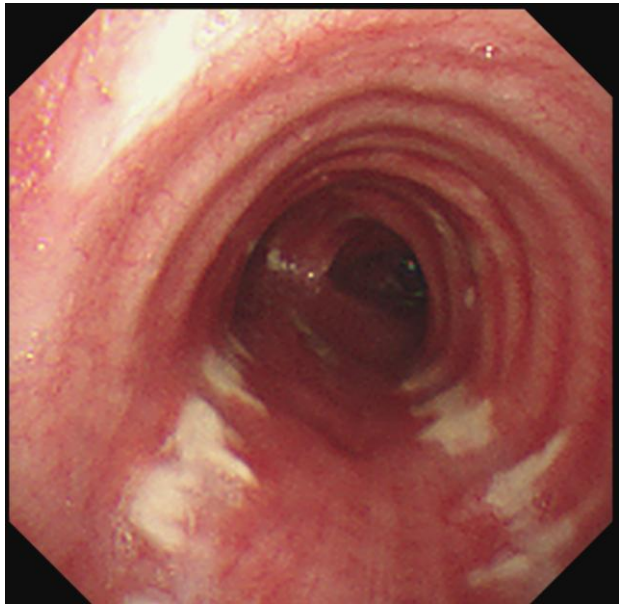
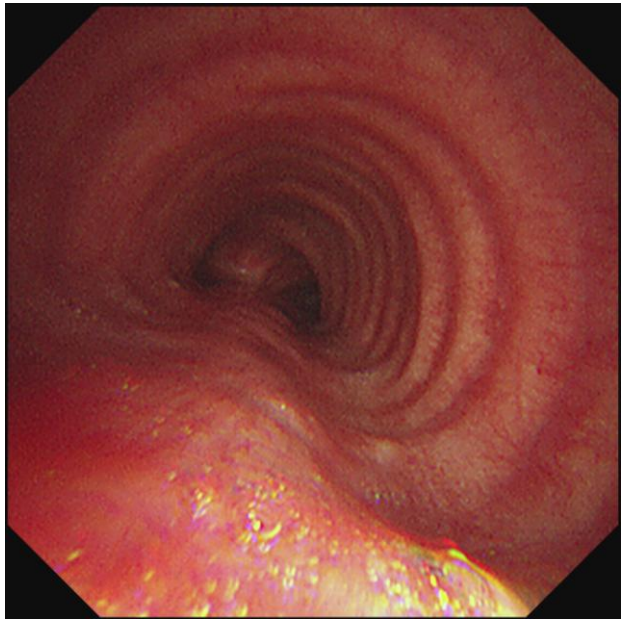


Voriconazole

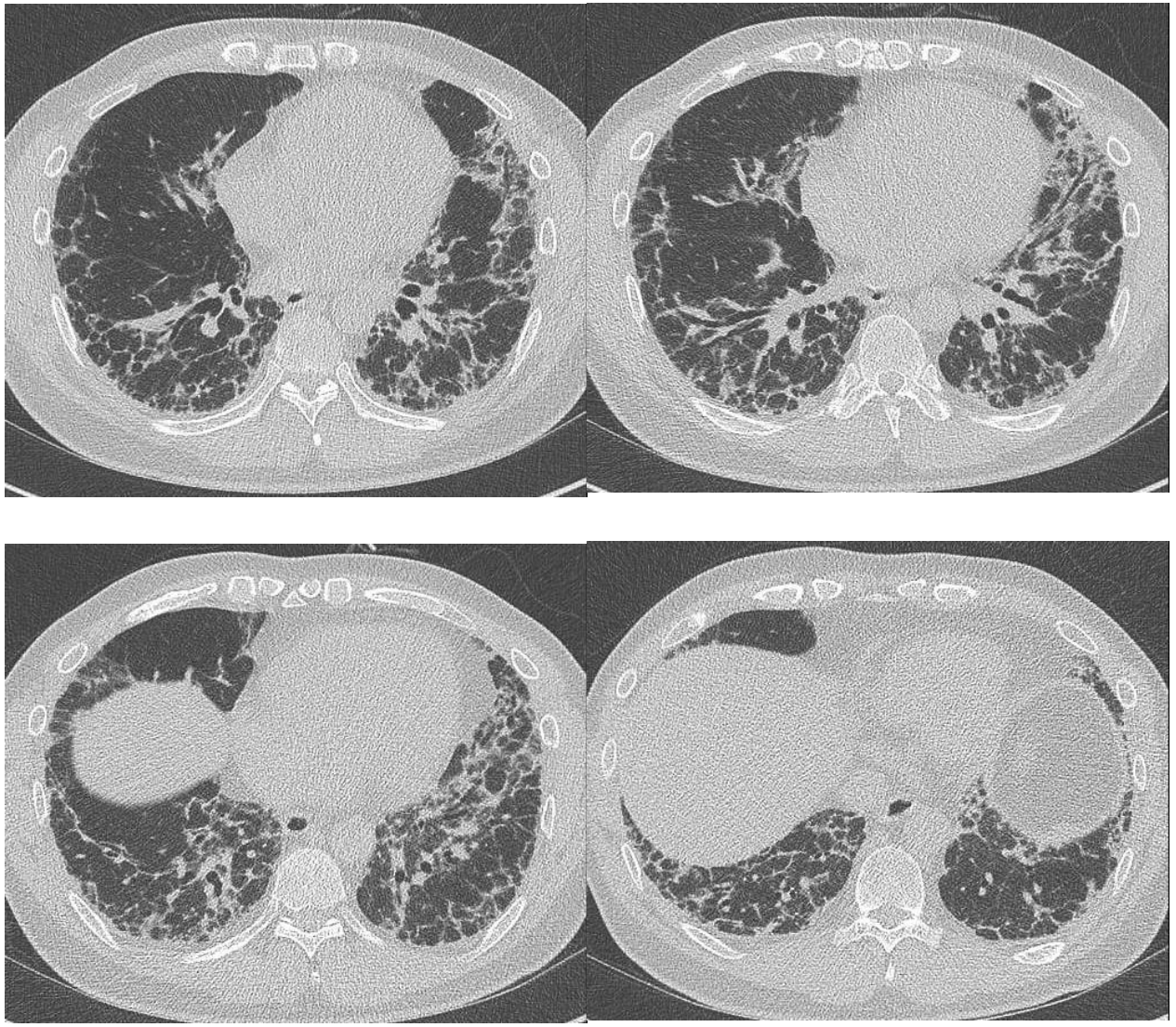


2024-08-02

Rituxmab
Cyclosporine
PD 2.5mg qd



2025-01-21



2024-06-12

2025-11-10

62/M

35PY exsmoker (4개월 전 금연)

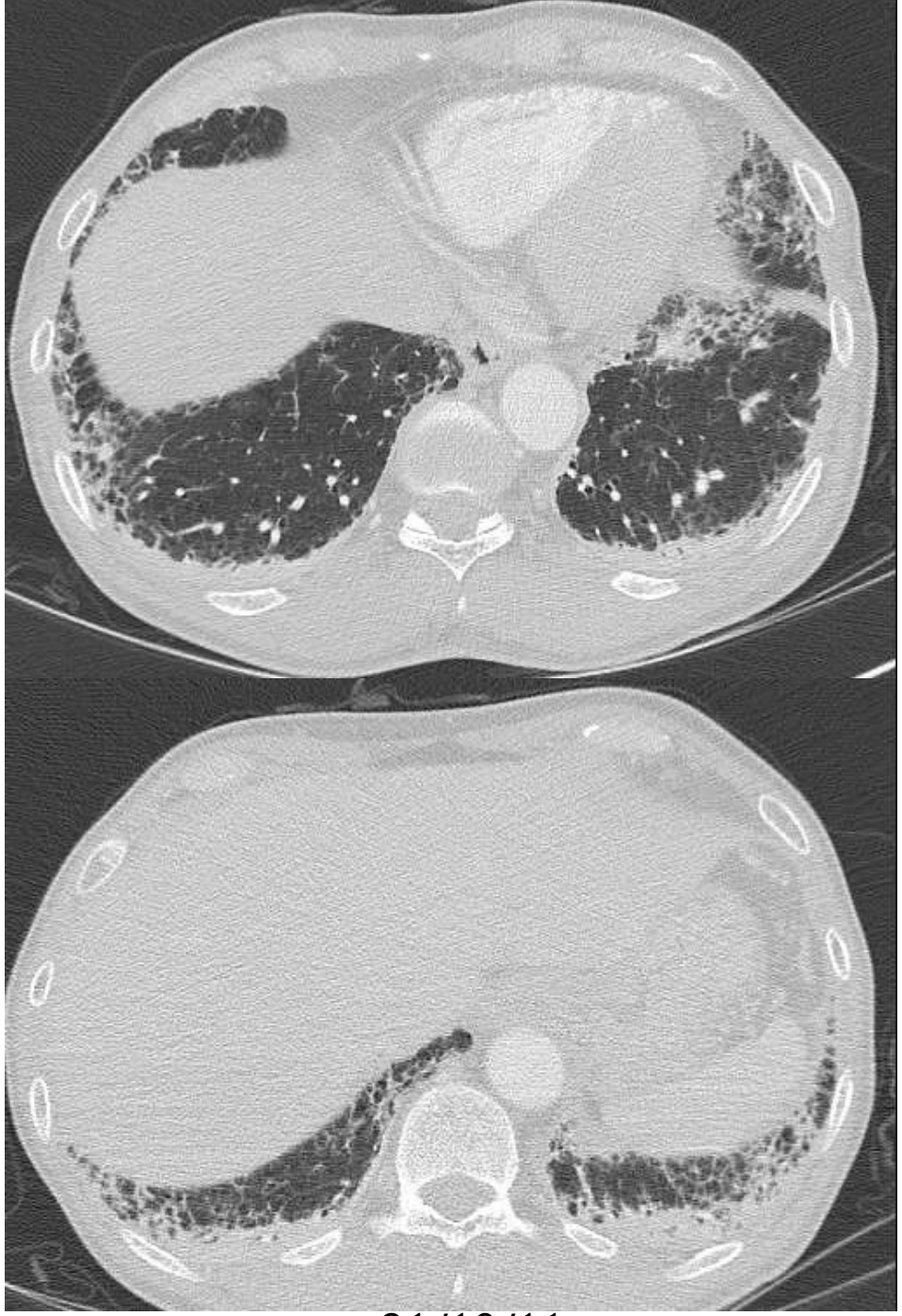
5년 전 RA 진단, 당시 DOE mMRC 1 있어 호흡기내과 consult

Opinion) RA-ILD 에 합당합니다. 우선 귀과적 치료하시면서 호흡기

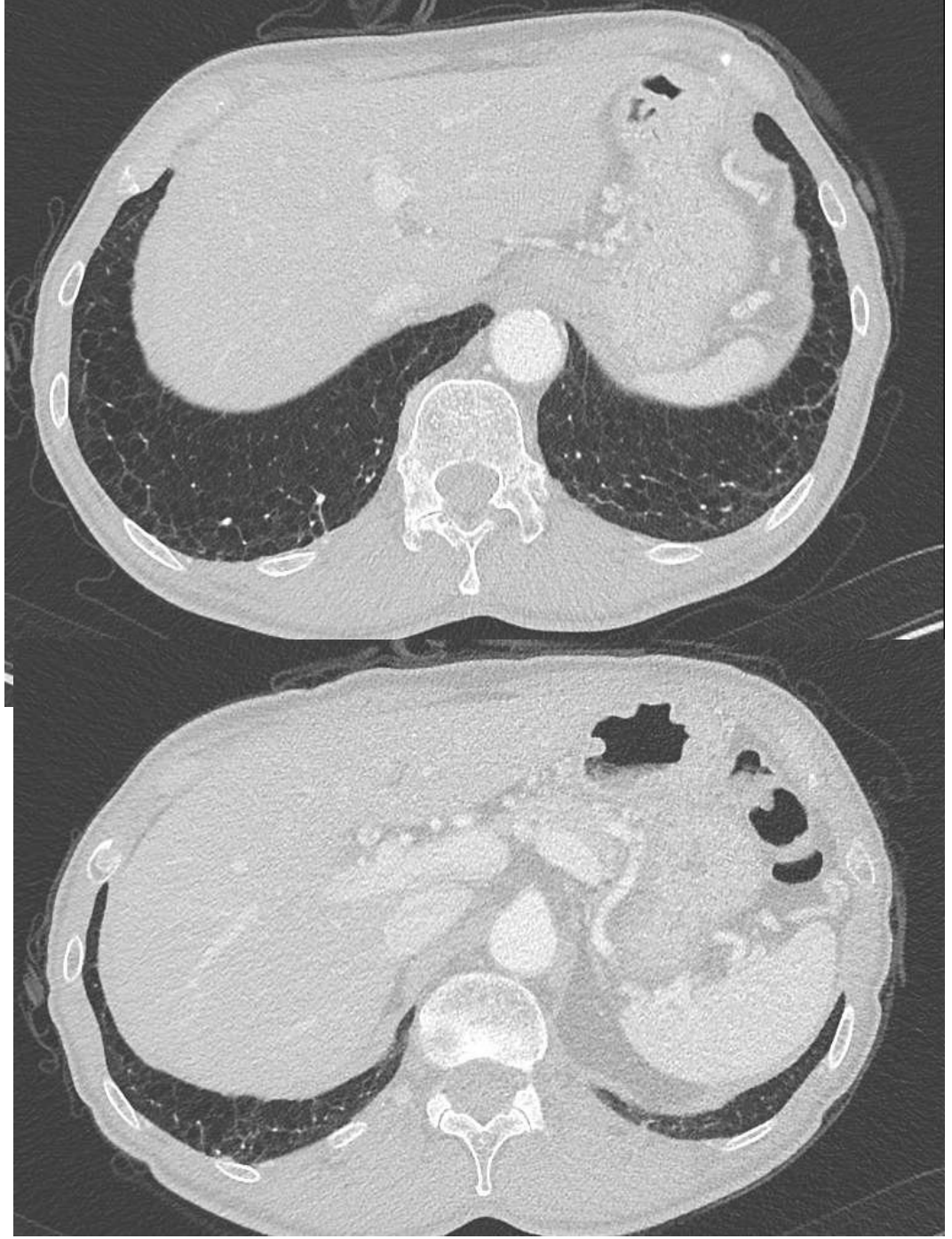
OPD FU 시켜주십시오.

⇒ 이후 호흡기는 오지 않고 류마티스내과에서 PD+ MTX 사용하다가

현재는 MTX 만 사용 중



21/10/11



26/05/14

Summary

- 2025년 ERS/ATS 에서는 Interstitial pneumonia 의 새로운 classification을 제안하였다.
- Progressive Pulmonary Fibrosis(PPF) 는 지난 1년 동안 증상, 폐기능, 영상의학적 소견 중 두 가지가 악화 시 진단할 수 있다.
- Hypersensitivity pneumonitis, CTD-ILD, unclassifiable ILD 등이 PPF 로 진행하는 경우가 많다.
- 악화되는 CTD-ILD는 면역억제제를 조절할 지 PPF 로 생각하고 항섬유화제를 추가할 지에 대해 개별적인 판단이 필요하다.

울산대학교병원
ULSAN UNIVERSITY HOSPITAL

감사합니다.

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| 울산대학교병원 | 호흡기내과 | 제갈양진