Respiratory Review of 2025
2025-04-12

Respiratory Review of 2025: Interstitial Lung Disease

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- Deep learning usage in imaging
- Precision medicine
- Pulmonary hypertension in ILD

Deep learning usage in imaging

ORIGINAL ARTICLE

Deep Learning–based Segmentation of Computed Tomography Scans Predicts Disease Progression and Mortality in Idiopathic Pulmonary Fibrosis

Muhunthan Thillai[1,2], Justin M. Oldham[3], Alessandro Ruggiero[1,2], Fahdi Kanavati[2], Tom McLellan[1], Gauri Saini[5], Simon R. Johnson[5], Francois-Xavier Ble[6], Adnan Azim[6], Kristoffer Ostridge[7,9], Adam Platt[7], Maria Belvisi[4,8], Toby M. Maher[4,10], and Philip L. Molyneaux[4,11]

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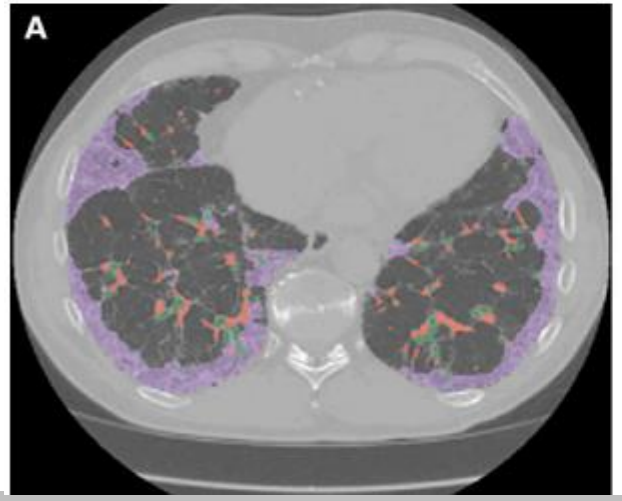
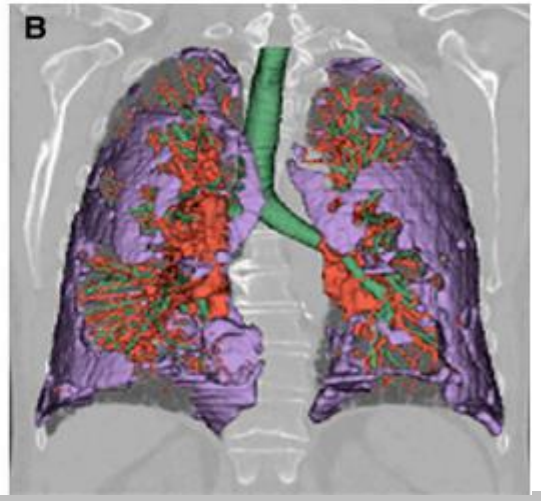
Am J Respir Crit Care Med . 2024 Aug 15;210(4):465-472

Objectives and Methods

- To develop automated imaging biomarkers using deep learning–based segmentation of CT scans→ lung function, prognosis markers for disease progression and mortality in IPF

- Methods
  - Segmentation processes for four anatomical biomarkers





1) Lung volume : open source
2) Airway volume: trained by radiologist
3) Vascular volume: classical methods
4) Fibrosis volume: trained by radiologist

Methods

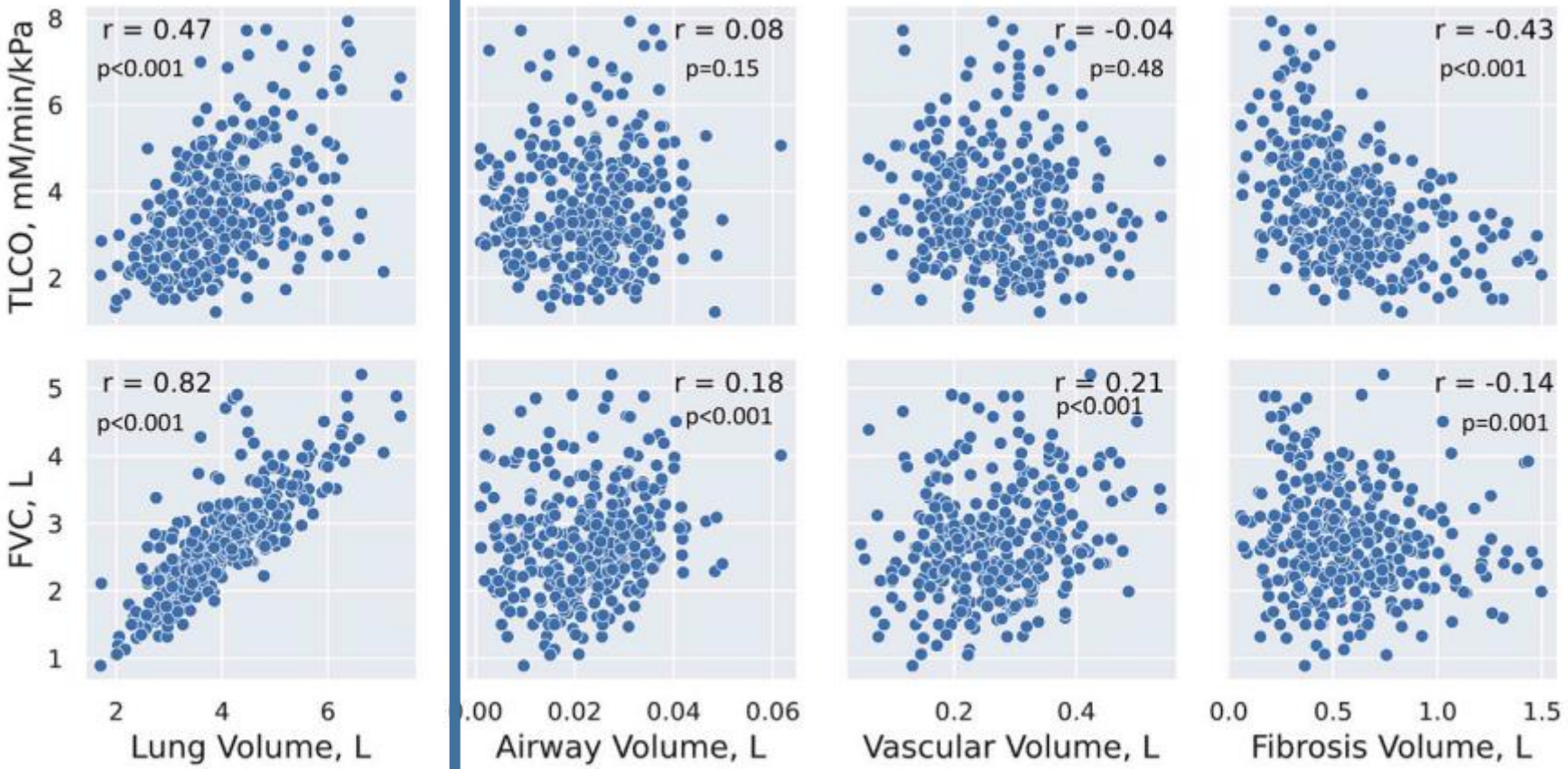
- PROFILE study : treatment –naïve patients
  - **P**rospective **O**bservation of **F**ibrosis in the **L**ung Clinical **Endpoints** study, UK
- Follow up : baseline 1, 3, 6, 12 month an then annual check until 3yr
- CT, PFT within 180 d

- PROFILE random divide to discovery / validation cohort
- Cambridge real world cohort : independent validation cohort

Table 1. Baseline Characteristics and Outcomes for the PROFILE and Cambridge Cohorts

Characteristic	PROFILE Discovery ($n = 223$)*	PROFILE Validation ($n = 223$)†	Cambridge Validation ($n = 195$)‡
Age, yr, mean (SD)	69.4 (8.2)	70.8 (8.2)	72.6 (7.7)
Male sex, *n* (%)	179 (80.3)	170 (76.2)	142 (85.0)
Ever-smoker, *n* (%)	163 (73.0)	142 (63.7)	153 (78.5)
Pulmonary function, mean (SD)			
FVC% predicted	75.5 (18.9)	79.3 (18.9)	77.2 (15.5)
D_{LCO}% predicted	43.5 (15.7)	44.7 (14.6)	50.1 (14.4)
Lung volume, L, mean (SD)	4.02 (1.13)	4.00 (1.01)	3.97 (1.12)
Airway volume, L, mean (SD)	0.02 (0.01)	0.02 (0.01)	0.07 (0.03)
Vascular volume, L, mean (SD)	0.27 (0.10)	0.25 (0.09)	0.23 (0.08)
Fibrosis volume, L, mean (SD)	0.63 (0.33)	0.54 (0.26)	0.57 (0.26)
Outcomes			
24-mo progression	116 (52.0)	101 (45.3)	NA
60-mo mortality	150 (67.3)	127 (57.0)	138 (70.8)

PFT and CT segmentations



Survival analysis (Baseline CT)

Baseline CT features and 5 –year survival (unadjusted)

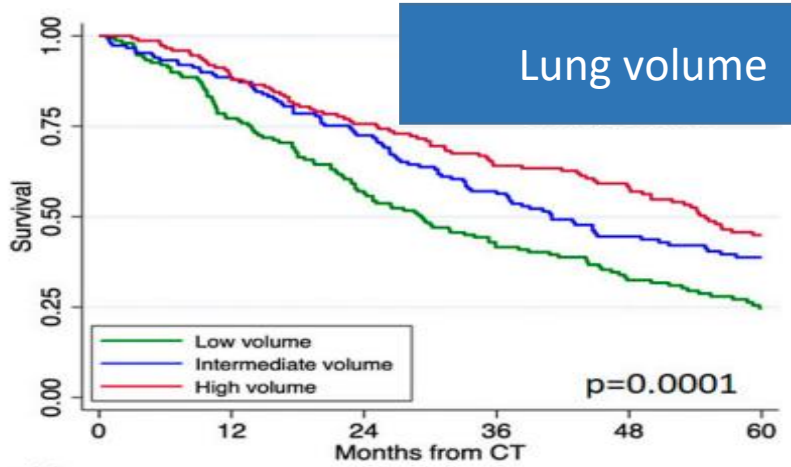
	PROFILE Discovery (*n* = 223)			PROFILE Validation (*n* = 223)			Cambridge Validation (*n* = 195)		
CT Measure	**HR**	**95% CI**	***P* Value**	**HR**	**95% CI**	***P* Value**	**HR**	**95% CI**	***P* Value**
Lung volume	0.98	0.96–0.99	0.001	0.97	0.96–0.99	0.003	0.98	0.96–0.99	0.005
Airway volume	1.90	0.46–7.82	0.374	10.09	2.06–49.35	0.004	1.82	0.96–3.45	0.068
Vascular volume	1.43	1.20–1.69	<0.001	1.31	1.09–1.59	0.005	1.23	1.01–1.50	0.042
Fibrosis volume	1.22	1.17–1.28	<0.001	1.22	1.15–1.30	<0.001	1.11	1.05–1.18	0.001

Baseline CT features and 5 –year survival (adjusted for baseline dz severity)

		Two-Year Progression-Free Survival			Five-Year Survival		
CT Measure	***n***	**HR**	**95% CI**	***P* Value**	**HR**	**95% CI**	***P* Value**
Lung volume	427	0.98	0.96–0.99	0.001	0.98	0.97–0.99	0.007
Airway volume	415	1.45	0.43–4.85	0.545	3.74	1.28–10.92	0.016
Vascular volume	426	1.30	1.12–1.51	0.001	1.37	1.20–1.57	<0.001
Fibrosis volume	427	1.17	1.12–1.22	<0.001	1.17	1.12–1.22	<0.001

KM survival and combined cohort

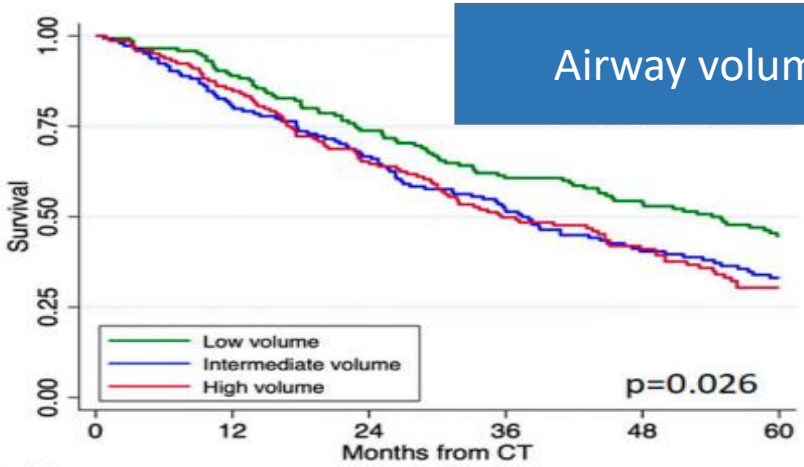
A



Lung volume

Number at risk

Low	149	115	85	61	45	30
Intermediate	149	132	108	83	56	46
High	148	131	112	94	80	58

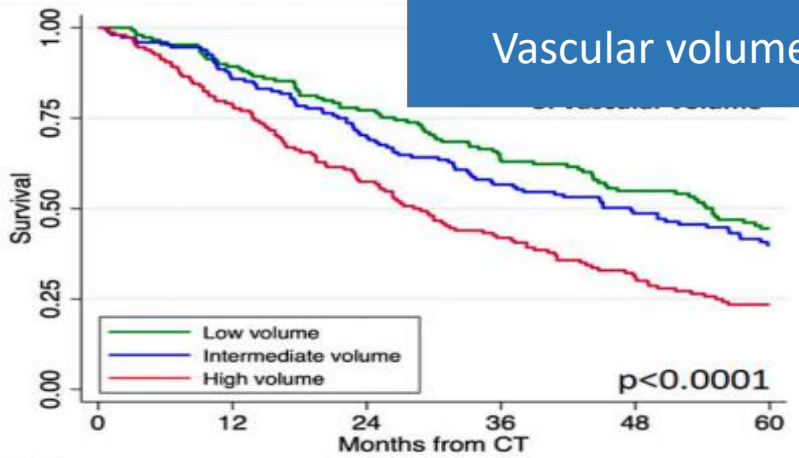


Airway volume

Number at risk

Low	145	129	107	87	75	58
Intermediate	144	116	96	74	53	40
High	144	123	94	70	48	32

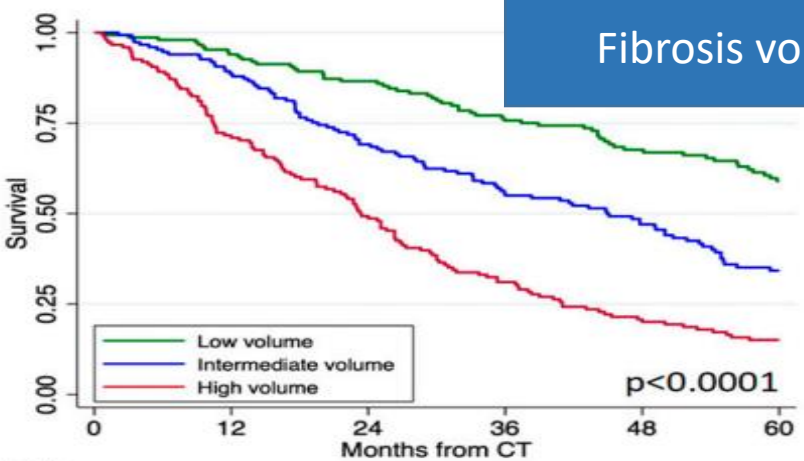
C



Vascular volume

Number at risk

Low	149	133	115	92	73	55
Intermediate	148	127	104	83	63	48
High	148	117	85	62	44	30



Fibrosis volume

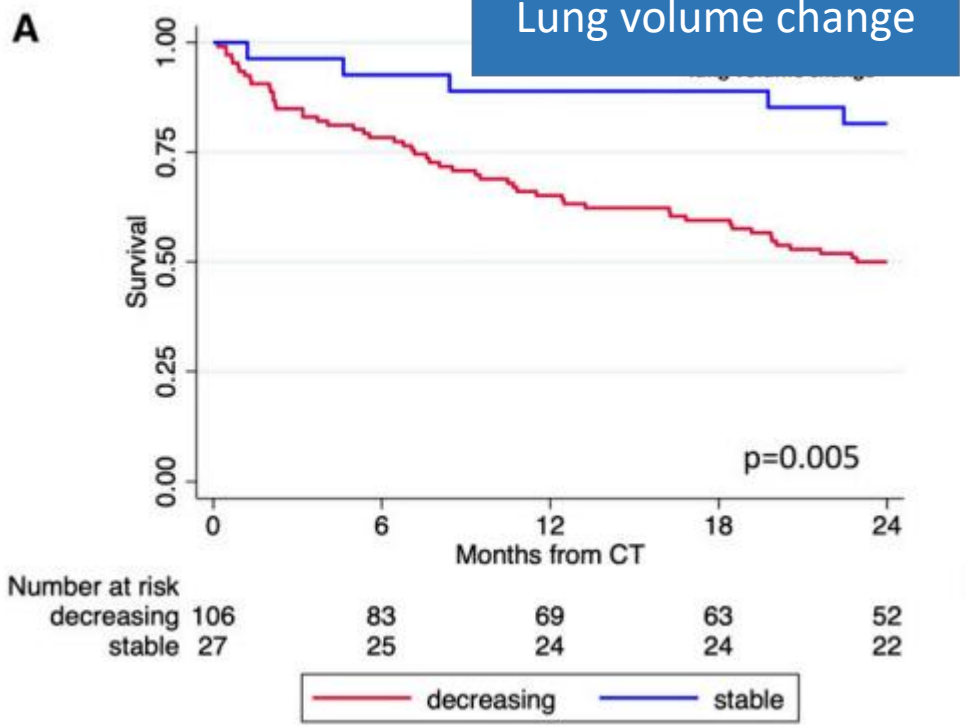
Number at risk

Low	149	140	129	110	88	73
Intermediate	149	132	103	82	63	40
High	148	106	73	46	30	21

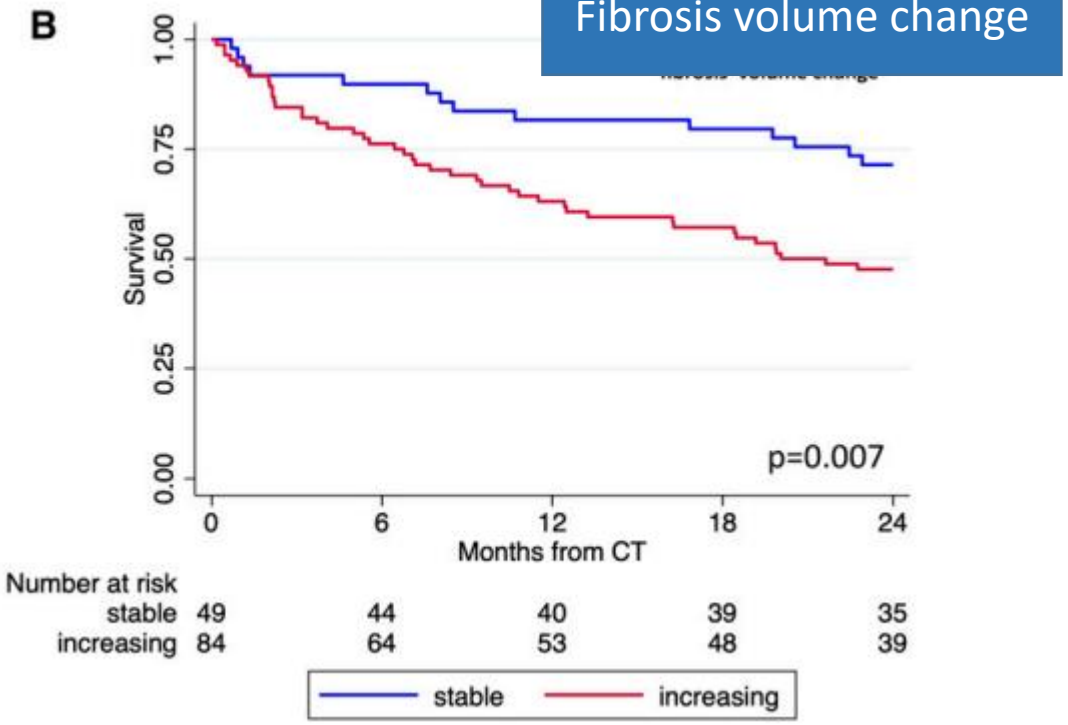
Longitudinal analysis

N=134, CT f/u 6-24m after baseline CT

Lung volume change



Fibrosis volume change



Conclusions

- Automate the segmentation and interpretation of CT scans
  - Prognostic information in clinical practice
  - Can be key endpoints or imaging biomarkers in clinical trials

[Diffuse Lung Disease Original Research]

CHEST

A Deep Learning-Based Radiomic Classifier for Usual Interstitial Pneumonia

Jonathan H. Chung, MD; Lydia Chelala, MD; Janelle Vu Pugashetti, MD; Jennifer M. Wang, MD; Ayodeji Adegunsoye, MD; Alexander W. Matyga, MS; Lauren Keith, PhD; Kai Ludwig, PhD; Sahar Zafari, PhD; Sahand Ghodrati, MD; Ahmadreza Ghasemiesfe, MD; Henry Guo, MD; Eleanor Soo, MBBS; Stephen Lyen, MBBS; Charles Sayer, MBBS; Charles Hatt, PhD; and Justin M. Oldham, MD

Objectives

- Does a deep learning (DL)-based classifier for UIP derived using CT scan features accurately discriminate radiologist-determined visual UIP ?

CHEST 2024; 165(2):371-380

Data sets and model training

TABLE 1] Data Sets Comprising Training, Validation, Performance, and Interstitial Lung Disease Clinical Cohorts (N = 3,155)

Source	Type	Training	Validation	Performance	Clinical
Lung Tissue Research Consortium	Publicly funded study	1,408	301	328	0
Chronic Obstructive Pulmonary Disease Genetic Epidemiology	Publicly funded study	176	34	50	0
Proprietary idiopathic pulmonary fibrosis clinical trial	Private clinical drug trial	350	73	81	0
Proprietary systemic sclerosis clinical trial	Private clinical drug trial	0	0	20	0
Segmed Insights	Private medical image repository	0	0	60	0
Medical Imaging Data Resource Center's RSNA International COVID-19 Open Radiology Database	Publicly funded study	0	0	16	0
National Lung Screening Trial (NSLT)	Publicly funded study	0	0	10	0
University of Chicago	Site-funded patient registry	0	0	0	160
University of California at Davis	Site-funded patient registry	0	0	0	85
Total		1,934	408	565	245

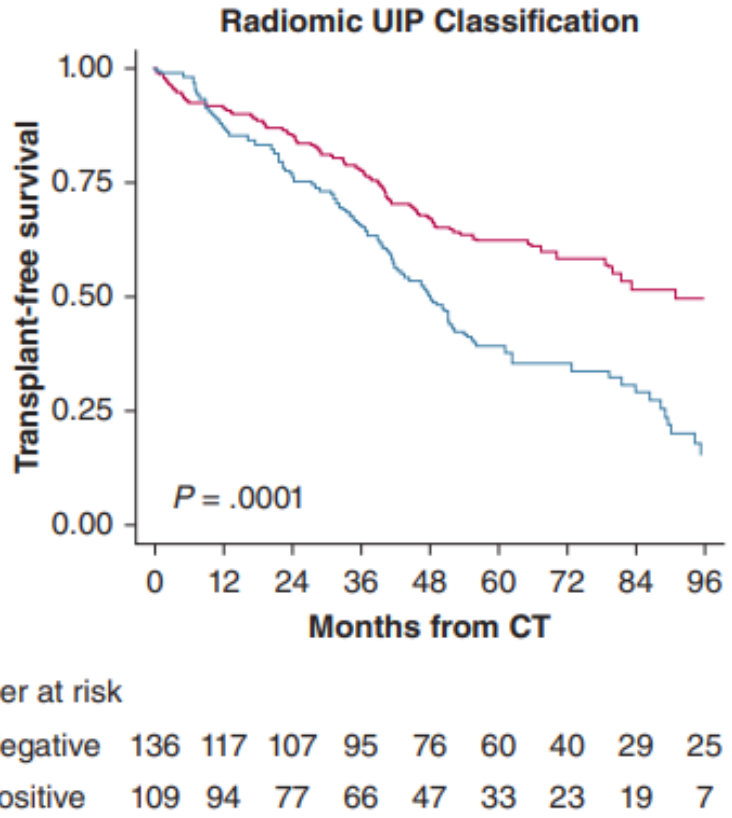
CNN (Wide Residual Network) based Wide Residual Networks
Learn about ILD patterns
Visual UIP: two radiologist

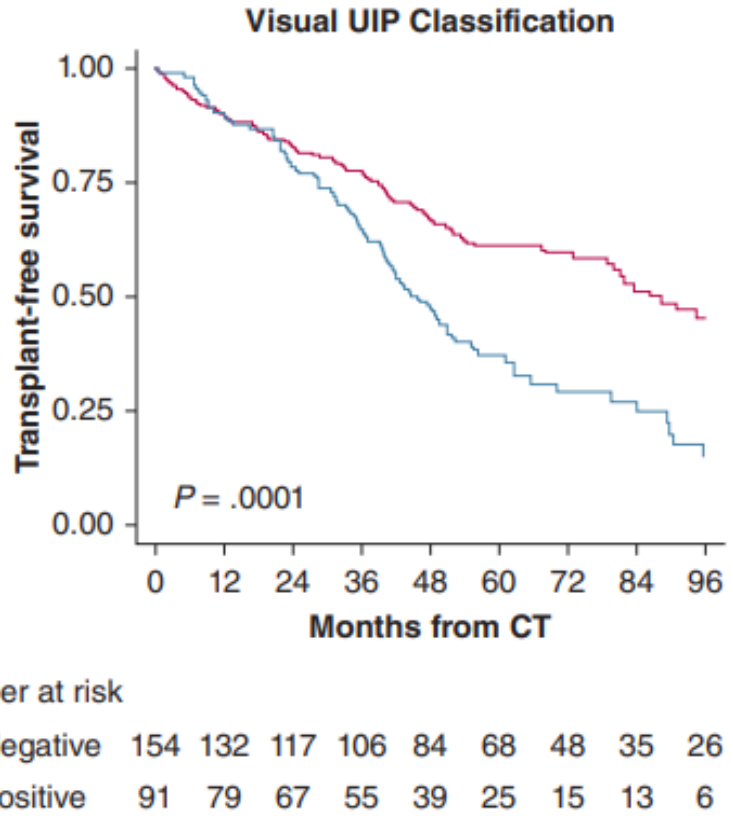
Test performance characteristics

TABLE 2 Test Performance Characteristics for a DL-Based UIP Classifier When Applied to an Independent Performance Cohort and Multicenter ILD Cohort

Classification and Test Performance Characteristic	Performance Cohort (n = 565)		ILD Clinical Cohort (n = 245)	
	Visual UIP Positive	Visual UIP Negative	Visual UIP Positive	Visual UIP Negative
DL UIP positive	65	68	74	35
DL UIP negative	5	427	17	119
Sensitivity	0.93		0.81	
Specificity	0.86		0.77	
PPV	0.49		0.68	
NPV	0.99		0.88	

Survival analysis





DL-based UIP: HR 1.69 (95% CI: 1.14–2.50)

Visual UIP: HR 1.68 (95% CI: 1.11–2.55)

Conclusions

- A DL-based classifier for UIP demonstrated good test performance across a wide range of UIP prevalence and similarly discriminated survival when compared with radiologist-determined UIP.
- This automated tool could efficiently screen for UIP in patients undergoing chest CT scan and identify a high-risk phenotype among those with known ILD

Precision medicine

CHEST

The Dawn of Precision Medicine in Fibrotic Interstitial Lung Disease

[1] Theodoros Karampitsakos; Bochra Tourki; and Jose D. Herazo-Maya, MD

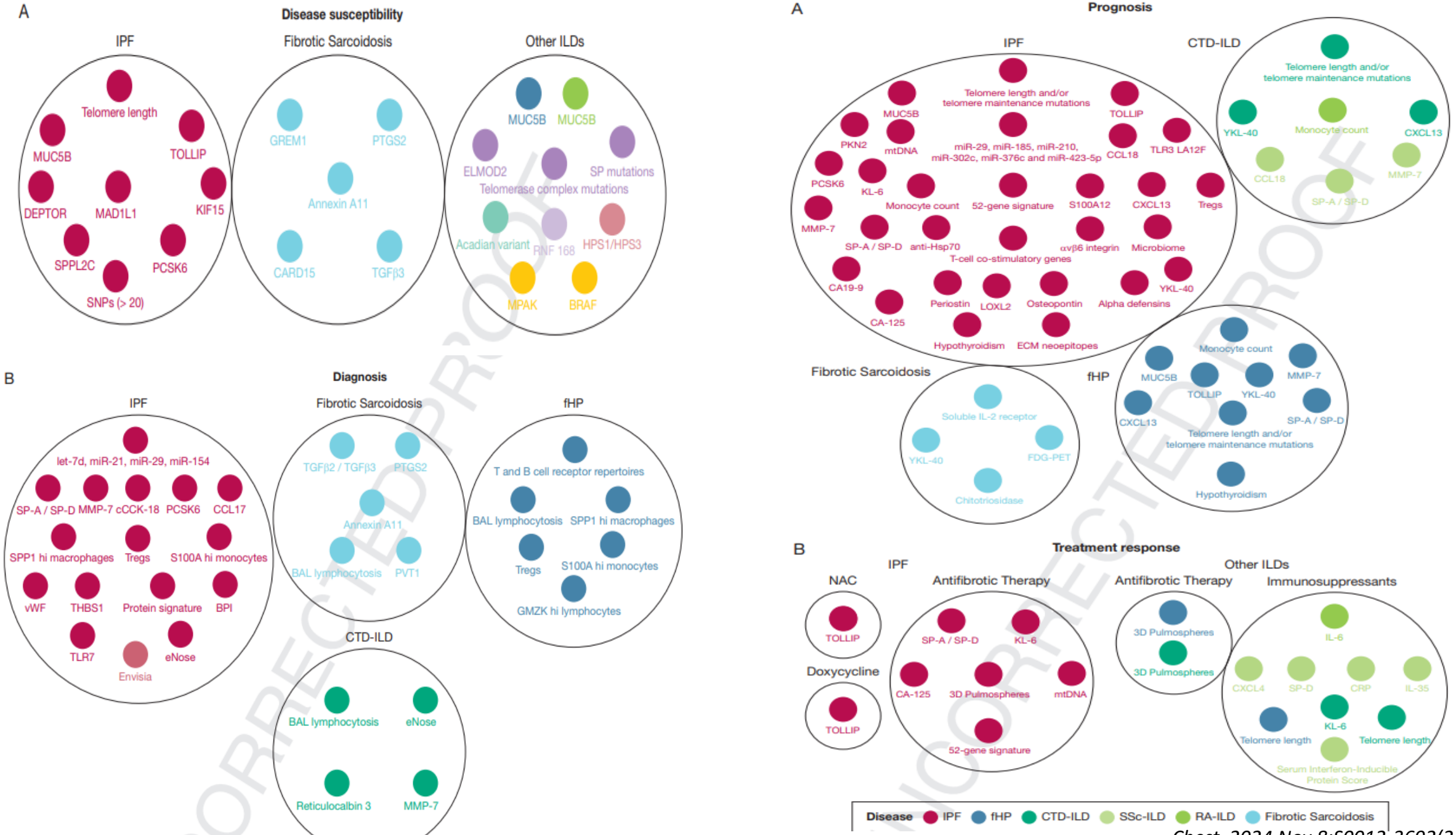
STATE OF THE ART

Genetics and Genomics of Pulmonary Fibrosis

Charting the Molecular Landscape and Shaping Precision Medicine

Ayodeji Adegunsoye[1,2*], Jonathan A. Kropski[3,4,5*], Juergen Behr[6,7], Timothy S. Blackwell[3,4,5], Tamera J. Corte[8,9,10], Vincent Cottin[11,12], Allan R. Glanville[13], Marilyn K. Glassberg[14], Matthias Griese[15], Gary M. Hunninghake[16,17], Kerri A. Johannson[18], Michael P. Keane[19], John S. Kim[20], Martin Kolb[22], Toby M. Maher[23,24], Justin M. Oldham[25], Anna J. Podolanczuk[26], Ivan O. Rosas[27], Fernando J. Martinez[28‡], Imre Noth[21*], and David A. Schwartz[29*]

Chest 2024 Nov 8:S0012-3692(24)05452-7.
Am J Respir Crit Care Med. 2024 Aug 15;210(4):401-423.



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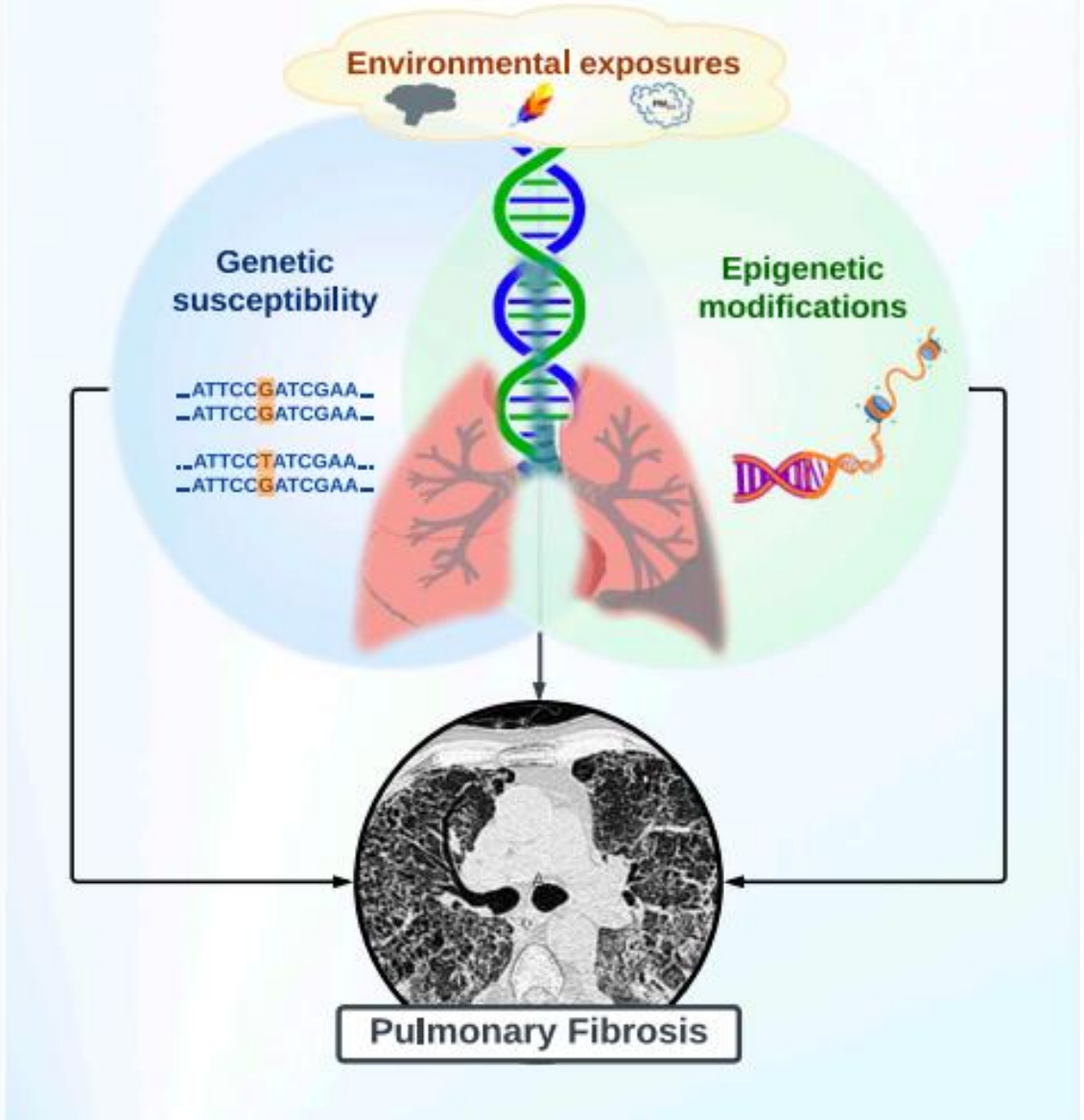

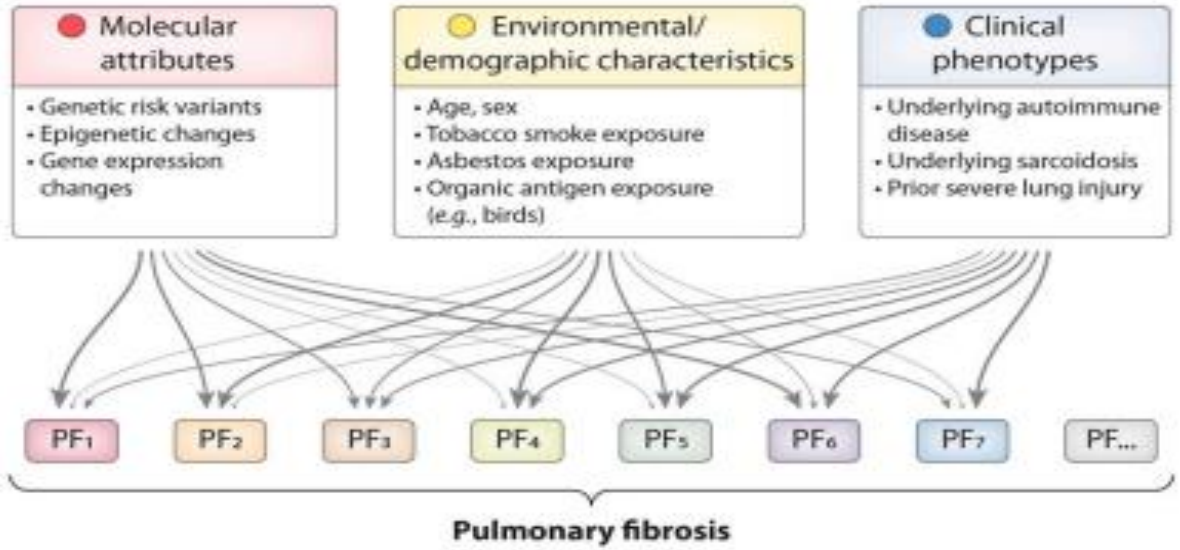

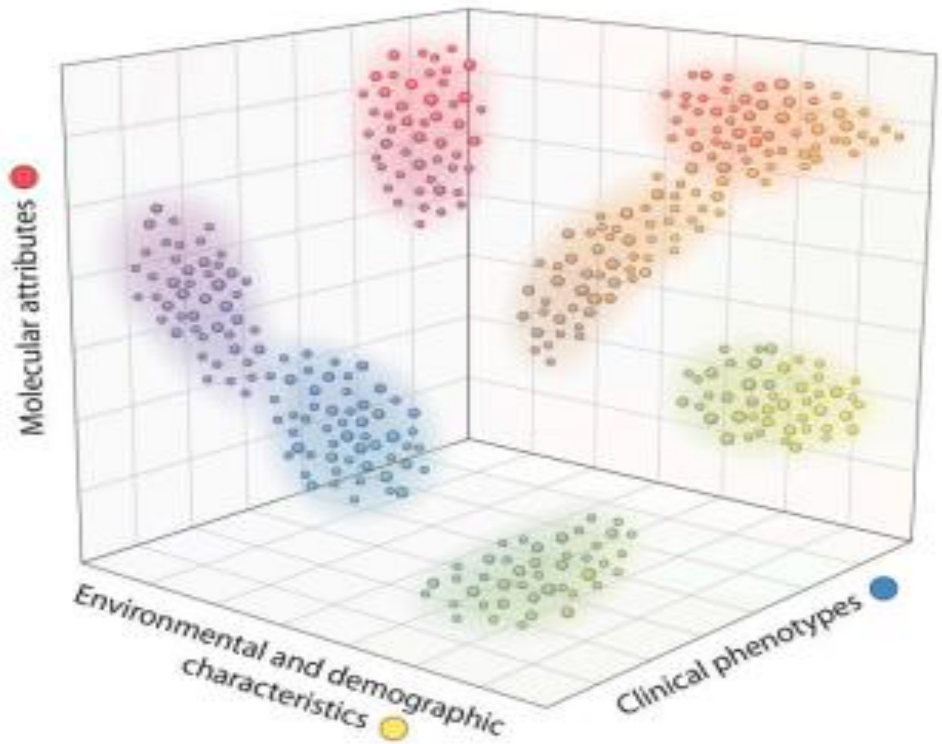

Am J Respir Crit Care Med. 2024 Aug 15;210(4):401-423.

Cluster analysis of blood biomarkers to identify molecular patterns in pulmonary fibrosis: assessment of a multicentre, prospective, observational cohort with independent validation

Hernan P Fainberg, Yuben Moodley, Isaac Triguero, Tamera J Corte, Jannie M B Sand, Diana J Leeming, Morten A Karsdal, Athol U Wells, Elisabetta Renzoni, John Mackintosh, Dino B A Tan, Roger Li, Joanne Porte, Rebecca Braybrooke, Gauri Saini, Simon R Johnson, Louise V Wain, Philip L Molyneaux, Toby M Maher, Iain D Stewart, R Gisli Jenkins

Objectives

To classify patients with pulmonary fibrosis according to blood biomarkers to differentiate distinct disease patterns, known as endotypes.

Lancet Respir Med 2024;12: 681–92

- **Cohorts:**
  - PROFILE (n=455): incident IPF or fibrotic NSIP (UK)
  - AIPFR (n=117): prevalent IPF (Australia)
- **Process**
  - 13 plasma biomarkers representing ECM turnover, epithelial stress, and thrombosis were measured by ELISA.
  - Unsupervised consensus clustering → 3 molecular endotypes.

Cluster	Dominant Biology	Biomarkers
BM (Basement Membrane)	ECM remodeling	↑ PRO-C4, PRO-C28, C3M, C6M
EI (Epithelial Injury)	Epithelial stress	↑ MMP-7, SP-D, CYFRA211, CA19-9, CA-125
XF (Crosslinked Fibrin)	Coagulation pathway	↑ X-FIB

  - A supervised machine learning model (XGBoost) validated clusters in AIPFR.
  - Associations with mortality and FVC% decline were evaluated using Cox and mixed-effects models.

Cohort baseline characteristics

	BM cluster	EI cluster	XF cluster
PROFILE cohort (n=455)			
Number of patients	248	109	98
Age, years	71·5 (8·7)	72·7 (8·5)	74·3 (6·9)
Gender			
Men	187 (75%)	82 (75%)	79 (81%)
Women	61 (25%)	27 (25%)	19 (19%)
Diagnosis			
Idiopathic pulmonary fibrosis	210 (85%)	101 (93%)	88 (90%)
Non-specific interstitial pneumonia	38 (15%)	8 (7%)	10 (10%)
European descent	238 (96%)	107 (98%)	93 (95%)
Ever smoker	176 (71%)	70 (64%)	69 (70%)
Ever treated with pulmonary fibrosis medication	95 (38%)	46 (42%)	35 (36%)
Pulmonary function at baseline			
FVC%		75·66 (18·09)	80·34 (19·39)
FVC%/DL_{CO}%	0·78 (0·28)	0·77 (0·20)	0·82 (0·25)
FEV_1/FVC*	80·72 (7·06)	81·81 (8·56)	81·55 (7·93)
Survival			
Alive at 5 years after baseline	115 (46%)	30 (28%)	33 (34%)
Median survival (95% CI), years	4·82 (4·35–4·99)	2·49 (2·39–3·78)	3·97 (3·57–4·53)

AIPFR cohort (n=117)			
Number of patients	93	8	16
Age, years	72·8 (8·2)	71·5 (7·9)	74·2 (6·1)
Gender			
Men	69 (74%)	5 (63%)	13 (81%)
Women	24 (26%)	3 (38%)	3 (19%)
Diagnosis			
Idiopathic pulmonary fibrosis	93 (100%)	8 (100%)	16 (100%)
Ever smoker	56 (60%)	4 (50%)	9 (56%)
Pulmonary function at baseline			
FVC%	88·47 (19·66)	79·94 (19·34)	88·96 (18·03)
FVC%/DL_{CO}%	0·23 (0·07)	0·23 (0·05)	0·28 (0·23)

Mortality of pt with PF (PROFILE cohort)

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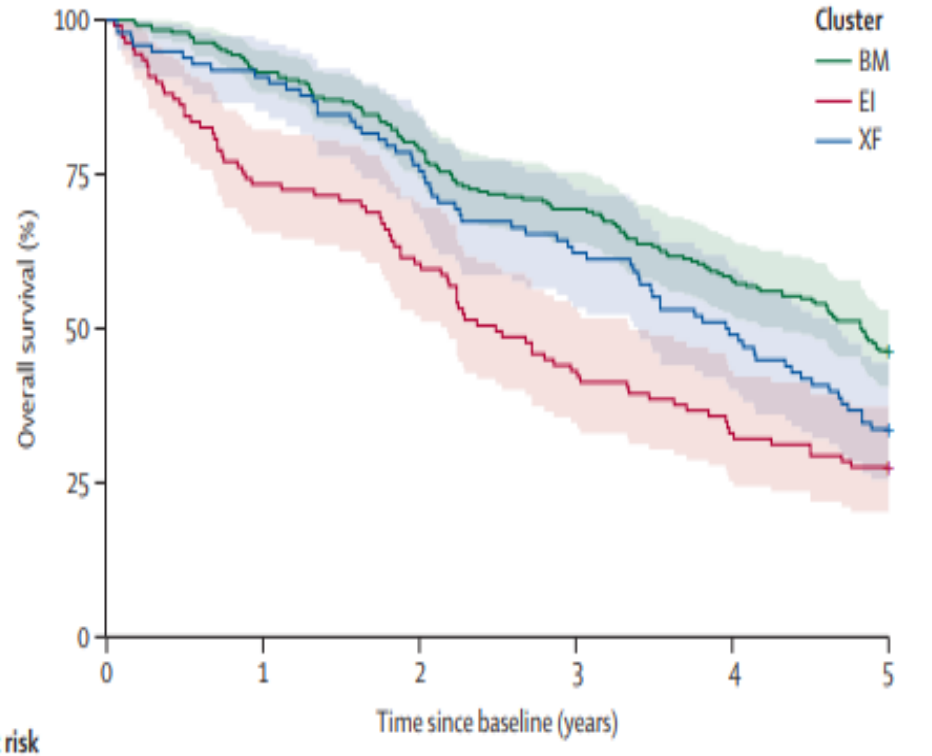


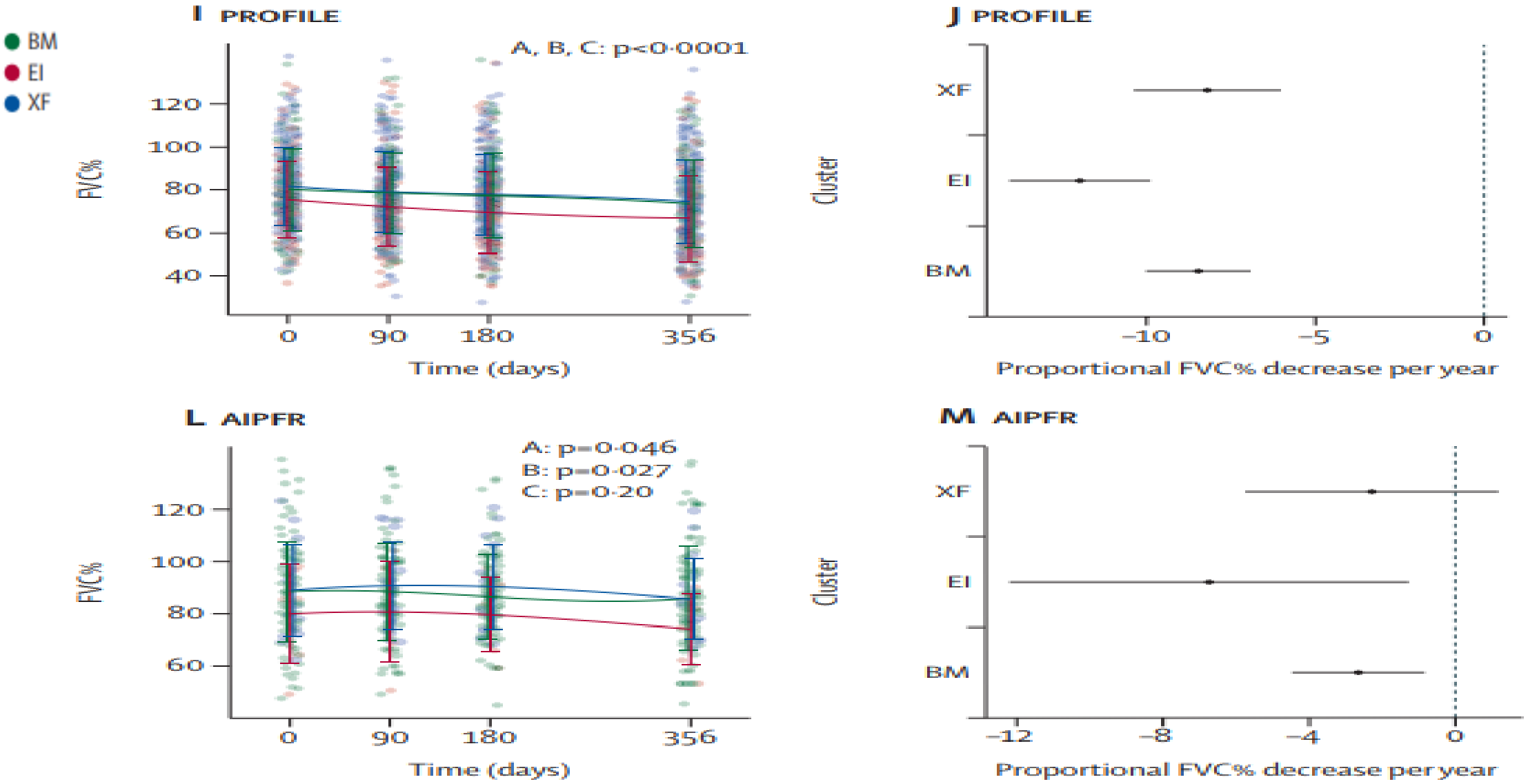
Number at risk (number censored)						
BM	248 (0)	227 (21)	196 (31)	172 (24)	144 (28)	115 (29)
EI	109 (0)	80 (29)	66 (14)	47 (19)	36 (11)	30 (6)
XF	98 (0)	89 (9)	75 (14)	61 (14)	48 (13)	33 (15)

	Adjusted HR (95% CI)	p value
Main clustering model		
BM (n=248)	1 (ref)	
EI (n=109)	1·88 (1·42–2·49)	0·0001
XF (n=98)	1·53 (1·13–2·06)	0·0058
Baseline FVC%		
Per 1% increase (n=455)	0·98 (0·97–0·98)	0·0001
Age		
Per 1-year increase (n=455)	0·97 (0·95–0·98)	0·0001
Gender		
Women (n=107)	1 (ref)	
Men (n=348)	1·20 (0·89–1·61)	0·23
Antifibrotic treatment		
No (n=334)	1 (ref)	
Yes (n=121)	1·01 (0·78–1·32)	0·92
Steriod treatment		
No (n=409)	1 (ref)	
Yes (n=46)	1·02 (0·69–1·52)	0·91

Number of events: 277; global p value (log-rank): $5{\cdot}9 \times 10^{-20}$

Akaike information criterion: 3079·58; concordance index: 0·68

Relative changes in FVC per 1 year (PROFILE and AIPFR)



Conclusions

- Blood biomarker clustering in pulmonary fibrosis identified three distinct blood biomarker signatures associated with lung function and prognosis, suggesting unique pulmonary fibrosis biomarker patterns.

- These findings support the presence of pulmonary fibrosis endotypes with the potential to guide targeted therapy development.

ORIGINAL ARTICLE

Molecular Endotypes of Idiopathic Pulmonary Fibrosis
A Latent Class Analysis of Two Multicenter Observational Cohorts

Manoj V. Maddali[1,2], Andrew R. Moore[1], Pratik Sinha[3,4], Chad A. Newton[5], John S. Kim[6], Ayodeji Adegunsoye[7], Shwu-Fan Ma[6], Mary E. Strek[7], Ching-Hsien Chen[8], Angela L. Linderholm[8], Rachel L. Zemans[9], Bethany B. Moore[9,10], Paul J. Wolters[12], Fernando J. Martinez[13*], Angela J. Rogers[1], Rishi Raj[1], Imre Noth[6], and Justin M. Oldham[9,11]

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Objectives

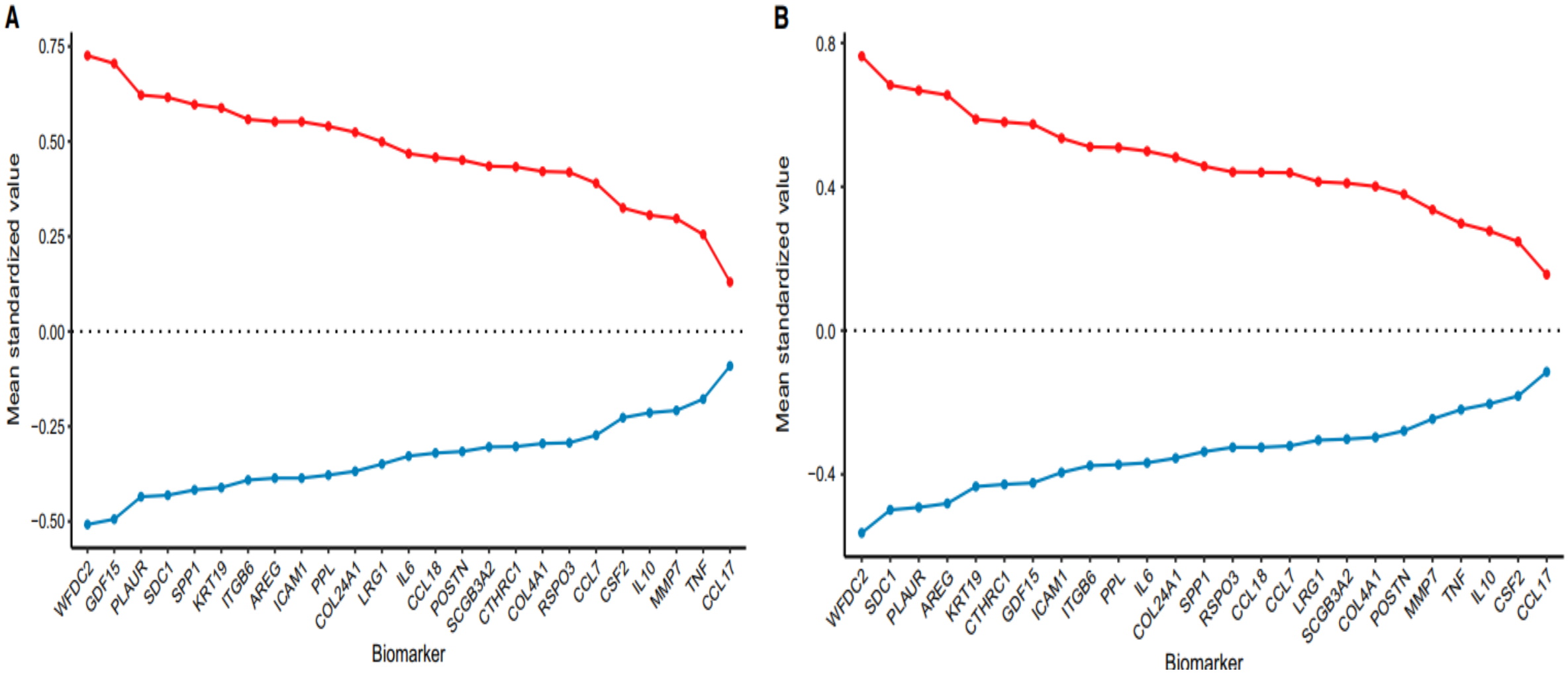
To identify and validate biologically driven **molecular endotypes** using latent class analysis (LCA), and evaluate their **prognostic and therapeutic implications.**

Am J Respir Crit Care Med . 2024 Aug 15;210(4):455-464.

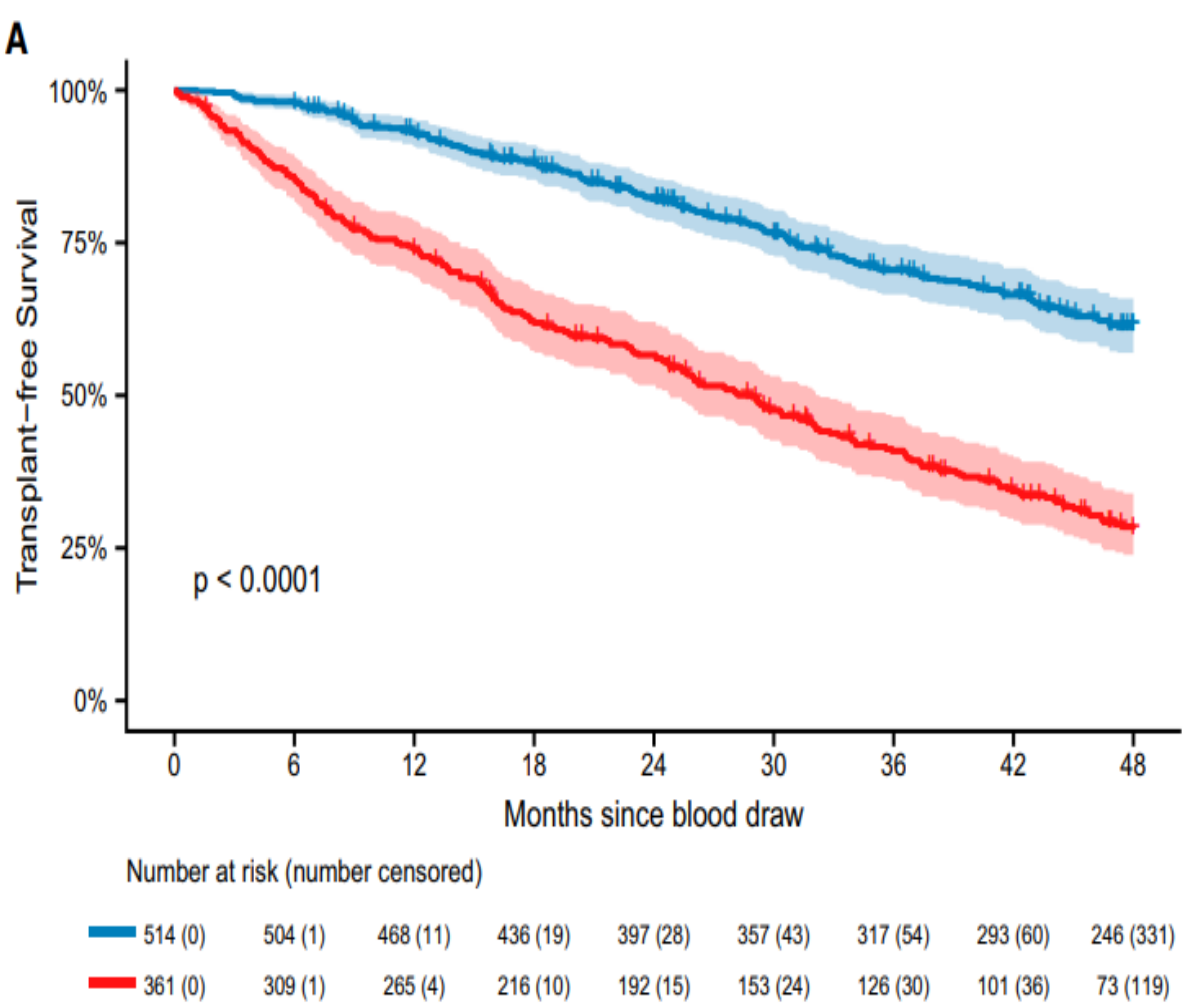
Methods

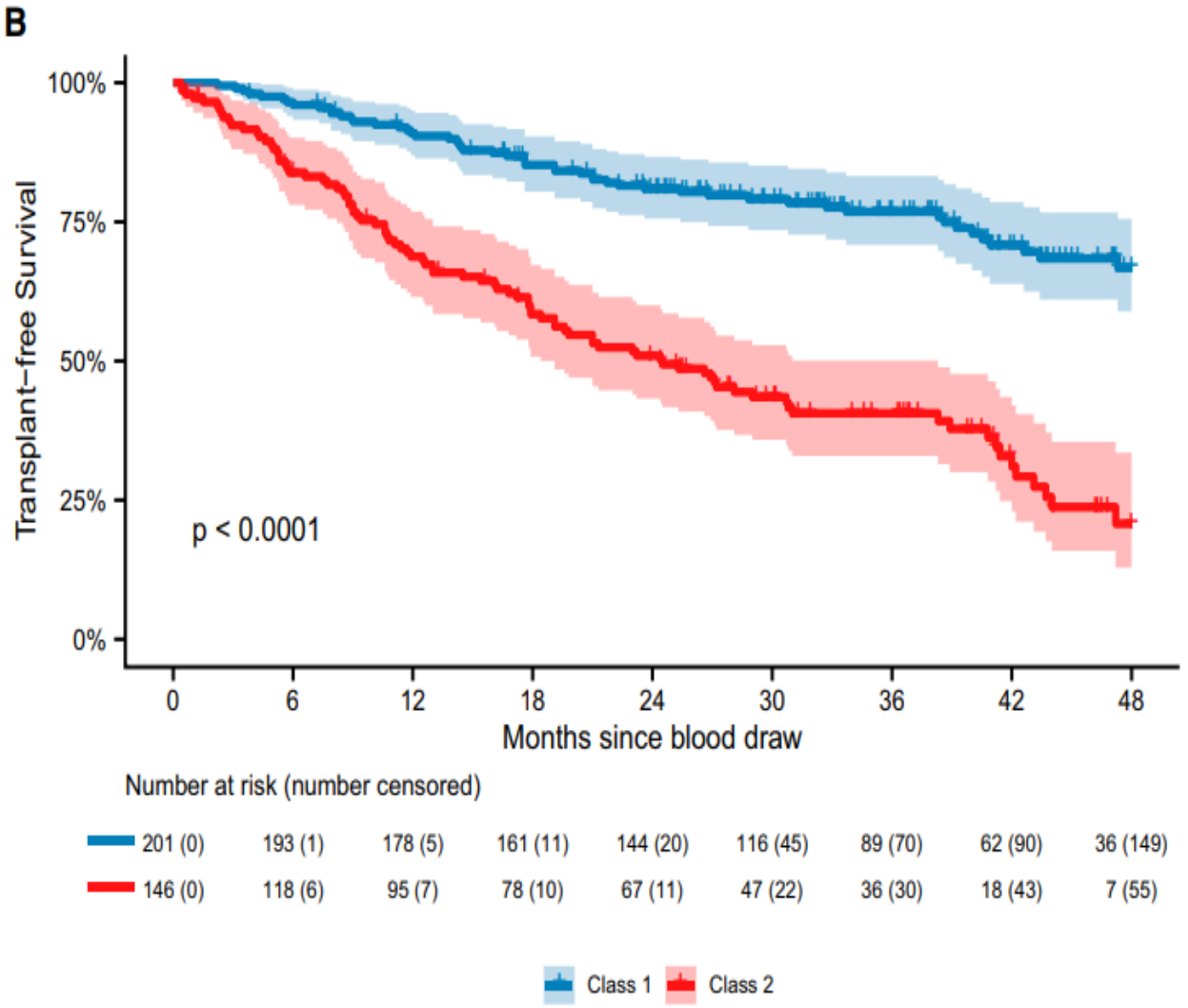
- Cohort
  - Dicovery: Pulmonary Fibrosis Foundation Patient Registry (PFF; March 2016 to June 2018) (n=875)
  - Validation: site-specific registries (n=347)
- Methods
  - LCA using 25 expert-selected plasma proteins (associated with fibrosis, ECM, epithelial injury)
  - Primary outcome: 4-year transplant-free survival (multivariable Cox)
  - Treatment analysis: Pooled cohort of antifibrotic-naive patients (n=555)

Biomarker cluster

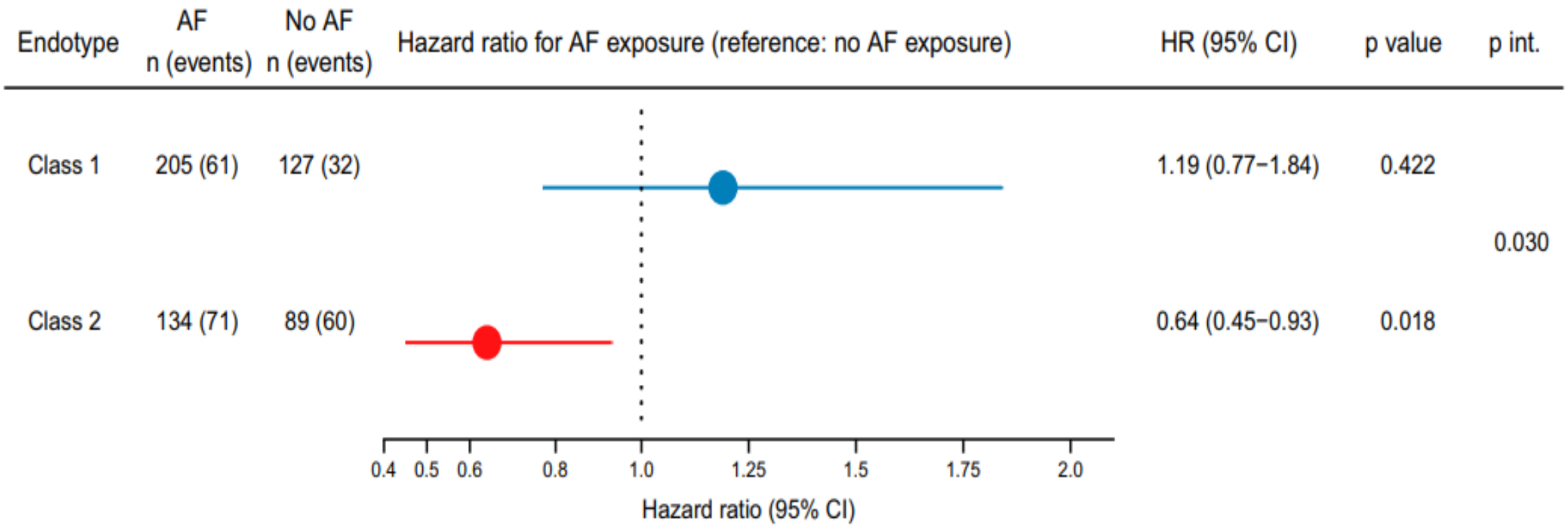


Four-year transplant-free survival curves





Differential Treatment Response



Conclusions

- Identifying two novel molecular endotypes of IPF with divergent clinical outcomes and responses to antifibrotic therapy.

- These endotypes could enable a precision medicine approach for future IPF clinical trials.

Pulmonary hypertension in ILD

Original research

Mild elevation of pulmonary vascular resistance predicts mortality regardless of mean pulmonary artery pressure in mild interstitial lung disease

Tomonori Sato [1], Taiki Furukawa [1,2], Ryo Teramachi [1,2], Jun Fukihara [3], Yasuhiko Yamano [3], Toshiki Yokoyama [3], Toshiaki Matsuda [3], Kensuke Kataoka [3], Tomoki Kimura [3], Koji Sakamoto [1], Makoto Ishii [1], Yasuhiro Kondoh [3]

Objectives

To evaluate the clinical significance of MPAP and PVR values for mortality in patients newly diagnosed with ILD.

Thorax 2024;79:422–429.

Methods

- Retrospective analysis of 854 ILD patients (Japan)
  - New diagnosed with ILD
- Undergoing right heart catheterization (RHC) between 2007–2018.
- Patients classified into six groups based on MPAP (≤20 / 20–25 / ≥25 mmHg) and PVR (≤2 / >2 Wood units).
- Cox proportional hazard models adjusted with the ILD-GAP Index.

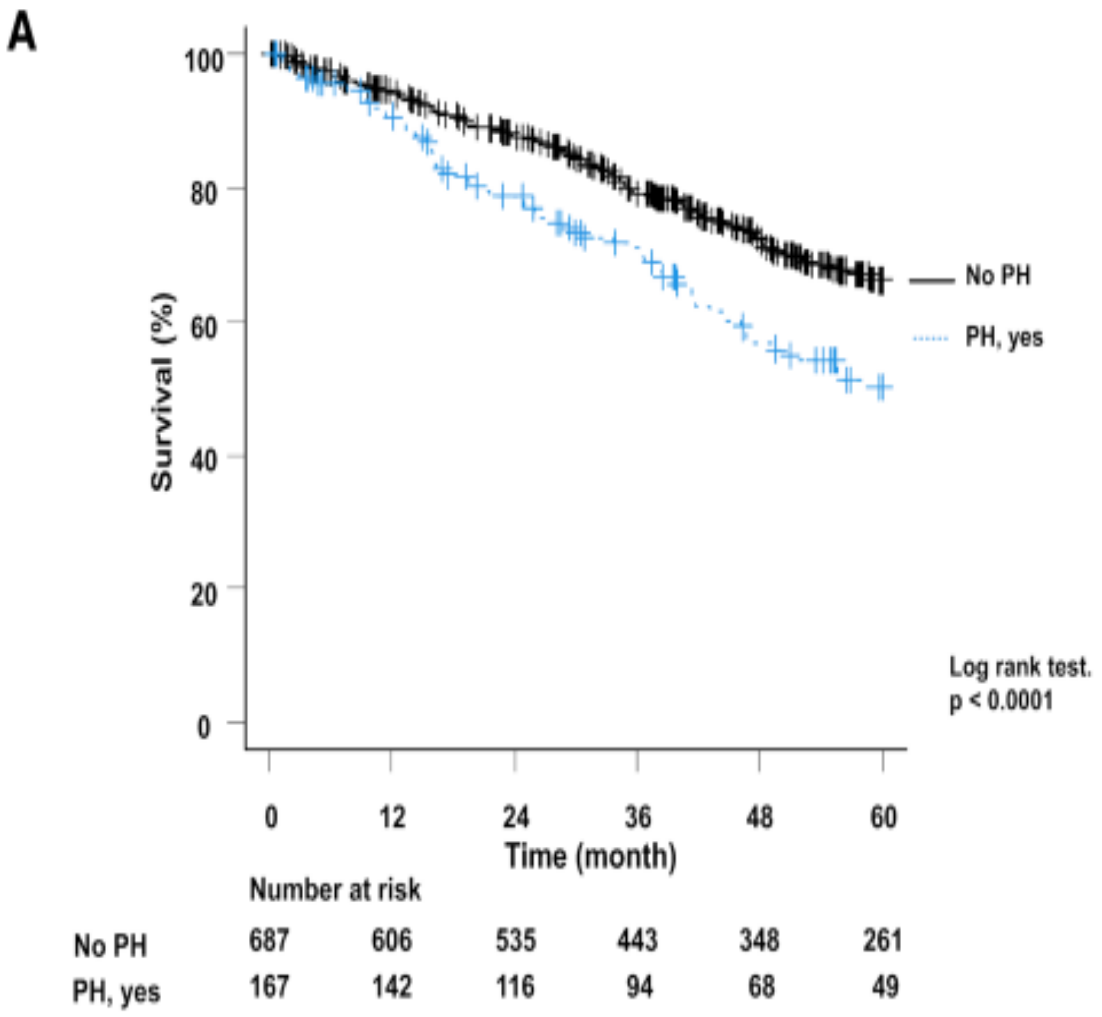
Table 1 Baseline characteristics based on haemodynamic definition of pulmonary hypertension (PH)

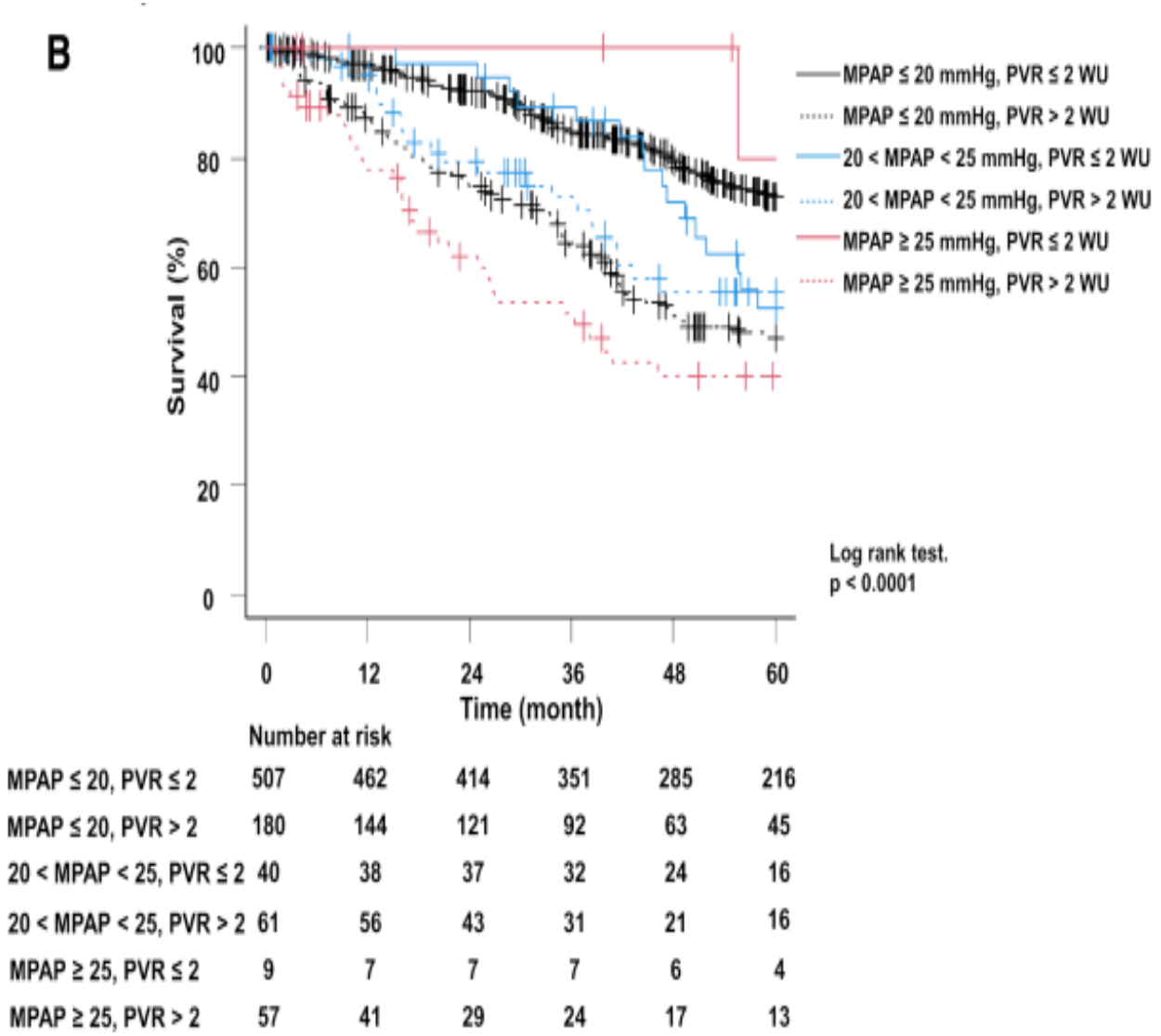
Characteristic	Total	MPAP≤20 mm Hg		20<MPAP<25 mm Hg		MPAP≥25 mm Hg	
		PVR≤2 WU	PVR>2 WU	PVR≤2 WU	PVR>2 WU	PVR≤2 WU	PVR>2 WU
PH		No	No	Yes	Yes	Yes	Yes
Subjects	854	507	180	40	61	9	57
Non-IPF in IIPs, %	234 (27.4)	161 (31.8)	44 (24.4)	9 (22.5)	10 (16.4)	2 (22.2)	8 (14.0)
CTD-ILD, %	171 (20.0)					.4)	19 (33.3)
Others, %	44 (5.2)					.1)‡‡	1 (1.8)§§
Pulmonary function test							
FVC % predicted	82.7 (68.6–97.3)¶¶					(73.8–102.2)	80.5 (55.0–93.4)
TLco % predicted	62.8 (48.9–79.5)‡‡‡					(49.7–77.5)	39.0 (33.3–52.3)††††
ILD-GAP Index‡‡‡‡							
0–1	258 (30.3)					.3)	11 (19.3)
2–3	383 (45.0)					.6)	17 (29.8)
4–5	176 (20.7)					.1)	23 (40.4)
>5	34 (4.0)						6 (10.5)
Haemodynamic parameters							
MPAP, mm Hg	16.0 (13.0–19.0)					(26.0–27.0)	29.0 (27.0–35.0)
PVR, Wood units (WU)	1.70 (1.22–2.25)					(1.27–1.79)	4.30 (3.23–5.66)
C.I., L/min/m^2	3.10 (2.74–3.54)	3.20 (2.81–3.62)	2.90 (2.57–3.20)	3.40 (2.98–3.77)	3.00 (2.62–3.33)	3.50 (3.05–4.10)	2.80 (2.47–3.42)
<2.2 L/min/m^2, %	25 (2.9)	8 (1.6)	11 (6.1)	0 (0)	3 (4.9)	0 (0)	3 (5.3)
PAWP, mm Hg	7.0 (5.0–10.0)	7.0 (5.0–9.0)	5.0 (3.0–7.0)	13.0 (12.0–14.3)	9.0 (7.0–10.0)	17.0 (14.0–19.0)	10.0 (6.0–13.0)
>15 mm Hg, %	19 (2.2)	1 (0.2)	0 (0)	6 (15.0)	1 (1.6)	6 (66.7)	5 (8.8)
RAP, mm Hg	4.0 (2.0–6.0)	4.0 (2.0–6.0)	3.0 (2.0–5.0)	9.0 (7.0–11.0)	6.0 (4.0–8.0)	9.0 (6.0–14.0)	6.0 (3.0–9.0)
SvO_2, %	72.1 (68.7–75.6)	73.2 (69.7–76.2)	70.1 (67.0–73.5)	73.0 (69.0–76.3)	71.7 (67.6–74.6)	74.2 (71.2–77.1)	68.1 (63.2–71.6)

Characteristic	Total	MPAP≤20 mm Hg		20<MPAP<25 mm Hg		MPAP≥25 mm Hg	
		PVR≤2 WU	PVR>2 WU	PVR≤2 WU	PVR>2 WU	PVR≤2 WU	PVR>2 WU
PH		No	No	Yes	Yes	Yes	Yes
Subjects	854	507	180	40	61	9	57

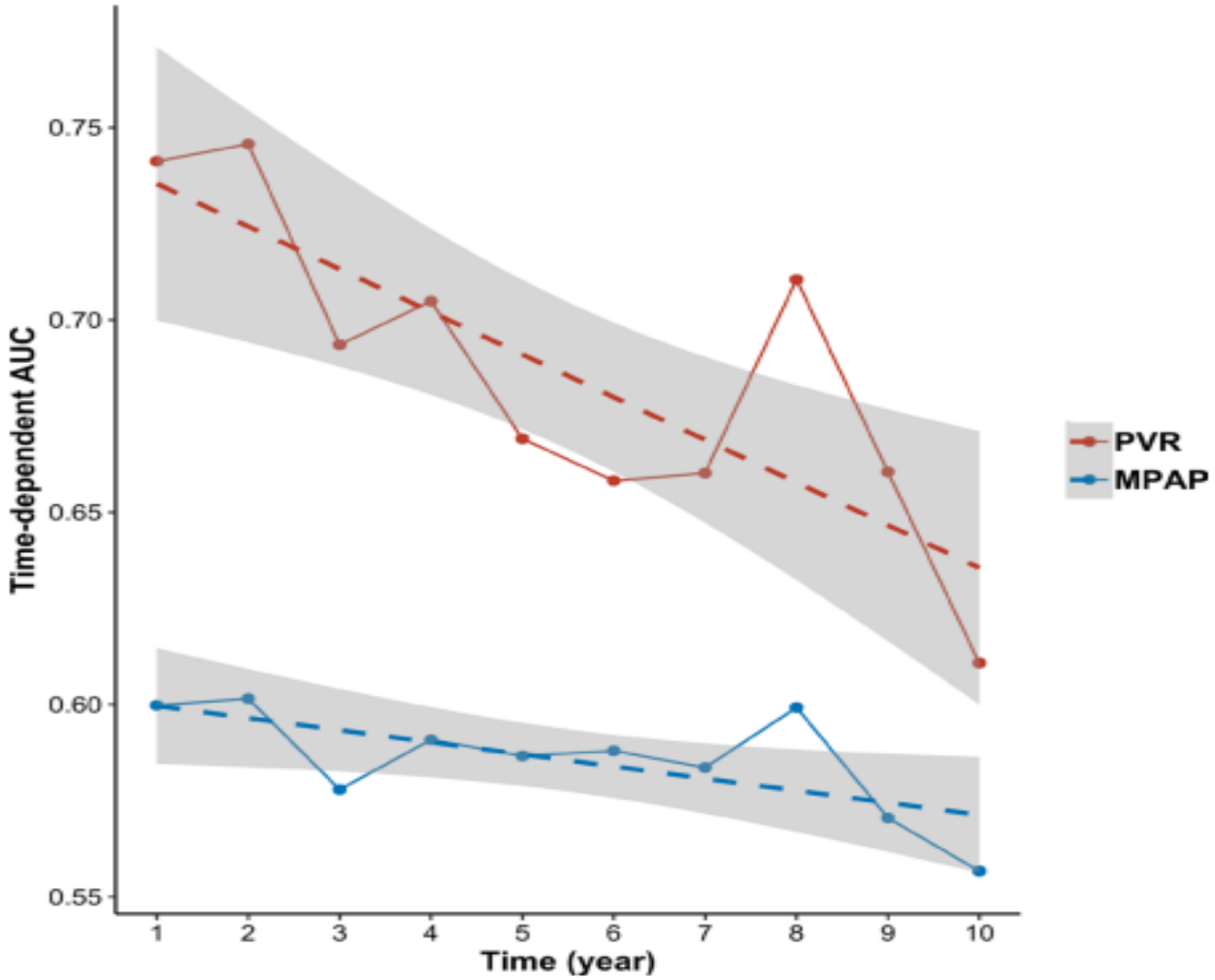
- PVR>2WU
  - Lower FVC (72.5% vs. 87.6%, p<0.0001)
  - Lower DLCO (51.4% vs 69.0%, p<0.0001)
  - Higher proportion of low CI (5.7% vs. 1.4%, P=0.0009)
  - Higher proportion of IPF (54.4% vs. 43.7% , P=0.0032)

Survival analysis





ROC curves for mortality



Conclusions

- PVR>2 WU is associated with higher mortality rate in patients with ILD, regardless of MPAP values.

- The simultaneous interpretation of PVR and MPAP is important in patients with ILD.

Original research

Survival analysis from the INCREASE study in PH-ILD: evaluating the impact of treatment crossover on overall mortality

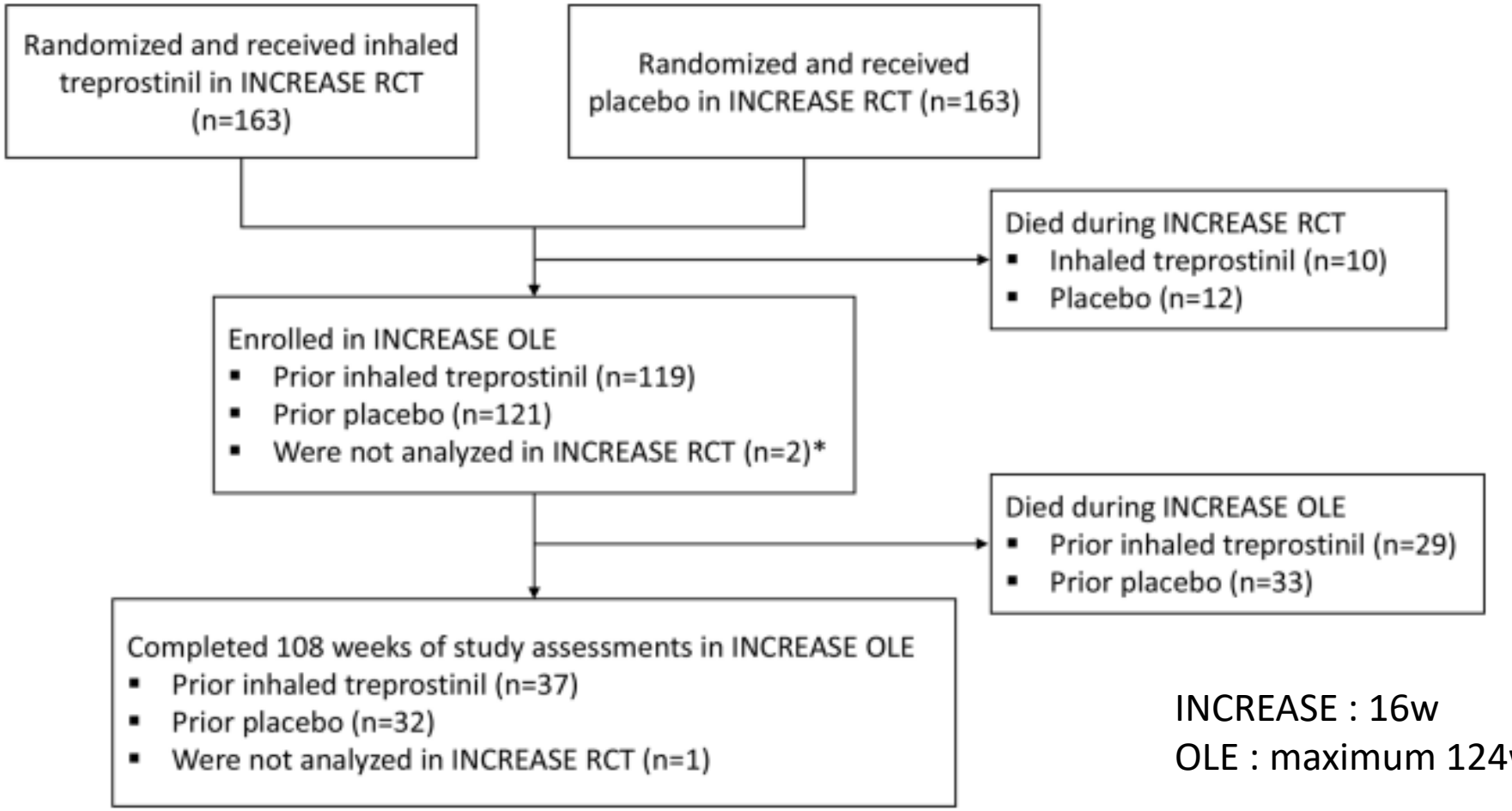
Steven D Nathan ,[1] Shilpa Johri,[2] Joanna M Joly,[3] Christopher S King,[1] Amresh Raina,[4] Colleen A McEvoy,[5] Dasom Lee,[6] Eric Shen,[6] Peter Smith,[6] Chunqin Deng,[6] Aaron B Waxman[7]

Objectives

To evaluate whether inhaled treprostinil has a long-term survival benefit in patients with pulmonary hypertension associated with interstitial lung disease (PH-ILD). (post-hoc analysis)

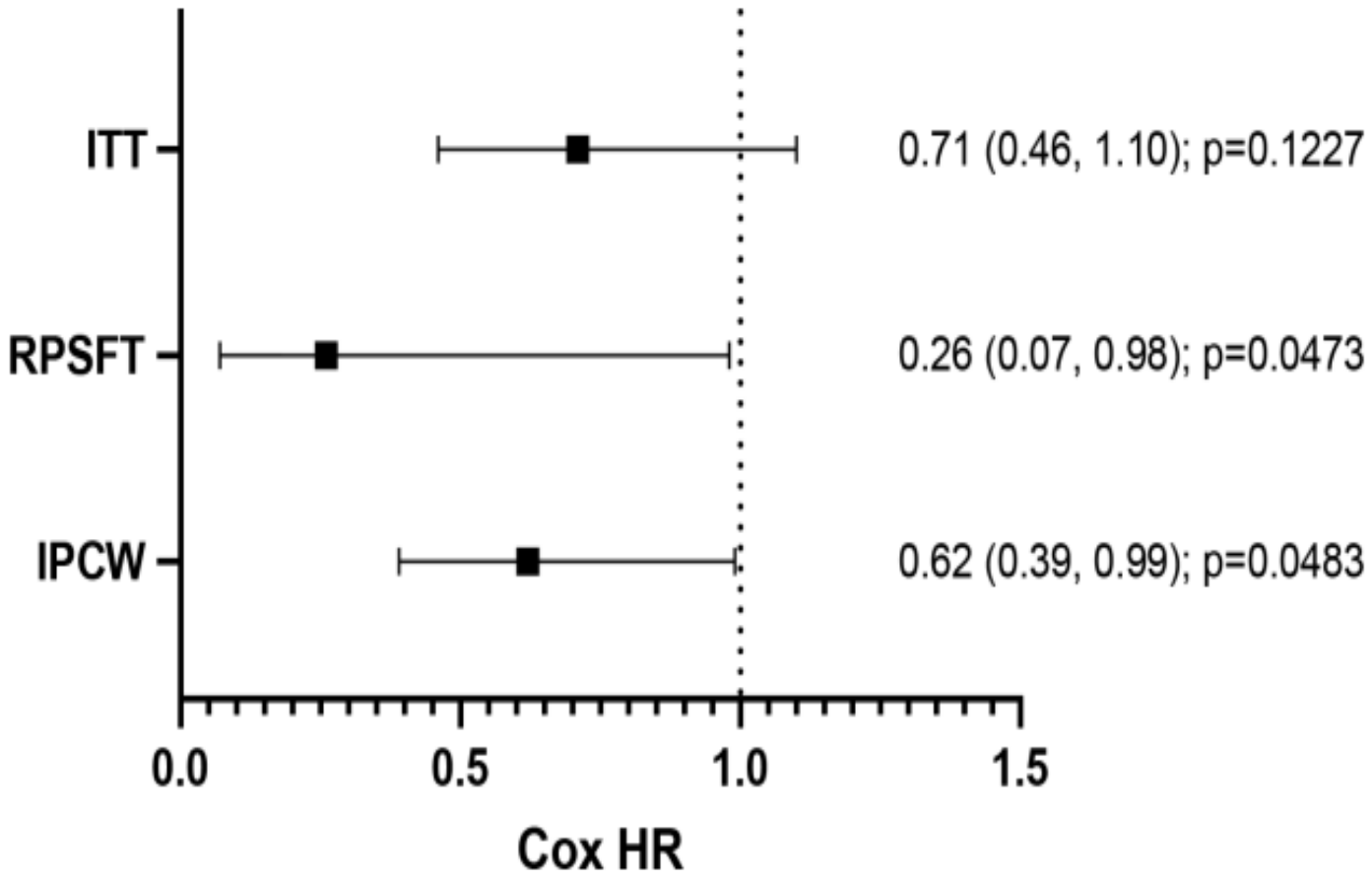
Thorax 2024;79:301–306

Methods



INCREASE : 16w
OLE : maximum 124w

Estimated overall survival HR

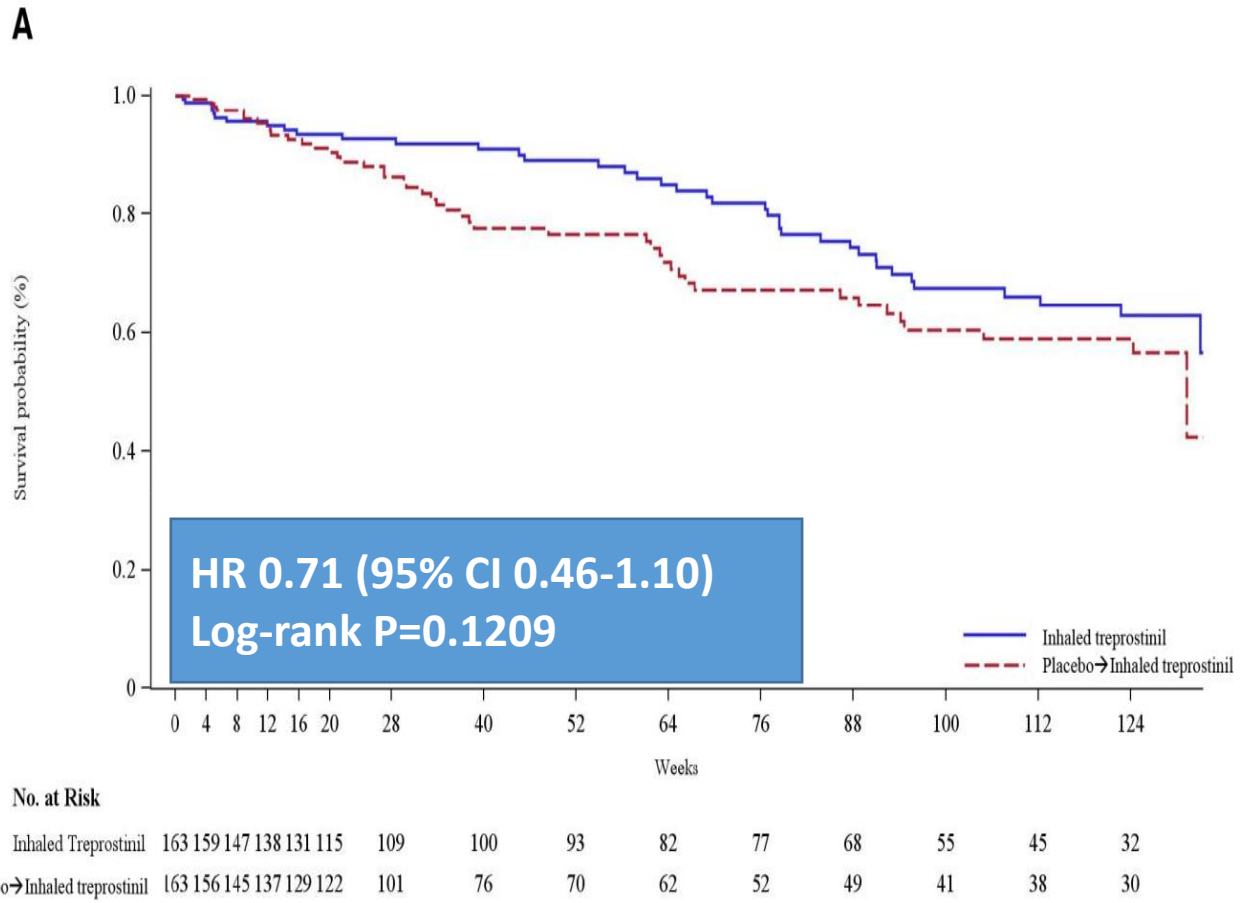


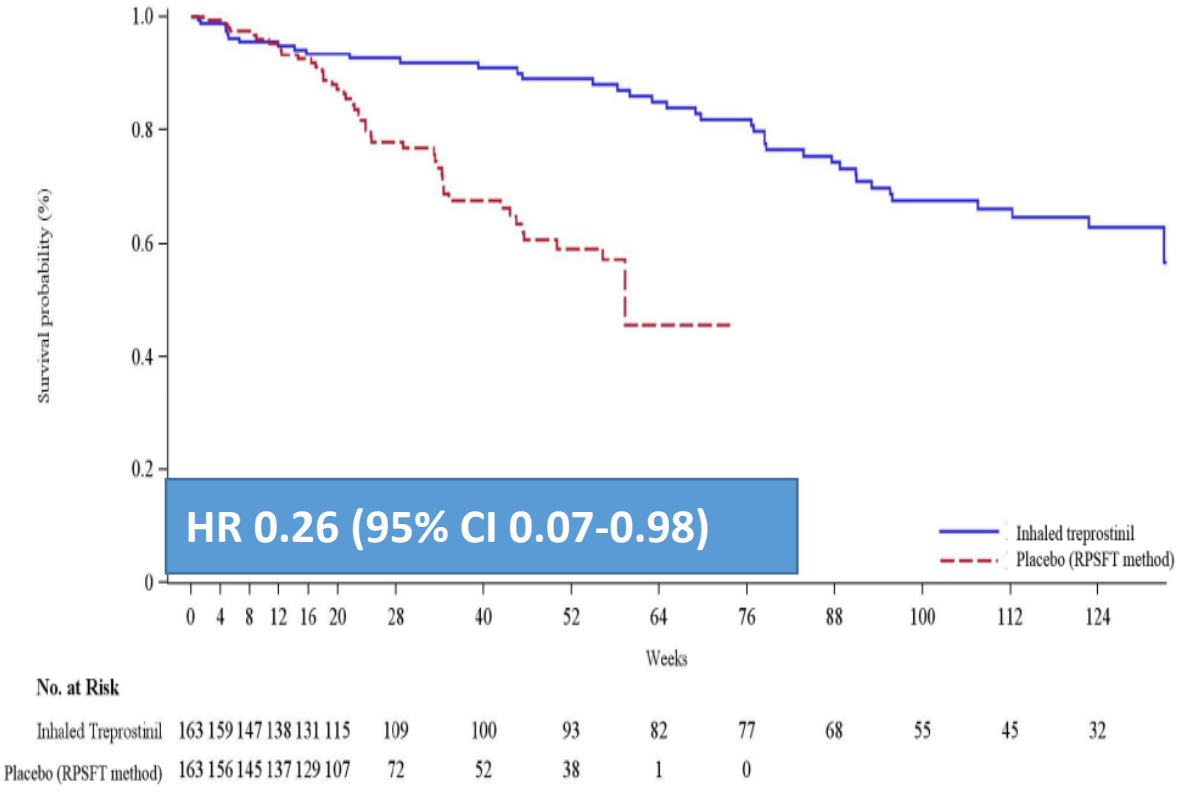
ITT: intention to treat

RPSFT :Rank-Preserving Structural Failure Time

IPCW: Inverse Probability of Censoring Weighting

Survival analysis





Conclusions

- The authors suggest a long-term survival benefit associated with inhaled treprostinil treatment in patients with PH-ILD.

- Two independent modelling techniques were used.

Summary

- Deep learning usage in imaging
  - Can be helpful in diagnosis or prognosis of ILD
- Precision medicine
  - Can be an important concept in ILD
  - Cluster analysis can be the bridge to detailed precision medicine.

- Pulmonary hypertension in ILD
  - PVR could be important in ILD
  - Inhaled treprostinil could improve survival in PH-ILD.

Thank you

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