

Group 3 PH
Associated with lung diseases
and/or hypoxia

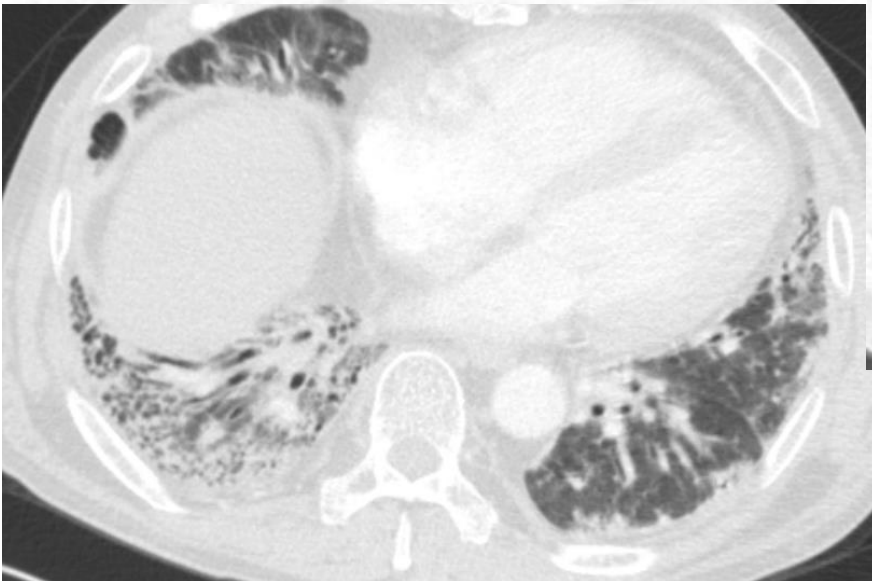
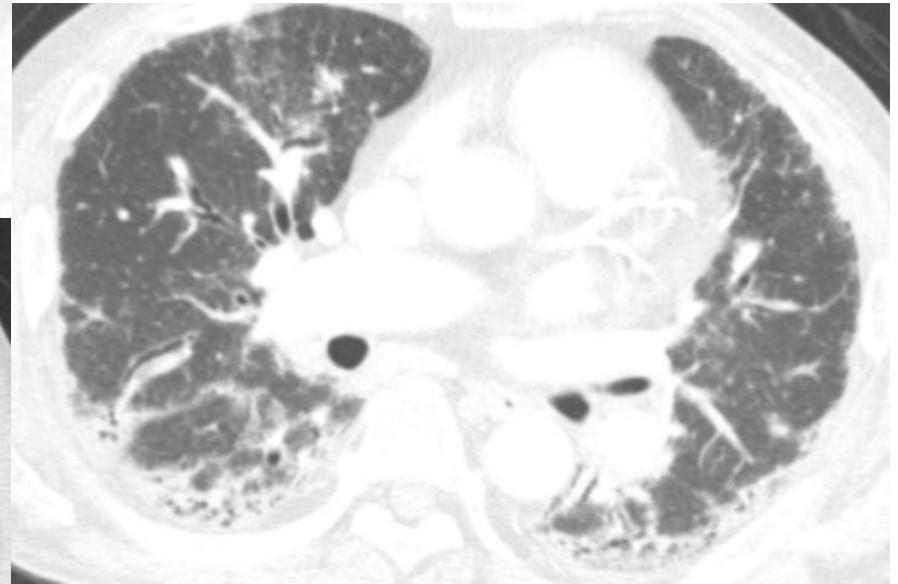
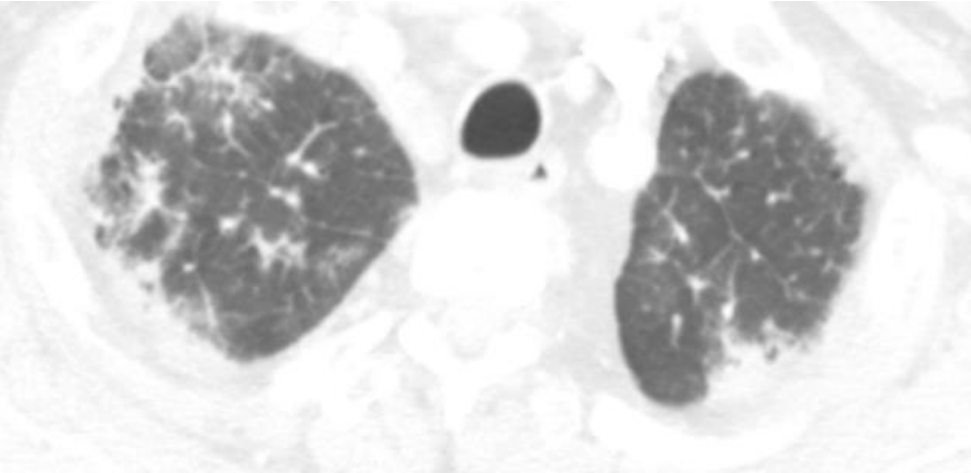
증례 토의

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장항제

Case review

57/M, Rheumatoid arthritis

Associated interstitial lung disease (UIP pattern)

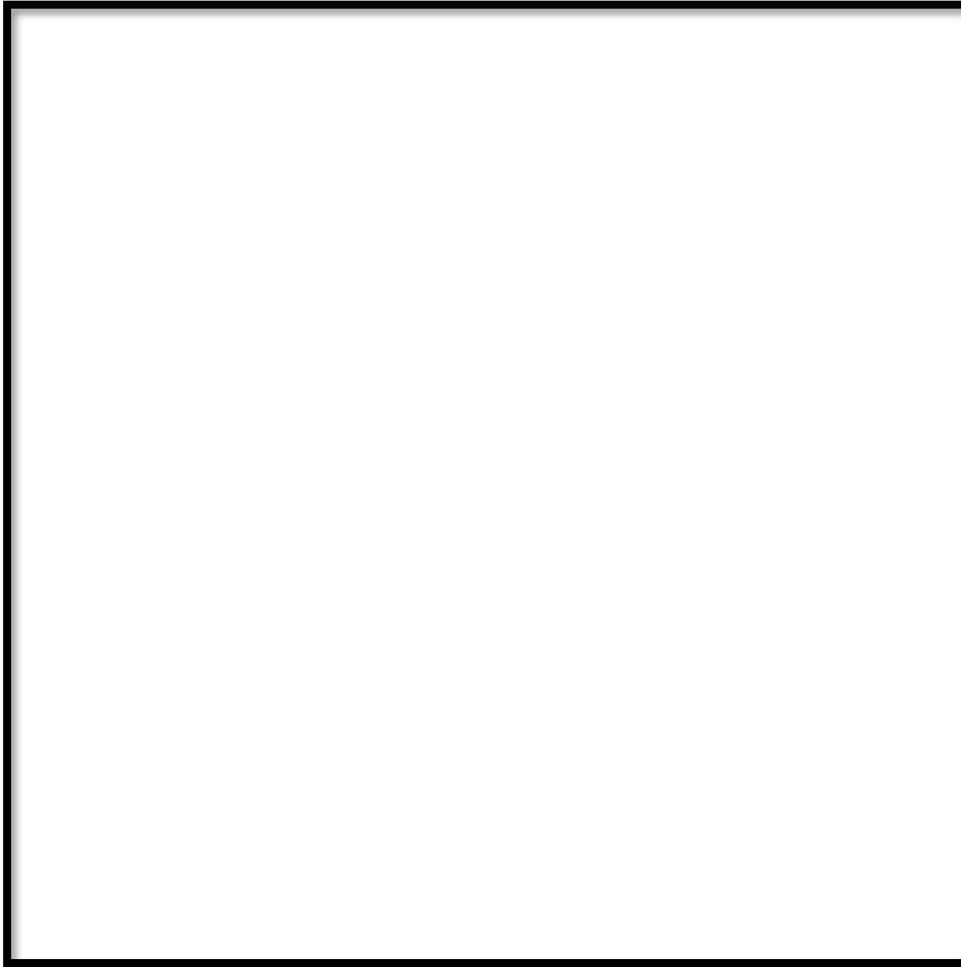




Previous PFT

		Ref	Pre	% Ref
Spirometry				
FVC	Liters	4.87	2.08	43
FEV1	Liters	3.79	1.79	47
FEV1/FVC	%	78	86	
FEF25-75%	L/sec	3.35	2.37	71
IsoFEF25-75	L/sec	3.35	2.37	71
PEF	L/sec	8.35	8.73	105
FET100%	Sec		7.60	
FIF50%	L/sec		6.29	
Diffusing Capacity				
DLCO	mL/mmHg/min	25.9	5.1	20
DL Adj	mL/mmHg/min	25.9	5.1	20
DLCO/VA	mL/mHg/min/L	4.09	1.83	45
DL/VA Adj	mL/mHg/min/L		1.83	
VA	Liters		2.81	
IVC	Liters		1.80	

Echocardiography

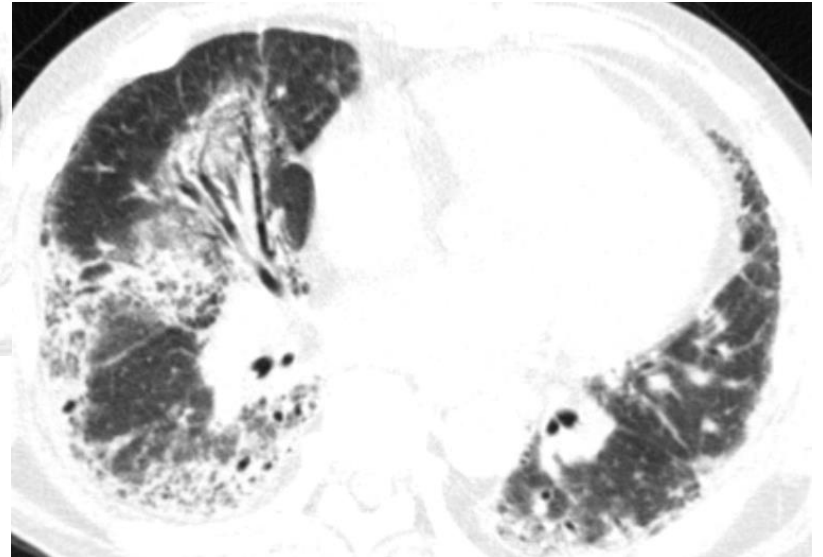
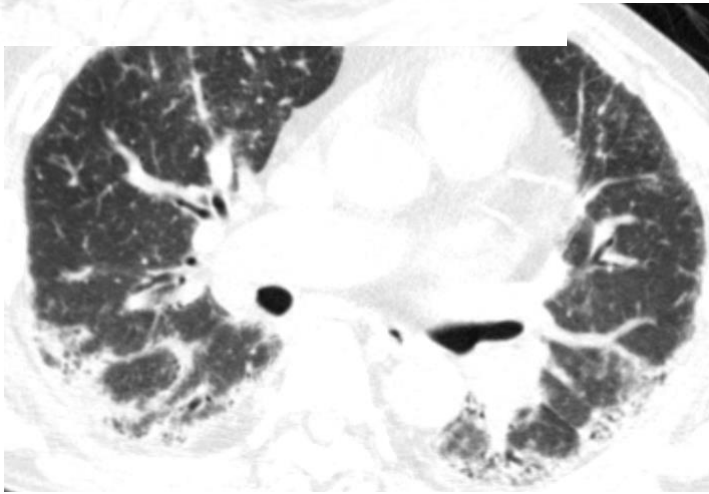
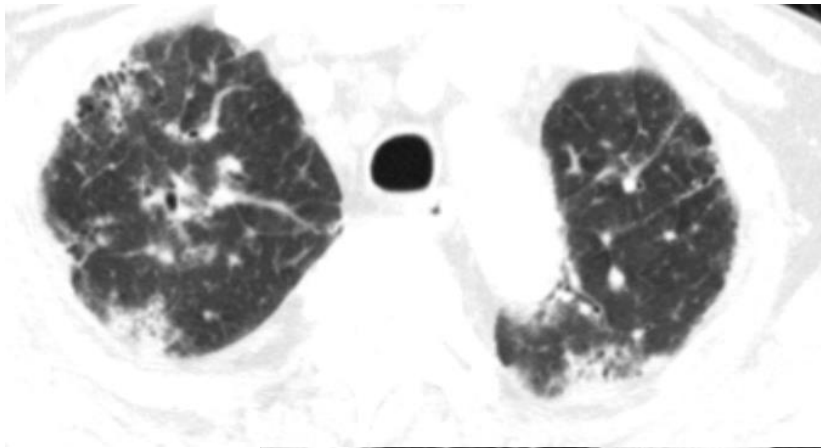


Peak TR velocity
=4.25 m/s

Worsening exertional dyspnea 2 weeks

- Cough ↑ ↑
- Non-purulent mucoid sputum ↑
- Febrile sense (+)
- **Elevated CRP 10.84 mg/dl**
- Procalcitonin 0.06 ng/ml

Pneumonia vs. ILD acute exacerbation



ILD AE >> pneumonia

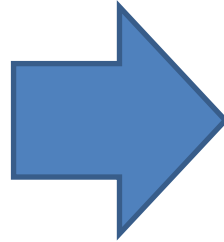
- Short-term High dose steroid
- 3 days pulse therapy (500mg/day)
- 1 mg/kg 7 days

- Cough , sputum, dyspnea → improved
- WHO Functional class III → II

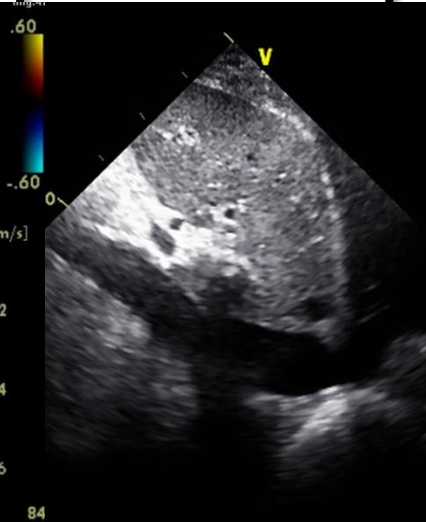
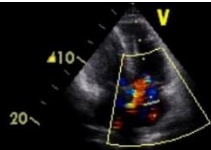
Worsening dyspnea again!

- Both **leg pitting edema** ++/++
- → Functional class IV
- Resting hypoxemia
94% on nasal prong 4 L/min
- Cough, sputum : same as before
- HR CT follow up : no interval change

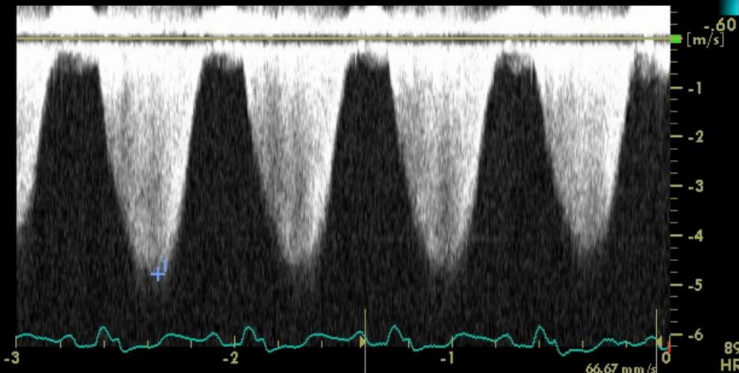
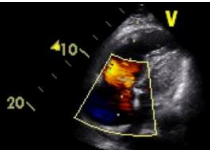
Echocardiography



1 TR Vmax 4.25 m/s
TR maxPG 72.37 mmHg
RVSP 82.37 mmHg



1 TR Vmax 4.78 m/s
TR maxPG 91.48 mmHg
RVSP 101.48 mmHg



Management

- Short term management for RV failure
: Low dose dobutamine infusion
Lasix IV → negative I/O balance
- **Sildenafil** 20 mg t i d PO
→ 5 days later → WHO **Fc III**
→ **Sildenafil** 40 mg t i d PO
→ 2 weeks later → WHO **Fc II**

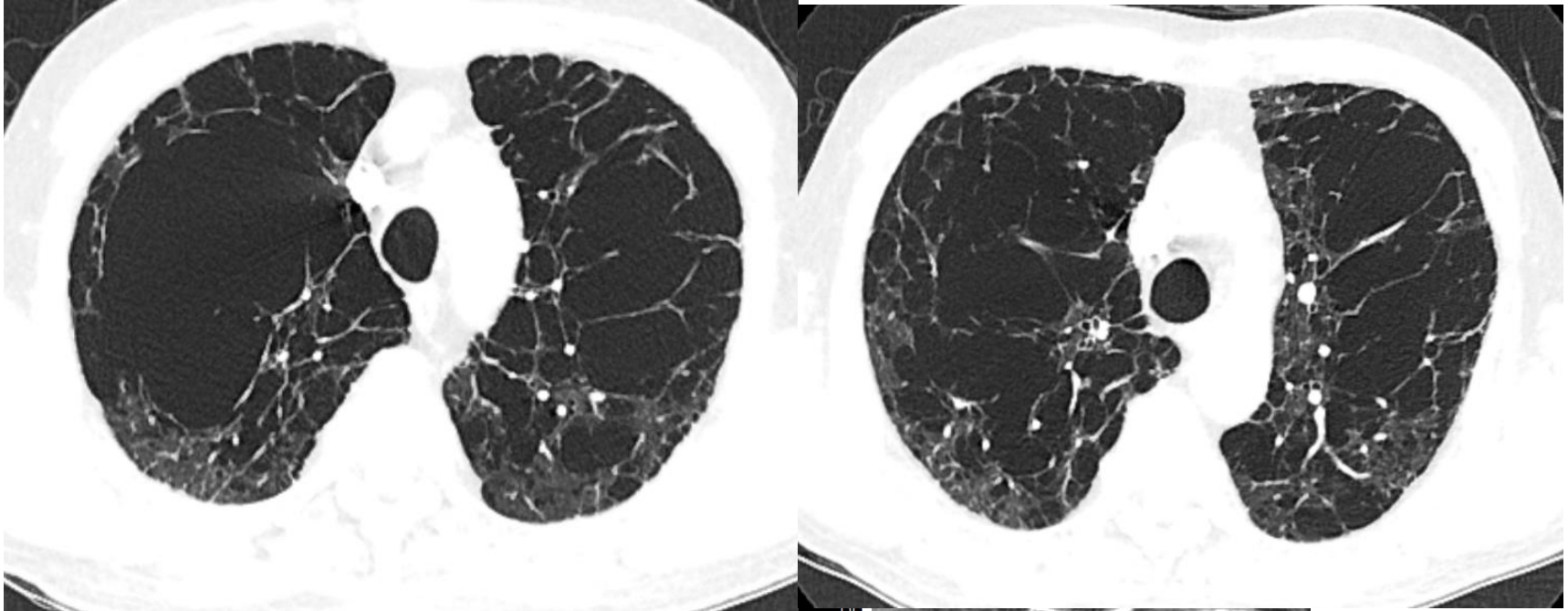
Consider Inhaled prostacycline → patient refused



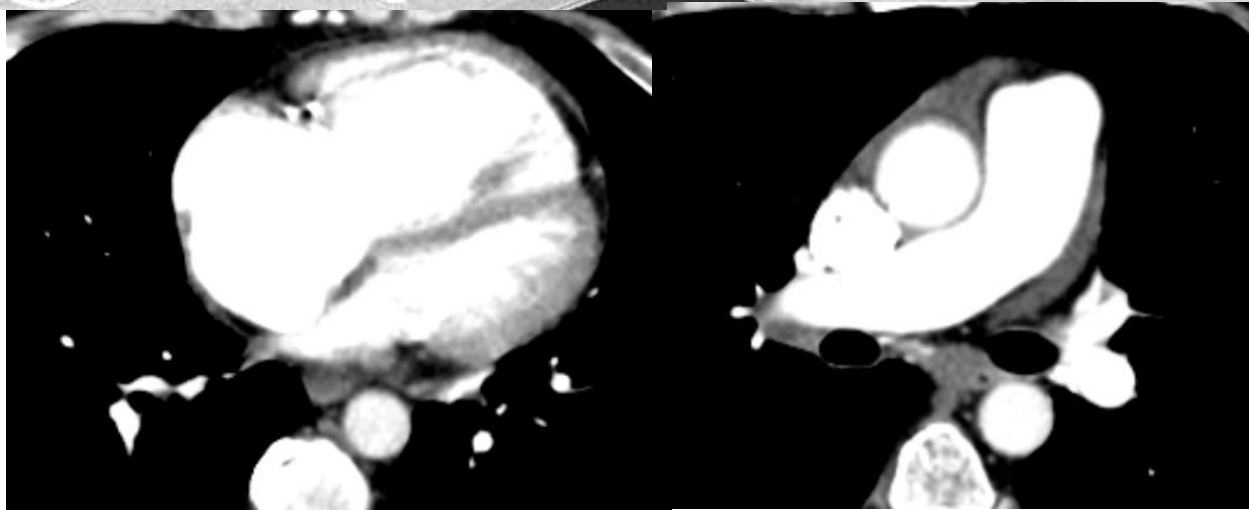
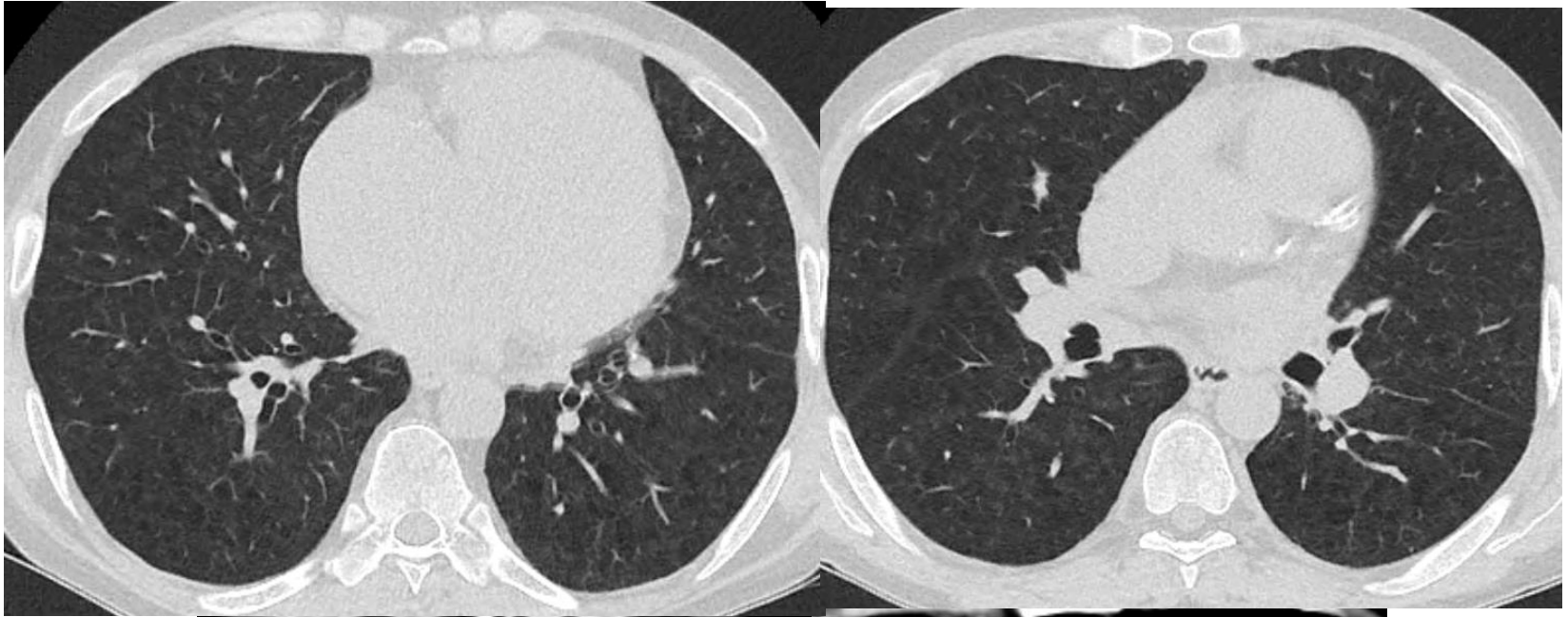
Discussion

- Sero-positive Rheumatoid arthritis
- Associated ILD (UIP pattern)
- PH due to chronic lung disease
- CTD (RA) associated PAH ?

Selection of good candidate for PAH target therapy ??



Selection of good candidate for PAH target therapy ??



2022 ESC/ERS recommendations for PH-LD

Recommendations	Class ^a	Level ^b
In patients with lung disease and suspected PH, it is recommended to optimize treatment of the underlying lung disease and, where indicated, hypoxaemia, sleep-disordered breathing, and/or alveolar hypoventilation	I	C
Inhaled treprostinil may be considered in patients with PH associated with ILD [734]	IIb	B
The use of ambrisentan is not recommended in patients with PH associated with IPF [740]	III	B
The use of riociguat is not recommended in patients with PH associated with IIP [181]	III	B
The use of PAH medication is not recommended in patients with lung disease and non-severe PH ^e	III	C

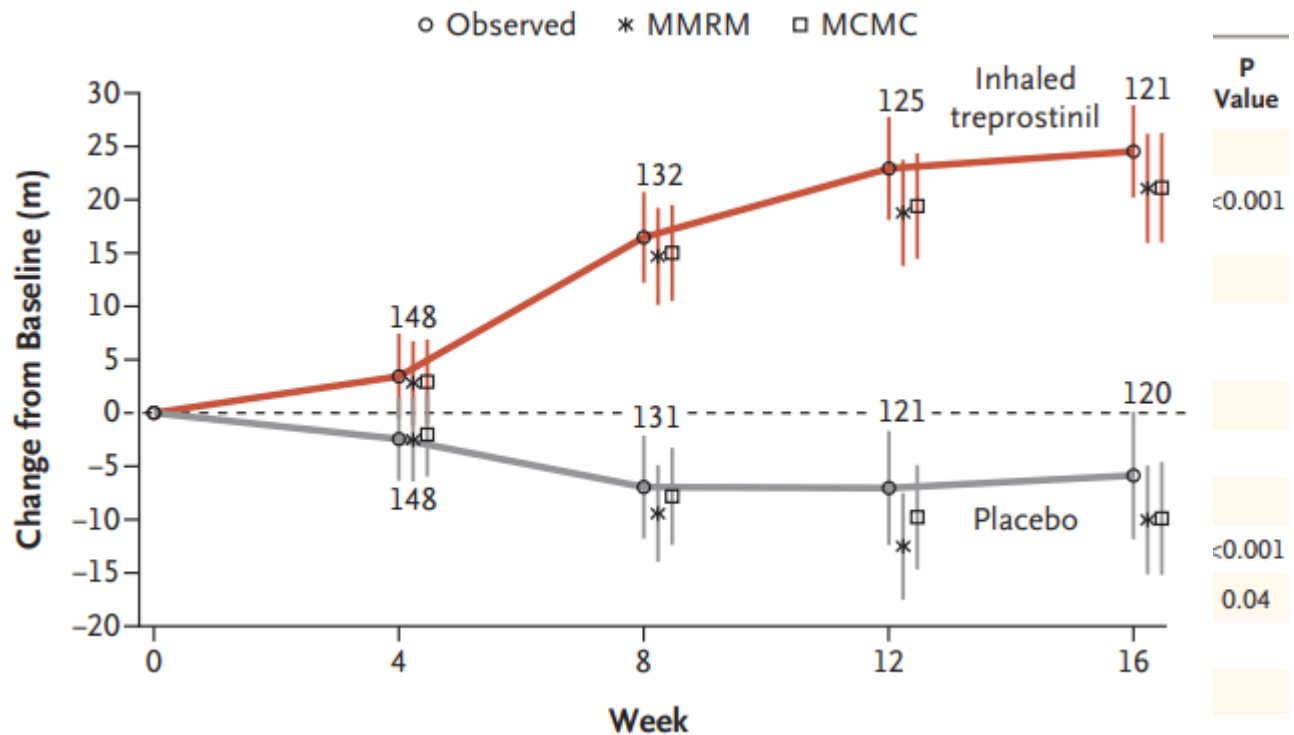
Recommendations	GRADE		Class ^a	Level ^b
	Quality of evidence	Strength of recommendation		
PDE5is may be considered in patients with severe PH associated with ILD (individual decision-making in PH centres)	Very low	Conditional	IIb	C
The use of PDE5is in patients with ILD and non-severe PH is not recommended	Very low	Conditional	III	C

Inhaled Treprostinil in PH-ILD: INCREASE trial

Multicenter, randomized, DB, placebo-controlled, **16-week trial**

Inclusion criteria

End Point
Primary end point
Change in peak 6-minute walk to wk 16 — m†
Secondary end points‡
Change in plasma concentration baseline to wk 16¶
Mean (±SD) change — p
Median — pg/ml
Range — pg/ml
Ratio to baseline
Occurrence of clinical worsening
Any event
Hospitalization for cardiac
Decrease in 6-minute walk baseline
Death from any cause
Lung transplantation



P Value
<0.001
<0.001
0.04

Death from any cause	4 (2.5)	4 (2.5)
Lung transplantation	2 (1.2)	0

Take-home message



Group 3 PH 의 기본적인 치료 전략은 기저 폐질환에 대한 최적화된 치료

Lung disease and non-severe PH 에 대해서는 일반적으로 PAH target medication 이 권장되지 않는다.

환자에 따라 선택적으로
Individual decision making
필요하다



다른 원인에 의한
PH 가능성도 고려하고
배제해야 한다

Cor pulmonale

- Acute



- Sub-acute



- **Chronic**

Hypoxic pulmonary
vasoconstriction



RV dilatation
vascular remodeling

+ Hypertrophy

Acute PTE
Air/Fat/amniotic
embolism

ARDS
Severe pneumonia
Aspiration

Chronic lung disease
: ILD
COPD
TBDL

검색하실 내용을 입력해주세요.



대한결핵 및 호흡기학회

The Korean Academy of
Tuberculosis and Respiratory Diseases

로그아웃 마이페이지 CONTACT US

학회소개

지회 및 연구회

학술행사

논문 및 간행물

뉴스레터

게시판

결핵과 전문의

자료실

지회 및 연구회
연구회



지회 및 연구회



연구회



폐혈관 연구회

알림

임원

회칙

게시판

일정

멤버

멤버관리

폐혈관연구회 송년모임

- 12월 14일 저녁 6시반
- 장소: 서래향 수서점 (중식당)
- 서울 강남구 광평로 51길 6-9 새롬빌딩
- 수서역 3번 출구에서 131 m

Thank you

