

폐혈관 School 2016: Case study (PAH)

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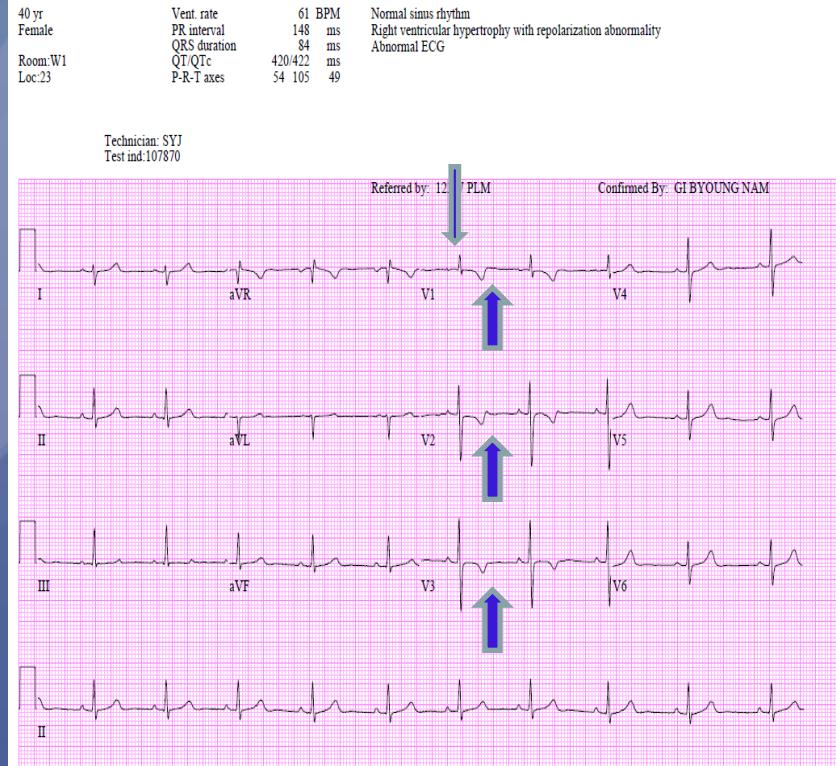
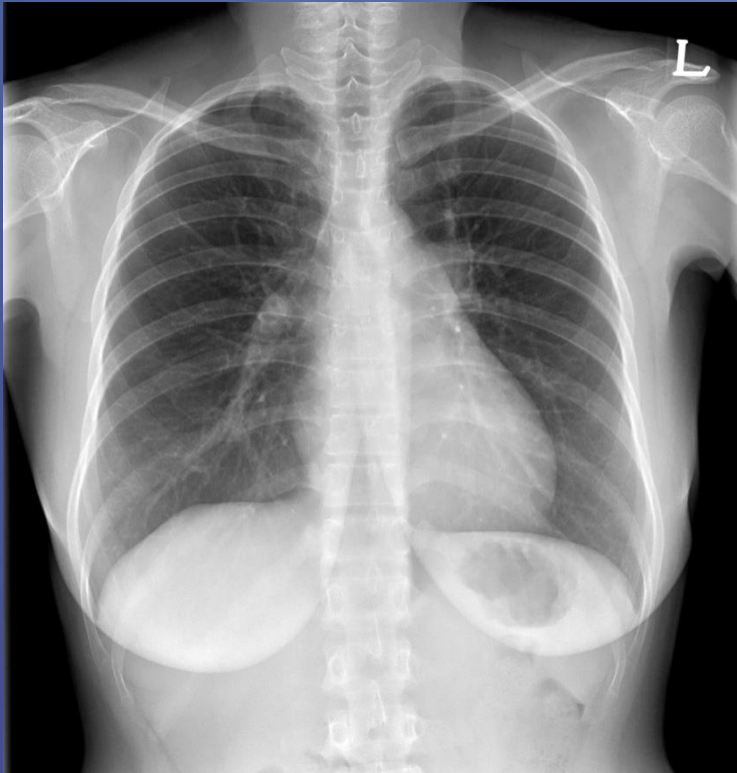
Case presentation

- IPAH: CCB responder
CCB nonresponder; IV & SQ treprostinil
- Connective tissue disease (SLE)-PAH
- CTEPH
- PH associated with pulmonary artery stenosis



Case 1 (F/40)

- C. C. : Dyspnea (WHO Fc II)
- P. M. Hx (-)
- Initial CXR & EKG:



Case 1 (F/40)

Lab.: CBC 6400-**13.5**-237K

BUN/Cr 11/0.7 mg/dL AST/ALT 22/14 IU/L T.B 0.8 mg/dL

HBsAg/Ab (-/-) HCV-Ab(-) **BNP 34 pg/mL**

FANA (-) RF factor (-) ANCA (-)

TSH/free T4 W.N.L

PFT: **FEV1/FVC 79% FEV1 90% pred. FVC 97% pred.**

DLco 75 % pred. DL/VA 82% pred.

6MWD 500m (SaO₂ 98%)



Case 1 echocardiography

Left Ventricle D-shaped LV

LVIDs	25 mm	LVIDd	41 mm	LA	28 mm	Aorta	29 mm
LVPWs	12 mm	LVPWd	7 mm	ESV	20 ml	EDV	67 ml
IVSs	12 mm	IVSd	9 mm	LVEF	70 %	LV Mass	97.7 g
P.E. (ant)	mm	P.E. (post)	mm	LV Mass Index	67.2 g/m ²		

Right Ventricle decreased

RVIDs	mm	RVIDd	27 mm
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Tricuspid Valve Normal

TR grade	2	TR jet area	5 cm ²	Peak TR Vel	4.2 m/s
PGsys(RV-RA)	71 mmHg	Peak E Vel	cm/s	Peak A Vel	cm/s

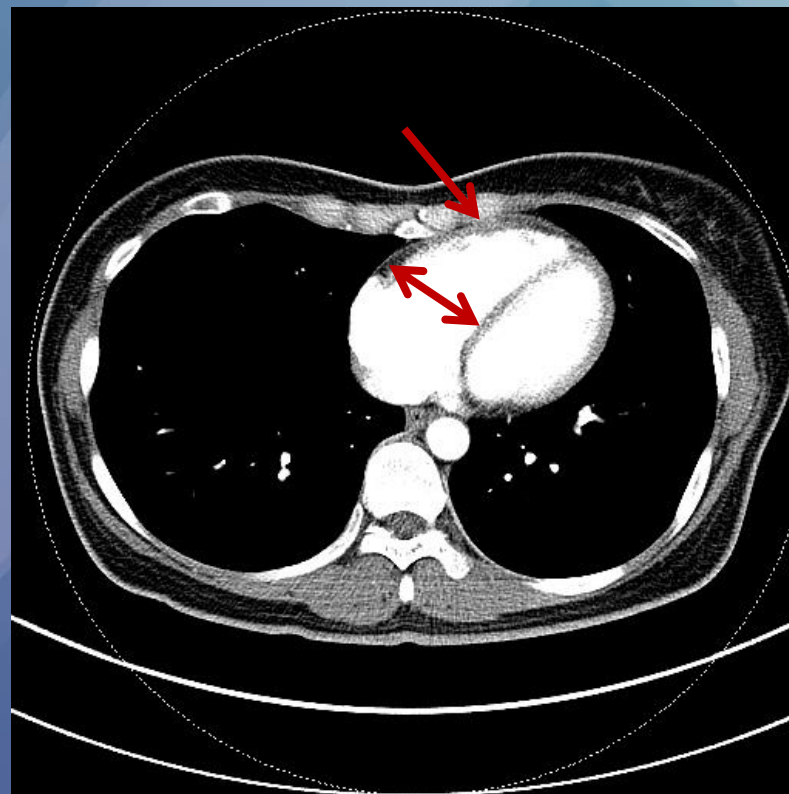
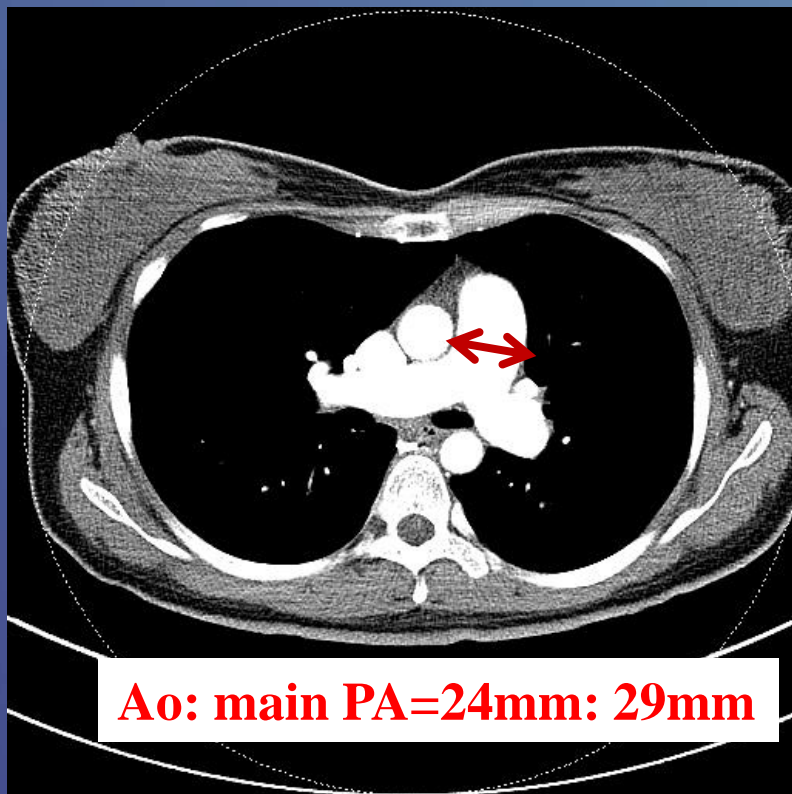
RV free wall thickness 8mm, main PA 27mm

Agitated saline test : no intracardiac shunt

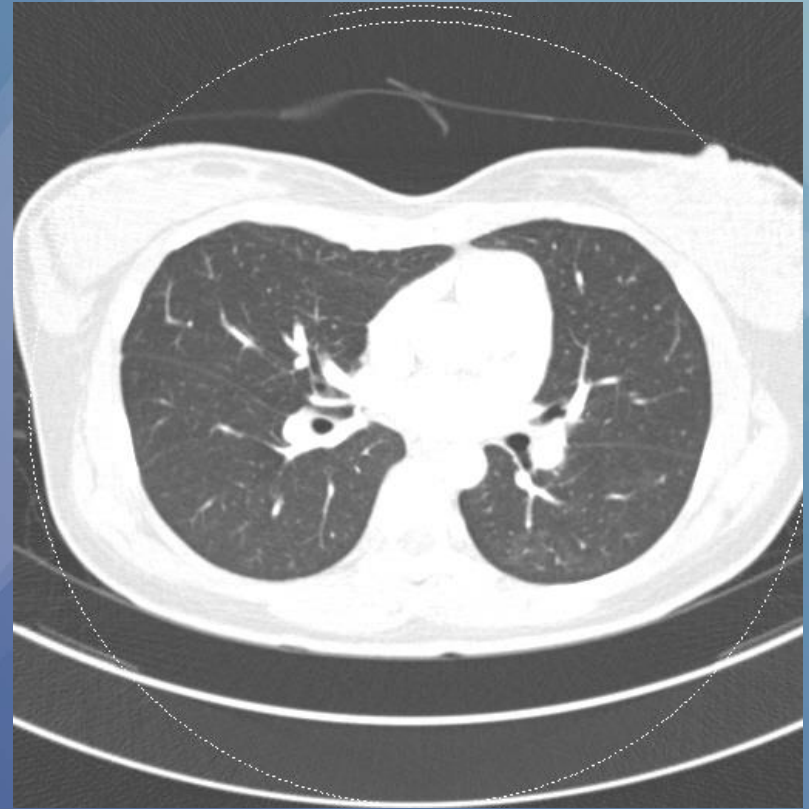
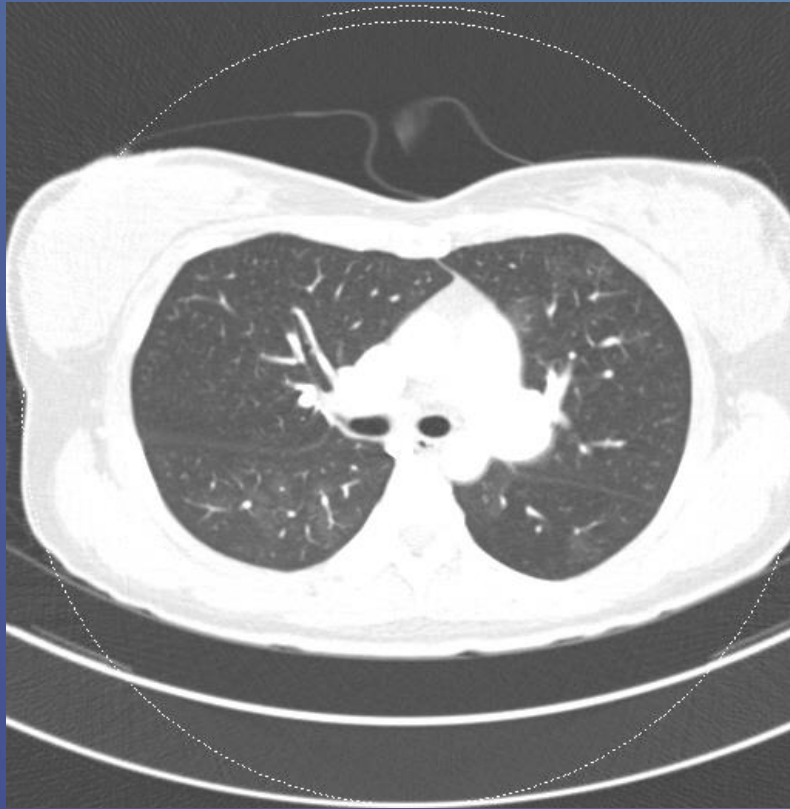
-> RV dysfunction with severe resting PH



Case 1; Chest CT



Case 1; Chest CT



Lobular GGO in both upper and superior segment of both lower lobes



Case 1; Perfusion scan

Patient Name: KIM, SOOHEE, F40
Study Date: 7/5/2011

Patient ID: 42187196

Study Name: Lung^Perfusion Scan

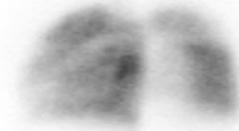
Lung Perfusion Scan
99mTc-MAA



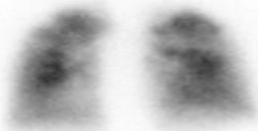
ANT



LAO



LPO



POST



RPO



RAO

Medium to large perfusion defects in BUL



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Case 1; RHC & acute vasodilator test

	Baseline	NO 10ppm	NO 20ppm		Peridipine 1.5mg
BP	121/68	118/64	124/74		97/52
sPAP/dPAP	69/28	28/9	16/8		40/12
Mean PAP	43	17	12		24
PCWP	2	2	2		2
CO	3.7	3.5	3.9		3.8
HR	71	60	69		82
PVR	886	366	246		484



Case 1; Diagnosis & treatment

- Dx: **IPAH, CCB responder**
- Tx: **nifedipine 30mg qd**

- 3 Month f/u
 - Dyspnea (-)
 - Echo: **TR Vmax 2.2m/s, PG 19 mmHg**
normalized RV contractility

- 5 year f/u with nifedipine 30mg qd without deterioration.



Case 1; f/u echo after 5 years

Left Ventricle Normal LV

LVIDs	26 mm	LVIDd	45 mm	LA	25 mm	Aorta	25 mm
LVPWs	14 mm	LVPWd	7 mm	ESV	25 mL	EDV	65 mL
IVSs	10 mm	IVSd	7 mm	LVEF	62 %	LV Mass	96.0 g
P.E. (ant)	mm	P.E. (post)	mm	LV Mass Index		63.6 g/m ²	

Right Ventricle preserved RV contractility

RVIDs	mm	RVIDd	21 mm
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Tricuspid Valve Normal

TR grade	tr	TR jet area	cm ²	Peak TR Vel	2.4 m/s
PGsys(RV-RA)	23 mmHg	Peak E Vel	cm/s	Peak A Vel	cm/s

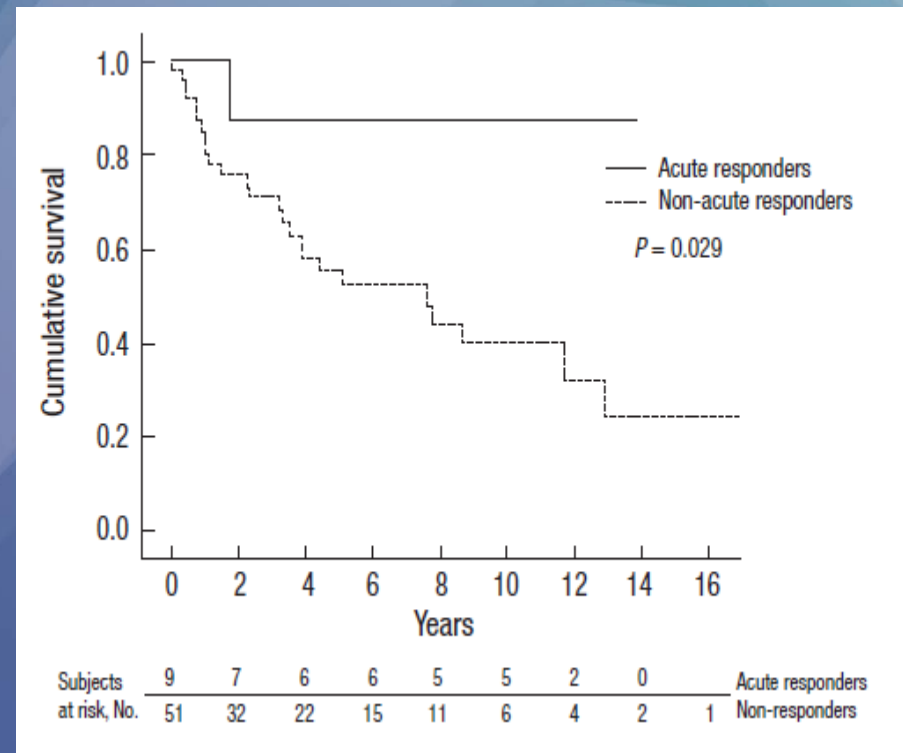
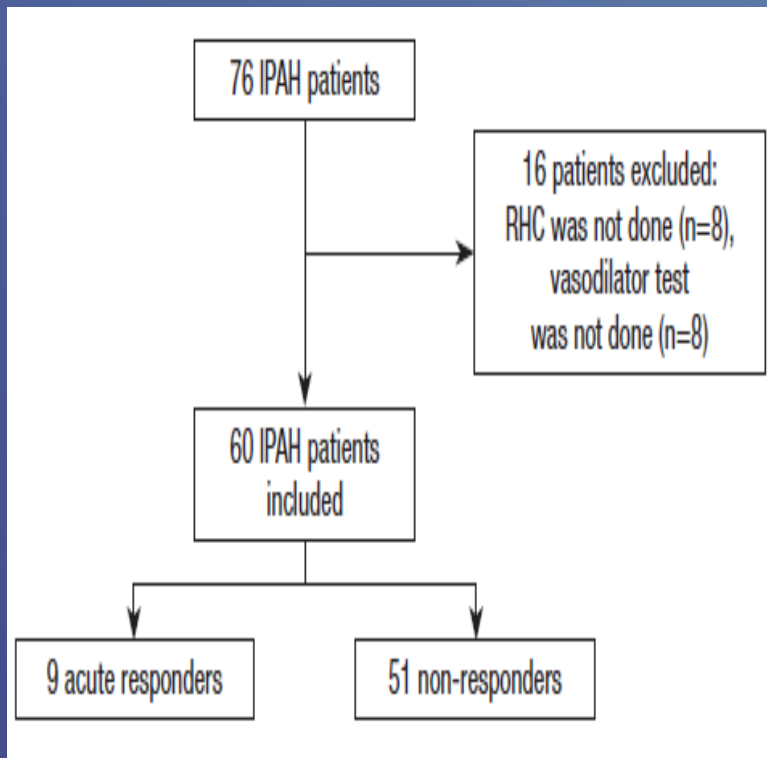
Normal RV size with relieved resting PH





CrossMark
click for updates

Clinical Characteristics and Survival of Korean Idiopathic Pulmonary Arterial Hypertension Patients Based on Vasoreactivity

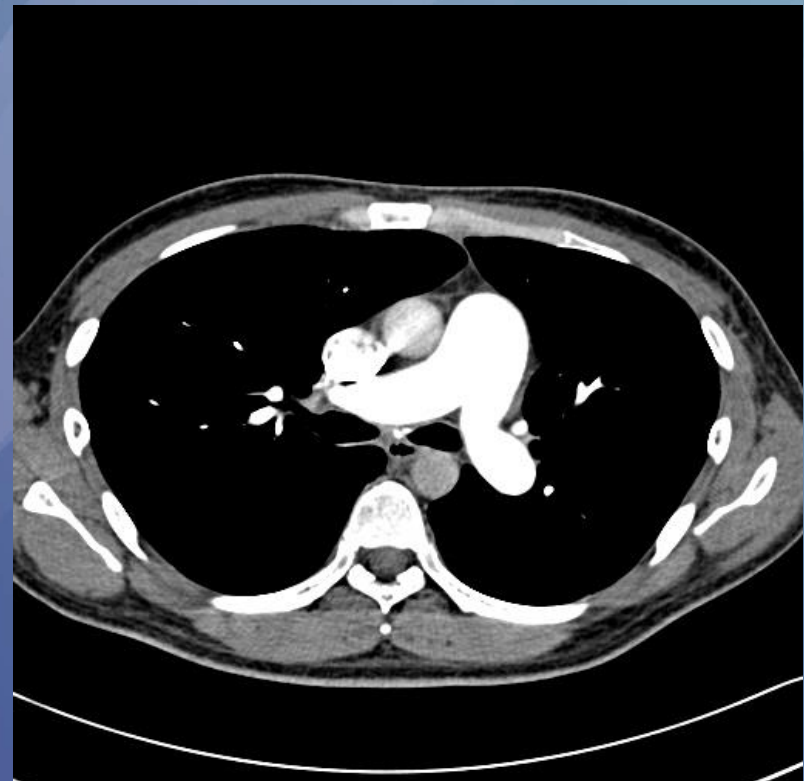
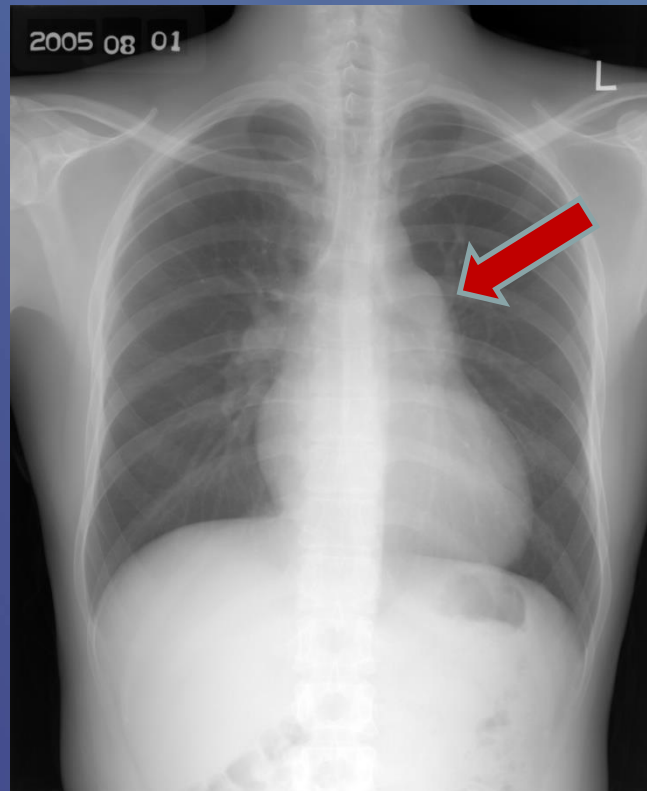


Characteristics of acute vasoreactivity positive IPAH patients

	All Patients (n=60)	Acute Responders (n=9)	Non- Responders (n=51)	P Value
Age at diagnosis, yr	33±13	38±16	32±13	0.355*
Onset of symptoms, mo	35±46	39±40	34±47	0.858*
WHO functional class I:II:III:IV, %	3:32:60:5	11:44:44:0	2:29:63:6	0.085‡
Six-minute walk distance, m	413±137	460±147	404±135	0.077*
Peak velocity of TR, m/sec	4.5±0.6	4.1±0.3	4.6±0.6	0.01*
Mean RAP, mm Hg	8±5 (n=30)	6±4 (n=8)	8±6 (n=22)	0.237*
Mean PAP, mm Hg	61±17	47±10	63±17	0.003*
Cardiac output, L/min	4.0±1.4 (n=57)	4.7±2.6	3.9±1.1 (n=48)	0.632*
PVR, Wood units	13.8±7.5 (n=41)	10.3±4.9	14.7±7.9 (n=32)	0.154*
SvO ₂ , %	67±9 (n=46)	70±5 (n=8)	66±9 (n=38)	0.191*

Case 2 (M/31)

- C. C : Dyspnea (WHO Fc III)
- P/Ex: both leg pretibial pitting edema
- Initial CXR & CT:



Echo : **TR Vmax 5.1 m/s, PG(RV-RA) 104 mmHg**

-> severe resting PH with severe RV dysfunction

PFT: FEV1/FVC 85% FEV1 88% pred. FVC 83% pred.

DLco 82 % pred.

6MWD 537m (SaO2 96%)

RHC: **sPAP/dPAP/mPAP 93/54/66 mmHg PCWP 13 mmHg**

CO 3.2 L/m (CI 2.0) PVR 13 WUs

(Non-responder to NO)

Assessment: IPAH

Treatment : sildenafil 25mg tid, inhaled iloprost



7 years after combination treatment with sildenafil and iloprost

S: dyspnea (Fc IV)

O: both pretibial pitting edema

BUN/Cr **21/1.59 mg/dL**

AST/ALT 35/20 IU/L

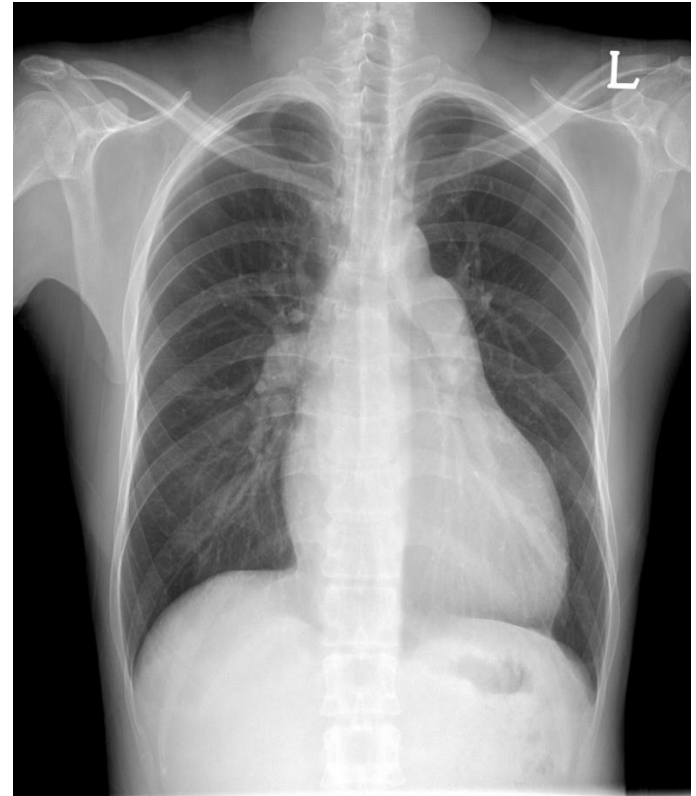
T. B **3.5 mg/dL**

BNP **1247 pg/mL**

6MWD 335m (95%)

Echo: **TR Vmax 5.4m/s**

PG_{sys}(RV-RA) **117 mmHg**



A: IPAH deterioration with RV failure

Plan: dobutamine, lasix

inhaled iloprost -> IV treprostinil 변경



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IV treprostinil to SC treprostinil

HD#6 C-line insertion

IV treprostinil 2ng/kg/min start

HD#9 IV treprostinil 14ng/kg/min up-titration

3시간 후

-> **nausea, dyspnea, BP 87/69 mmHg, SaO2 75%**

IV treprostinil 12 ng/kg/min down-titration

→ BP 105/78mmHg, PR 103/m, RR 20/m

HD#12 IV treprostinil 12ng/kg/m

-> SC treprostinil 12ng/kg/m

HD#14 SC treprostinil 20ng/kg/m

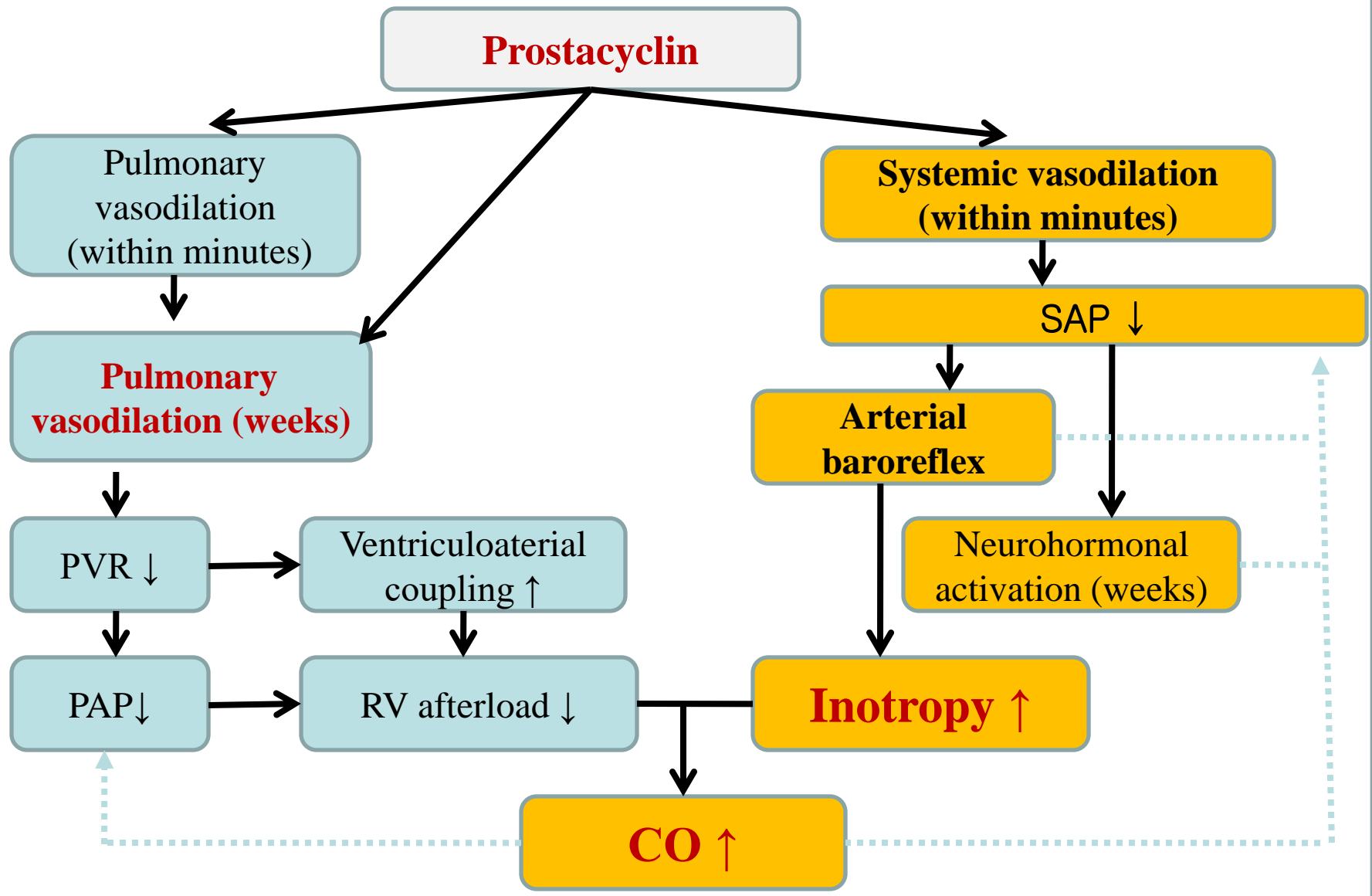
HD#18 discharge

Outpatients up-titration 2ng/kg/m per 2 weeks

Up to 40ng/kg/min



Hemodynamic effects of IV prostanooids



Adverse events of SC treprostinil

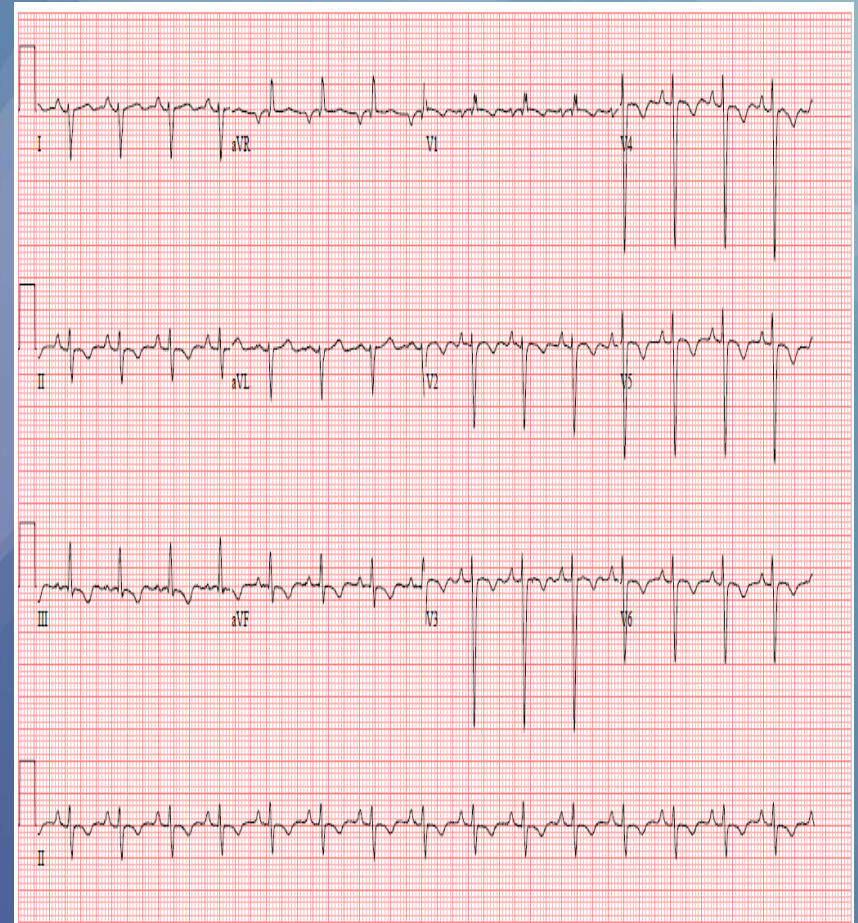
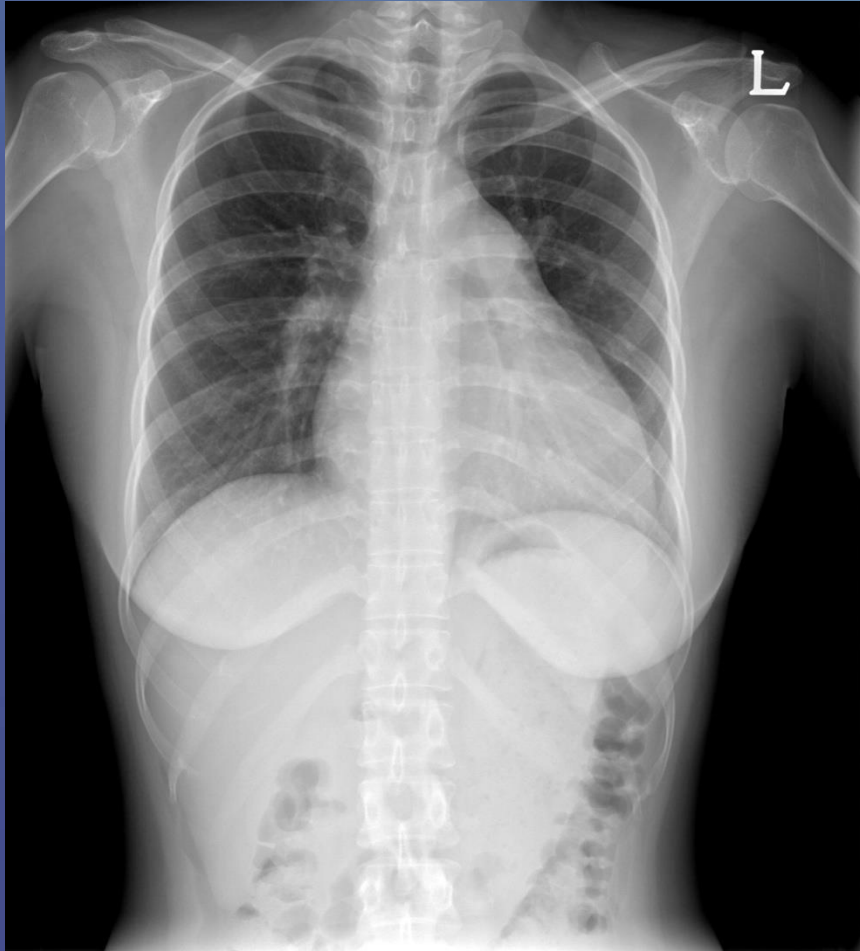
Events	Treprostinil (n=233) (%)	Placebo (n=236) (%)	p Value
Infusion site pain	85	27	<0.0001
Infusion site reaction	83	27	<0.0001
Infusion site bleeding/bruising	34	44	ns
Headache	27	23	ns
Diarrhea	25	16	0.009
Nausea	22	18	ns
Rash	14	11	ns
Jaw pain	13	5	0.0001
Vasodilation	11	5	0.01
Dizziness	9	8	ns
Edema	9	3	0.002



Case 3 (F/31)

- C. C : DOE (WHO Fc II) onset; 1 month ago
- P. I: 20YA photosensitivity
 - 8YA polyarthritis (wrist, PIP, MCP) and morning stiffness
 - dry mouth, recurrent oral ulcer
 - 3YA **Raynaud's phenomenon**
- P.M. Hx: **Hypothyroidism**
- R. O. S: cough (-), sputum (-) hemoptysis (-) chest pain (-)
- P/Ex.: BP 104/75 mmHg PR 100/min
 - jugular vein distension (-)
 - RHB without murmur
 - Pretibial pitting edema (-)

Case 3 (F/31)



Case 3 (F/31)

Lab.: CBC 5500-15.1-153K

PT (INR) 1.03 aPTT 23.8s

BUN/Cr 8/0.48 mg/dL Protein/albumin 8.5/3.5 g/dL

AST/ALT 49/36 IU/L T.B 0.6 mg/dL

CRP 1.16 mg/dL ESR 74 mm/hr

BNP 535 pg/mL

U/A albumin (-) RBC (-) WBC (-)

ABGA 7.46-24-104-18.0-98%

TSH/free T4 5.7uU/mL, 1.5 ng/dL

HBsAg/Ab (-/-) HCV-Ab(-)



Case 3 (F/31)

FANA speckled 1:1280 anti-ds-DNA 13.6 IU/mL
anti-U1RNP (+) anti-SSA(Ro)/SSB(La) (+/equivocal)
C3/C4/CH50 197mg/dL/15.1 mg/dL/47.2U/ml

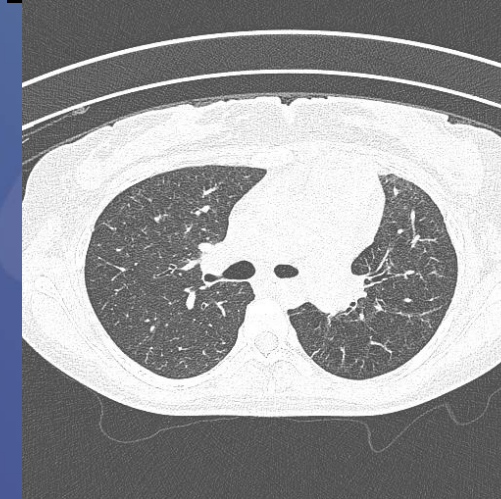
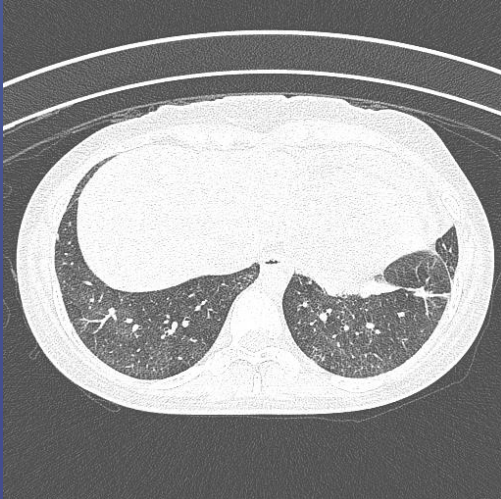
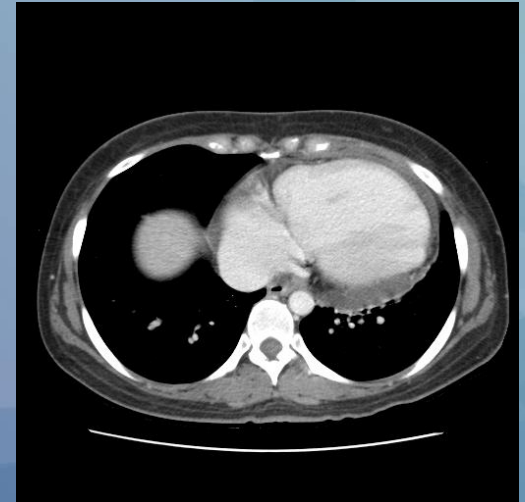
RF factor (-) ANCA (-)

Lupus anticoagulant (-) ACA IgG/IgM (-/+)

Beta2-GPI IgG/IgM (-/-)



Case 3 (F/31)



Case 3 (F/31)

Echo : LV EF 68% E/E' 9 D-shape LV

TR Vmax 4.2 m/s, PG(RV-RA) 71 mmHg

RV wall thickness 6mm

-> severe resting PH with RV dysfunction

PFT: **FEV1/FVC 82% FEV1 55% pred. FVC 52% pred.**

DLco 42 % pred.

6MWD 200m (SaO₂ 84%)

RHC: **sPAP/dPAP/mPAP 90/51/57 mmHg** PCWP 10 mmHg

CO 2.37 L/m (CI 2.4) PVR 19 WU



Case 3 (F/31) Diagnosis and treatment

SLE-PAH with ILD

Bosentan 62.5 mg bid and mPD 1mg/kg

	2014.01.07	2014.01.18	2014.03.05
TRVmax	4.2	3.3	
PGsys(RV-RA)	71	44	
BNP	535		26
6MWD	200		440
FVC	55% pred.		73% pred.
DLco	42% pred.		51% pred



Case 3 (F/31) 2 year after bosentan

Left Ventricle Normal LV

LVIDs	24 mm	LVIDd	41 mm	LA	32 mm	Aorta	30 mm
LVPWs	14 mm	LVPWd	9 mm	ESV	23 mL	EDV	69 mL
IVSs	14 mm	IVSd	10 mm	LVEF	67 %	LV Mass	123.5 g
P.E. (ant)	mm	P.E. (post)	mm	LV Mass Index	79.6 g/m ²		

Right Ventricle preserved RV contractility

RVIDs	mm	RVIDd	mm
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Tricuspid Valve normal

TR grade	tr	TR jet area	cm ²	Peak TR Vel	2.6 m/s
PGsys(RV-RA)	27 mmHg	Peak E Vel	cm/s	Peak A Vel	cm/s

Normal RV size with relieved resting PH



Case 4. (64/M)

Chief complaint : dyspnea (WHO II)

Present illness

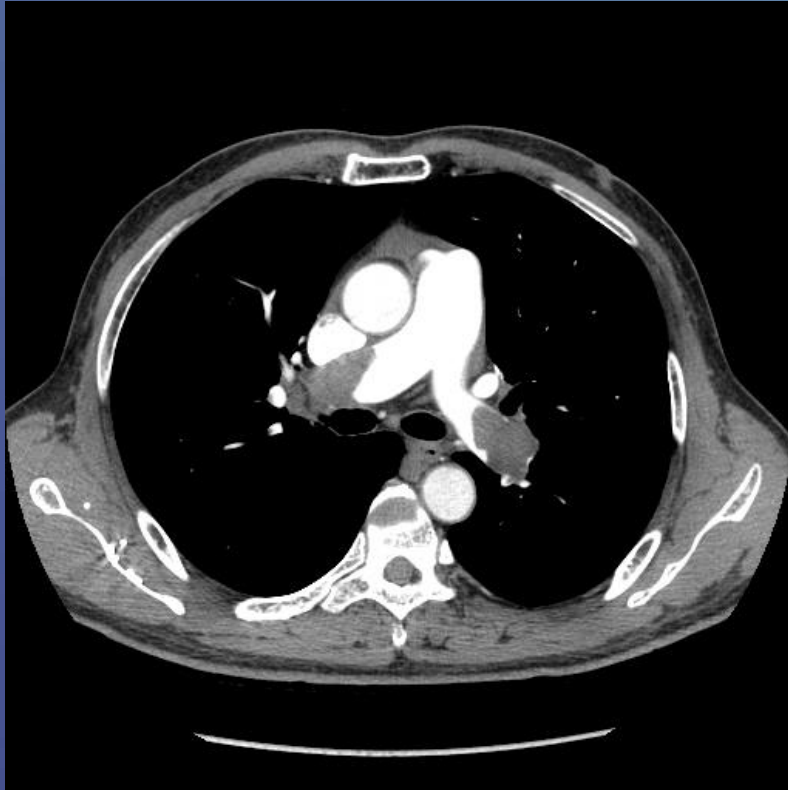
내원 9년 전 실신으로 외부병원 입원하여 acute PE with DVT로 thrombolysis 치료 시행 후 warfarin 복용함.

내원 2년 전 심장초음파상 우심기능이상 동반한 폐고혈압으로 sildenafil 복용

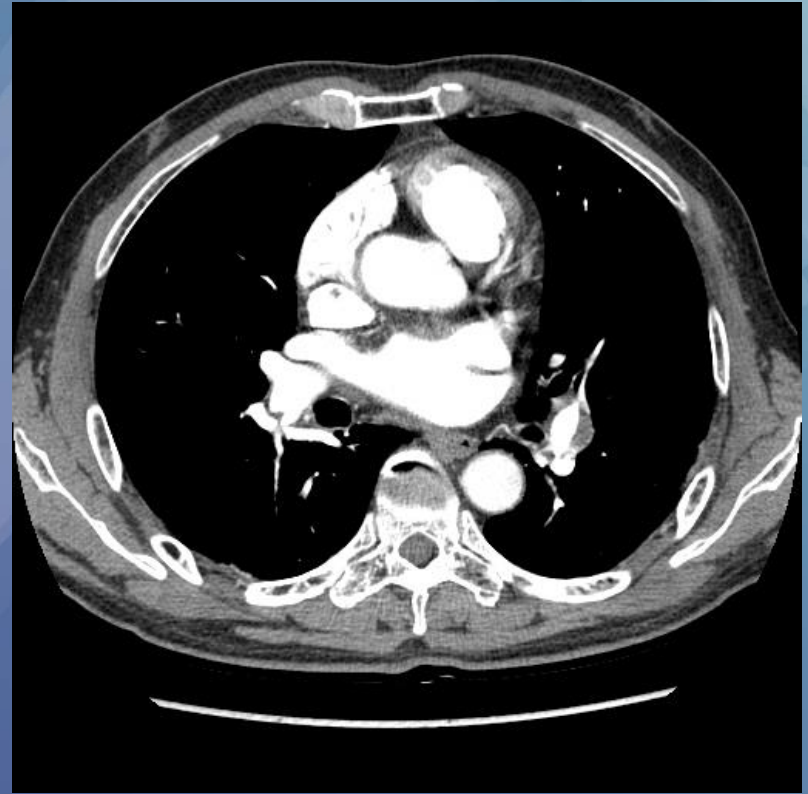
내원 3개월 전 검진 위내시경검사에서 조기위암으로 수술 위해 내원



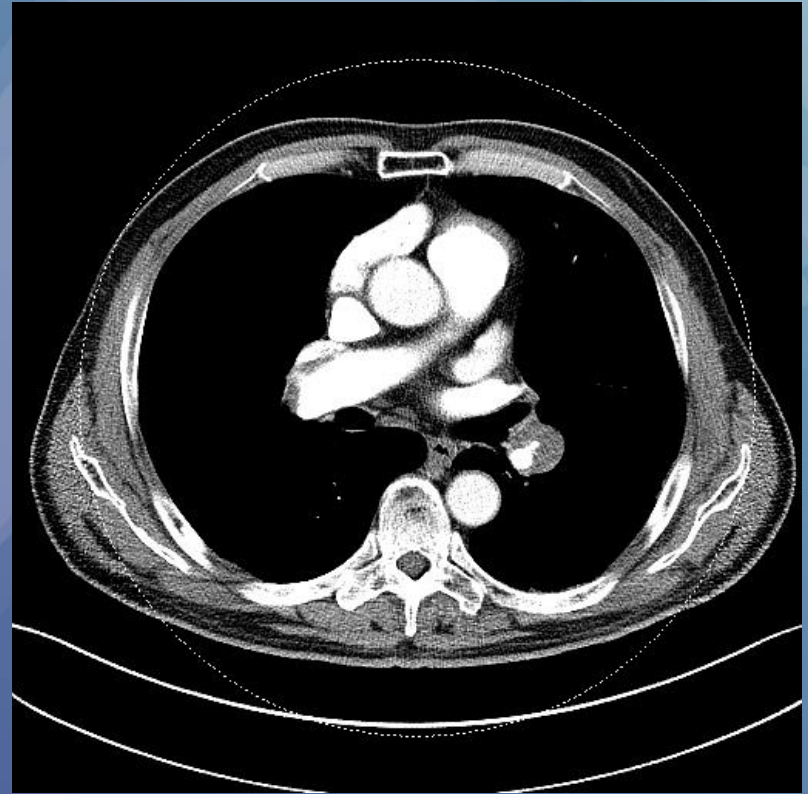
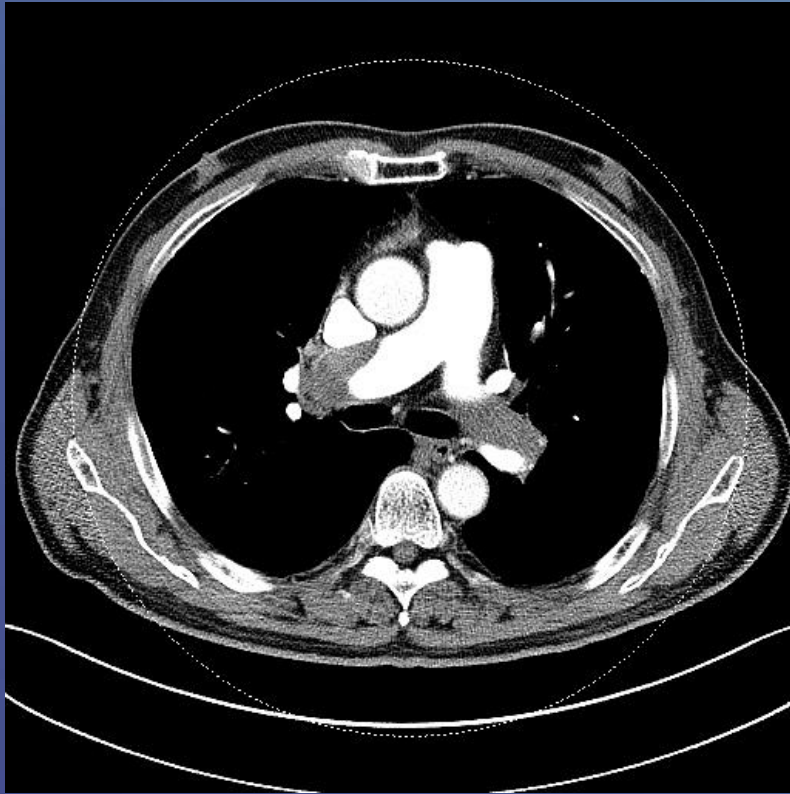
Case 4; Chest CT (9 years ago)



Case 4; Chest CT (after thrombolysis)



Case 4; Chest CT(1 week ago)



Case 4; Preoperative echo

Left Ventricle D-shaped LV

LVIDs	34 mm	LVIDd	48 mm	LA	40 mm	Aorta	37 mm
LVPWs	13 mm	LVPWd	10 mm	ESV	39 mL	EDV	115 mL
IVSs	13 mm	IVSd	9 mm	LVEF	66 %	LV Mass	159.7 g
P.E. (ant)	mm	P.E. (post)	mm	LV Mass Index	88.1 g/m ²		

Right Ventricle decreased contractility(Ts 4.4 cm/sec)

RVIDs	mm	RVIDd	mm
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Tricuspid Valve mild tethering

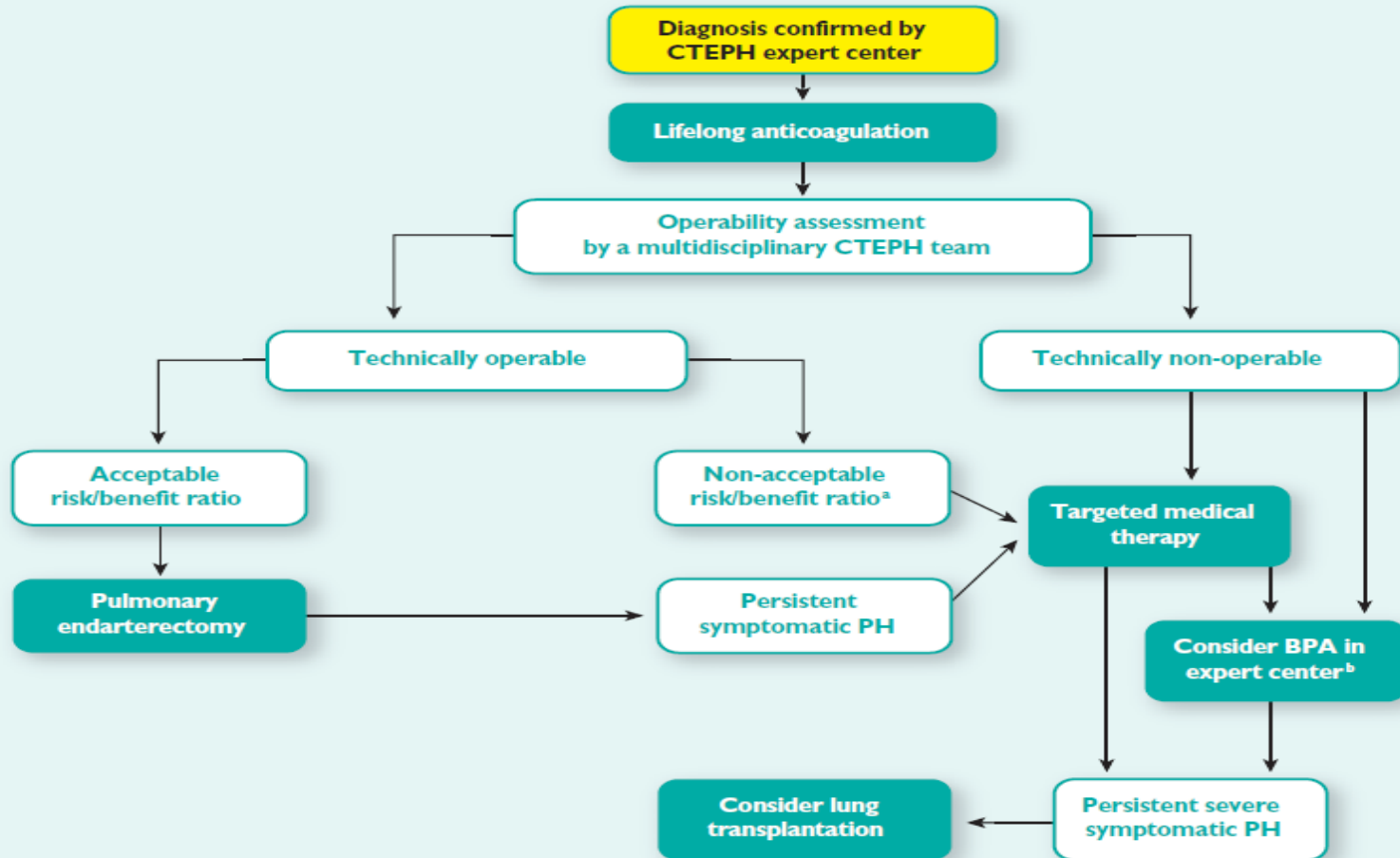
TR grade	2	TR jet area	6 cm ²	Peak TR Vel	4.4 m/s
PGsys(RV-RA)	77 mmHg	Peak E Vel	cm/s	Peak A Vel	cm/s

Case 4; Preoperative RHC

		max	min	mean
Assumed VO ₂ (ml/min/m ²)			227.5	
Hemoglobin (g/dL)			16.9	
Aorta	Pressure	98	57	74
Aorta(PV)	Saturation		97	
Aorta(PV)	PaO ₂		85	
SVC	Saturation		62	
IVC	Saturation		63	
MV O ₂			62.25	
RA	Pressure	14	9	9
RV	Pressure	82	6	14
MPA	Pressure	88	22	42
	Saturation		59	
	PaO ₂		25	
PCW(LA)	Pressure	12	11	10

PBF(Q _p) (L/min)	2.55
SBF(Q _s) (L/min)	2.79
Q _p /Q _s	0.92
Total PVR (Wood units)	12.54

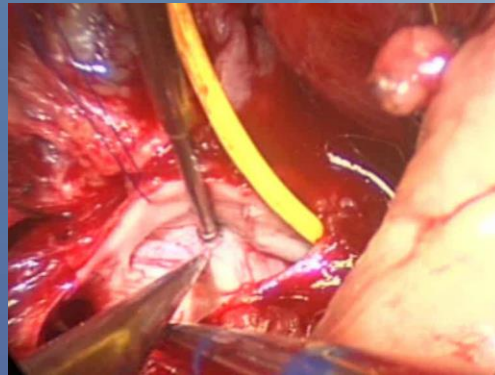
Treatment of CTEPH



2015 ESC/ERS guidelines for PH



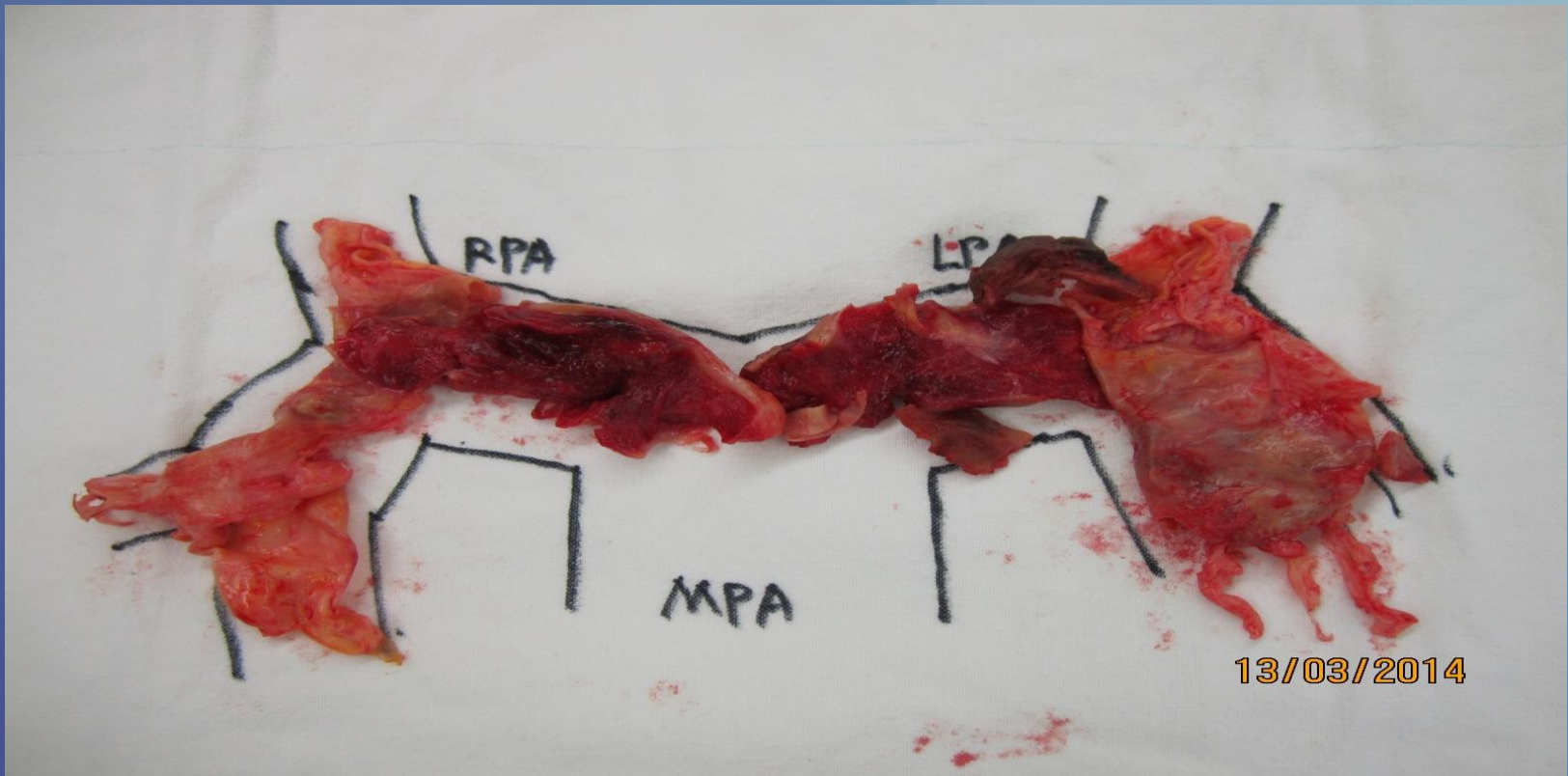
Pulmonary endarterectomy (PEA)



- 정중 흉골절개술
- 인공심폐기 통한 체외순환
- 저체온 (20° C)
- 심장 정지 (20 분)
- 폐동맥의 혈전 제거 및 내막 박리



Case 4: PEA

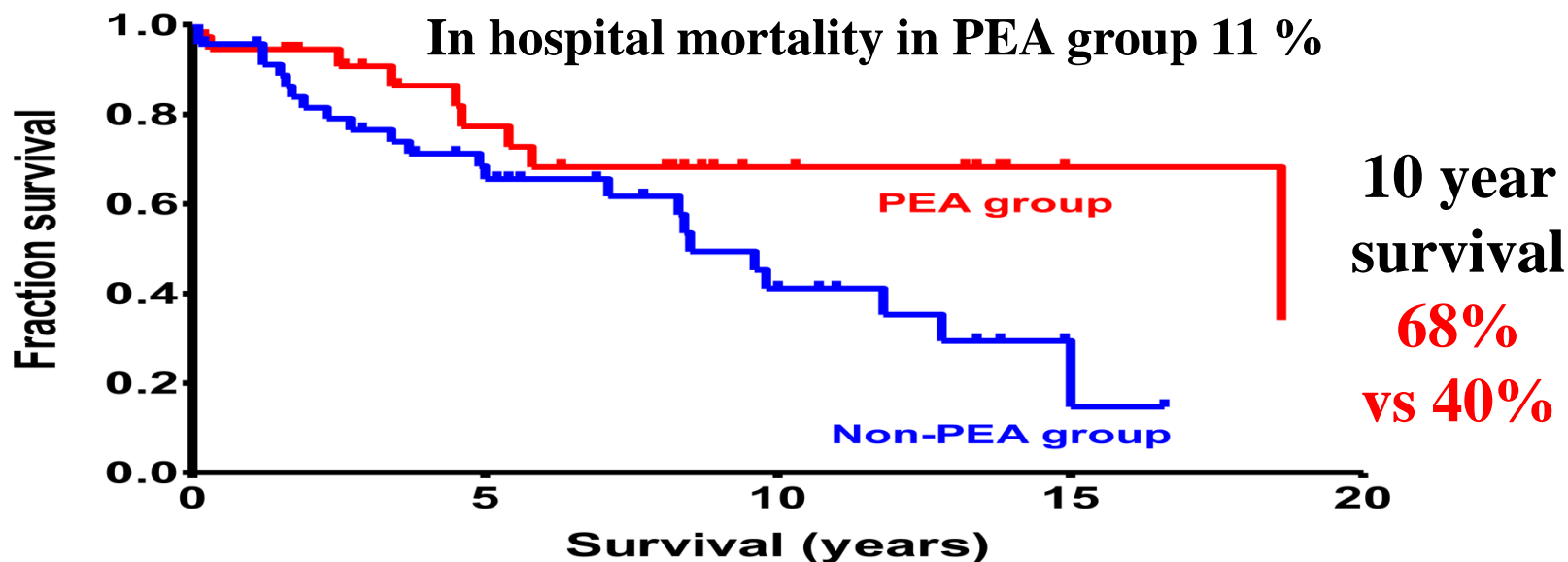


Tricuspid Valve mild tethering

TR grade	1	TR jet area	cm ²	Peak TR Vel	3.5 m/s
PGsys(RV-RA)	49 mmHg	Peak E Vel	cm/s	Peak A Vel	cm/s



Survival of CTEPH patients in AMC

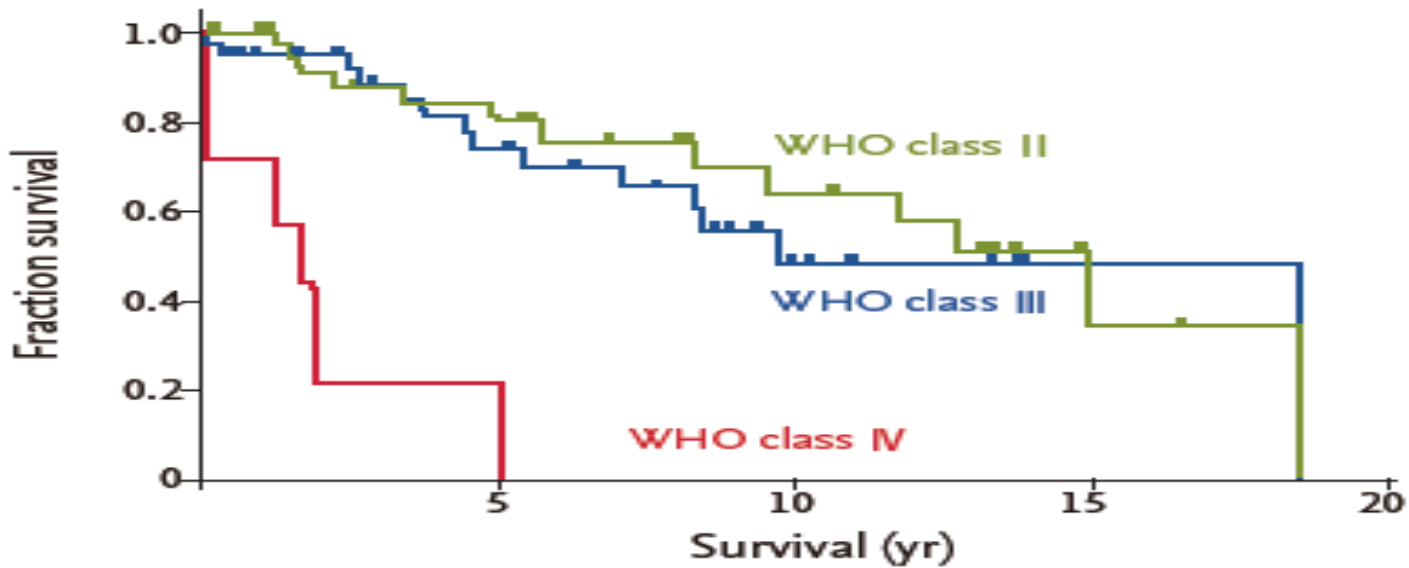


Number at risk	0yr	1yr	3yr	5yr	10yr	15yr	Death
PEA group	37	30	21	17	8	2	9 patients
Non-PEA group	51	42	29	23	9	1	24 patients

Kim et al. Korean J Intern Med 2016



Survival of CTEPH patients based on WHO FC in AMC



Number at risk	0yr	1yr	3yr	5yr	10yr	15yr	Death
WHO class II	40	35	24	20	11	3	12 patients
WHO class III	41	31	25	19	7	1	15 patients
WHO class IV	7	5	1	0			6 patients

Kim et al. Korean J Intern Med 2016



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Case 5. (55/F)

Chief complaint : dyspnea (WHO III) onset: 5 YA

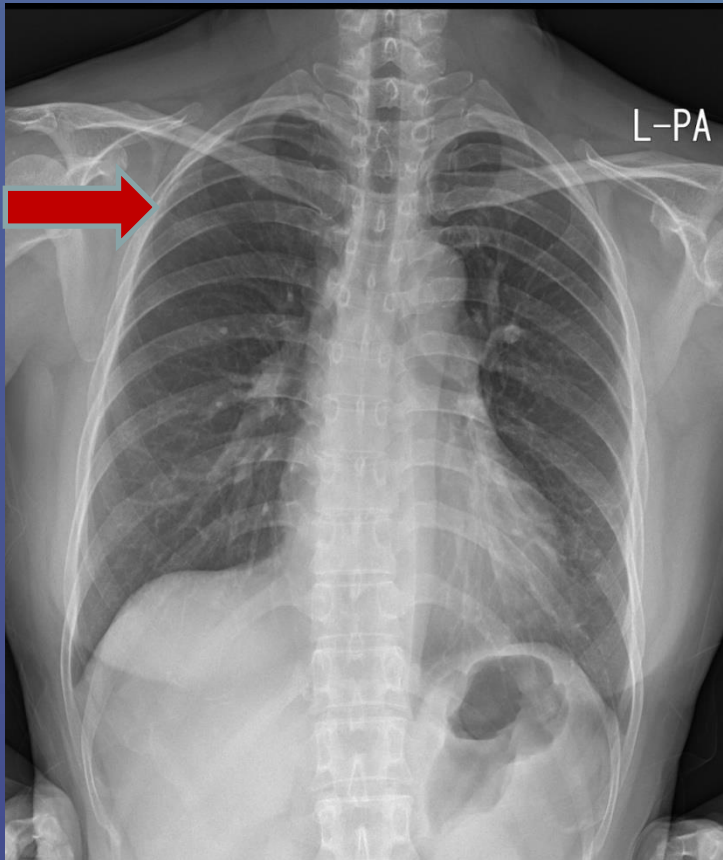
R.O. S.: dry mouth

P. M.Hx. : HTN

S. Hx: smoking (-)



Case 5 (55/F)

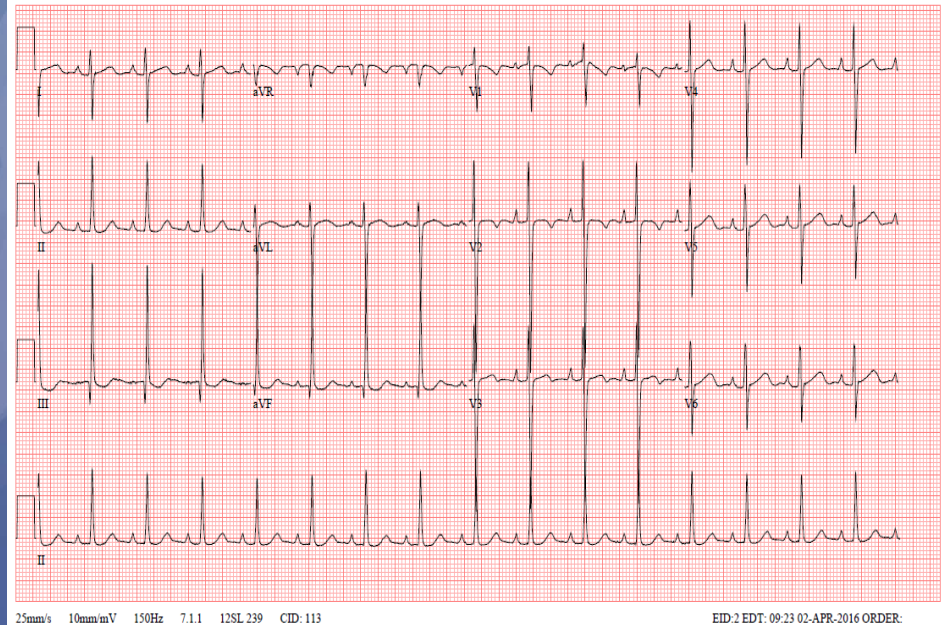


55 yr	Vent. rate	95	BPM	Normal sinus rhythm
Female	PR interval	156	ms	Right atrial enlargement
	QRS duration	96	ms	Rightward axis
Room:	QT/QTc	360/452	ms	Pulmonary disease pattern
Loc:113	P-R-T axes	26 103 22		Abnormal ECG

Technician: YSH
Test no:43650

Referred by: 124W

Confirmed By: GIBYOUNG NAM



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Case 5 (55/F)

SPIROMETRY

(S2002)

	(S2001)			Bronchodilator		
	Pred.	Meas.	%Pred.	Meas.	%Pred.	%Chg.
FVC	3.12	2.28	73	2.29	73	0
FEV ₁	2.48	1.70	69	1.76	71	4
FEV ₁ /FVC	79	75	95	77		
FEF _{25-75%}	2.50	1.34	54	1.66	66	24
PEF	5.43	6.08	112	5.96	110	-2

LUNG VOLUMES

(S2015)

	Pred	Meas.	%Pred.
TLC	4.44	3.48	78
VC	2.80	2.37	85
FRC	2.64	1.84	70
ERV	0.92	0.73	79
RV	1.72	1.11	65

DIFFUSING CAPACITY

	Pred.	Meas.	%Pred.
DL	18.2	11.3	62
VA(TLC)		3.09	
DL/VA	4.35	3.67	84
Corrected	18.2	11.5	63
Hb	Date 2016012 14.1 g/dl		

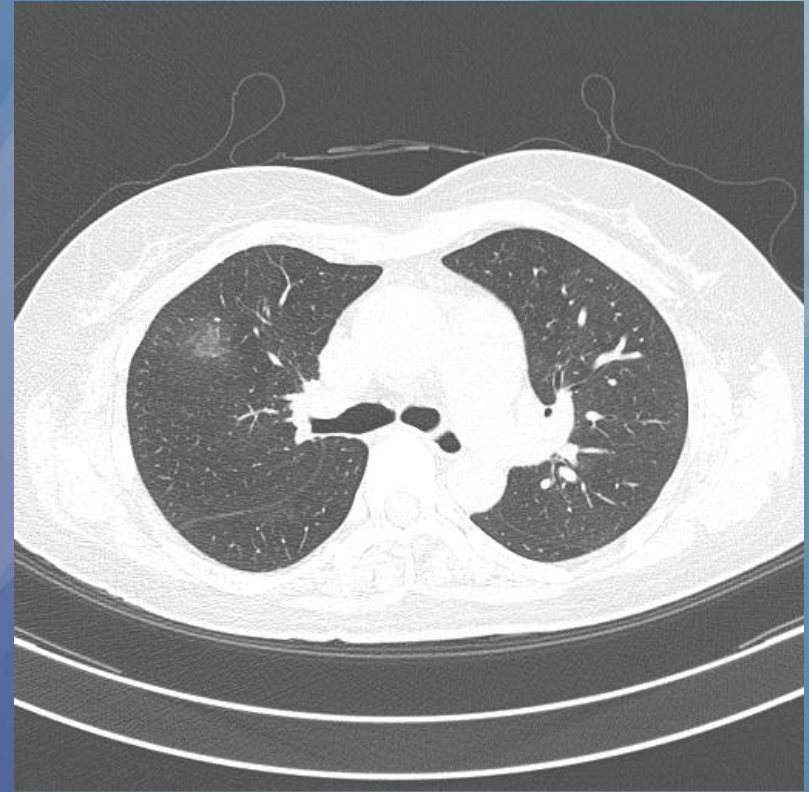
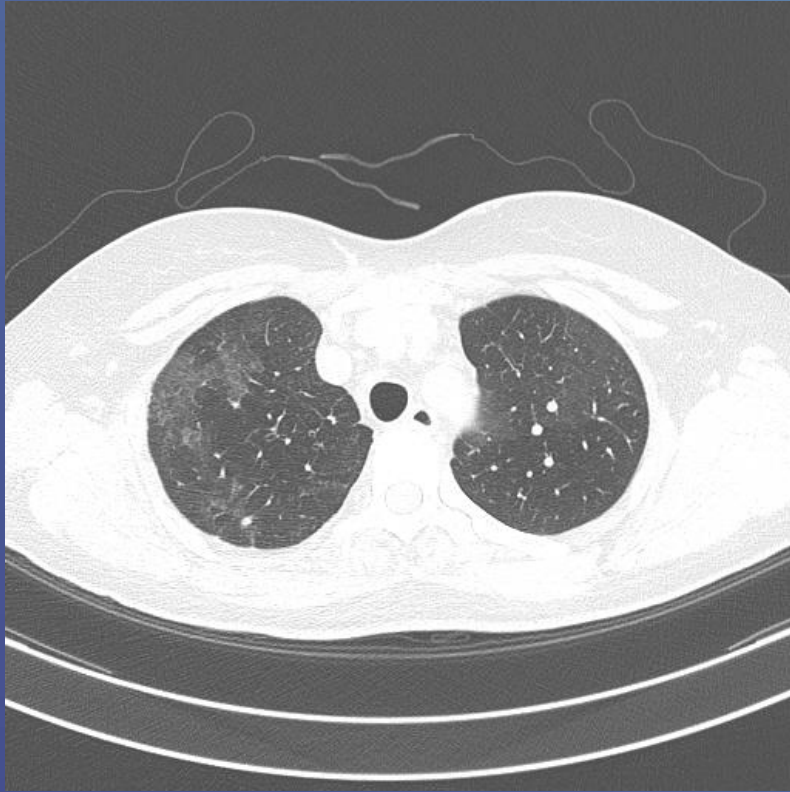
TEST(t) O₂

LPM

Pulse	SaO ₂	DISTANCE	RPE
108	96		
110	95		
112	95		
113	95		
115	94		
115	94		
119	94		
118	93		
117	92		
117	91		
118	90		
118	89		
117	88	335	5



Case 5 (55/F) HR chest CT



Multifocal ill-defined patchy ground-glass opacity, multiple small irregular nodules with partial calcification in right upper lobe.

Case 5 (55/F) Echocardiography

Left Ventricle D-shaped LV

LVIDs	21 mm	LVIDd	35 mm	LA	29 mm	Aorta	32 mm
LVPWs	15 mm	LVPWd	9 mm	ESV	10 mL	EDV	38 mL
IVSs	11 mm	IVSd	9 mm	LVEF	74 %	LV Mass	89.0 g
P.E. (ant)	mm	P.E. (post)	mm	LV Mass Index	57.8 g/m ²		

Right Ventricle decreased contractility

RVIDs	mm	RVIDd	mm
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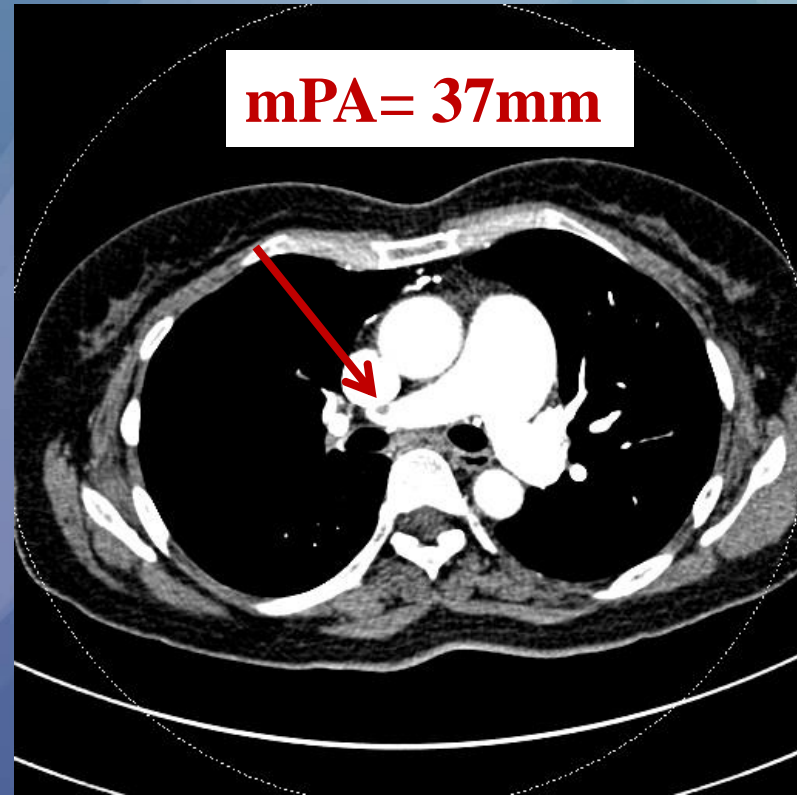
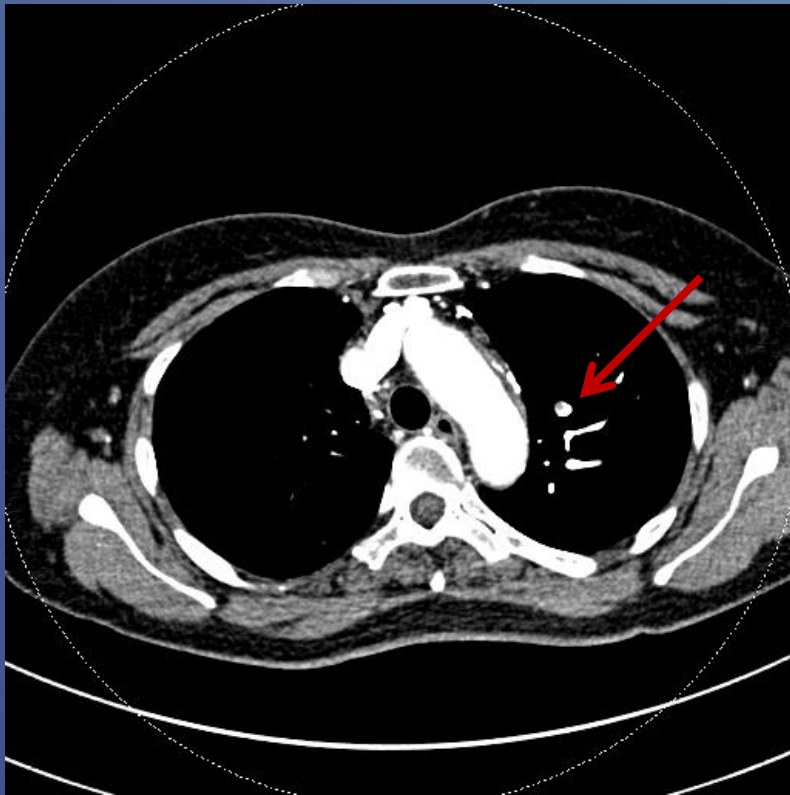
Tricuspid Valve incomplete closure

TR grade	3	TR jet area	11 cm ²	Peak TR Vel	5.4 m/s
PGsys(RV-RA)	117 mmHg	Peak E Vel	cm/s	Peak A Vel	cm/s

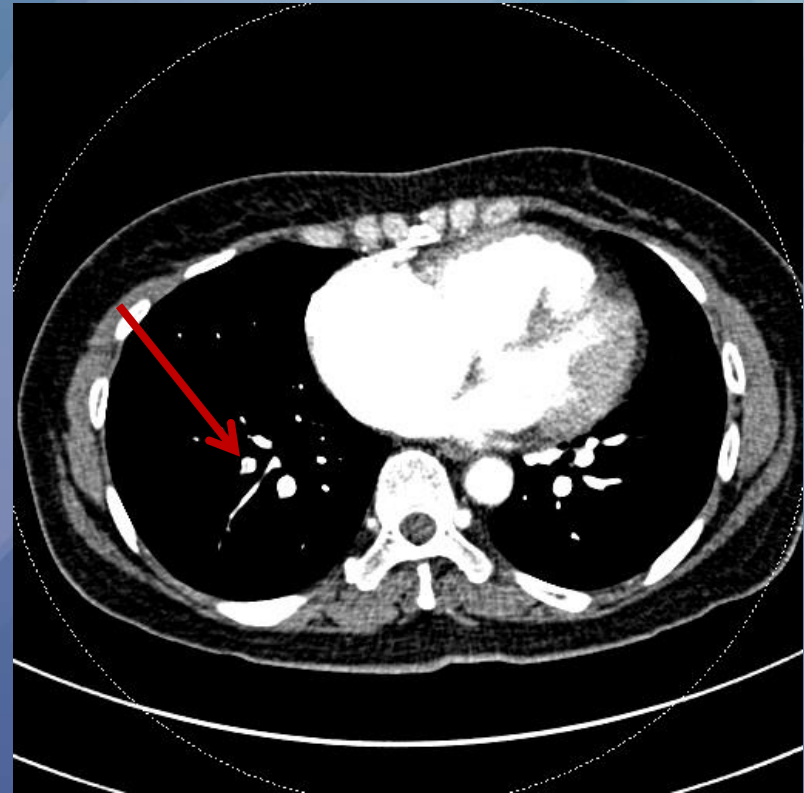
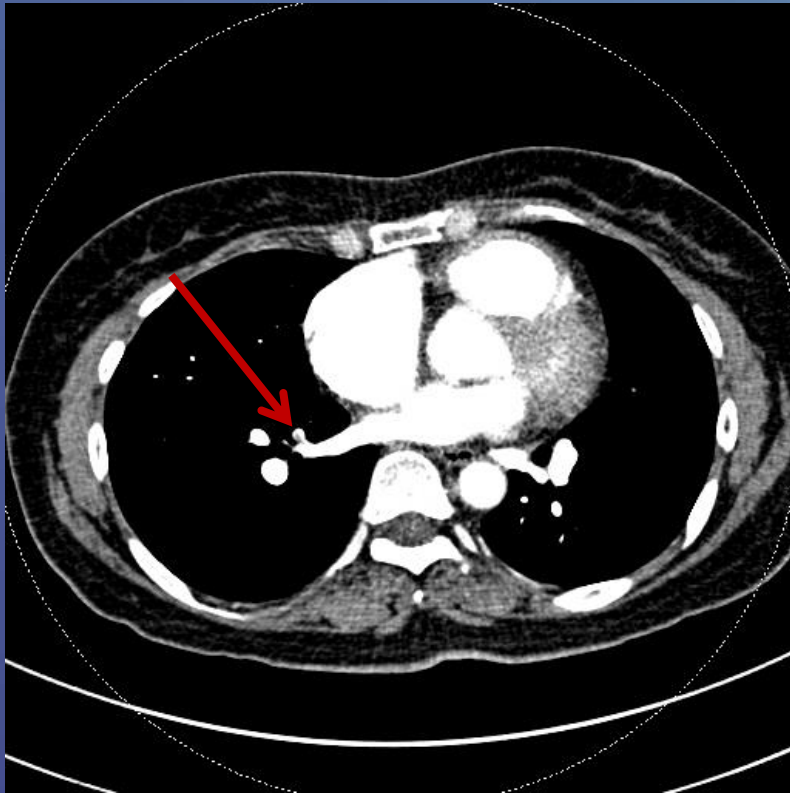
Enlarged RV and thickened RV wall (7mm) with RV dysfunction and severe resting pulmonary hypertension, C/W chronic RV pressure overloading



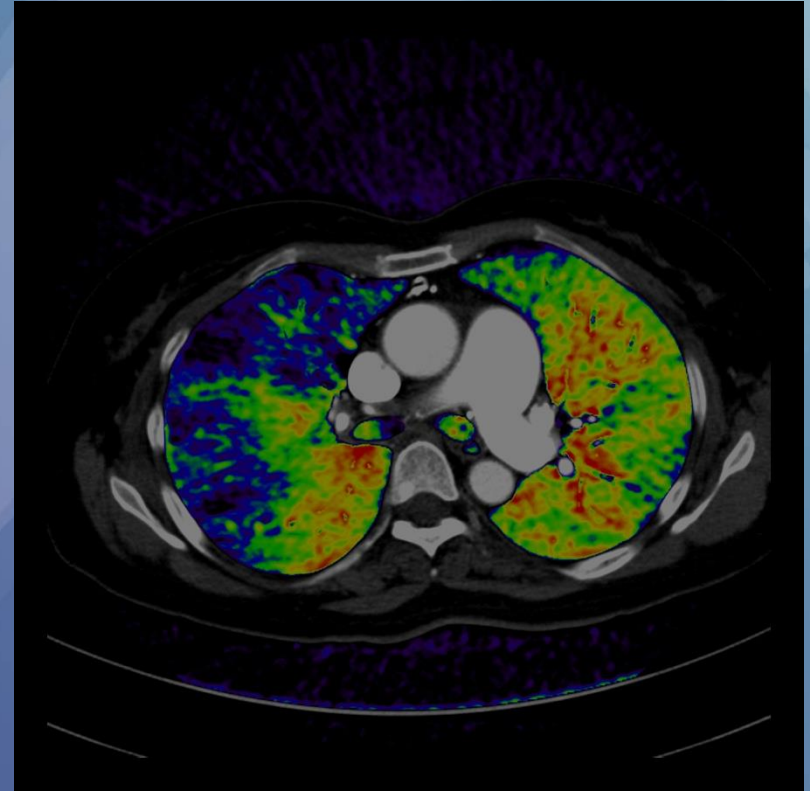
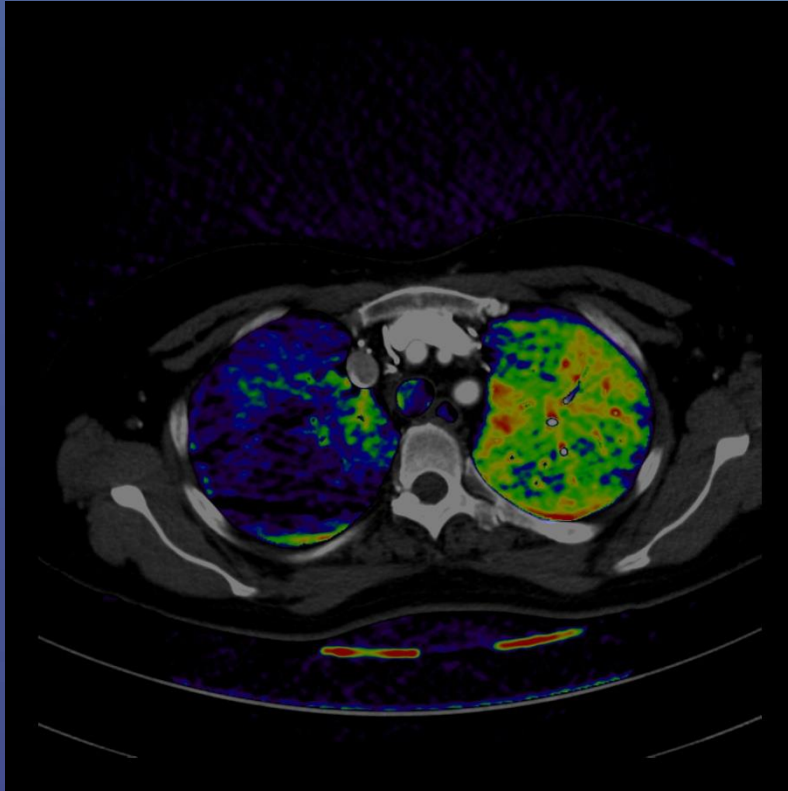
Case 5 (55/F) Chest CT



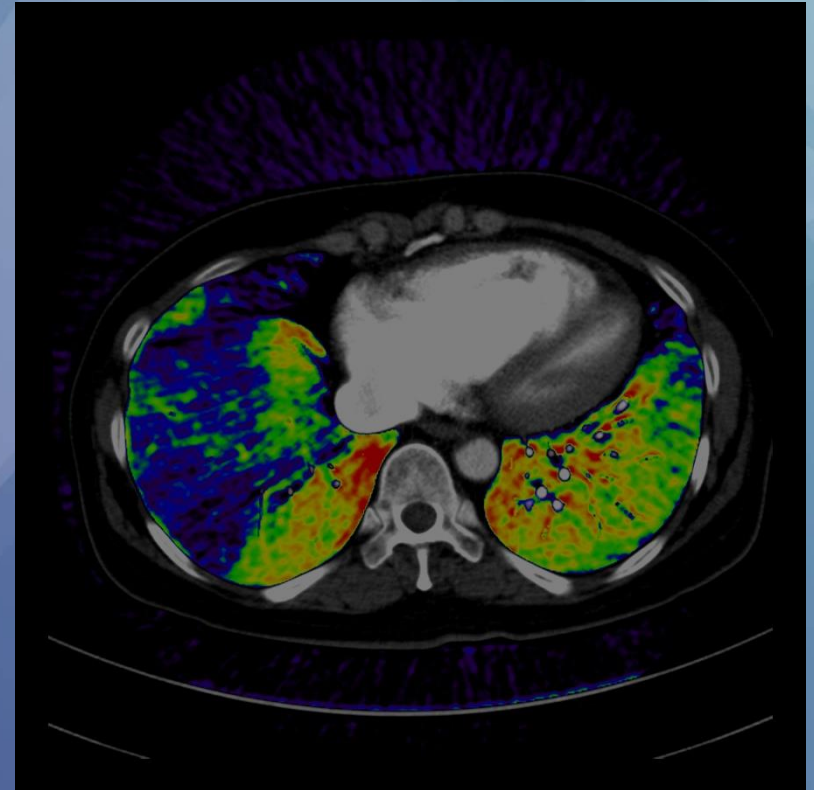
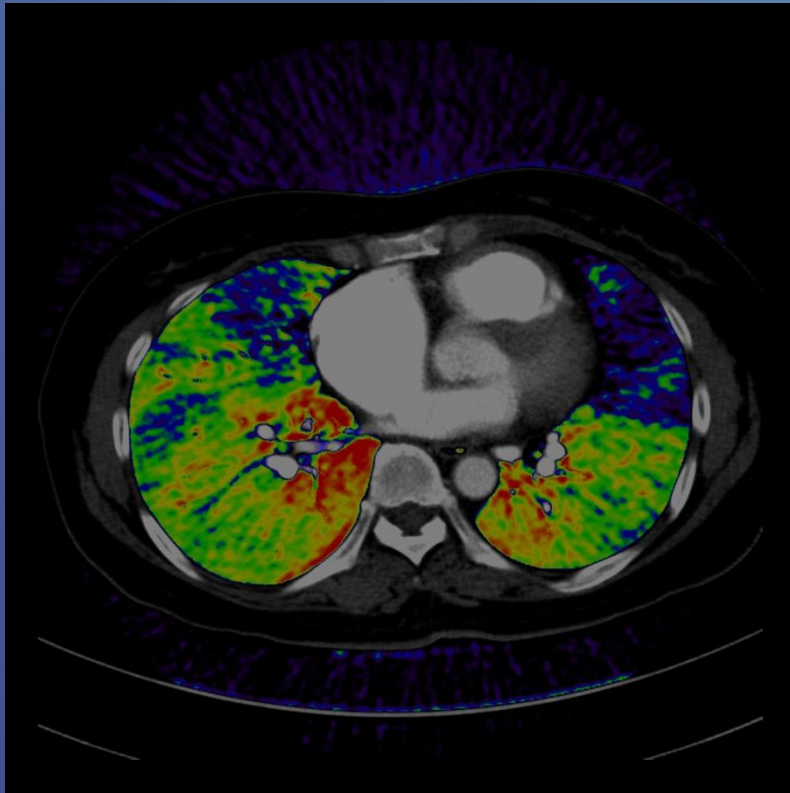
Case 5 (55/F) Chest CT



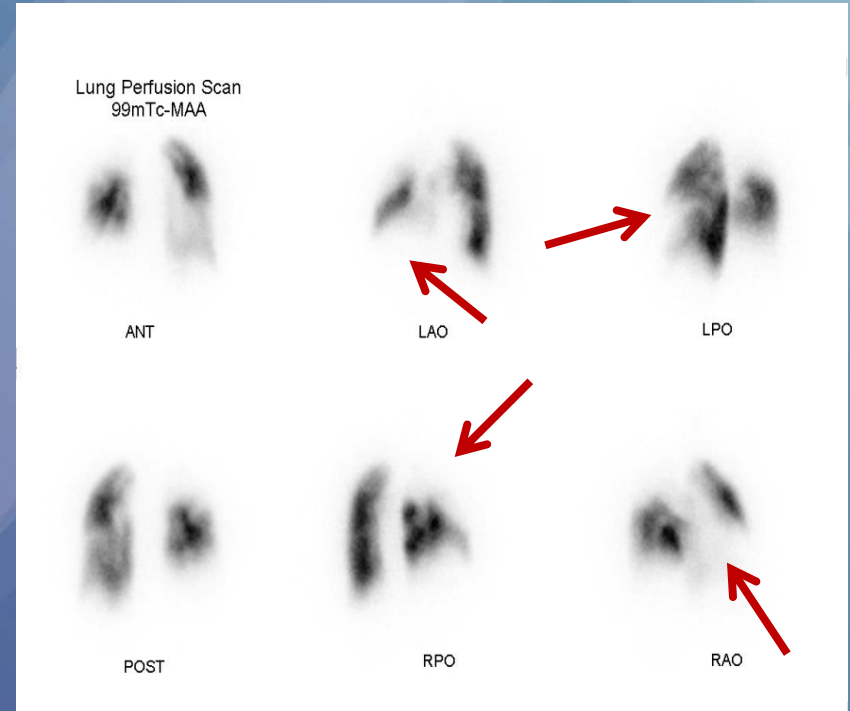
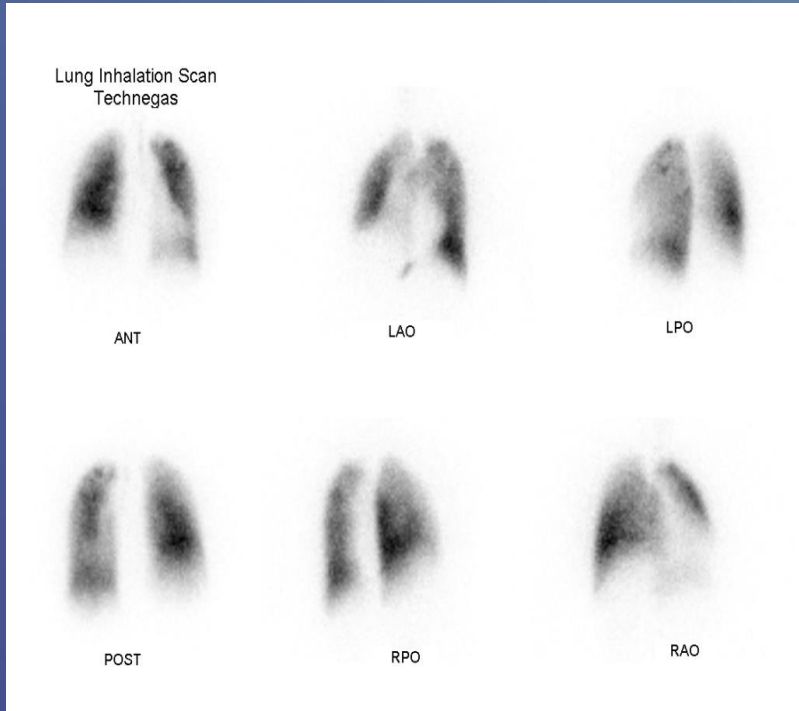
Case 5 (55/F) Chest CT perfusion



Case 5 (55/F) Chest CT perfusion

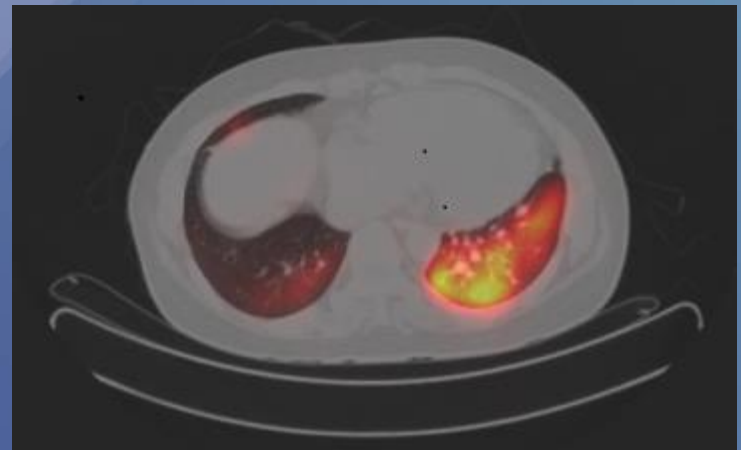
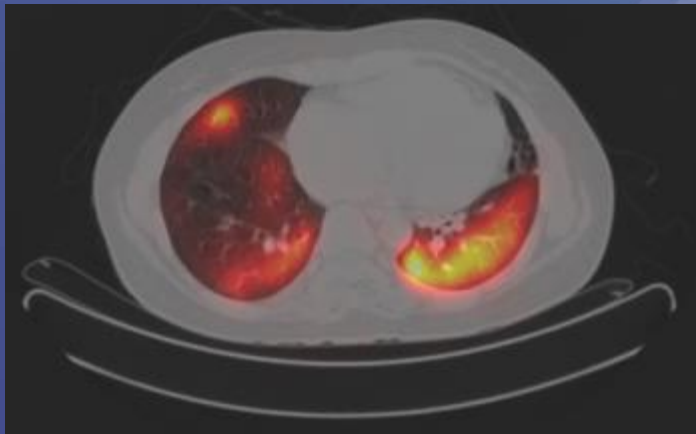
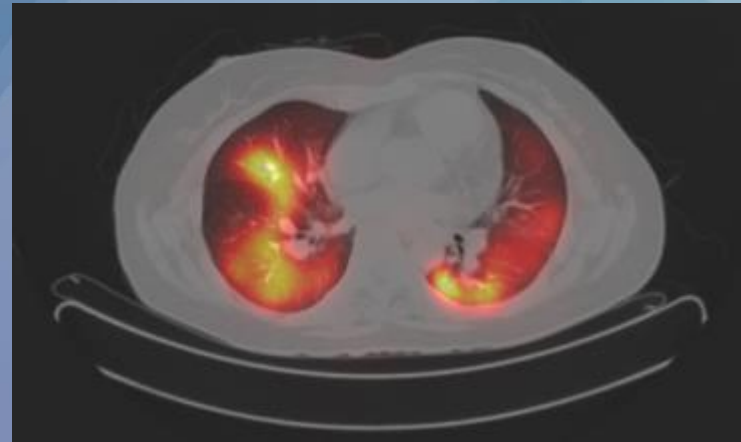
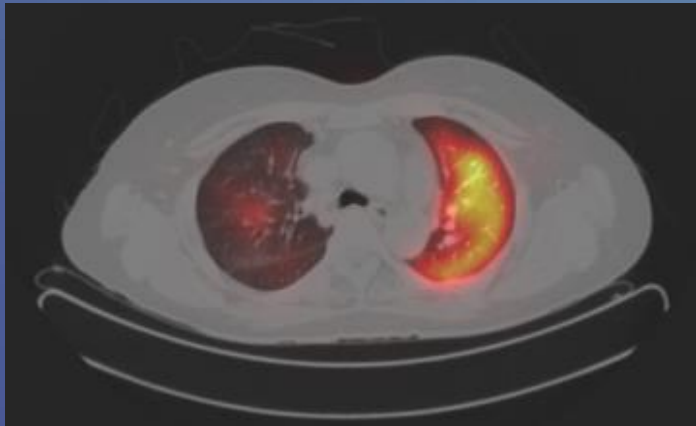


Case 5 (55/F) V/Q scan



Case 5: Fusion perfusion SPECT/CT

6 months after anticoagulation



Case 5: RHC with Pulmonary angiography

RHC

mRA 5 mmHg

sPAP/dPAP/mPAP

91/23/48 mmHg

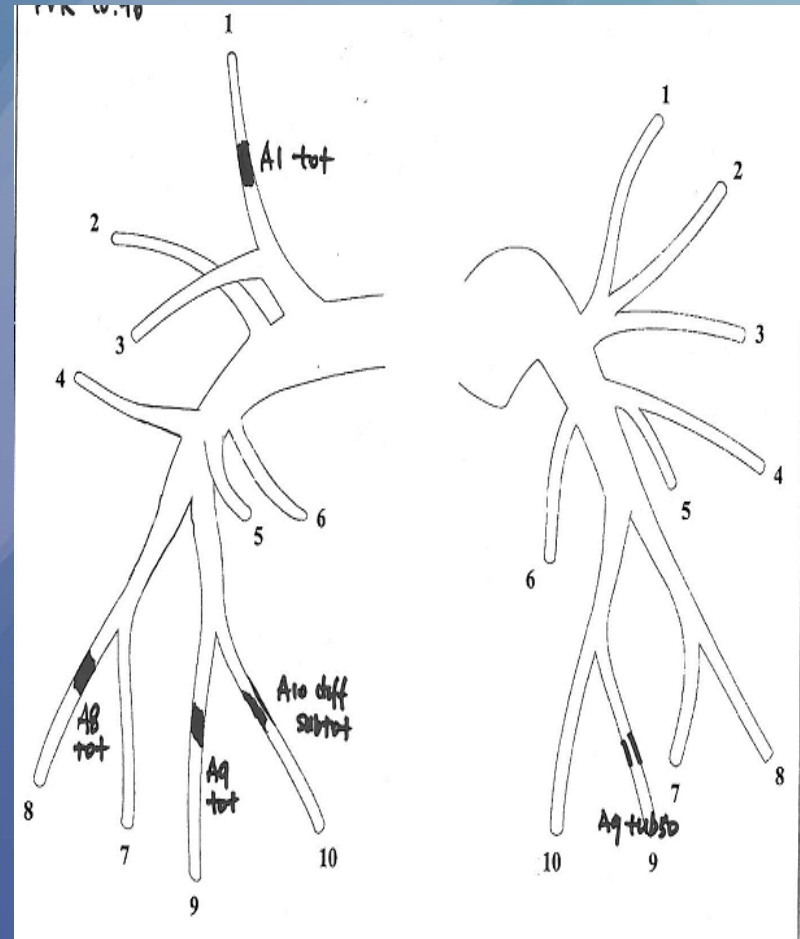
PCWP 5 mmHg

CO 3.88 L/m

CI 2.56 L/min/m²

PVR 11 Wood units

Mixed SvO₂ 67%



Case 5: Balloon pulmonary angioplasty (BPA)



Pulmonary artery intervention in AMC

Case 6. 51/M Stonecutter

C. C> : DOE (WHO Fc II)

PFT: FEV1/FVC 64% FEV1 78% pred. FVC 96% pred.
DLco 91 % pred.

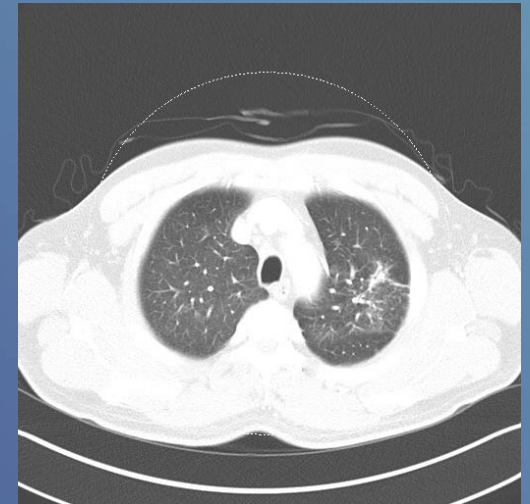
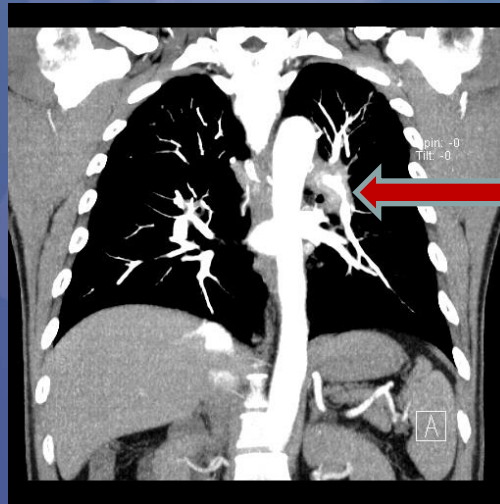
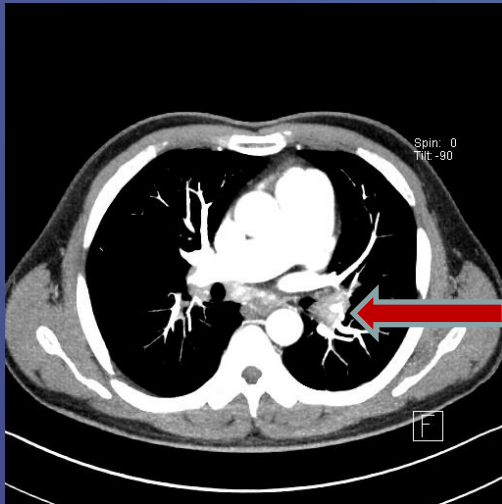
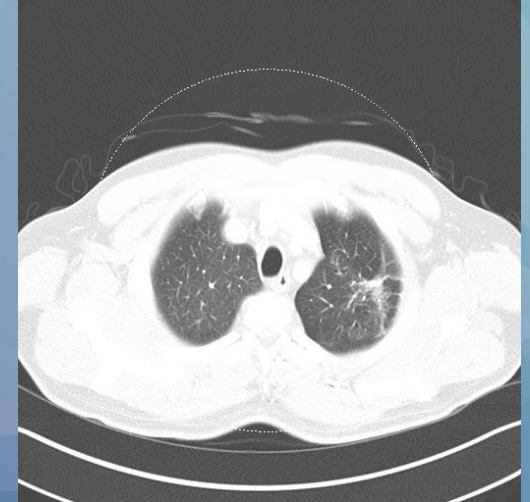
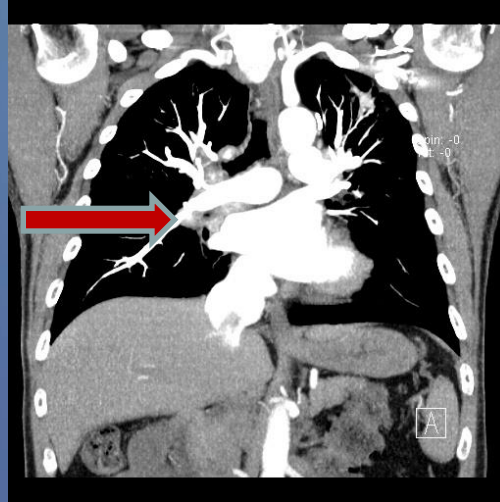
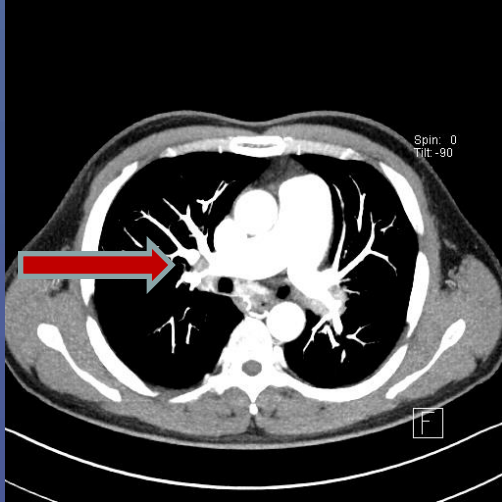
6MWD 490m (SaO2 94%)

Echo : **TR Vmax 3.9 m/s, PG(RV-RA) 61 mmHg**
-> severe resting PH with RV dysfunction

RHC: **sPAP/dPAP/mPAP 54/20/32 mmHg** PCWP 8 mmHg
CO 4.23 L/m PVR 4.78 WU

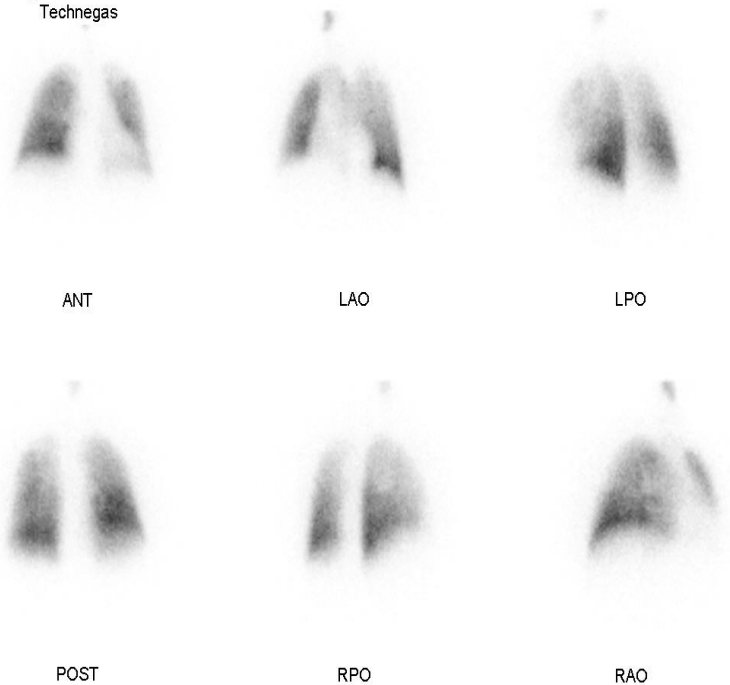


Case 6; Chest CT



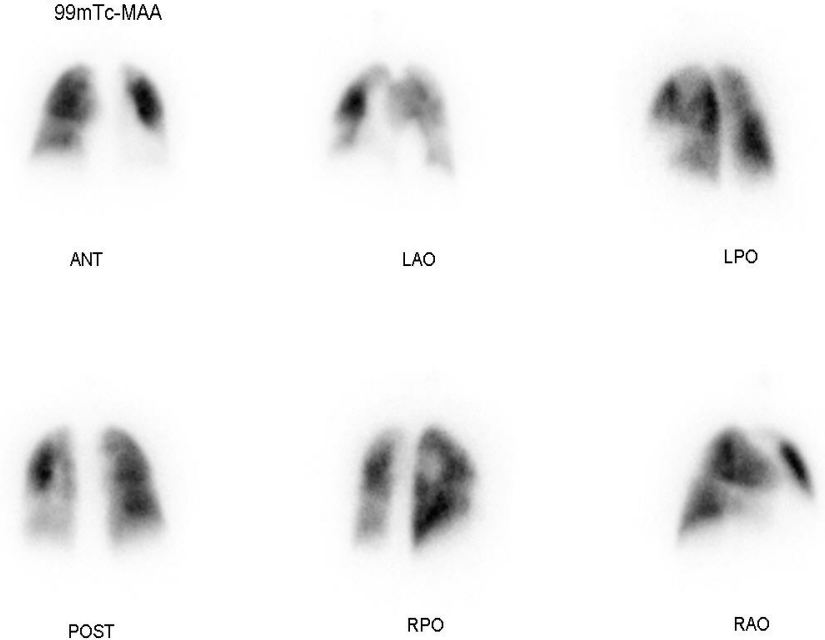
Case 6; V/Q scan

Lung Inhalation Scan
Technegas

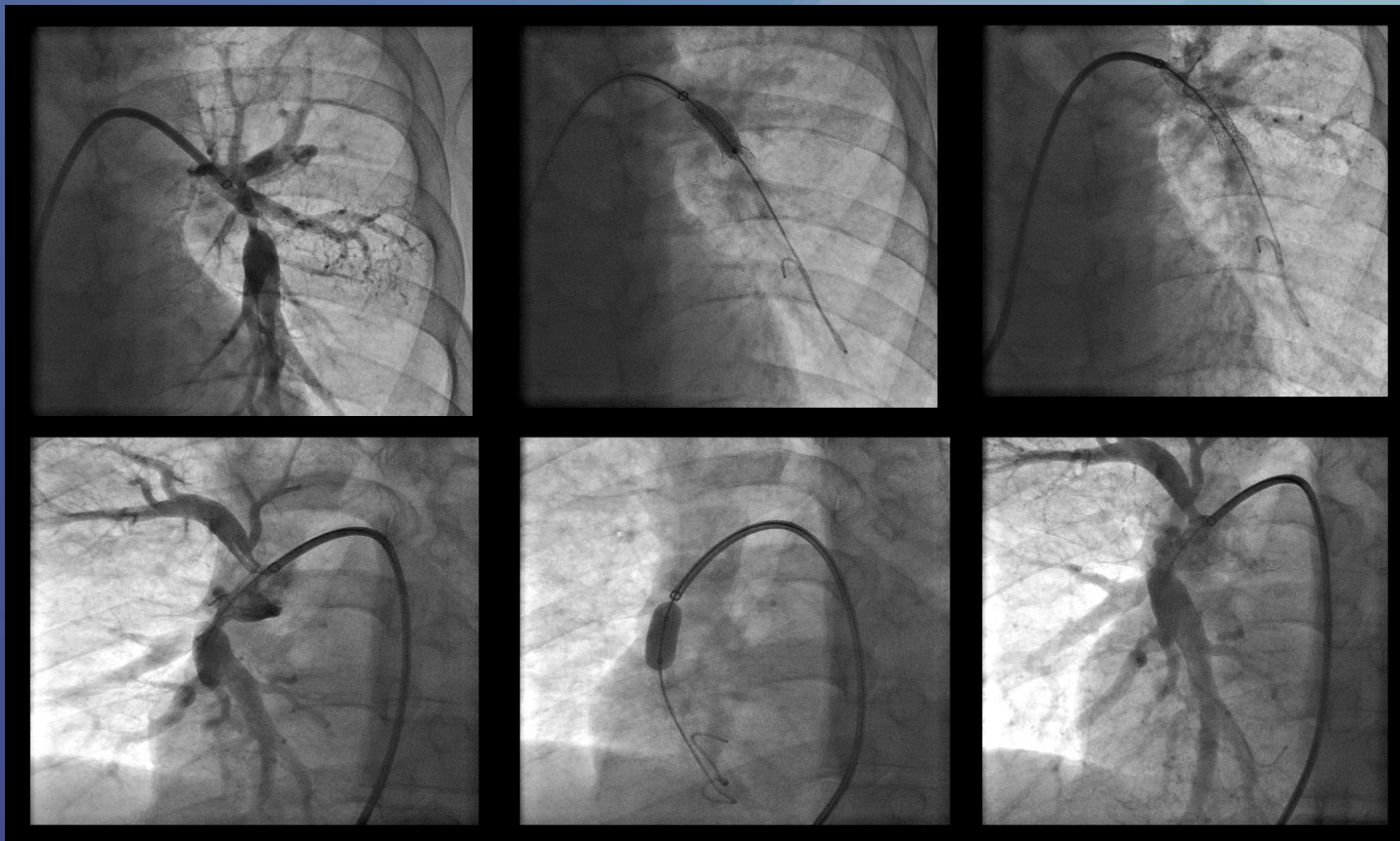


P

Lung Perfusion Scan
99mTc-MAA



Case 6. Pulmonary artery balloon dilatation and stent insertion



Case 7 (F/55)

C.C.: DOE (Fc II)

P. M. Hx: Takayasu's arteritis

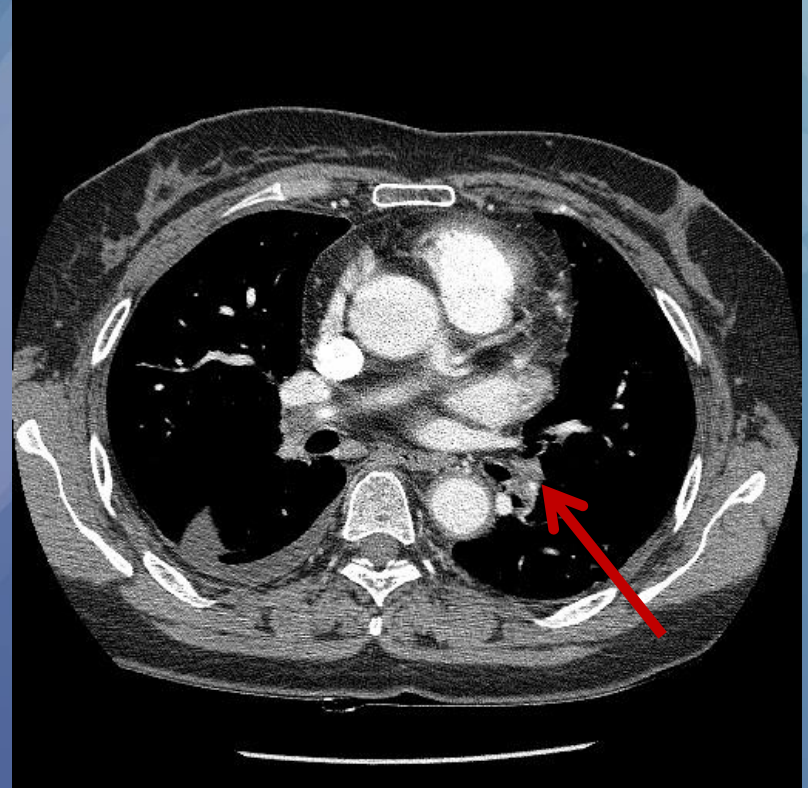
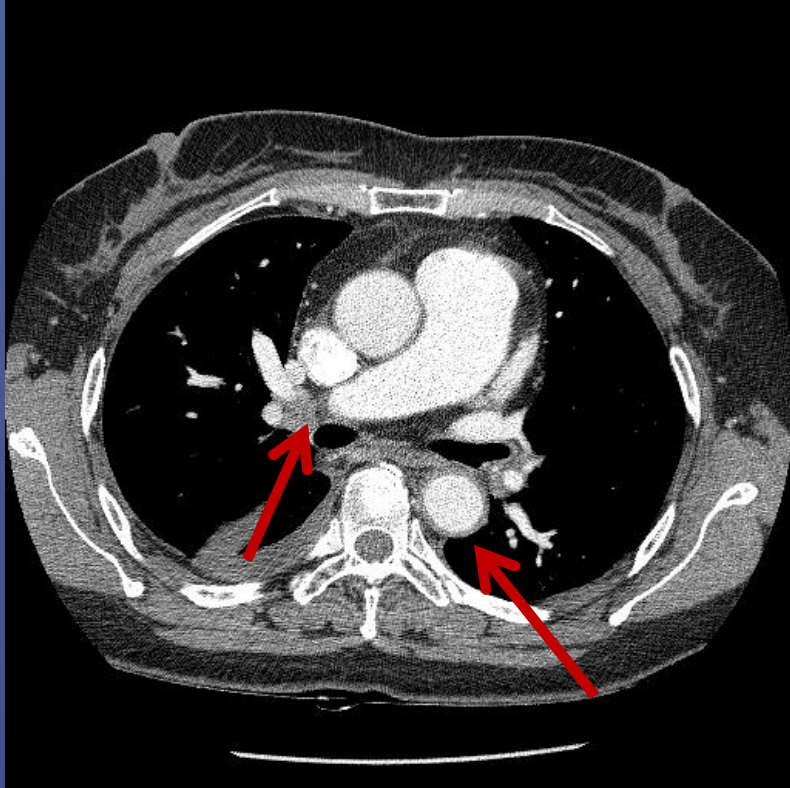
**Echo : TR Vmax 3.0 m/s, PG(RV-RA) 36 mmHg
-> borderline PH**

**PFT: FEV1/FVC 77% FEV1 2.11 L (76% pred.) FVC 2.73 (79% pred.)
TLC 3.74 L (76% pred.)**

Dlco 45 % pred.

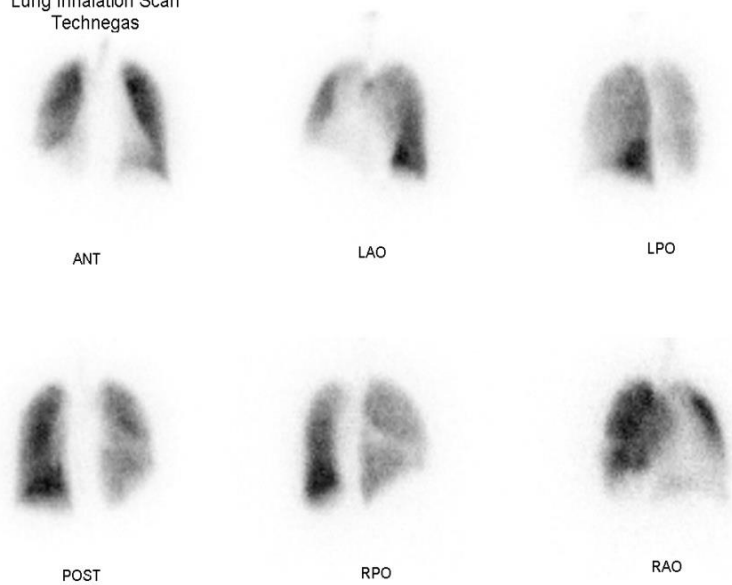


Case 7; Chest CT

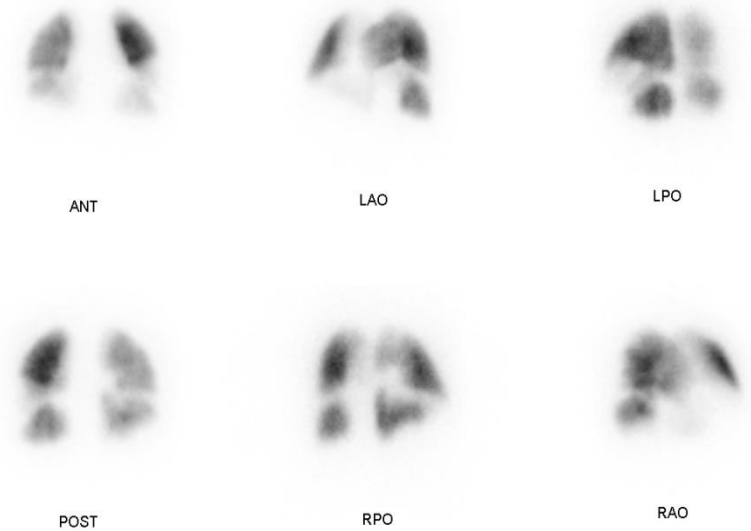


Case 7 (F/55) V/Q scan

Lung Inhalation Scan
Technegas



Lung Perfusion Scan
99mTc-MAA



Case 7 (F/55) RHC and PA

RHC

mRA 8 mmHg

sPAP/dPAP/mPAP

60/18/33 mmHg

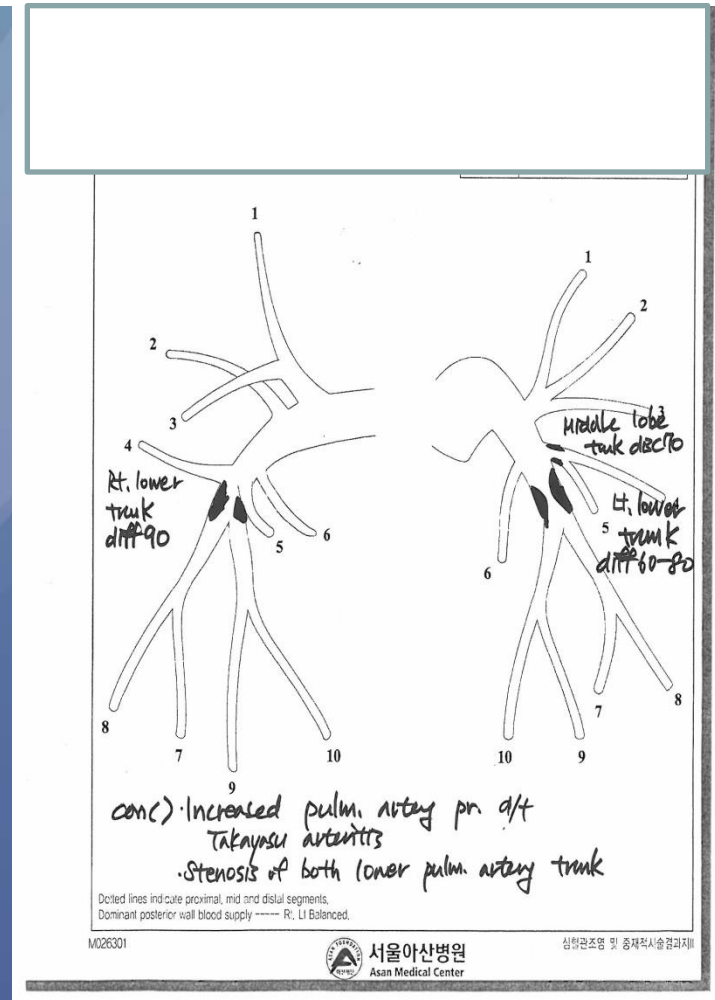
PCWP 14 mmHg

CO 6.01 L/m

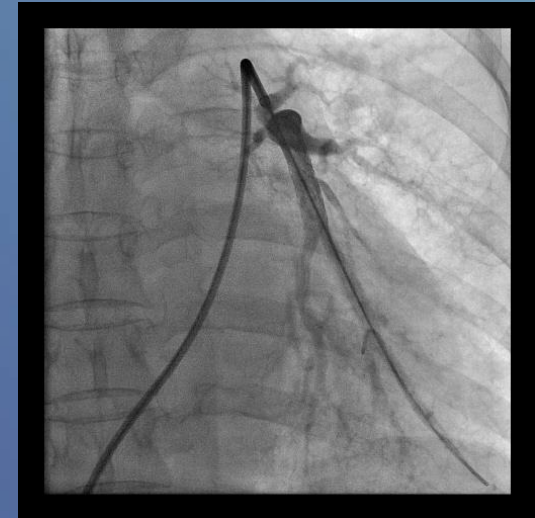
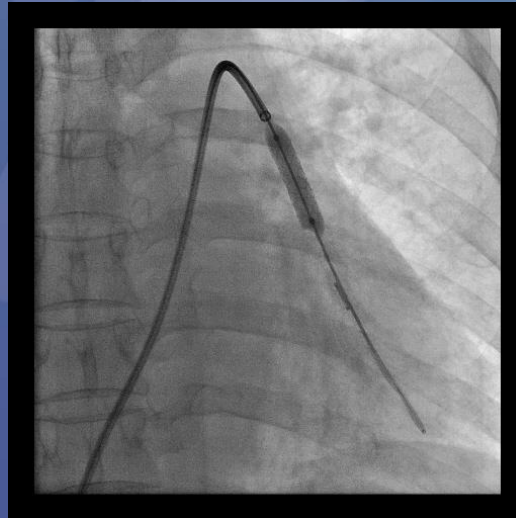
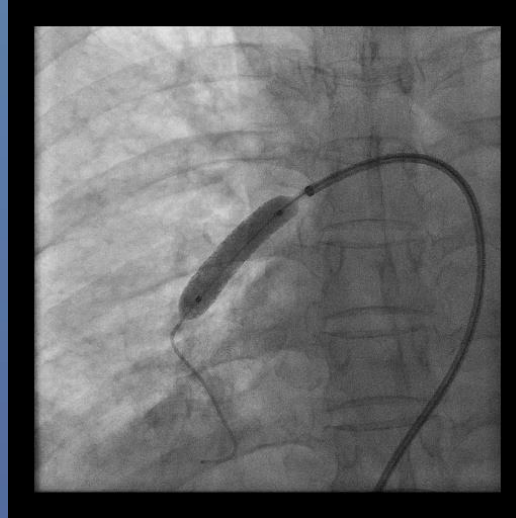
CI 3.52 L/min/m²

PVR 3.50 Wood units

Mixed SvO₂ 76%



Case 6. Pulmonary artery balloon dilatation and stent insertion



Case 6. 6MWT pre-and post- stenting

TEST(I) O2		LPM		TIME
Pulse	SaO ₂	DISTANCE	RPE	
80	95			resting
				—
				test
92	93			30"
94	91			1'
103	87			1'30"
106	86			2'
107	84			2'30"
107	85			3'
110	84			3'30"
111	85			4'
110	84			4'30"
107	84			5'
106	85			5'30"
105	85	375	4	6'
Comment				

TEST(I) O2		LPM		TIME
Pulse	SaO ₂	DISTANCE	RPE	
66	98			resting
				—
				test
87	95			30"
98	94			1'
109	91			1'30"
110	90			2'
115	90			2'30"
116	88			3'
121	88			3'30"
122	88			4'
124	88			4'30"
125	88			5'
127	87			5'30"
128	86	543	4	6'
Comment				

