

PAH cases

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Center
Pulmonary Hypertension & Venous Thrombosis Center
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Case 1 (F/30)

C. C. : DOE (WHO FC III)

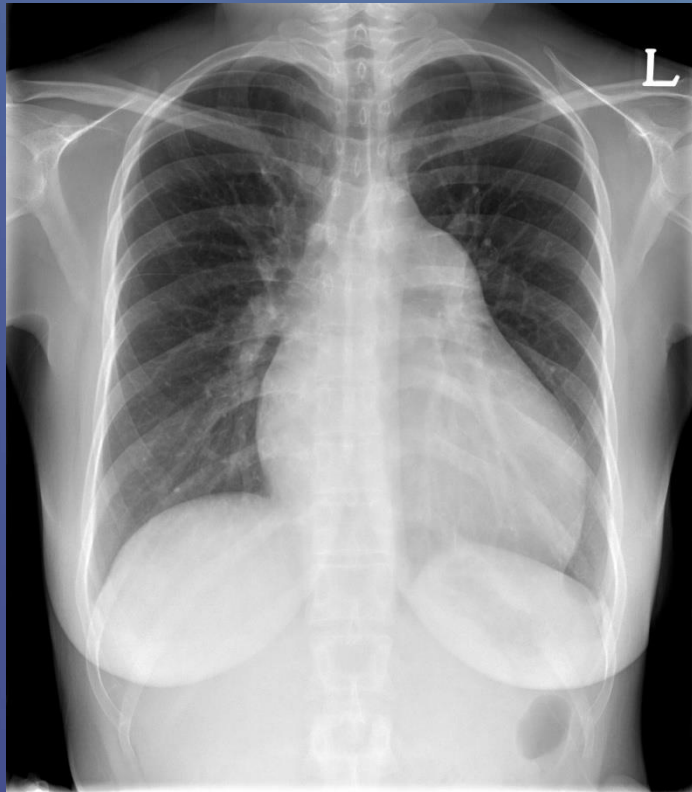
P. I: 약 2개월 전부터 약 200m 정도 걸으면 호흡곤란 및 가슴 답답한 증상
1차례 syncope 발생한 후 조금만 걸어도 호흡곤란 발생

- P. M. Hx. : Hypothyroidism으로 Synthroid 복용
- F. Hx: None
- S. Hx.: nonsmoker

- P/Ex: BP 120/100 mmHg PR 96/m RR 20/m

Case 1 (F/30) Chest PA/lateral

2012.07



Case 1 (F/30) ECG

30 yr
Female
Room:
Loc:118

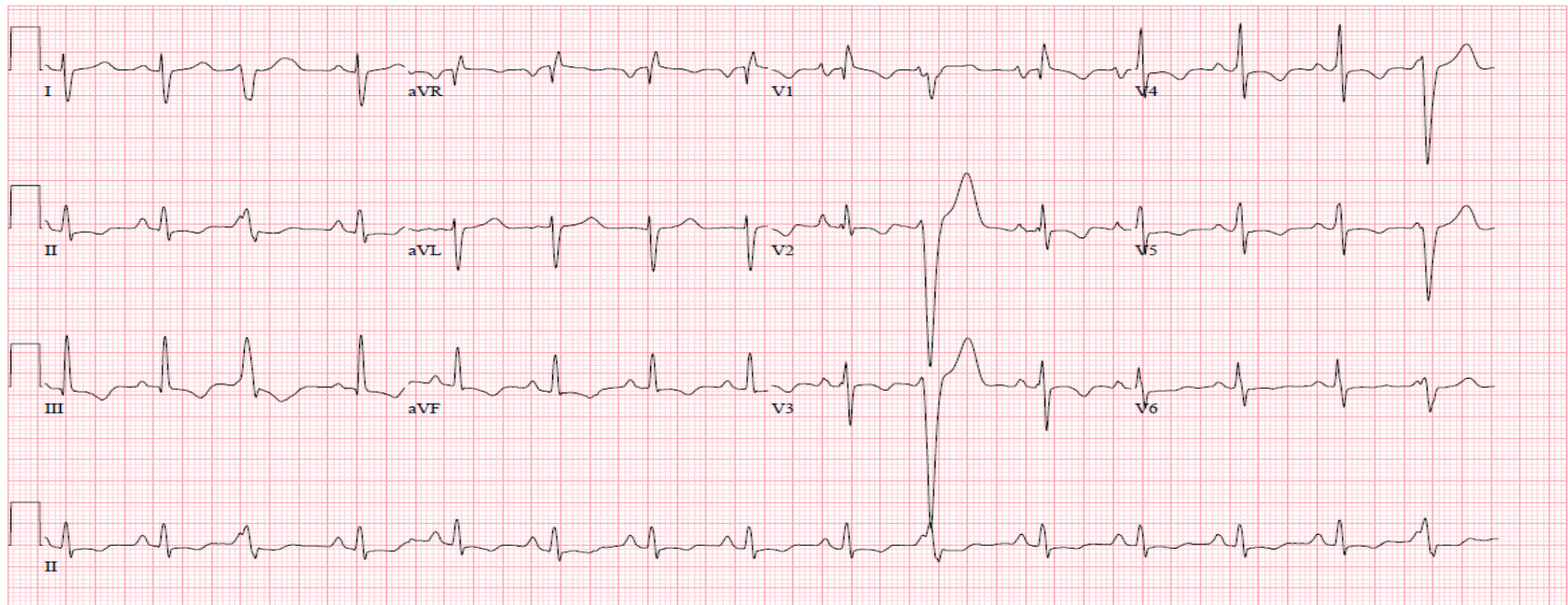
| | | |
|--------------|---------|-----|
| Vent. rate | 90 | BPM |
| PR interval | 156 | ms |
| QRS duration | 106 | ms |
| QT/QTc | 428/523 | ms |
| P-R-T axes | 55 121 | -30 |

Sinus rhythm with frequent Premature ventricular complexes
Possible Left atrial enlargement
Right axis deviation
Incomplete right bundle branch block
Right ventricular hypertrophy
ST & T wave abnormality, consider inferior ischemia
ST & T wave abnormality, consider anterolateral ischemia
Prolonged QT
Abnormal ECG

Technician: KEY
Test ind:95080

Referred by: CV

Confirmed By: GI BYOUNG NAM



25mm/s 10mm/mV 40Hz 7.1.1 12SL 239 CID: 118

EID:2 EDT: 17:07 25-JUN-2012 ORDER:

Page 1 of 1

Case 1 (F/40)

Lab.: CBC 6000-14.0-144K

BUN/Cr 13/0.7 mg/dL Uric acid 6.0 mg/dL

Protein/albumin 5.4/3.0 g/dL AST/ALT 36/28 IU/L

T.B 2.0 mg/dL

Na/K/Cl/CO2 138/3.9/105/28 mmol/L

BNP 898 pg/mL

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

FANA (-) RF factor (-)

TSH/free T4 1.9 uU/mL (0.4-5.0)/1.9 ng/dL (0.8-1.9)

Case 1 (F/40) Echocardiography

Left Ventricle D-shaped LV

| | | | | | | | |
|------------|--------------|-------------|--------------|---------------|------------------------------|---------|----------------|
| LVIDs | 27 mm | LVIDd | 44 mm | LA | 33 mm | Aorta | 27 mm |
| LVPWs | 17 mm | LVPWd | 10 mm | ESV | 13 ml | EDV | 42 ml |
| IVSs | 14 mm | IVSd | 11 mm | LVEF | 69 % | LV Mass | 159.1 g |
| P.E. (ant) | mm | P.E. (post) | 6 mm | LV Mass Index | 98.6 g/m ² | | |

Right Ventricle dilated RV size and decreased RV contractility(8 mm)

[2012/07/02 - 2D+Doppler Echocardiography]

Conclusion)

1. Markedly dilated RV and severe RV dysfunction with severe resting PH
2. Minimal pericardial effusion

| | | | | | |
|----------------------|-------------------|--------------|--------|-------------|-----------------|
| DTI S/E'A' (lateral) | / / cm/s | MR grade | | MR jet area | cm ² |
| MVA(2D/PHT) | / cm ² | PG(max/mean) | / mmHg | PISA | mm |

Tricuspid Valve tethering

| | | | | | |
|--------------|----------------|-------------|---------------------------|-------------|----------------|
| TR grade | 4 | TR jet area | 18 cm ² | Peak TR Vel | 4.4 m/s |
| PGsys(RV-RA) | 77 mmHg | Peak E Vel | cm/s | Peak A Vel | cm/s |

Pulmonic Valve

| | | | | | |
|----------|----------|--------------|----------------|-------------------|----------------|
| PR grade | 2 | PR Peak Vel | 3.8 m/s | PR ED Vel | 2.6 m/s |
| Peak Vel | m/s | PG(max/mean) | / mmHg | Pul vein flow S/D | / cm/s |

Case 1 (F/40) PFT

SPIROMETRY

(S2001)

| | Pred. | Meas. | %Pred. |
|-----------------------|-------|-------|--------|
| FVC | 3.68 | 3.12 | 85 |
| FEV ₁ | 3.17 | 2.66 | 84 |
| FEV ₁ /FVC | 87 | 85 | 98 |
| FEF _{25-75%} | 3.46 | 3.04 | 88 |
| PEF | 6.45 | 7.41 | 115 |

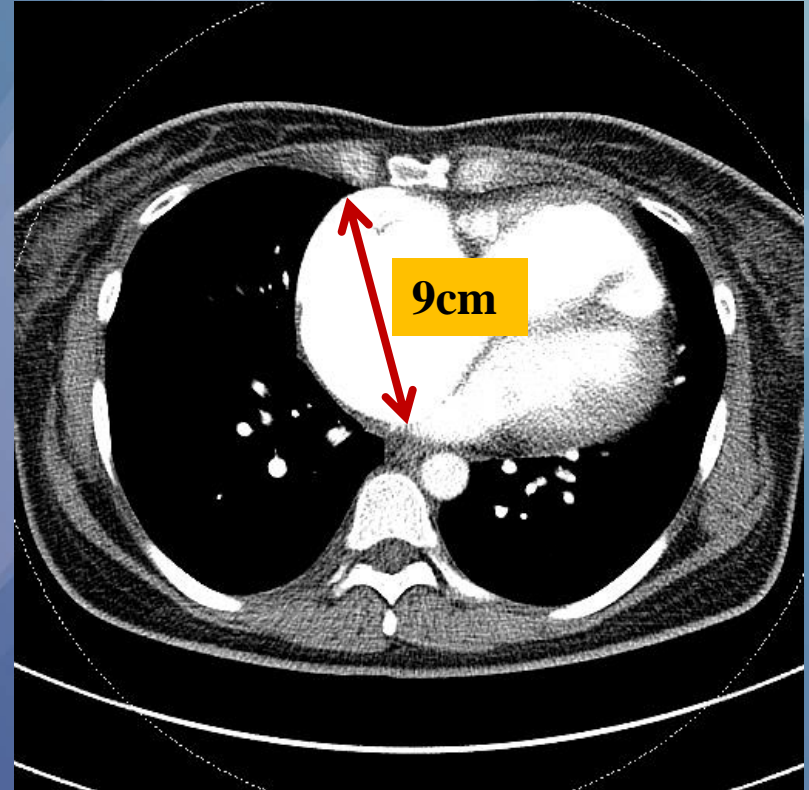
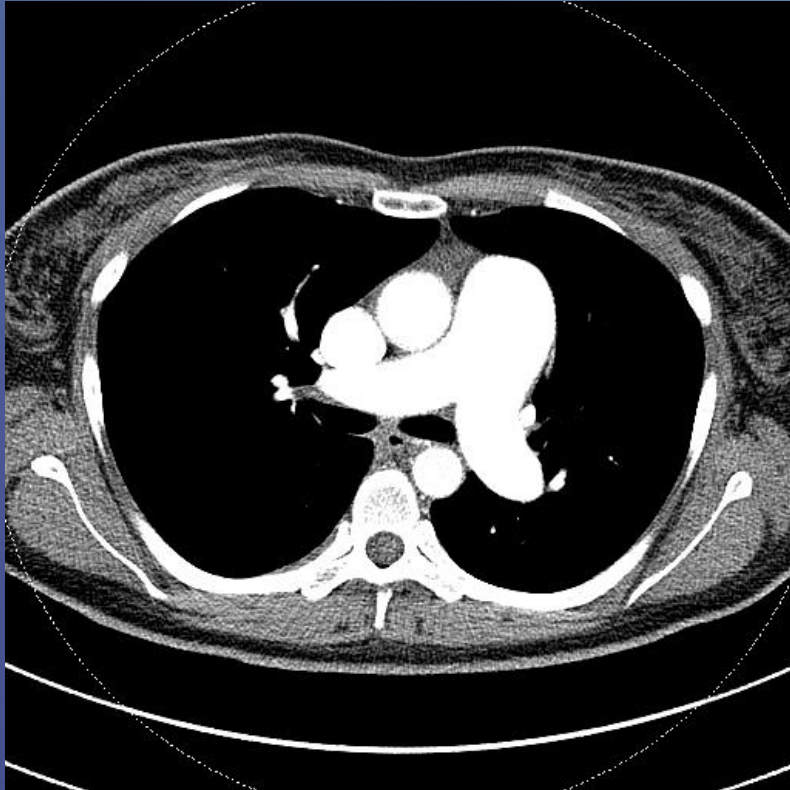
DIFFUSING CAPACITY

| | Pred. | Meas. | %Pred. |
|-----------|---------------------------|-------|--------|
| DL | 23.9 | 15.1 | 63 |
| VA(TLC) | | 4.14 | |
| DL/VA | 5.01 | 3.65 | 73 |
| Corrected | 23.9 | 15.4 | 64 |
| Hb | Date 2012070 14.0 g/dl | | |

6-minute Walk Distance Test

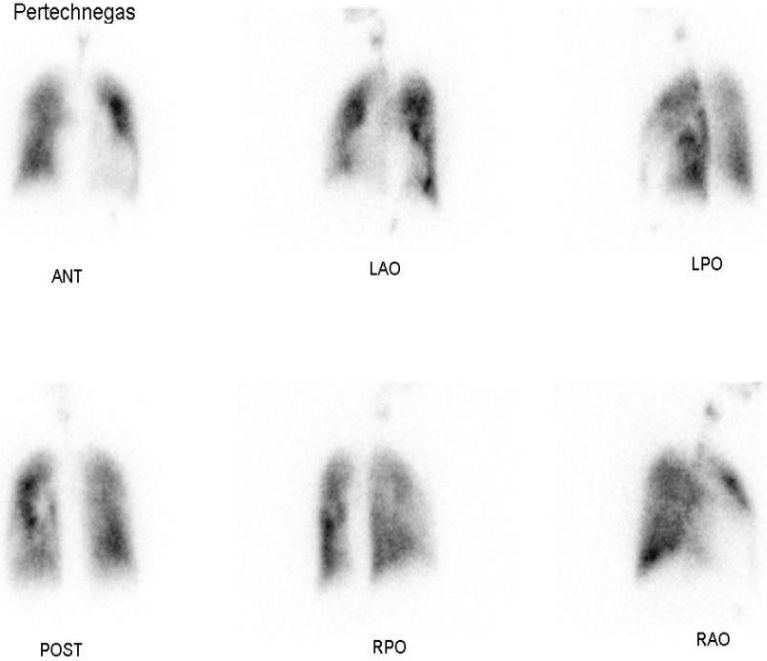
| 진료구분 | 입원 | | |
|---------|------------------|----------|-----|
| 상병명 | | | |
| | TEST(I) O2 | LPM | |
| Pulse | SaO ₂ | DISTANCE | RPE |
| 73 | 99 | | |
| | | | |
| 90 | 98 | | |
| 97 | 99 | | |
| 100 | 99 | | |
| 107 | 99 | | |
| 110 | 98 | | |
| 111 | 97 | | |
| 115 | 95 | | |
| 120 | 96 | | |
| 124 | 96 | | |
| 125 | 96 | | |
| 112 | 97 | | |
| 98 | 97 | 240 | 10 |
| Comment | 5분에서 머지러워서 휴식 | | |

Case 1; chest CT

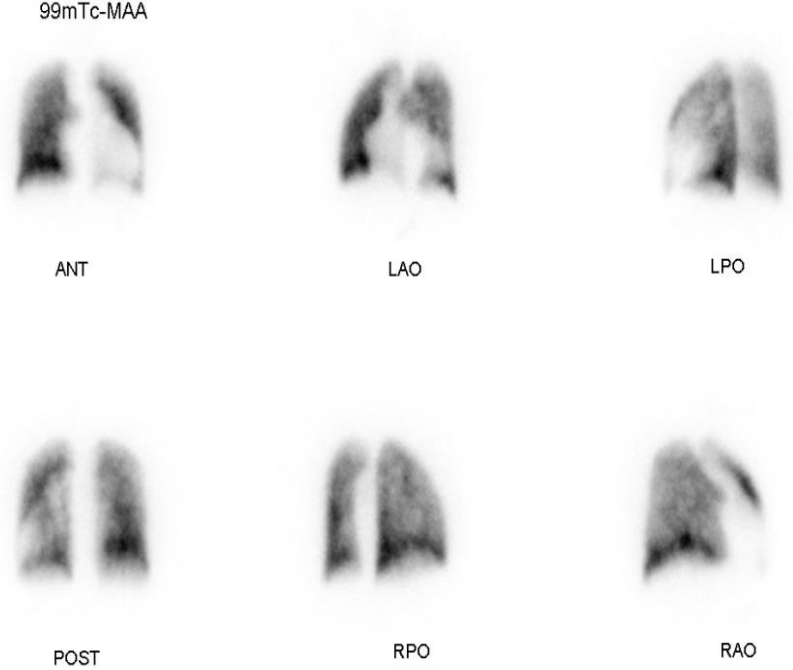


Case 1; V/Q scan

Lung Inhalation Scan
Pertechnegas



Lung Perfusion Scan
99mTc-MAA



Case 1; RHC & acute vasodilator test

| | Baseline | NO 10ppm | NO 20ppm | NO 40ppm |
|----------------------|-------------|-------------|-------------|-------------|
| BP | 124/91 | 118/64 | 124/74 | 97/52 |
| sPAP/dPAP | 87/45 | 95/42 | 95/44 | 95/44 |
| Mean PAP | 61 | 61 | 61 | 61 |
| PCWP | 18 | | | |
| mRAP | 13mmHg | | | |
| CO | 2.8 | 2.8 | 2.8 | 2.8 |
| CI | 1.7 | 1.7 | 1.7 | 1.7 |
| PVR (WU) | 15.1 | 17.6 | 17.9 | 17.9 |
| SvO ₂ (%) | | | | |

Case 1; Diagnosis & treatment

- **Assessment: IPAH, Acute vasoreactivity test (-), High risk**
- **Tx: Initial combination treatment**
(ambrisentan 5mg Qd, sildenafil 25mg tid)
Warfarin

3-month after ambrisentan+sildenafil

| | 2012.07 | 2012.11 |
|---------------------|----------------|----------------|
| WHO FC | III | II |
| TRVmax | 4.4 | |
| PGsys(RV-RA) | 77 | |
| BNP | 898 | 570 |
| 6MWD | 240 | 430 |

12-months after treatment

S: dyspnea (Fc III- IV), leg edema, fatigue, Syncope (+)

| | 2012.07 | 2012.11 | 2013.08 |
|--------------------------|------------|------------|------------|
| WHO FC | III | II | III |
| TRVm ax | 4.4 | | 4.6 |
| PGsys(RV-RA) | 77 | | 85 |
| BNP | 898 | 570 | 709 |
| 6MWD | 240 | 430 | 395 |

- Tx: Volibris + sildenafil -> bosentan + sildenafil**

3-months after bosentan and sildenafil

| | 2013.08 | 2014.02 |
|---------------------|------------|------------|
| WHO FC | III | III |
| TRVmax | 4.6 | 4.5 |
| PGsys(RV-RA) | 85 | 81 |
| BNP | 709 | 430 |
| 6MWD | 395 | 405 |

1-year after bosentan and sildenafil

| | Baseline (2012.07) | After 2 year (2014.12) |
|-----------|-----------------------|---------------------------|
| BP | 124/91 | 103/74 |
| sPAP/dPAP | 87/45 | 91/37 |
| Mean PAP | 61 | 55 |
| PCWP | 18 | 7 |
| mRAP | 13 | 6 |
| CO | 2.8 | 2.6 |
| CI | 1.7 | 1.5 |
| PVR (WU) | 15.1 | 18.4 |
| ScVO2 | 57% | 62% |

6-month after macitentan and sildenafil

| | 2014.12 | 2015.06 |
|---------------------|------------|------------|
| WHO FC | III | II |
| TRVmax | 4.5 | 3.2 |
| PGsys(RV-RA) | 81 | 41 |
| BNP | 430 | 422 |
| 6MWD | 405 | 420 |

6-month after macitentan and sildenafil

| | 2014.12 | 2015.06 |
|------------------------|--------------|--------------|
| BP | 103/74 | 94/67 |
| sPAP/dPAP | 91/37 | 80/27 |
| Mean PAP | 55 | 48 |
| PCWP | 7 | 7 |
| mRAP | 6 | |
| CO | 2.6 | 4.2 |
| CI | 1.5 | 2.5 |
| PVR (WU) | 18.4 | 9.7 |
| SvO₂ | 62% | 72% |

2-year after macitentan and sildenafil

| | 2015.06 | 2016.02 | 2017.05 |
|---------------------|------------|------------|------------|
| WHO FC | II | II | II |
| TRVmax | 3.2 | | 3.8 |
| PGsys(RV-RA) | 41 | | 58 |
| BNP | 422 | | |
| NT-proBNP | | 653 | 822 |
| 6MWD | 420 | 445 | 450 |

2.5-year after macitentan and sildenafil

| | 2015.06 | 2017.11 |
|------------------------|--------------|--------------|
| BP | 94/67 | 100/62 |
| sPAP/dPAP | 80/27 | 88/33 |
| Mean PAP | 48 | 51 |
| PCWP | 7 | 10 |
| CO | 4.2 | 4.3 |
| CI | 2.5 | 2.6 |
| PVR (WU) | 9.7 | 9.4 |
| SvO₂ | 72% | 70% |

Case 2. (72/M)

Chief complaint : dyspnea (WHO FC III)

Present illness : Dyspnea for 10 days
Syncope on 2 days ago

P. M. Hx: Stomach cancer and renal cell carcinoma
-> **s/p Distal gastrectomy and radical nephrectomy**

S. Hx. : smoking (-)

P/Ex.: **BP 94/52 mmHg PR 110 /m RR 22/m BMI 24**

Chest PA and EKG

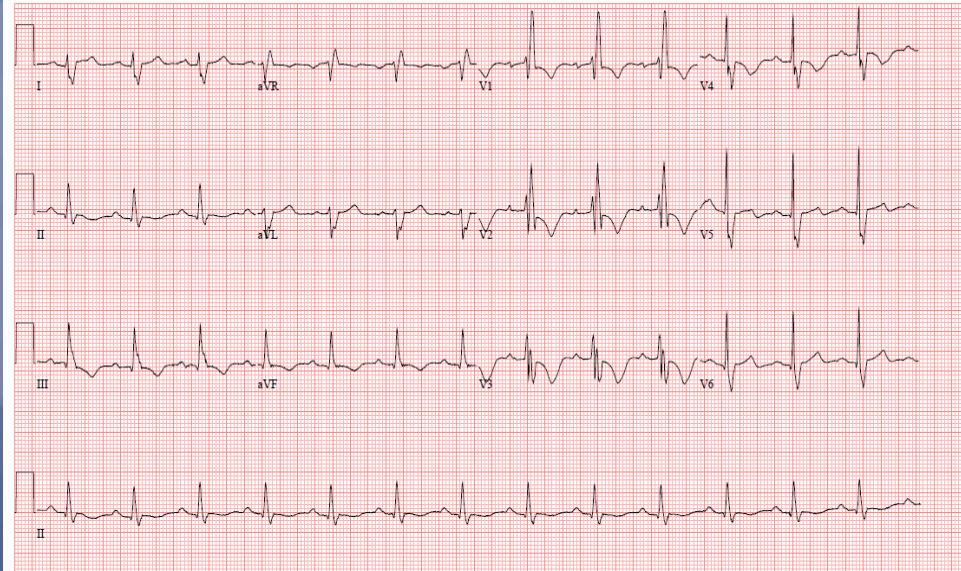


| | | | | |
|---------|--------------|---------|-----|--|
| Male | Vent. rate | 80 | BPM | Normal sinus rhythm |
| | PR interval | 202 | ms | Right bundle branch block |
| Room: | QRS duration | 140 | ms | Cannot rule out inferior infarct, age undetermined |
| Loc:117 | QT/QTc | 448/516 | ms | Abnormal ECG |
| | P-R-T axes | 50 103 | -30 | |

Technician:
Test ind:

Referred by:

Newly Acquired



25mm/s 10mm/mV 150Hz 7.1.1 12SL 241 HD CID: 117

EID/Newly Acquired EDT: ORDER:

Initial Laboratory data

CBC WBC 4300 /mm³ , Hb 13.9 g/dL , PLT 163×10³/mm³

Coagulation battery PT 11.3s (1.03 INR) aPTT 23.3 s

Chemical & electrolyte battery

Ca/P (mg/dL) 10.6/2.4

Glucose (mg/dL) 116

BUN/Cr (mg/dL) 24/1.3

Protein/albumin (mg/dL) 7.0/3.5

AST/ALT (IU/L) 49/86

ALP (IU/L) 86 T.bil (mg/dl) 0.9

Na/K/Cl/HCO₃ (mEq/L) 139/4.7/109/18

CK-MB 2.5 ng/mL

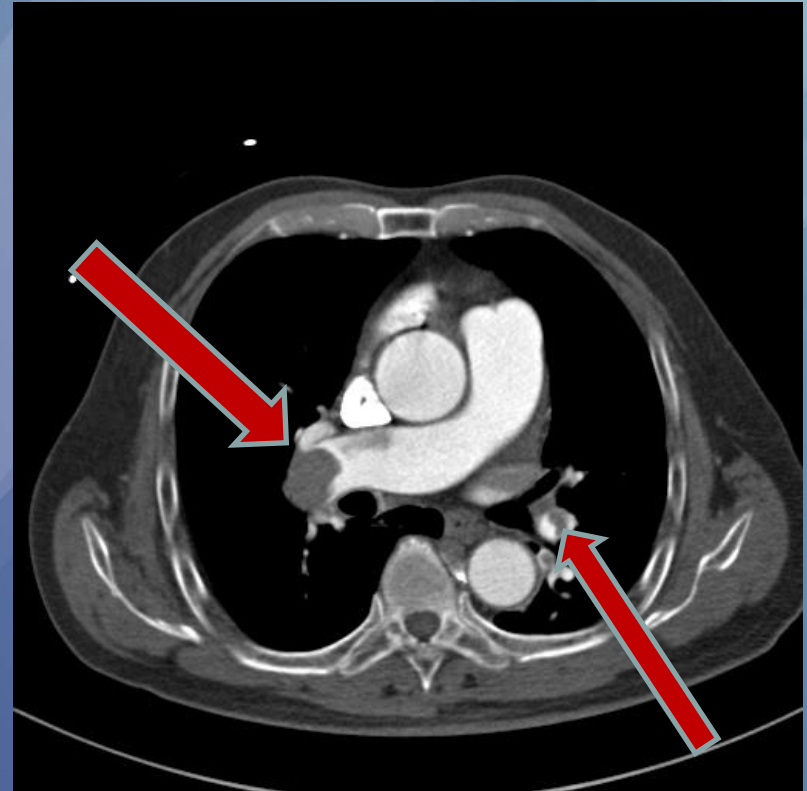
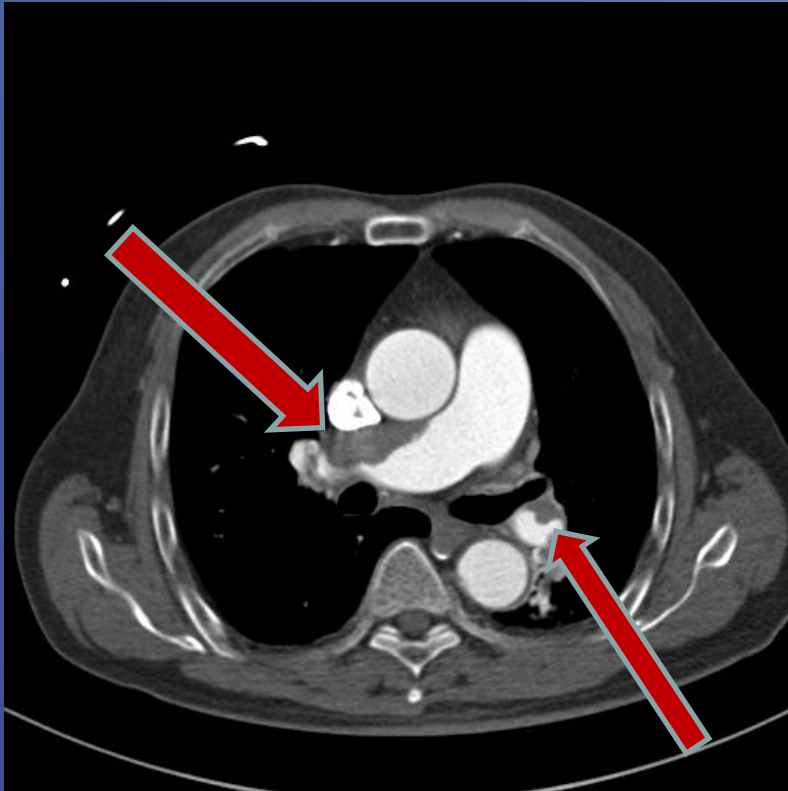
Tn-I 0.06 ng/mL

D-dimer 3.08 ug/ml

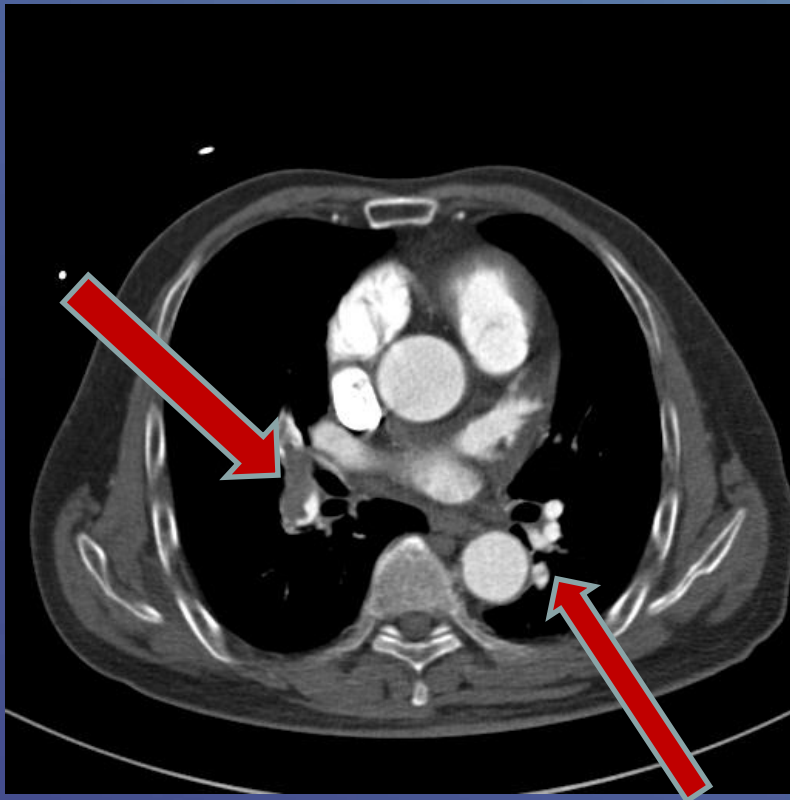
BNP 235 pg/mL

ABGA 7.46-28-77-20.0-96% lactic acid 0.8 mmol/L

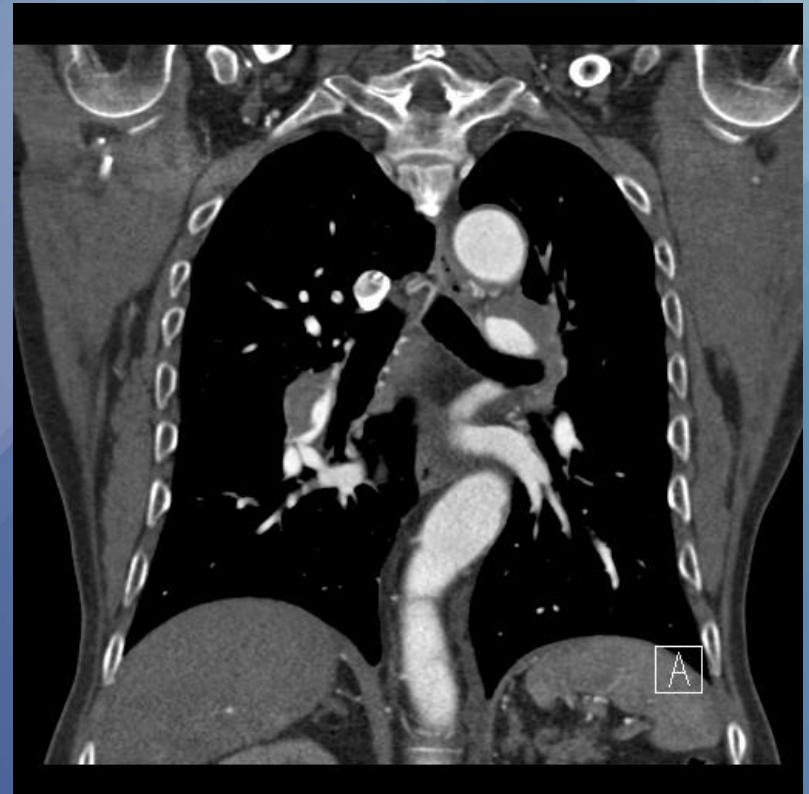
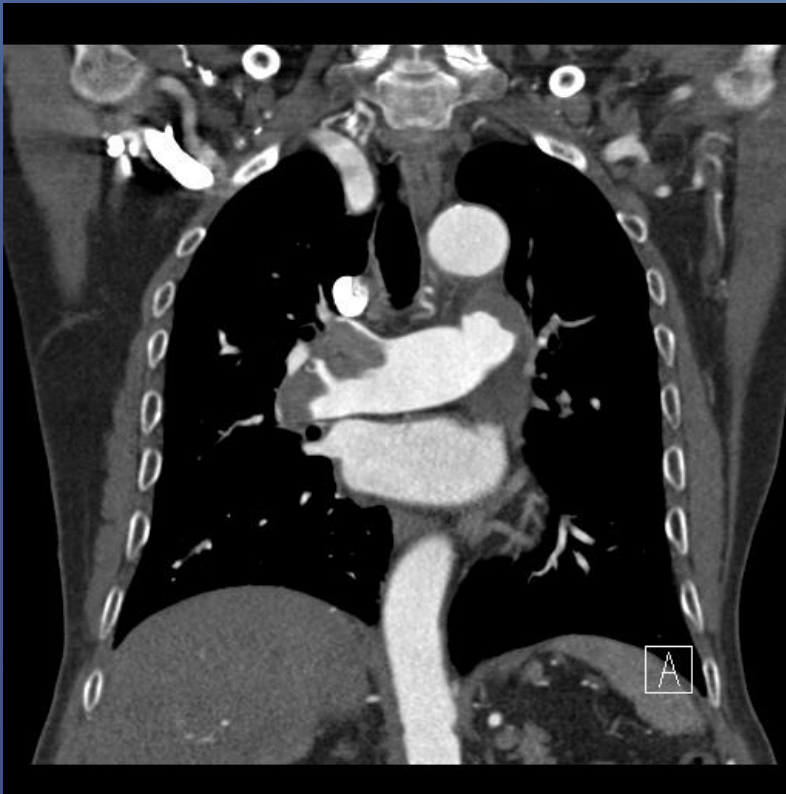
Chest CT



Chest CT



Chest CT



Echocardiography

Left Ventricle D-shaped LV

| | | | | | | | |
|------------|--------------|-------------|--------------|---------------|--------------|------------------|----------------|
| LVIDs | 29 mm | LVIDd | 37 mm | LA | 45 mm | Aorta | 37 mm |
| LVPWs | 14 mm | LVPWd | 10 mm | ESV | 19 ml | EDV | 46 ml |
| IVSs | 13 mm | IVSd | 10 mm | LVEF | 59 % | LV Mass | 113.0 g |
| P.E. (ant) | mm | P.E. (post) | mm | LV Mass Index | | g/m ² | |

Right Ventricle decreased RV

| | | | |
|-------|----|-------|--------------|
| RVIDs | mm | RVIDd | 42 mm |
|-------|----|-------|--------------|

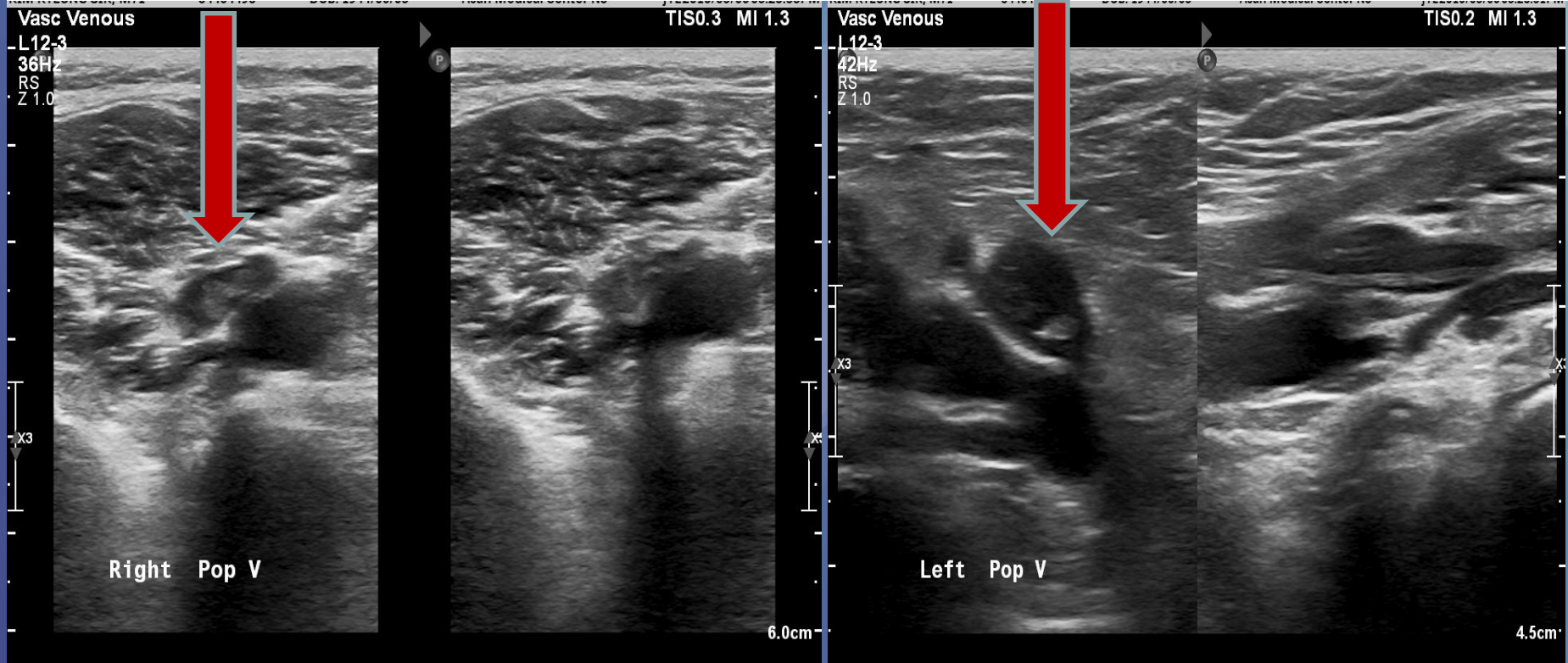
Tricuspid Valve tethering

| | | | | | |
|--------------|----------------|-------------|---------------------------|-------------|----------------|
| TR grade | 3 | TR jet area | 10 cm ² | Peak TR Vel | 4.4 m/s |
| PGsys(RV-RA) | 77 mmHg | Peak E Vel | cm/s | Peak A Vel | cm/s |

Conclusion)

1. Echogenic round mass in right pulmonary artery with RV dilatation and dysfunction ;C/W pulmonary thromboembolism
2. **Severe resting pulmonary hypertension with normal RV wall thickness, suggesting acute RV pressure overloading**

Lower extremity US



HD#1-7

**A: Acute pulmonary embolism with DVT
(Intermediate-high risk group)**

**Treatment: Anticoagulation with LMWH (D1-D4)
-> rivaroxaban 15mg bid (D5->)**

Echo after 9 days anticoagulation

Left Ventricle D-shaped LV

| | | | | | | | |
|------------|-------|-------------|-------|---------------|-----------------------|---------|---------|
| LVIDs | 26 mm | LVIDd | 45 mm | LA | 30 mm | Aorta | 39 mm |
| LVPWs | 15 mm | LVPWd | 9 mm | ESV | 25 mL | EDV | 66 mL |
| IVSs | 15 mm | IVSd | 9 mm | LVEF | 62 % | LV Mass | 133.5 g |
| P.E. (ant) | mm | P.E. (post) | 2 mm | LV Mass Index | 78.8 g/m ² | | |

Right Ventricle decreased contractility

| | | | |
|-------|----|-------|-------|
| RVIDs | mm | RVIDd | 41 mm |
|-------|----|-------|-------|

Tricuspid Valve tethering

| | | | | | |
|--------------|---------|-------------|-------------------|-------------|---------|
| TR grade | 2 | TR jet area | 5 cm ² | Peak TR Vel | 3.6 m/s |
| PGsys(RV-RA) | 52 mmHg | Peak E Vel | cm/s | Peak A Vel | cm/s |

1. Disappeared echogenic round mass in right pulmonary artery
2. **RV dysfunction with moderate resting pulmonary hypertension**
; decreased pulmonary artery pressure compared with previous study

Echo after 3 months anticoagulation

Left Ventricle D-shaped LV

| | | | | | | | |
|------------|--------------|-------------|--------------|---------------|------------------------------|---------|----------------|
| LVIDs | 28 mm | LVIDd | 47 mm | LA | 34 mm | Aorta | 35 mm |
| LVPWs | 13 mm | LVPWd | 8 mm | ESV | 26 ml | EDV | 66 ml |
| IVSs | 13 mm | IVSd | 10 mm | LVEF | 61 % | LV Mass | 143.5 g |
| P.E. (ant) | mm | P.E. (post) | mm | LV Mass Index | 83.3 g/m ² | | |

Right Ventricle decreased contractility

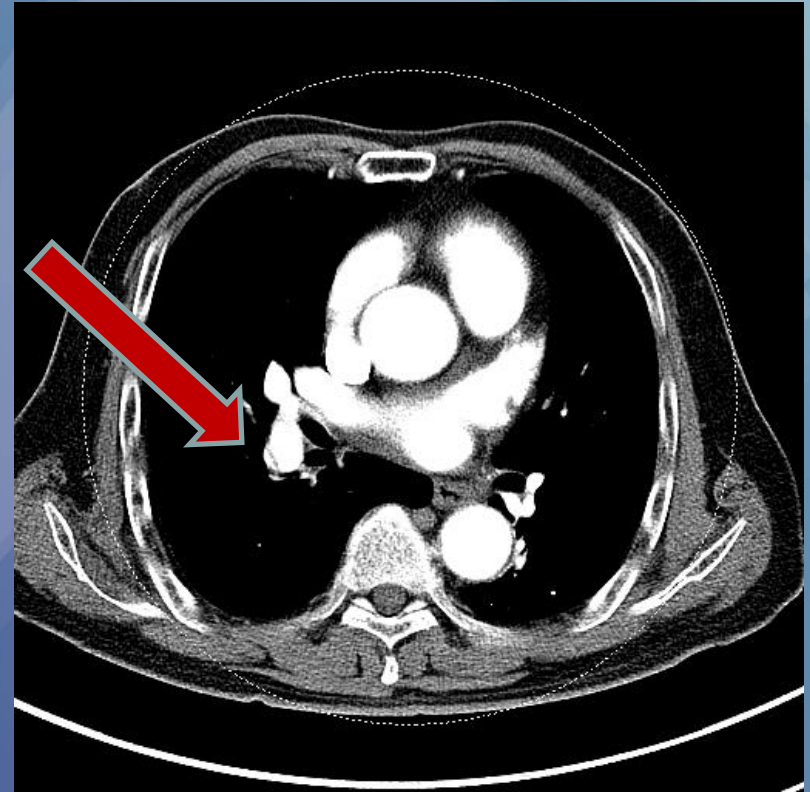
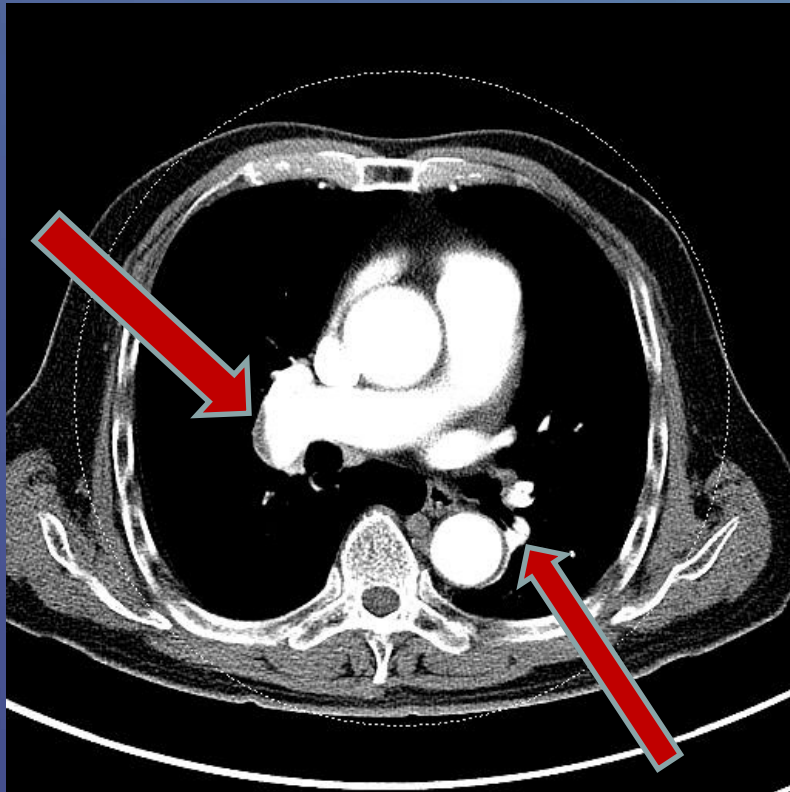
| | | | |
|-------|--------------|-------|--------------|
| RVIDs | 33 mm | RVIDd | 37 mm |
|-------|--------------|-------|--------------|

Tricuspid Valve mild tethering

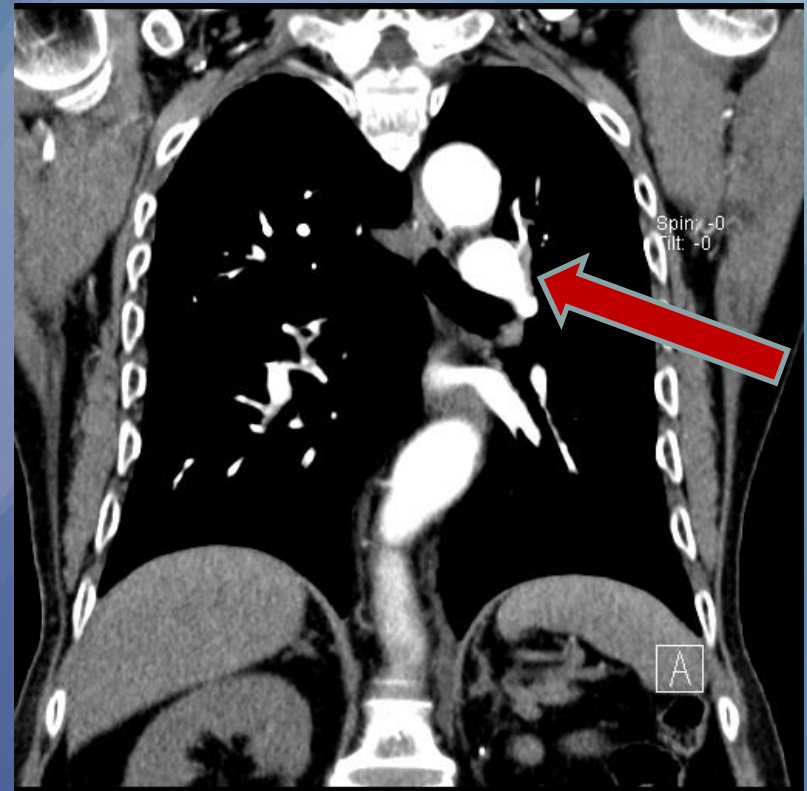
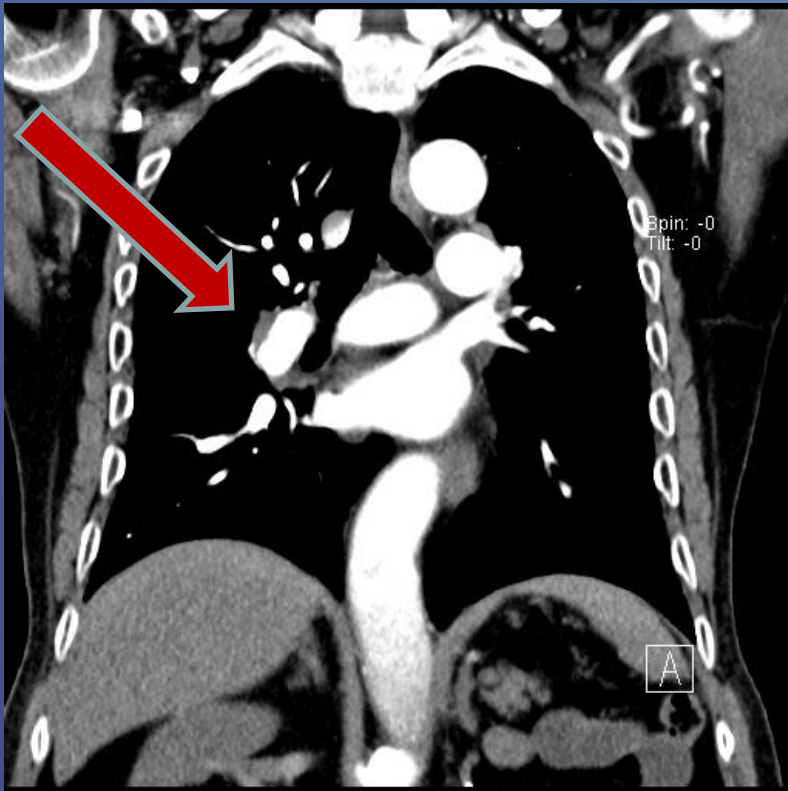
| | | | | | |
|--------------|----------------|-------------|--------------------------|-------------|----------------|
| TR grade | 1-2 | TR jet area | 5 cm ² | Peak TR Vel | 4.0 m/s |
| PGsys(RV-RA) | 64 mmHg | Peak E Vel | cm/s | Peak A Vel | cm/s |

Slightly improved RV dysfunction with severe resting pulmonary hypertension ;increased pulmonary artery pressure compared with the previous study(2016.5.11)

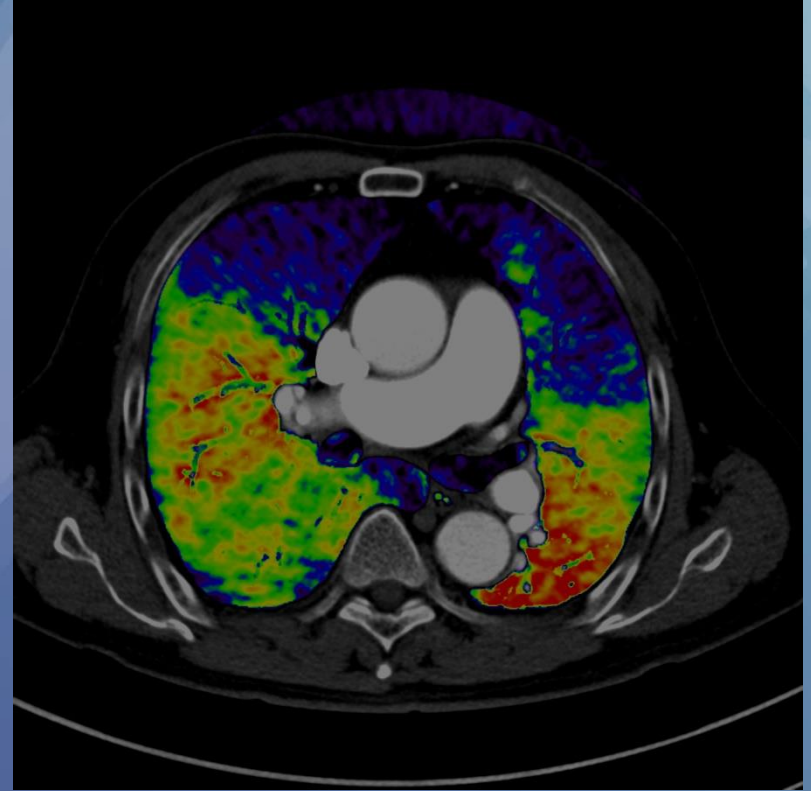
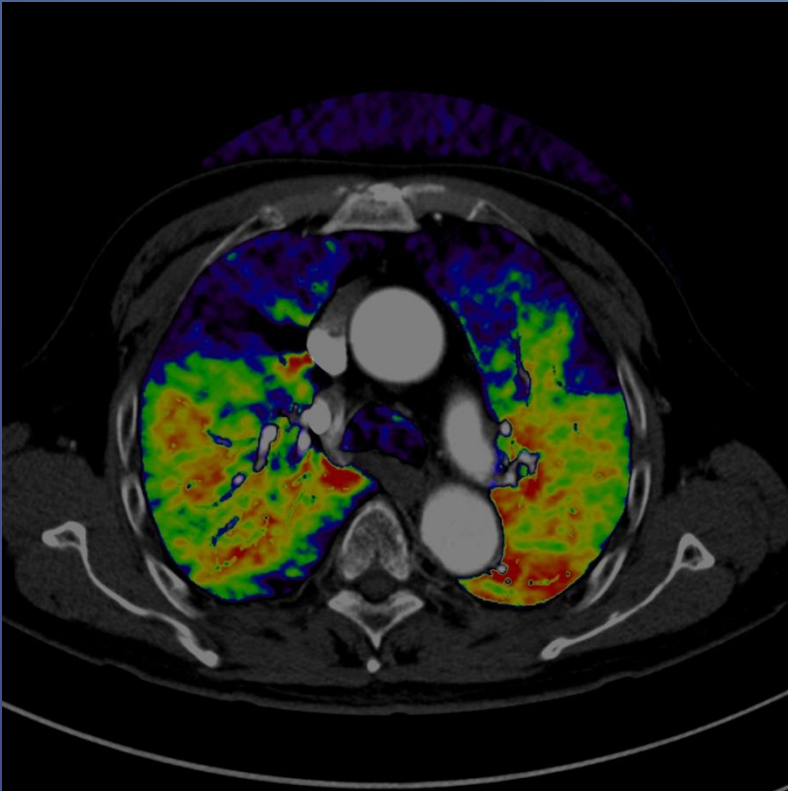
CT after 3-months anticoagulation



CT after 3-months anticoagulation

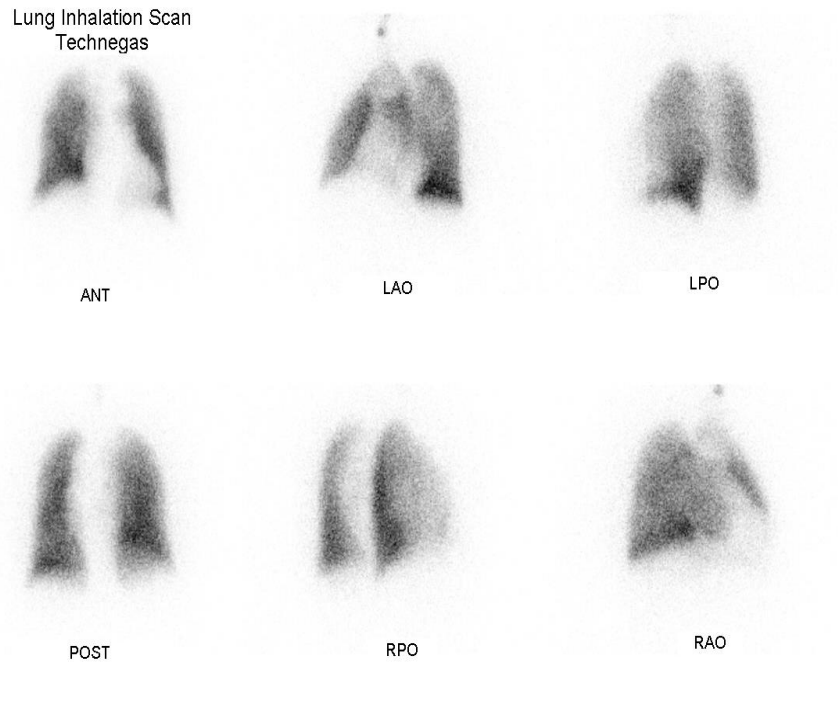


CT after 3-months anticoagulation

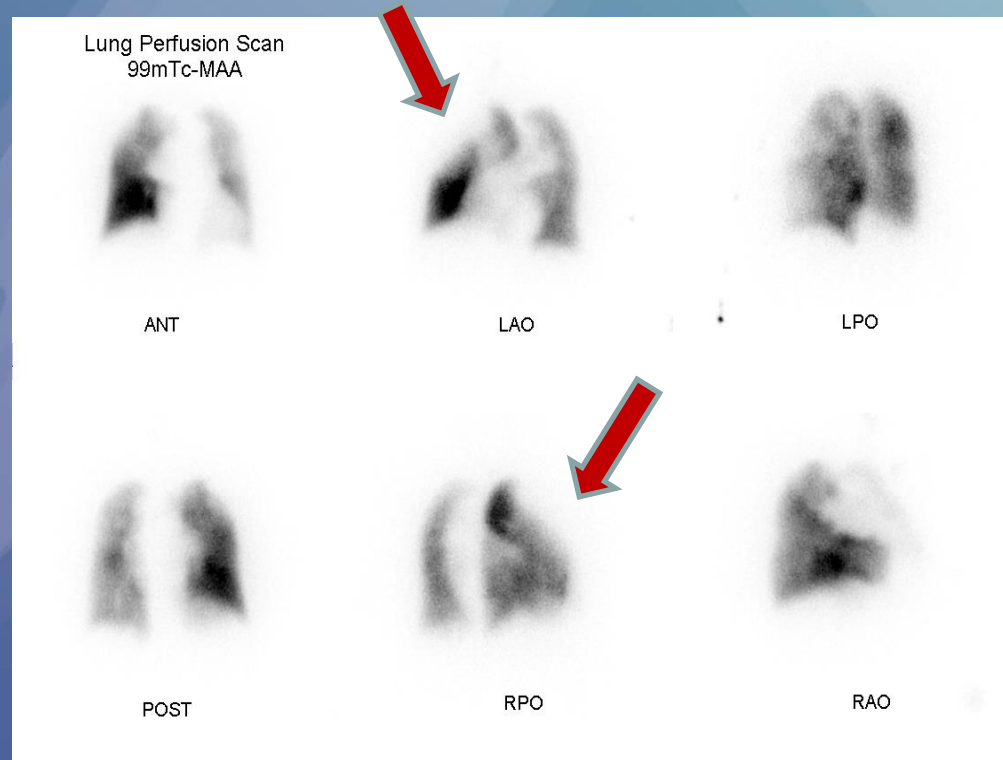


V/Q scan after 5-months anticoagulation

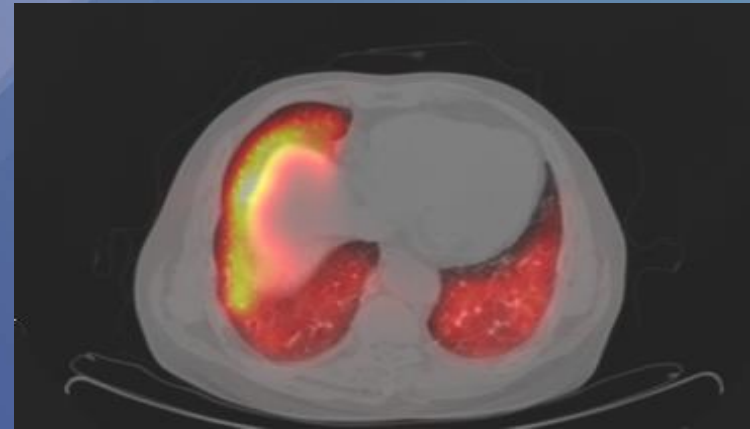
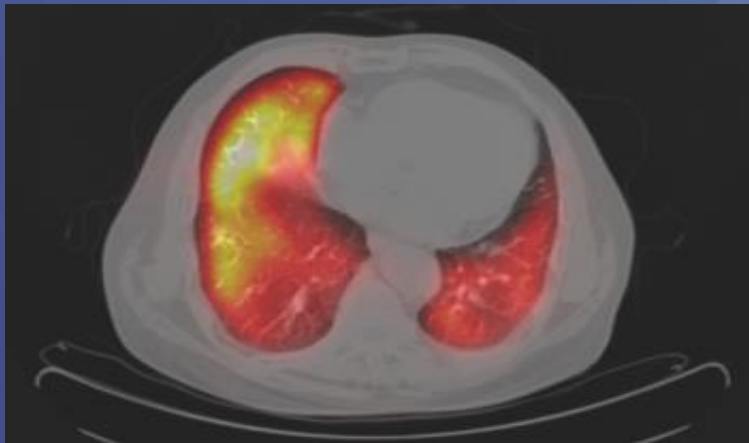
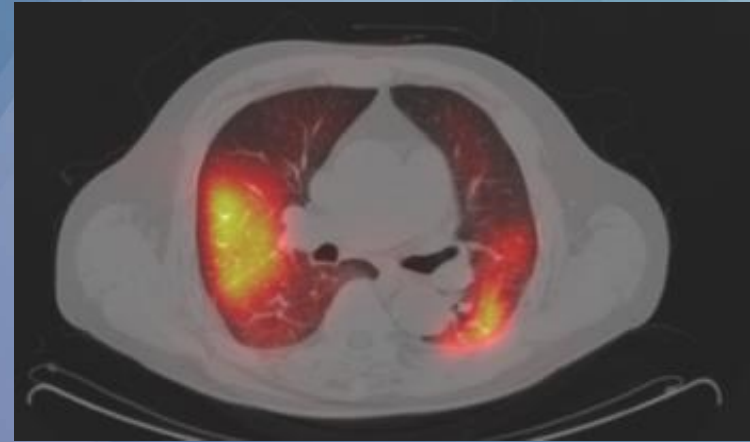
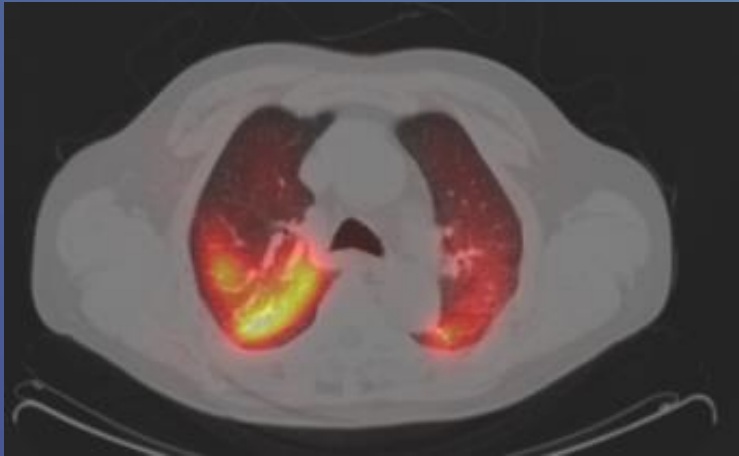
Lung Inhalation Scan
Technegas



Lung Perfusion Scan
99mTc-MAA



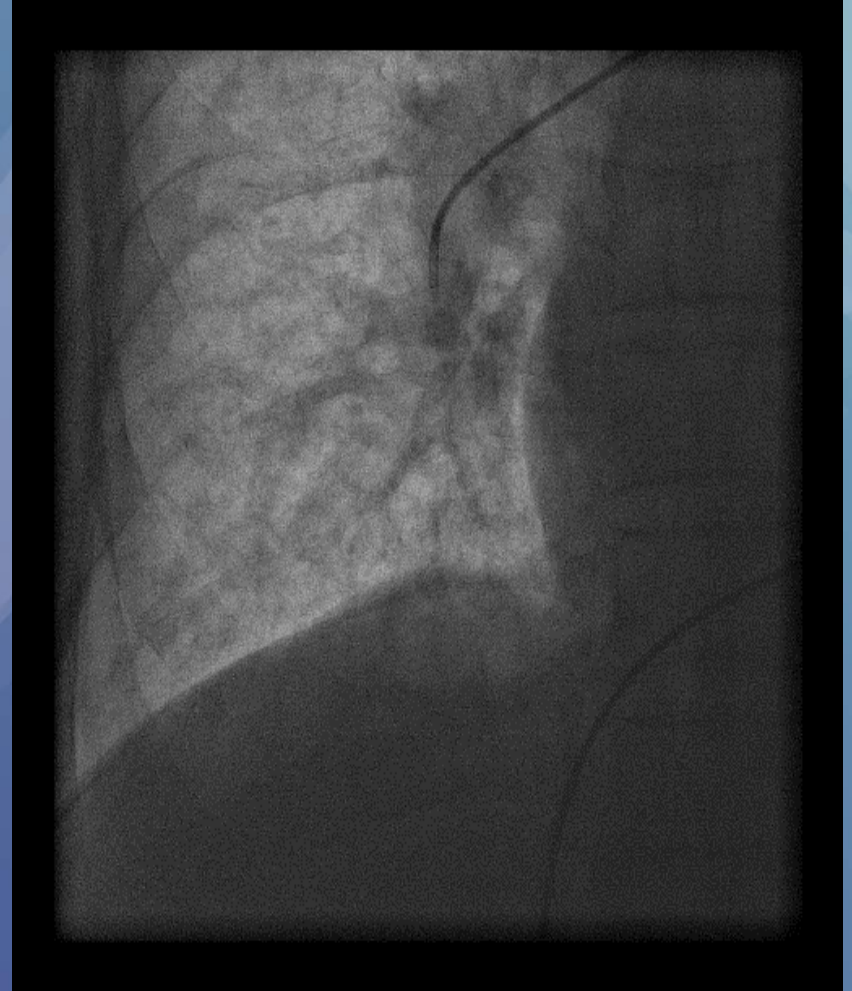
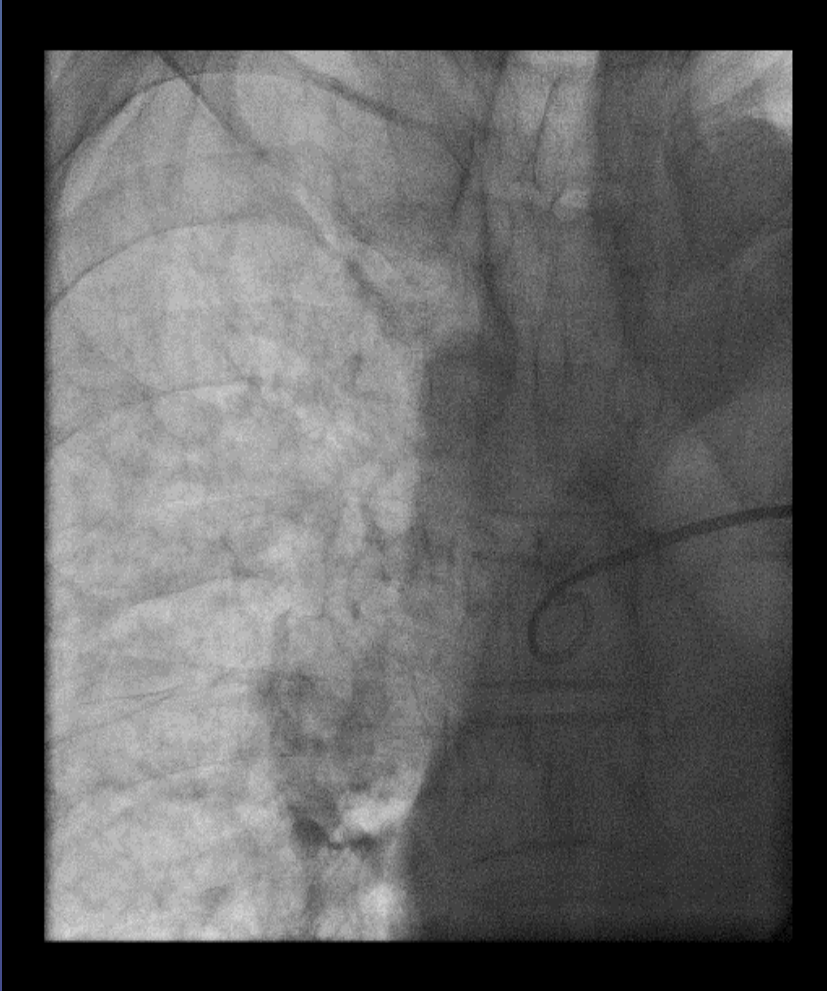
Fusion perfusion SPECT/CT after 5-month anticoagulation



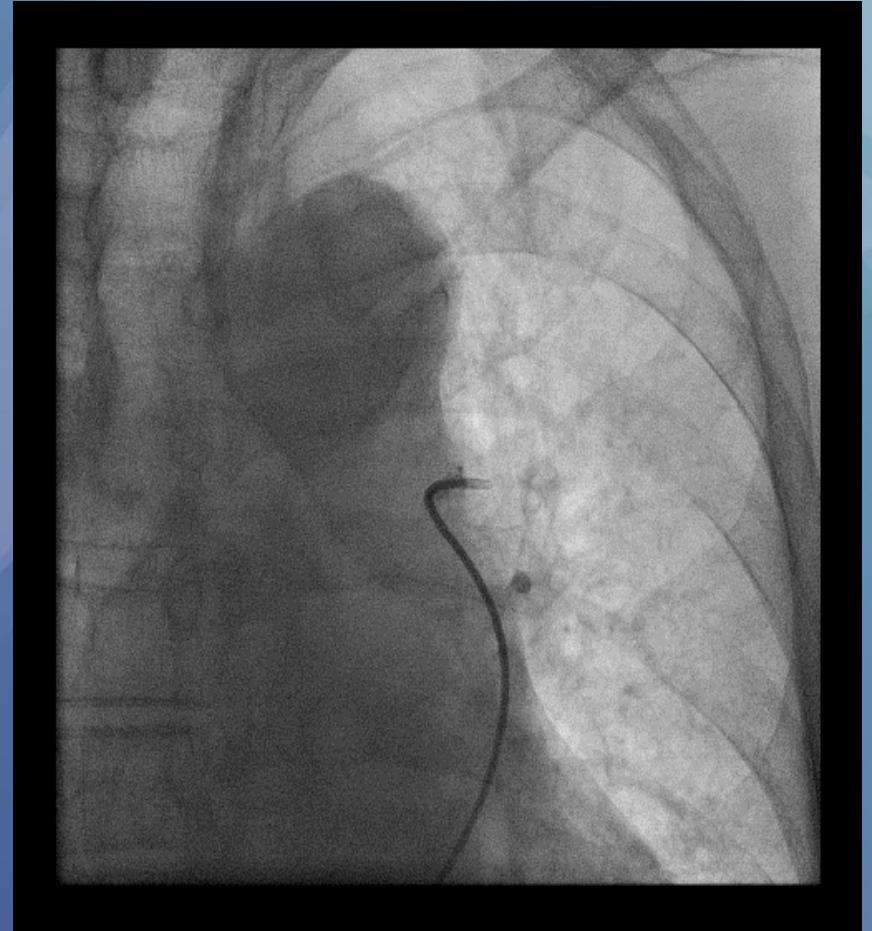
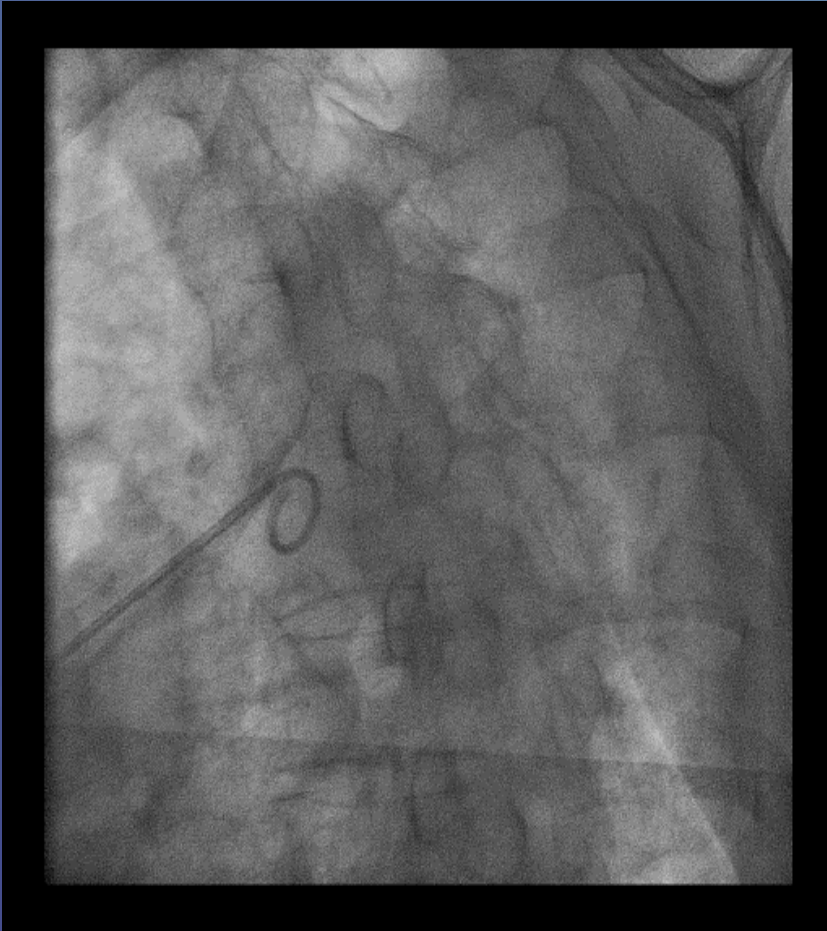
RHC

| | |
|------------------------|---------------|
| BP | 154/86 |
| mRAP | 5 |
| sPAP/dPAP | 50/16 |
| Mean PAP | 28 |
| PCWP | 9 |
| CO | 4.7 |
| CI | 2.7 |
| PVR (WU) | 4.05 |
| SvO₂ | 69% |

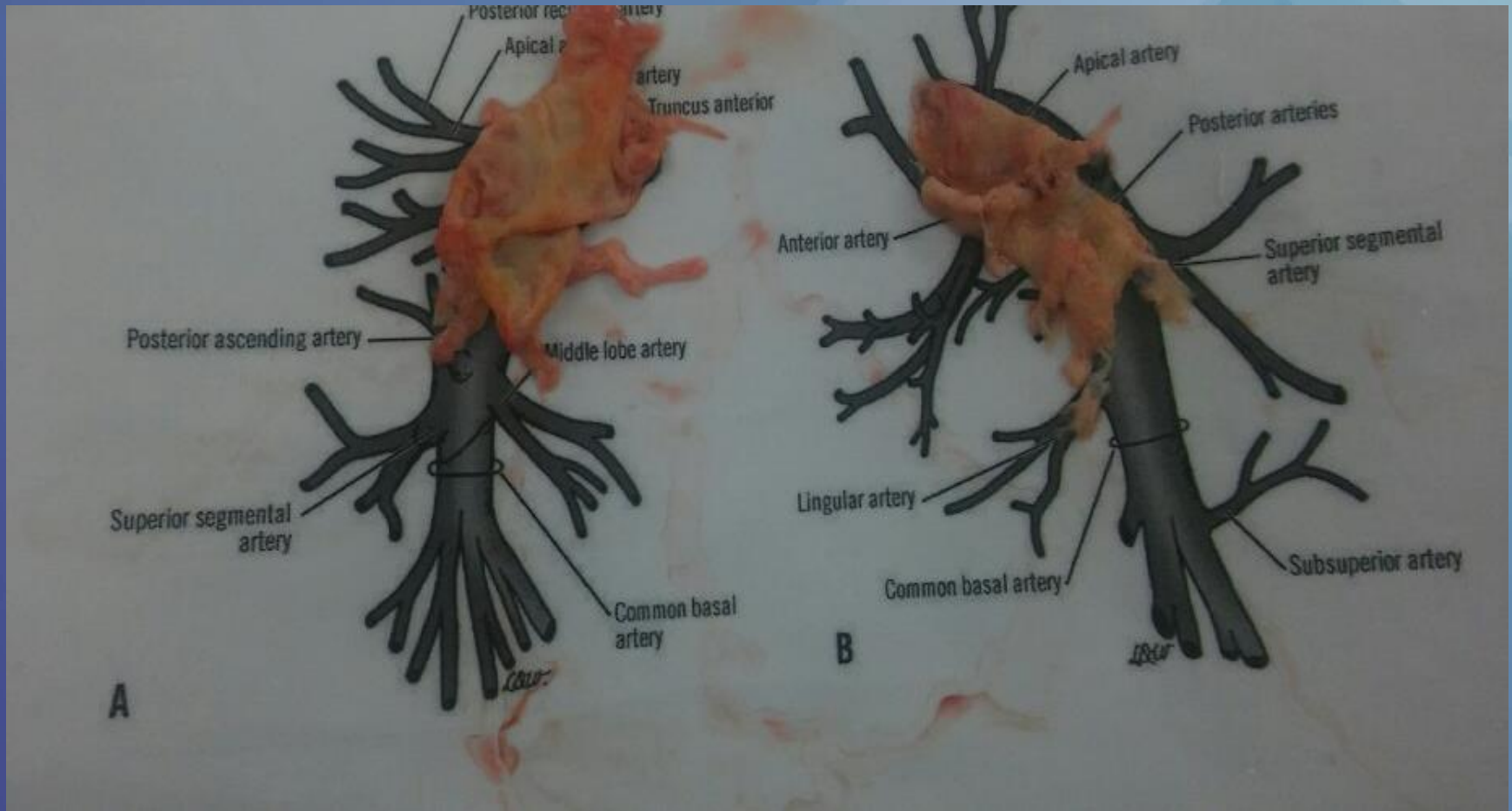
Right pulmonary angiography



Left pulmonary angiography

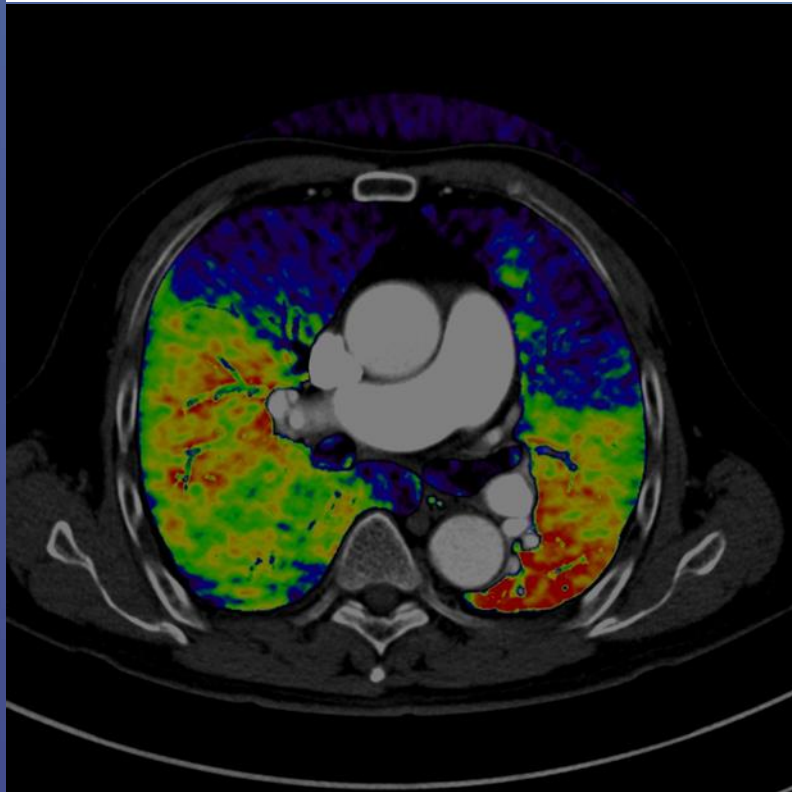


Pulmonary endarterectomy (PEA)

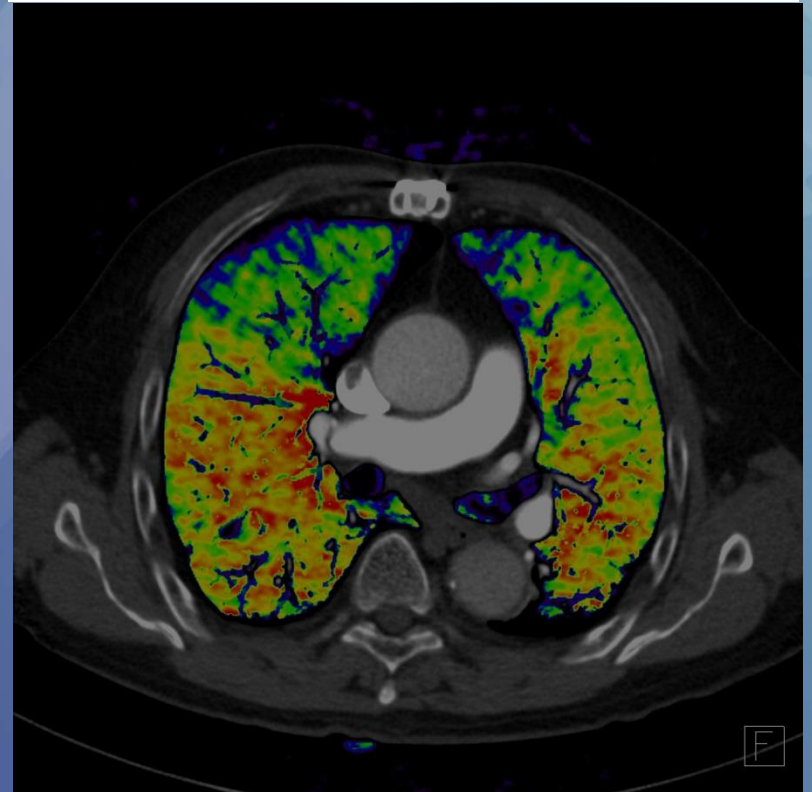


1yr after PEA

Pre-PEA



Post-PEA

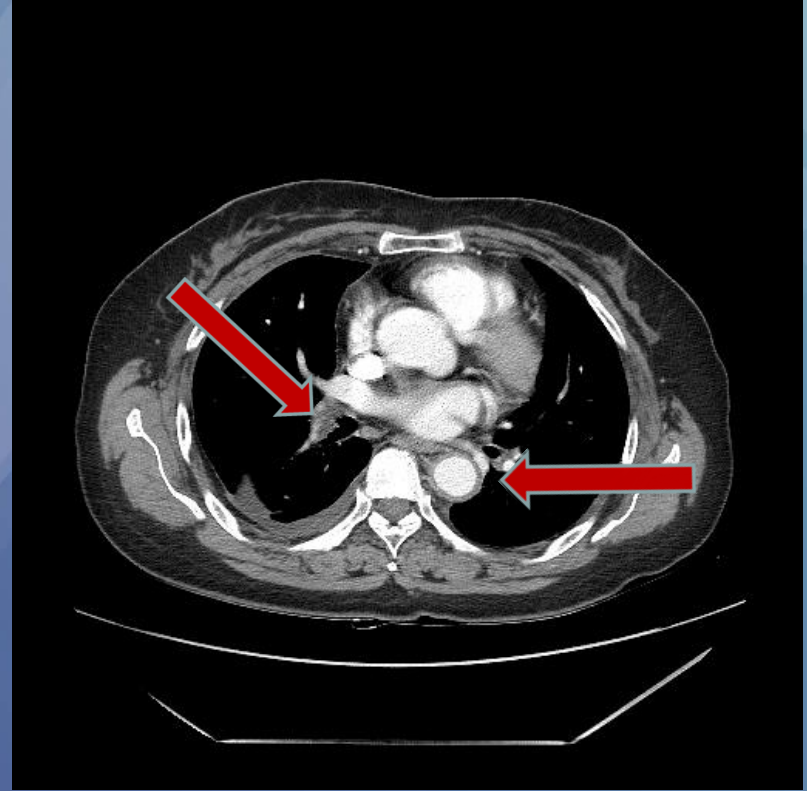
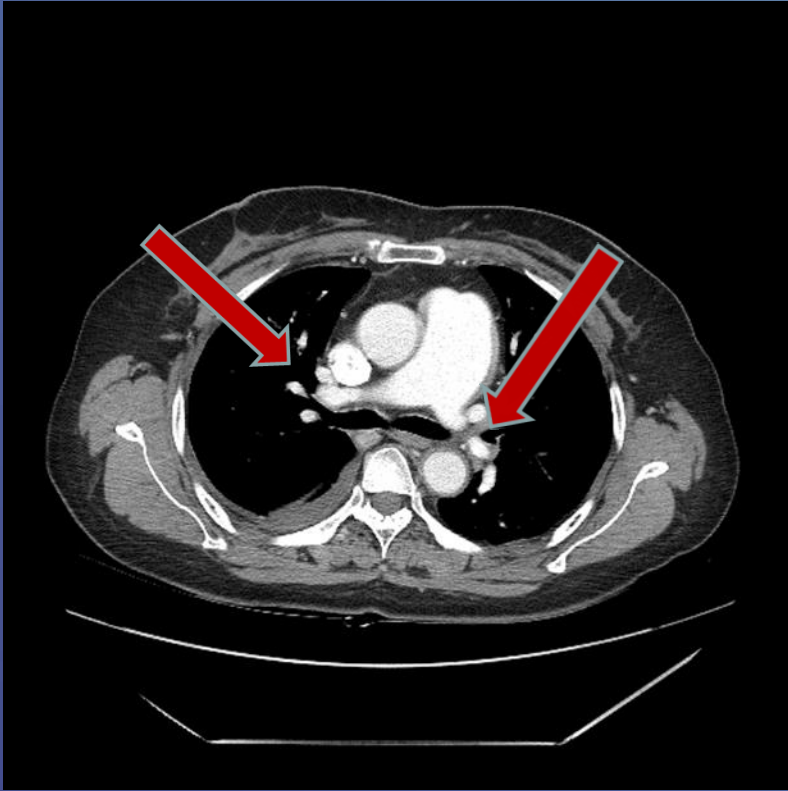


1yr after PEA

| | 2016.12 | 2018.03 |
|---------------------|------------|------------|
| WHO FC | II | I |
| TRVmax | 3.0 | 2.2 |
| PGsys(RV-RA) | 36 | 19 |
| BNP | 30 | 31 |
| 6MWD | 415 | 476 |
| Nadir SaO2 | 92% | 97% |

예비 증례

54/F Takayasu's arteritis



Echocardiography

1. Narrowing of right pulmonary artery with mild stenosis and thickened ascending aorta;

C/W Takayasu arteritis

2. Borderline resting pulmonary hypertension (TR Vmax 3.0 m/s)

3. Normal LV size and systolic function

V/Q scan

Lung Inhalation Scan
Technegas



ANT



LAO



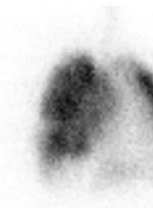
LPO



POST



RPO



RAO

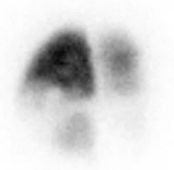
Lung Perfusion Scan
99mTc-MAA



ANT



LAO



LPO



POST

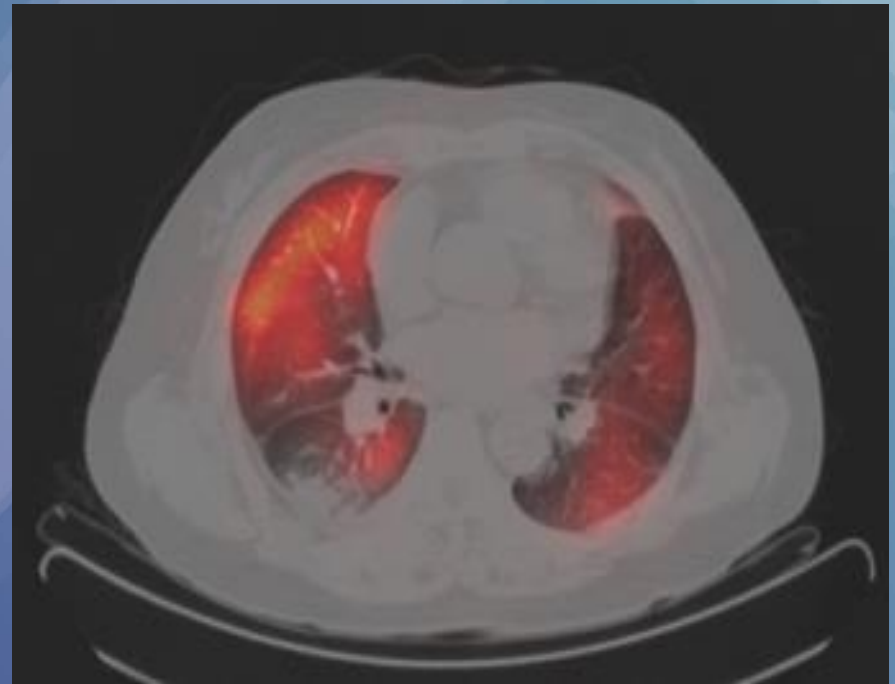
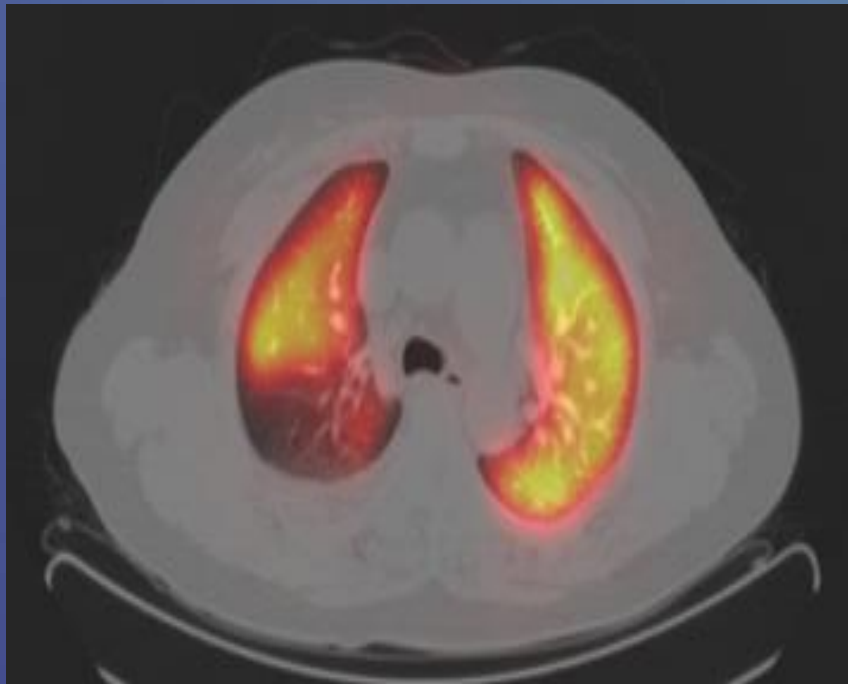


RPO



RAO

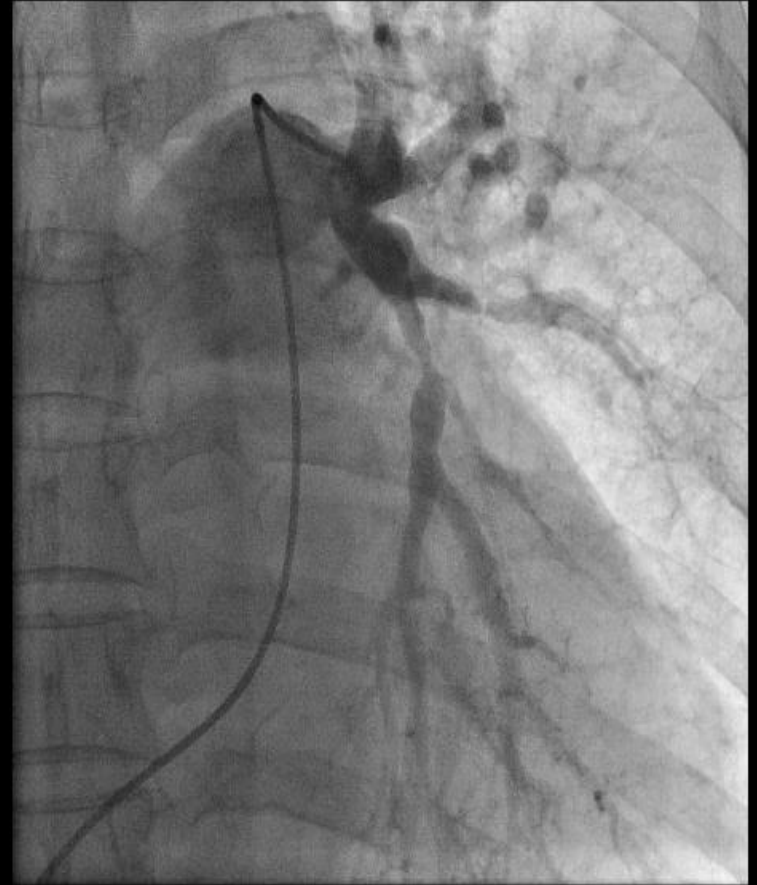
Fusion perfusion SPECT/CT



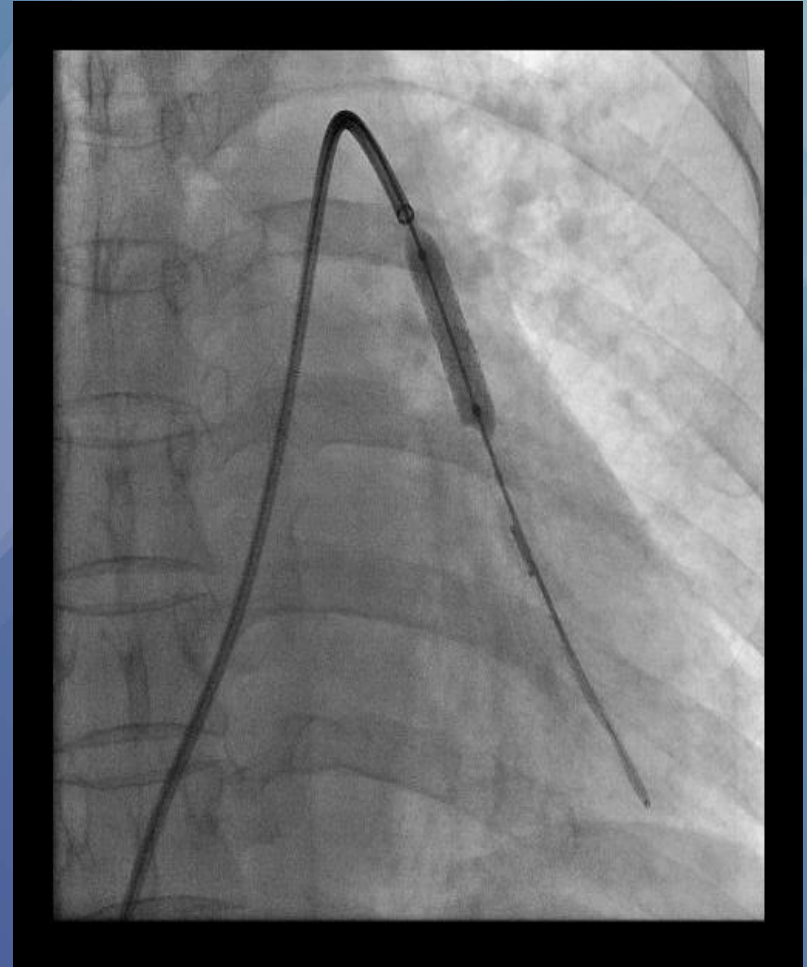
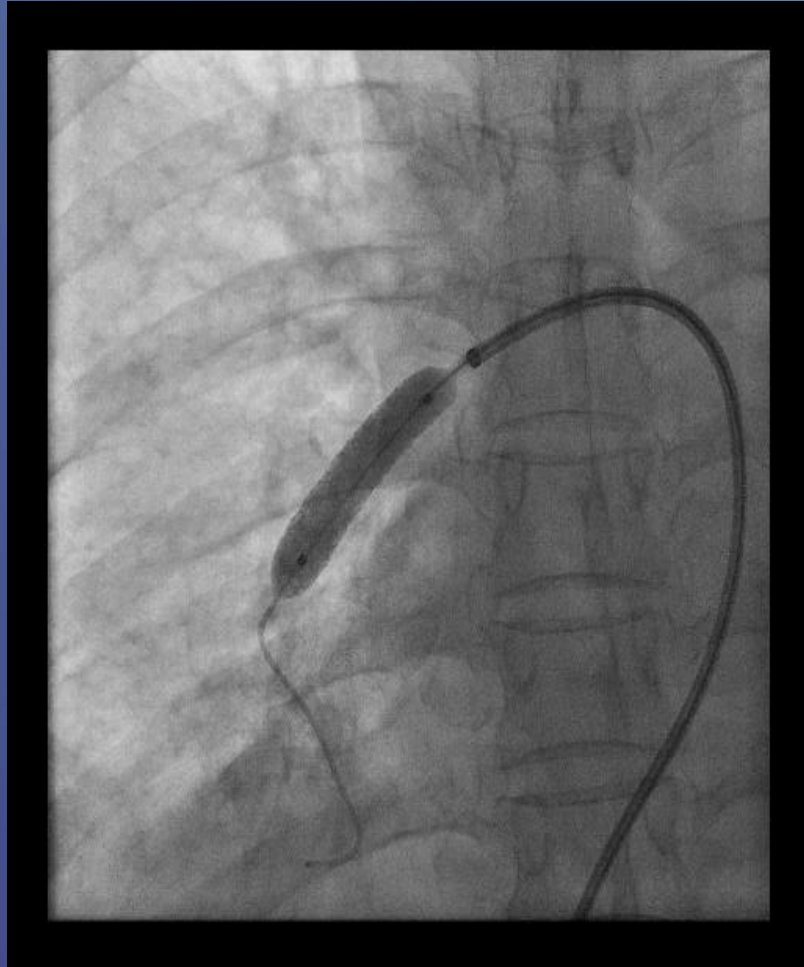
RHC

| | |
|------------------------|---------------|
| BP | 154/86 |
| mRAP | 8 |
| sPAP/dPAP | 60/18 |
| Mean PAP | 33 |
| PCWP | 14 |
| CO | 6.01 |
| CI | 3.52 |
| PVR (WU) | 3.50 |
| SvO₂ | 76% |

Pulmonary angiography



BPA and PA stenting



After BPA and PA stenting

Pre-BPA

Lung Perfusion Scan
99mTc-MAA



ANT



LAO



LPO



POST



RPO



RAO

Post-BPA



ANT



LAO



LPO



POST

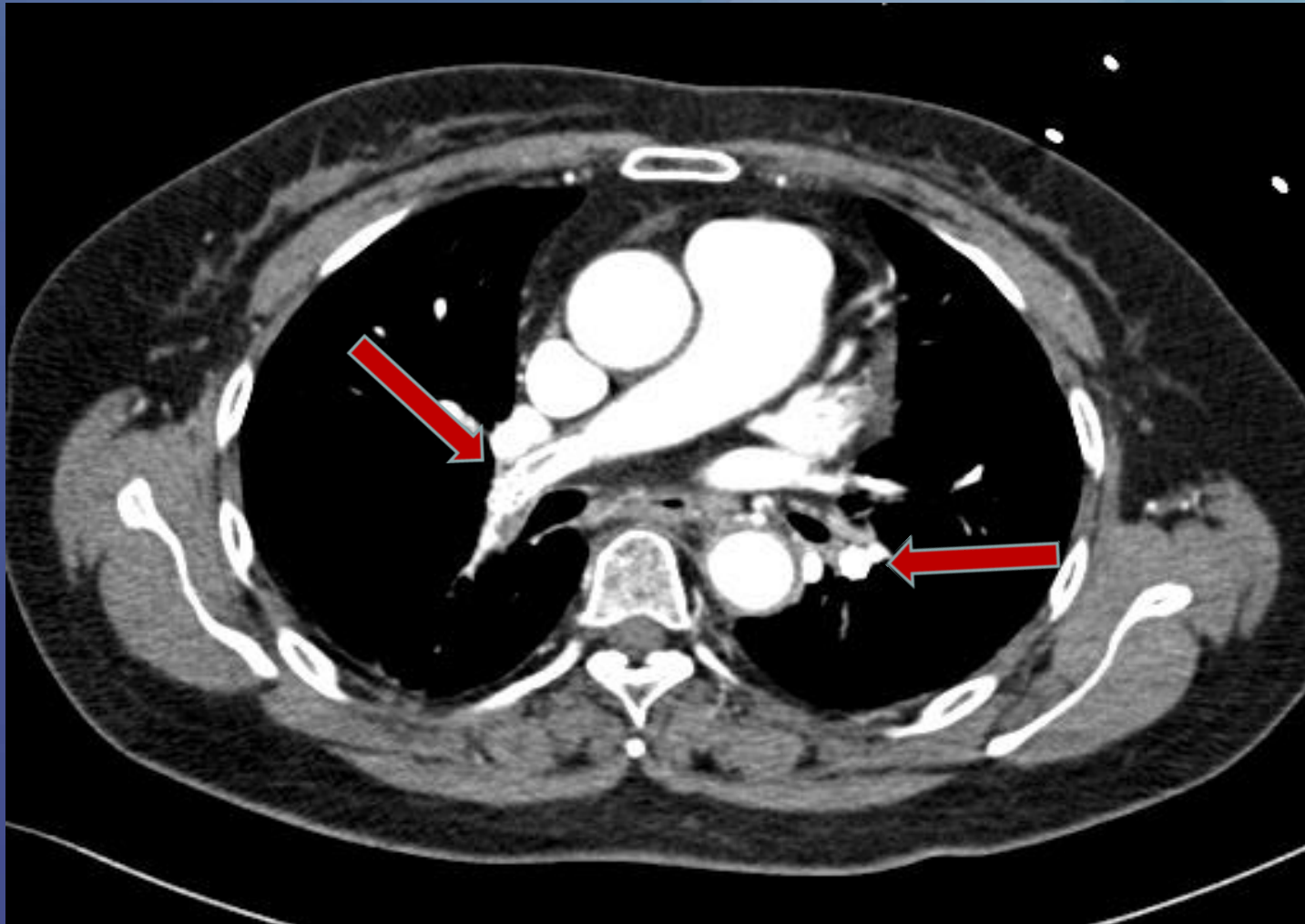


RPO



RAO

2-year after BPA and stenting



2-year after BPA and stenting

| | 2016.05 | 2018.03 |
|---------------------|------------|------------|
| WHO FC | II | I |
| TRVmax | 3.0 | 3.0 |
| PGsys(RV-RA) | 36 | 36 |
| BNP | 11 | |
| 6MWD | 375 | 518 |
| Nadir SaO2 | 84% | 89% |

Case. (68/M)

Chief complaint : dyspnea (mMRC III)

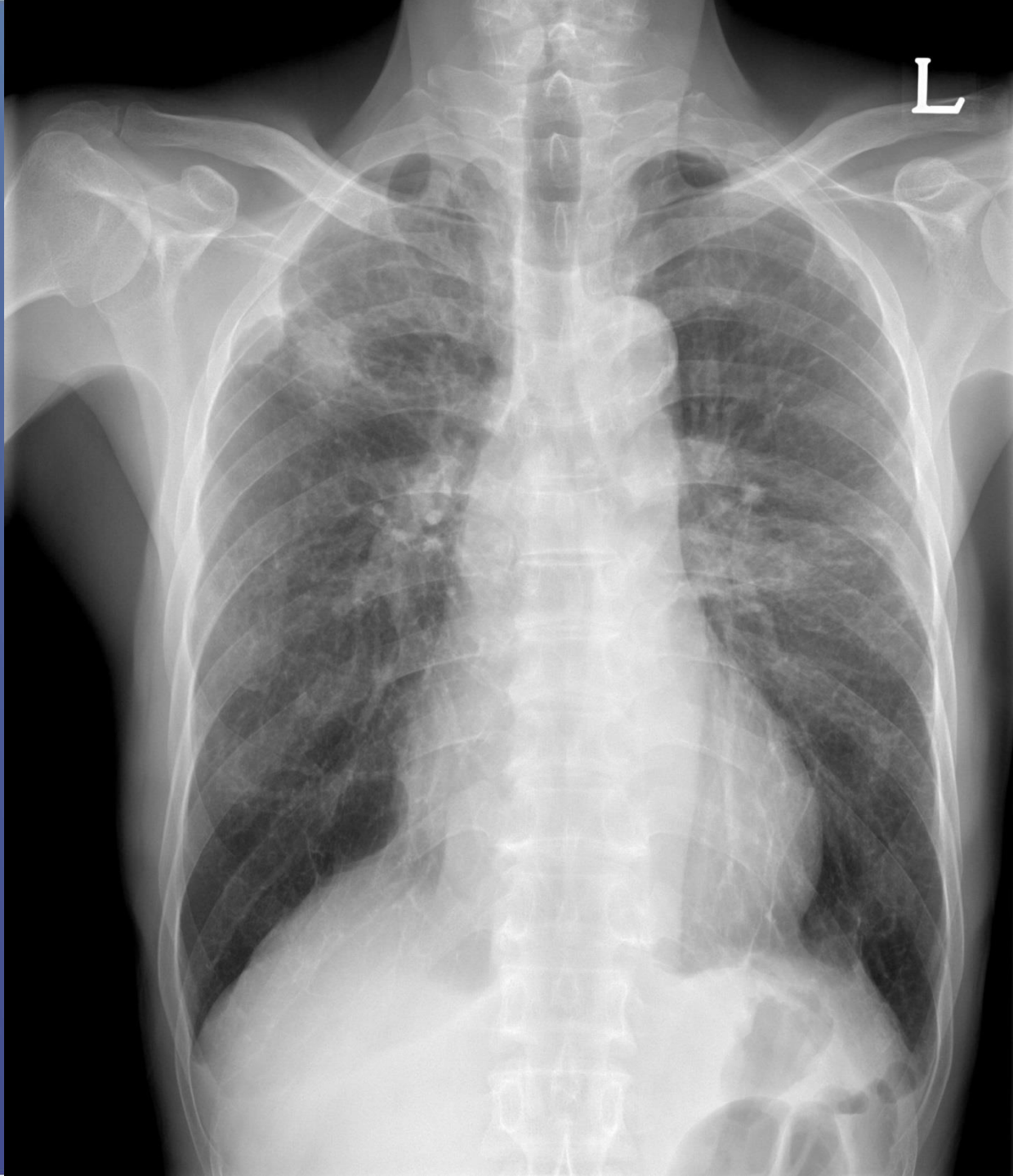
P. M. Hx: Pulmonary TBc (2013)

S. Hx. : smoking : 30 pack years

Echocardiography: **TR Vmax 4.5 m/s**

severe pulmonary hypertension

Dilated RV with RV dysfunction



Pulmonary function tests

SPIROMETRY

(S2002)

| | (S2001) | | | Bronchodilator | | |
|-----------------------|---------|-------|--------|----------------|--------|-------|
| | Pred. | Meas. | %Pred. | Meas. | %Pred. | %Chg. |
| FVC | 4.56 | 3.72 | 82 | 3.72 | 82 | -0 |
| FEV ₁ | 3.37 | 2.46 | 73 | 2.49 | 74 | 1 |
| FEV ₁ /FVC | 71 | 66 | 93 | 67 | | |
| FEF _{25-75%} | 2.63 | 1.68 | 64 | 1.86 | 71 | 11 |
| PEF | 7.94 | 5.44 | 68 | 5.50 | 69 | 1 |

LUNG VOLUMES

(S2015)

| | Pred | Meas. | %Pred. |
|-----|------|-------|--------|
| TLC | 6.83 | 6.09 | 89 |
| VC | 4.17 | 3.72 | 89 |
| FRC | 3.99 | 4.13 | 104 |
| ERV | 1.42 | 1.76 | 123 |
| RV | 2.57 | 2.37 | 92 |

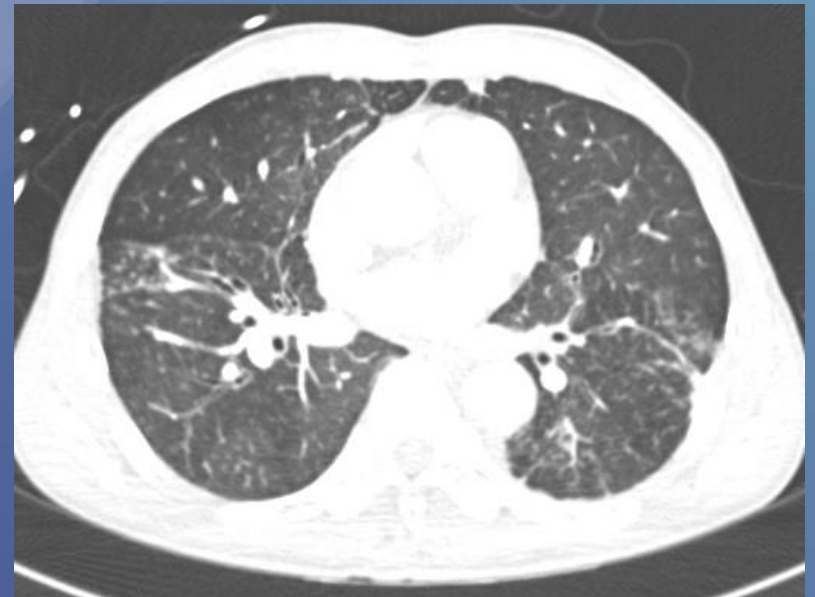
DIFFUSING CAPACITY

| | Pred. | Meas. | %Pred. |
|-----------|----------------------------|-------|--------|
| DL | 22.9 | 13.4 | 58 |
| VA(TLC) | | 4.61 | |
| DL/VA | 3.57 | 2.91 | 81 |
| Corrected | 22.9 | 13.0 | 57 |
| Hb | Date 2016061: 15.6 g/dl | | |

TEST(D) O₂

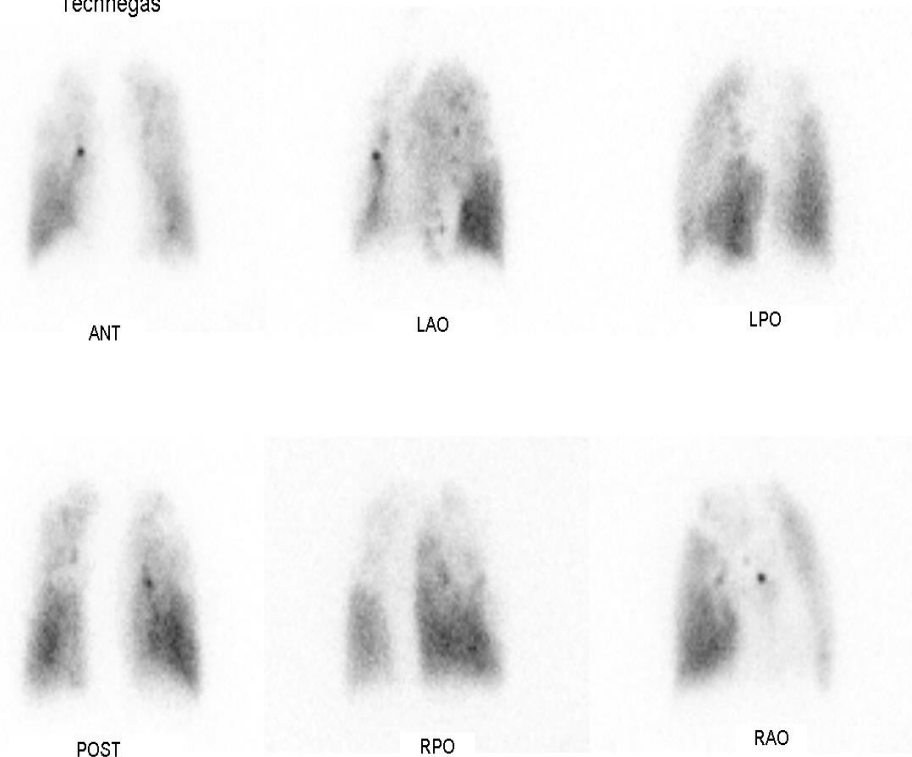
LPM

| Pulse | SaO ₂ | DISTANCE | RPE |
|-------|------------------|----------|-----|
| 72 | 97 | | |
| 85 | 95 | | |
| 89 | 94 | | |
| 89 | 94 | | |
| 92 | 94 | | |
| 93 | 94 | | |
| 96 | 93 | | |
| 99 | 93 | | |
| 102 | 94 | | |
| 103 | 95 | | |
| 105 | 94 | | |
| 106 | 94 | | |
| 110 | 94 | 425 | 7 |

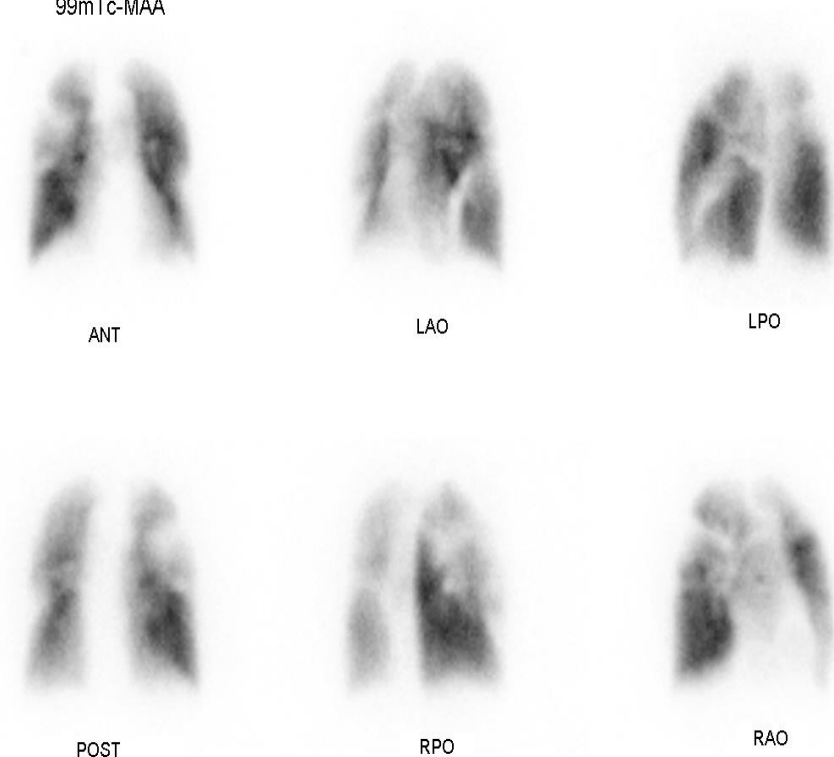




Lung Inhalation Scan
Technegas



Lung Perfusion Scan
99mTc-MAA



V/Q matching defects in in RUL, RML, and LUL

**V/Q mismatching large perfusion defects in RLL superior segment
and LLL anteromedial basal and lateral basal segments**

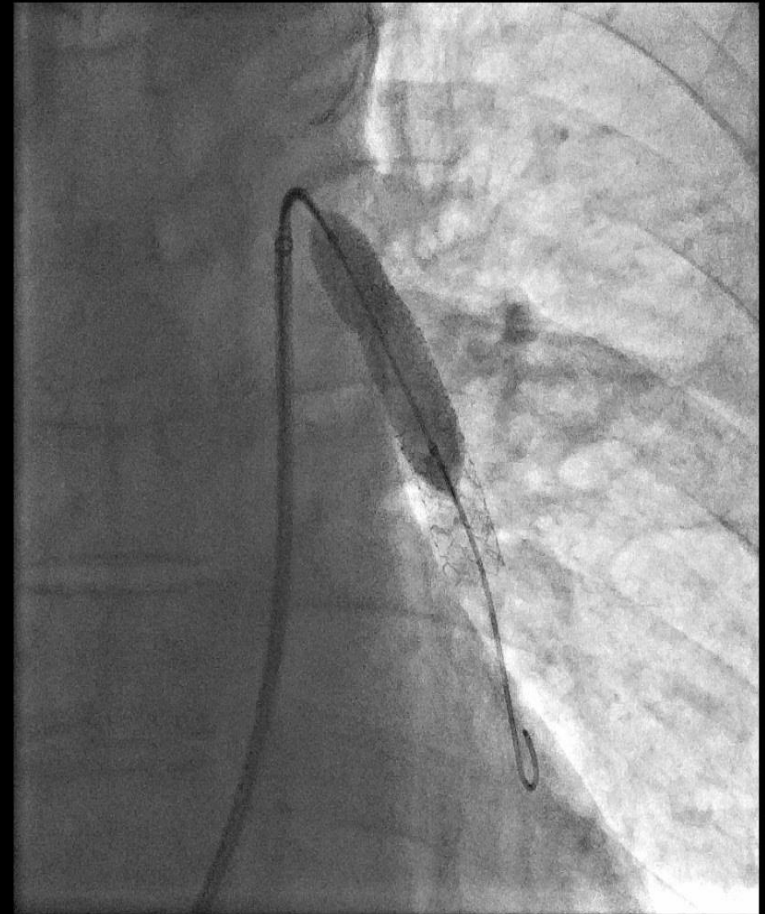
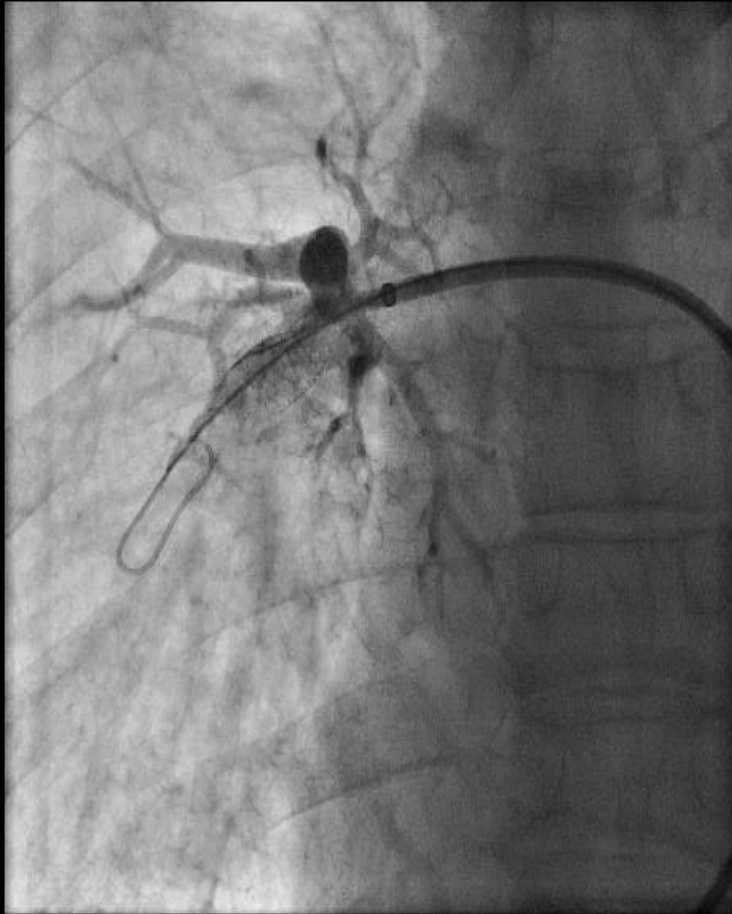
RHC

| | |
|------------------------|---------------|
| BP | 138/95 |
| mRAP | 7 |
| sPAP/dPAP | 88/26 |
| Mean PAP | 50 |
| PCWP | 9 |
| CO | 4.19 |
| CI | 2.51 |
| PVR (WU) | 10.42 |
| SvO₂ | 74% |

Pulmonary angiography



BPA and PA stenting



1-year after BPA and stenting

| | 2016.06 | 2017.04 |
|---------------------|------------|------------|
| WHO FC | III | II |
| TRVmax | 4.5 | 3.0 |
| PGsys(RV-RA) | 81 | 36 |
| 6MWD | 425 | 515 |
| Nadir SaO2 | 93% | 92% |

Case. (76/M)

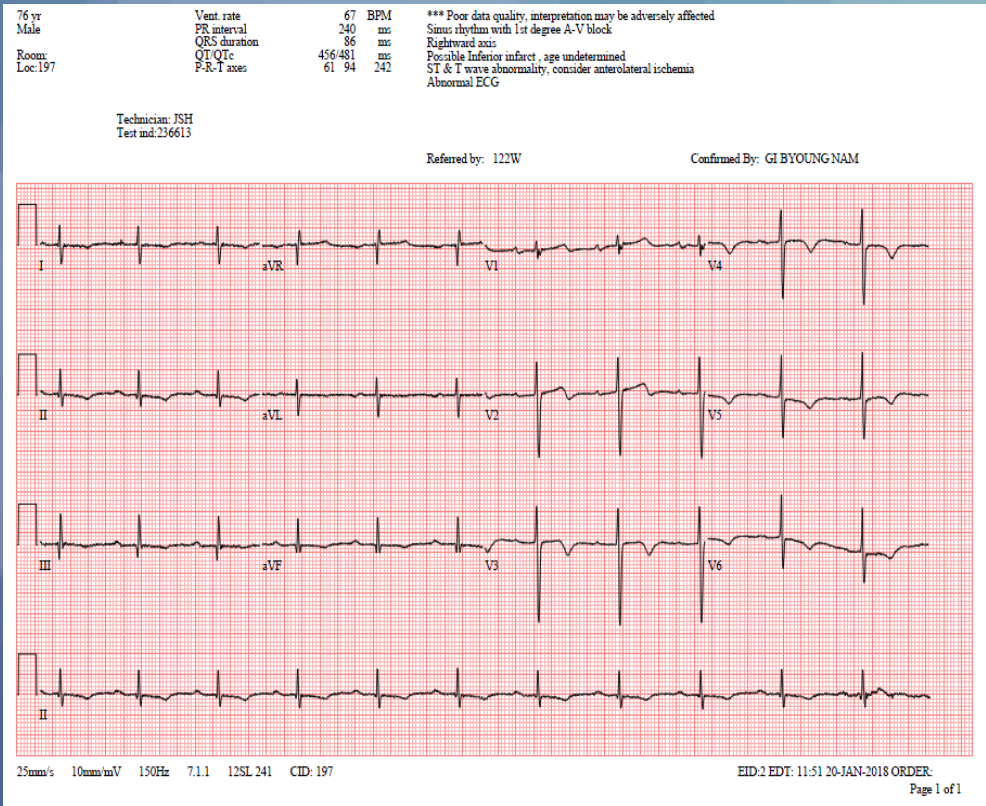
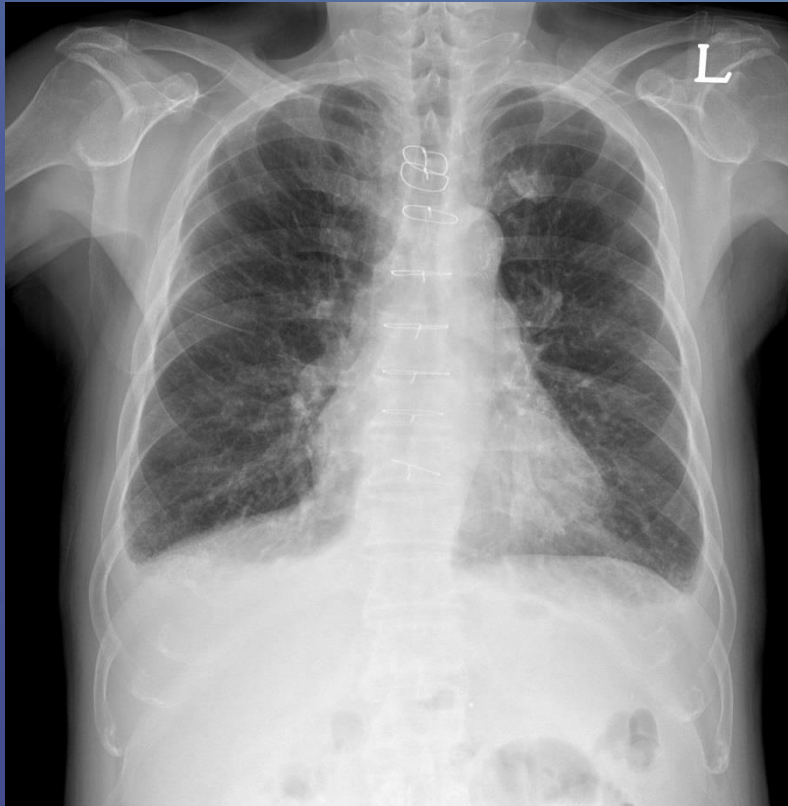
Chief complaint : dyspnea (WHO FC IV)

**P. M. Hx: DM (+) HTN(+) 3VD -> s/p CABG (2007)
COPD on LTOT (2008)**

S. Hx. : smoking : 60 pack years

**Echocardiography: TR Vmax 4.1-4.5 m/s
severe pulmonary hypertension**

CXR & EKG



Pulmonary function tests

SPIROMETRY

(S2002)

| | (S2001) | | | Bronchodilator | | |
|-----------------------|---------|-------|--------|----------------|--------|-------|
| | Pred. | Meas. | %Pred. | Meas. | %Pred. | %Chg. |
| FVC | 3.78 | 3.62 | 96 | 3.59 | 95 | -1 |
| FEV ₁ | 2.48 | 1.85 | 75 | 1.81 | 73 | -2 |
| FEV ₁ /FVC | 70 | 51 | 73 | 50 | | |
| FEF _{25-75%} | 2.05 | 0.65 | 32 | 0.73 | 36 | 12 |
| PEF | 6.39 | 5.72 | 90 | 5.83 | 91 | 2 |

LUNG VOLUMES

(S2015)

| | Pred | Meas. | %Pred. |
|-----|------|-------|--------|
| TLC | 5.71 | 5.89 | 103 |
| VC | 3.18 | 3.74 | 117 |
| FRC | 3.64 | 3.87 | 106 |
| ERV | 1.10 | 1.72 | 157 |
| RV | 2.54 | 2.15 | 85 |

DIFFUSING CAPACITY

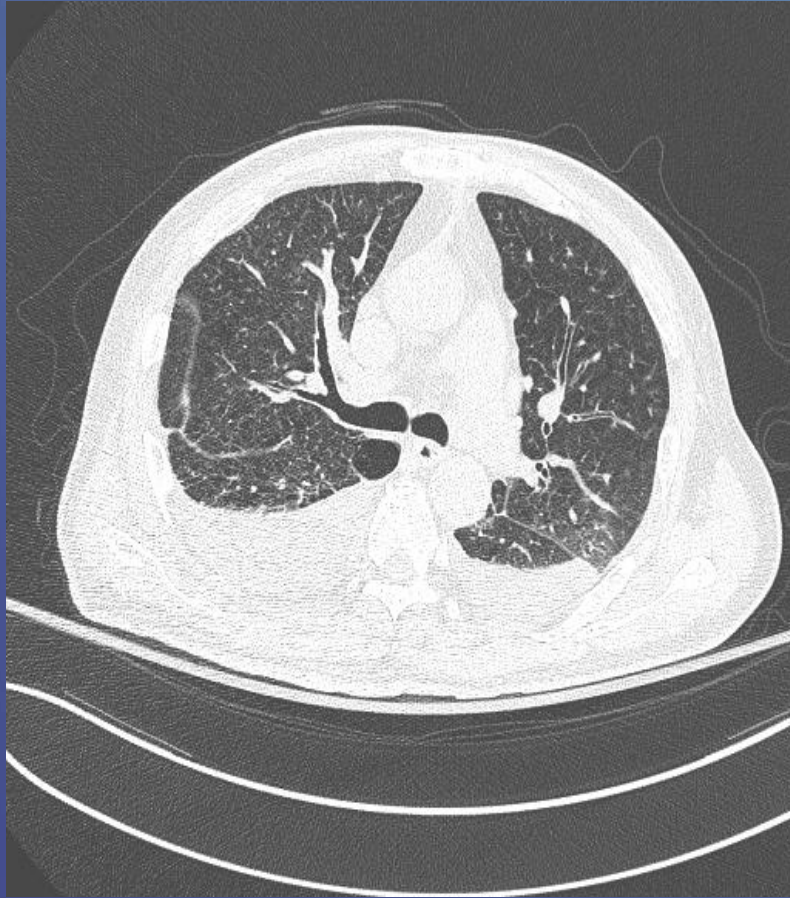
| | Pred. | Meas. | %Pred. |
|-----------|---------------------------|-------|--------|
| DL | 16.5 | 2.9 | 18 |
| VA(TLC) | | 5.11 | |
| DL/VA | 3.66 | 0.58 | 16 |
| Corrected | 16.5 | 2.9 | 18 |
| Hb | Date 2017111 14.3 g/dl | | |

TEST(I) O₂

LPM

| Pulse | SaO ₂ | DISTANCE |
|-------|------------------|----------|
| 85 | 92 | |
| | | |
| 89 | 87 | |
| 91 | 84 | |
| 94 | 85 | |
| 99 | 85 | |
| 99 | 85 | |
| 96 | 85 | |
| 96 | 86 | |
| 95 | 87 | |
| 93 | 87 | |
| 101 | 88 | |
| 96 | 89 | |
| 95 | 90 | 120 |

HR chest CT



RHC

| | |
|------------------------|---------------|
| BP | 147/97 |
| mRAP | 16 |
| sPAP/dPAP | 84/36 |
| Mean PAP | 56 |
| PCWP | 18 |
| CO | 2.52 |
| CI | 1.52 |
| PVR (WU) | 15.64 |
| SvO₂ | 48% |

Case. (74/F)

Chief complaint : dyspnea (WHO FC IV)

P. M. Hx: **DM (+) HTN(+)**

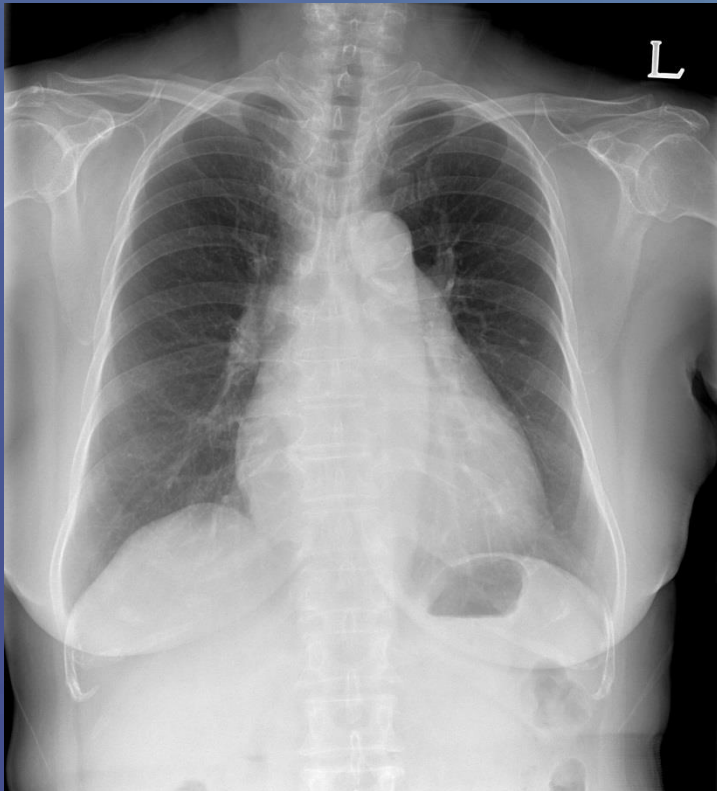
Severe TR, AF

-> s/p TV annuloplasty & Maze operation (2014.05)

S. Hx. : smoking (-)

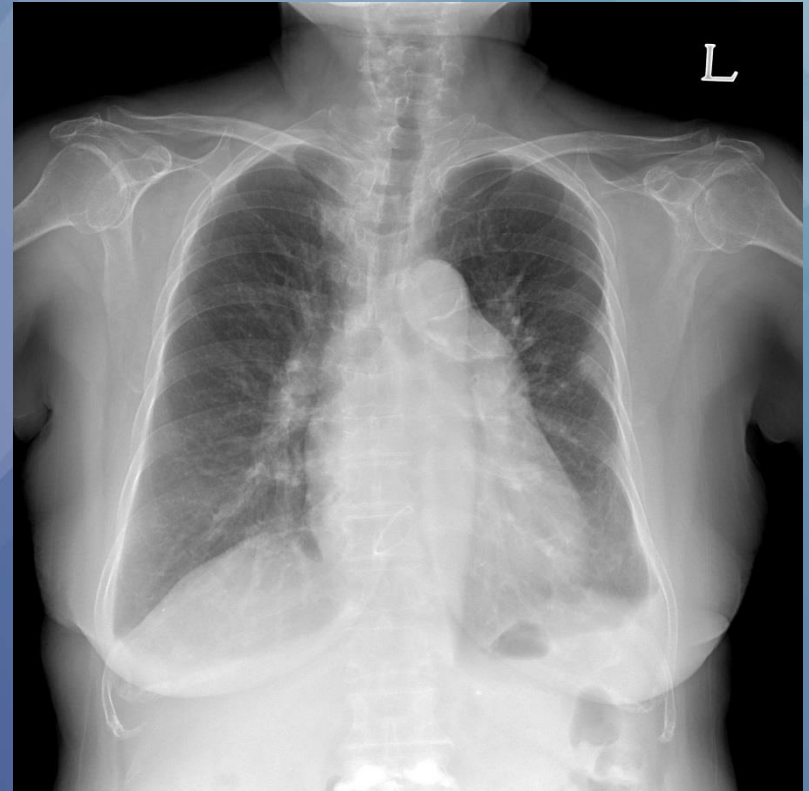
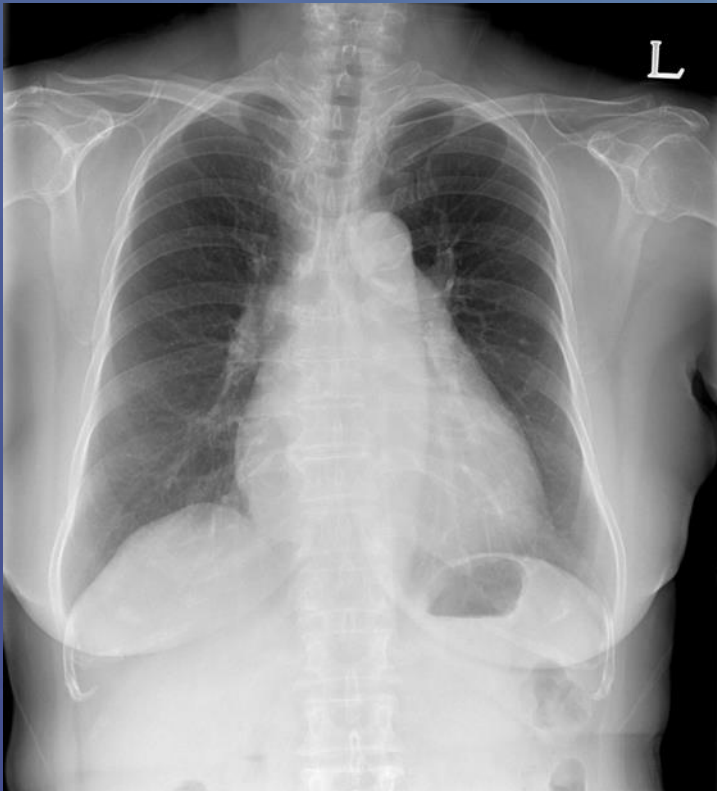
Height 148cm weight 61kg BMI 28

Preoperative CXR & spirometry



| | <u>SPIROMETRY</u> | | | (S2002) | | |
|-----------------------|-------------------|-------|--------|----------------|--------|-------|
| | (S2001) | | | Bronchodilator | | |
| | Pred. | Meas. | %Pred. | Meas. | %Pred. | %Chg. |
| FVC | 2.67 | 1.95 | 73 | | | |
| FEV ₁ | 1.95 | 1.30 | 67 | | | |
| FEV ₁ /FVC | 76 | 66 | 87 | | | |
| FEF _{25-75%} | 1.99 | 0.71 | 36 | | | |
| PEF | 4.85 | 3.63 | 75 | | | |

4-year after operation



Echocardiography

Left Ventricle Normal LV

| | | | | | | | |
|------------|--------------|-------------|--------------|---------------|------------------------------|---------|----------------|
| LVIDs | 28 mm | LVIDd | 46 mm | LA | 45 mm | Aorta | 27 mm |
| LVPWs | 13 mm | LVPWd | 9 mm | ESV | 34 ml | EDV | 93 ml |
| IVSs | 16 mm | IVSd | 9 mm | LVEF | 63 % | LV Mass | 138.4 g |
| P.E. (ant) | mm | P.E. (post) | mm | LV Mass Index | 90.5 g/m ² | | |

Right Ventricle preserved contractility (Tricuspid annulus DTI S velocity=15.2 cm

| | | | |
|-------|----|-------|----|
| RVIDs | mm | RVIDd | mm |
|-------|----|-------|----|

Tricuspid Valve S/P TVP

| | | | | | |
|--------------|----------------|-------------|---------------------------|-------------|----------------|
| TR grade | 3 | TR jet area | 11 cm ² | Peak TR Vel | 4.3 m/s |
| PGsys(RV-RA) | 74 mmHg | Peak E Vel | 135 cm/s | Peak A Vel | 18 cm/s |

- 1.Moderate TR with severe resting pulmonary hypertension :increased pulmonary artery pressure compared with previous study(2015.10.12)**
- 2.Moderate functional MR**

Pulmonary function tests

SPIROMETRY

(S2002)

| | (S2001) | | | Bronchodilator | | |
|-----------------------|---------|-------|--------|----------------|--------|-------|
| | Pred. | Meas. | %Pred. | Meas. | %Pred. | %Chg. |
| FVC | 2.49 | 1.50 | 60 | 1.76 | 71 | 17 |
| FEV ₁ | 1.73 | 0.85 | 49 | 1.05 | 61 | 24 |
| FEV ₁ /FVC | 75 | 56 | 75 | 60 | | |
| FEF _{25-75%} | 1.78 | 0.30 | 17 | 0.78 | 44 | 165 |
| PEF | 4.57 | 2.90 | 63 | 4.10 | 90 | 41 |

LUNG VOLUMES

(S2015)

| | Pred | Meas. | %Pred. |
|-----|------|-------|--------|
| TLC | 3.85 | 3.26 | 85 |
| VC | 1.98 | 1.54 | 78 |
| FRC | 2.48 | 1.93 | 78 |
| ERV | 0.65 | 0.21 | 32 |
| RV | 1.83 | 1.72 | 94 |

DIFFUSING CAPACITY

| | Pred. | Meas. | %Pred. |
|-----------|---------------------------|-------|--------|
| DL | 13.4 | 7.6 | 57 |
| VA(TLC) | | 2.51 | |
| DL/VA | 3.86 | 3.05 | 79 |
| Corrected | 13.4 | 9.1 | 68 |
| Hb | Date 2018040. 9.8 g/dl | | |

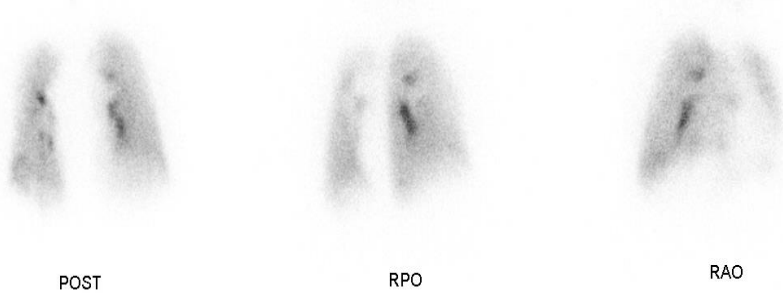
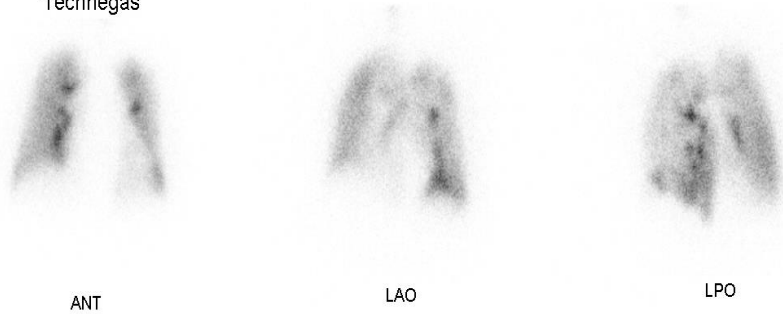
TEST(D) O₂

LPM

| Pulse | SaO ₂ | DISTANCE |
|-------|------------------|----------|
| 52 | 95 | |
| | | |
| 60 | 98 | |
| 63 | 98 | |
| 67 | 98 | |
| 69 | 98 | |
| 70 | 98 | |
| 72 | 98 | |
| 72 | 98 | |
| 73 | 98 | |
| 75 | 98 | |
| 75 | 98 | |
| 75 | 97 | |
| 76 | 98 | 320 |

V/Q scan

Lung Inhalation Scan
Technegas



Lung Perfusion Scan
99mTc-MAA



RHC

| | |
|------------------------|---------------|
| BP | 127/56 |
| mRAP | 16 |
| sPAP/dPAP | 65/17 |
| Mean PAP | 36 |
| LVEDP | 20 |
| CO | 6.17 |
| CI | 4.03 |
| PVR (WU) | 3.12 |
| SvO₂ | 71% |

Case (38/M)

P. I.: 20YA Chronic HBV hepatitis

LC with portopulmonary hypertension

-> 외부병원 liver transplantation w/u

**-> RHC (mPAP 46 mmHg, PCWP 5 mmHg
PVR 4 WUs)**

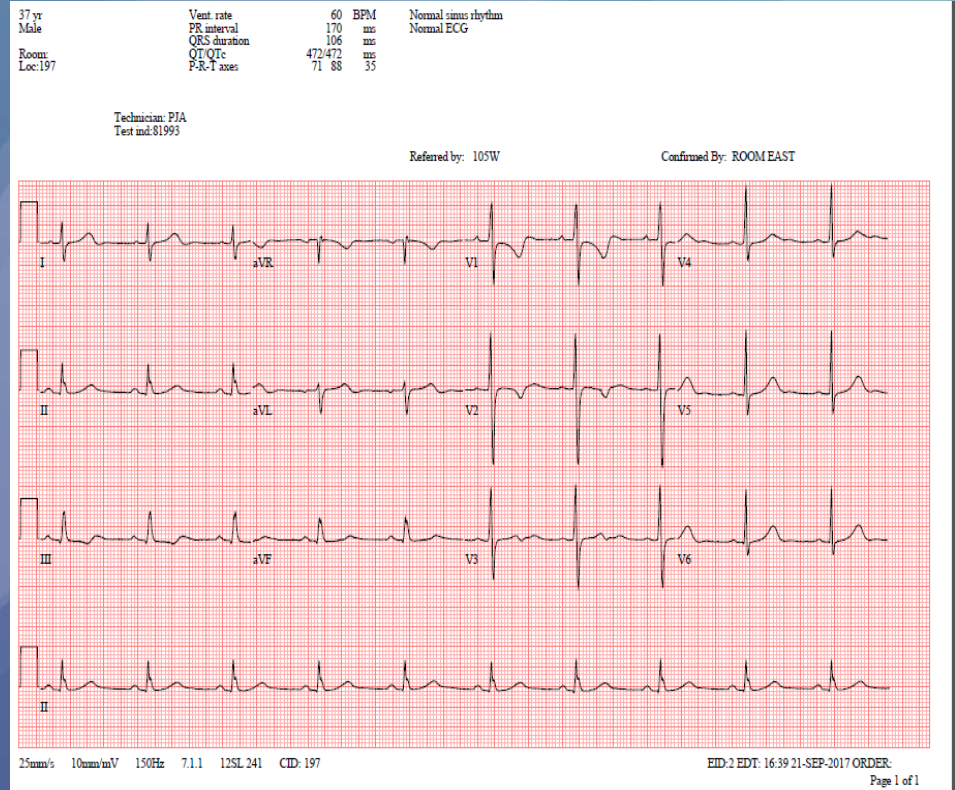
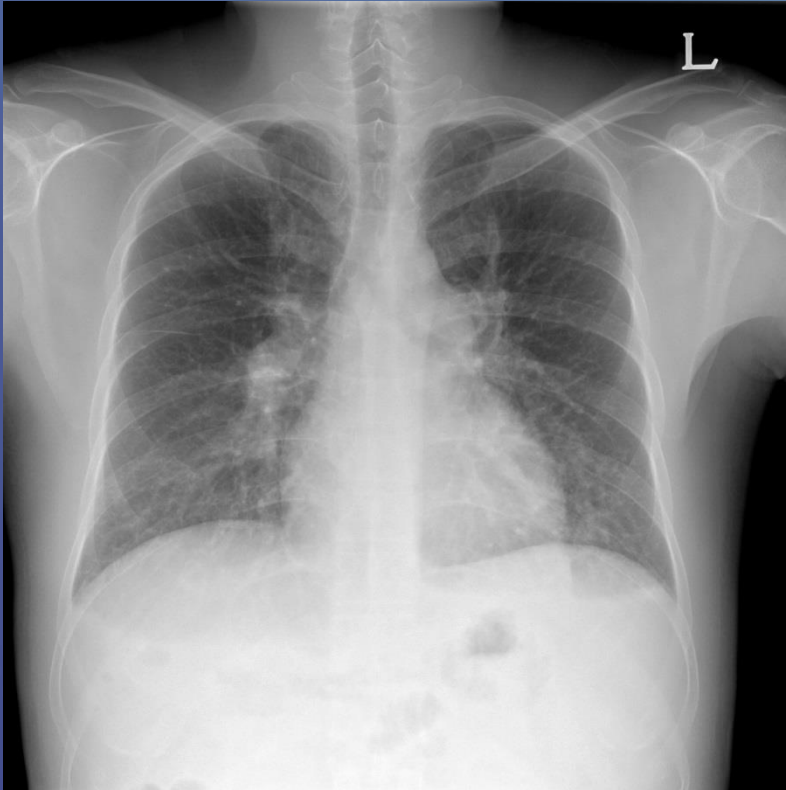
**portopulmonary hypertension으로
macitentan 복용**

Echo: D-shape LV (EF 68%) LA size 45mm

TR Vmax 4.5 m/s sPAP 86mm Hg

Preserved RV contractility

Chest PA and EKG



Initial Laboratory data

CBC WBC 9000 /mm³ , Hb 15.9 g/dL , **PLT 74×10³/mm³**

Coagulation battery **PT 15.4s (59.9 % , 1.34 INR)**

aPTT 37.6 s

Chemical & electrolyte battery

Cr (mg/dL) 1.2 eGFR 108 mol/L

Protein/**albumin** (mg/dL) 6.1/**2.6**

AST/ALT (IU/L) 43/34

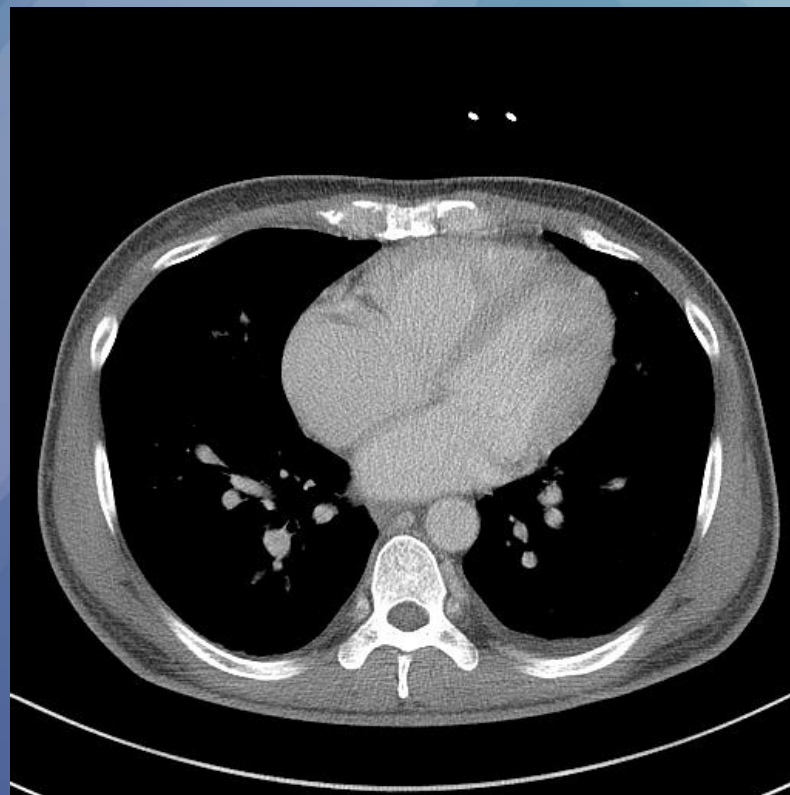
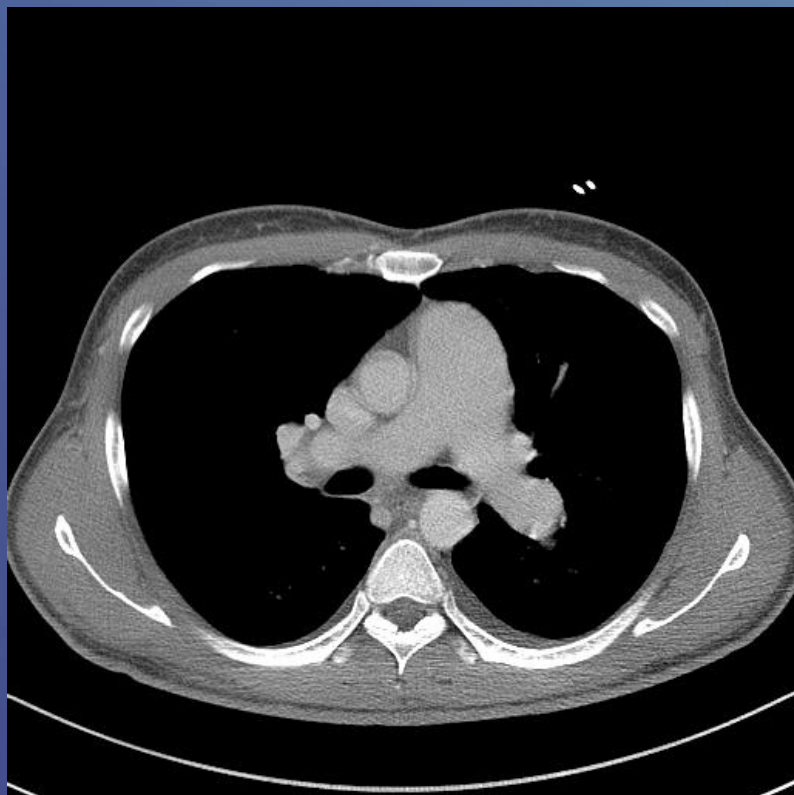
ALP (IU/L) 96

T.bil (mg/dl) 3.2

Na/K/Cl/HCO₃ (mEq/L) 133/4.5/107/20

BNP 53 pg/mL

Chest CT



RHC

| | 2017.05 |
|------------------------|----------------|
| BP | 129/64 |
| sPAP/dPAP | 71/27 |
| Mean PAP | 45 |
| PCWP | 10 |
| mRAP | 3 |
| CO | 10.64 |
| CI | 5.54 |
| PVR (WU) | 2.55 |
| SvO₂ | 79% |

1 week after macitentan+sildenafil

- **ER visit d/t dyspnea**

**Echo: TR Vmax 4.6 m/s sPAP 85mm Hg
Mild RV dysfunction**

Assessment: PoPH with high CO

Plan: stop macitentan, sildenafil

lasix 40mg qd, spironolactone 100mg qd

1-month after diuretics

S: dyspnea (FC I)

O: Echo: TR Vmax 3.4 m/s sPAP 46mm Hg

Decreased PA pressure

6-month after diuretics

S: dyspnea (FC I)

O: Echo: TR Vmax 5.2 m/s sPAP 108mm Hg

Increased PA pressure

Deteriorated RV contractility

F/U RHC

| | 2017.05 | 2018.01 |
|------------------------|--------------|--------------|
| BP | 129/64 | 116/62 |
| sPAP/dPAP | 71/27 | 85/35 |
| Mean PAP | 45 | 51 |
| PCWP | 10 | 8 |
| mRAP | 3 | 7 |
| CO | 10.64 | 9.08 |
| CI | 5.54 | 5.04 |
| PVR (WU) | 2.55 | 4.91 |
| SvO₂ | 79% | 83% |

3-month after ambrisentan

S: dyspnea (FC I)

O: Echo: TR Vmax 4.3 m/s sPAP 74mm Hg

Decreased PA pressure

Improved RV contractility