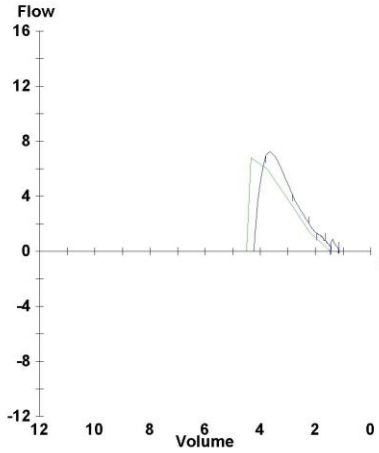
CTEPH:
Complication salvage case

Speaker: Huang Wen-Pin
Institute: Cheng-Hsin General hospital
Taipei Taiwan
(台北振興醫院)

LFT on 2016-10-18

病歷號：[redacted]
姓　名：[redacted]
性　別：Female　齡：47
身　高：159 體　重：56.3
病　房：51-58
日　期：2016 10 18

Pulmonary Function Report



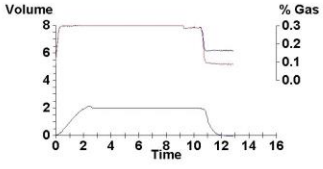
Spirometry

		Ref	Pre Meas	Pre % Ref	Post Meas	Post % Ref	Post % Chg
FVC	Liters	3.03	3.10	102			
FEV1	Liters	2.53	2.61	103			
FEV1/FVC	%	81	84				
FEF25-75%	L/sec	2.91	2.71	93			
PEF	L/sec	6.79	7.21	106			
PIF	L/sec						
VC	Liters	3.03	3.10	102			
FVL ECode			000000				

Lung Volumes

		Ref	Pre Meas	Pre % Ref	Post Meas	Post % Ref	Post % Chg
Vt	Liters		1.19				
VC	Liters	3.03	3.10	102			
TLC	Liters	4.48	4.22	94			
RV	Liters	1.45	1.12	77			
FRC PL	Liters	2.57	2.09	81			
ERV	Liters	1.14	0.58	51			
IC	Liters	1.89	2.14	113			

Diffusion

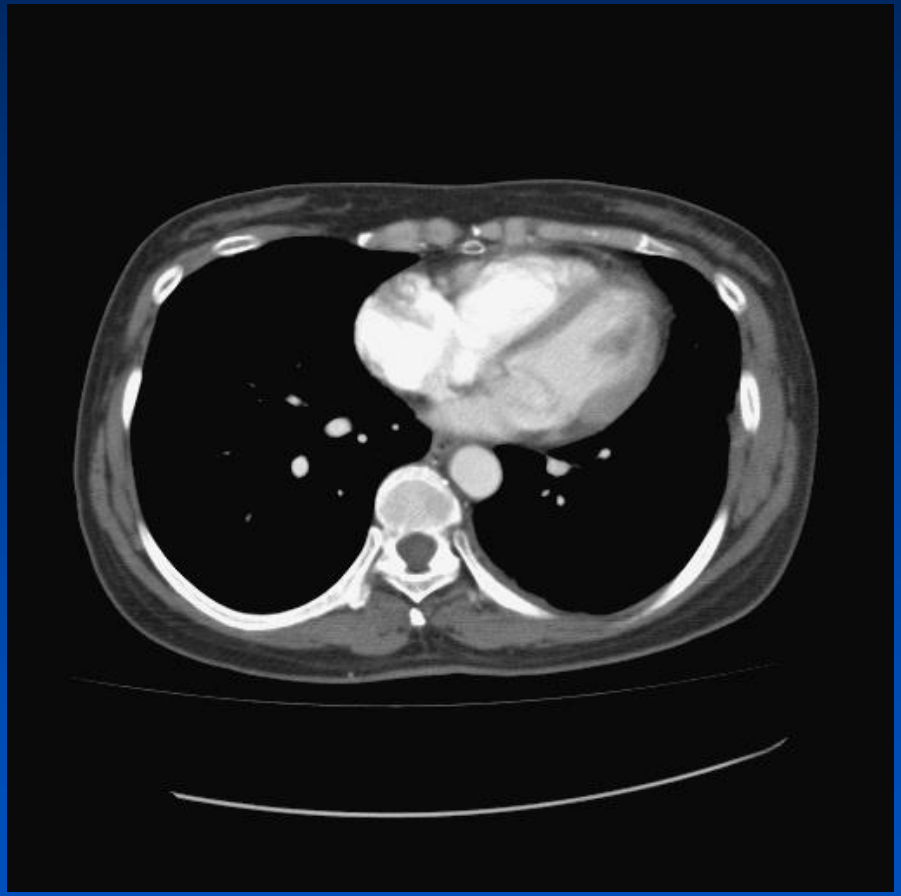


		Ref	Pre Meas	Pre % Ref	Post Meas	Post % Ref
DLCO	mL/mmHg/min	24.8	15.8	64		
DL Adj	mL/mmHg/min		15.8			
VA	Liters	4.48	3.60	80		
DLCO/VA	mL/mHg/min/L	5.01	4.40	88		
DL/VA Adj	mL/mHg/min/L		4.40			
IVC	Liters		2.17			

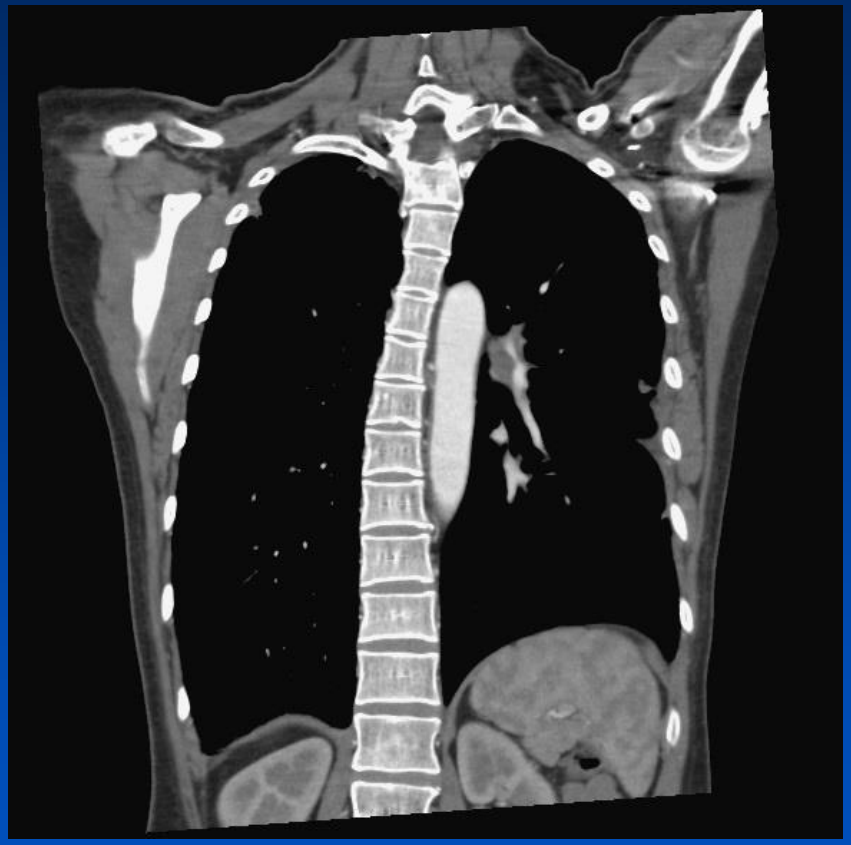
Chest CT on 2016/10/17

- Multiple thrombi in bilateral main, right upper and bilateral lower pulmonary arteries.
- Multiple patch ground-glass and small nodular opacities in peripheral LUL and LLL. R/I infecti ous process. Advise follow-up.
- Multiple fibrotic foci in RUL and left lung. Sugg estive of chronic inflammation.
- Mild pleural effusion, left, and pleural thickenig of bilateral upper chest.

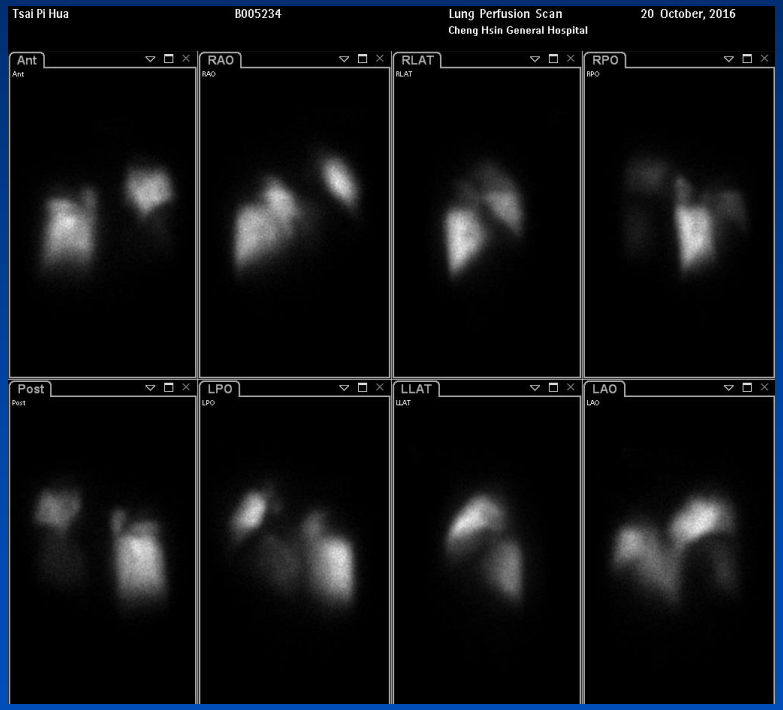
Chest CT on 2016/10/17

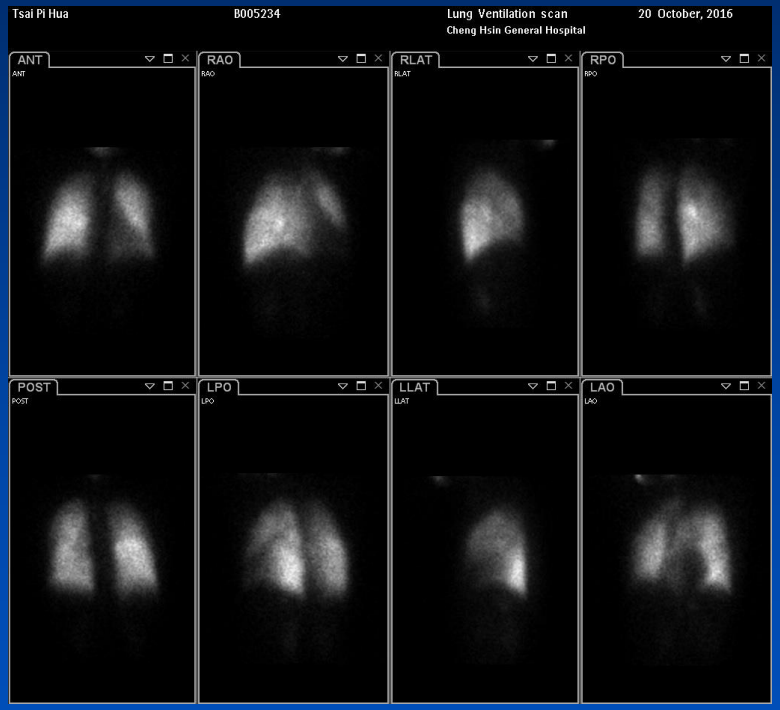


Chest CT on 2016/10/17



V/Q scan on 2016-10-20





Coronary angiography on 2016-10-27

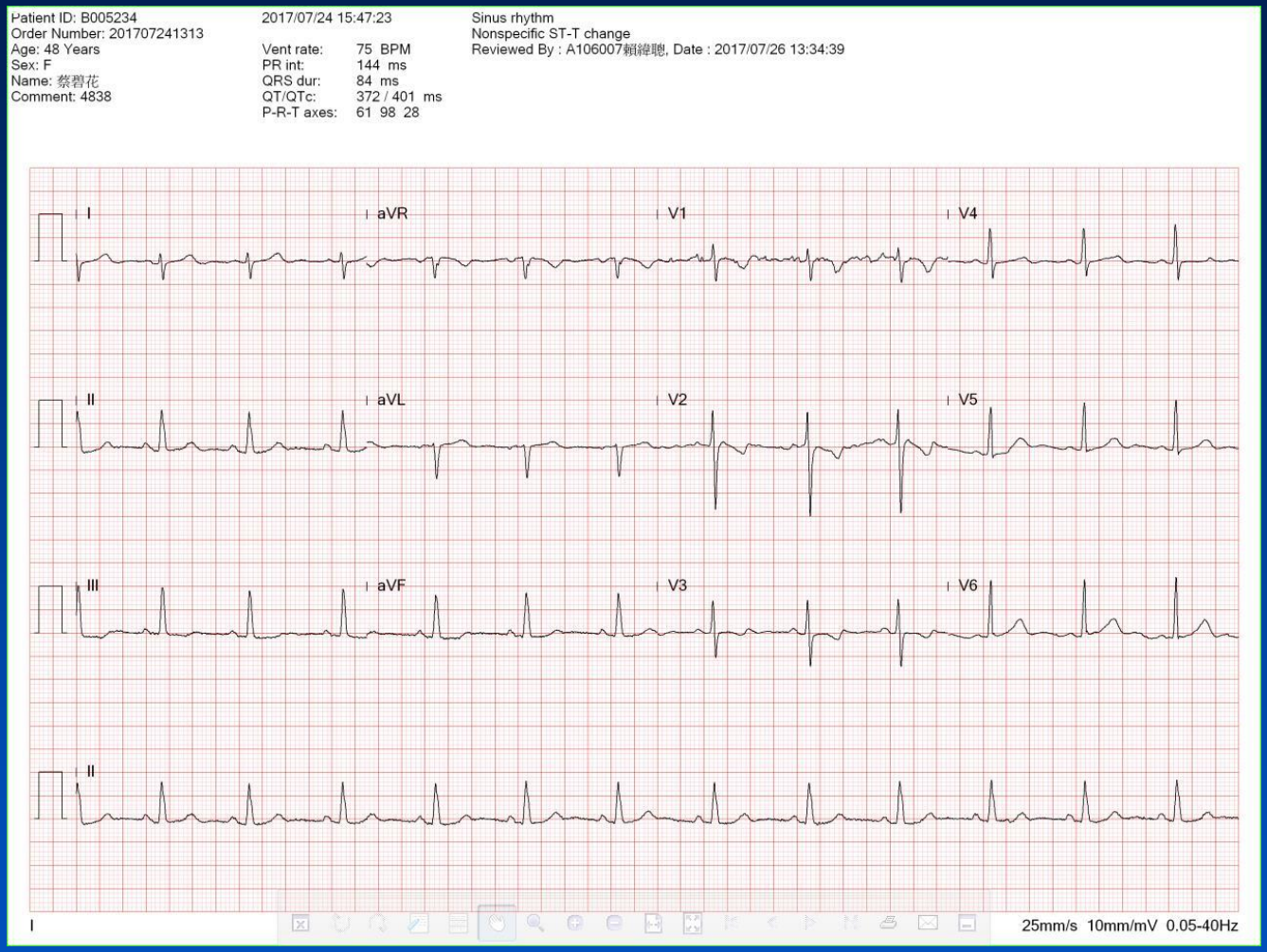
- CAG: LM: Normal
- LAD: Normal
- LCX: Normal
- RCA: Normal
- LVG: Normal LV systolic function, LVEF= 70%, No MR, LVEDP= 12 mmhg

- Vasodilatror test: Negative. [mPAP: 33 --> 27 mmhg]
- CO= 4.76 CI= 3.03
- AOP: 130/78/102 O2 Sat 90.4 %
- PAP: 69/13/33 O2 Sat: 72.3 %
- LVP: 136/3/12 RVP: 71/3/9 PCWP: 14/10/9 RAP: 13/9/8
- PVR: 403/80
- Dx: Pulmonary hypertension Group 4 (CTEPH).

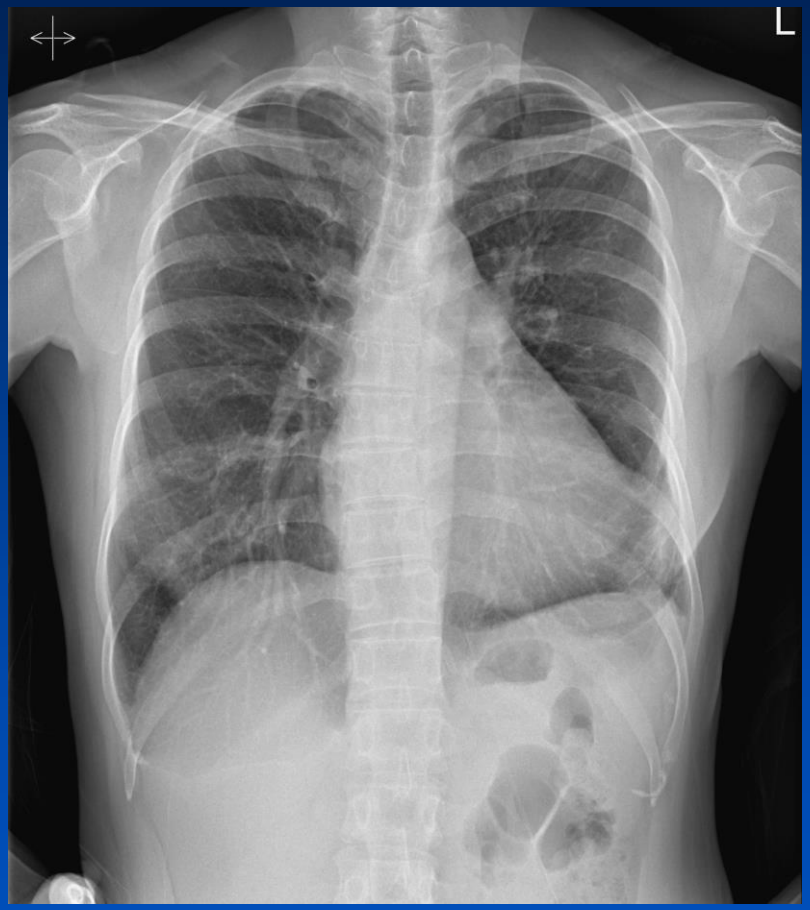
V/Q scan on 2016-10-20

- The lung ventilation study (inhalation of radiotracer with at most 10% of 40 mCi Tc-99m DTPA aerosols)
  - There is normal distribution of radiotracer in both lung fields.
  - In addition, some central hot spots over bil. peri-hilar regions are also seen, most likely due to central localized deposition of aerosol particles in the large airways.
- Follow-up Tc-MAA lung scan
  - mismatched perfusion defects over both lungs, esp. bil. upper & left lower lungs.
- Imp: Mismatched V/P defects in both lungs may suggest high probability for pul. embolism, esp. bil. upper & left lower lungs.

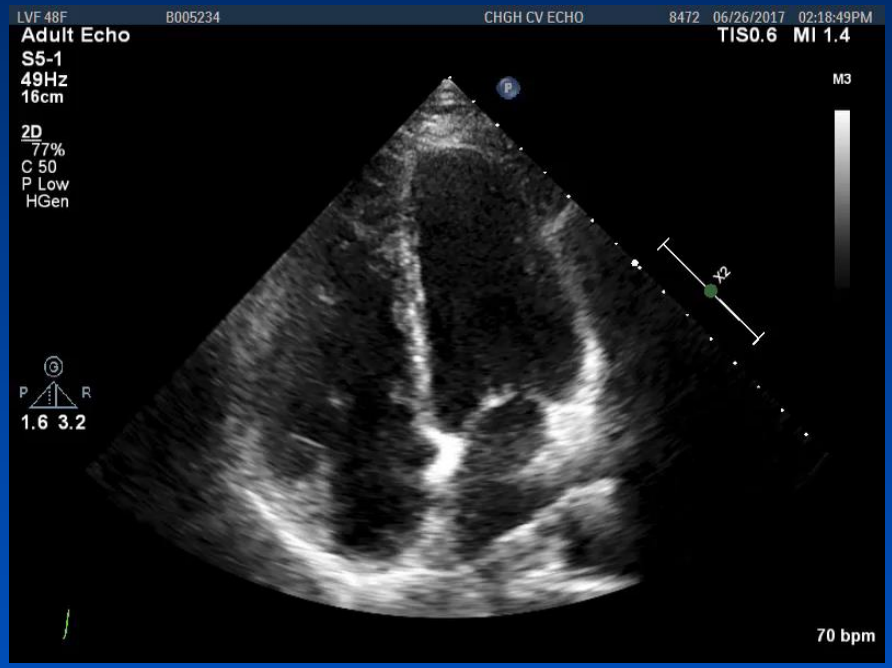
ECG
2017-7-24

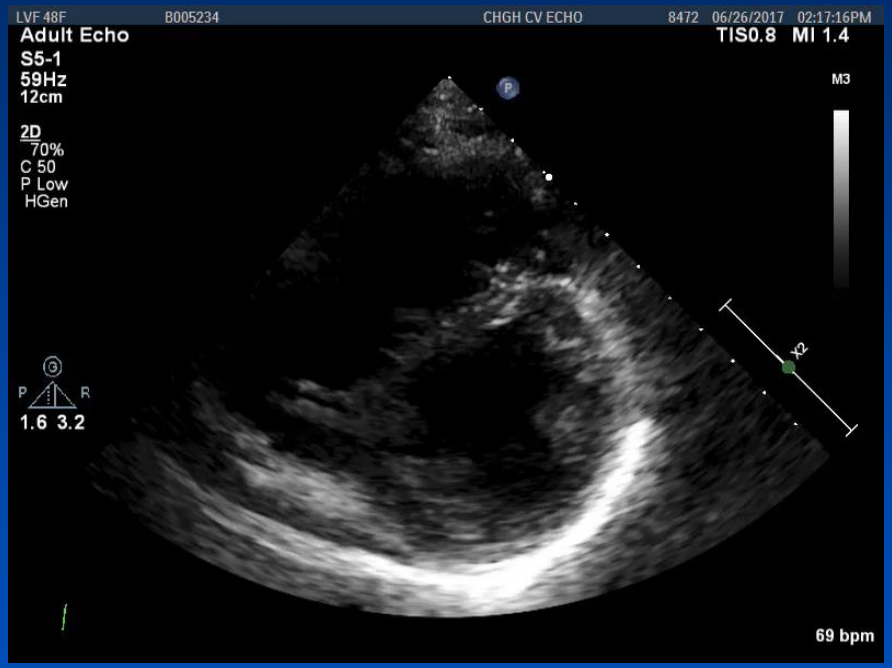

Patient ID: B005234
Order Number: 201707241313
Age: 48 Years
Sex: F
Name: 蔡碧花
Comment: 4838
2017/07/24 15:47:23
Vent rate: 75 BPM
PR int: 144 ms
QRS dur: 84 ms
QT/QTc: 372 / 401 ms
P-R-T axes: 61 98 28
Sinus rhythm
Nonspecific ST-T change
Reviewed By : A106007賴緯聰, Date : 2017/07/26 13:34:39
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
II
I
25mm/s 10mm/mV 0.05-40Hz

CXR on 2017-6-26



Echocardiography on 2017-6-26

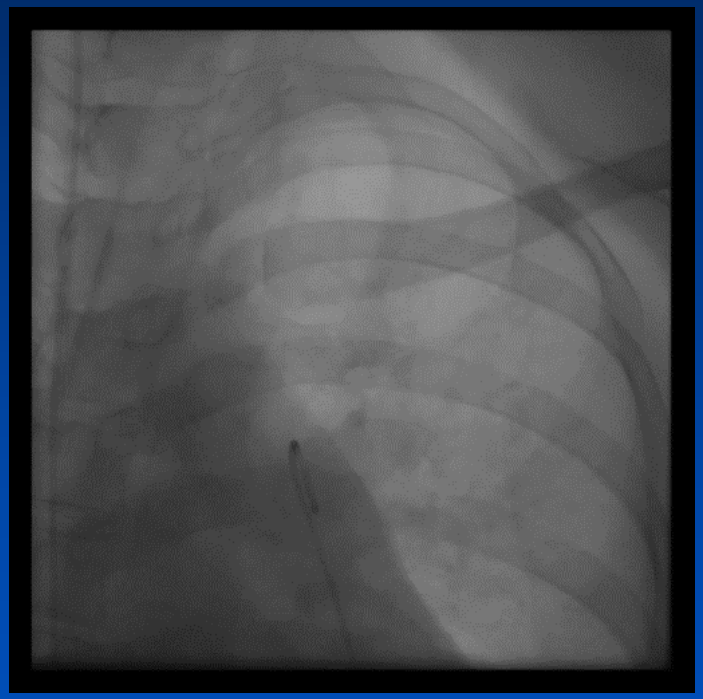


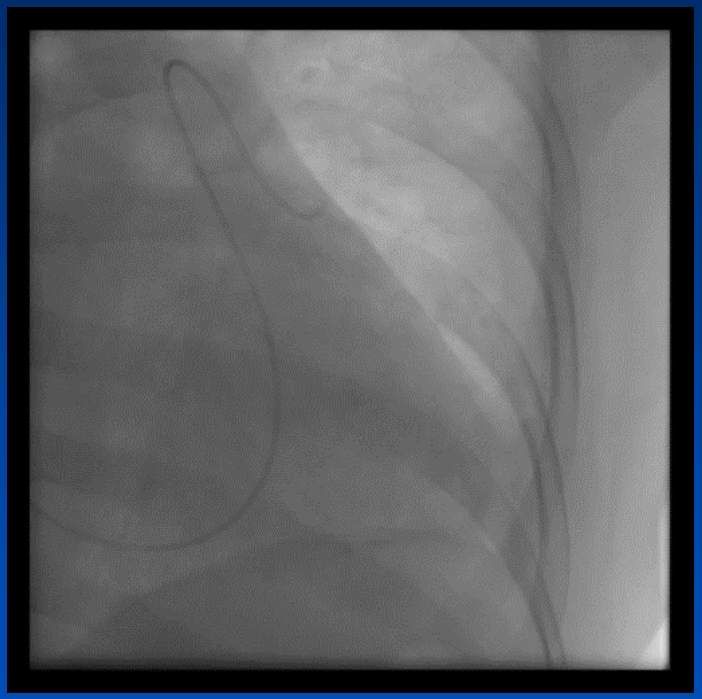


Pulmonary arterial angiography on 2017-7-25

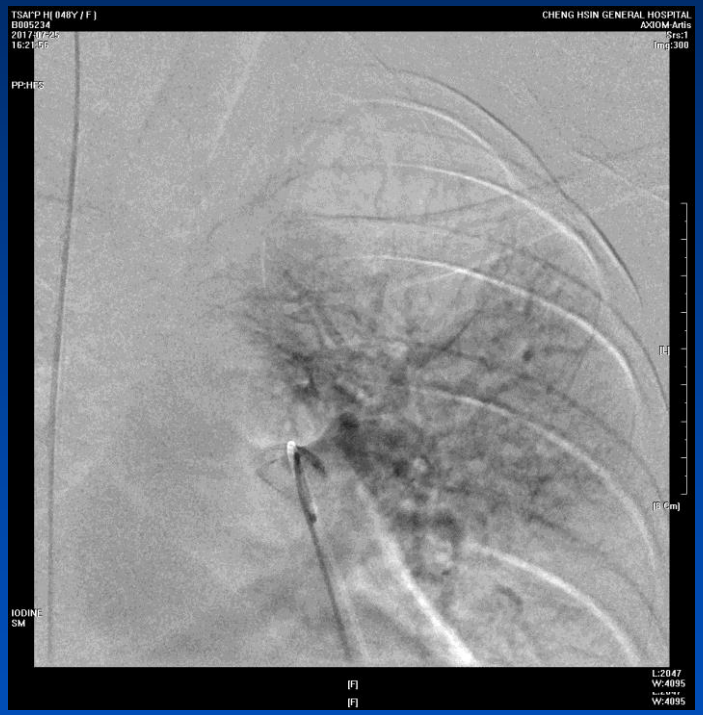
- PA angiography:
  - Right side PA: A2 near total occlussion, A5 critical stenosis, A 9 80% stenosis.
  - Left side PA: A3 70% stenosis, A6 total occlussion, A5 90% st enosis. A7,A8, A10 total occlusion, A9 89% stenosis. .
- Right heart study data:
- CO= 4.64 CI= 2.94
- O2 Sat : PA 77.2 % AO 97.0 %
- Pressure:
  - RA 9/6/6 RV 87/1/11, PA 80/12/36, PCWP: 10/10/12, LV NA AOP: 111/73/86
  - PVR 413.8/80, SVR= 1379/80 TPVR= 620.7/80 TSVR= 1482 /80
- Dx: CTEPH (PH Group IV)
- Tx: BPA

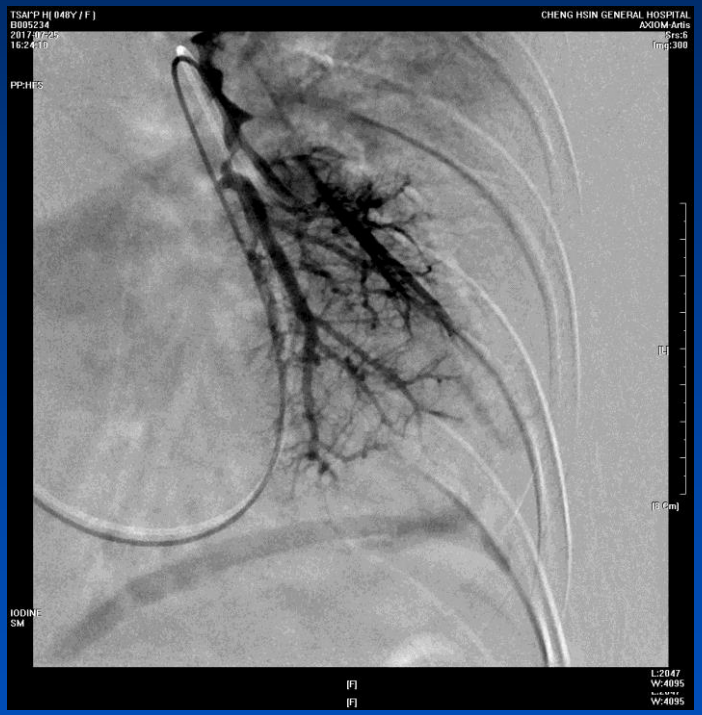
Pulmonary arterial angiography on 2017-7-25



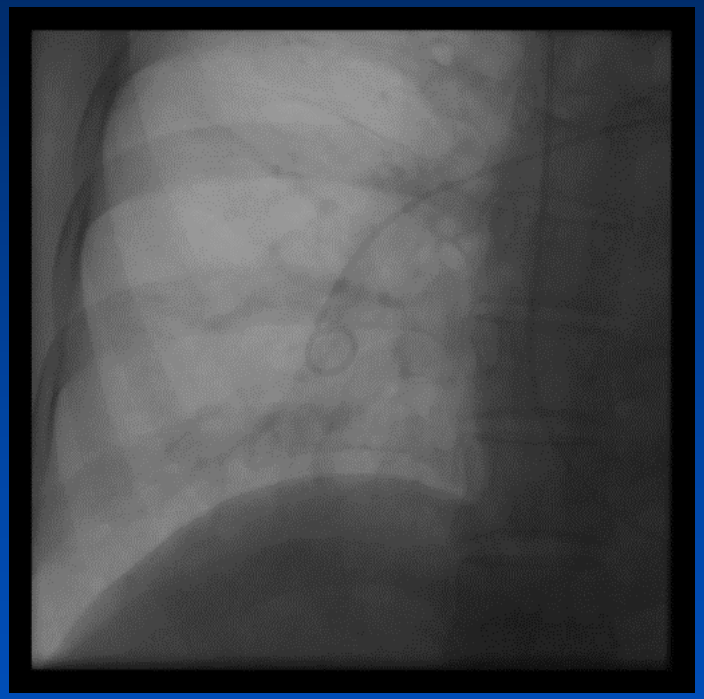


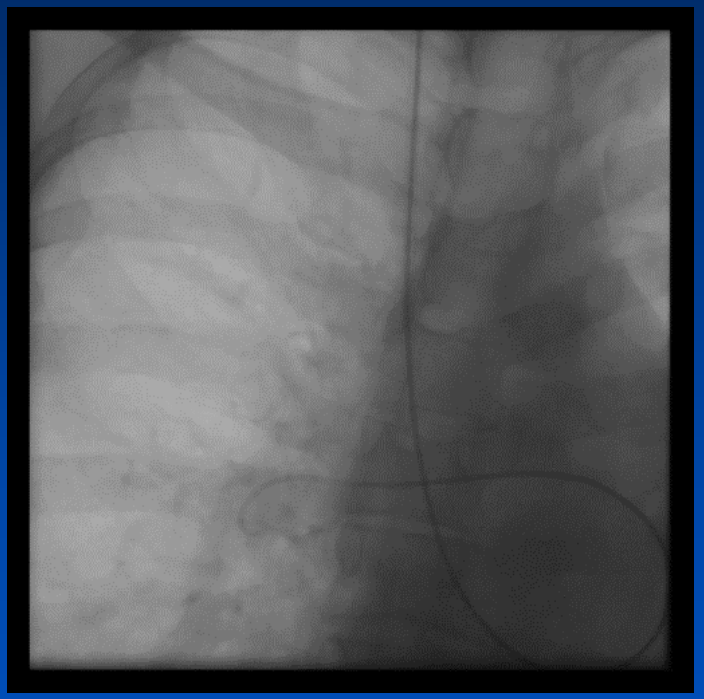
Pulmonary arterial angiography on 2017-7-25



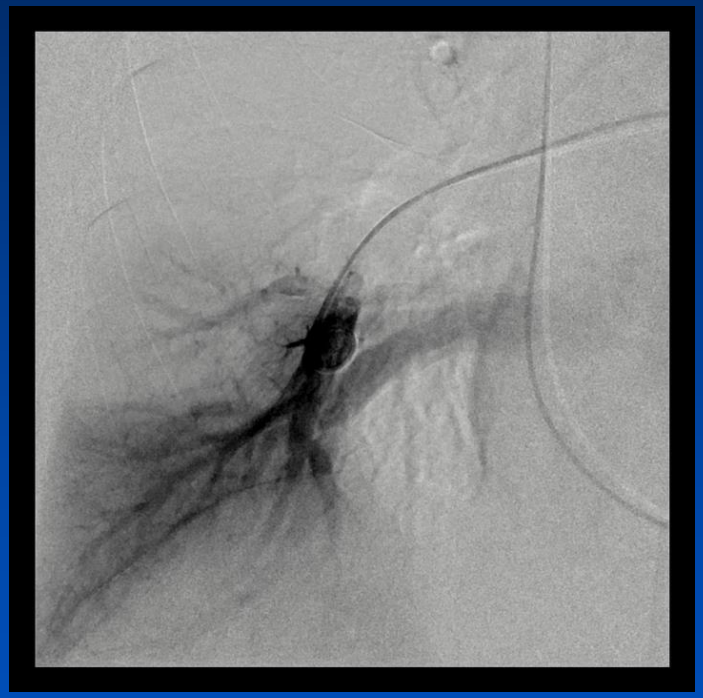


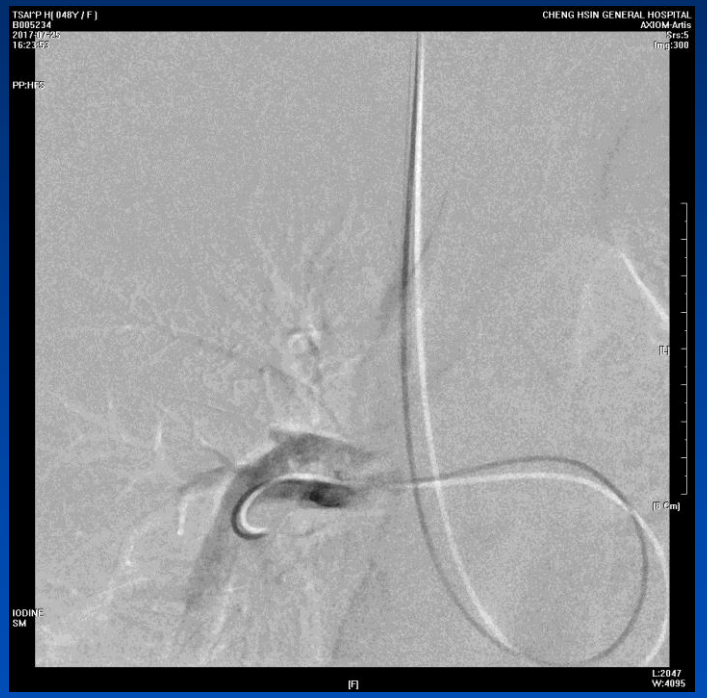
Pulmonary arterial angiography on 2017-7-25





Pulmonary arterial angiography on 2017-7-25

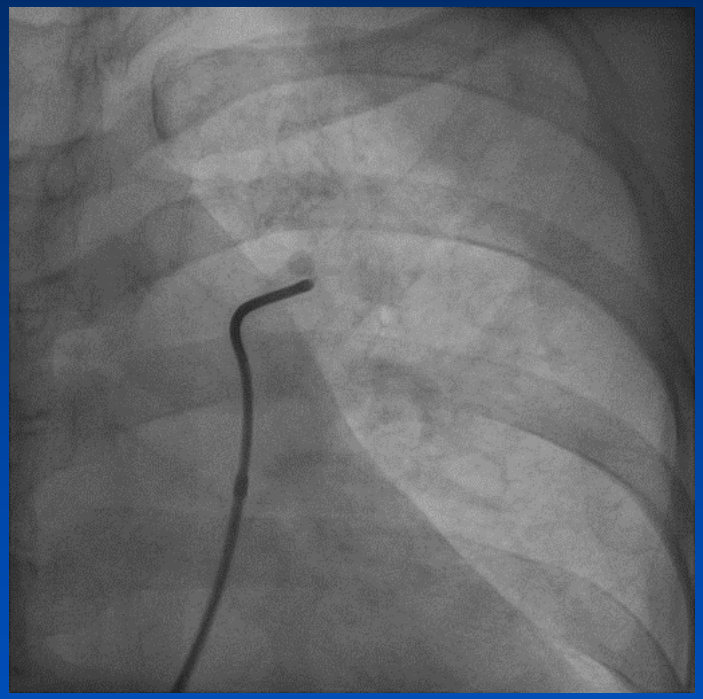


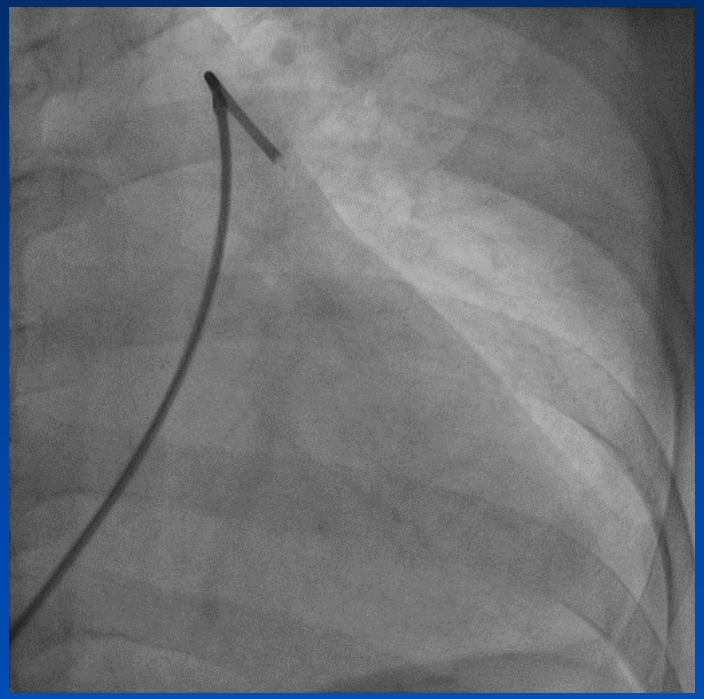


Balloon pulmonary angioplasty on 2017-8-19

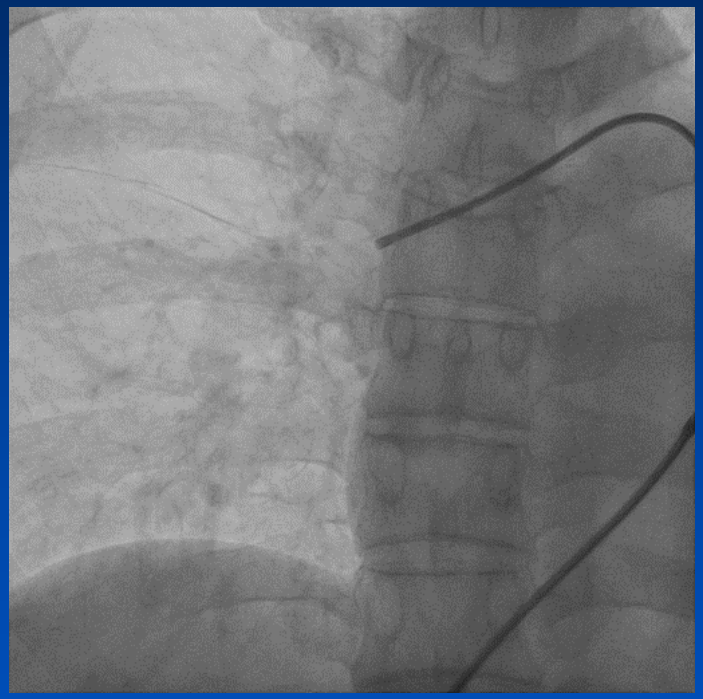
- Sheath :9 Fr Daig sheath and 6 Fr 75 cm Teru mo sheath
- Guiding cath: MP 6 Fr
- Gudewire: 0.014 BMW wire x 1
- Balloon: 2.0 x 20 mm mini-trek x 1, 3.0 x 20 mm trek x 1, 4.0 x 20 mm Emerge x 1
- Right heart study data: PAP(L't) : 59/32/41 P AP(R't): 55/14/29

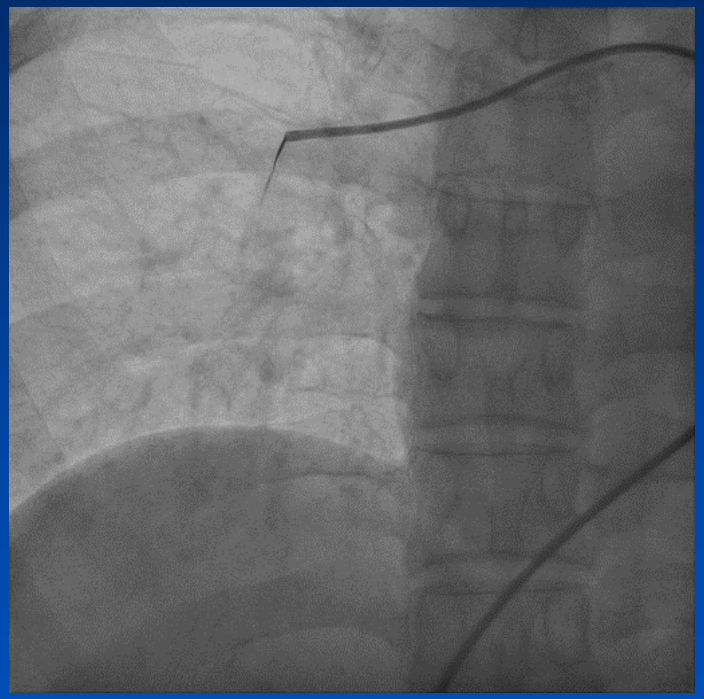
BPA on 2017-8-19



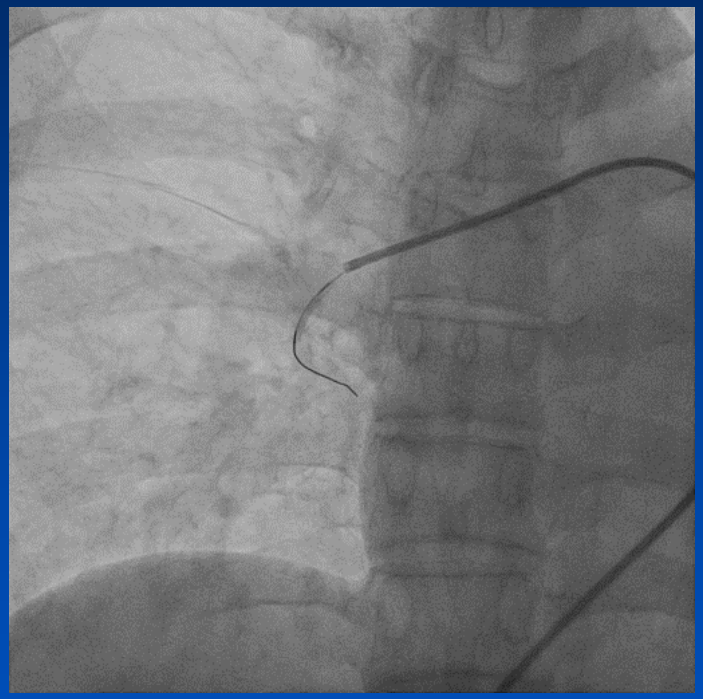


BPA on 2017-8-19

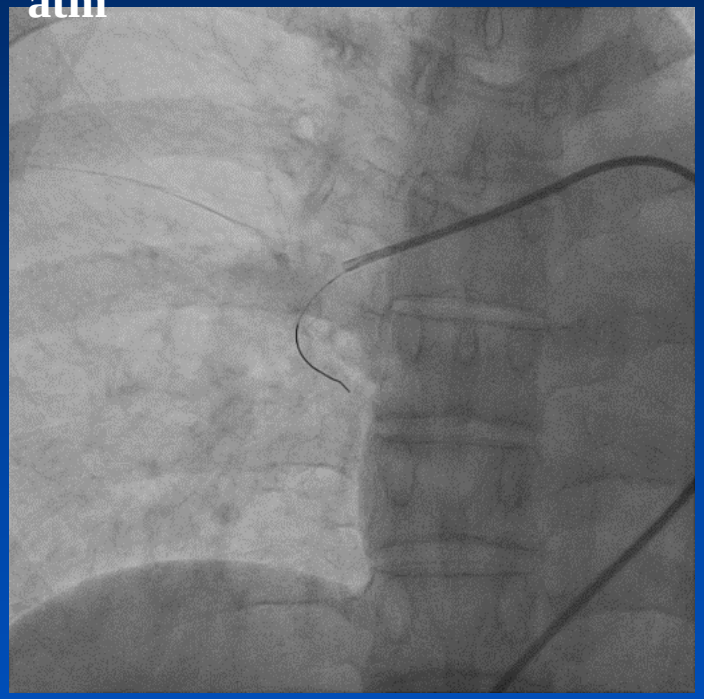




BPA on 2017-8-19

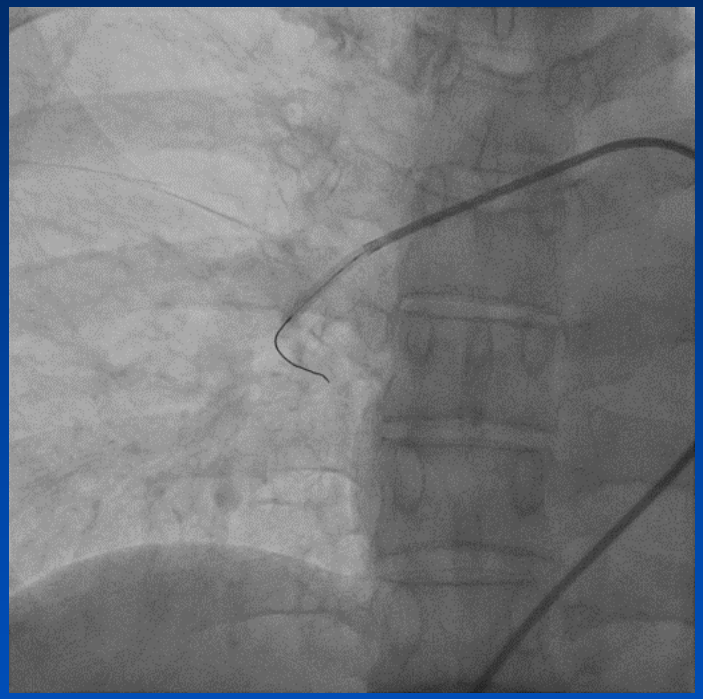


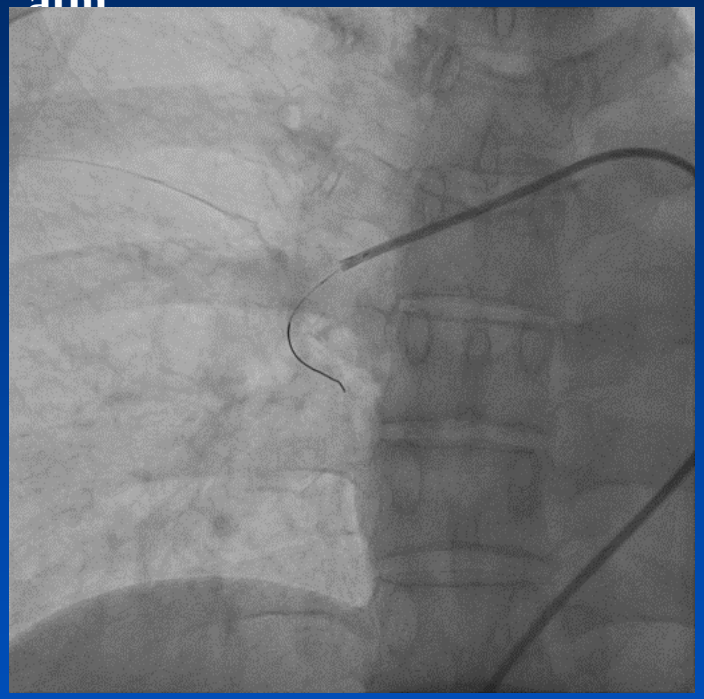
Right A5: 2.0 x 20mm balloon with 8 atm



BPA on 2017-8-19

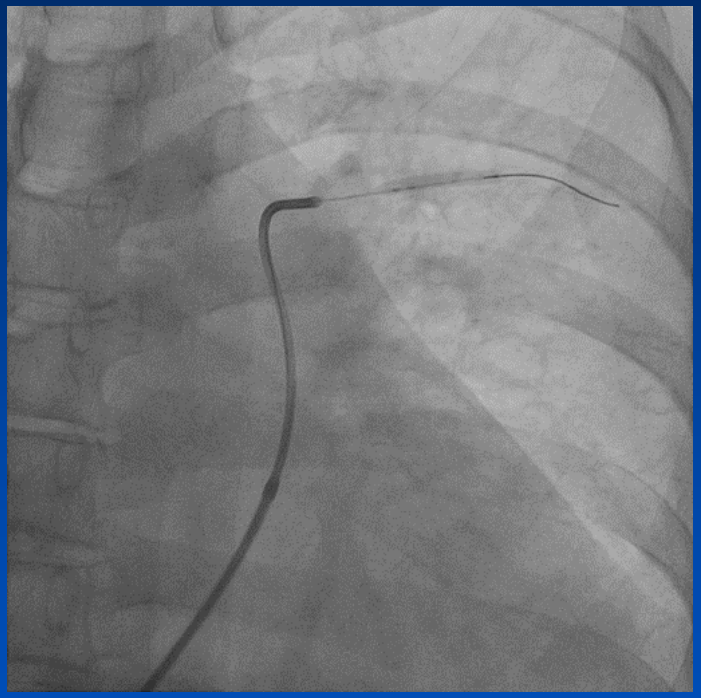
Right A5: 3.0 x 20 mm balloon with 8 atm

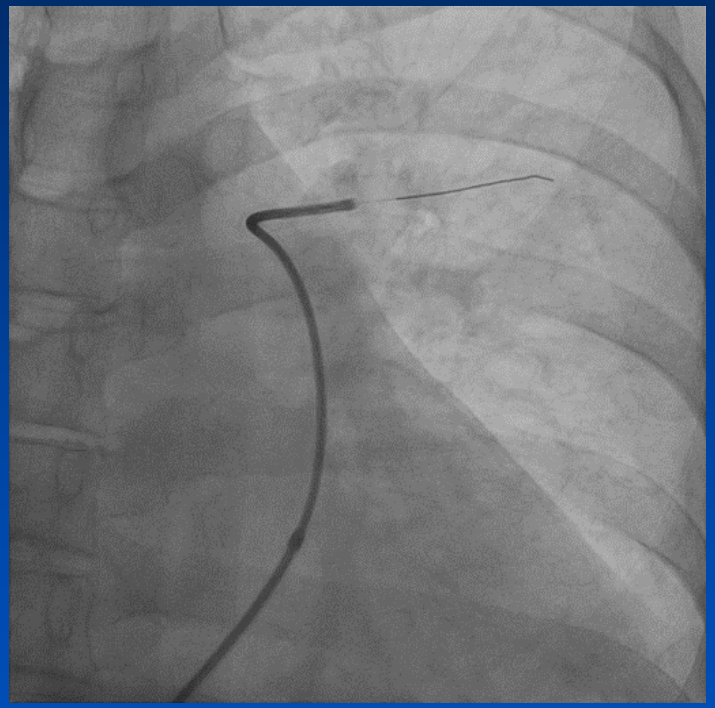




BPA on 2017-8-19

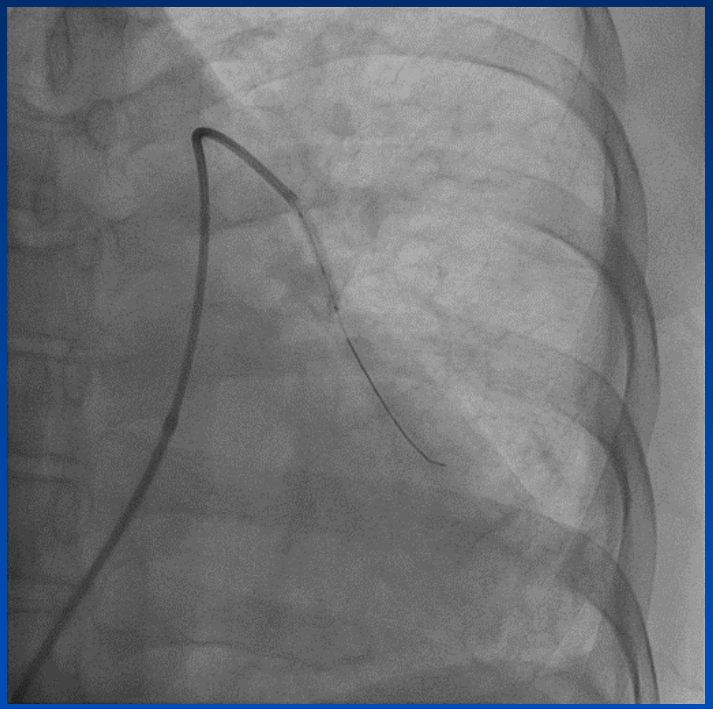
L't side A1+2: 2.0 x20 mm and 3.0x20 mm balloon with 8 atm

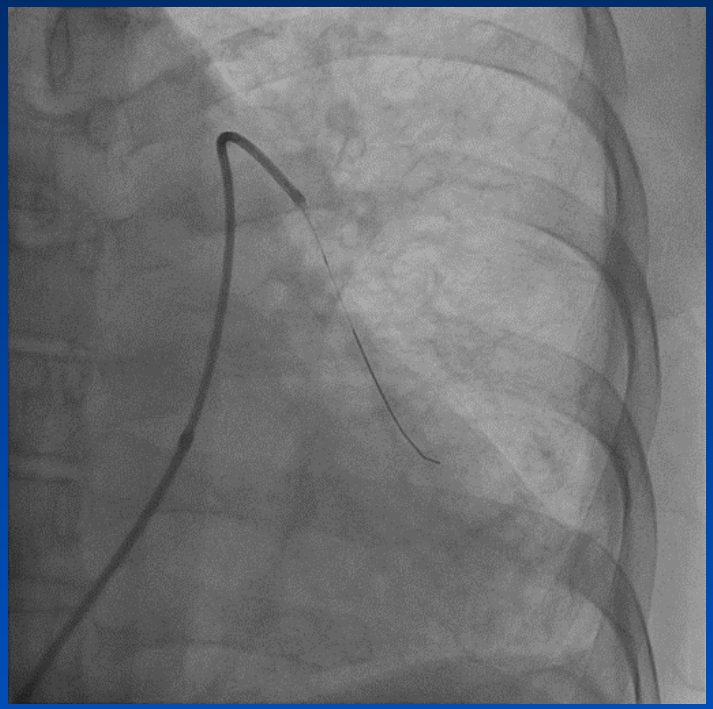




BPA on 2017-8-19

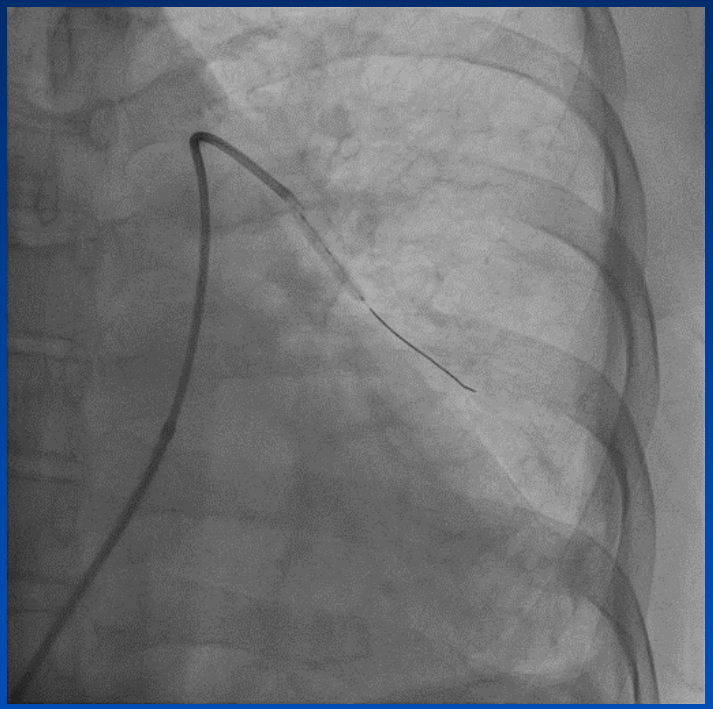
L't A5: 4.0 x 20 mm balloon with 10 atm.

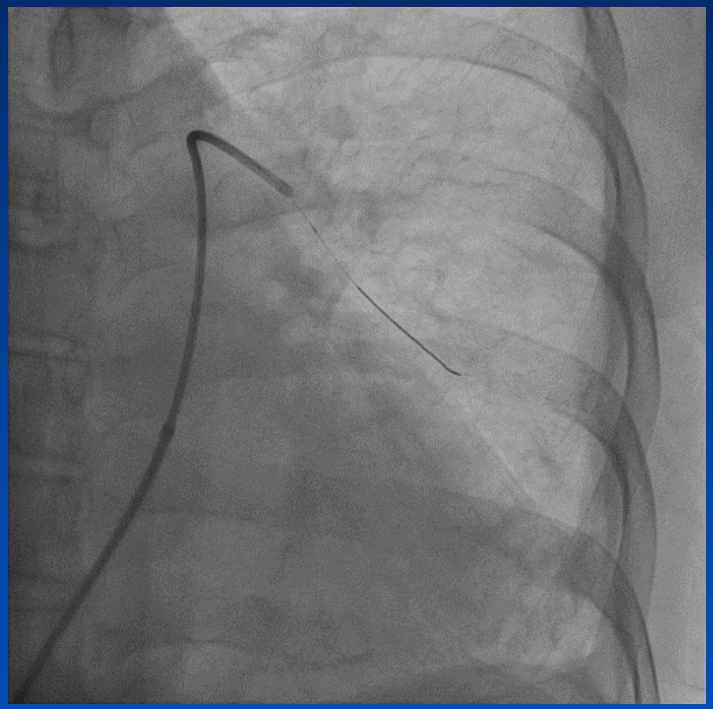




BPA on 2017-8-19

L't A5: 4.0 x 20 mm balloon with 10 atm.

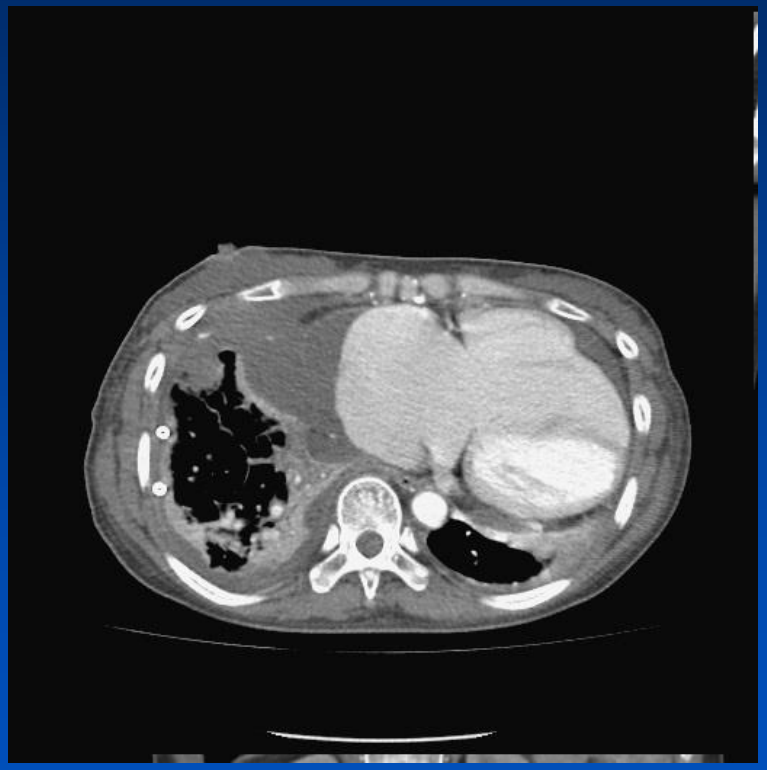




Chest CT on 2018/03/01

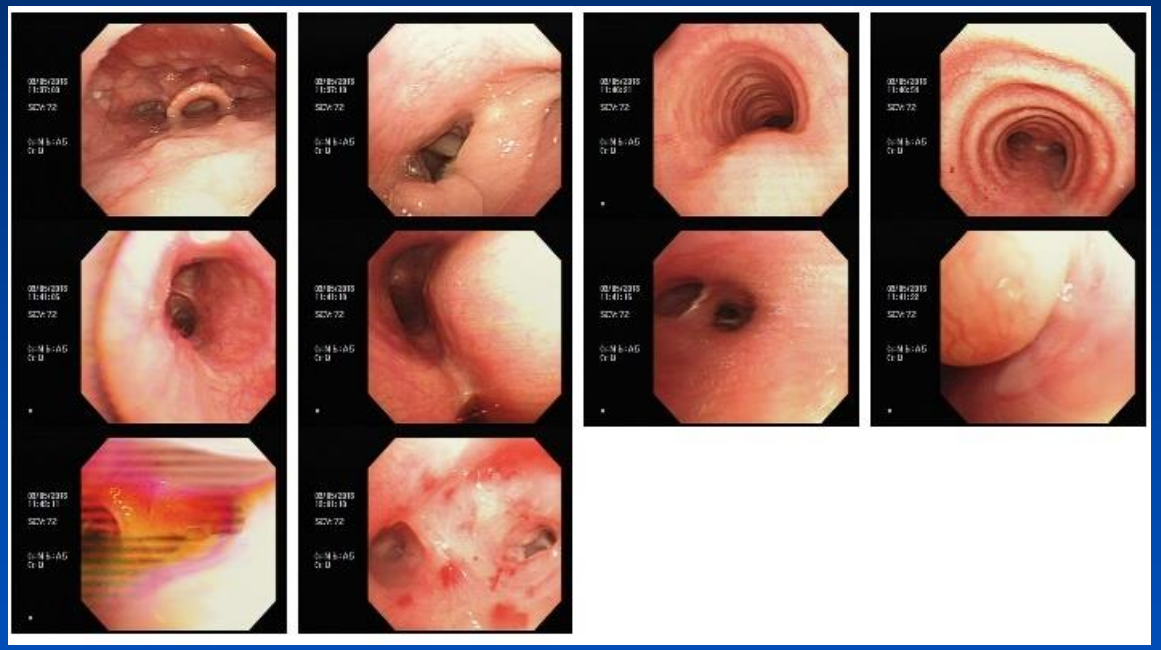
- Irregular poor enhancing mass is depicted at pulmonary trunk and right pulmonary artery, and right atrium. and extension to right hilum and subcarina region.
  Compatible with pulomnary artery intimal sarcoma.
- Post pigtail drainage of right pleura, and post operation of pericarium is noticed. Increase of right pericardiac eff -usion and bilateral pleural effusion is depicted, esp. lob -luated fluid in right pleural and right major and minor fis -sure. Seeding cannot be ruled out.
- Focal passive atelectasis at right lower lung is depicted. Infiltraton of right lower lung is depicted. Either due to lymphangitic spreading or post radiation change.
- Mosaic enhancement of liver parenchyma is noticed. Congestion due to venous return compromised conside -red.
- Subcuatneous edema of trunk is noticed.

Chest CT on 2018/03/01



Bronchoscope on 2018-3-8

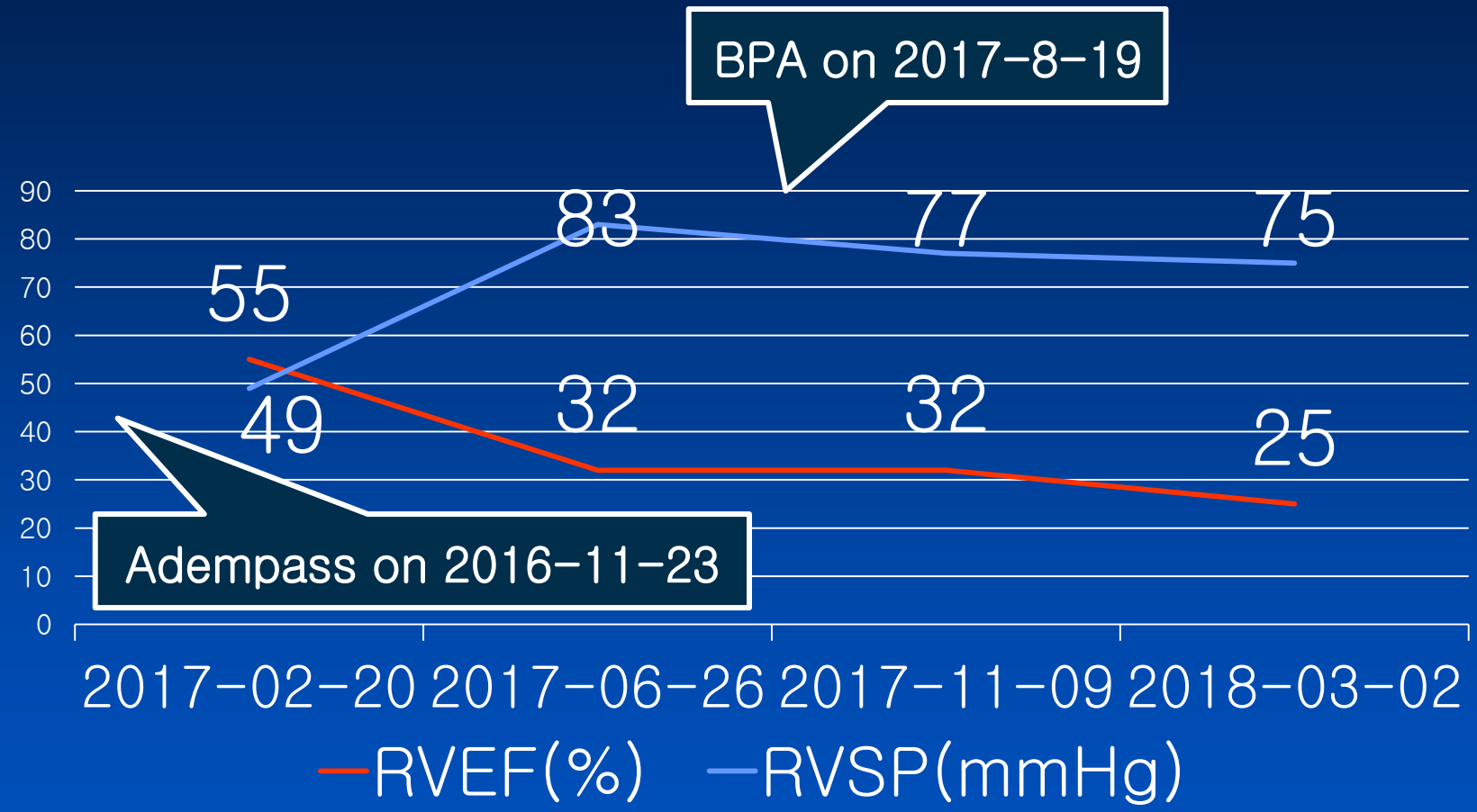
- Normal vocal cord, t rachea & carina
- Left bronchus: essential negative findings
- Right bronchus:
  - 2 protruding sub mucosa nodules with smooth surf ace over RUL br onchus
  - a tumor growth with total obstruc tion over B3 orifi ce



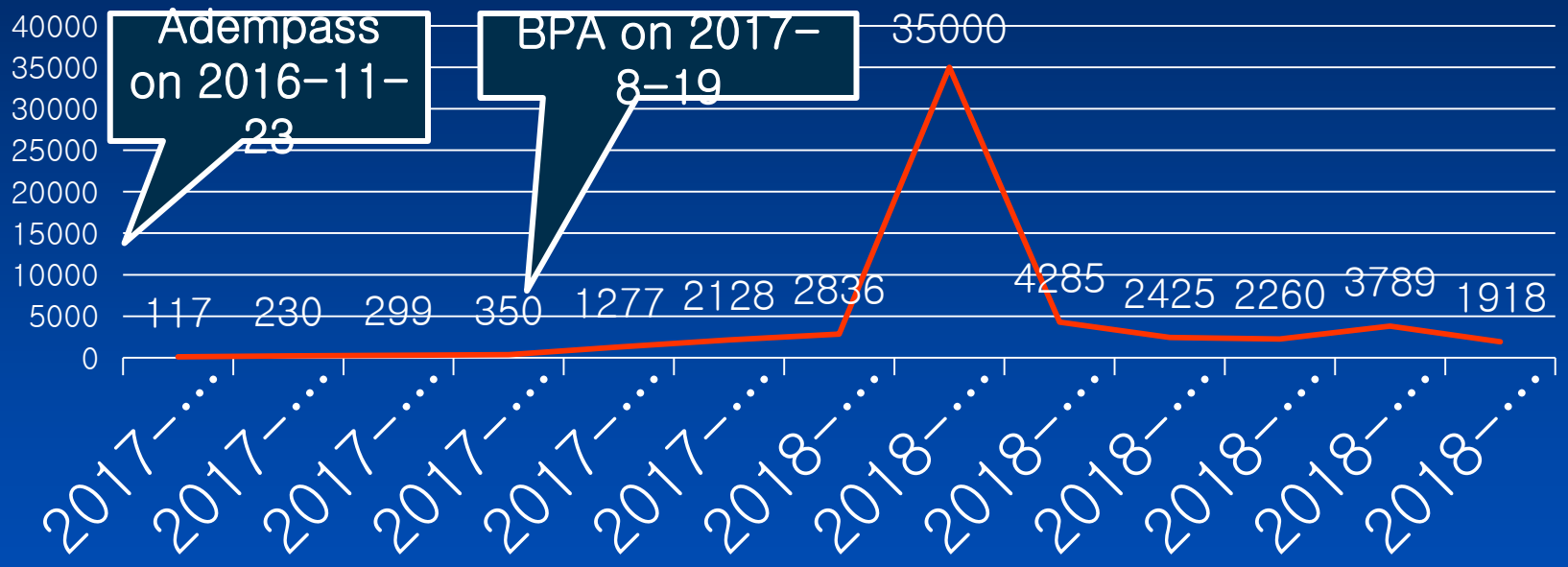
Pathologic report on 2018/03/08

- Lymph node, subcarina, right: anthracosis
- Lymph node, hilar, right: spindle cell sarcoma
- GROSS:
  - The specimen submitted for frozen section contains two lymph node s tissue in fresh state. The first one is right subcarinal lymph node a nd the second specimen is hilar lymph node. Grossly, they are gray and elastic firm and measures 0.7x0.5x0.3 cm in size.
- All for frozen section.
  - The frozen section of the right subcarinal lymph node reveals anthra cosis only, the hilar lymph node shows metastatic spindle cell malign ancy.
  - Microscopically, after fixation, the permanent section of subcarinal L N confirms the diagnosis of anthracosis. The hilar lymph node shows spindle cell tumor with hyperchromatic nuclei and frequent mitoses. I HC study reveals the tumor cells are negative to CK, TTF-1, p40, S-100, CD163, CDK4 and DOG-1 but positive to CD31, MDM2. It is compatible with a metastatic pulmonary sarcoma and a pulmonary a rtery intimal sarcoma is highly suspected.

Echocardiography

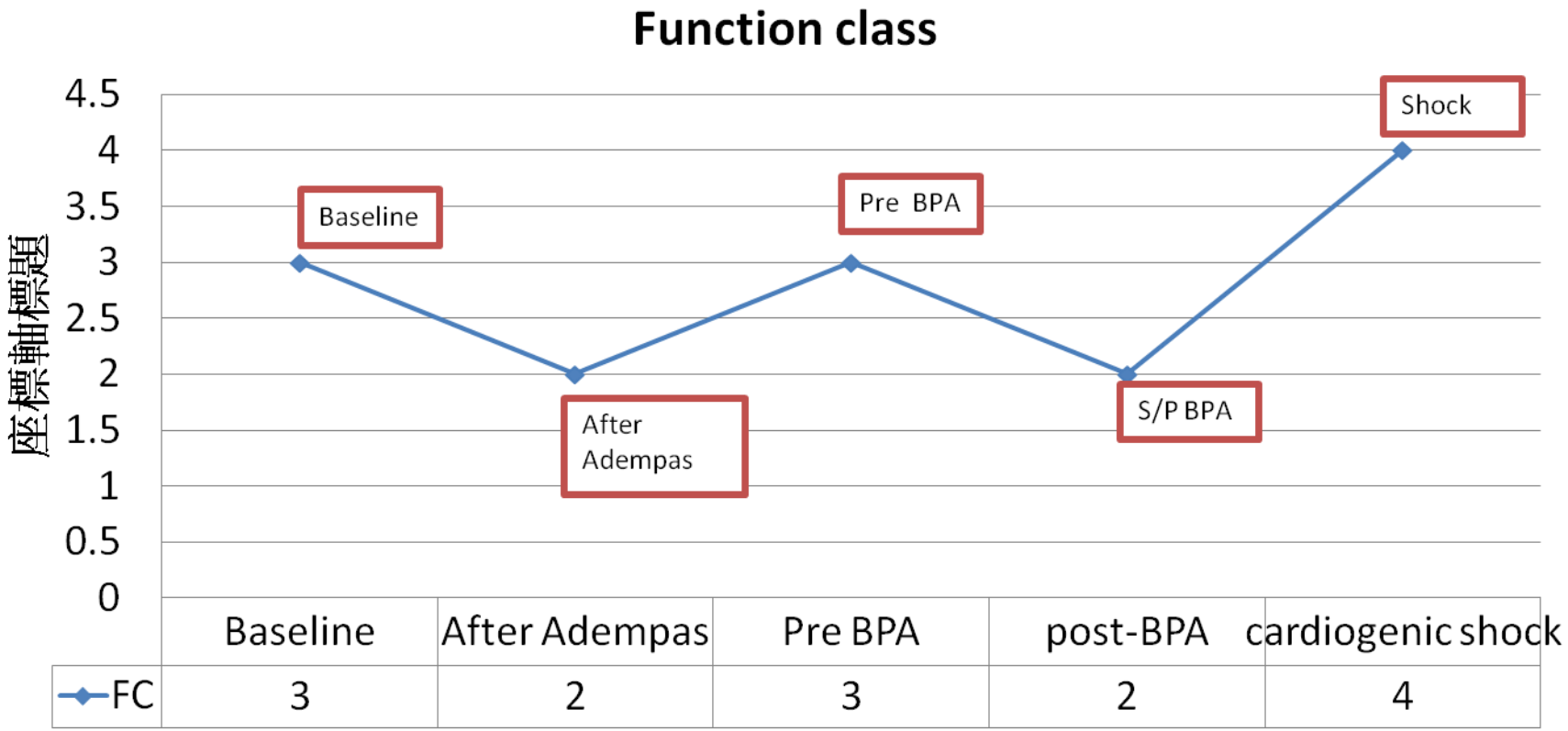


BNP


Adempass on 2016-11-23
BPA on 2017-8-19
40000
35000
30000
25000
20000
15000
10000
5000
0
117
230
299
350
1277
2128
2836
35000
4285
2425
2260
3789
1918
2017-…
2017-…
2017-…
2017-…
2017-…
2017-…
2018-…
2018-…
2018-…
2018-…
2018-…
2018-…
2018-…

6MWD

	6MWD	Borg sale
Adempass since 2016/11/23		
2017/3/20	400	0->2
2017/6/26	360	1->3
BPA on 2017-8-19		
2017/11/9	430	0->3


Function class
座標軸標題
4.5
4
3.5
3
2.5
2
1.5
1
0.5
0
Baseline
After Adempas
Pre BPA
S/P BPA
Shock
Baseline
After Adempas
Pre BPA
post-BPA
cardiogenic shock
FC
3
2
3
2
4

Discussion Points

- Primary pulmonary angiosarcoma and CTEPH
- Balloon pulmonary angioplasty (BPA)

- Tumoral PH includes pulmonary micro - embolism and pulmonary thrombotic microangiopathy.

- Diagnosis is difficult and often delayed, with high mortality.

- Improved survival is reported with some cancer therapies so early recognition Is imperative

Pulmonary artery angiosarcoma

Sarcomas are rare tumours of mesenchymal origin that originate in bones or in soft tissues. Of these, angiosarcomas arise from endothelial cells and comprise 2% of all sarcomas [68]. Pulmonary artery angiosarcomas typically arise from the pulmonary trunk [69] and can metastasise to the lung and mediastinal lymph nodes, as well as through haematogenous dissemination. They are important albeit rare mimics of pulmonary thromboembolic disease.

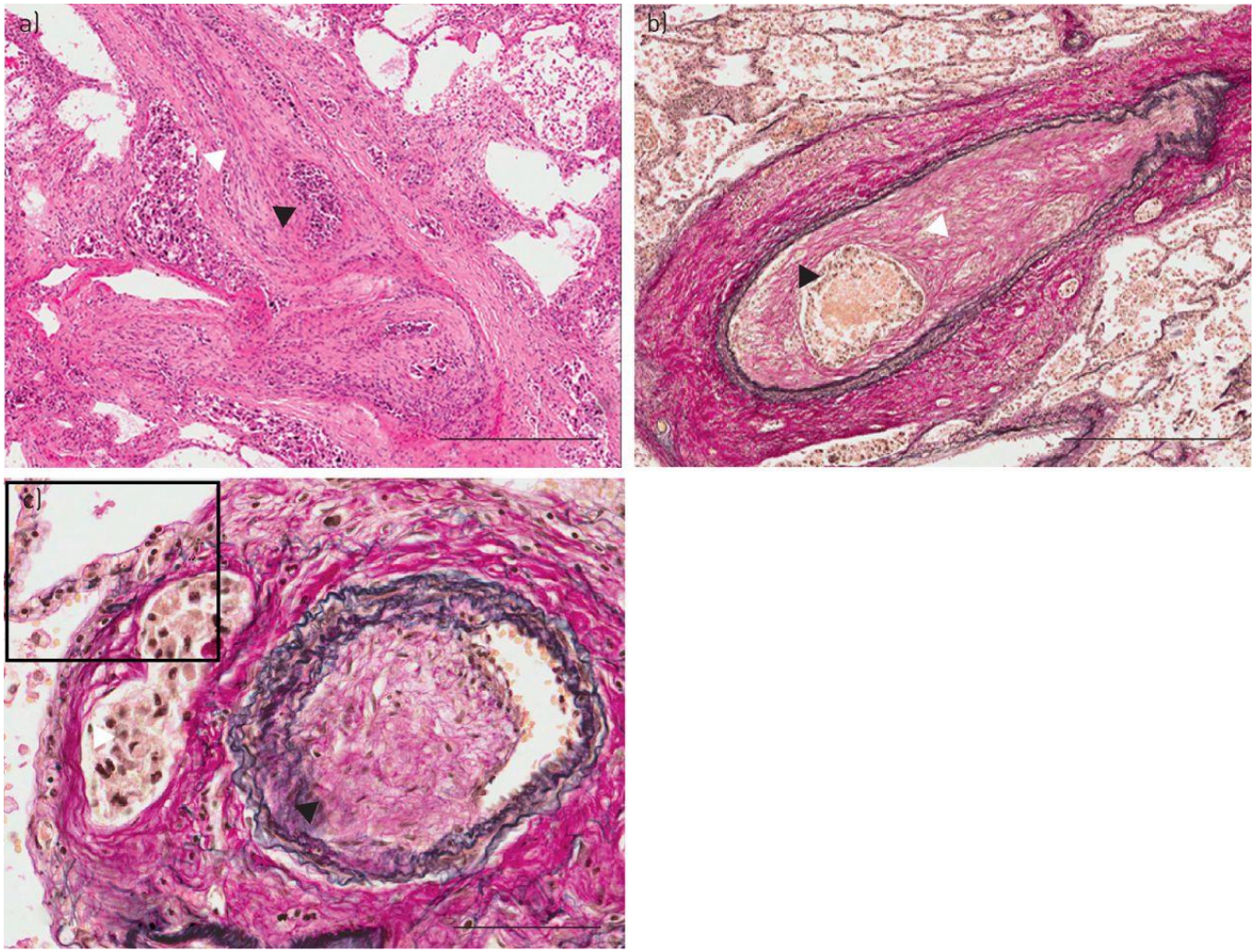
Recognised associations with angiosarcoma include exposure to radiotherapy, genetic mutations including BRCA, neurofibromatosis type 1 and Kippel Trelawney syndrome, and patients with immunosuppression including HIV [68].

The clinical presentation of pulmonary artery angiosarcomas is nonspecific. The most common symptoms are dyspnoea, pleuritic chest pain, cough and haemoptysis [70].

CT findings include low attenuation filling defects, enhancement of the mass in the lumen of the artery and extravascular spread of the lesion [71]. Gadolinium-enhanced magnetic resonance imaging may help differentiate between thrombotic masses and vascular tumours: unlike thromboemboli, angiosarcomas exhibit heterogeneous enhancement with gadolinium [69]. The use of FDG-PET/CT can help differentiate malignant from benign pulmonary artery lesions [71].

Surgical treatment is preferred for patients with primary pulmonary angiosarcoma, and is only an option in the 50% presenting without metastatic disease [4]. Radical resection offers a median survival of 37±20 months, compared to 11±3 months following subtotal resection or debulking [72]. Without surgery, the mean survival is 1.5 months. Chemotherapy may improve survival, but evidence remains limited [72]. Inhibitors of angiogenesis may be more promising future therapies [73].

Lung histology from a case of pulmonary tumour thrombotic microangiopathy related to severe pulmonary hypertension. a) Post mortem section showing occlusion of a medium-sized pulmonary arterial lumen by fibrointimal proliferation of fibroblasts and collagen...



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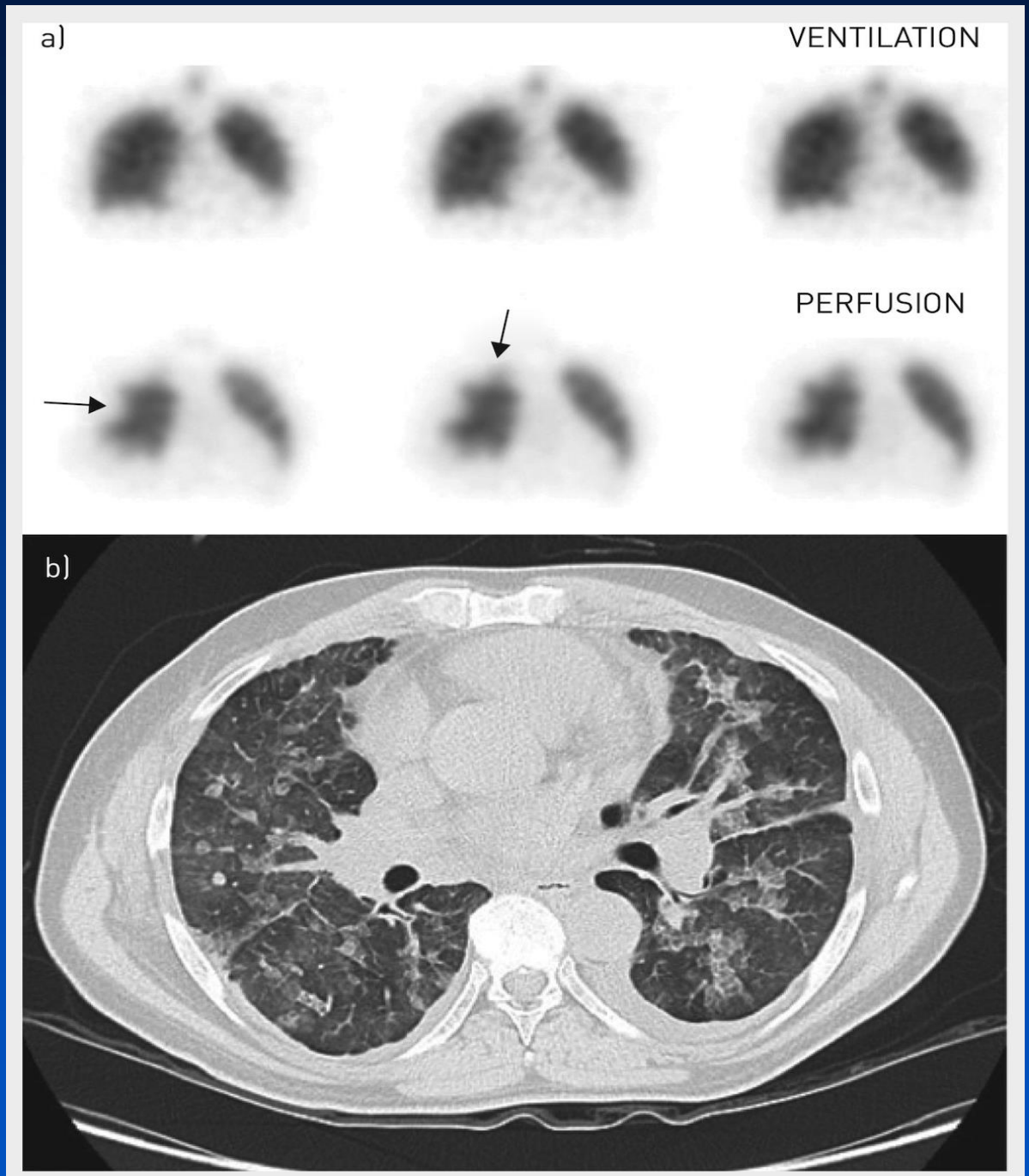
a)
VENTILATION
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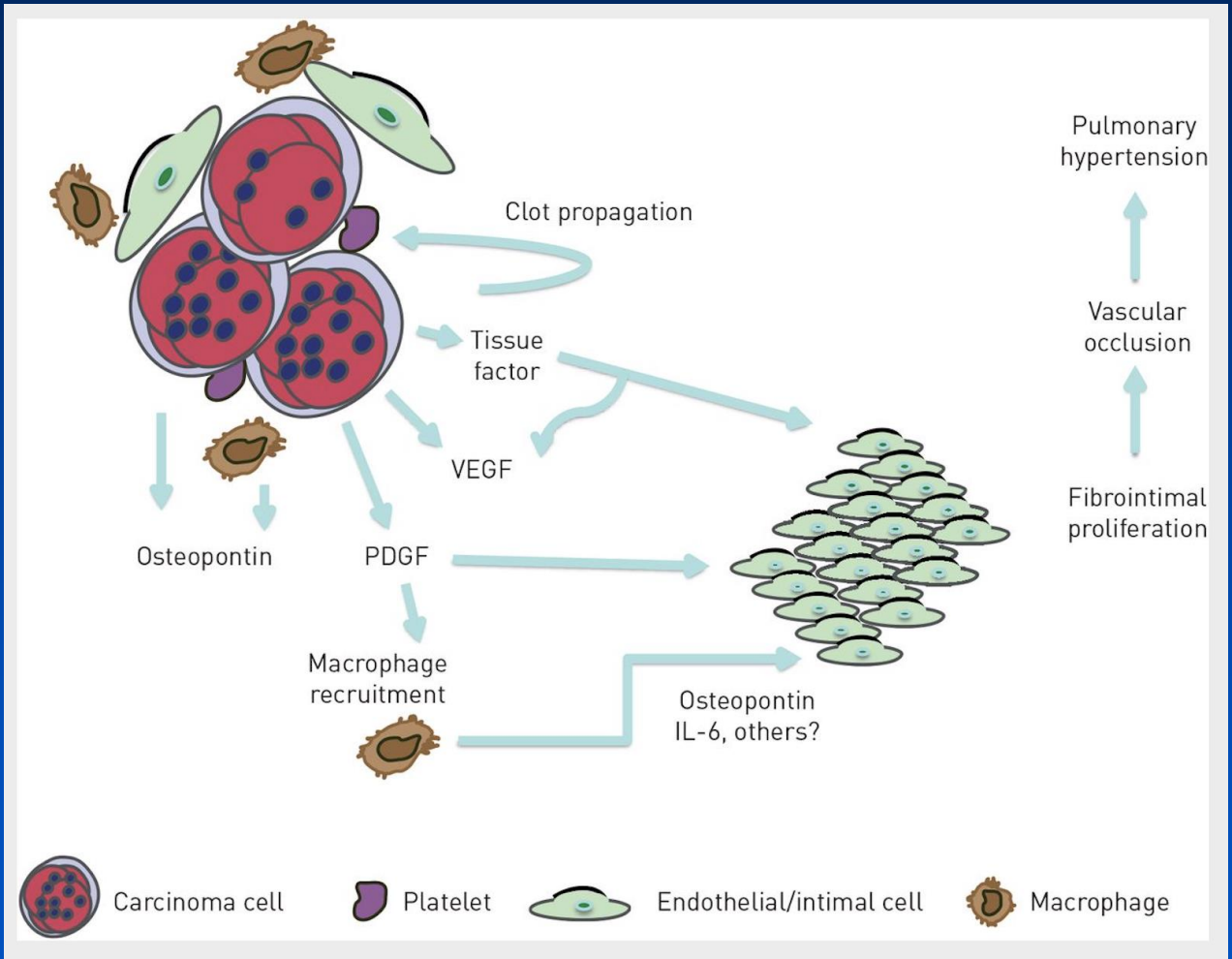
b)

Proposed mechanisms for fibrointimal proliferation in pulmonary tumour thrombotic microangiopathy



CMAJ PRACTICE CME

CASES

Primary pulmonary artery angiosarcoma mimicking pulmonary embolism in a 66-year-old man with dyspnea

Philippe Desmarais MD, Mikhael Laskine MD MSc, Christine Caporuscio MD

A 66-year-old man consulted his family physician because of progressive dyspnea. He had a history of hypertension and dyslipidemia, but no personal or family history of cardiac or respiratory diseases. He was a lifelong nonsmoker and had been a professional endurance swimmer. After his professional career, he started a swimming school, worked as a trainer and continued to swim on a regular basis. Over the last three years, he noticed a progressive decrease in his endurance. One month before consultation, he experienced a substantial increase in dyspnea and new onset of pleuritic pain localized over the left side of his chest.

The patient was referred to a cardiologist. Results of dipyridamole sestamibi scanning and transthoracic echocardiography were normal. Coronary computed tomography (CT) angiography showed multiple pulmonary emboli, including phology and adequate left and right heart function. The systolic pressure of the pulmonary artery was elevated at 47 mm Hg.

We diagnosed unprovoked multiple pulmonary emboli with mild pulmonary hypertension. The patient was discharged with a prescription for warfarin, a follow-up visit and referral for an outpatient colonoscopy to investigate the anemia.

The patient's symptoms were not improved one month later. Repeat CT angiography showed an unchanged obstructive lesion in the left pulmonary artery despite the anticoagulation treatment (Figure 2). The lesion measured 5.3 × 2.6 cm and was heterogeneously hypodense with intrinsic contrast enhancement. The filling defects in the left lobar and lingular arteries had resolved. The left pleural effusion had markedly increased, and there was now mild dilatation of the right ventricle. These findings suggested a possible malignant process

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The authors have obtained patient consent.

Correspondence to:
Philippe Desmarais, philippe.desmarais.1@umontreal.ca

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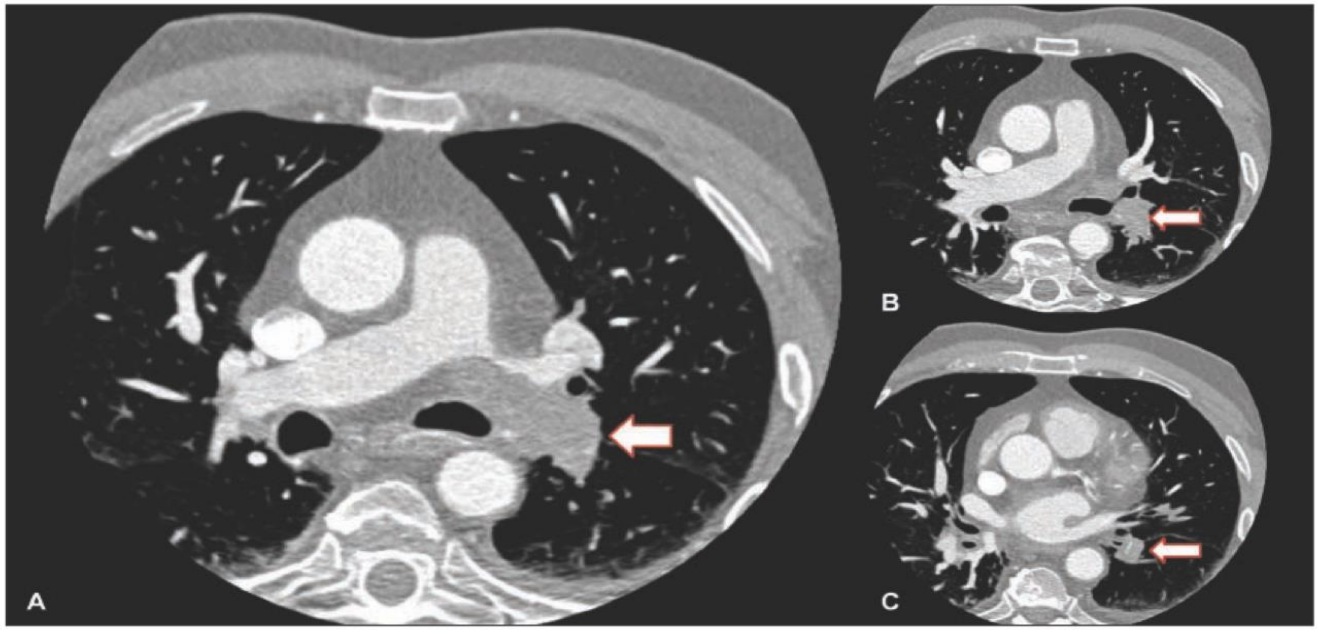

Figure 1: Coronary computed tomography angiograms in a 66-year-old man, showing multiple filling defects (arrows) in the left arterial pulmonary circulation, with a main lesion at the level of the left pulmonary artery.

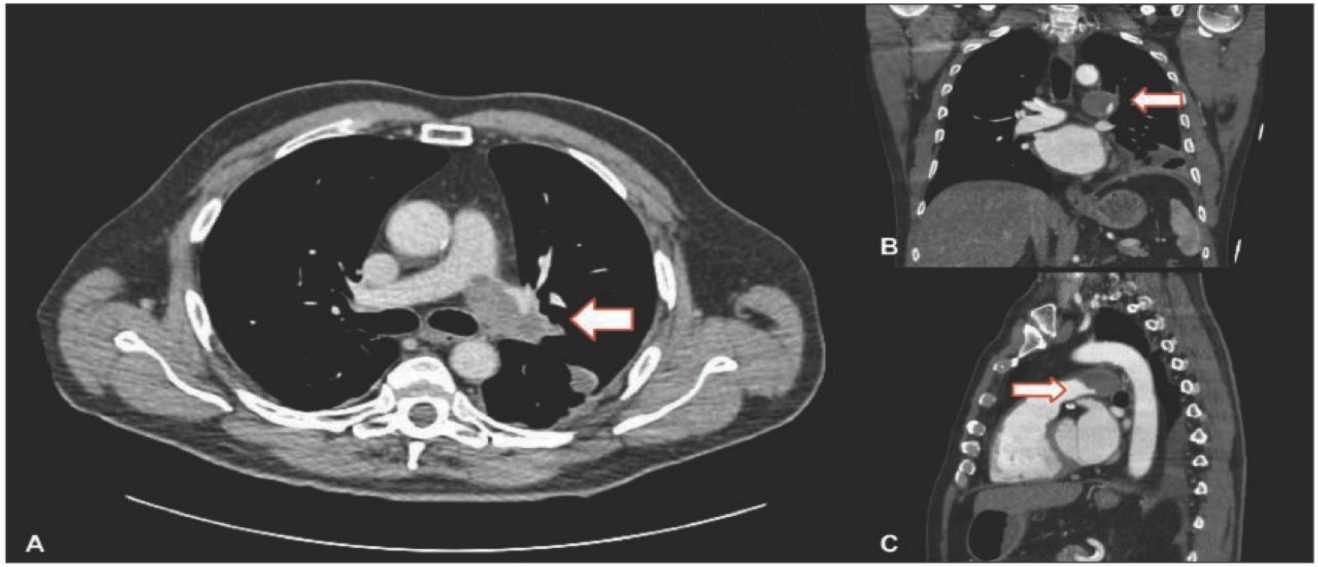

Figure 2: Computed tomography angiograms of the patient's thorax after four weeks of warfarin treatment. A large obstructive lesion (arrows) is visible in the left pulmonary artery, extending into the pulmonary trunk. The lumen of the left pulmonary artery is almost completely obliterated by the lesion.

Clinical presentation

The clinical presentation of pulmonary artery angiosarcomas is nonspecific. Most common symptoms are dyspnea, pleuritic chest pain, cough and hemoptysis.[6,8] Patients may have signs of chronic pulmonary hypertension. Because the radiologic findings resemble those of subacute pulmonary embolism, angiosarcomas are often misdiagnosed as pulmonary embolism.[6,8]

Treatment and Prognosis

Radical surgical resection offers the best prognosis, with a median survival of 36.5 ± 20.2 months.[10] Subtotal resection or debulking of the tumour offers a median survival of 11 ± 3 months.[10] Unfortunately, many patients are not surgical candidates because as many as 50% present with metastatic disease.[4] Without surgery, the mean survival is 1.5 months.[8] Chemotherapy may prolong the median survival time, but supporting evidence is limited.[4,8,10]

Take-home Message

- Primary pulmonary artery angiosarcoma is an important albeit rare mimics of CTEPH
- Most common symptoms are dyspnea, cough, pleuritic pain and hemoptysis.
- Accurate Image diagnosis is utmost important and MRI may be helpful for differetial diagnosis, multidisciplinary approach at PH expert center is best strategy for caring CTEPH patients.
- Early surgical treatment is the only cure for primary pulmonary artery angiosarcoma but balloon pulmonary angioplasty (BPA) may be a palliative strategy for improving pulmonary hypertension symptom.

Thanks for your attention