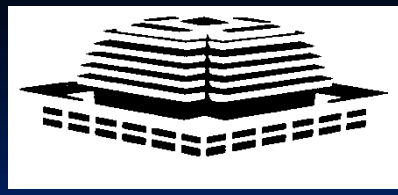


ONASSIS
CARDIAC
SURGERY
CENTER

CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

Case presentation

- The most frequent and characteristic complication of BPA is pulmonary injury.
- Pulmonary vessel injury caused by the guidewire, guiding catheter, balloon dilation, or contrast medium injection at high pressure may play a role in inducing pulmonary injury in BPA.
- It is necessary to determine how to dilate the lesion to achieve maximal therapeutic efficacy and reduce the risk of pulmonary vessel injury, which could potentially become lethal. To dilate the lesion, balloon size, vessel size, the number of organised thrombi and patient haemodynamics must be considered. Balloon size is the only one of these factors we can control.

- Dilatation using a larger balloon appears to decrease residual stenosis at the lesion site; however, it also increases the probability of damage to the vessel wall in case of overdilatation, which may result in an oozing haemorrhage.
- Considering these circumstances, we dilate the lesion sequentially in two sessions. A smaller balloon relative to the actual vessel size is selected for the initial BPA session to reduce the risk of pulmonary vessel injury and restore minimal blood flow to the occluded or stenotic pulmonary vessels. At the second session, a balloon catheter of 100-120% the size of the reference vessel diameter indicated on the angiogram is selected to optimise the dilatation of the lesion

Case Report

- Female 60 years old.
- P.H. due to chronic Thromboembolic disease, firstly diagnosed in January 2007.

Symptoms

Dyspnea (**WHO III**), exercise intolerance, chest pain.

Desaturation during effort < 70% (!)

History

1995: **Myelodysplastic syndrome** with thrombocytosis,
treated with hydroxyurea & clopidogrel until Aug 2007.
2007: **Acute myeloid leukemia**.
Hickman catheter implantation for chemotherapy (**anthracyclines**).
2004: **Splenectomy**.
2008: **Vein thrombosis**. Chronic anticoagulation therapy.
Thrombus in the right atrium.
Hickman catheter removal.
Two hospitalizations for suspected **Pulmonary Embolism**

- **Physical examination**

  Accentuated pulmonary component of S2,
  a right ventricular S3. Murmurs over lung fields.
- **Laboratory Tests**

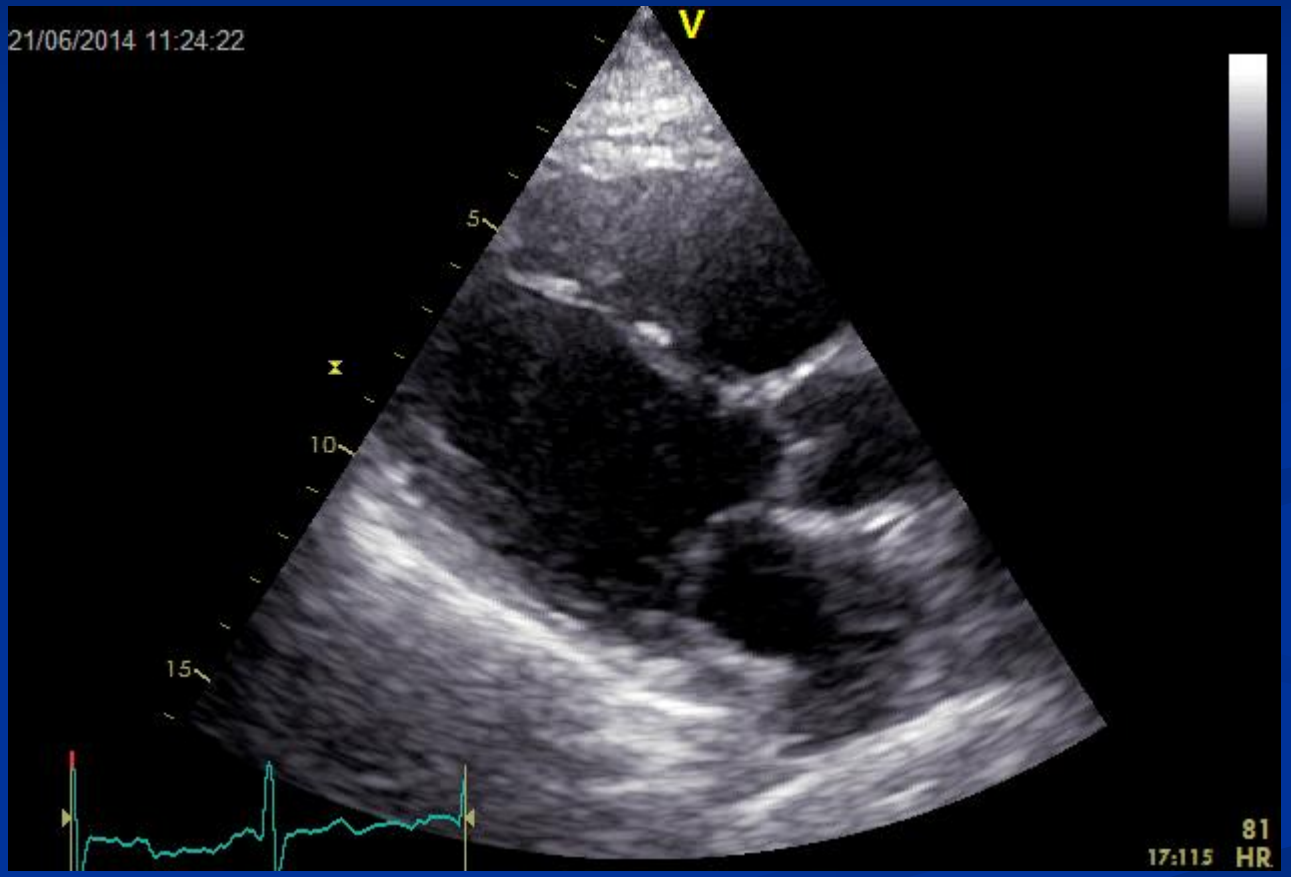
  NT-proBNP: 3221 pg/ml, uric acid 7,4 mg/ dl
  HIV negative, PLT: 800.000
- **Pulmonary function test**

  Normal.
- **Venous Ultrasonography**

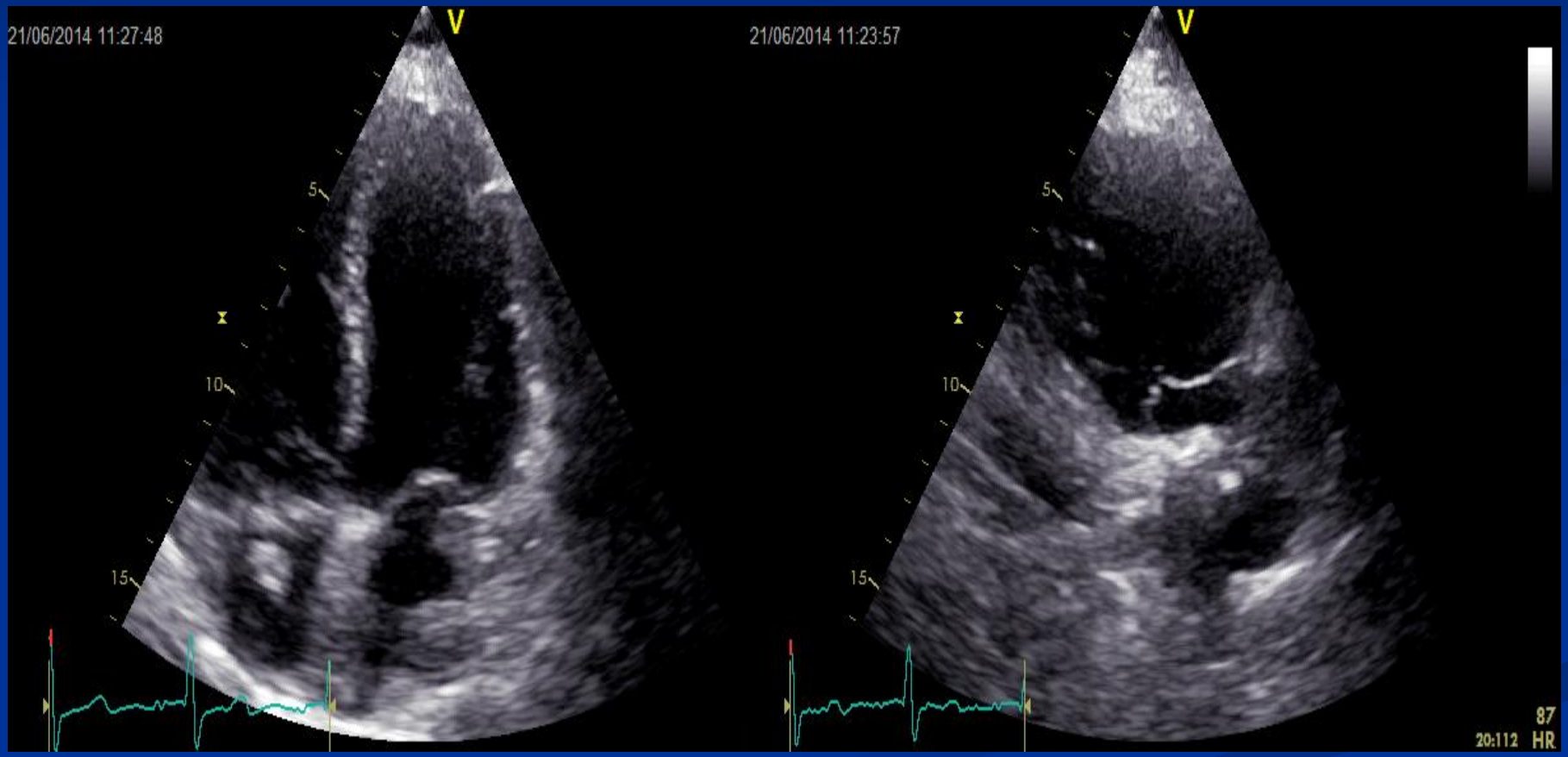
  without evidence of DVT.
- **6MWT**: 212m, reduction of SaO2 from 89% to 69%.
- **Echocardiogram**

  RV dilatation, RVEF & LVEF reduction,
  RVSP: 55mmHg,
  Right atrial thrombus (3,0 x 2,0 cm).

Echo (1)

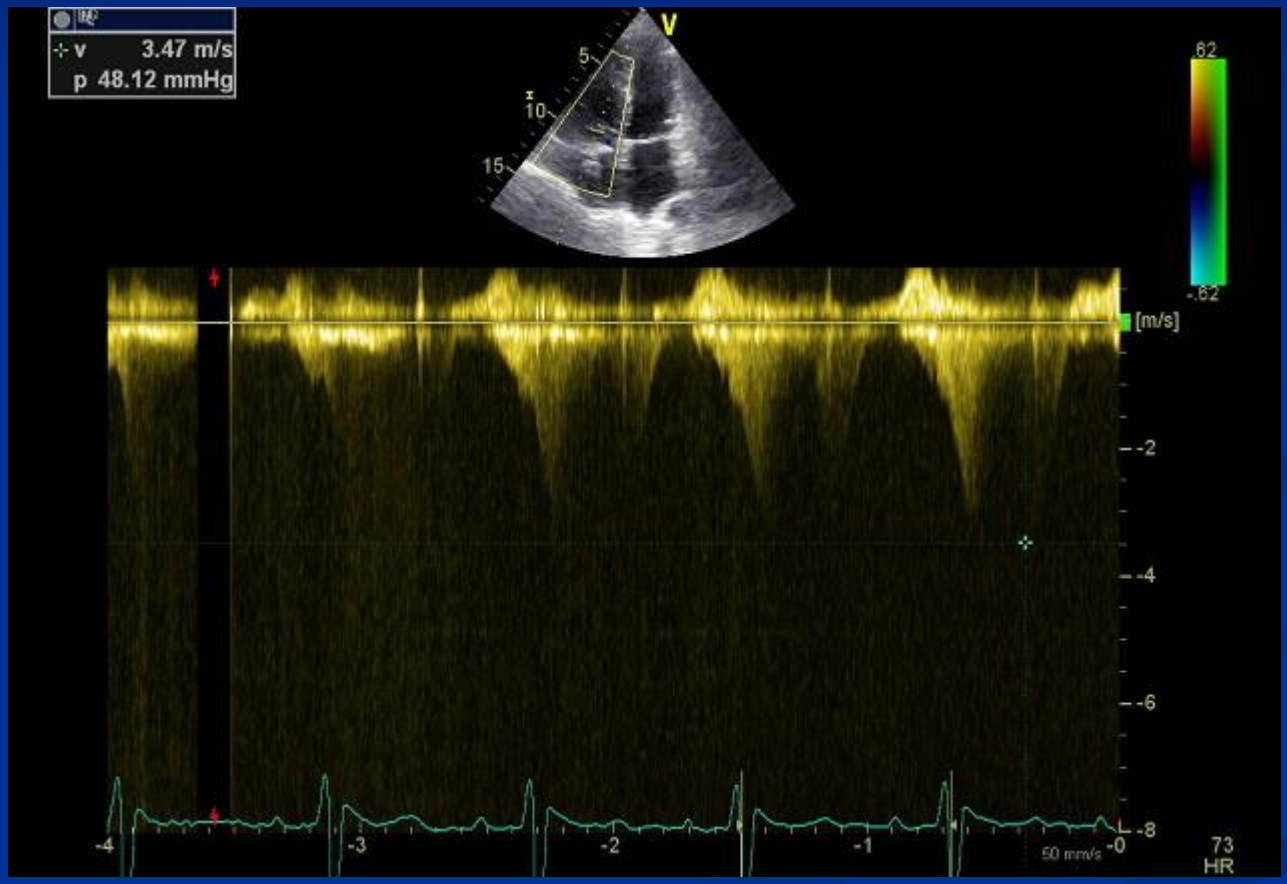


Echo (2)



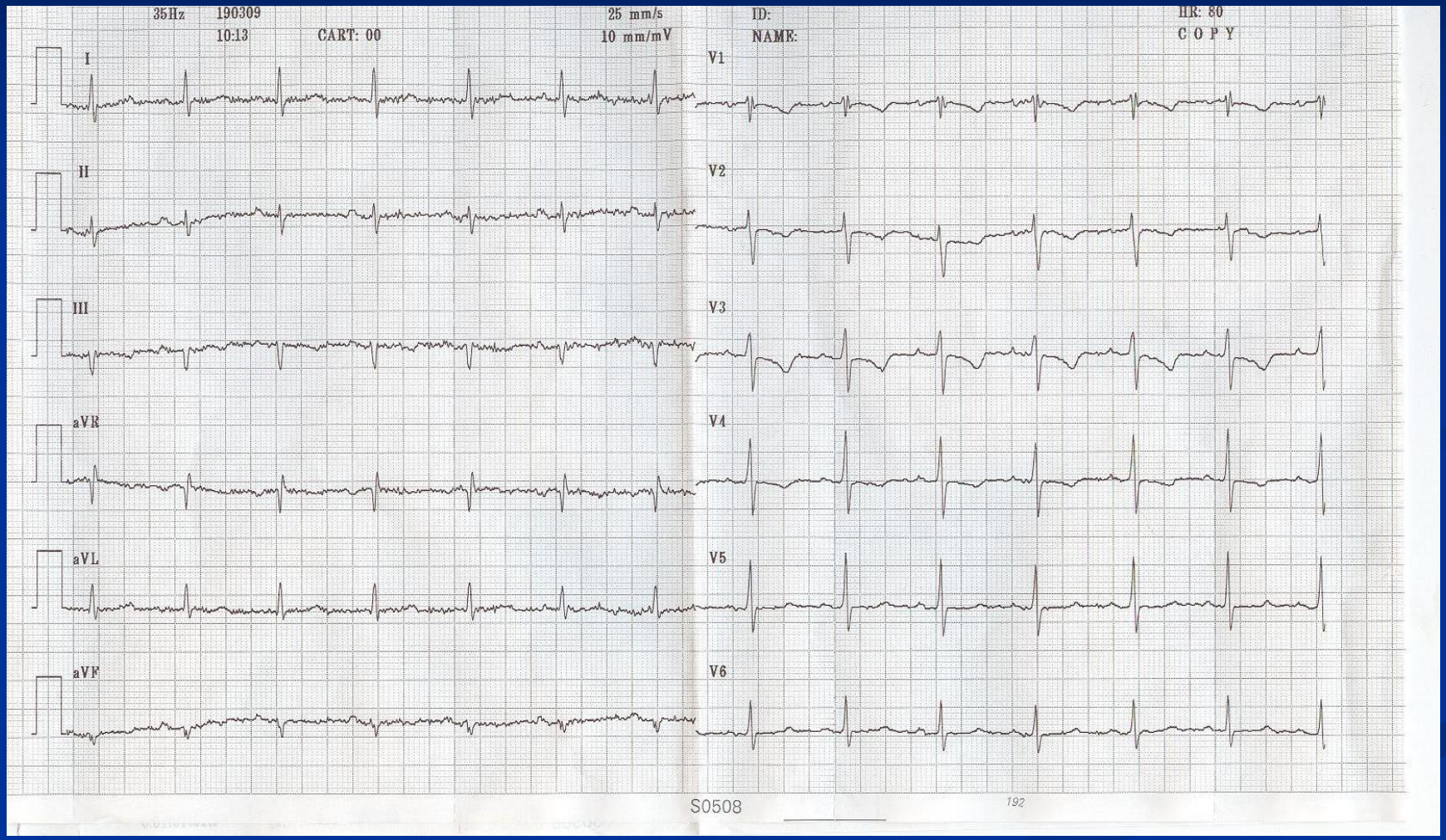
Calcified thrombus inside RA

Echo (3)

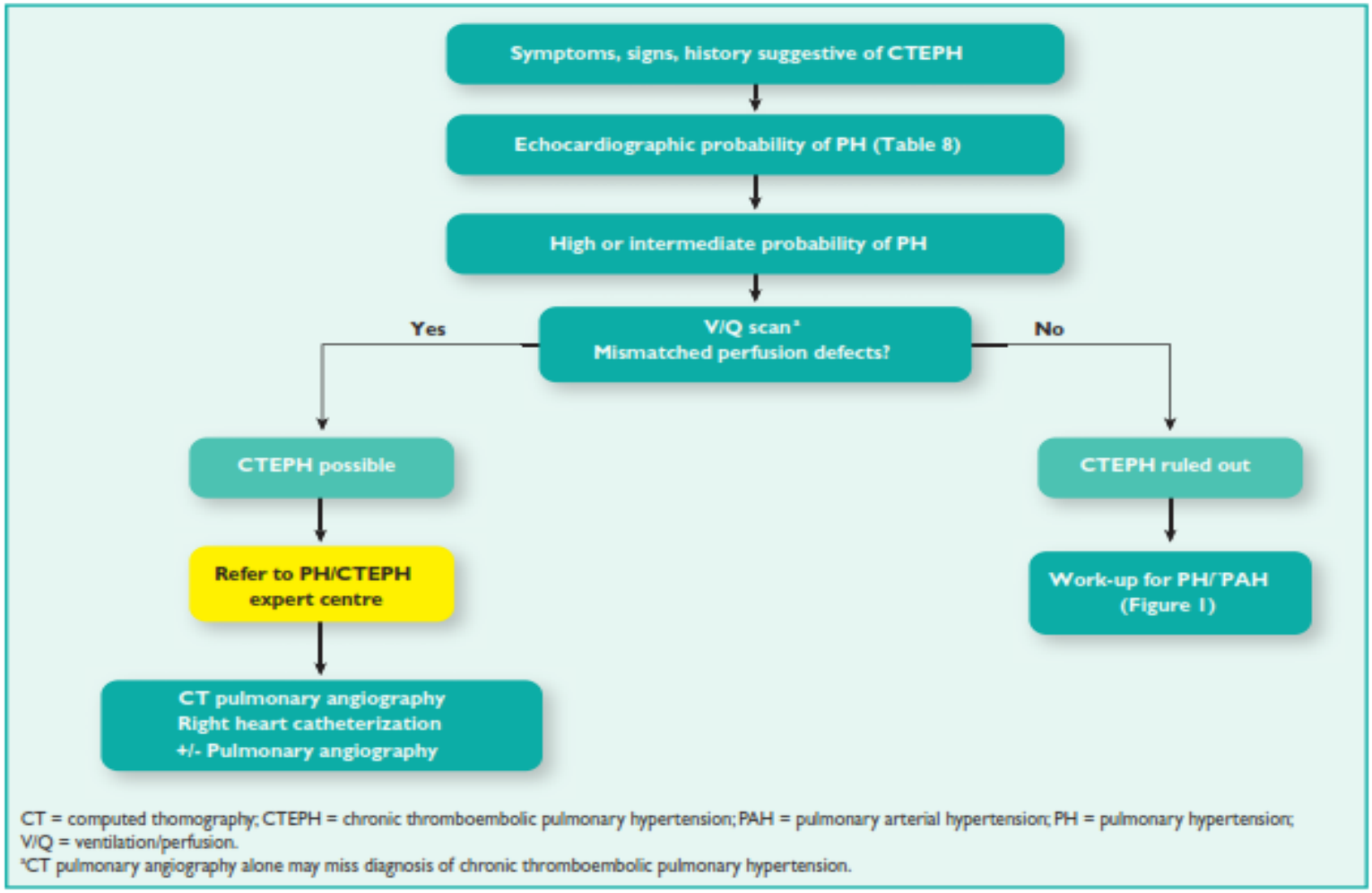


RVSP ~ 55 mmHg

ECG


35Hz
190309
10:13
CART: 00
25 mm/s
10 mm/mV
ID:
NAME:
HR: 80
C O P Y
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
S0508
192

Diagnostic Algorithm for CTPH



CT = computed thomography; CTEPH = chronic thromboembolic pulmonary hypertension; PAH = pulmonary arterial hypertension; PH = pulmonary hypertension; V/Q = ventilation/perfusion.
[a]CT pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.

- **Right Heart Catheterization**

  PAP 58/23/37 mmHg, RV 58/7 mmHg,

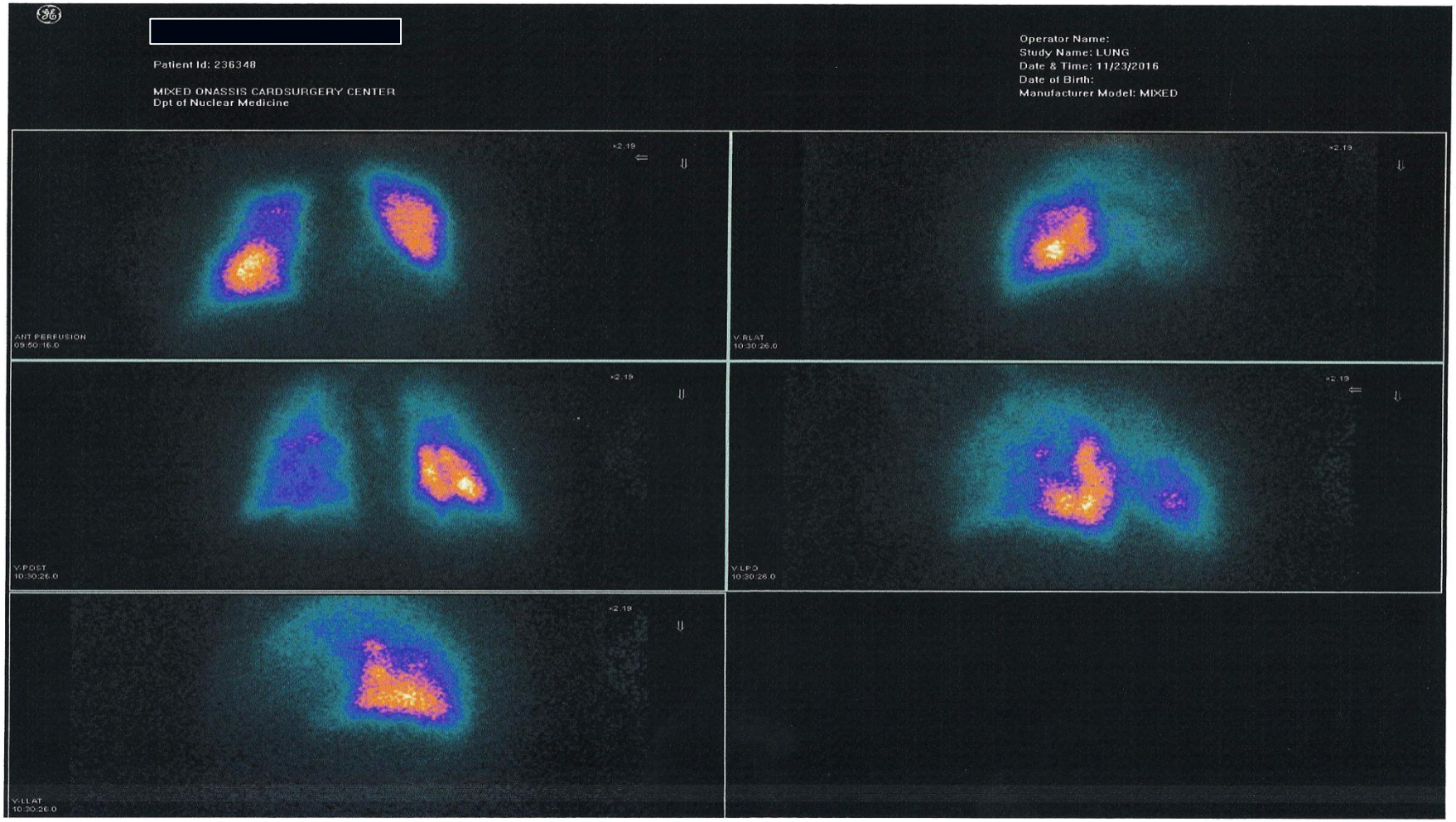
  RAPmean 14 mmHg, **PCWP 14 mmHg**,

  C.O. 3.2 l/min, **C.I. 1.76 l/min/m2**,

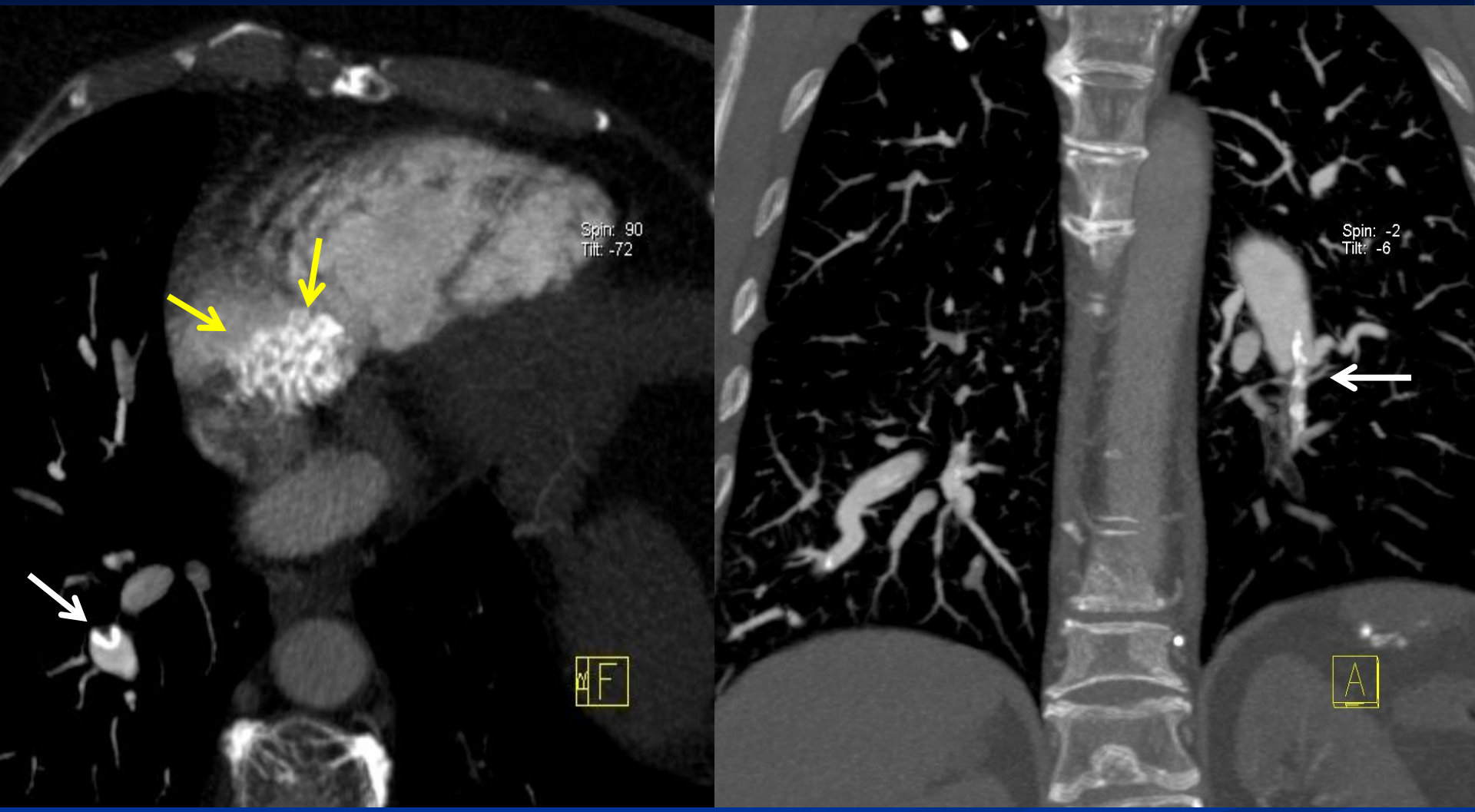
  PVR 9.5 WU AO sat 87%, PA sat 58%

- **V/Q lung scanning**.
- **Chest CT**.
- **Biplane Pulmonary Angiography**.

V/Q lung Scan

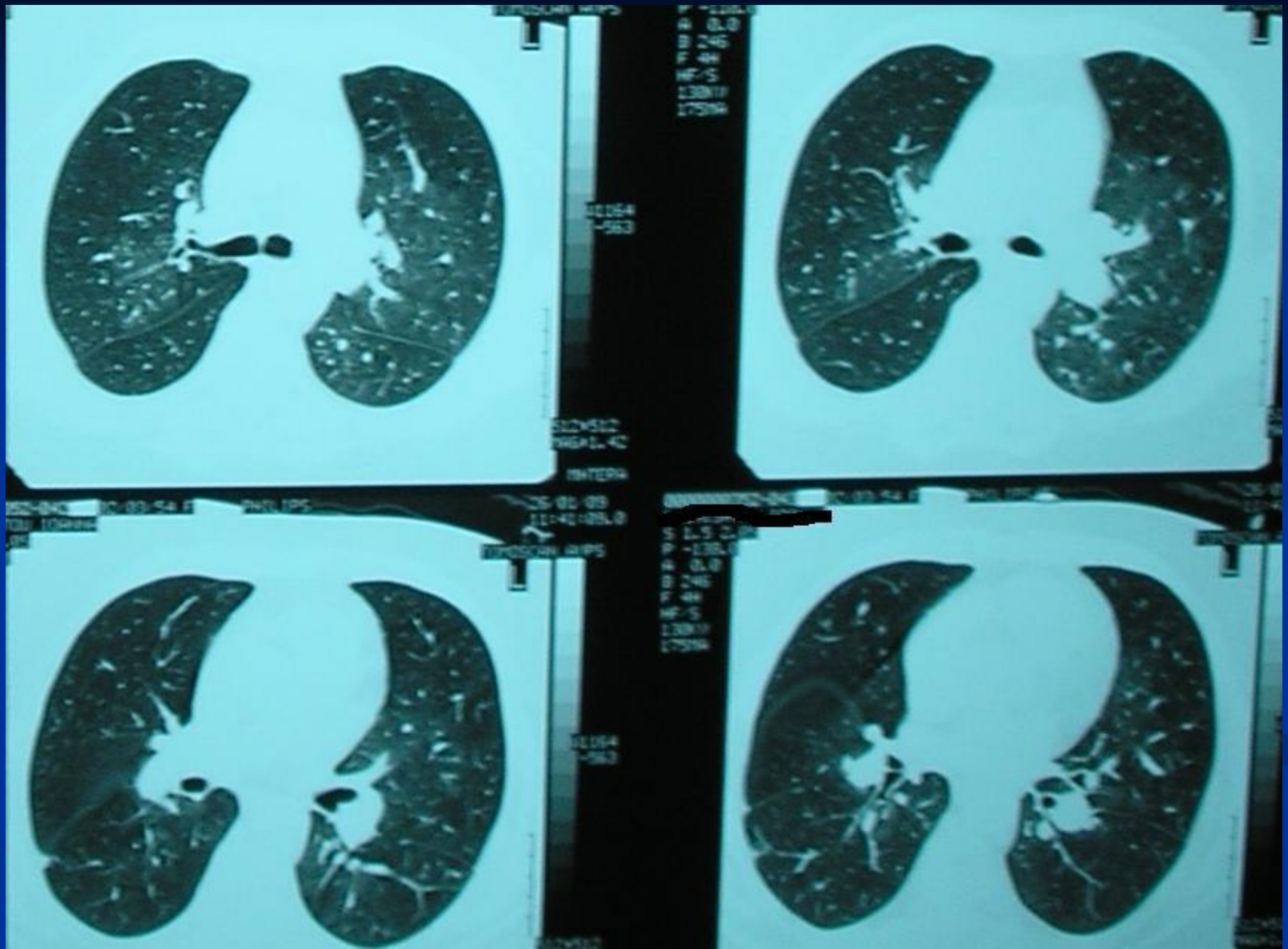

[redacted]
Patient Id: 236348
MIXED ONASSIS CARDSURGERY CENTER
Dpt of Nuclear Medicine
Operator Name:
Study Name: LUNG
Date & Time: 11/23/2016
Date of Birth:
Manufacturer Model: MIXED
ANT PERFUSION
09:50:16.0
V: RLAT
10:30:26.0
V: POST
10:30:26.0
V: LPO
10:30:26.0
V: LLAT
10:30:26.0

LUNG CT



Yellow arrows: calcified thrombus inside RA
White arrows: calcified thrombi inside PA

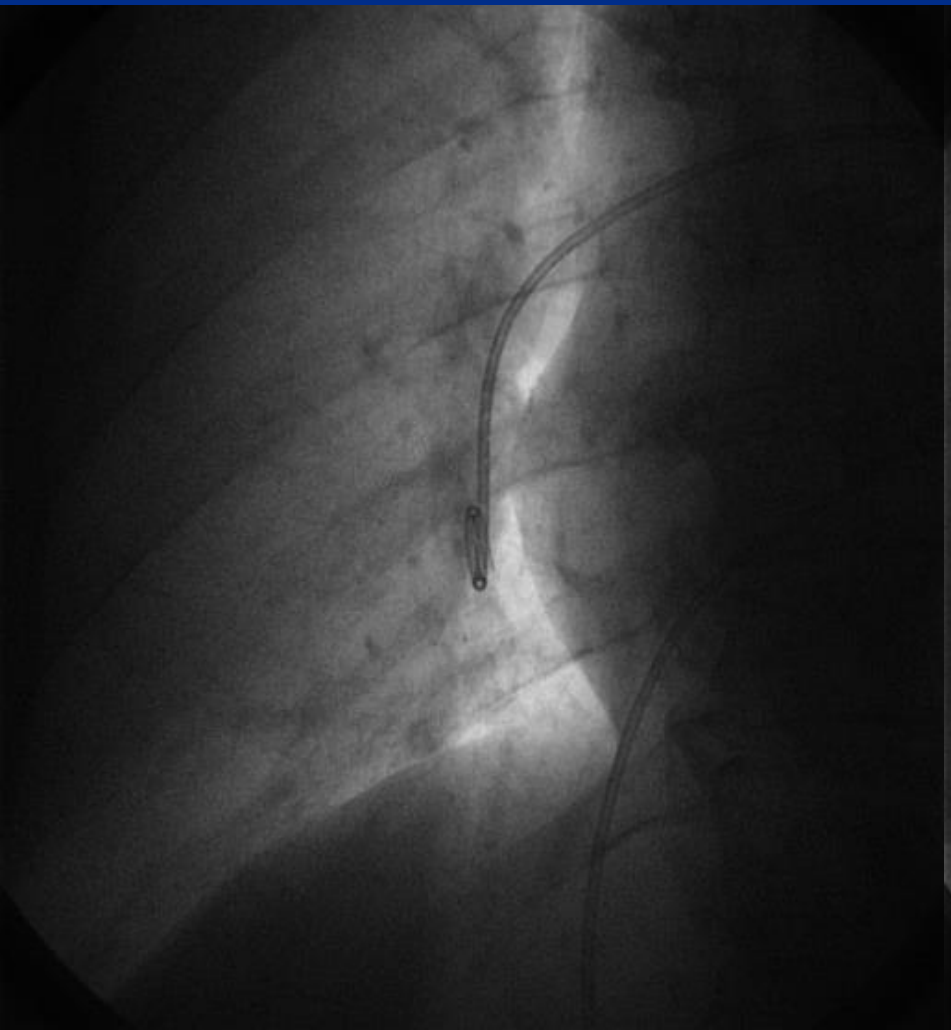
LUNG CT (Mosaic appearance)

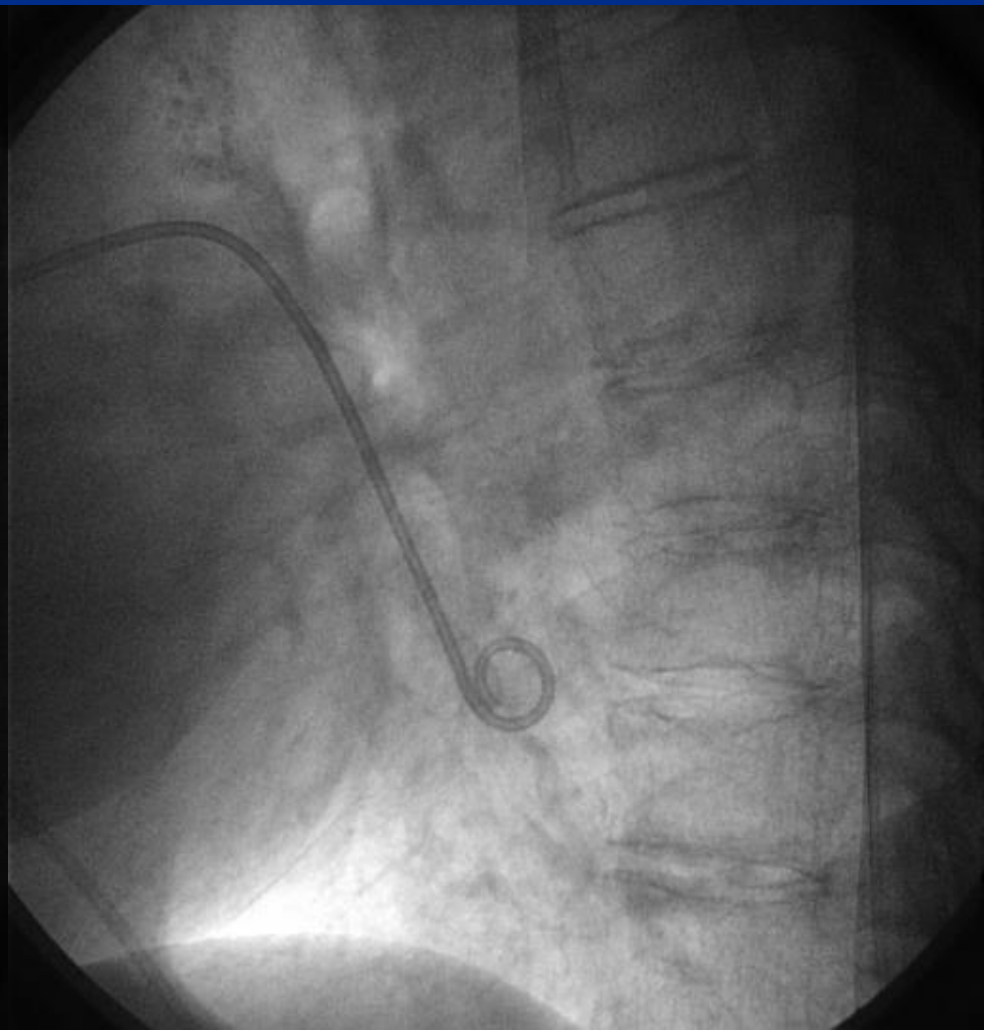


PULMONARY ANGIOGRAPHY
RPA

AP VIEW

LAO

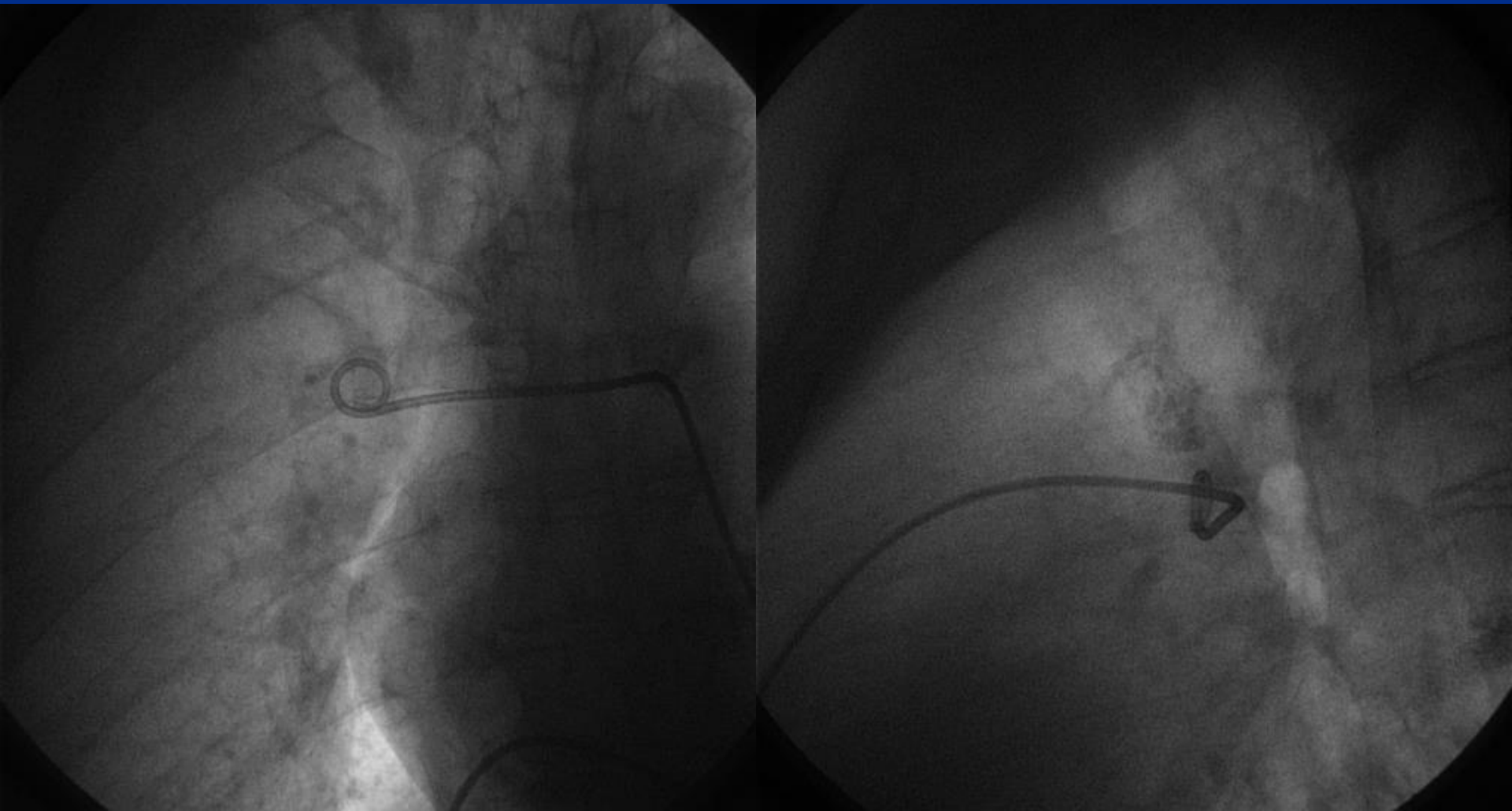




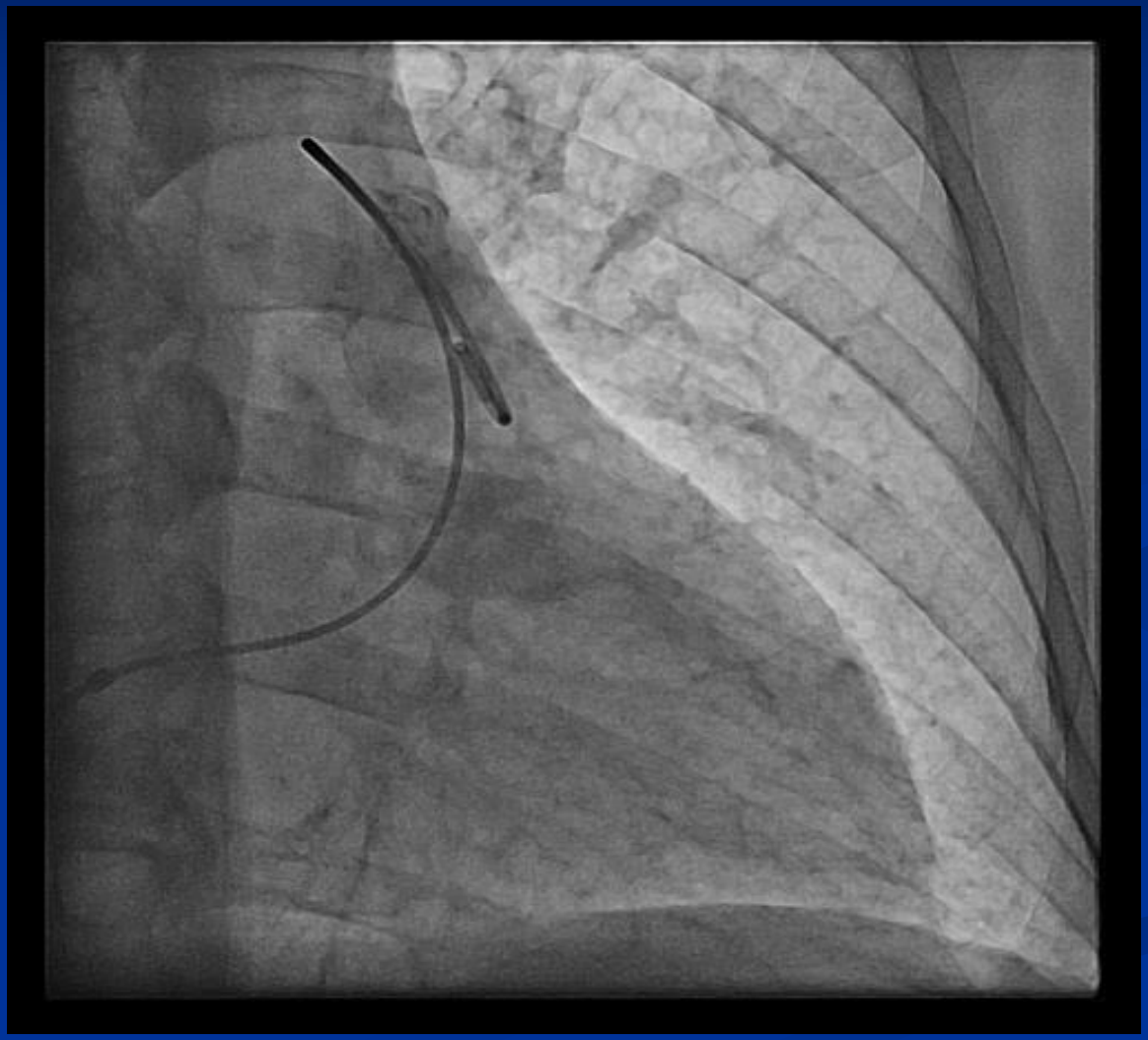
PULMONARY ANGIOGRAPHY
RPA (Upper lobe)

AP

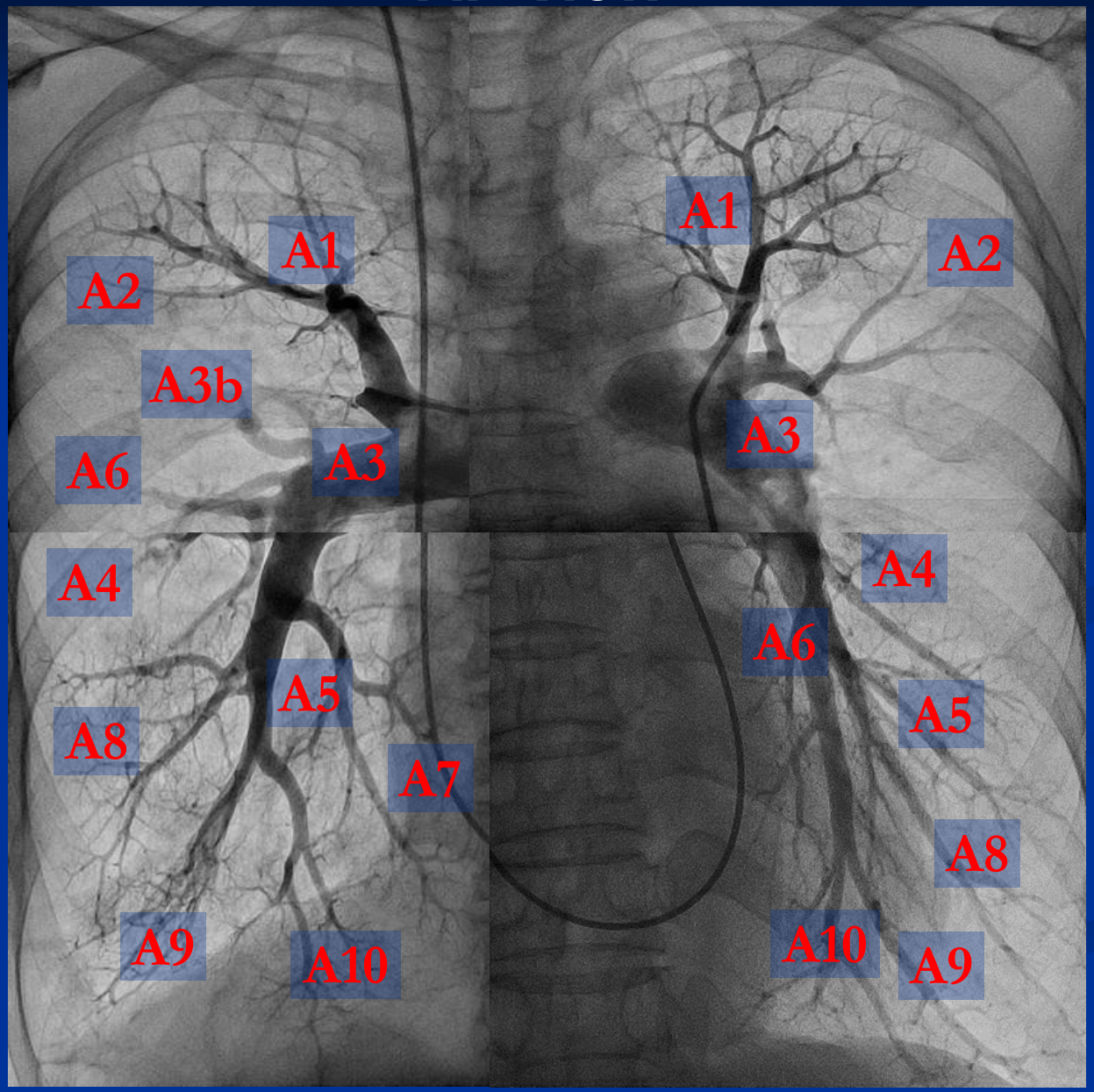
LAO



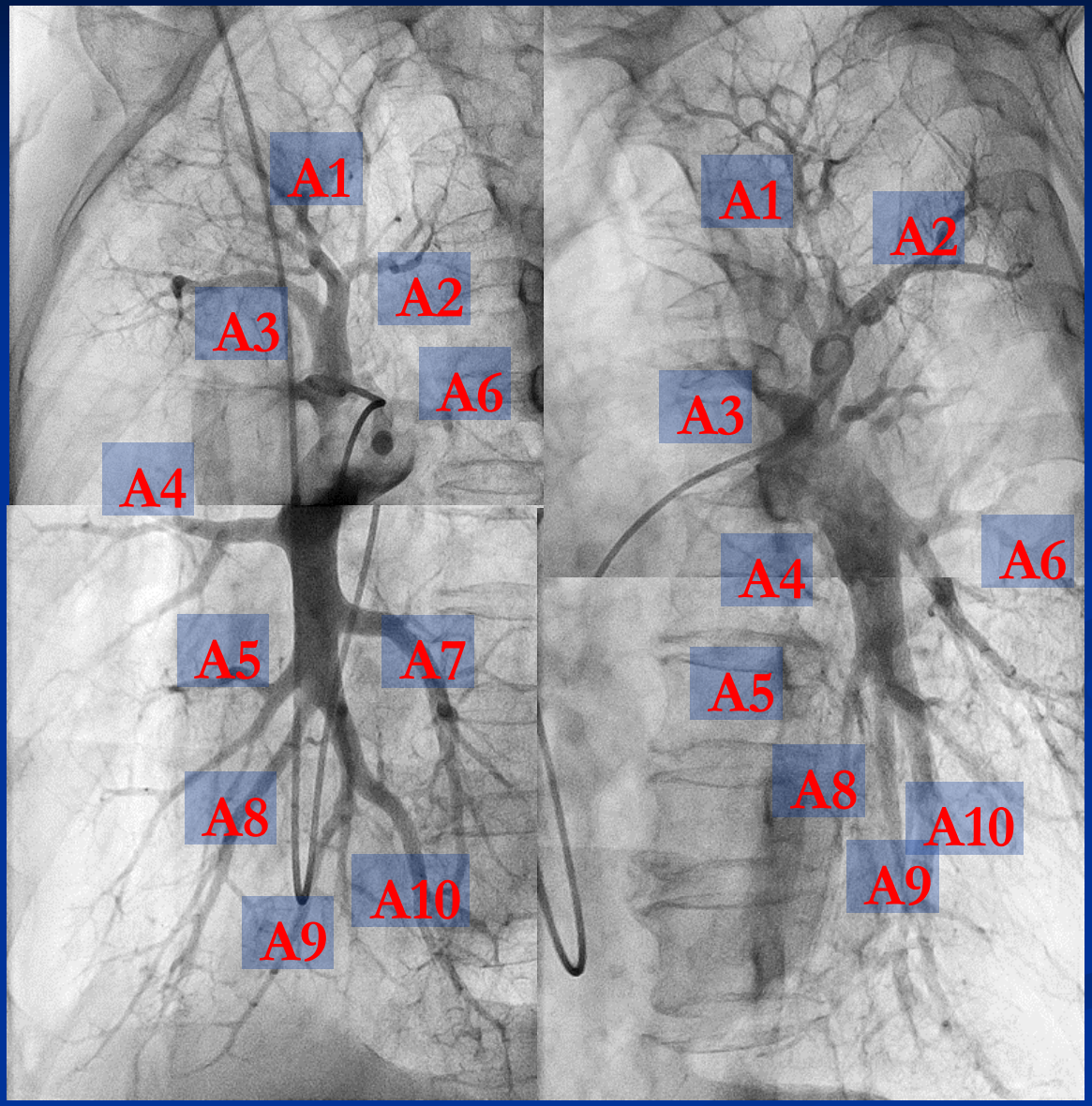
PULMONARY ANGIOGRAPHY
LPA



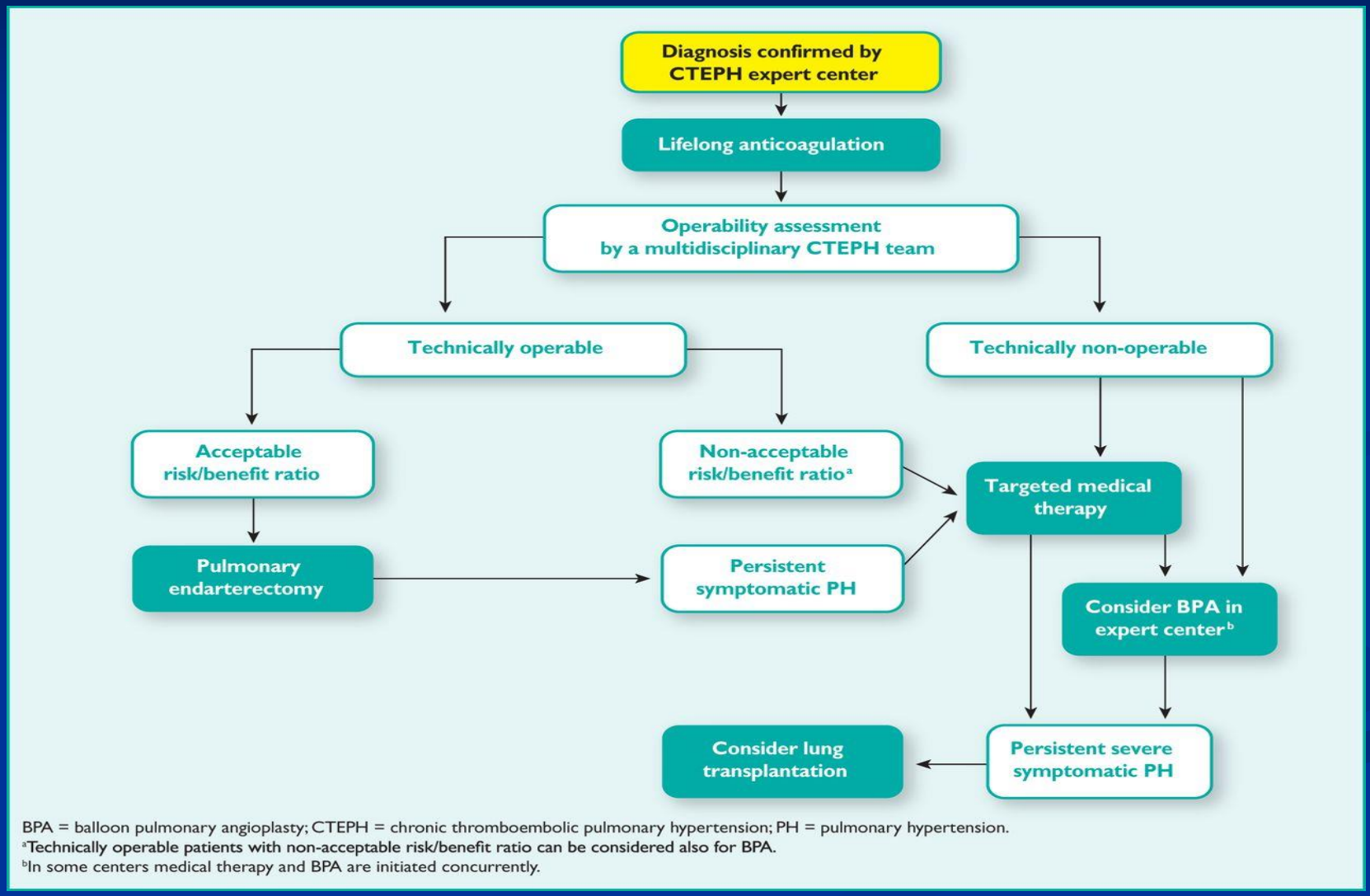
Pulmonary Arteries Anatomy
AP view



Pulmonary Arteries Anatomy
LAO 60^0 view



Treatment algorithm for chronic thromboembolic pulmonary hypertension. 2015 ESC/ERS Guidelines



BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic pulmonary hypertension; PH = pulmonary hypertension.
[a]Technically operable patients with non-acceptable risk/benefit ratio can be considered also for BPA.
[b]In some centers medical therapy and BPA are initiated concurrently.

Table 34

Recommendations for chronic thromboembolic pulmonary hypertension

Recommendations	Class[a]	Level[b]	Ref.[c]
In PE survivors with exercise dyspnoea, CTEPH should be considered	IIa	C	449
Life-long anticoagulation is recommended in all patients with CTEPH	I	C	91
It is recommended that in all patients with CTEPH the assessment of operability and decisions regarding other treatment strategies should be made by a multidisciplinary team of experts	I	C	91
Surgical PEA in deep hypothermia circulatory arrest is recommended for patients with CTEPH	I	C	91
Riociguat is recommended in symptomatic patients who have been classified as having persistent/recurrent CTEPH after surgical treatment or inoperable CTEPH by a CTEPH team including at least one experienced PEA surgeon	I	B	441
Off-label use of drugs approved for PAH may be considered in symptomatic patients who have been classified as having inoperable CTEPH by a CTEPH team including at least one experienced PEA surgeon	IIb	B	437–440
Interventional BPA may be considered in patients who are technically non-operable or carry an unfavourable risk:benefit ratio for PEA	IIb	C	57,444–446,448
Screening for CTEPH in asymptomatic survivors of PE is currently not recommended	III	C	417

BPA = balloon pulmonary angioplasty; CTEPH = chronic thromboembolic pulmonary hypertension; PAH = pulmonary arterial hypertension; PE = pulmonary embolism; PEA = pulmonary endarterectomy.

- **TREATMENT**

Dual *antiplatelet & anticoagulation* therapy, phospholipase A2 inhibitor (*anagrelide*),

Bosentan therapy as a bridge to pulmonary endarterectomy (PEA). (Riociguat not available at the time of diagnosis)

Referred to PEA expert center (Kerckhoff Klinik, Bad Nauheim, Germany): rejected twice due to comorbidities (mainly hematological disease)

Case Report

- **CTEPH**. (Chronic Thromboembolic Pulmonary Hypertension)
- Thrombocytosis, MDS (in remission).
- Right Heart Thrombus.
- Rejected for PEA
- Consider BPA

C.T.E.P.H.

- Chronic ThromboEmbolic Pulmonary Hypertension is one of the important clinical variants of P.H., and
- it ***is the only curable form of the disease***.

Book Reviews: Pulmonary Hypertension.
N Engl J Med 2008, 359:20.

Balloon Pulmonary Angioplasty (BPA)-Indications

- Unsuitable cases for PEA (surgically inaccessible lesions, surgically accessible but inoperable because of comorbidities, and cases of residual or recurrent pulmonary hypertension after PEA)

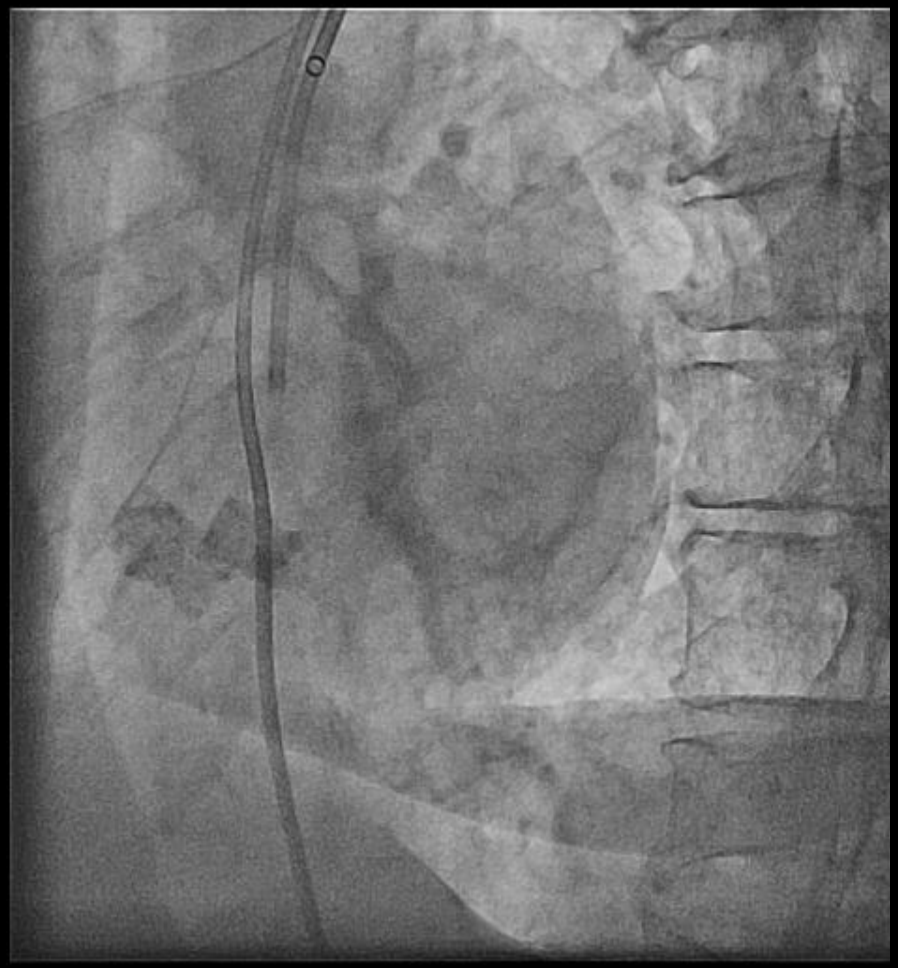
- Cases in which conventional therapy is insufficient (WHO functional class ≥II after conventional therapy, mean PAP ≥30 mmHg, or PVR ≥300 dyne · s · cm^{-5})

Balloon Pulmonary Angioplasty (BPA)- Contraindications

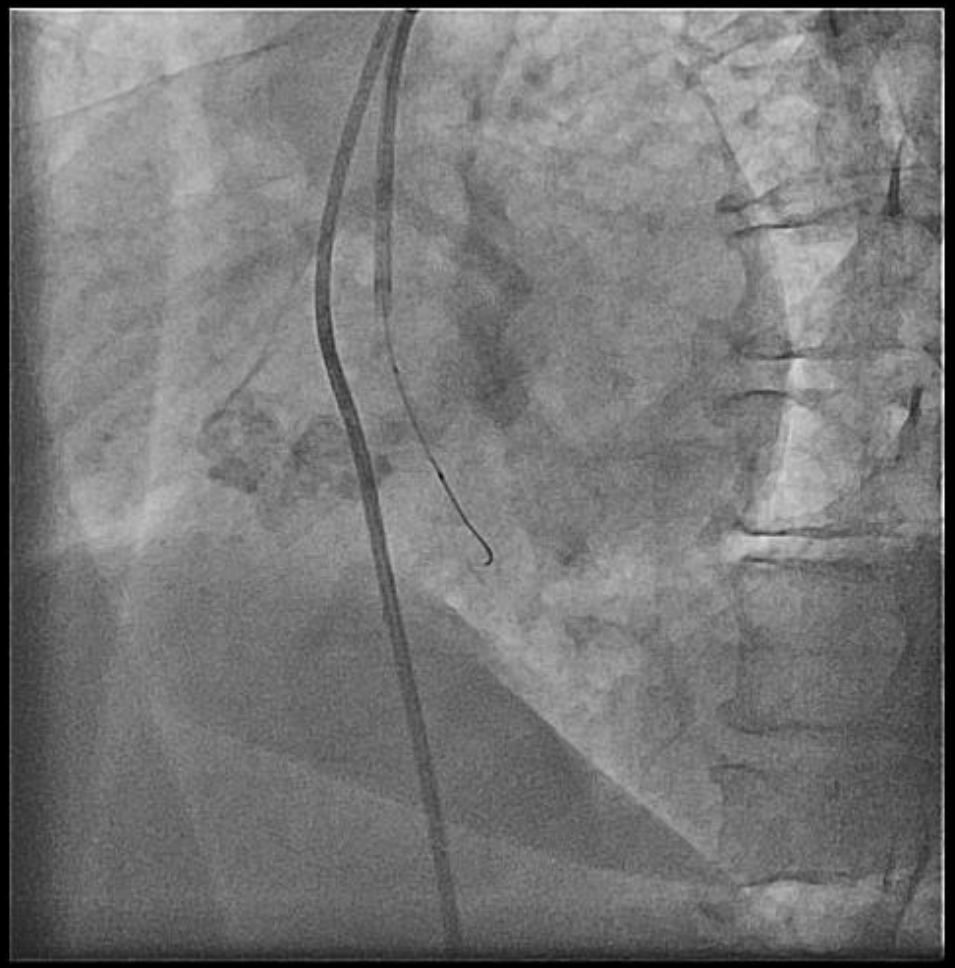
- Contraindications of BPA include iodine allergy, as the use of a contrast medium is essential in BPA.
- Additionally, in cases with renal dysfunction, the benefits of performing BPA must be weighed against the risks.
- Severity of pulmonary hypertension may not necessarily be a contraindication of BPA. Although previous reports have indicated a higher mean PAP at baseline is associated with more frequent complications, the patient prognosis will be worse without effective treatment in cases with severe hemodynamics. BPA can be expected to have more powerful effect in these patients

PULMONARY BALLOON ANGIOPLASTY (12/2016)

R A5 segment



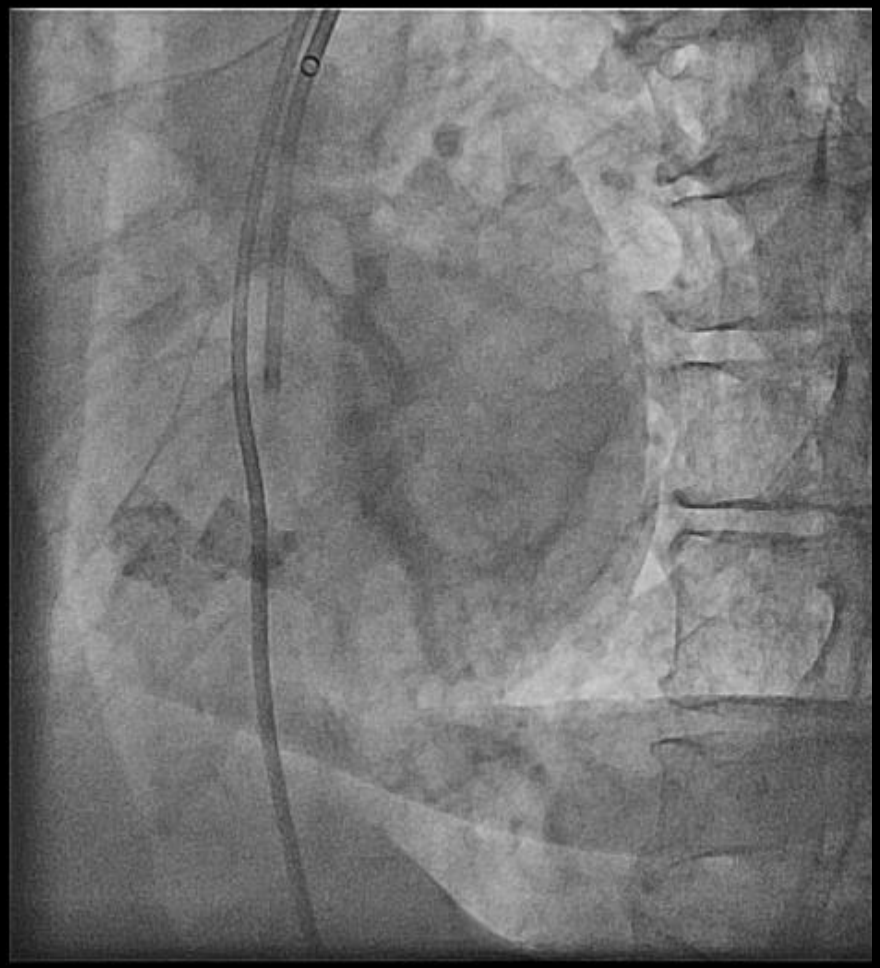
balloon 2.0X20 mm

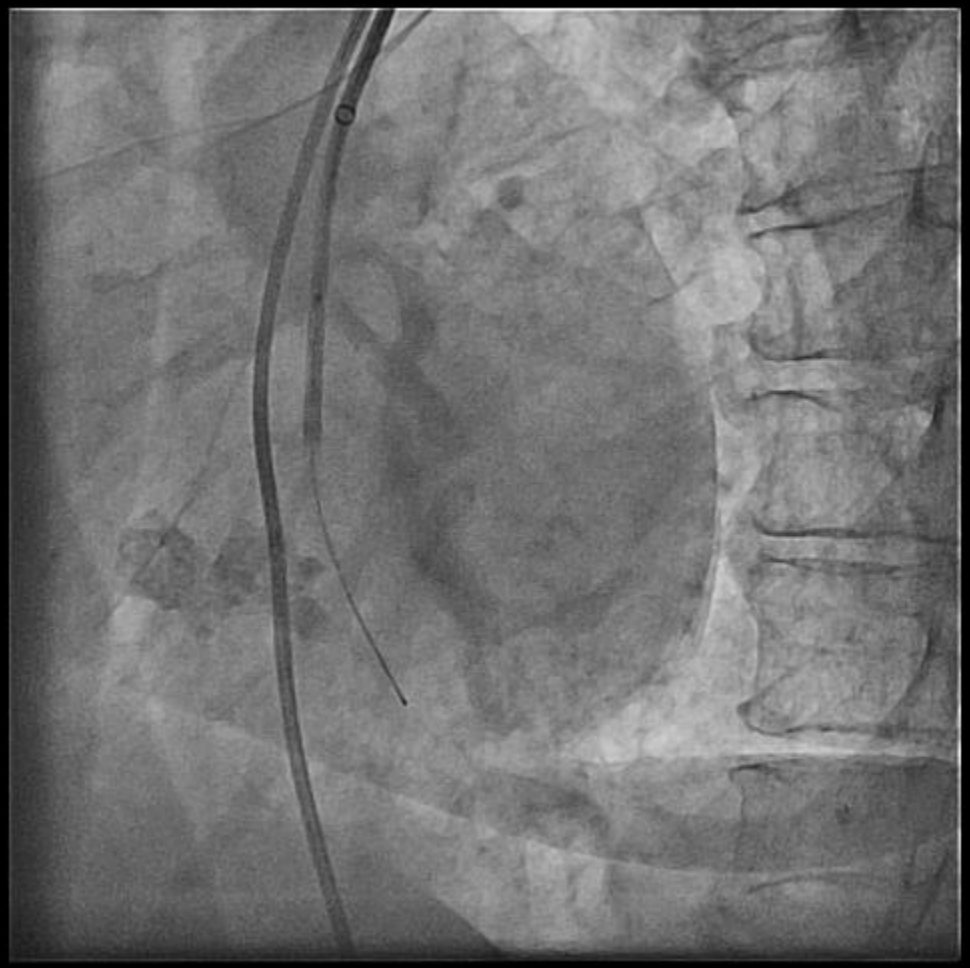


R A5 segment

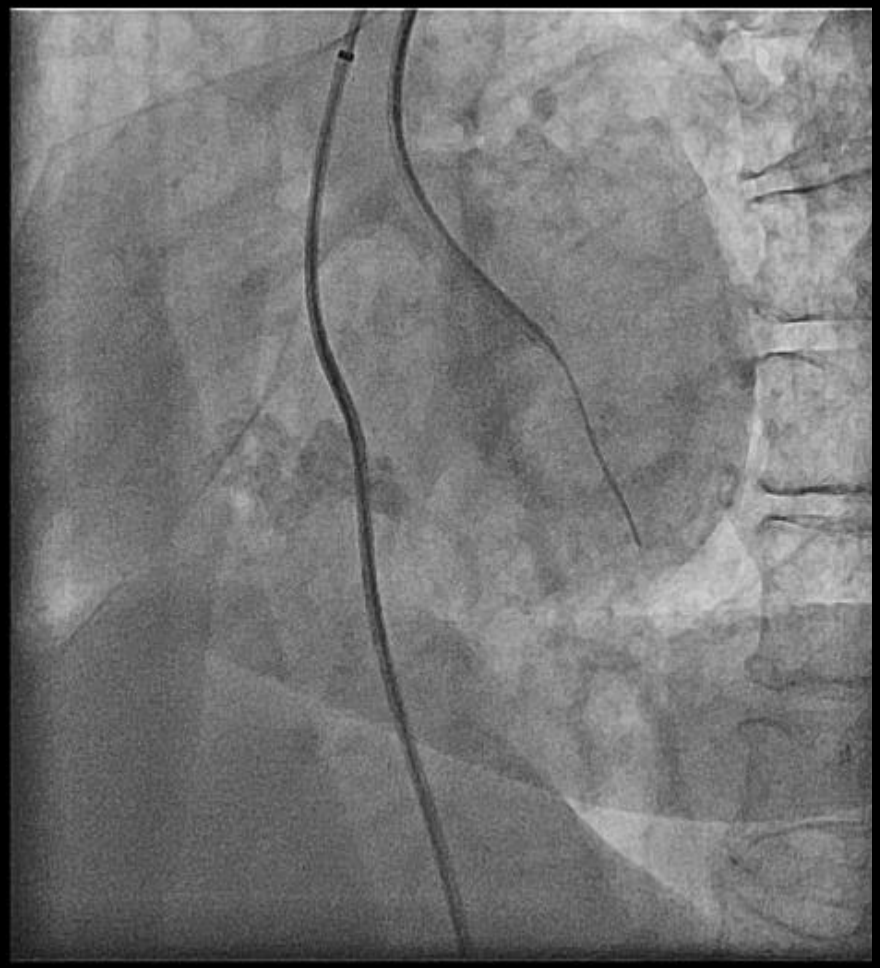
PRE

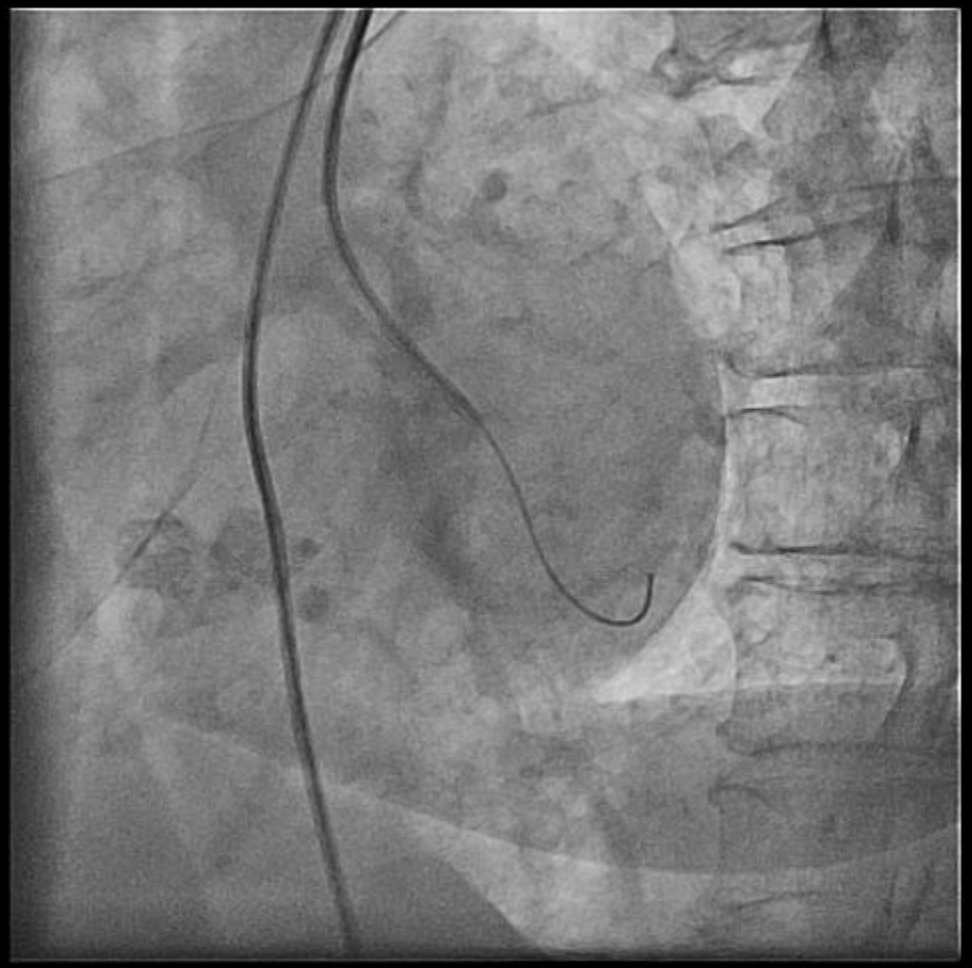
AFTER BALLOON





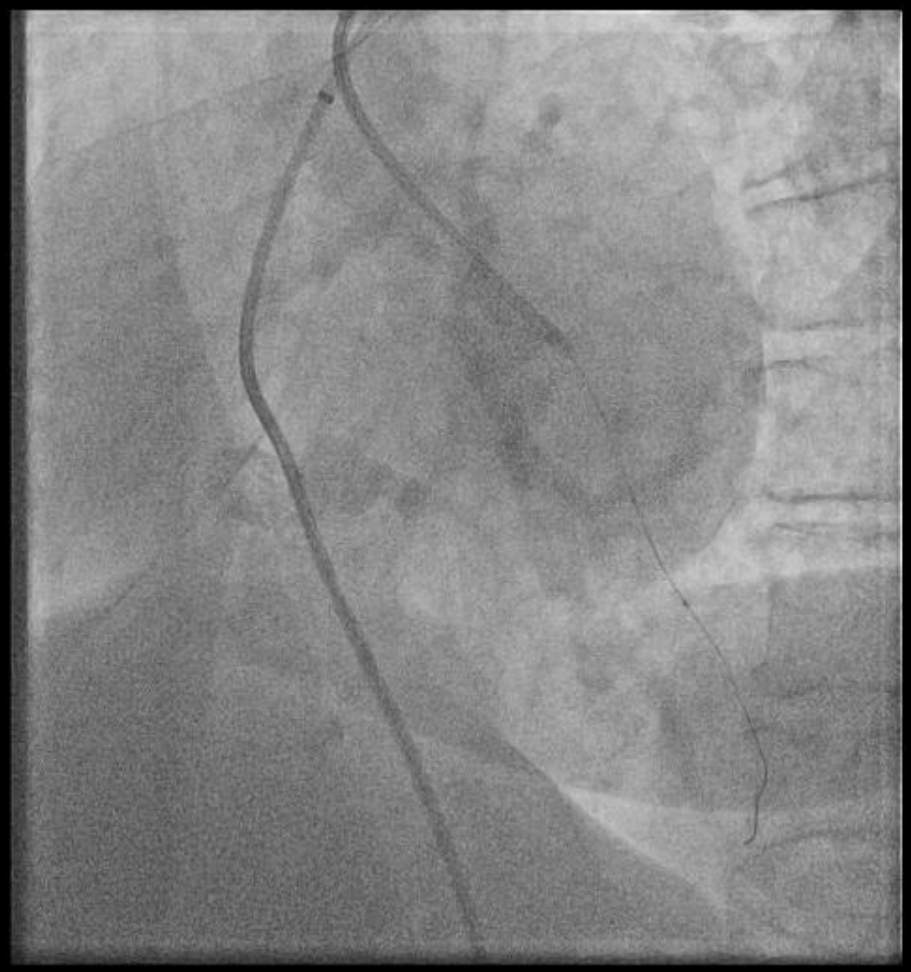
R A10 segment



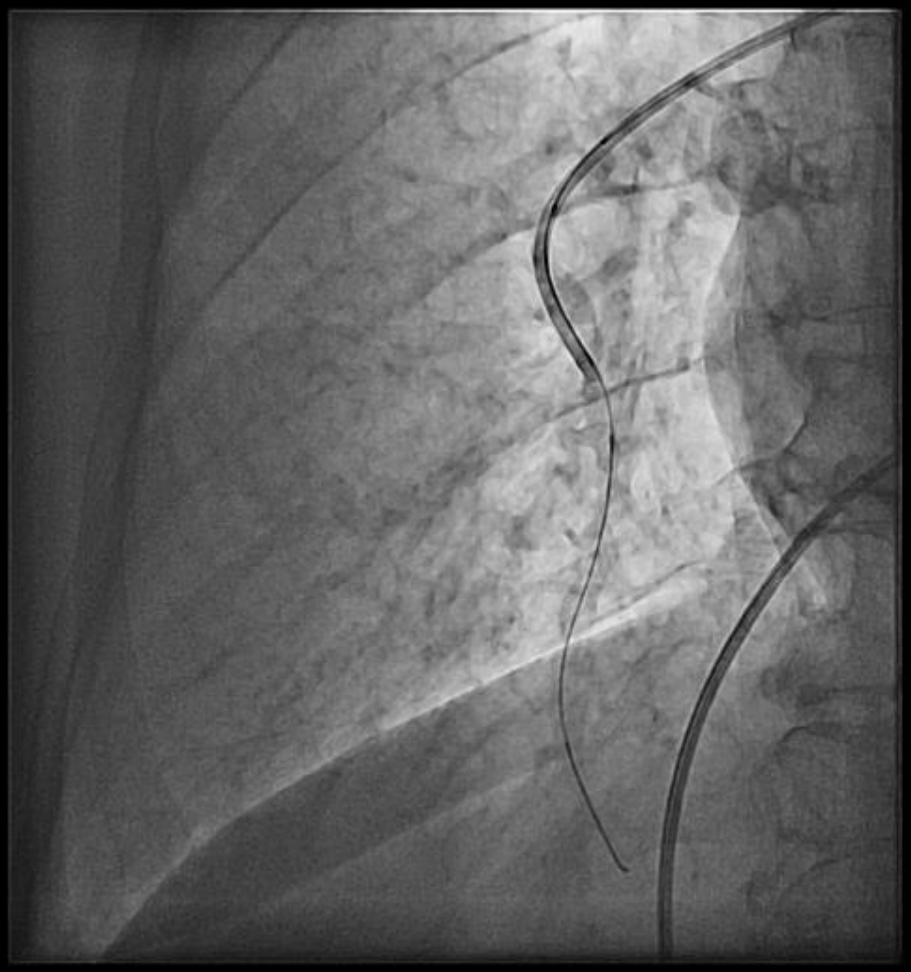


R A10 segment

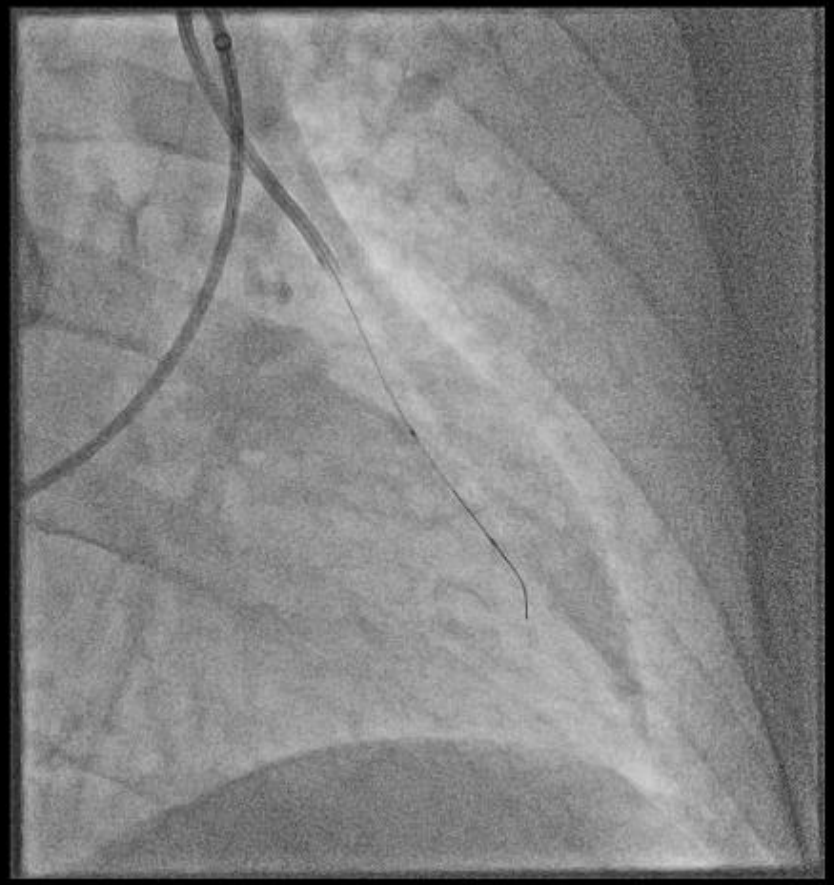
Balloon (LAO 60^0)



Post Balloon (AP view)

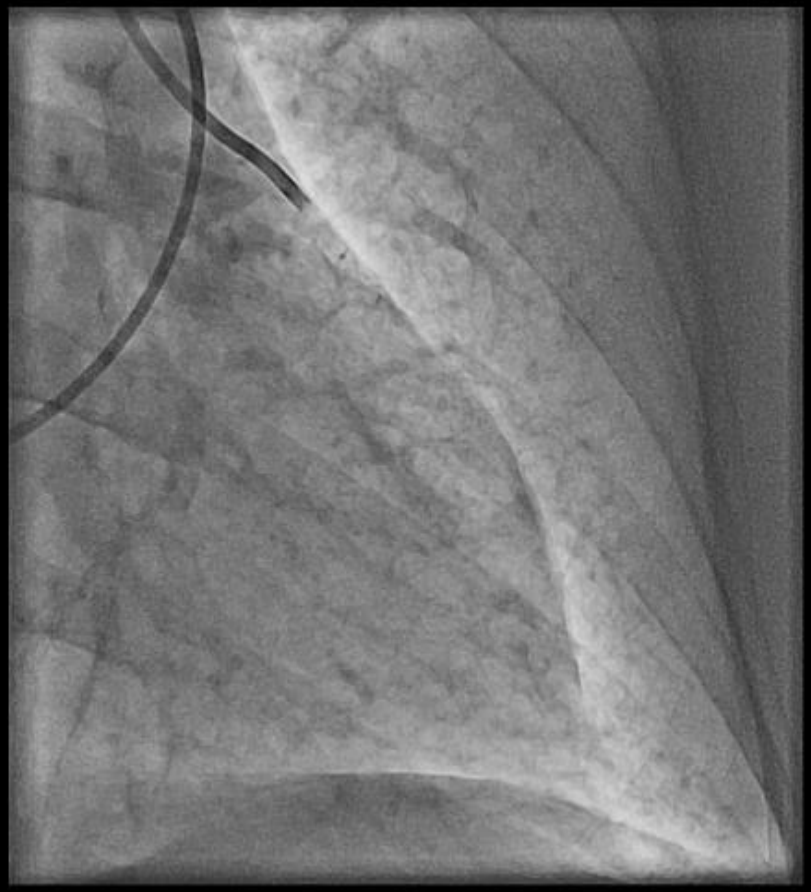


L A5b segment

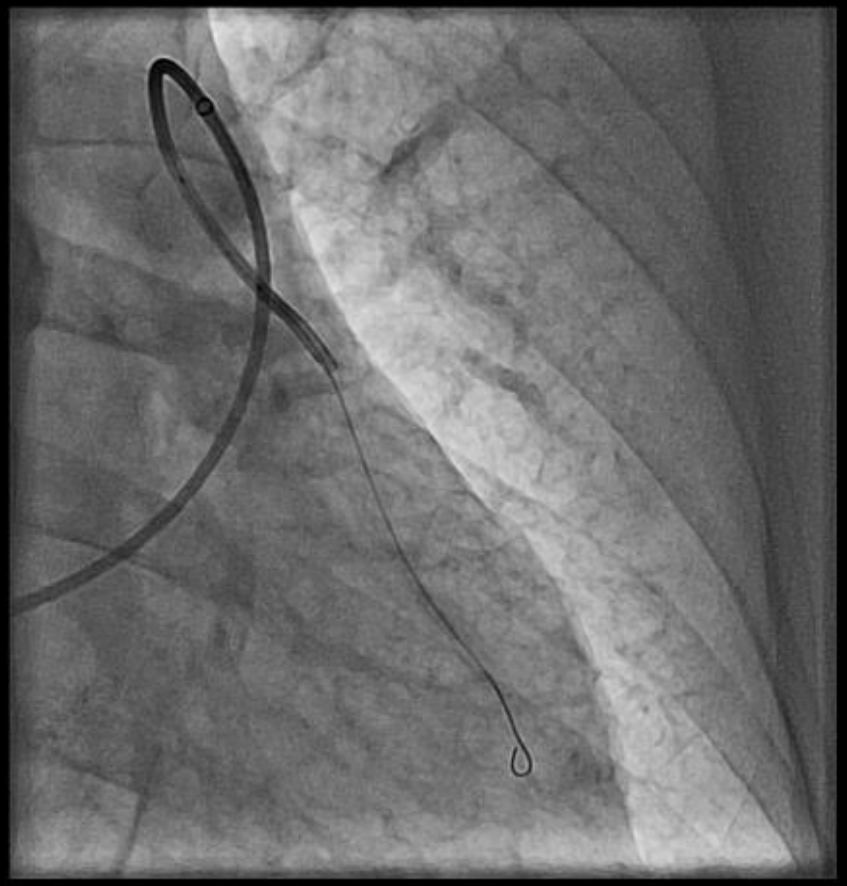


L A5b segment

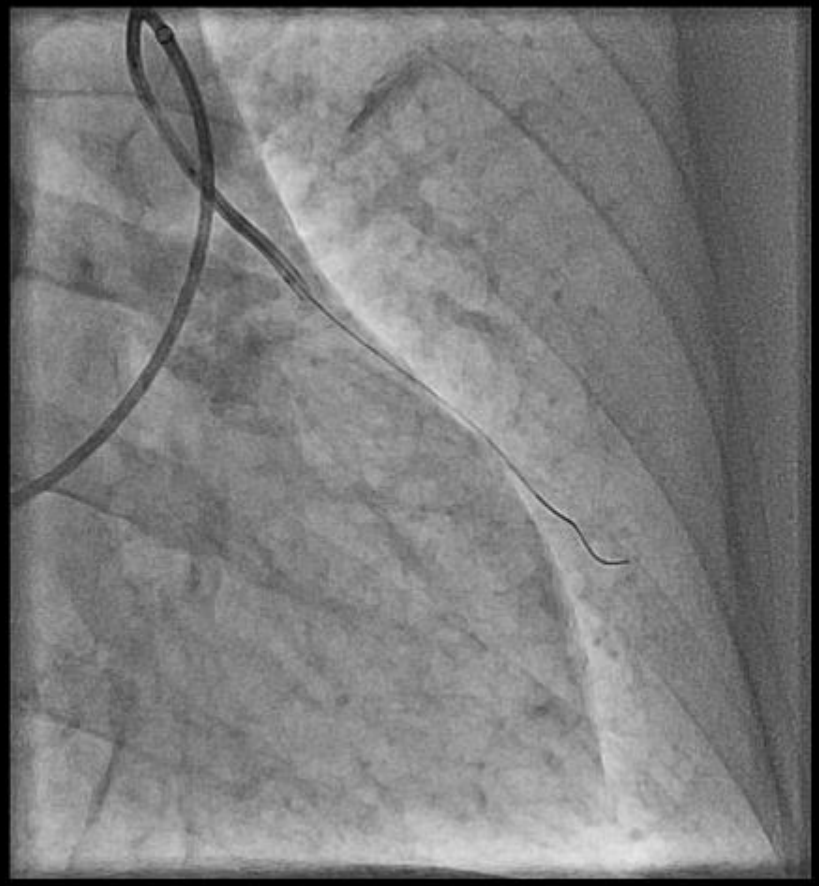
PRE



After Balloon

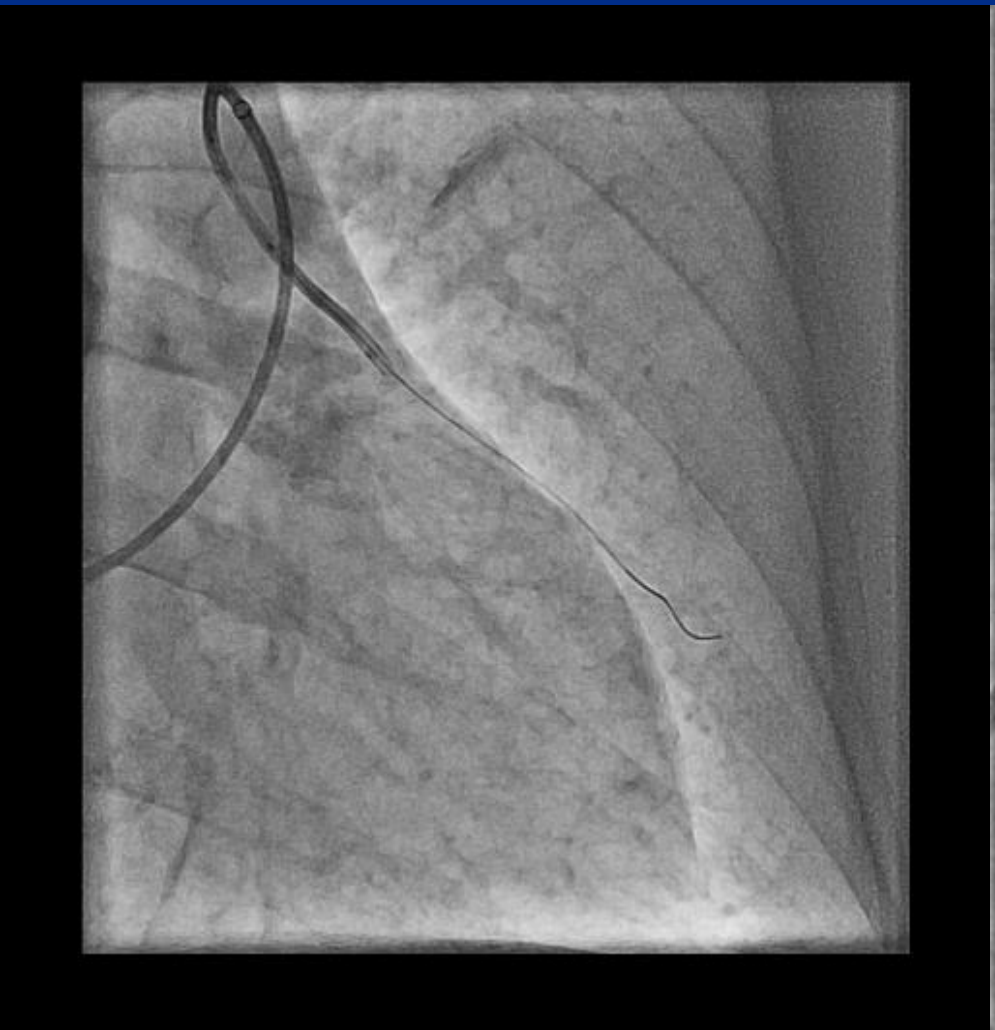


L A5a segment



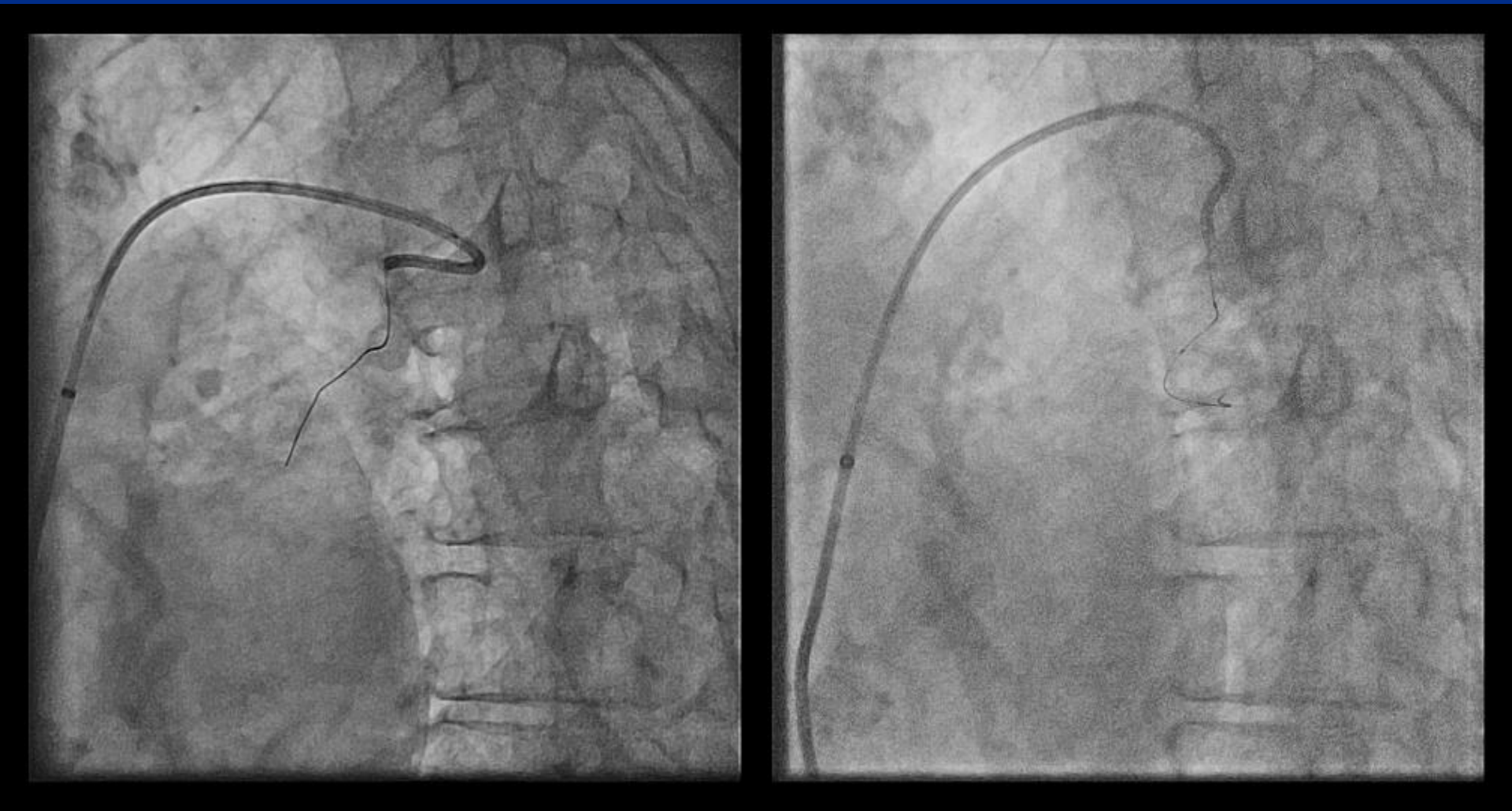
L A5a segment

PRE



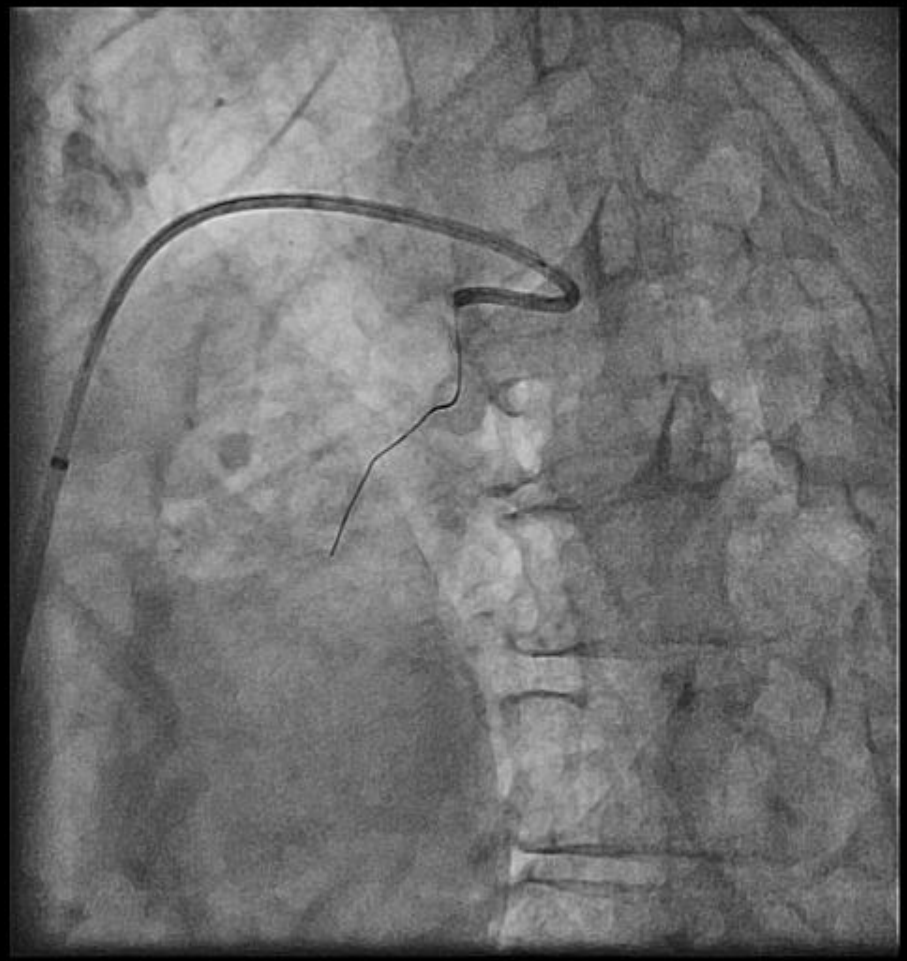
After Balloon

L A3 segment

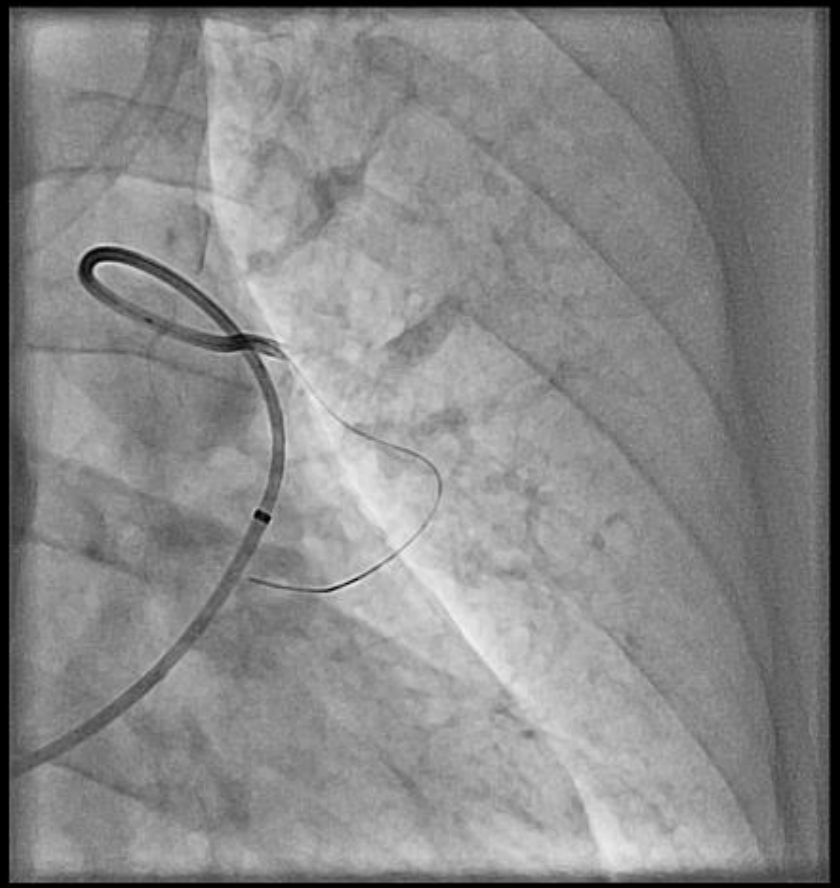


L A3 segment

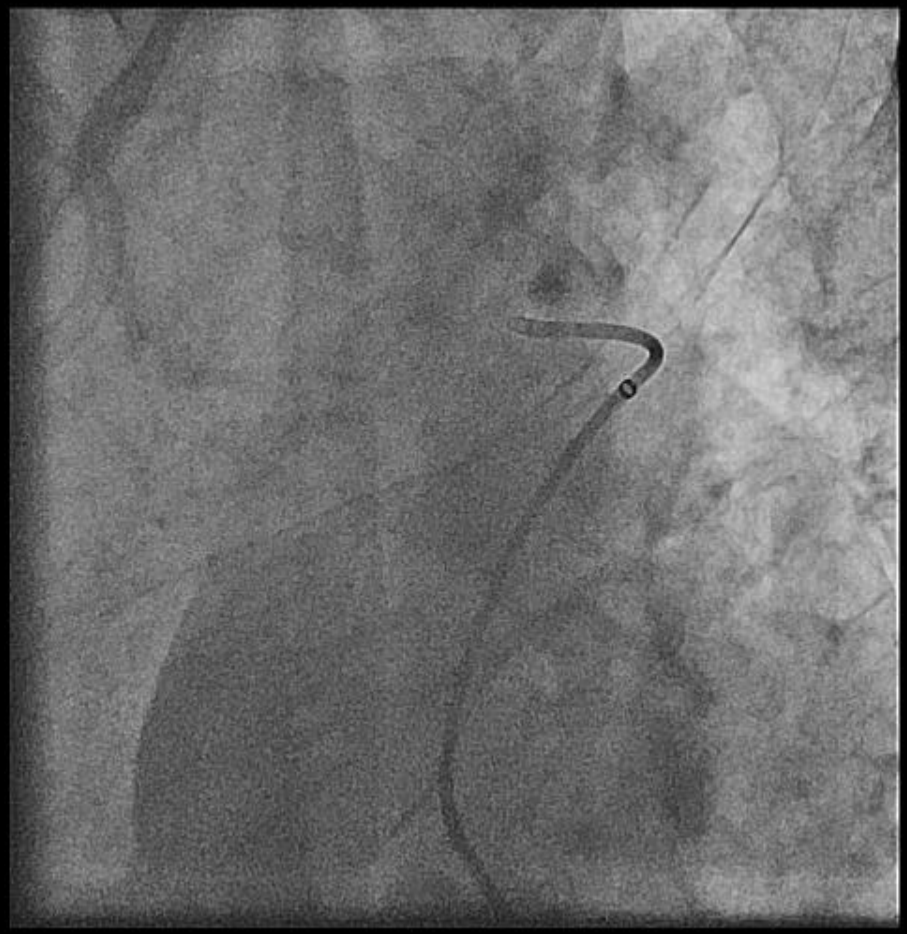
PRE (LAO 60^0)

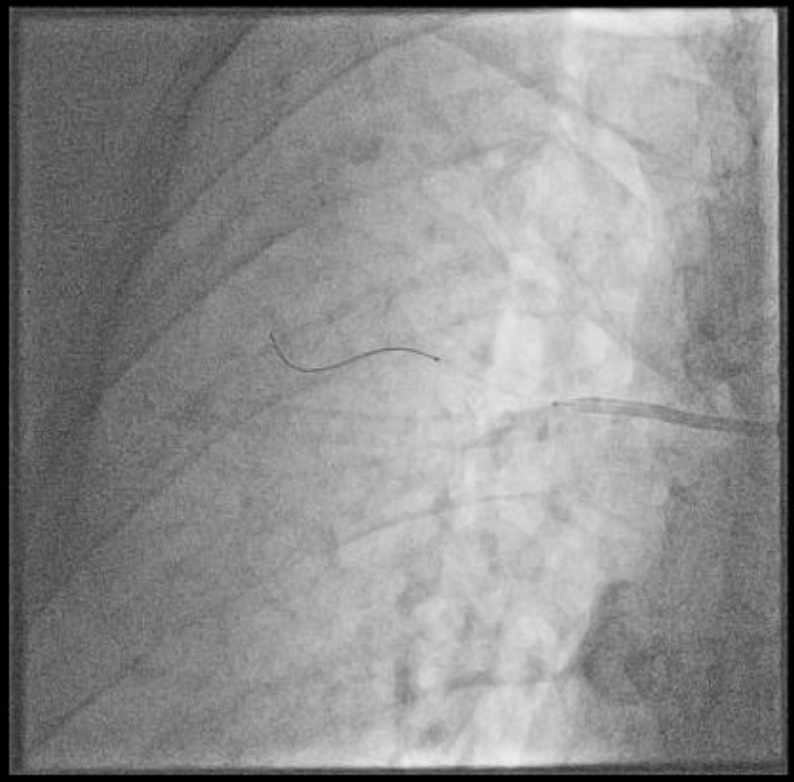


POST Balloon (AP view)



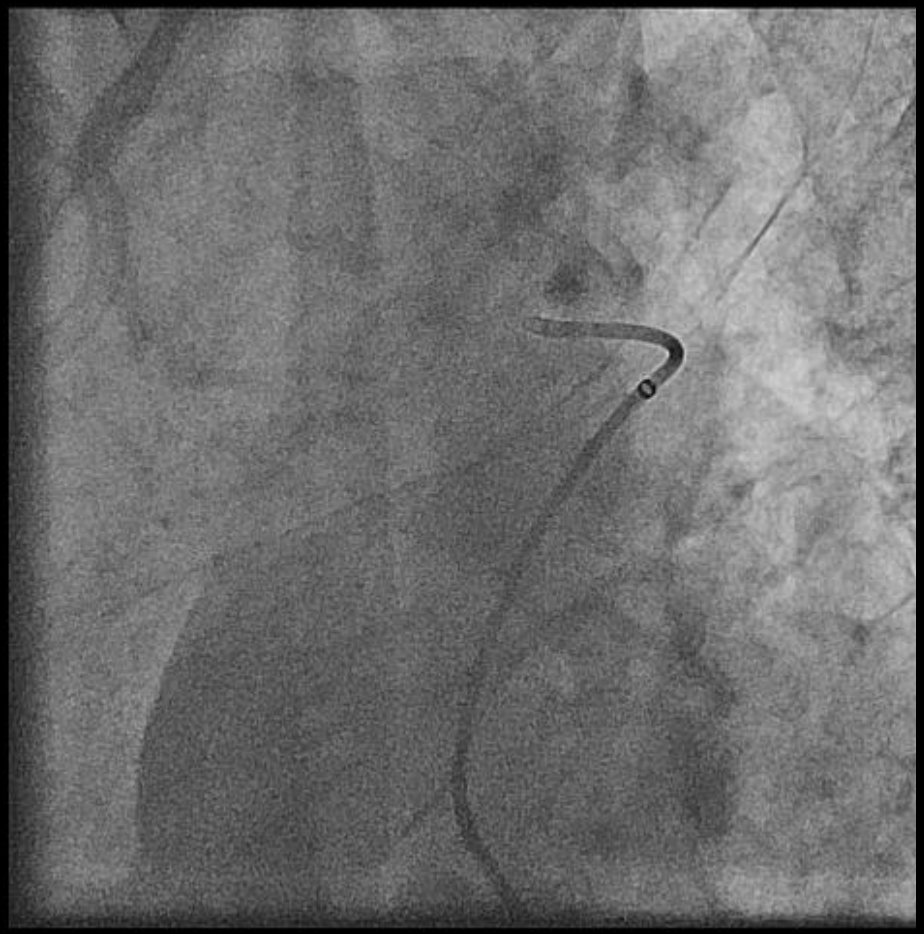
R A3 branch



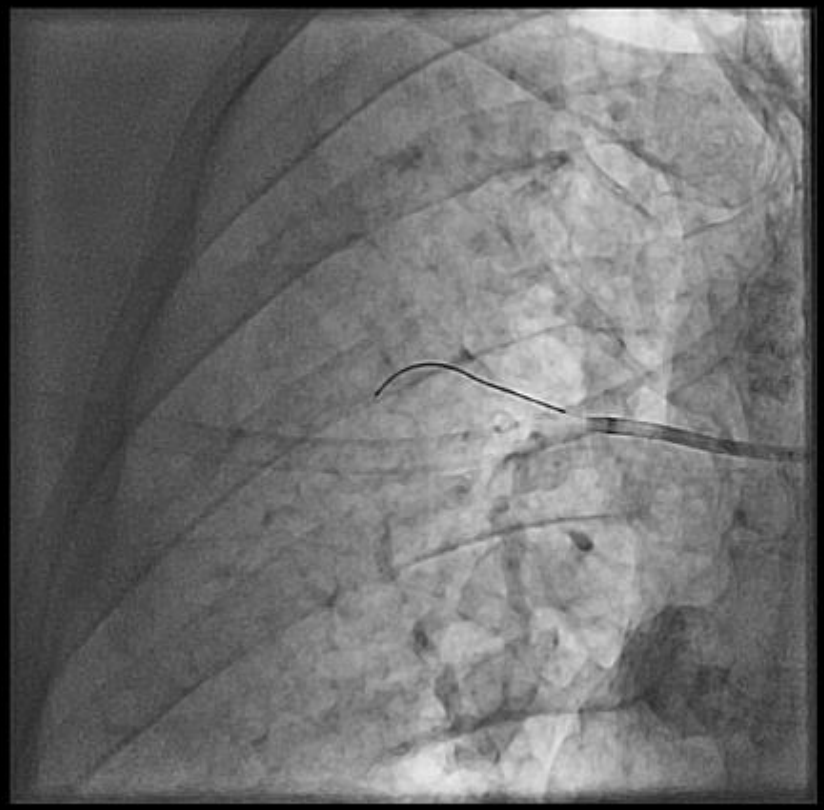


R A3 branch

PRE



POST BALLOON



- Totally 15 lesions treated in two sessions
- Saturation at the beginning: 83% - 89% on O_2
- Saturation at the end 95%-97% on O_2
- mean PA decreased from 37 mmHg to 31 mmHg
- Scheduled for another sessions:

1. Efforts to reopen total occlusions in left lower lobe
2. Optimization of results in already treated lesions with a larger balloon
3. Angioplasty in untreated lesions

- There is no limitation regarding the number of lobes targeted in one session.
- The maximum time of radiographic fluoroscopy in a single session should be limited to 60 min.
- As a consequence, lesions from 4–10 sites could generally be treated in a single session.
- To enhance the therapeutic effect of BPA, it is important to make the area of reperfusion large, which requires repeated treatment with four to five sessions per patient depending on the treatment goal

Complications of BPA

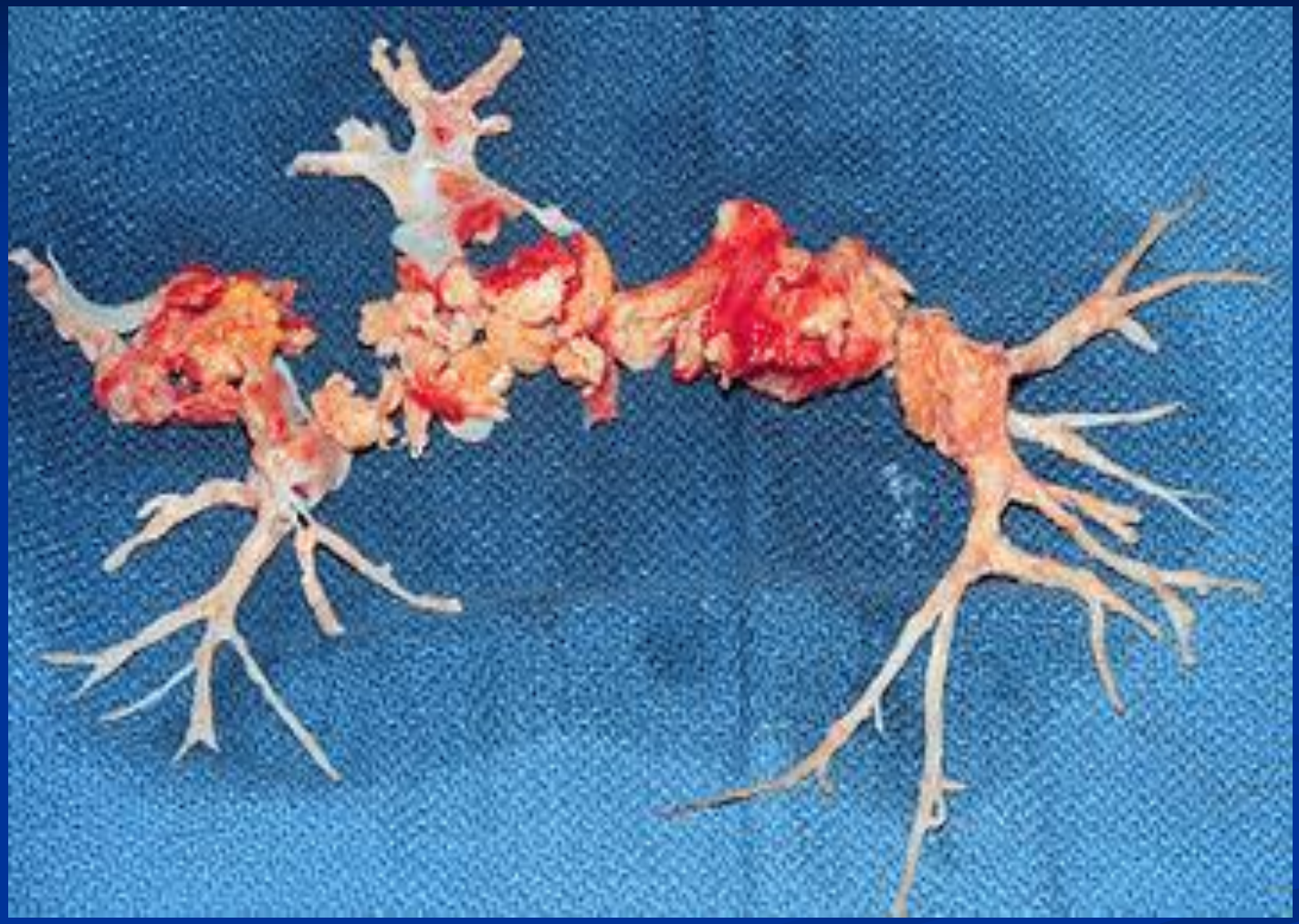
Complications	Diagnostic Criteria
Reperfusion pulmonary injury	Hemoptysis
	Chest radiographic opacities
	Chest computed tomographic opacities
Pulmonary artery perforation	
Pulmonary artery rupture	

Conclusions

- BPA is an alternative therapeutic option in inoperable patients with CTPH
- A very demanding technique in complex anatomy
- Biplane is mandatory
- Improved strategies to overcome the complications associated with BPA must be established
- Randomized control trials to evaluate the superiority of BPA over drug therapy and studies on the cost-effectiveness of BPA procedures are needed
- Long-term survival and efficacy need to be clarified.

THANK YOU

Thromboendarterectomy - CTEPH



Chronic Myeloproliferative Disorders

- PHT : the most prominent cardiac pathology.

Int J Cardiol, 2004; 97:213-220.

- Thromboembolic complications in thrombocythemia.
- Inappropriate state of platelet activation.

Semin Thromb Hemost. 1997;23:391-402.

- Medical agents: Phospholipase A2 Inhibitor, antiplatelet therapy & hydroxyurea .

SPLENECTOMY & C.T.E.P.H.

- Pathophysiologic mechanisms poorly defined.
- Development of a prothrombotic state.

  Abnomal erythrocytes remain in the peripheral circulation → abnormal exposure of phosphatidylserine on the surface of erythrocytes → activation of the coagulation process.

- Splenectomy was found to be an independent risk factor for CTEPH.

  Bonderman D, Maurer G, et al.:
  Thromb Haemost 2005; 93:512-516.

- CTEPH associated with splenectomy is distal.

  Jais X, Simonneau G, Humbert M et al.:
  Thorax 2005; 60:1031-1034.

Right Heart Thrombi

- The prevalence in unselected pts with P.E. : 4%.

- Associated with increased early mortality.
- Untreated mobile thrombi: death rate 80-100%.

- The treatment of choise is controversial.
- Thrombolysis / embolectomy more effective.
- Anticoagulation alone less effective.

ESC Guidelines, 2008.

Treatment of Right Heart Thromboemboli

Retrospective analysis of 177 cases (1966-2000)

Treatment		*Mortality Rate*
none	9 %	100 %
anticoagulation	35 %	28,6 %
embolectomy	35,6 %	23,8 %
thrombolysis	19,8 %	11,3 %

Chest 2002; 121:806-814.

Lung Computer Tomography (CT)

- Organised thrombus (eccentric).
  Abrupt narrowing & tapering of P.A., pouch defects.
  Auger W, et al: Clin Chest Med 2007; 28:255-69.

- Mosaic perfusion pattern of lung parenchyma.
- Central pulmonary artery enlargement.
- RA & RV enlargement.

- Detection of mediastinal pathology (unilateral occlusion of central pulmonary artery) or parenchymal lung disease.