CTEPH
surgical treatment

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1 Pulmonary arterial hypertension (PAH)

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2
  - 1.2.2 ALK1, endoglin (with or without hereditary haemorrhagic telangiectasia)
  - 1.2.3 Unknown
- 1.3 Drugs and toxins induced
- 1.4 Associated with (APAH)
  - 1.4.1 Connective tissue diseases
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
  - 1.4.6 Chronic haemolytic anaemia
- 1.5 Persistent pulmonary hypertension of the newborn

1′ Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

2 Pulmonary hypertension due to left heart disease

- 2.1 Systolic dysfunction
- 2.2 Diastolic dysfunction
- 2.3 Valvular disease

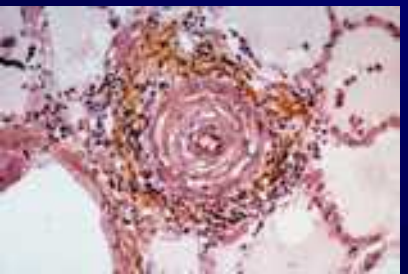
3 Pulmonary hypertension due to lung diseases and/or hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental abnormalities

4 Chronic thromboembolic pulmonary hypertension

5 PH with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders: myeloproliferative disorders, splenectomy.
- 5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoural obstruction, fibrosing mediastinitis, chronic renal failure on dialysis



Clinical Classification of PH

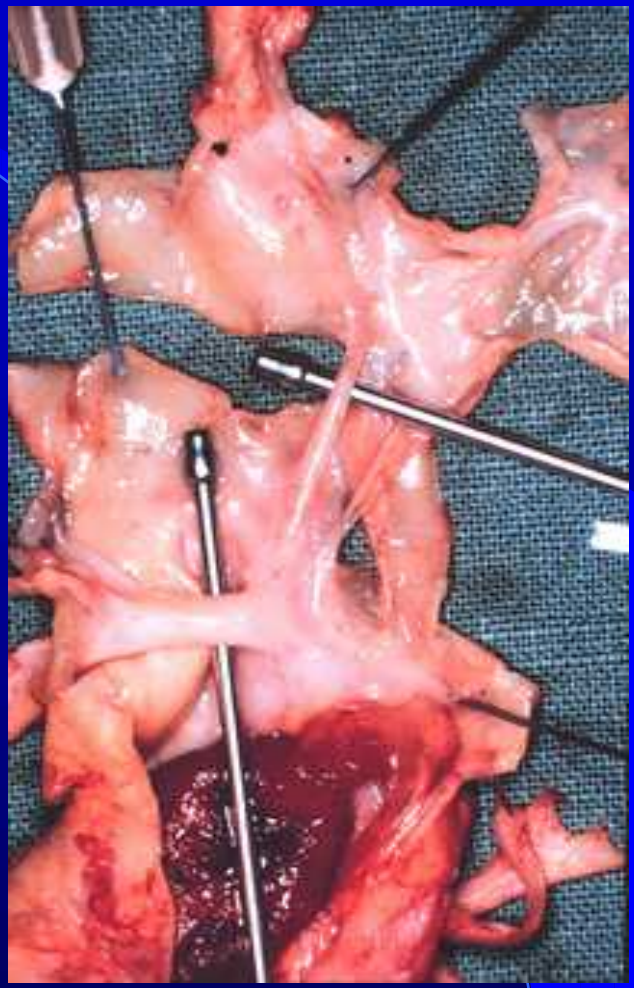
G Simonneau et al JACC 2009

Incidence

- The true incidence of CTEPH is difficult to evaluate
  - Very few studies have analyzed the long-term outcome of patients presenting with acute PE
  - Large proportion of patients (70%-80%) have had PE that remains undiagnosed before death or until PH develops
- 0.5% to 5% of patients can develop CTEPH after the first episode of acute PE
- CTEPH can occur in 1/3 of patients with recurrent PE
- Rough estimates of >5,000 cases/ year in the US

Definition of CTEPH

- CTEPH is symptomatic PH (mPAP $\geq$ 25 mmHg) with persistent perfusion defects despite 3-6 months of adequate anti-coagulation
- CTEPH is a disease with:
  - a mechanical component judged amenable to surgery
  - and variable small vessel disease



Pathophysiology

- Natural course of acute PE is complete resolution of the acute embolus
- CTEPH is a sequelea of Acute Pulmonary Embolism (PE)
- Fibrous organization of the clots

?

HOWEVER

40 to 70% of all CTEPH had previous PE (Humbert 2006- European CTEPH Registry)

Consequences -1

- Obstruction of lobar, segmental, or subsegmental PA branches by unresolved thrombo-emboli
- Modifications of the non obstructed PA territories
- Systemic pulmonary circulation develops from:
  - ✓ Bronchial arteries
  - ✓ Pleural adhesions
  - ✓ Internal mammary arteries

Mechanisms of pulmonary hypertension

PH is due to:

- **Obstruction of PA bed by thrombo-embolic material**
- **Development of vasculopathy in the distal unobstructed PA territories because of overflow redistribution**

Consequences -2

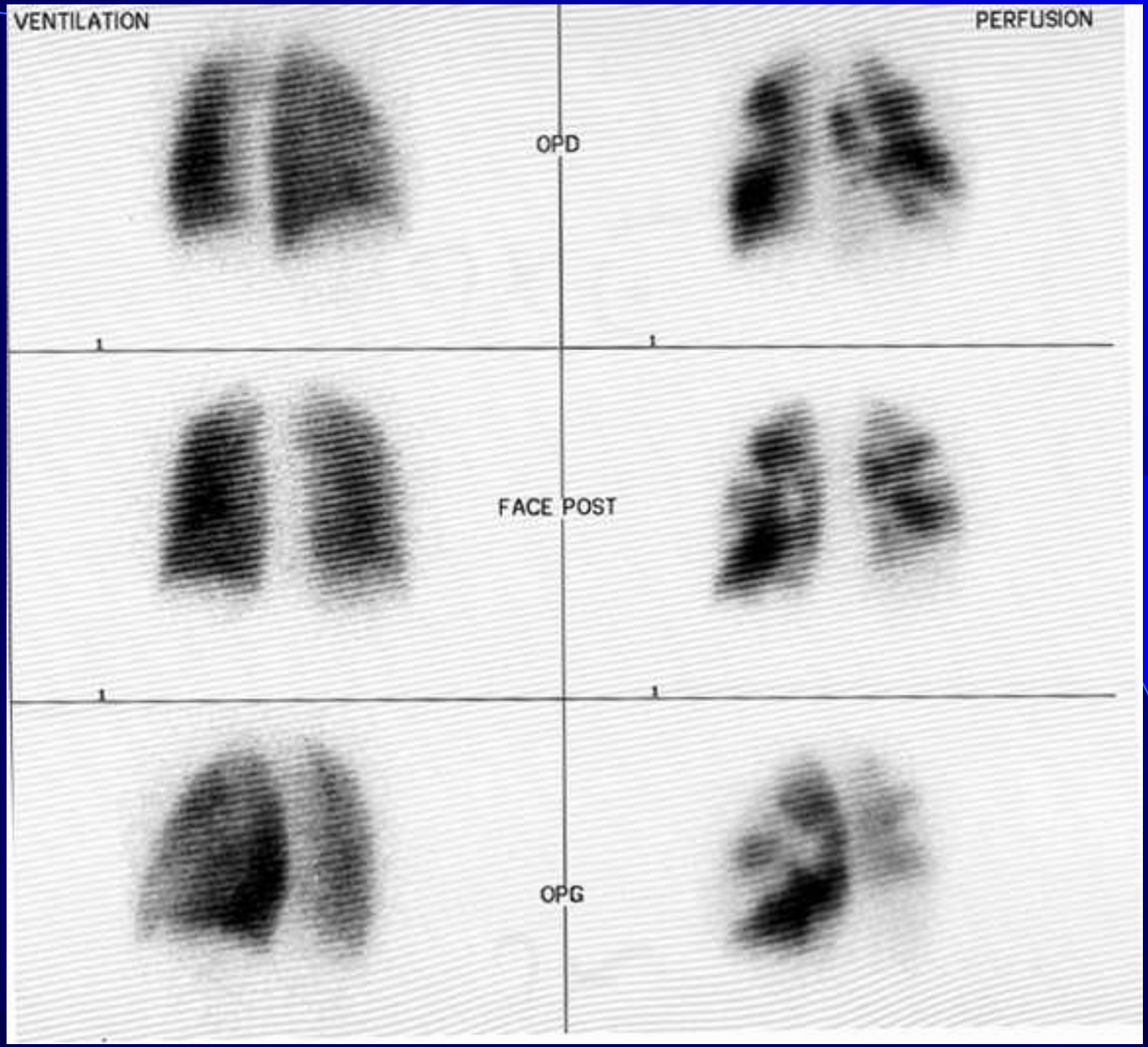
- Progressive aggravation of pulmonary hypertension is rarely due to recurrent PE
  - Typically PH progresses despite anticoagulation
- Aggravation of pulmonary hypertension is mainly due to the development of a distal vasculopathy in the unobstructed PA territory
- Changes in the unobstructed PA bed can lead to an inoperable situation or can dramatically increase the risk of surgery
- Pulmonary endarterectomy is better earlier than later in the course of the disease

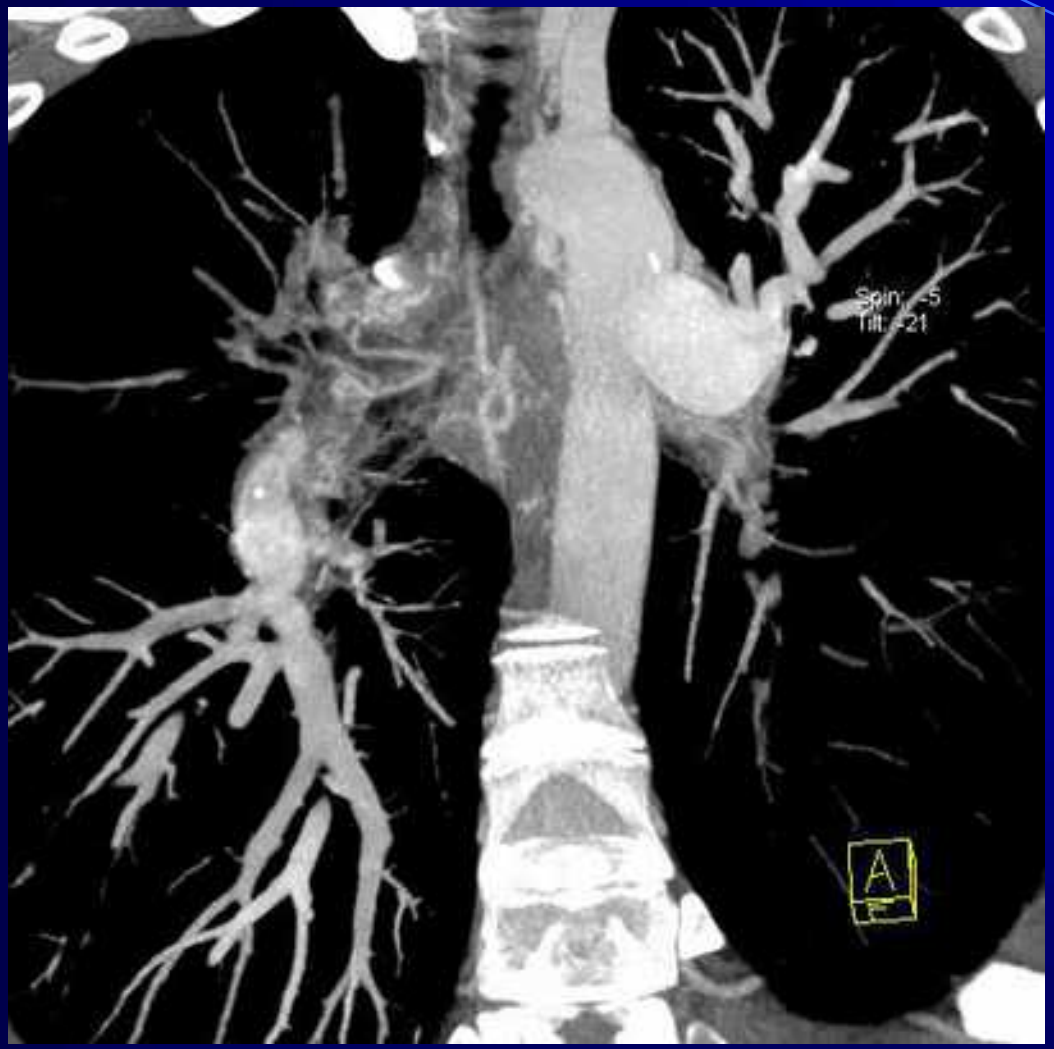
Mechanisms of dyspnea

- Pulmonary hypertension
- Ventilation perfusion mismatch

Preoperative work-up

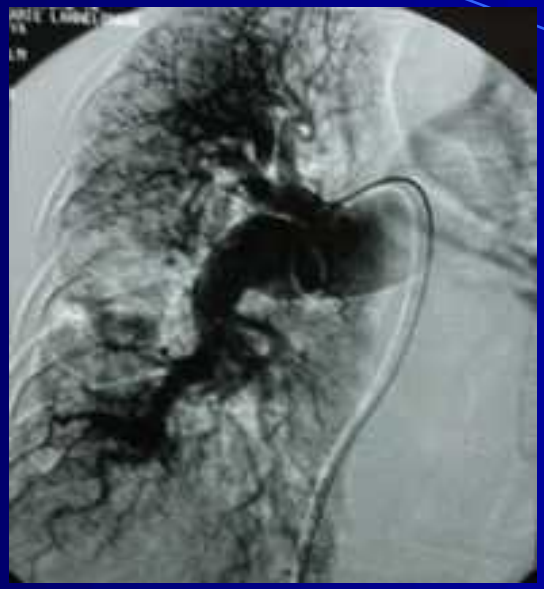
- Nuclear scan Ventilation and Perfusion
- Angiography
- CT scan with reconstruction
- Right catheterization
- Correlation between anatomic obstruction and hemodynamic severity

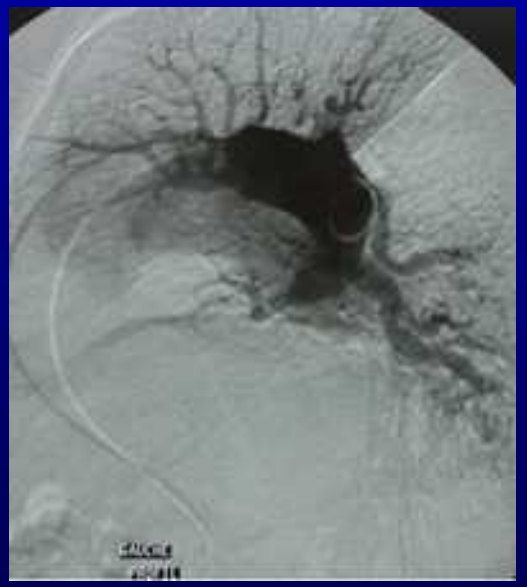

VENTILATION
PERFUSION
OPD
FACE POST
OPG

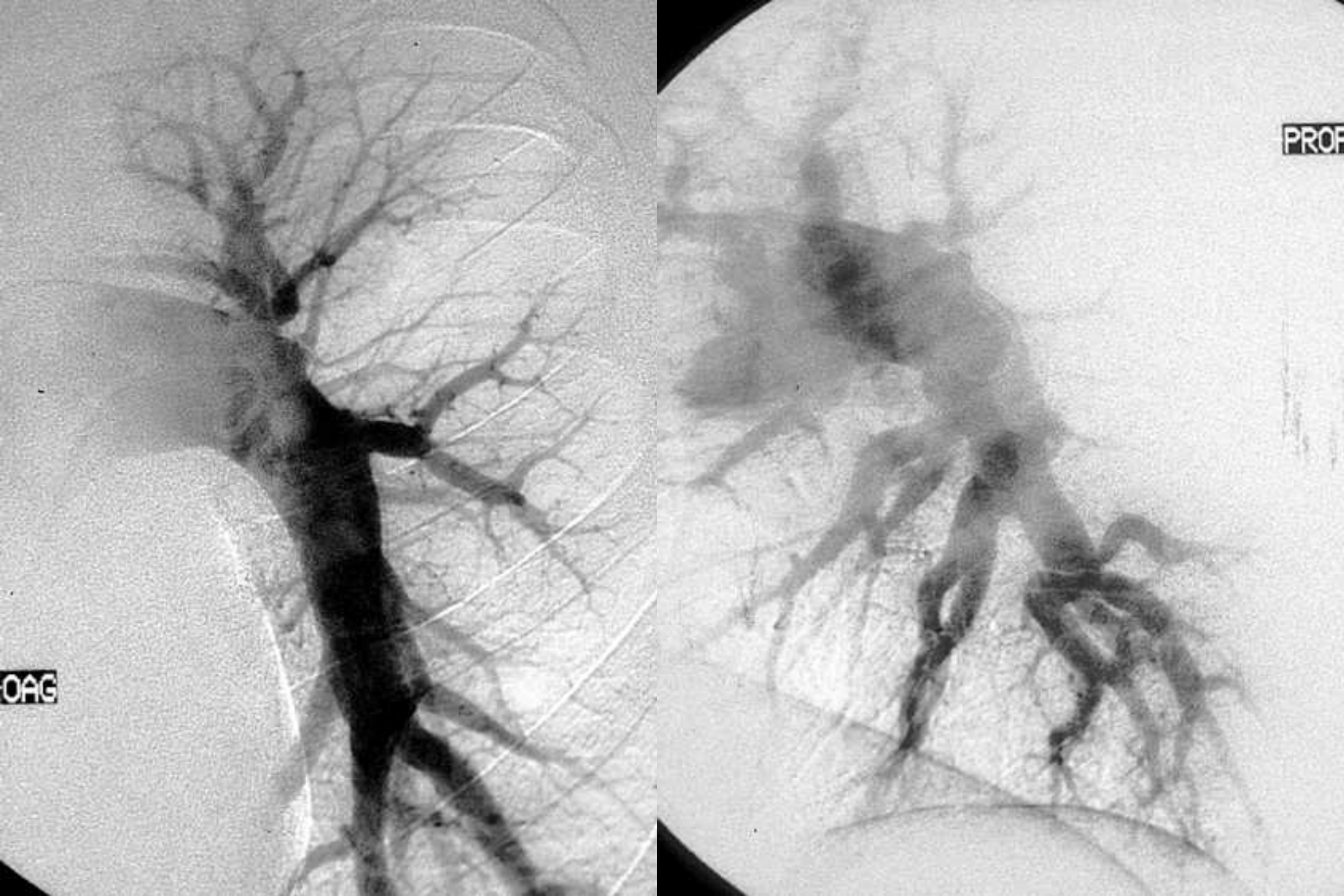

A


Spin: -90
Tilt: 0
L

Pulmonary Angiogram





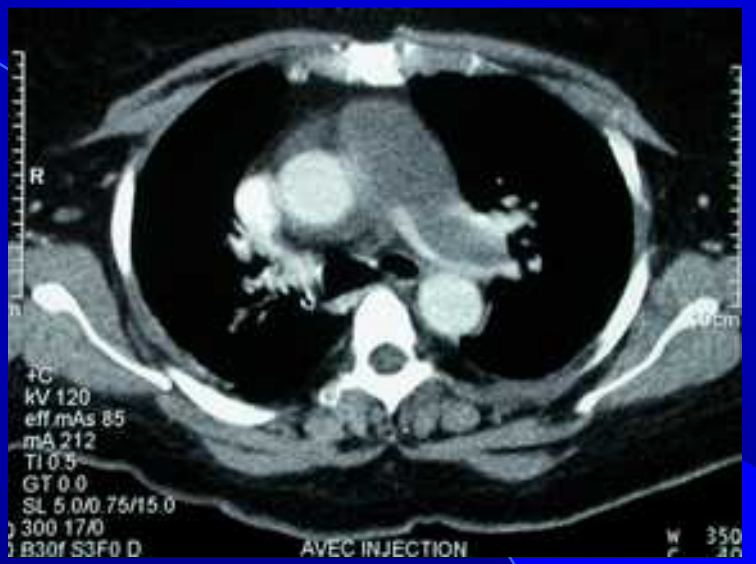

OAG
PROF

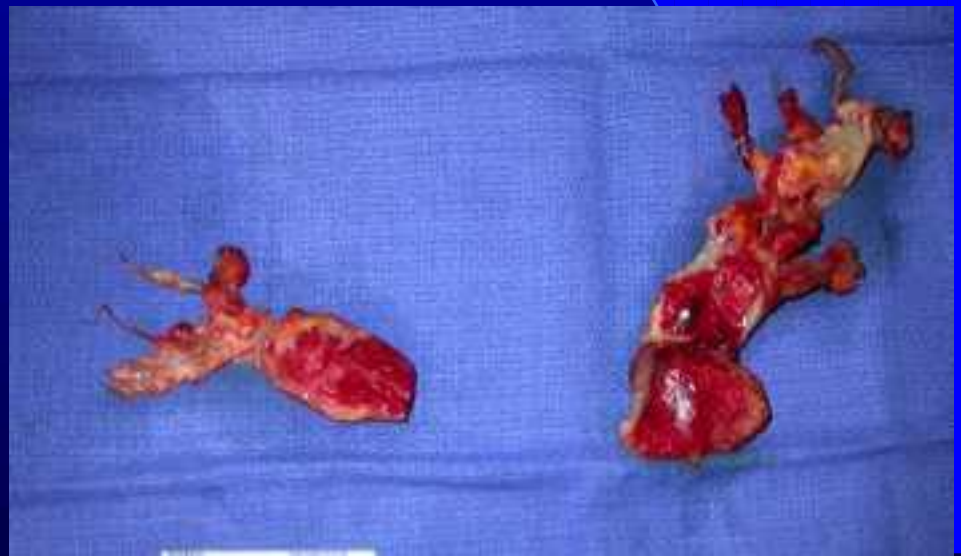
CTEPH

947 Dynes.s.cm-5

PAH

2100 Dynes.s.cm-5





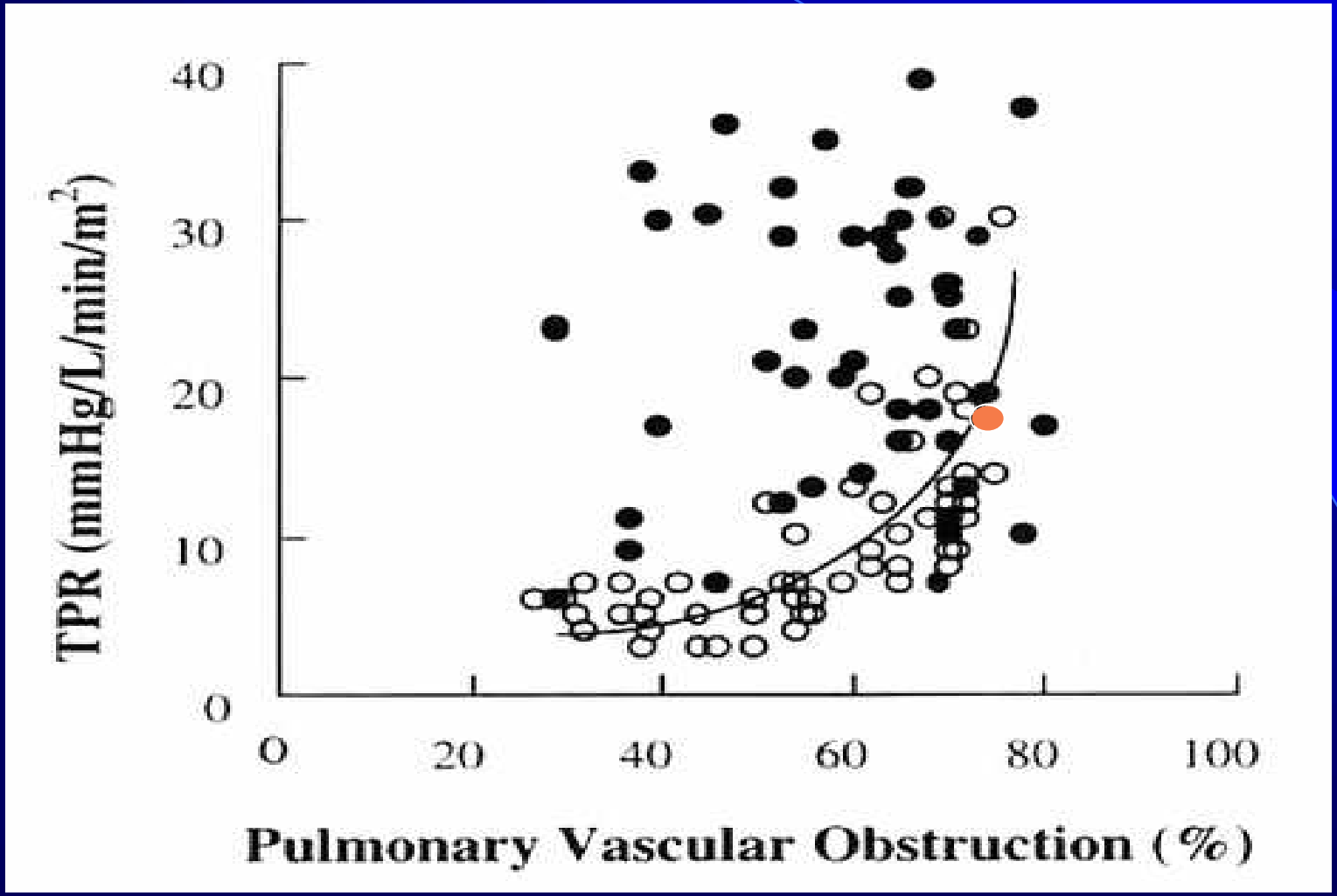
JAMIESON CLASSIFICATION

- Type I Disease (20 %) Thrombus + Fibrotic Tissue
- Type II Disease (70 %) Thickened intima
- Type III Disease (10 %) Distal disease

Indwelling catheters
Ventriculo-atrial shunts
PM wires

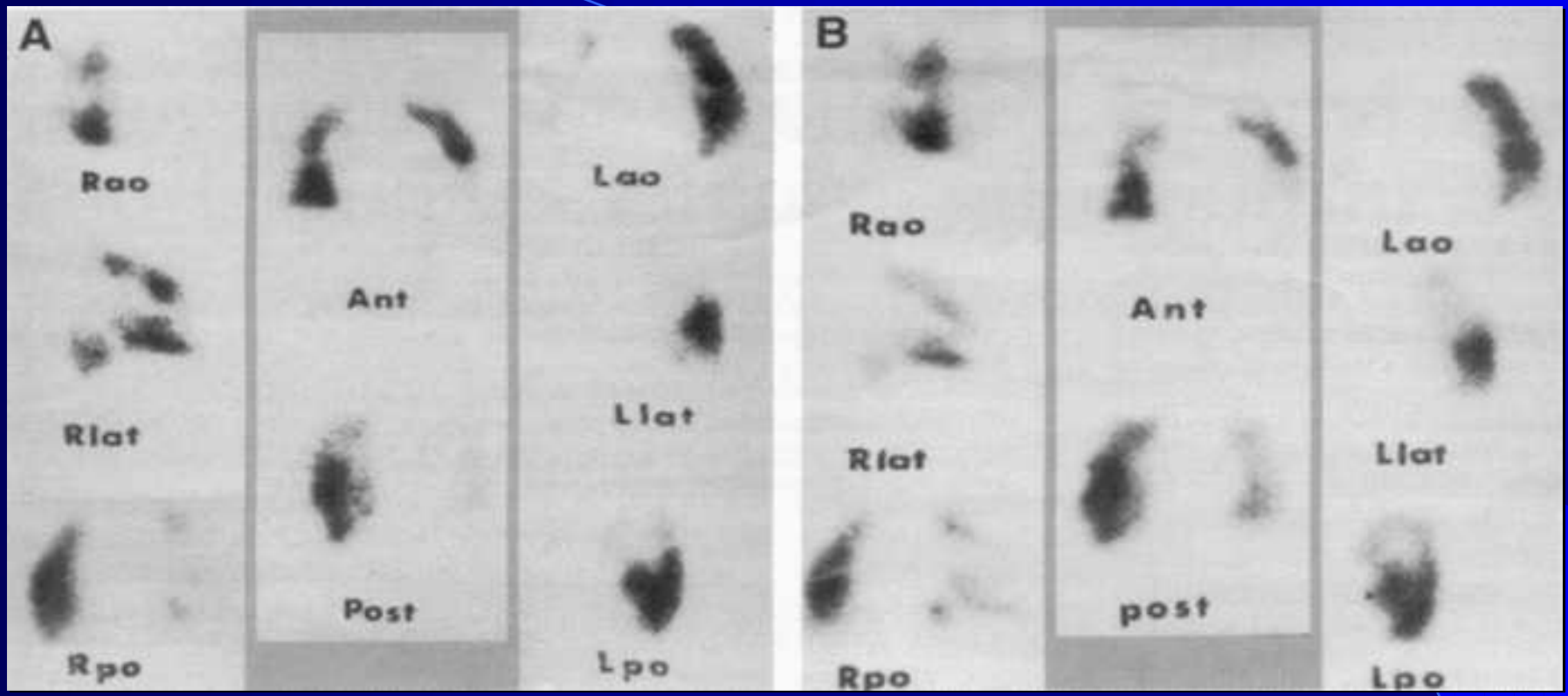
- Type IV Disease PHT

Correlation between anatomic obstruction and PVR



The right time for PEA

- Early prior developing arteriolitis
- After a 3 month anticoagulant therapy time
- As first line when anatomical obstruction is correlated to hemodynamic severity
- Hemodynamic severity and distal disease ??



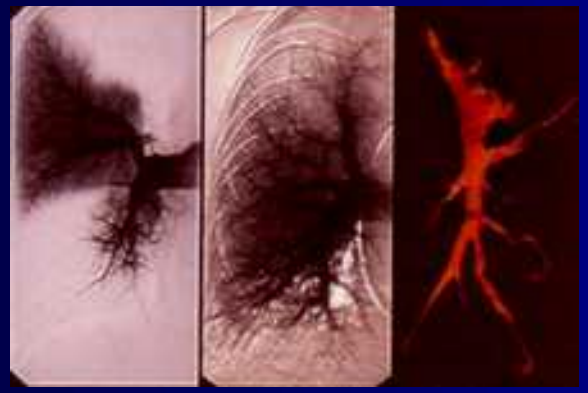
08/1993

NYHA FC 2
6'WD 440 m
Mean PAP 48 mmHg
CO 5.6 l/min
TPR 686 dyn.sec.cm^{-5}

02/1995

NYHA FC 4
6'WD 210 m
Mean PAP 54 mmHg
CO 3.4 l/min
TPR 1271 dyn.sec.cm^{-5}

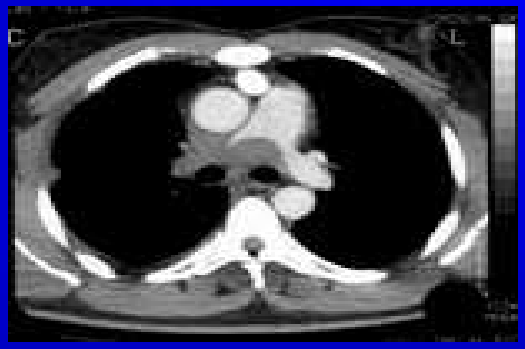
Postembolic PHT



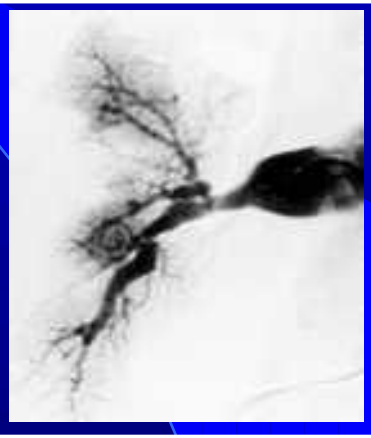
PA Angiosarcoma

Hydatic cyst

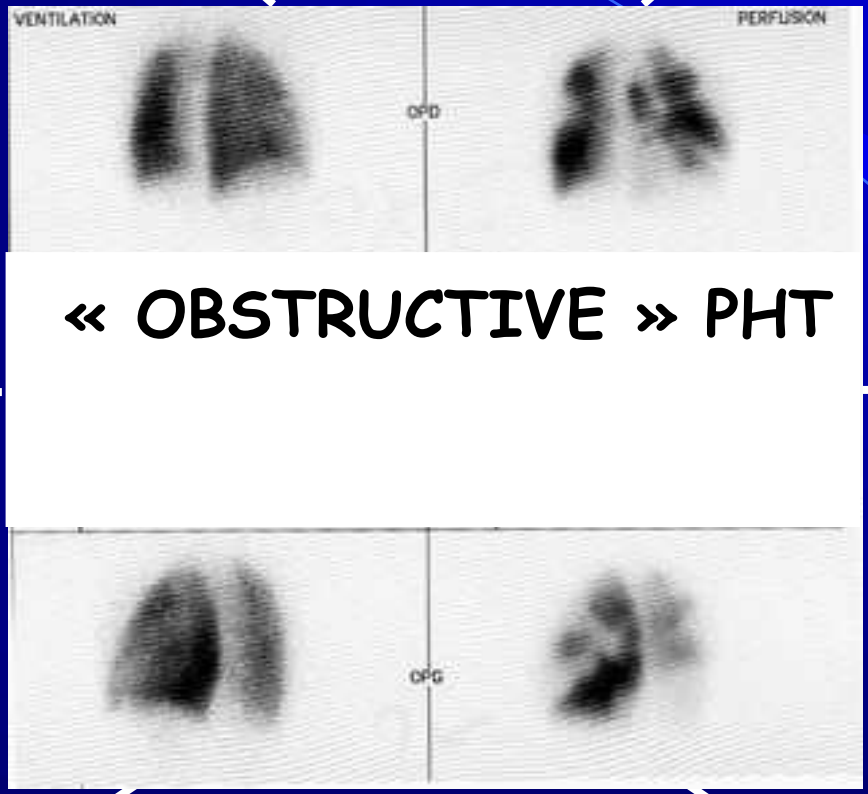
Fibrosing mediastinitis



Pulmonary arteritis



Teratoma - leiomyoma

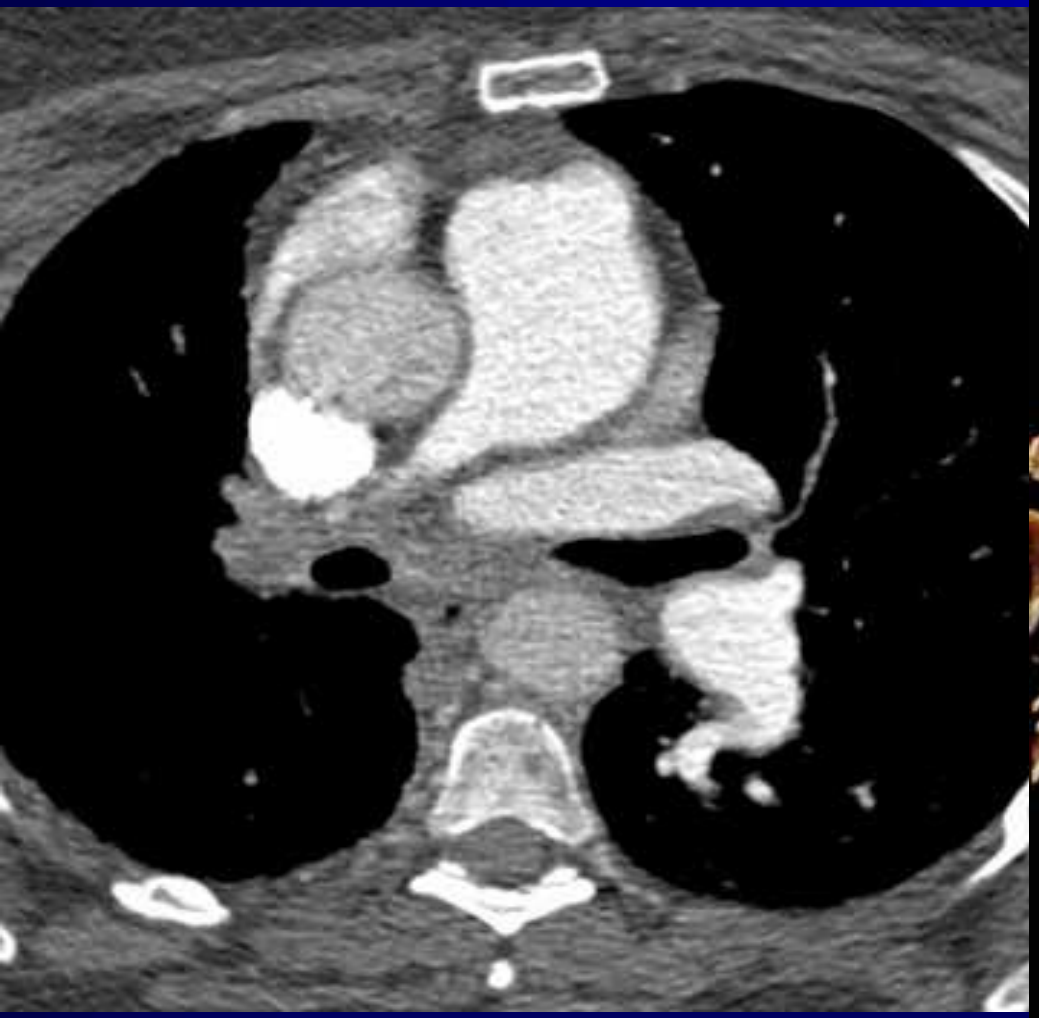


« OBSTRUCTIVE » PHT

Angiosarcoma of the pulmonary arteries

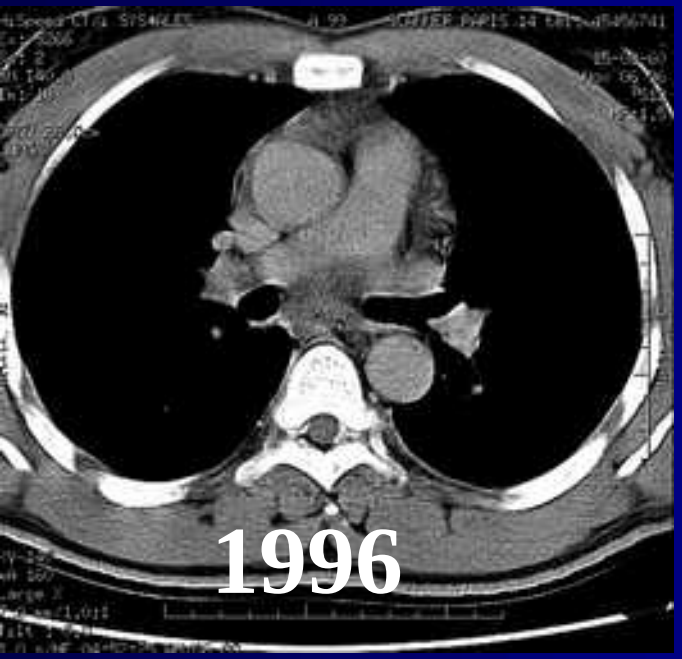
- Progressive dyspnea
- Tumor originating from the main PA
- PET scan +++

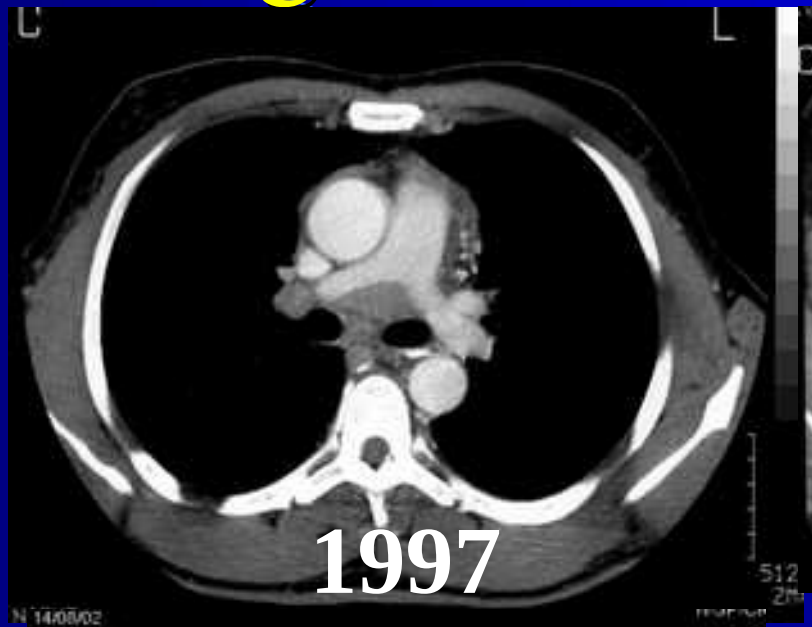
Multisliced CT-scan

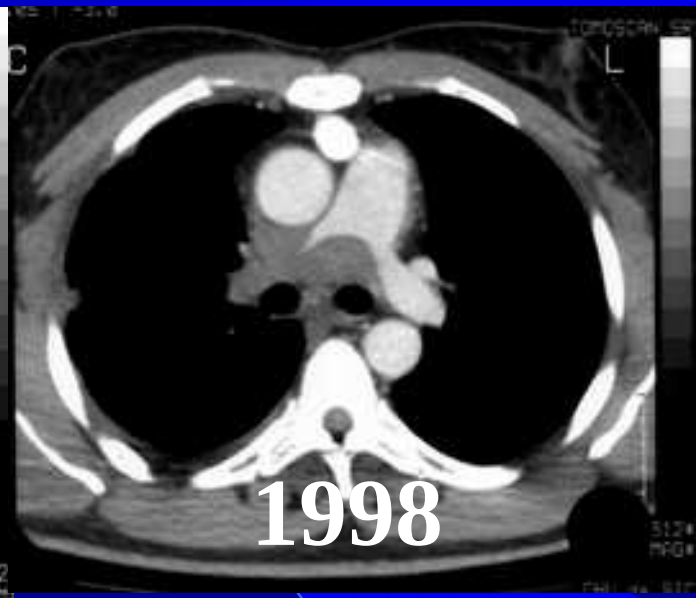


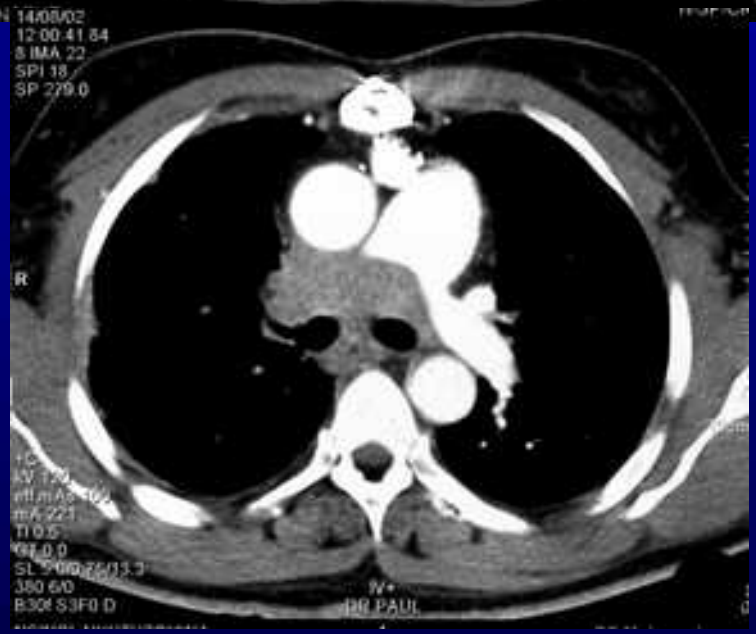
hypoplasic right PA, hypoplasic right lung

Fibrosing mediastinitis



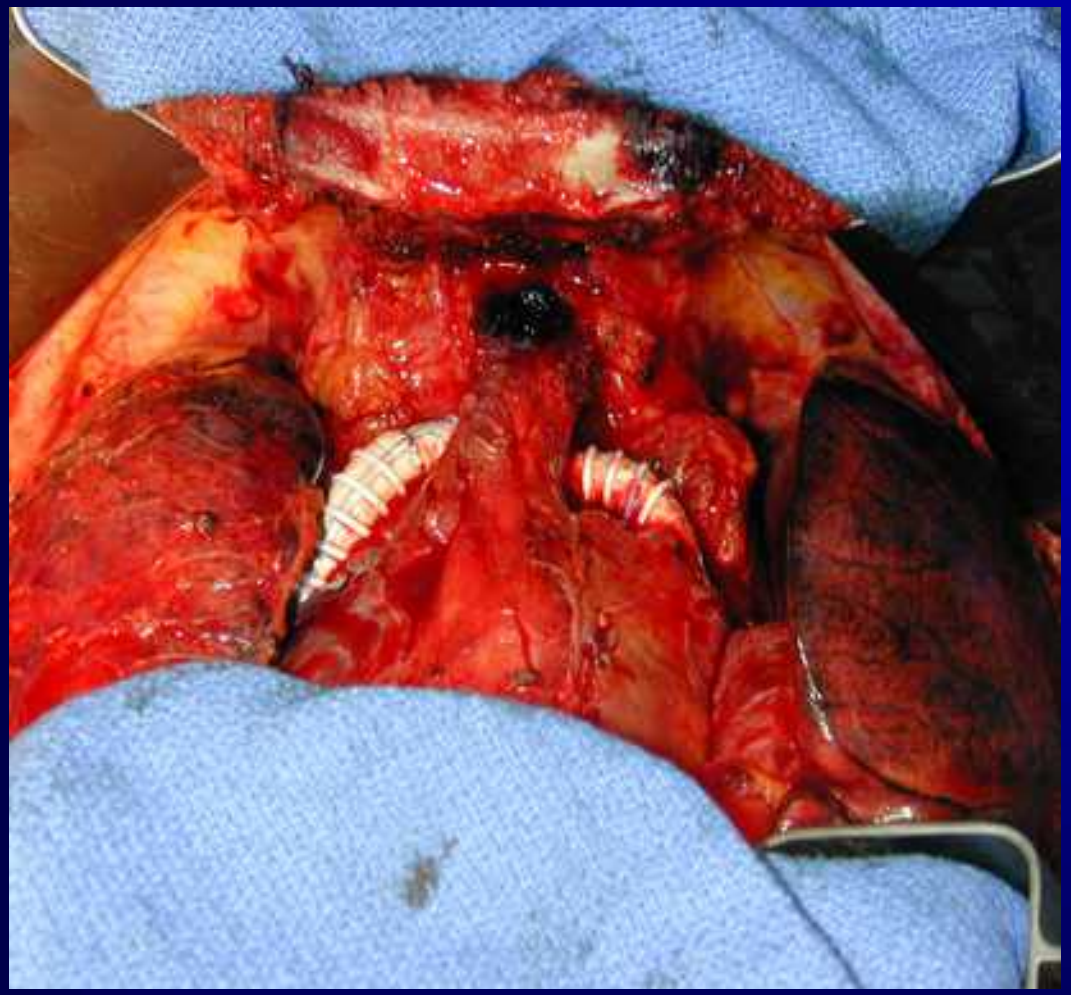


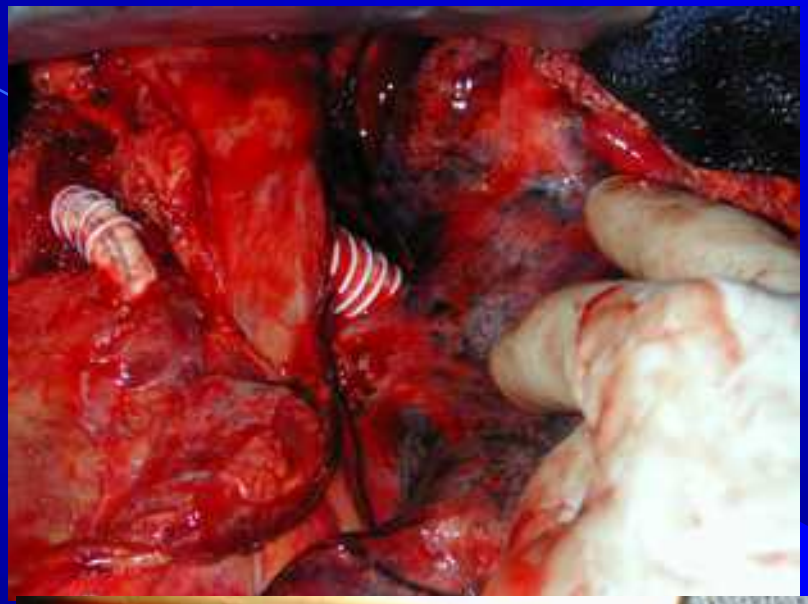


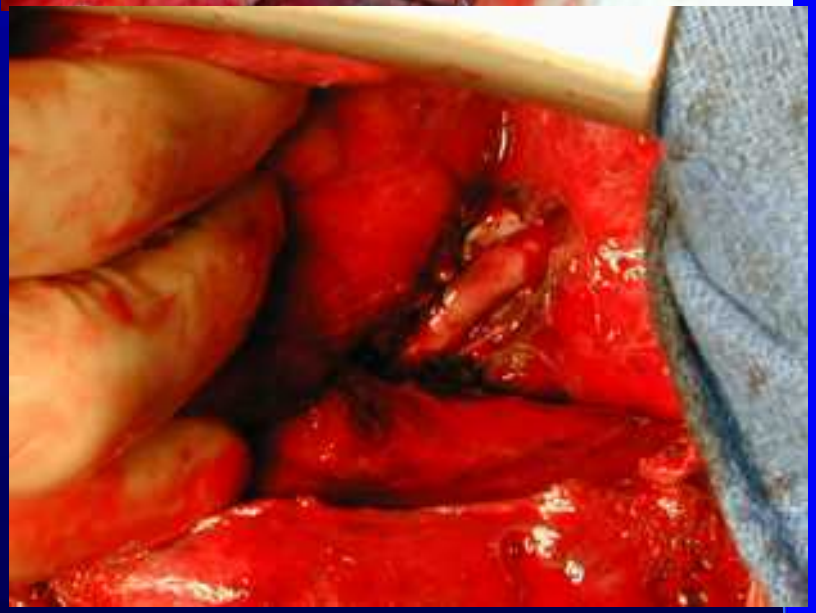


2002…
PHT

2002 : Double PTFE bypass

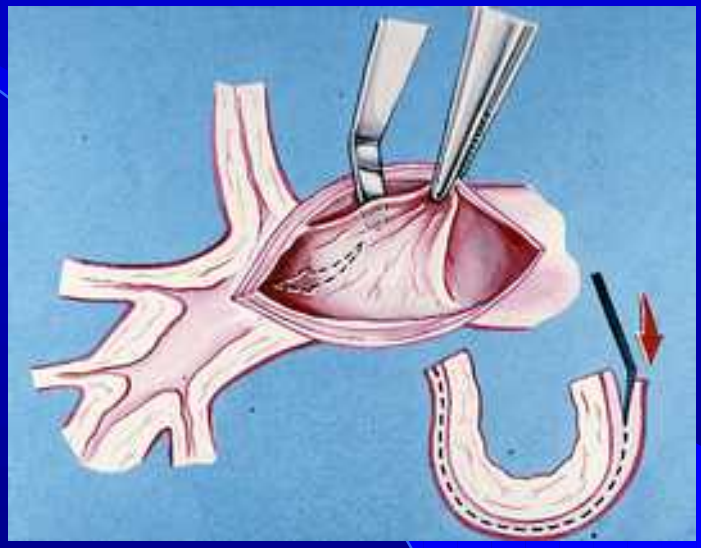


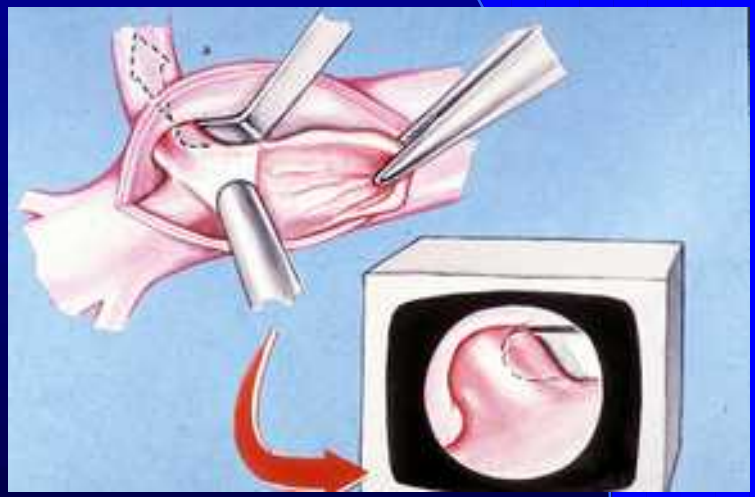




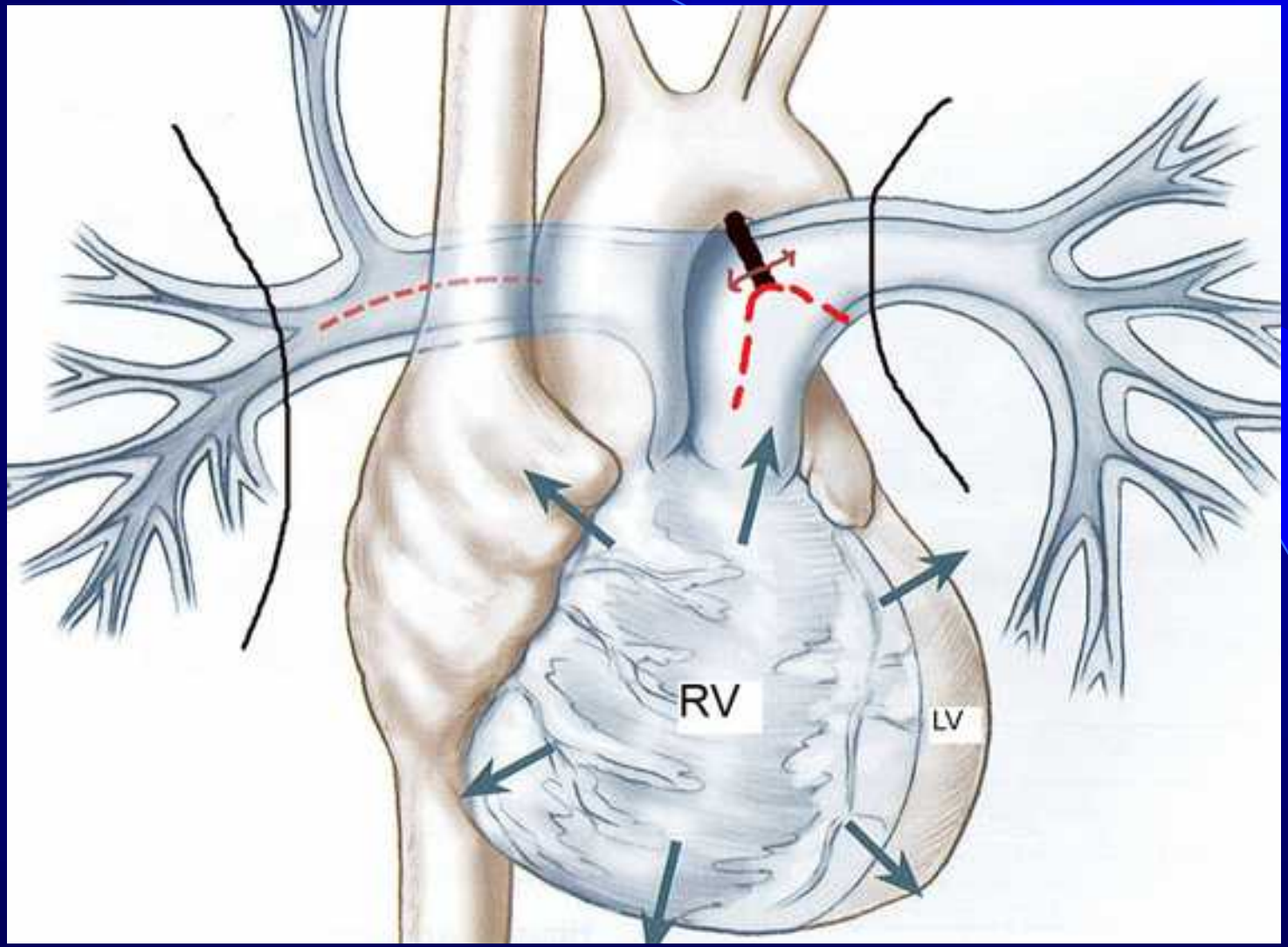
ENDARTERECTOMY

- Proximal arteriotomy
- Identification of the correct plane
  - Proximally
  - Posterior aspect of the PA
- Respect the arteriotomy line
- Eversion endarterectomy





Pulmonary artery incisions



Operative Strategy

- Deep Hypothermia
- Aortic Clampage
- Right Endarterectomy - circulatory arrest
- Reperfusion 15 *min.*
- Left Endarterectomy - circulatory arrest
- Reperfusion- Aortic unclamping
- Rewarming & CPB weaning


RA
10cm

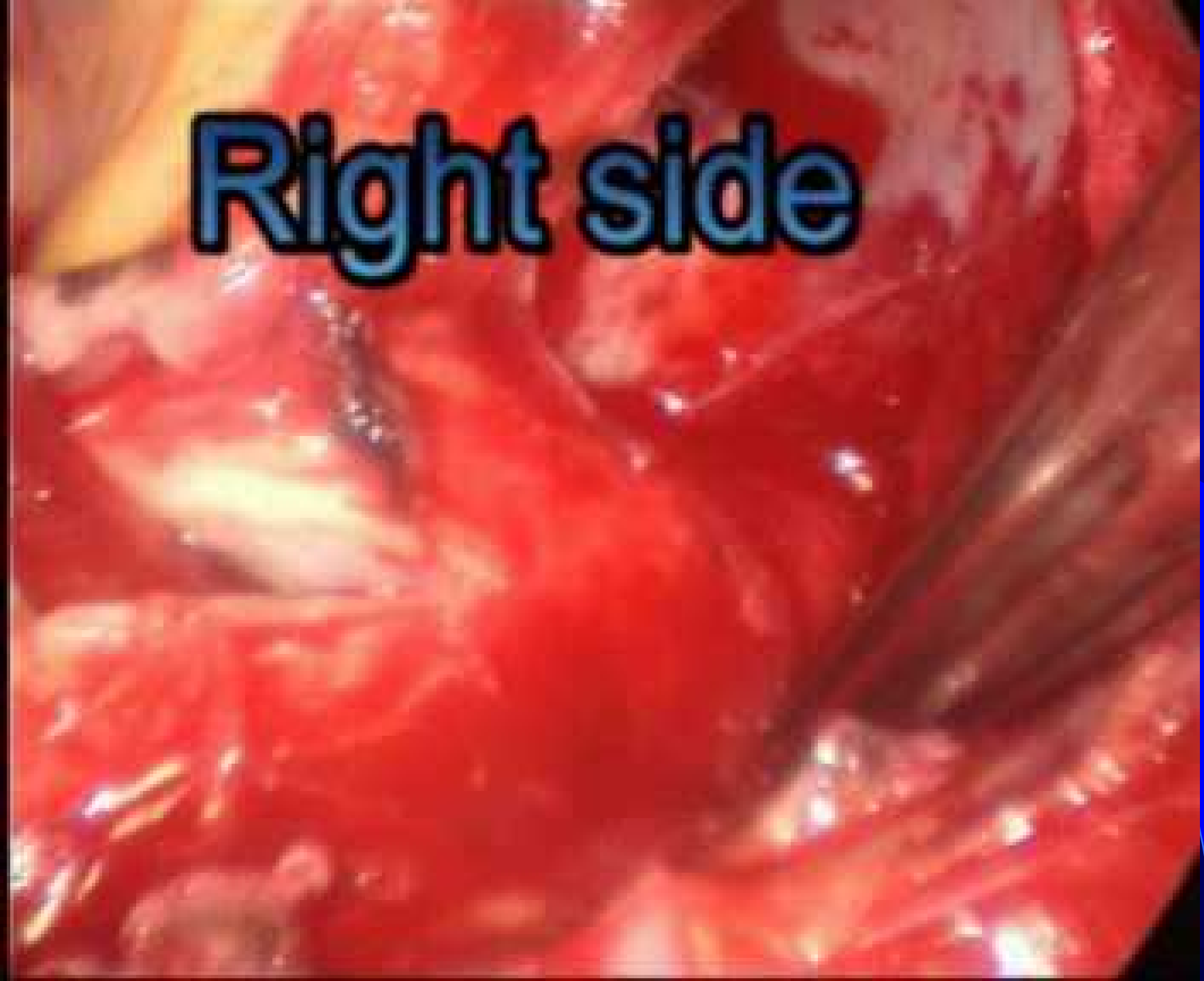

R
10cm
A
+C
kV 120

Endovascular Procedure

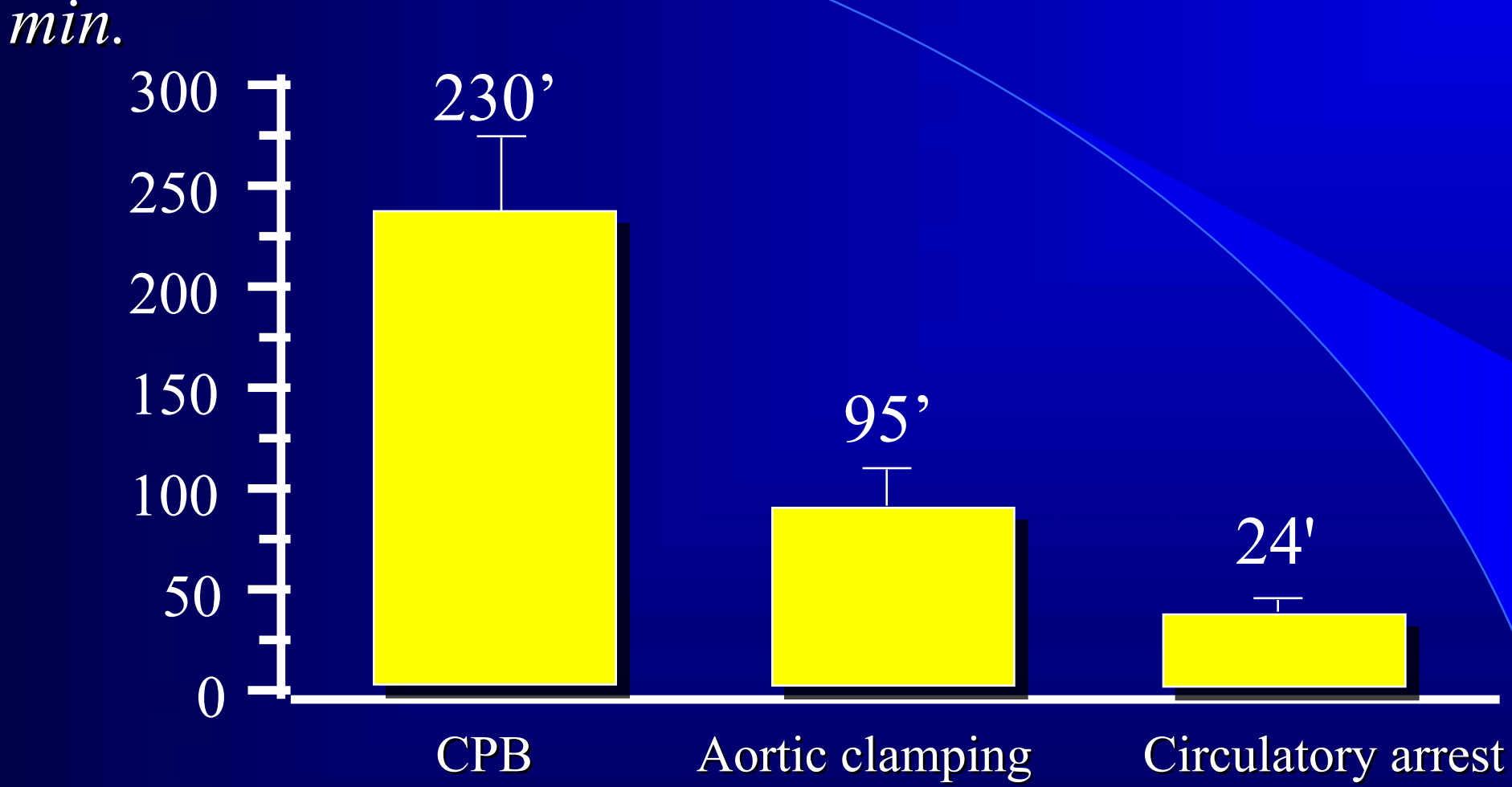
- Proximal initiation of endarterectomy
- Need of distal obstruction relief

→ **Video assistance provides**

- **Light**
- **Distal visualization**
- **View for assistant surgeons**


Right side

Duration of the procedure

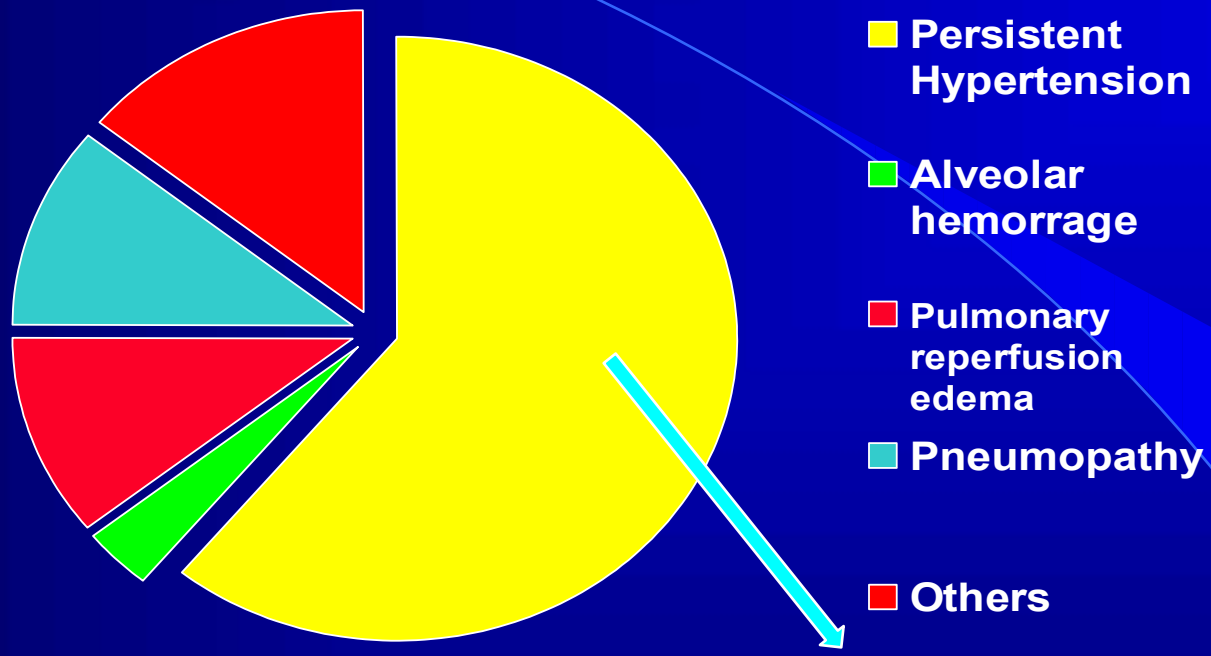


Postoperative complications

- Post-reperfusion pulmonary oedema (40 ECLS)
- Right heart failure secondary to persistently high pulmonary pressure
- Nosocomial pneumonia
- Haemoptysis: 12 bronchial artery embolisations
- Rethrombosis of an endarterectomised area occurs only rarely

Bronchial embolisation

Causes of death

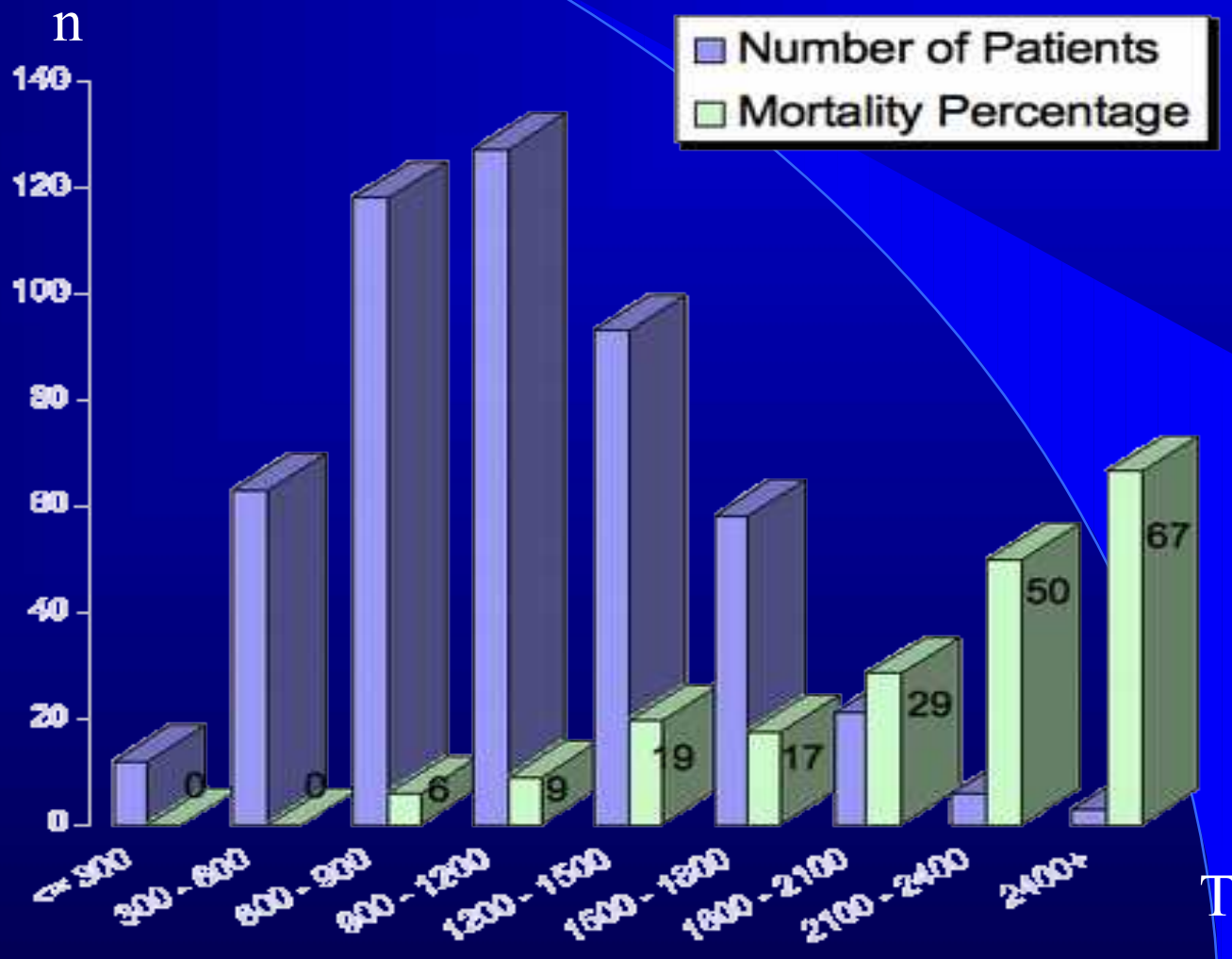


Pulmonary endarterectomy

Mortality increases with preoperative resistance,

p<0,001

(OR : 1,761 95% CI 1,45 – 2,13)

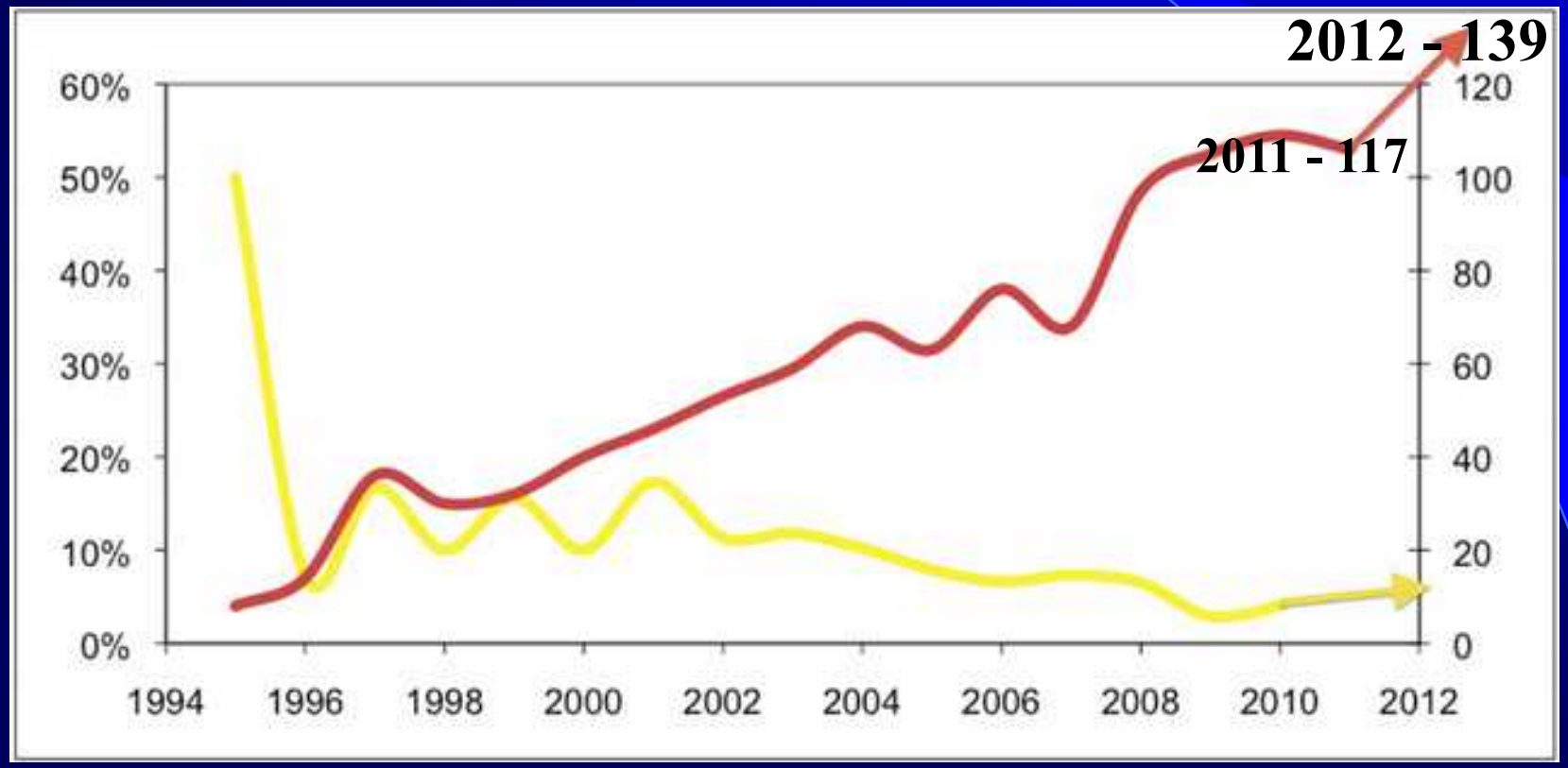


Pulmonary endarterectomy

Postoperative mortality

n = 1350

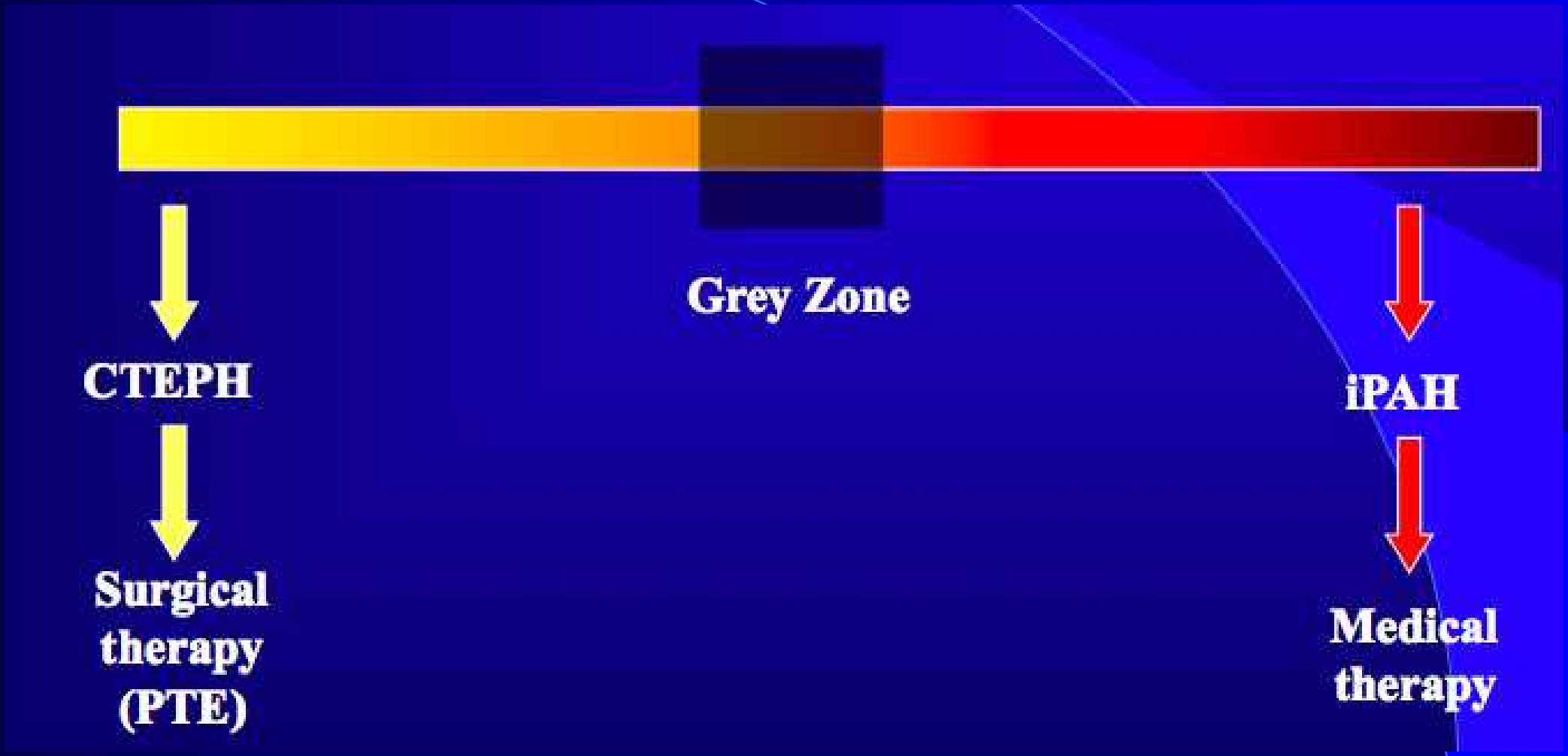
n/year



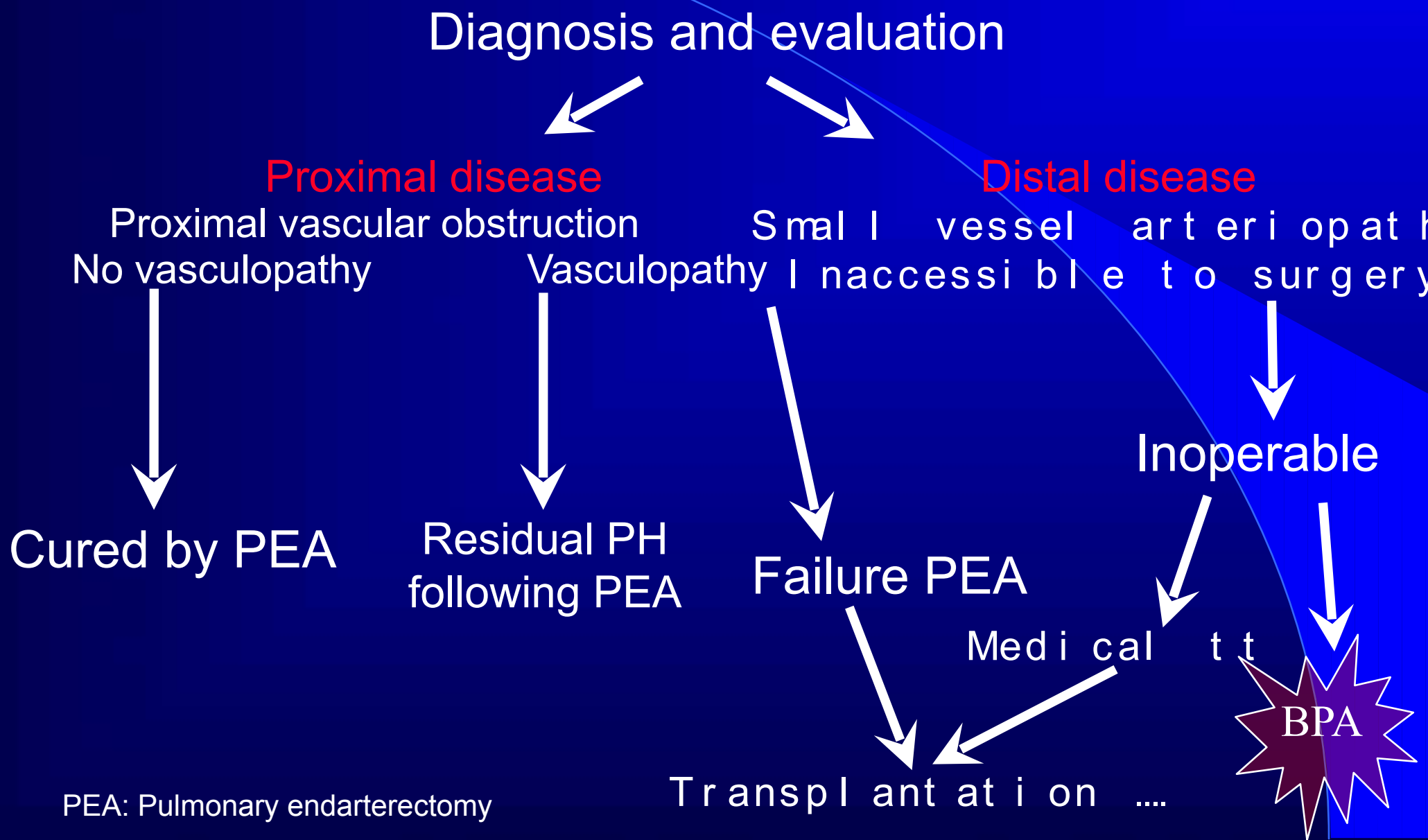
January 1995 to december 2012

Current mortality drop in hemodynamically severe patients

- Better patients selection
  - Proximal disease on imaging
  - Anatomical obstruction correlated with PVR
  - Eliminate patients with PAH + in-situ thrombosis +++
- Better postoperative management
  - Low cardiac output
  - Postoperative use of ECMO
  - Bronchial arteries embolization

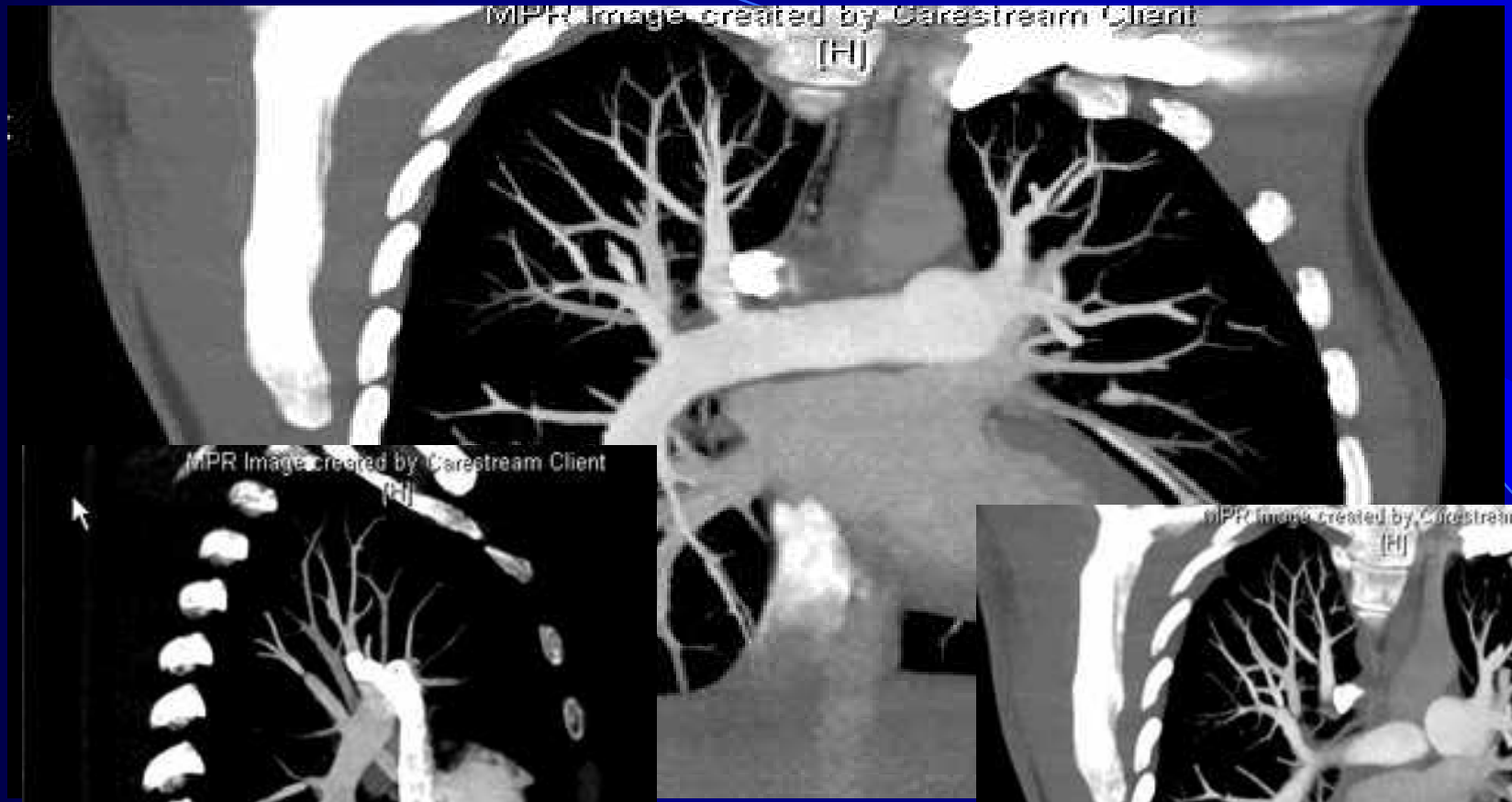

Grey Zone
CTEPH
Surgical therapy (PTE)
iPAH
Medical therapy

A new weapon in the armamentorium : BPA (?)



PEA: Pulmonary endarterectomy

CT Scan


MPR Image created by Carestream Client
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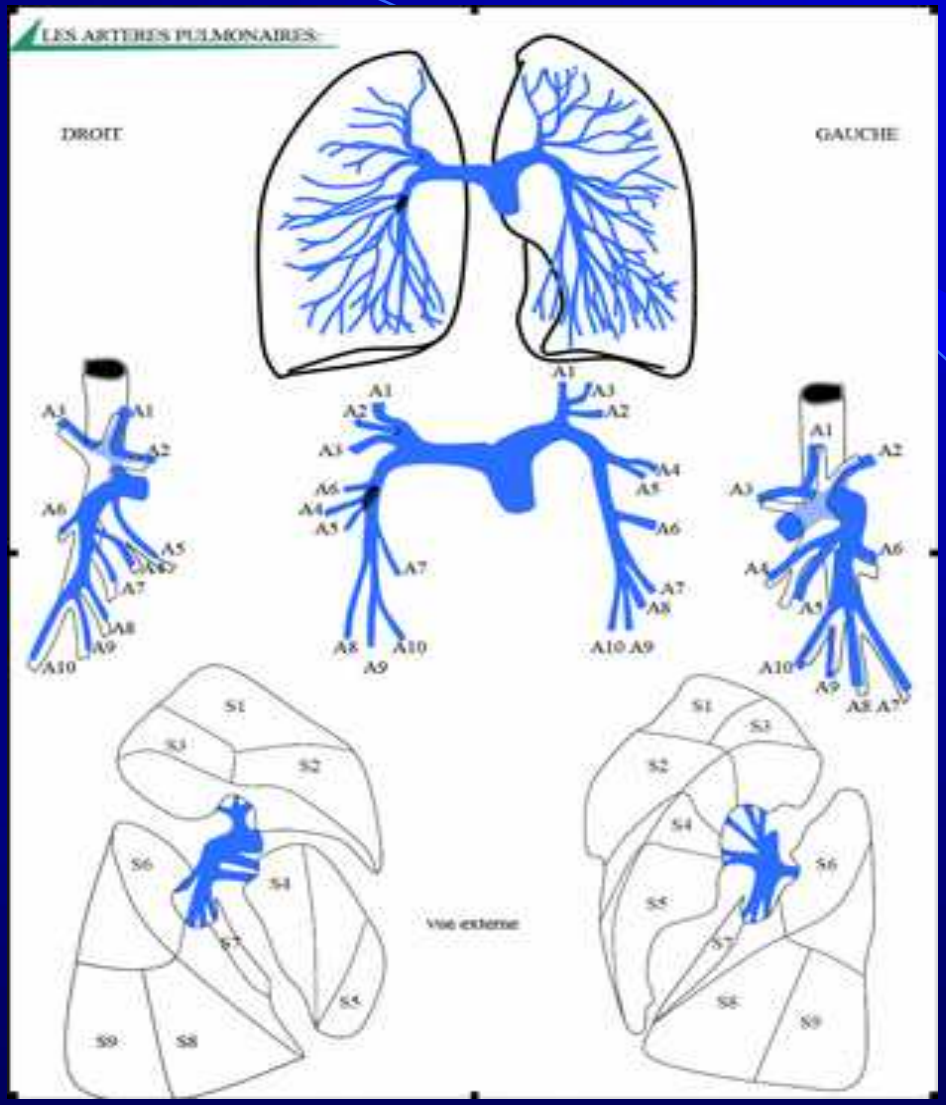
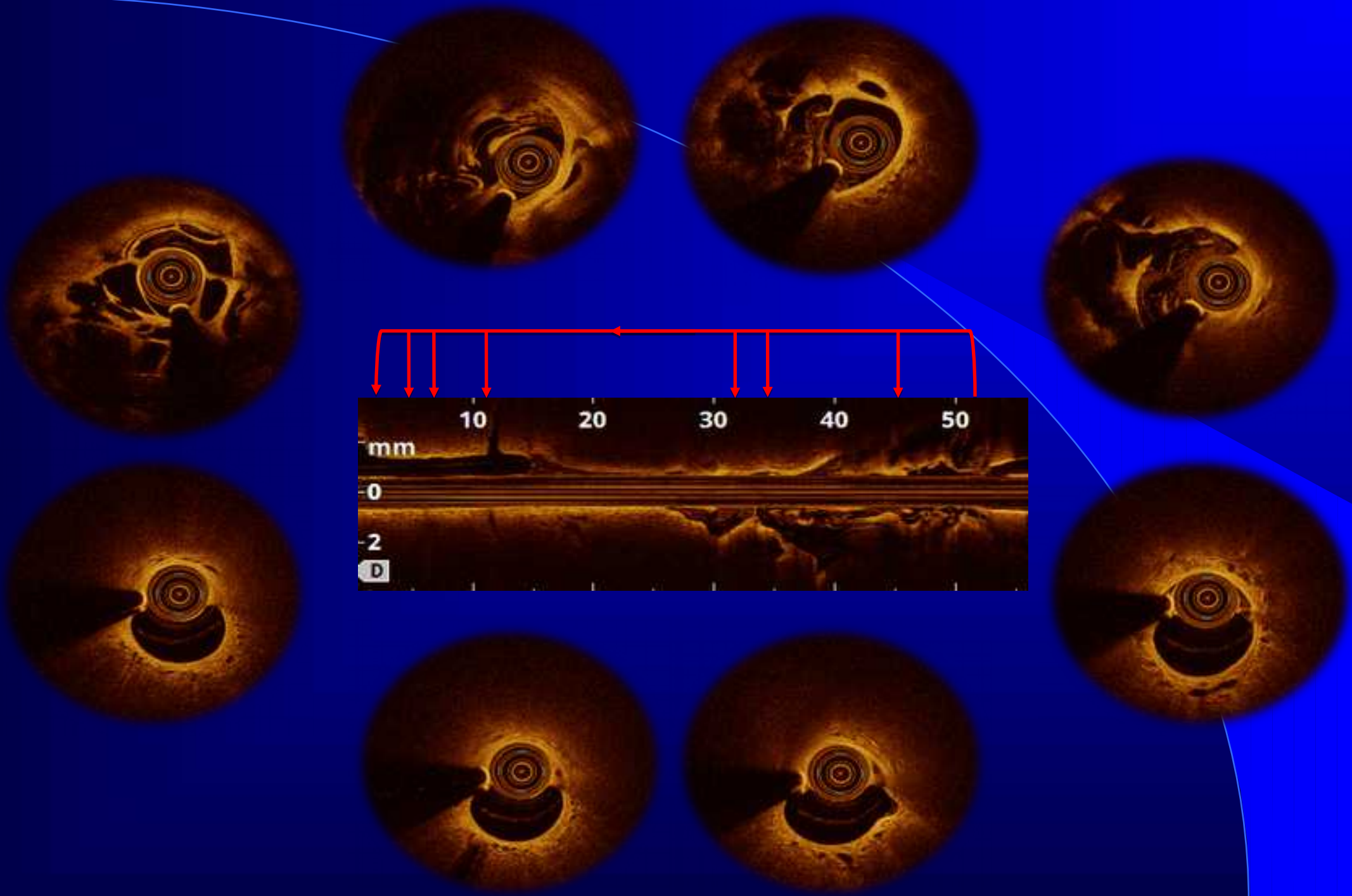
Pulmonary artery segmentation



First session 2/17/2014

- Ballon Angioplasty associated with OCT
- LLL : branches A8 +A9 + A 10


10
20
30
40
50
mm
0
2
D

Conclusion

- PEA is the ideal treatment for postembolic PHT
- Large experience required for:
  - Indication for surgery
  - Surgical procedure
  - Management of postoperative course
- Early operation avoids development of vasculopathy

Conclusion

- Operability is determined by experts in CTEPH
- Large indications for PEA (70%)
- Nuclear scan
- True history of PE
- Correlation between hemodynamics and imaging
- Mortality rate of PEA is less than 3%
- Bridge therapy is very debatable

ECMO for PRIMARY GRAFT FAILURE

- Double Lung Transplantation 5
  - for IPF (n=2), CTEPH (n=1), bronchiectasy (n=1), PH (n=1)
  - 4 postoperative death
  - 1 patient alive after a mean follow up of 2 months

- Heart-lung Transplantation 11
  - for PAH (n=7), Eisenmenger (n=2), CTEPH (n=2)
  - 4 postoperative deaths
  - 7 patients alive after a mean follow up of 10 months

ECMO AS A BRIDGE TO HEART-LUNG Tx (n = 4)

- Urgent procedures
- ECMO needed after
  - TEP failure (n=2)
  - Endstage PH (n=2, procedure under local anesthesia)
- 3 patient alive

Marie LANNELONGUE & Antoine BECLERE
PEA for International Patients

- All inclusive — **50 000 Euros**
- Work-up surgical and hospital fees
- Average hospital stay — 18 days
- Low cost local accomodation for family members — Non included

URGENT PROCEDURES

- Double Lung Transplantation 12
  - for PPH (n=9), CTEPH (n=1), IPF (n=2)
  - 4 postoperative death
  - 8 patients alive after a mean follow up of 6 months

- Heart-lung Transplantation 16
  - for PPH (n=8), CTEPH (n=1), IPF (n=1), pulmonary veins stenosis (n=1)
  - 5 postoperative death
  - 11 patients alive after a mean follow up of 10 months

Factors predicting efficiency

- History of acute PE or DVT
- Honey moon period (period between acute PE and the development of severe PH)
- Proximal disease on angiography
- Developed systemic circulation
- Anatomical obstruction correlating with PVR +++