# Thromboembolic Pulmonary Hypertension: Diagnosis and Management

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# Background - classification of PH and surgical options

**1. Pulmonary Arterial Hypertension**
- Idiopathic PAH (IPAH)
- Familial PAH (FPAH)
- related to
  connective tissue diseases
  HIV infection
  portal hypertension
  anorexigens
  congenital heart diseases
- PVOD/PCH

**2. PH with left heart dysfunction**
- atrial or ventricular
- valvular

**3. PH with hypoxia**
- COPD
- interstitial lung disease
- sleep disordered breathing
- developmental abnormalities

**4. PH due to chronic thrombotic and/or embolic disease**

**5. Miscellaneous**
- sarcoid
- histiocytosis X

# Background

•Improvement following acute pulmonary embolism is usually sufficient to restore normal pulmonary hemodynamics, gas exchange, and exercise tolerance.

•A minority of patients develop chronic thromboembolic pulmonary hypertension (CTEPH) following a single or recurrent episode of acute pulmonary embolism)

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# Natural History of CTEPH

- Honeymoon period after acute PE
- Usually present in their 40s
- Later presents with dyspnea, hypoxemia & RV dysfunction
- Death usually due to RV failure

*Riedel M, Stanek V, Widimsky J, et al. Longterm follow-up of patients with pulmonary thromboembolism. Late prognosis and evolution of hemodynamic and respiratory data. Chest 1982;81:151–8.*

# Epidemiology of

## Chronic Thrombo-Embolic PH (CTEPH)

Prospective study of acute PE survivors:

- N=305
- Clot lysis is the norm
- Incidence of symptomatic CTEPH
  - 3.1% at 1 year
  - 3.8% at 2 years

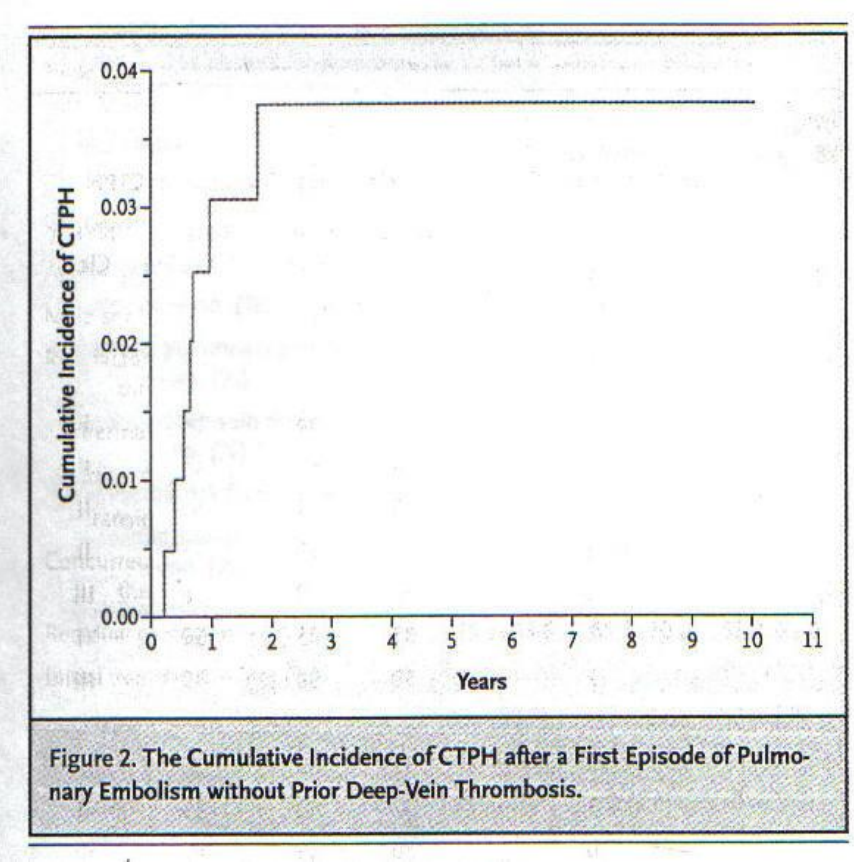



Figure 2. The Cumulative Incidence of CTPH after a First Episode of Pulmonary Embolism without Prior Deep-Vein Thrombosis.

50% of patients with CTEPH have no history of DVT/PE.

5% have prothrombotic tendency.

Pengo et al. NEJM. 2004;350:2257-2264

# Incidence

- 0.5% to 3.8% of pts after an acute PE & in upto 10% of those with a history of recurrent PE will develop CTEPH.
- A prospective follow-up study of 78 survivors of acute pulmonary embolism, Four patients (5.1%) developed definite CTEPH, and 3 of these subsequently underwent successful PEA.

  *(Ribeiro A, Lindmarker P, Johnsson H, Juhlin-Dannfelt A, Jorfeldt L. Pulmonary embolism: one-year follow-up with echocardiography Doppler and five-year survival analysis. Circulation. 1999;99:1325–1330.)*
- The only identifiable risk factors for persistent pulmonary hypertension were an age 70 years and a systolic pulmonary artery pressure 50 mm Hg at the initial presentation.
- In another prospective follow-up study of 223 patients who presented with acute pulmonary embolism, the incidence of symptomatic CTEPH was 3.1% at 1 year and 3.8% at 2 years (Pengo V, Lensing AW, Prins MH, Marchiori A, Davidson BL, Tiozzo F,Albanese P, Biasiolo A, Pegoraro C, Iliceto S, Prandoni P. Incidence of chronic thromboembolic pulmonary hypertension after pulmonary embolism. *N Engl J Med.* 2004;350:2257-2264. )

## Progression of CTEPH

Acute or recurrent PTE in pulmonary arteries

↓

Organisation these thrombi

↓

Occurence in situ thrombus due to slow blood flow in obstructed pulmonary arteries

↓

Occurence of arteritis in non obstructed small distal pulmonary arteries(remodelling)

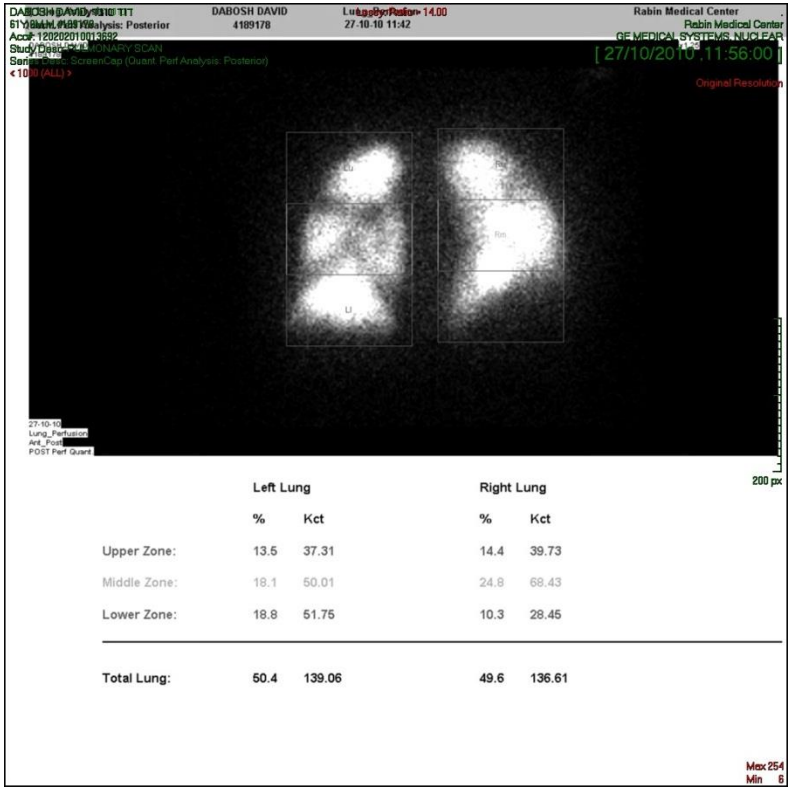
↓

Increased PVR, pulmonary hypertension

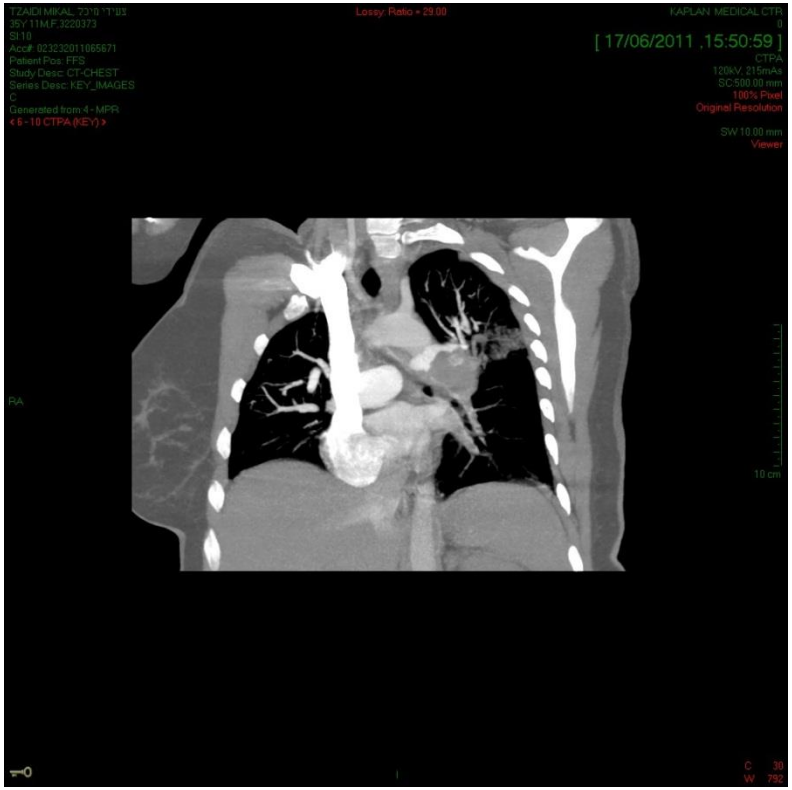
↓

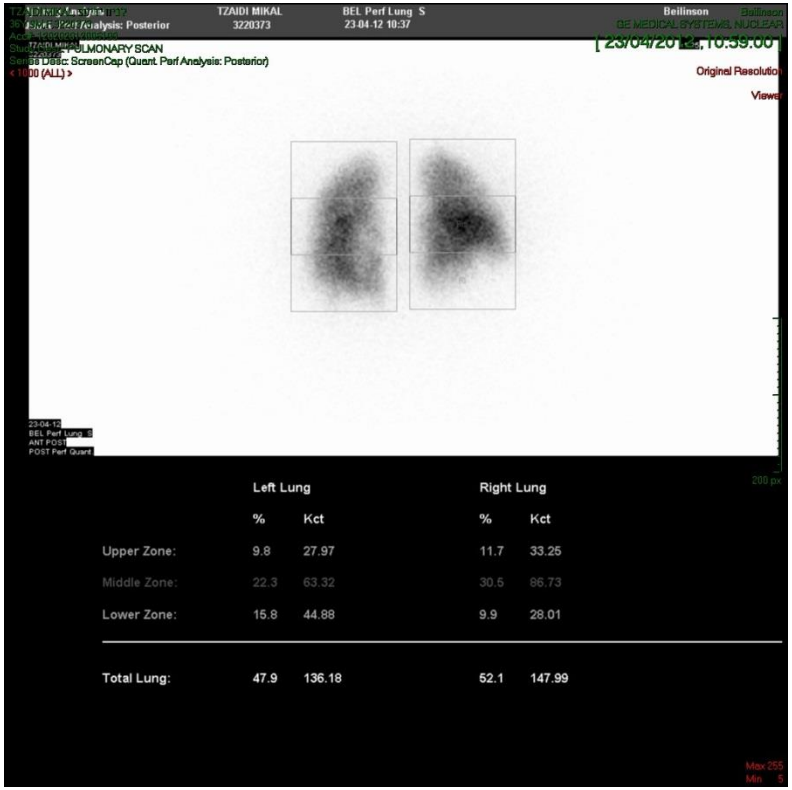
CTEPH

# Perfusion scan

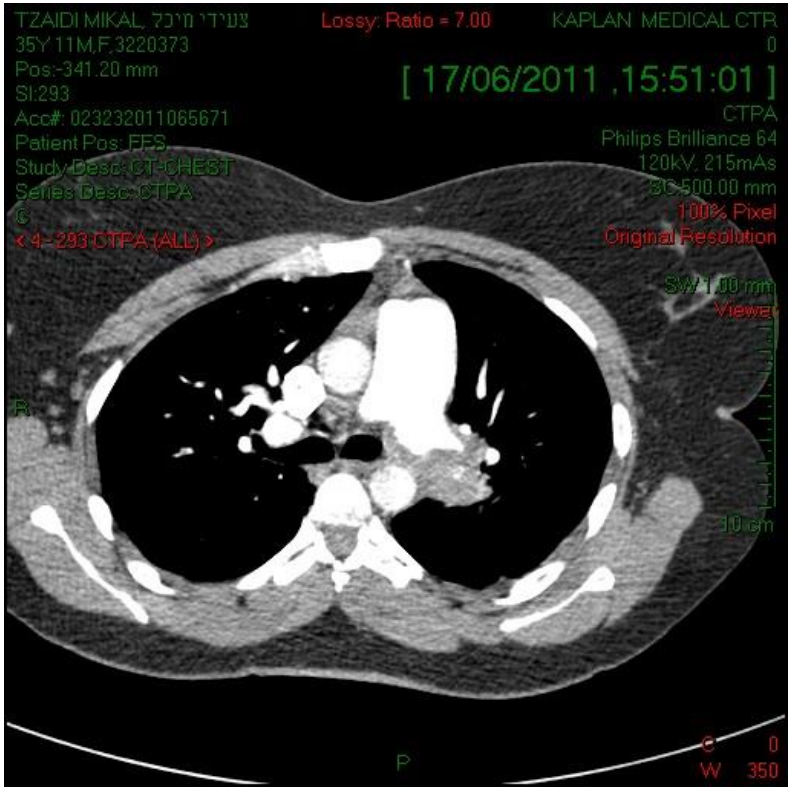


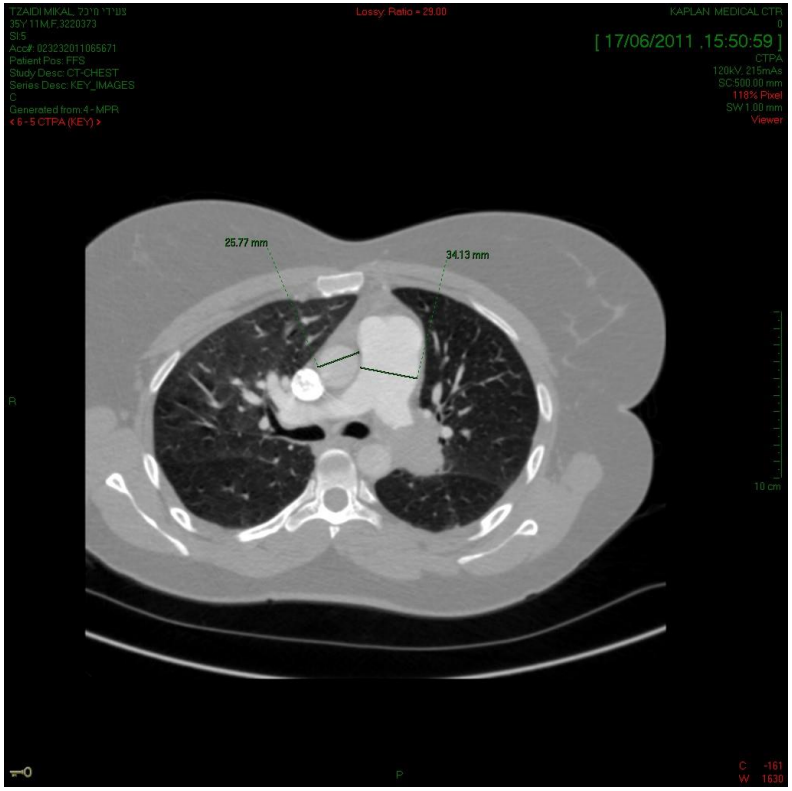

- Ventilation-perfusion (V/Q) lung scanning represents the initial imaging procedure of choice
- At least one (usually several) segmental or larger mismatched ventilation-perfusion defects.




| | Left Lung | | Right Lung | |
|---|---|---|---|---|
| | % | Kct | % | Kct |
| Upper Zone: | 9.8 | 27.97 | 11.7 | 33.25 |
| Middle Zone: | 22.3 | 63.32 | 30.5 | 86.73 |
| Lower Zone: | 15.8 | 44.88 | 9.9 | 28.01 |
| Total Lung: | 47.9 | 136.18 | 52.1 | 147.99 |


TZAIDI MIKAL, צעירדי מיכל
35Y 11M,F,3220373
Pos:-341.20 mm
SI:293
Acc#: 023232011065671
Patient Pos: FFS
Study Desc: CT-CHEST
Series Desc: CTPA
C
< 4 - 293 CTPA (ALL) >
Lossy: Ratio = 7.00
KAPLAN MEDICAL CTR
0
[ 17/06/2011 ,15:51:01 ]
CTPA
Philips Brilliance 64
120kV, 215mAs
SC:500.00 mm
100% Pixel
Original Resolution
SW:1.00 mm
Viewer
R
10 cm
P
C 0
W 350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Series Desc: KEY_IMAGES
C
Generated from:4 - MPR
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KAPLAN MEDICAL CTR
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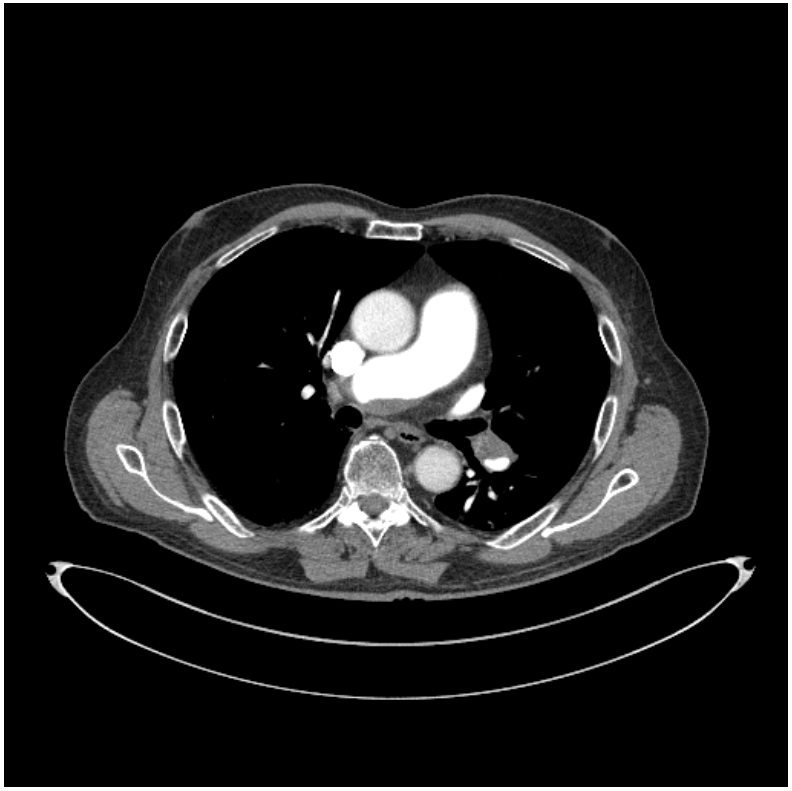
CTPA
120kV, 215mAs
SC:500.00 mm
118% Pixel
SW:1.00 mm
Viewer
25.77 mm
34.13 mm
R
10 cm
P
C -161
W 1630

# CT-angio

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CTPA
120kV, 215mAs
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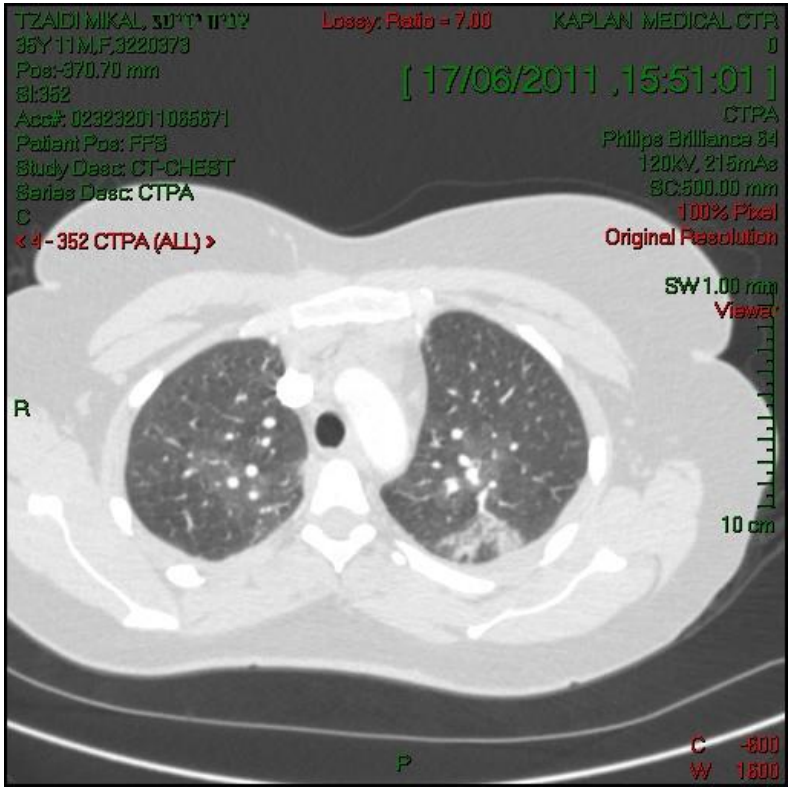
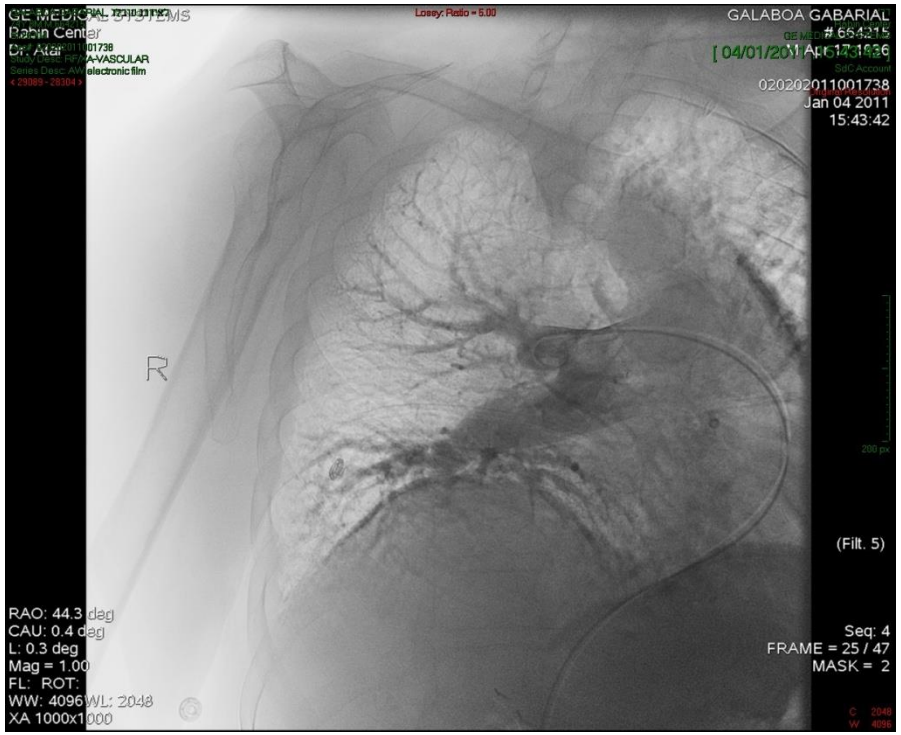
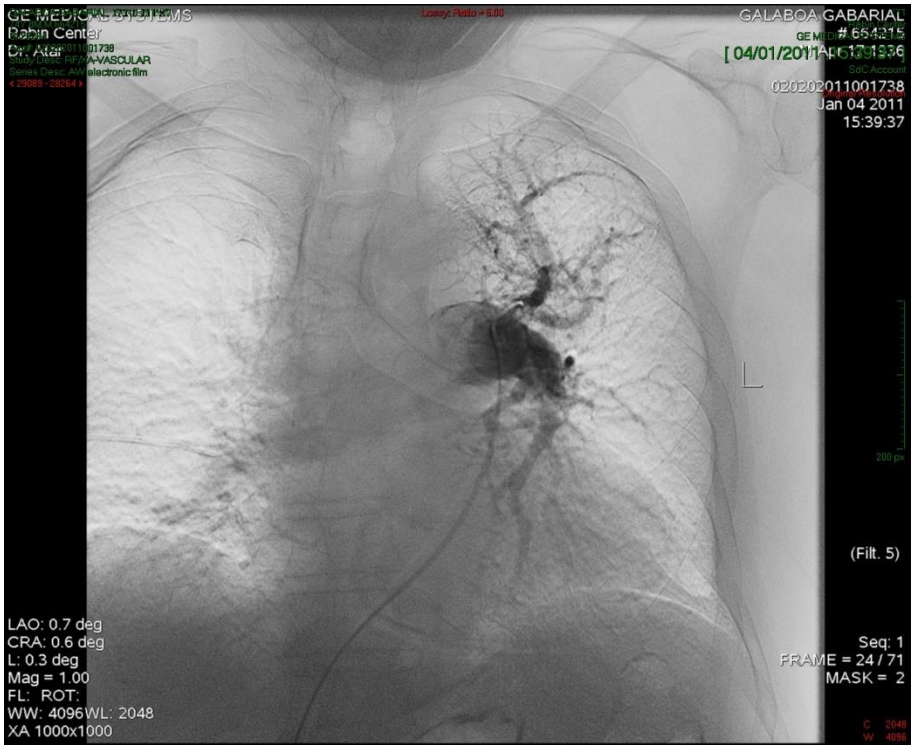
SW 1.00 mm
Viewer
Generated from 4 - MPR
< 6 - 1 CTPA (KEY) >
A
10 cm


TZAIDI MIKAL, צדיכי מיכל
35Y 11M,F,3220373
Pos:-370.70 mm
SI:352
Acc#: 023232011065671
Patient Pos: FFS
Study Desc: CT-CHEST
Series Desc: CTPA
C
< 4 - 352 CTPA (ALL) >
Lossy: Ratio = 7.00
KAPLAN MEDICAL CTR
[ 17/06/2011 ,15:51:01 ]
CTPA
Philips Brilliance 64
120kV, 215mAs
SC:500.00 mm
100% Pixel
Original Resolution
SW 1.00 mm
Viewer
R
10 cm
P
C -600
W 1600

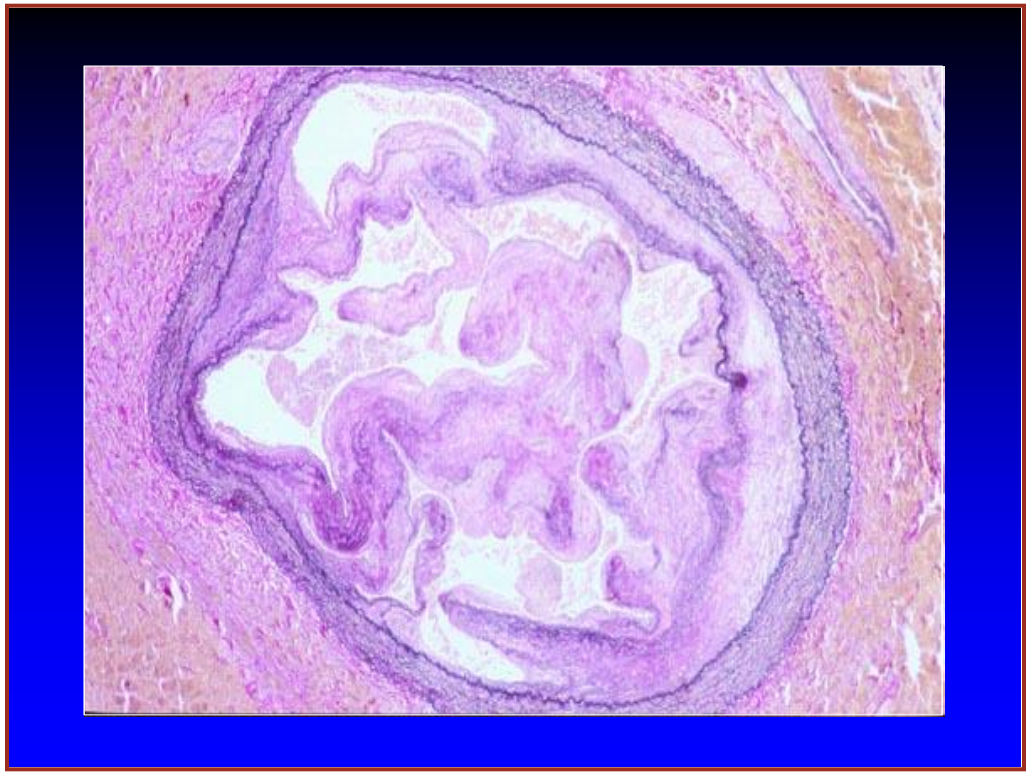
# Pulmonary angiography



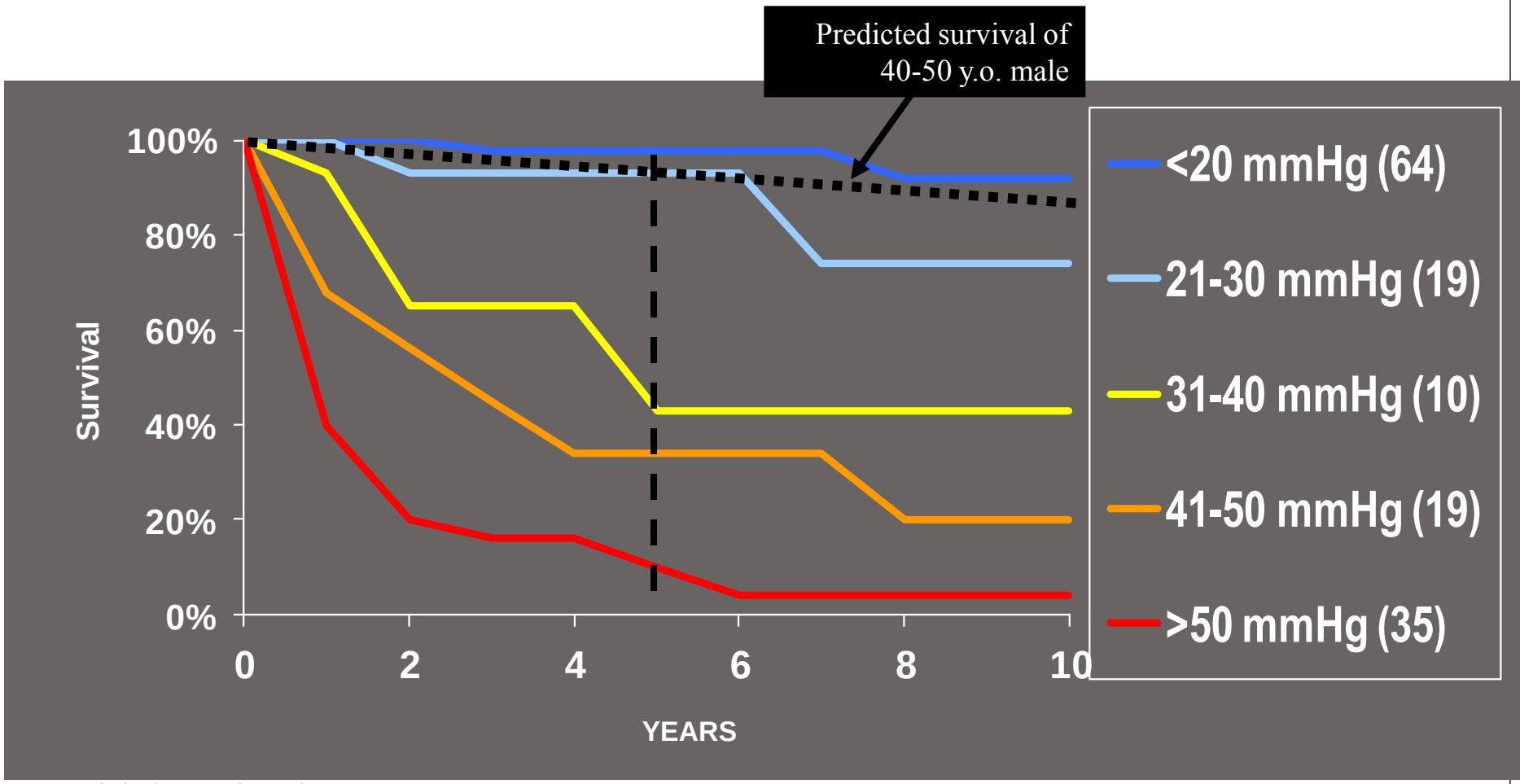
# Pulmonary angiography

# Histology



- Not blood clot!
- Fibrous organisation incorporated in vessel wall.
- Combination of obstruction and remodelling of small vessels.

# Prognosis of CTEPH *Stratified by mean PAP (n=147)*




Reidel et al. Chest. 1982;81:151-158

# PULMONARY THROMBOENDARTERECTOMY

## History

- MOSER, 1970: FIRST SUCCESSFUL OPERATION USING CARDIOPULMONARY BYPASS.
- BRAUNWALD, 1970: COMMENCED A PTE PROGRAM AT THE UNIVERSITY OF CALIFORNIA SAN DIEGO (UCSD).
- SABISTON, 1977: ANALYSIS OF 18 OPERATIONS REPORTED IN THE WORLD LITERATURE.
- CABROL 1978, DAILY 1980, DOR 1981, UTLEY 1982.
- CHITWOOD, 1984: WORLD LITERATURE REVIEWED (85 CASES, 22% MORTALITY).
- WINKLER, 1990: PERFUSION TECHNIQUE OF PROFOUND HYPOTERMIA AND CIRCULATORY ARREST FOR PTE.
- JAMIESON (UCSD), 2008: MORE THAN 2500 CASE.

**Pulmonary Endarterectomy India – Affordable Cost And World Class In India**

- Pulmonary thromboendarterectomy is considered in symptomatic patients who have hemodynamic or ventilatory impairment at rest or with exercise.
- The mean pulmonary vascular resistance in patients undergoing surgery is 800 to 1000 dyn·sec·cm.
- Thromboendarterectomy is also considered in patients who have normal or nearly normal pulmonary hemodynamics at rest but in whom marked pulmonary hypertension develops during exercise.

# Pulmonary thromboendarterectomy

- The most effective therapy –

- Pulmonary thromboendarterectomy.

*(J Am Coll Cardiol 2009;54:Suppl:S67-S77.)*

- Improvement in hemodynamics after pulmonary thrombo-endarterectomy causes

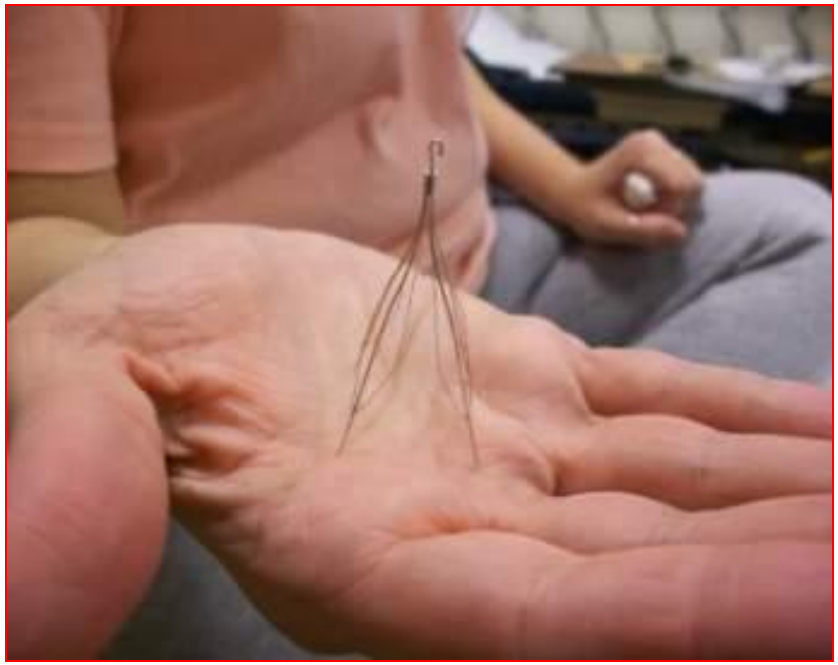
  - Reverse right ventricular remodeling

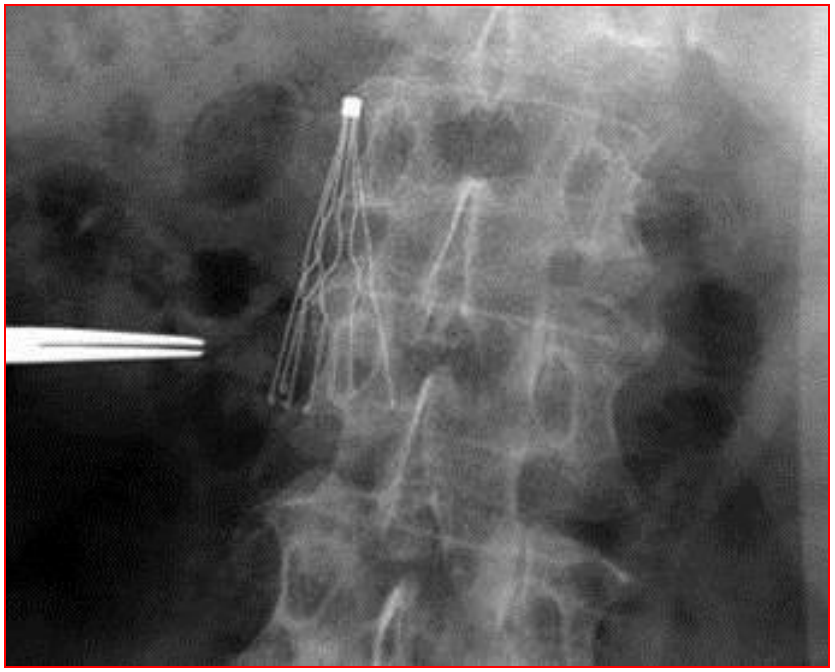
- The beneficial effect usually persists, unless small-vessel arteriopathy or recurrent pulmonary embolism develops.

*(Long-term outcome after pulmonary endarterectomy. Am J Respir Crit Care Med 2008;178:419-24.)*

# PULMONARY THROMBOENDOARTERECTOMY

## *CAVAL FILTER*



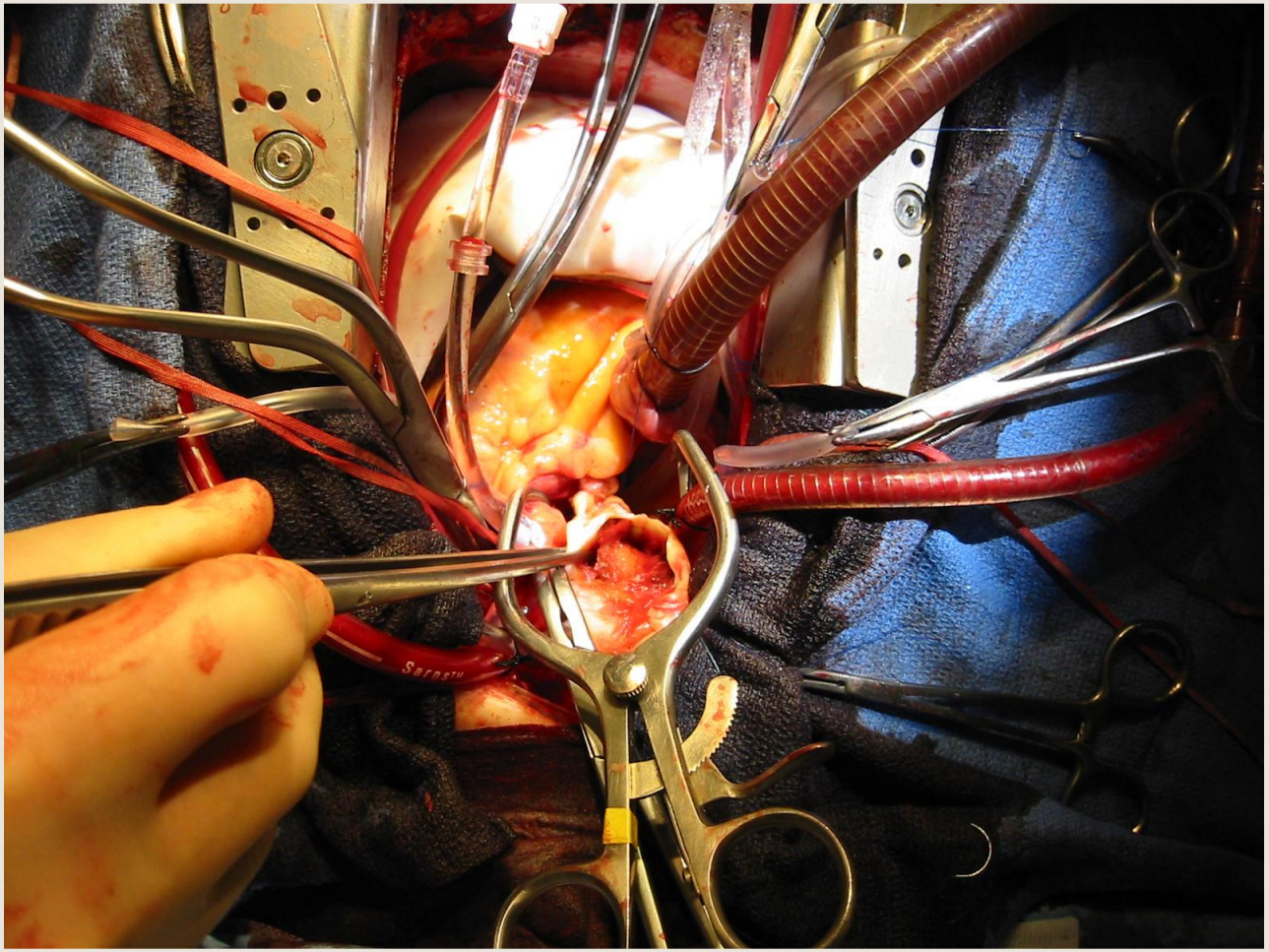


CAVAL FILTER (GREENFIELD FILTER) ROUTINELY PLACED BEFORE THE OPERATION

# Indicators of increased risk

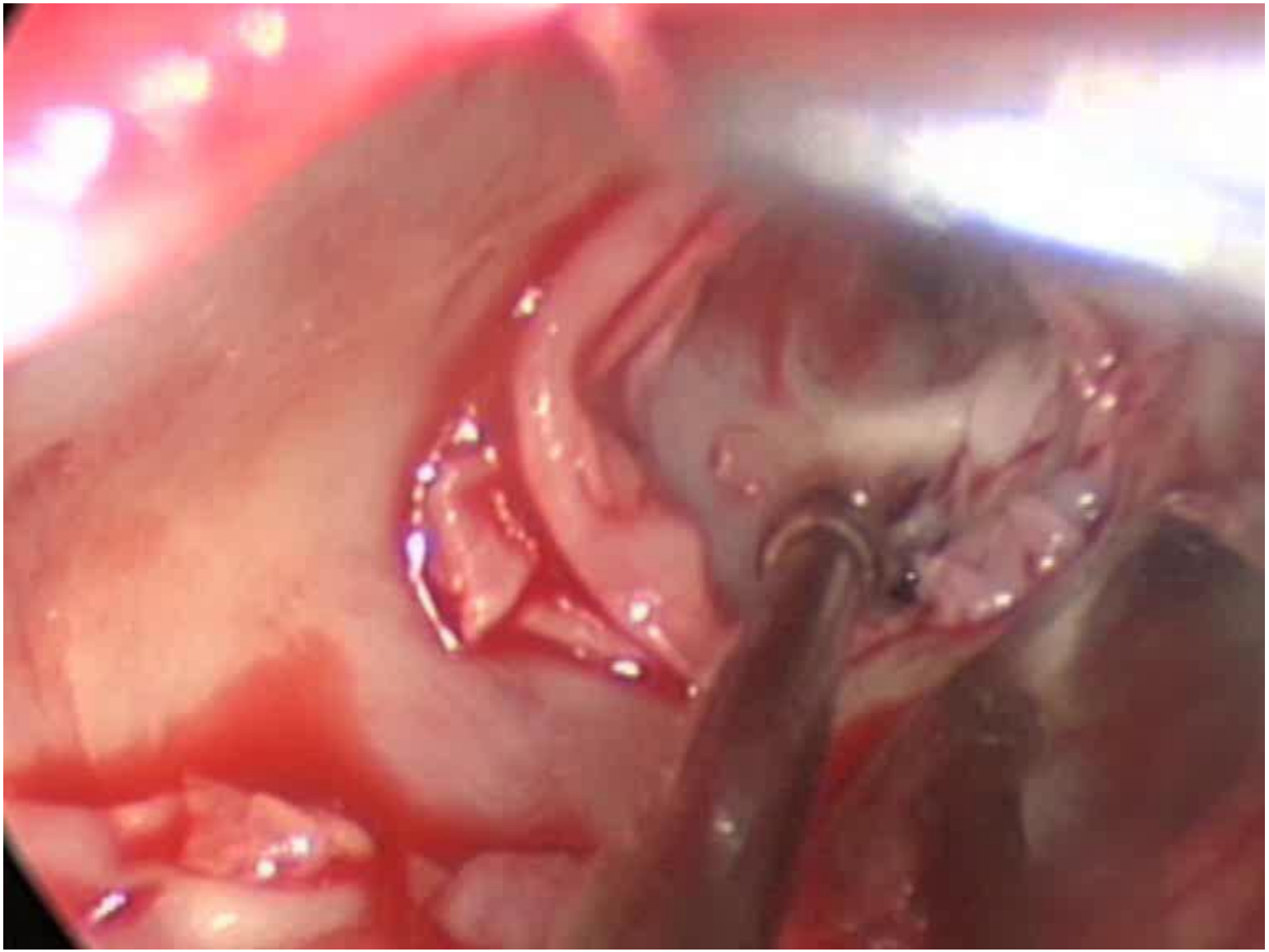
- Distal disease, type 3 in Jamieson classification.
- Increasing PA pressure and PVR, especially when out of proportion to disease on imaging.
- PVR > 1200 dynes.
- Chronicity of disease, small vessel disease.
- Complete occlusion.
- Lack of webs on imaging.
- Comorbidity – COPD.

# Surgical technique: not thrombectomy –but endarterctomy



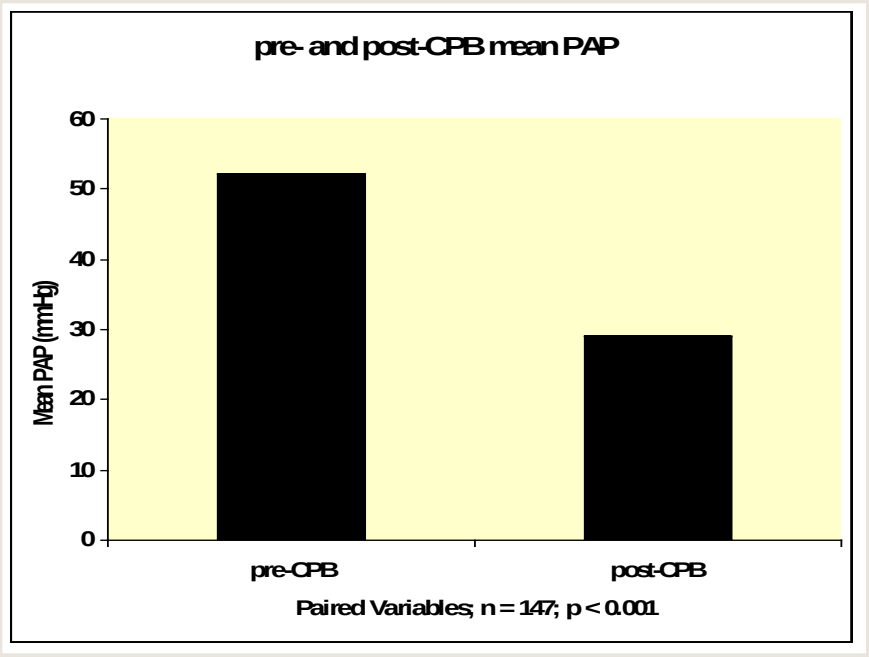
- Pulmonary endarterectomy is performed during circulatory arrest, removing obstructive material from each pulmonary artery, and its lobar and segmental branches, (20–30 branches in total), and is the only way to reduce pulmonary vascular resistance by at least 50%.
- The operation is performed entirely through a median sternotomy and through the pericardium without having to open the pleura or to dissect the pulmonary artery outside the pericardium.

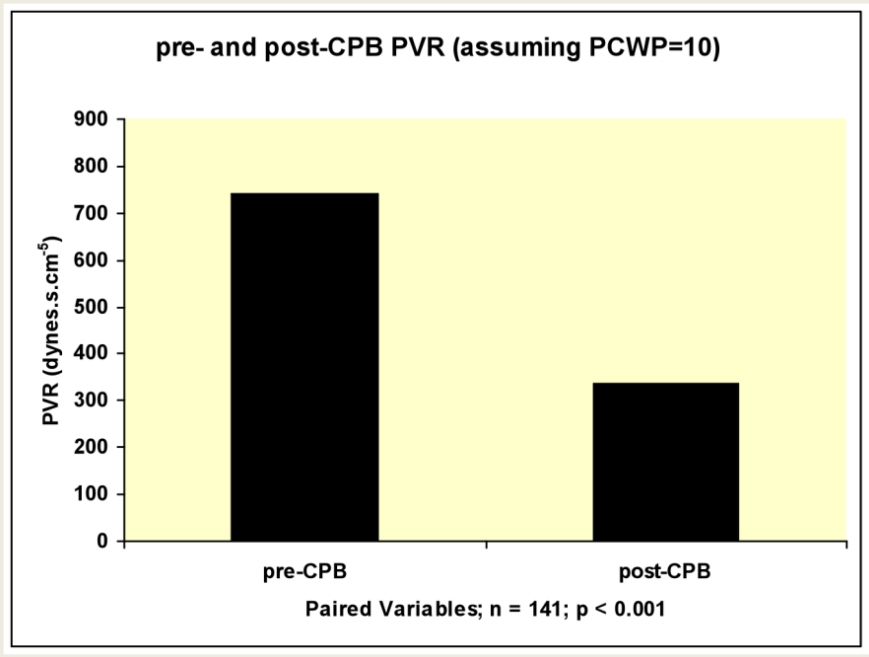
*Circulation. 2006;113:2011-2020*



# The specimen

# Results


pre- and post-CPB mean PAP
Mean PAP (mmHg)
60
50
40
30
20
10
0
pre-CPB
post-CPB
Paired Variables; n = 147; p < 0.001

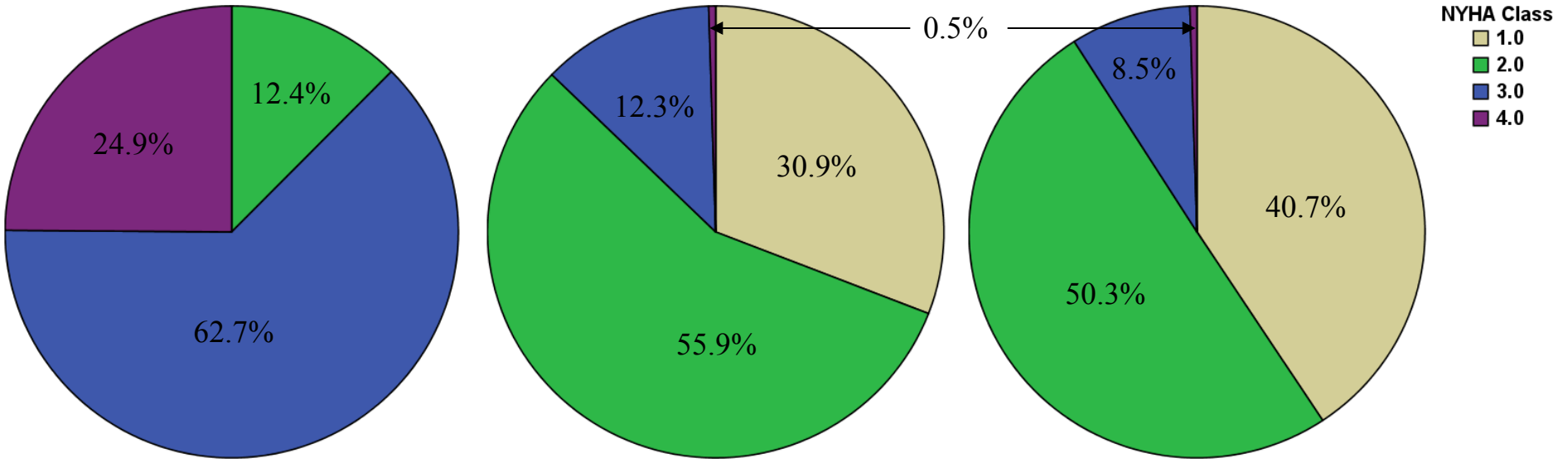

pre- and post-CPB PVR (assuming PCWP=10)
PVR (dynes.s.cm-5)
900
800
700
600
500
400
300
200
100
0
pre-CPB
post-CPB
Paired Variables; n = 141; p < 0.001

# NYHA Class

Pre-op

3 months

12 months




$p < 0.001$ vs preop

$p < 0.001$ vs 3 months

# Conclusions

- CTEPH is more common and often missed
- Clinical clues- PHT with previous PE or DVT
- Diagnosis is made by V\Q scan and CT-angio
- Surgery is the best treatment with high success rate
- Medical therapy should be reserved for inoperable cases or preoperatively

# Thank you