University of Pavia - School of Medicine
Foundation I.R.C.C.S. Policlinico "San Matteo"
Cardiac Surgery Unit - Chronic Thromboembolic Pulmonary Hypertension Centre
Pavia, Italy
Prof. Andrea M. D'Armini, M.D.

CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION
DIAGNOSIS – TREATMENT – CLINICAL CASE

FINANCIAL DISCLOSURE

Last three years

AOP Orphan Pharmaceuticals GmbH
Janssen Pharmaceutical
MSD

GENERAL CONSIDERATIONS

CTEPH is a two-cause disease

- Mecanichal obstruction → PEA and/or BPA are the therapeutic options

- Microvascular disease → specific medical therapy (Riociguat & Treprostinil plus Bosentan by 648)

EVALUATION PROCESS
DIFFERENT STEPS

- PEA's assessment
- Specific medical therapy
- BPA's assessment

REFERENCE

Terapia chirurgica dell'ipertensione polmonare cronica tromboembolica mediante endoarteriectomia polmonare

Andrea Maria D'Armini, Giorgio Zanotti, Matteo Pozzi, Salvatore Nicolardi, Corrado Tramontin, Vito G. Ruggieri, Marco Morsolini, Fabrizio Tancredi, Cristian Monterosso, Mario Viganò, a nome del Pavia Pulmonary Endarterectomy Study Group (vedi Appendice)

Cattedra e Divisione di Cardiochirurgia, Università degli Studi di Pavia, IRCCS Policlinico San Matteo, Pavia

(G Ital Cardiol 2006; 7 (7): 454-463)

REFERENCE

Tabella 2. Esami diagnostici di primo livello.

Test diagnostico	Razionale	Quadro tipico
ECG	Evidenziare dilatazione/sovraccarico ventricolare	Precoce: solitamente normale Tardivo: segni di dilatazione/ipertrofia VD e dilatazione AD
Rx torace	Visualizzare l'ombra cardiaca e gli ambiti polmonari (parenchimale/vascolare)	Precoce: solitamente normale Tardivo: area avascolare, dilatazione dell'arteria polmonare, trama vascolare irregolare (aumentato flusso bronchiale) e asimmetrica, esiti cicatriziali pleurici
Ecocardiogramma	Stimare le pressioni arteriose polmonari, valutare anatomia e funzione delle principali strutture cardiache, ricercare la presenza di shunt intracardiaci	Dilatazione/disfunzione delle camere cardiache destre, insufficienza tricuspidale destra, aumento delle pressioni polmonari, movimento paradosso del SIV, disfunzione VS, possibile comunicazione interatriale (FOP)

AD = atrio destro; ECG = elettrocardiogramma; FOP = forame ovale pervio; Rx = radiografia; SIV = setto interventricolare; VD = ventricolo destro; VS = ventricolo sinistro.

(G Ital Cardiol 2006; 7 (7): 454-463)

REFERENCE

Tabella 3. Esami diagnostici di secondo livello.

Test diagnostico	Razionale	Quadro tipico
Scintigrafia V/P	Diagnosi differenziale IPCTE vs IPI	Uno o più segmenti polmonari con "mismatch" ventilo-perfusorio
Cateterismo cardiaco destro	Misurare le pressioni in AP e IC, calcolare le RVP	Pressioni polmonari prossime a valori sistemici o sovrasistemiche, IC < 2.0 1/min/m^2 e RVP notevolmente elevate (spesso > 1000 dynes*s*cm^{-5})
PFR/EGA	Escludere/valutare cause parenchimali di IP, valutare la funzione respiratoria	Non anomalie significative/lieve quadro restrittivo (dovuto a precedenti infarti polmonari), ipossiemia, alcalosi respiratoria compensatoria, ridotta DL_{co}

AP = arteria polmonare; DL_{co} = capacità polmonare di diffusione per il monossido di carbonio; EGA = emogasanalisi; IC = indice cardiaco; IP = ipertensione polmonare; IPCTE = ipertensione polmonare cronica tromboembolica; IPI = ipertensione polmonare idiopatica; PFR = prove di funzionalità respiratoria; RVP = resistenze vascolari polmonari; V/P = ventilo-perfusoria.

(G Ital Cardiol 2006; 7 (7): 454-463)

REFERENCE

Tabella 4. Esami diagnostici di terzo livello.

Test diagnostico	Razionale	Quadro tipico
Angiografia polmonare	Valutare l'anatomia arteriosa polmonare, l'estensione/localizzazione degli emboli cronici (per porre diagnosi, valutare l'elegibilità all'intervento e prevedere l'outcome operatorio)	Irregolarità intimali, difetti "a tasca", bande o "reti" intravascolari, riduzione repentina del calibro vascolare o amputazione dello stesso
TC torace	Fase vascolare: identica a quella angiografica	Quadro vascolare: identico a quello angiografico
	Fase parenchimale/mediastinica: valutare il parenchima polmonare, escludere cause estrinseche di IP	Quadro parenchimale: perfusione a mosaico ed esiti cicatriziali subpleurici Quadro mediastinico: dilatazione delle arterie bronchiali

IP = ipertensione polmonare; TC = tomografia computerizzata.

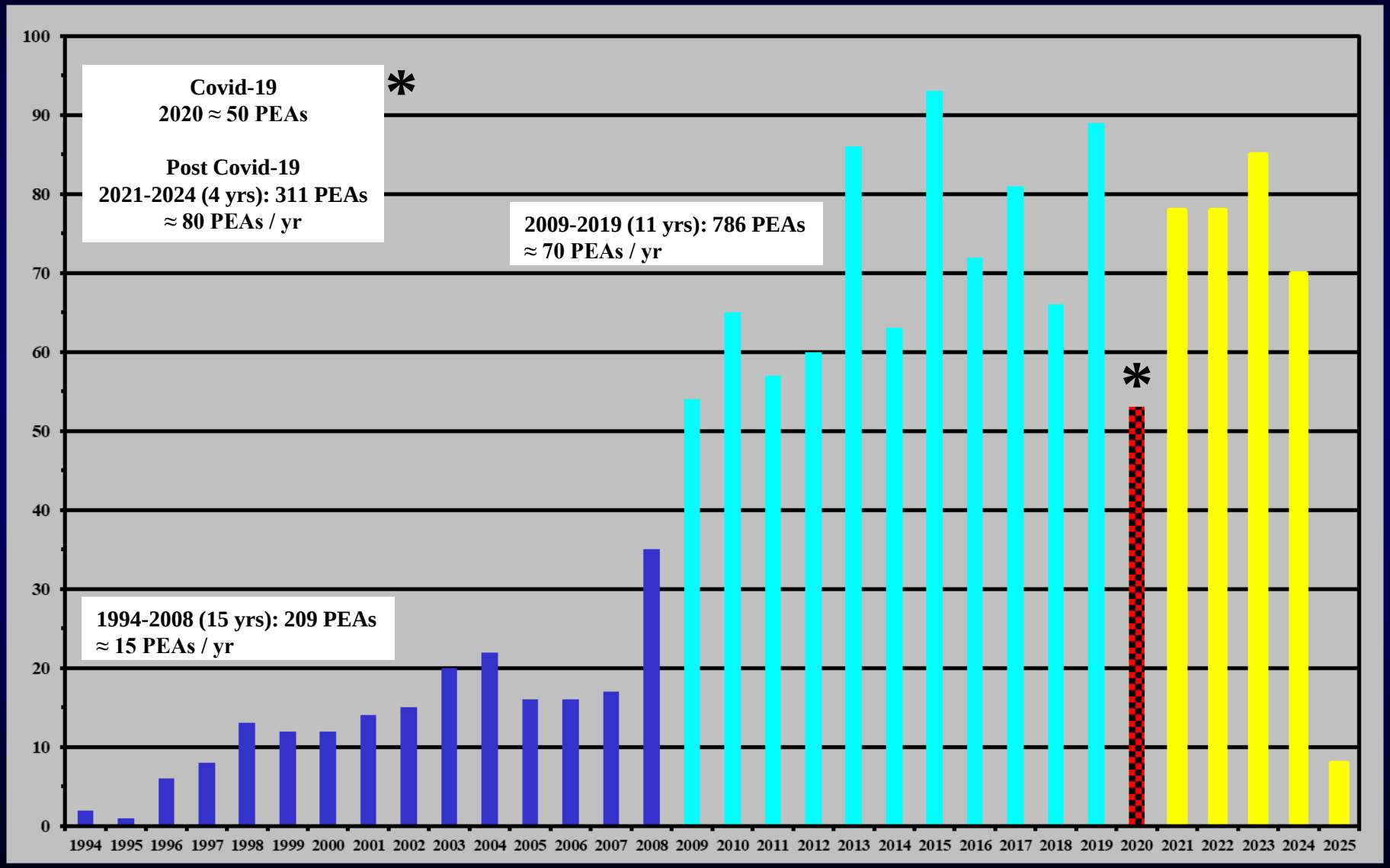
(G Ital Cardiol 2006; 7 (7): 454-463)

OUR PROGRAM

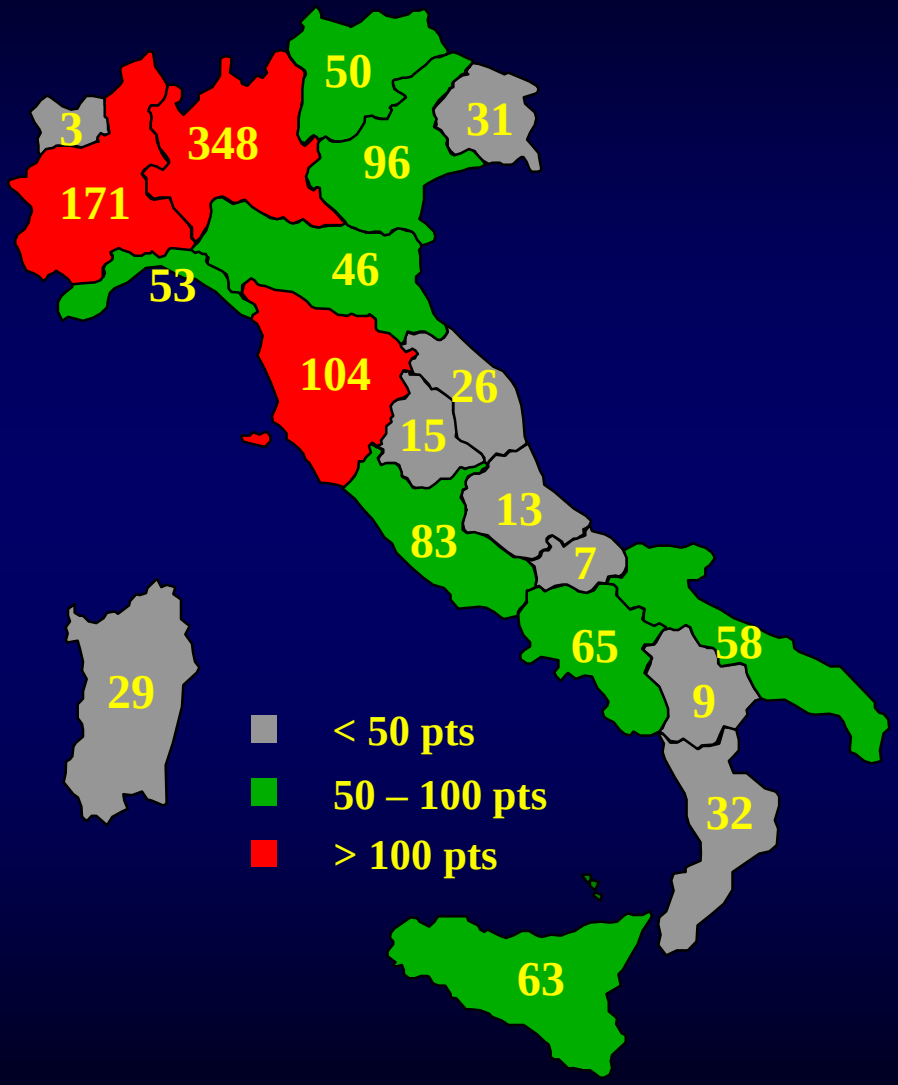
- National and international referral program
- Begin: April 1994
- January 2025: 1367 PEAs performed

AMOUNT OF PATIENTS

1367 PEAs



PATIENTS' REFERRAL

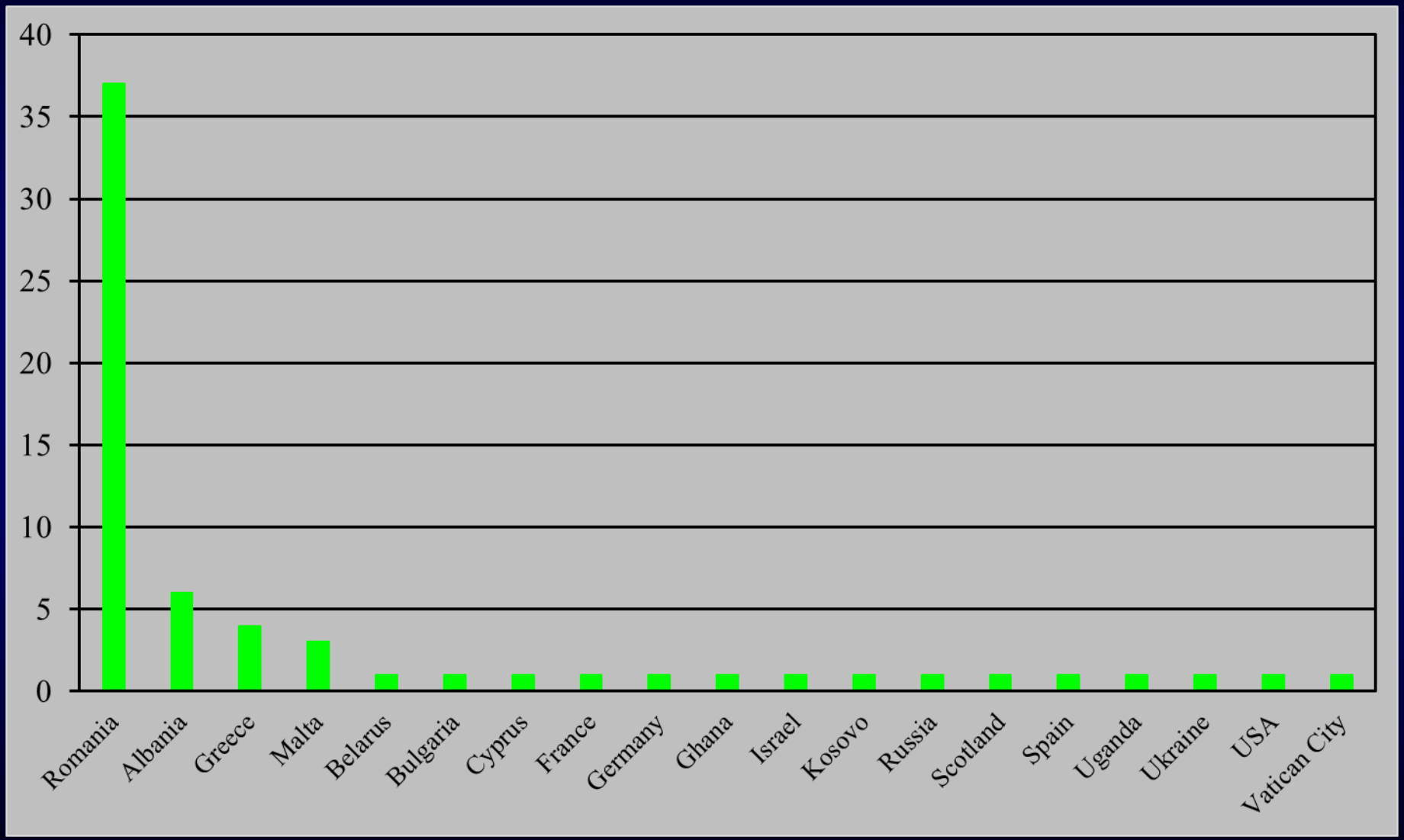


FROM 4/1994 TO 1/2025
1302 PEAs

LOMBARDY vs. OTHERS
27% vs. 73%

OUTSIDE ITALY PATIENTS' REFERRAL

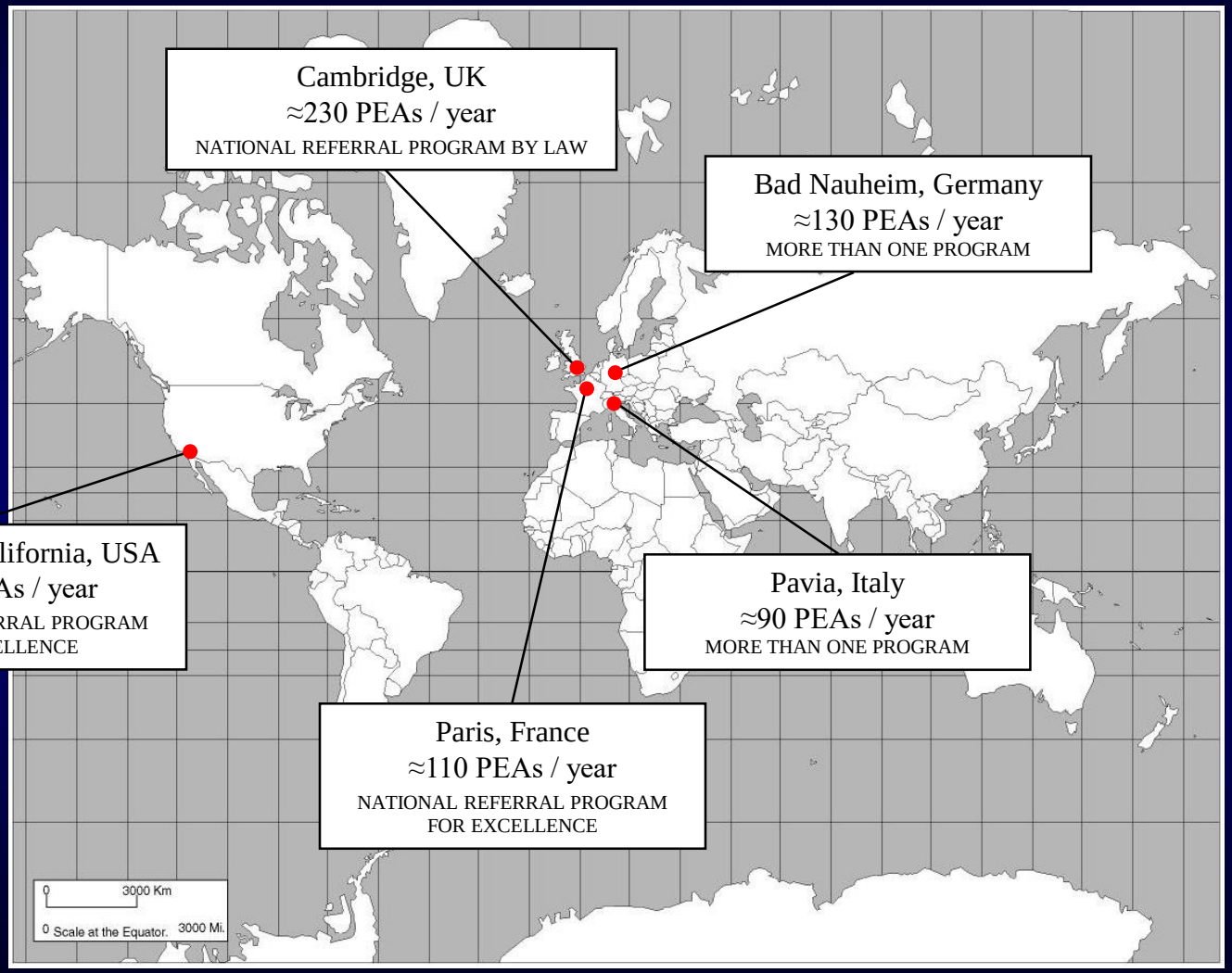
FROM 10/2001 TO 1/2024 - 65 PEAs



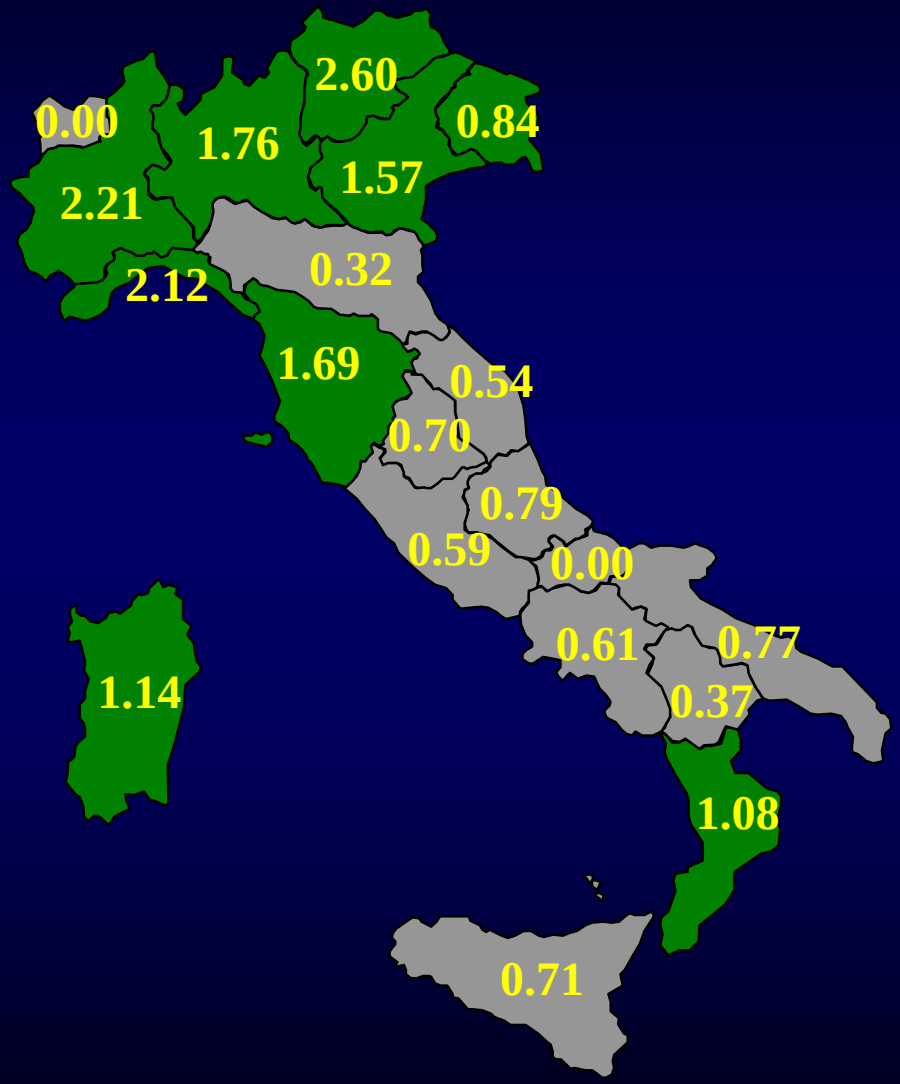
OUTSIDE ITALY PATIENTS' REFERRAL DIVIDED BY PERIOD OF ACTIVITY

PERIOD	YEARS	REFERRAL
1994 - 2008	15	2/209 (1.0%) **[FIRST PATIENT - 2001]**
2009 - 2018	10	10/697 (1.4%)
2019 - 2023	5	38/383 (9.9%) **[2023 - 19/85 - 22.4%]**
2024 - 2025	2	15/78 (19.2%) **[2024 - 15/70 - 21.4%]**
1994 - 2025	32	65/1367 (4.8%)

MAIN WORLD PEA CENTERS 2019



PATIENTS' REFERRAL



FROM 2019 TO 2023 → 345 PEAs
PTS / 10^6 / Year POPULATION

- ≤ 1 pt / 10^6 / Year
- 1 – 3 pts / 10^6 / Year
- ≥ 3 pts / 10^6 / Year

Update date population at 01/01/2024 (ISTAT)

GENERAL CONSIDERATIONS

Better diagnosis → more evaluated patients at a earlier stage

Better treatment → more operated patients with better results

INDICATIONS FOR SURGERY

NYHA FUNCTIONAL CLASS

ACQUIRED CARDIOVASCULAR DISEASE

(J Thorac Cardiovasc Surg 2011;141:702-10)

Surgical management and outcome of patients with chronic thromboembolic pulmonary hyperte… international prospective registry

Eckhard Mayer, MD,[a] David Jenkins, FRCS,[b] Jaroslav …
Jaap Kloek, MD,[e] Bart Meyns, MD,[f] Lars Bo Ilkjaer, MI…
Irene Lang, MD,[h] Joanna Pepke-Zaba, MD,[b] Gerald Sir…

Study Design

This prospective registry was designed to include newly diagnosed (≤6 months) consecutive patients with CTEPH in participating centers in Europe and Canada, from February 2007 to January 2009. The registry proto-

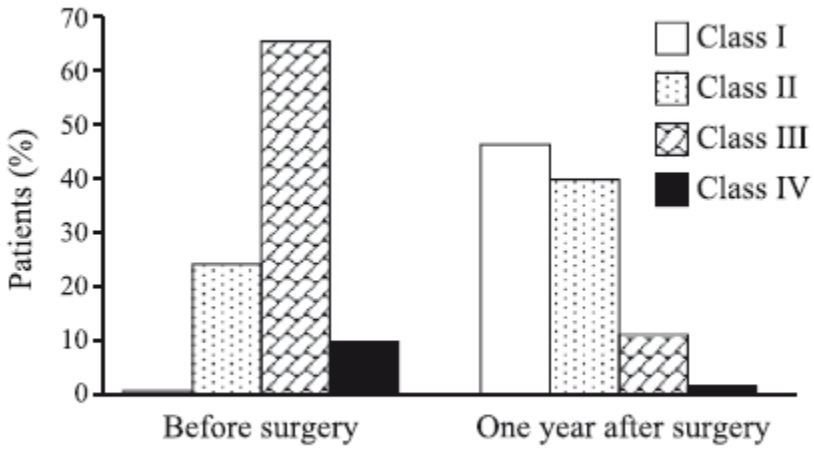


FIGURE 1. NYHA functional class before surgery and within 1 year after surgery (n = 221).

NYHA FUNCTIONAL CLASS

	INTERNATIONAL REGISTRY 2007-2008	PAVIA 2007-2008	PAVIA 2017-2018
II	25%	5%	19%
III	65%	50%	56%
IV	10%	45%	25%

NYHA FUNCTIONAL CLASS

Long-Term Outcome of Patients With Chronic Thromboembolic Pulmonary Hypertension

Results From an International Prospective Registry

Marion Delcroix, MD; Irene Lang, MD; Joanna Pepke-Zaba, MD; Pavel Jansa, MD; Andrea M. D'Armini, MD; Repke Snijder, MD; Paul Bresser, MD; Adam Torbicki, MD; Soren Mellemkjaer, MD; Jerzy Lewczuk, MD; Iveta Simkova, MD; Joan A. Barbera, MD; Marc de Perrot, MD; Marius M. Hoeper, MD; Sean Gaine, MD; Rudolf Speich, MD; Miguel A. Gomez-Sanchez, MD; Gabor Kovacs, MD; Xavier Jais, MD; David Ambroz, MD; Carmen Treacy, BSc; Marco Morsolini, MD; David Jenkins, MD; Jaroslav Lindner MD; Philippe Dartevelle, MD; Eckhard Mayer, MD; Gerald Simonneau, MD

Circulation. 2016;133:859-871

NYHA FUNCTIONAL CLASS

NYHA class IV is one of the most important predictor of death increasing mortality by 4 to 5 times

Table 4. Independent Correlates of Mortality for Operated and Not-Operated Patients

	Operated (n=346)			Not-Operated (n=219)		
Covariate	HR	95% CI	*P* Value	HR	95% CI	*P* Value
NYHA class III vs I–II				2.43	1.00–5.89	0.0489
NYHA class IV vs I–II	4.16	1.49–11.62	0.0065	4.76	1.76–12.88	0.0021

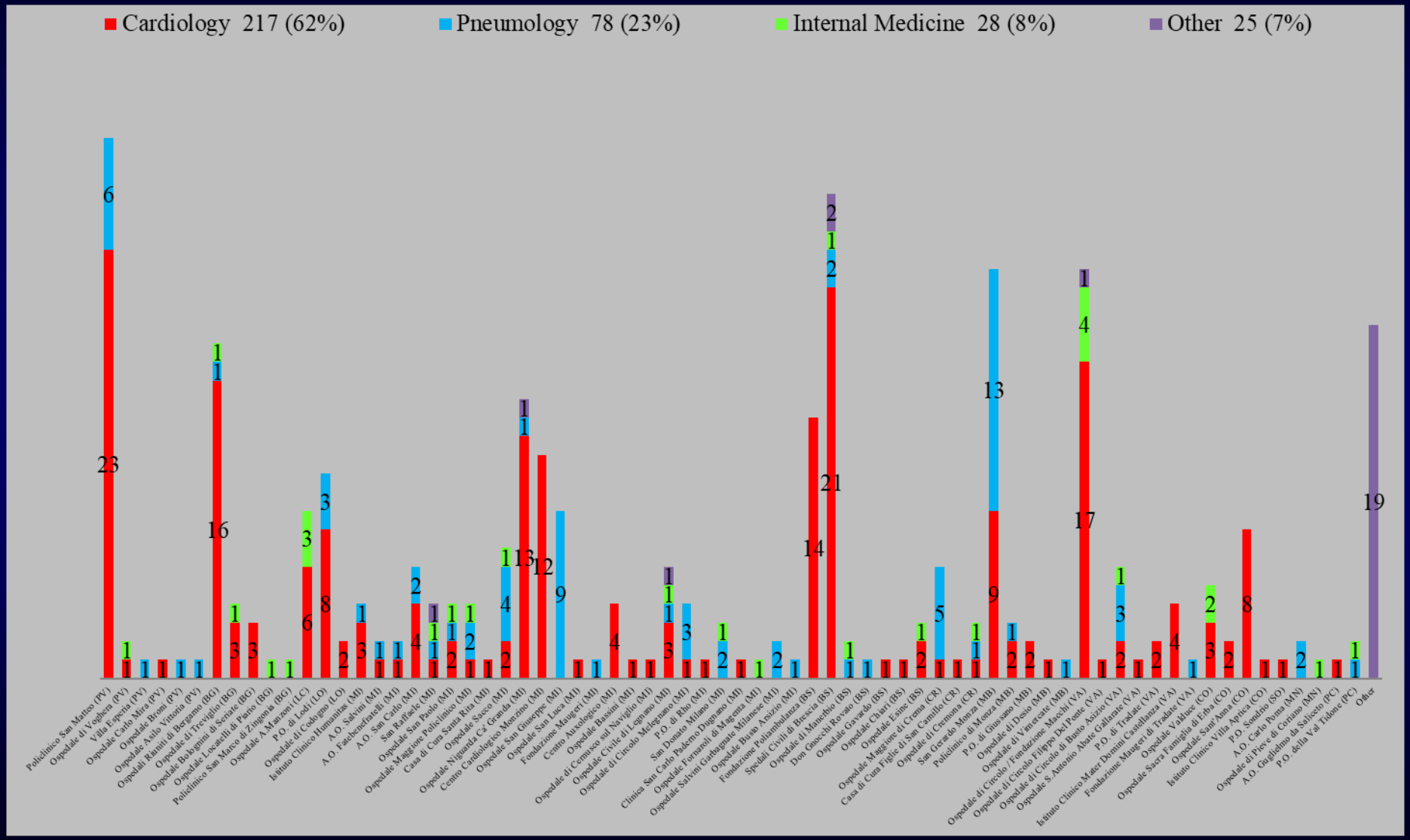
Circulation. 2016;133:859-871

HOSPITAL MORTALITY DIVIDED BY FUNCTIONAL CLASS

NYHA	HOSPITAL MORTALITY
II – 17%	2/225 (0.9%)
III – 47%	27/644 (4.2%)
IV – 36%	65/498 (13.1%)
II & III & IV	94/1367 (6.9%)

PTS' REFERRAL FROM 71 HOSPITAL IN LOMBARDIA

FROM 4/1994 TO 1/2025 - 348 PEAs



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GENERAL CONSIDERATIONS

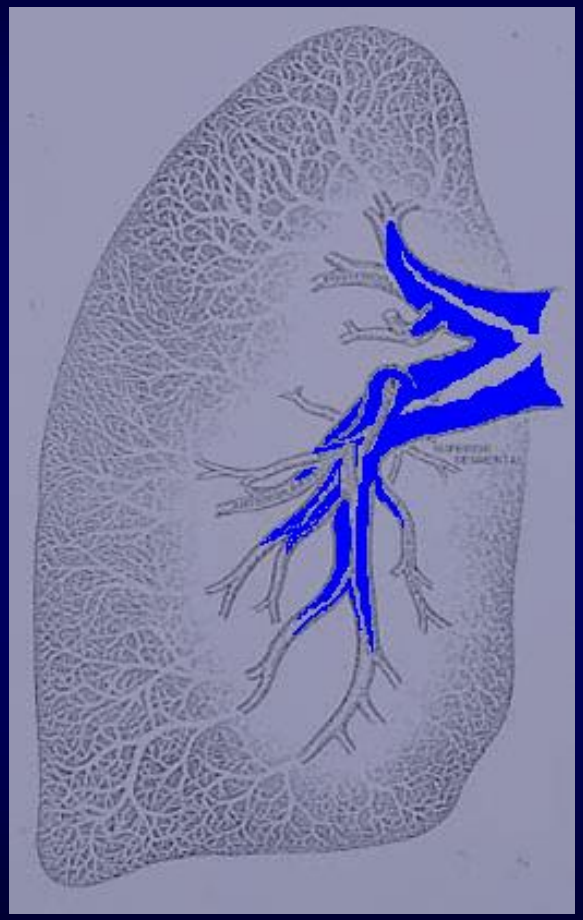
Better diagnosis → more evaluated patients at a earlier stage

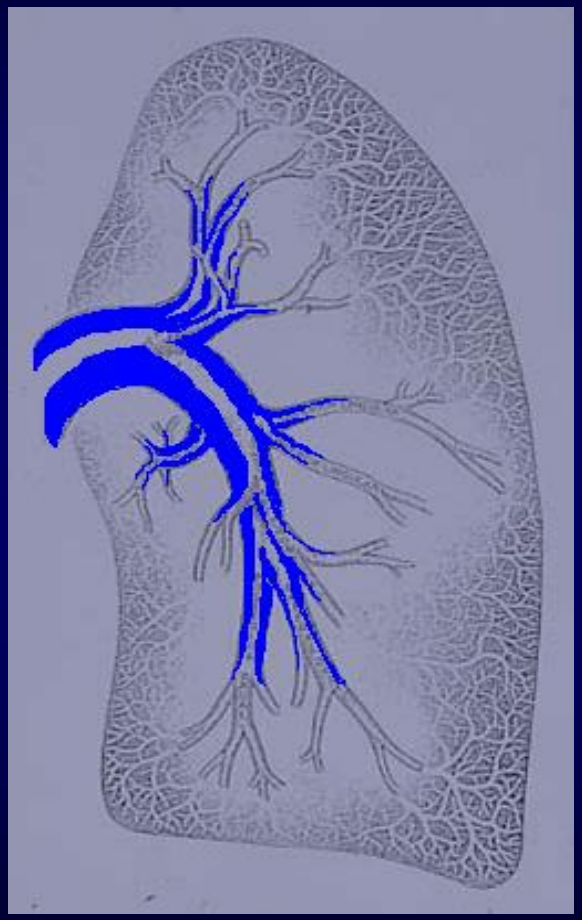
Better treatment → more operated patients with better results

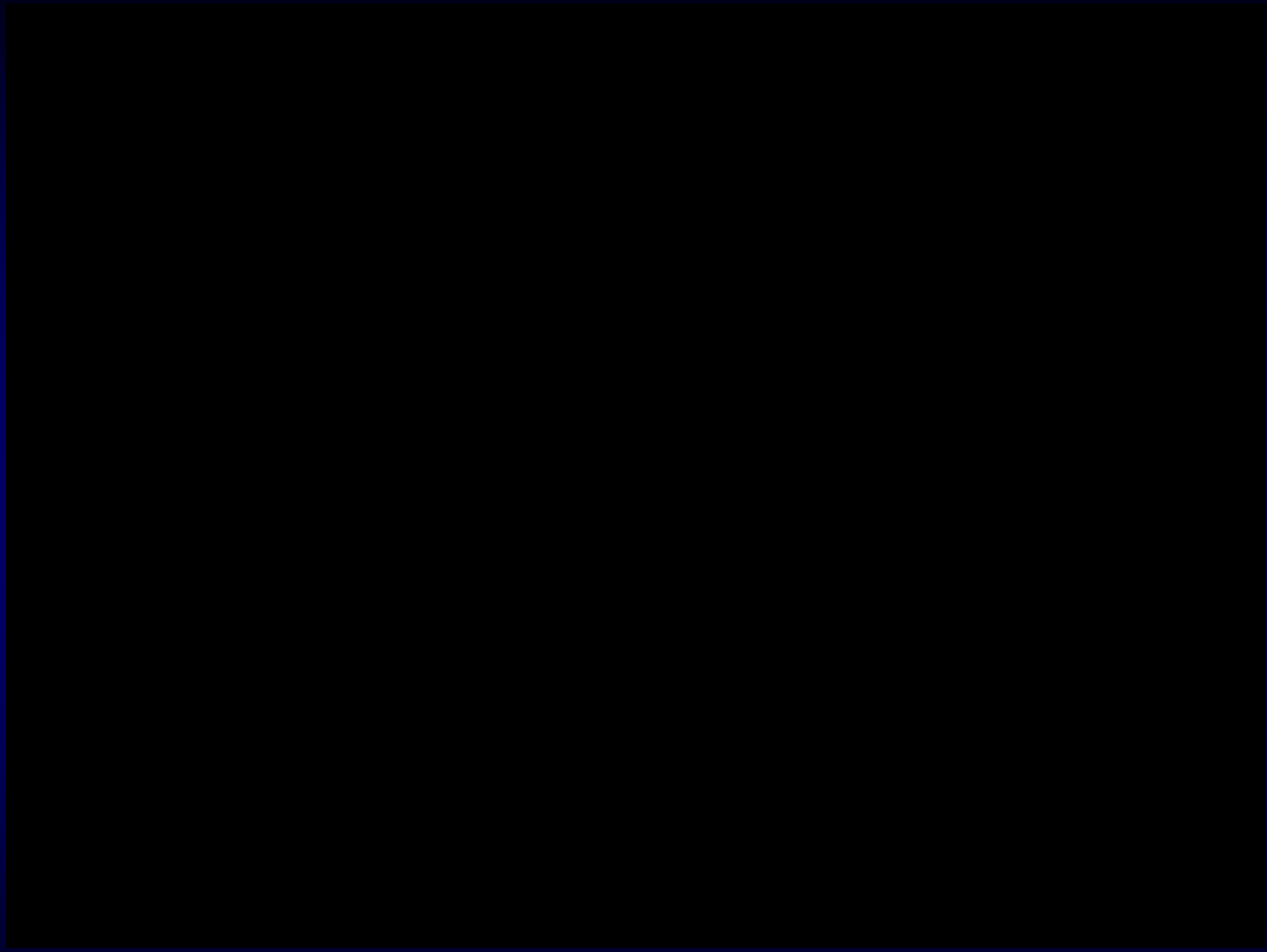
OPERABILITY ASSESSMENT

- Gold-standard and curative procedure for CTEPH is PEA
- Different operability percentage in different Centers
- Second opinion is mandatory before judge a patient inoperable

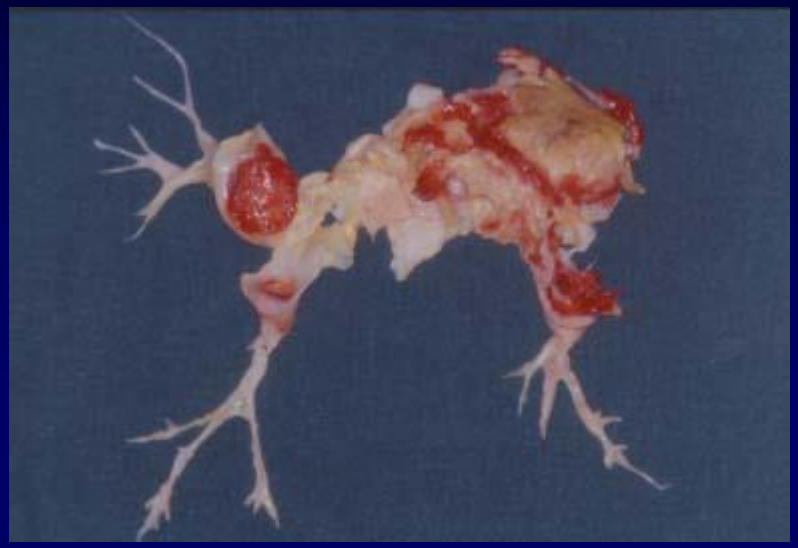
PROXIMAL LESIONS



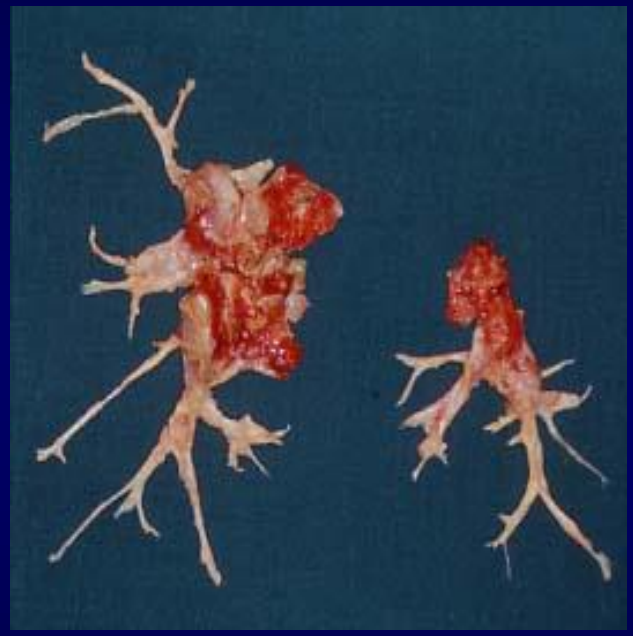




PROXIMAL SURGICAL SPECIMENS

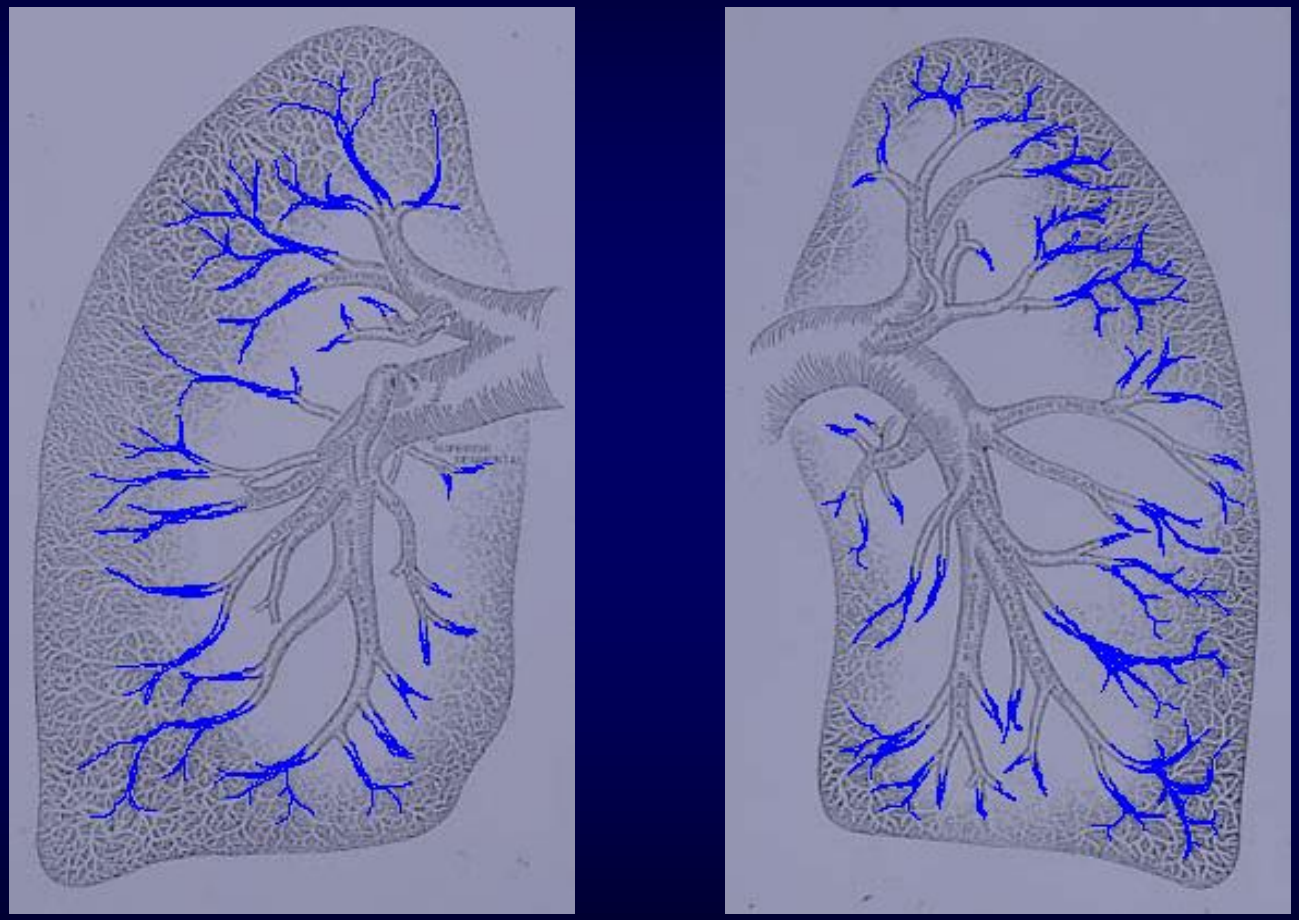


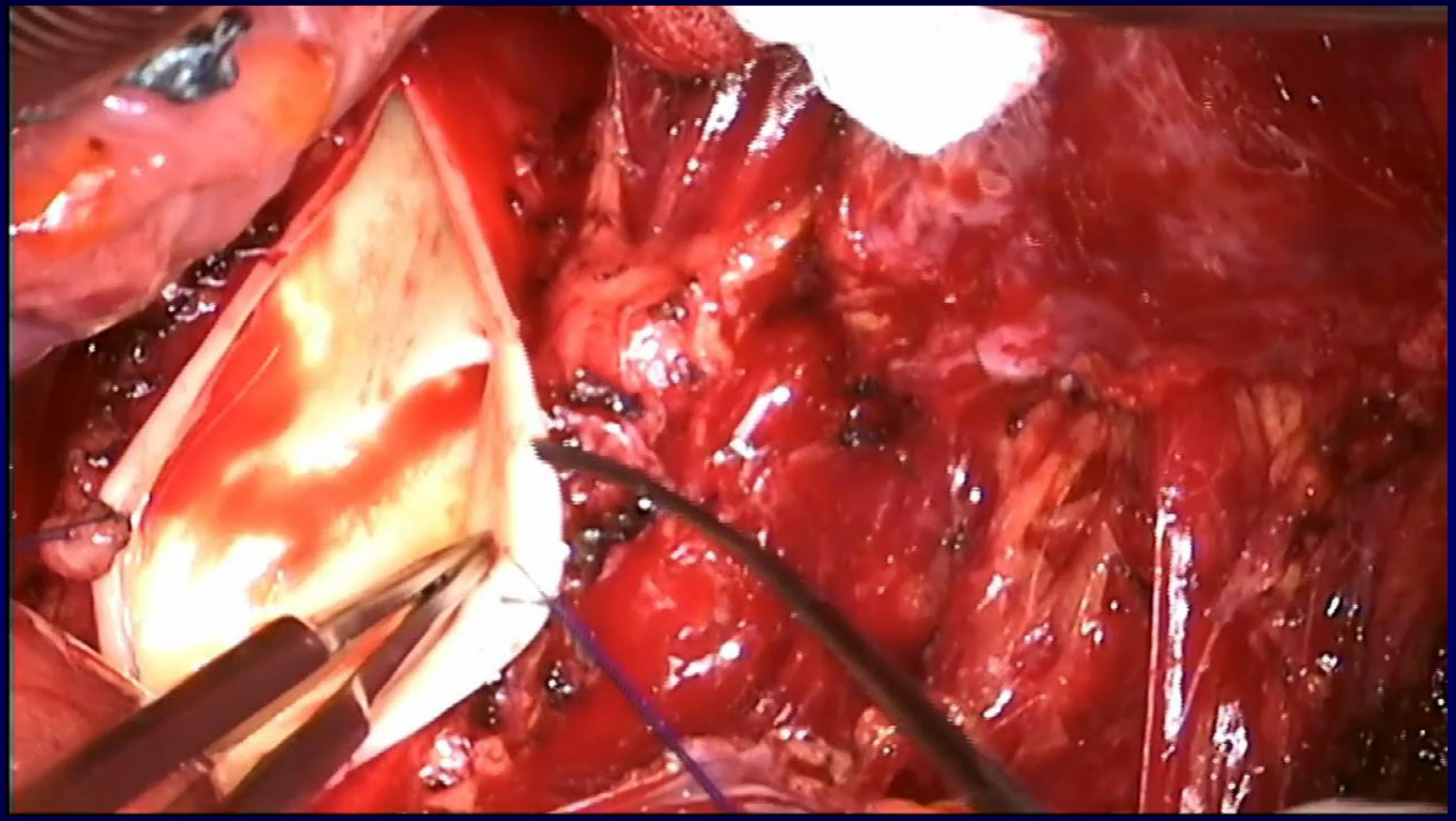
mPAP	**43**	**→**	**20**	**(-53%)**
CO	**3.3**	**→**	**6.9**	**(+109%)**
PVR	**994**	**→**	**220**	**(-78%)**



mPAP	**50**	**→**	**25**	**(-50%)**
CO	**2.6**	**→**	**4.4**	**(+69%)**
PVR	**1385**	**→**	**364**	**(-74%)**

DISTAL LESIONS





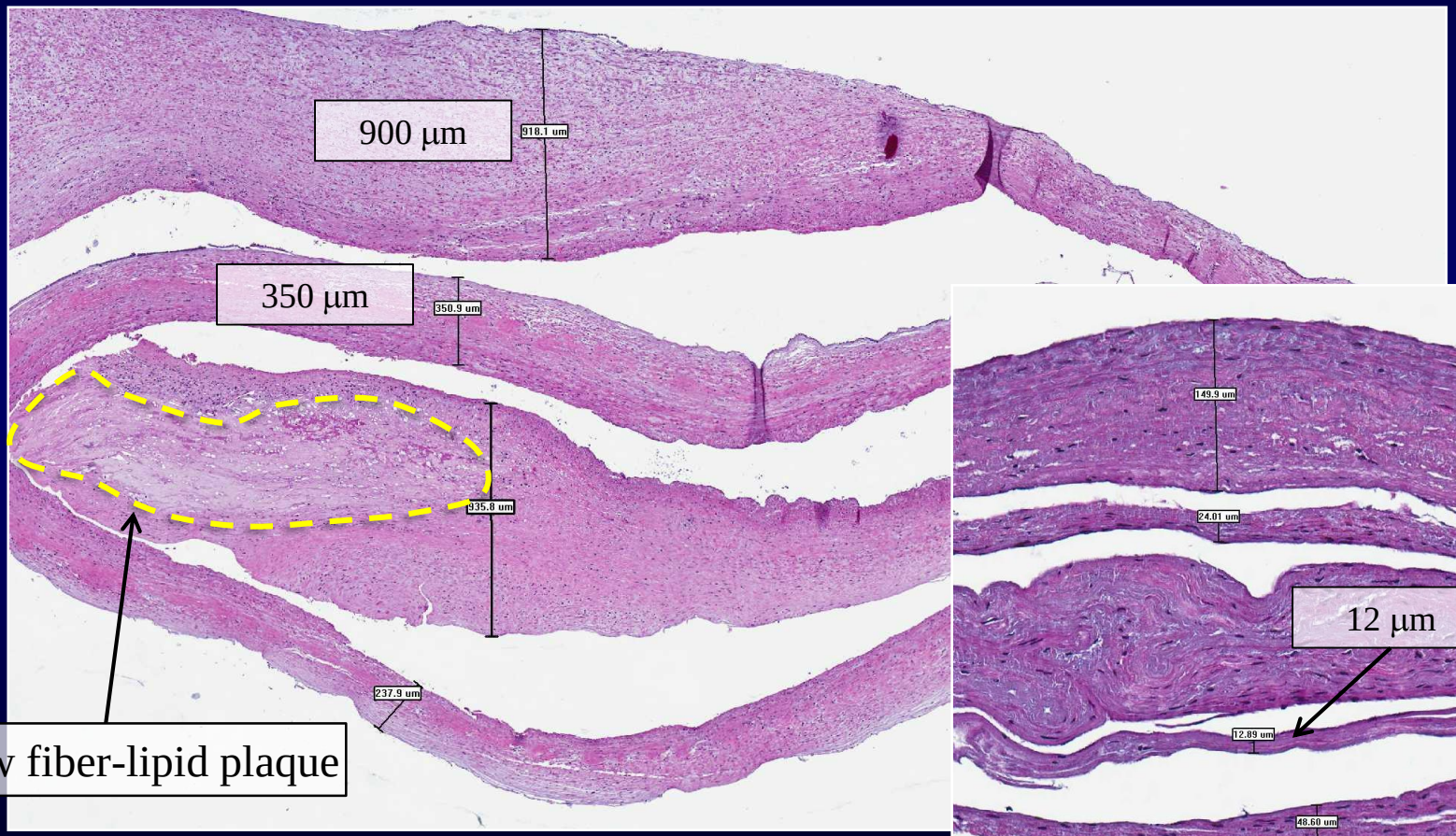
CORRECT ARTERIAL DISSECTION PLANE

Reverse Ariadne's thread

SURGICAL SPECIMENS

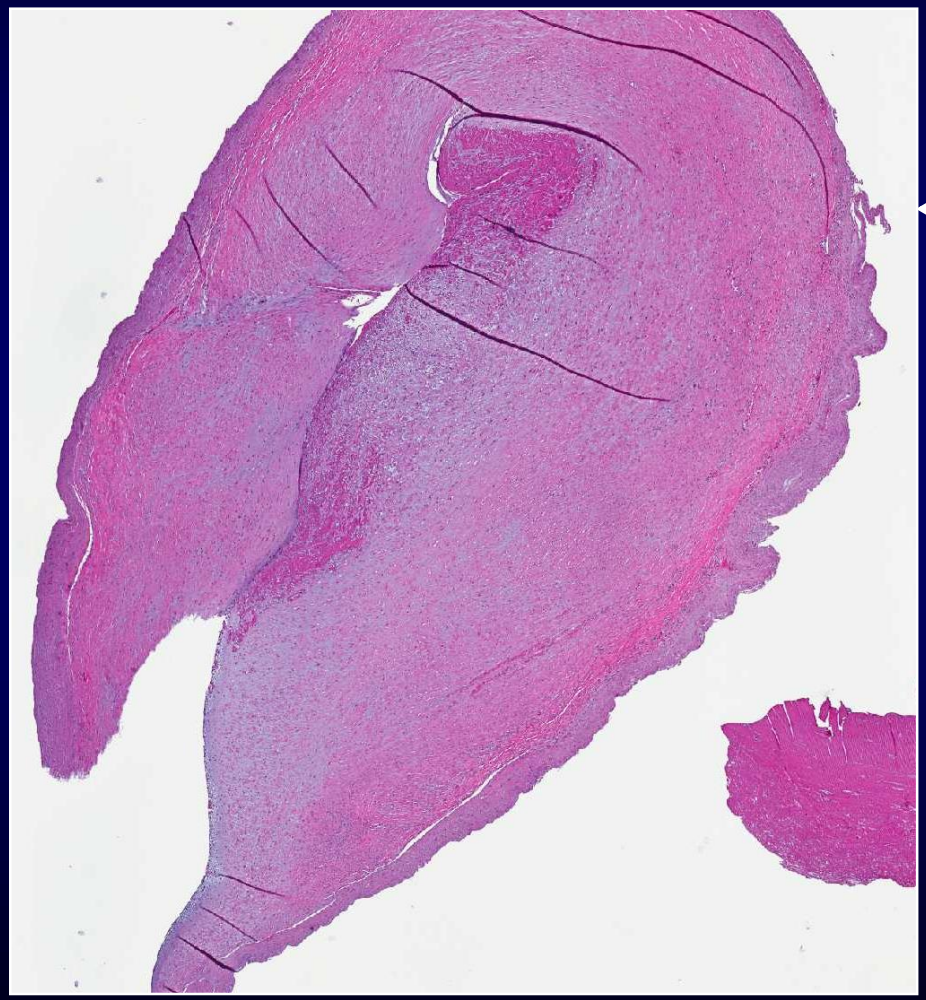
TRICKS AND TIPS

Proximal dissection for the clearance of distal obstructions



TRICKS AND TIPS

Proximal dissection for the clearance of distal obstructions



DISTAL LESIONS

Pulmonary endarterectomy for distal chronic thromboembolic pulmonary hypertension

Andrea M. D'Armini, MD,[a,b] Marco Morsolini, MD, PhD,[a] Gabriella Mattiucci, MD,[a,b] Valentina Grazioli, MD,[a,b] Maurizio Pin, MD,[b] Adele Valentini, MD,[c] Giuseppe Silvaggio, MD,[b] Catherine Klersy, MD, MSc,[d] and Roberto Dore, MD[c]

(J Thorac Cardiovasc Surg 2014;148:1005-12)

DISTAL LESIONS

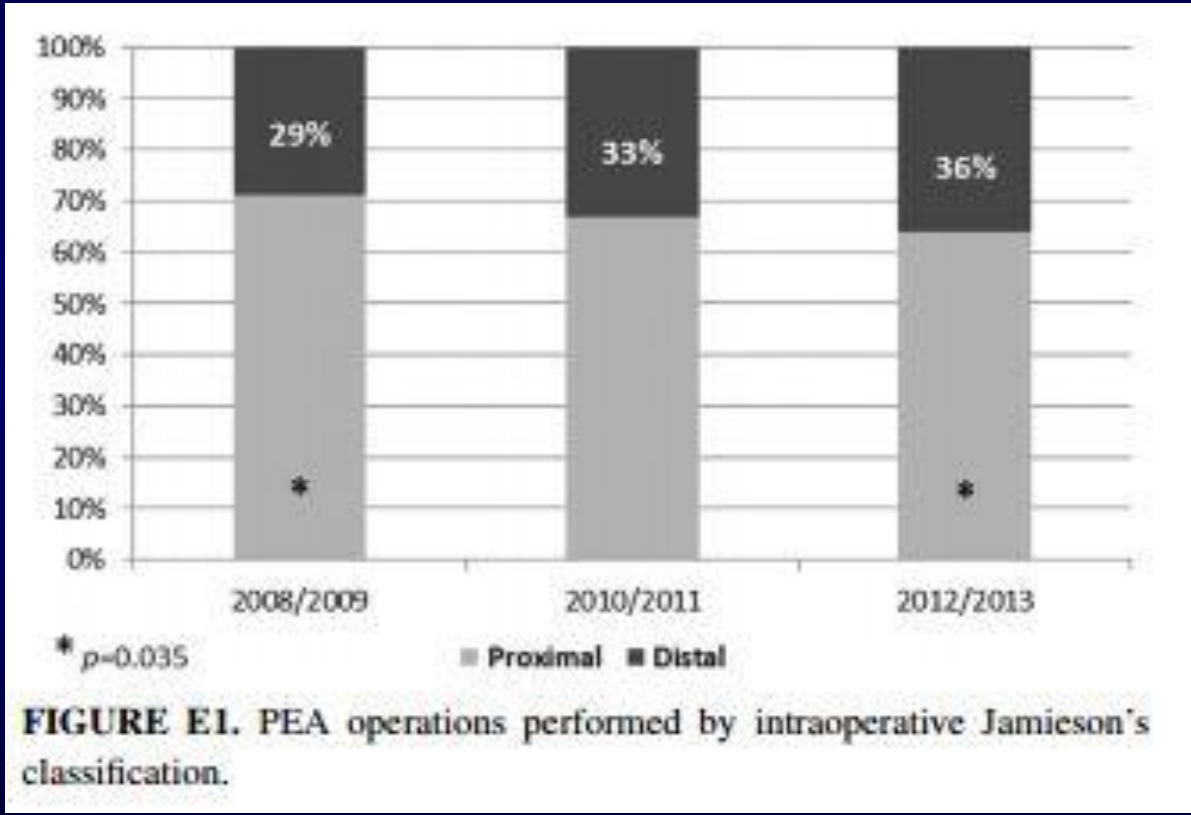


FIGURE E1. PEA operations performed by intraoperative Jamieson's classification.

DISTAL LESIONS

TABLE 3. Hemodynamic time course after pulmonary endarterectomy

	Proximal	Distal
Mean pulmonary arterial pressure (mm Hg)		
Preoperative	44 ± 10	46 ± 11
At discharge	22 ± 7	24 ± 6
3-mo follow-up	24 ± 9	25 ± 7
12-mo follow-up	23 ± 7	24 ± 8
P value*	<.001	<.001
PVR (dyne·s·cm^{-5})		
Preoperative	876 ± 392	926 ± 337
At discharge	251 ± 146	295 ± 161
3-mo follow-up	270 ± 175	300 ± 139
12-mo follow-up	243 ± 115	300 ± 224
P value*	<.001	<.001
Cardiac output (L/min)		
Preoperative	3.9 ± 1.3	3.7 ± 1.2
At discharge	5.0 ± 1.2	4.7 ± 1.2
3-mo follow-up	5.2 ± 1.1	5.0 ± 1.2
12-mo follow-up	5.0 ± 1.1	4.7 ± 1.0
P value*	<.001	<.001

PVR, Pulmonary vascular resistance. *Each time point versus preoperative. Test of interaction: $P = .975$ (mean pulmonary arterial pressure); $P = .777$ (PVR); $P = .825$ (cardiac output).

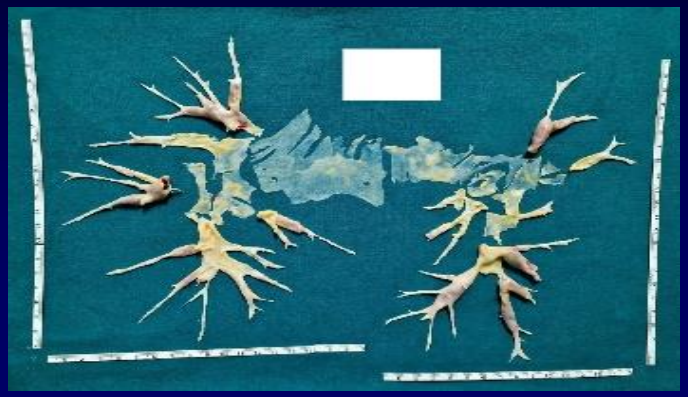
DISTAL LESIONS

TABLE 4. Partial pressure of oxygen in arterial blood, modified Bruce exercise test, and 6-minute walking distance time course after pulmonary endarterectomy

	Proximal	Distal
Arterial partial pressure of oxygen (mm Hg)		
Preoperative	65 ± 12	66 ± 11
3-mo follow-up	82 ± 13	80 ± 11
12-mo follow-up	80 ± 11	80 ± 11
P value*	<.001	<.001
Modified Bruce exercise test (m)		
Preoperative	51 (0-143)	52 (0-102)
3-mo follow-up	495 (182-658)	435 (143-586)
12-mo follow-up	520 (261-709)	474 (225-620)
P value*	<.001	<.001
6-min walking distance (m)		
Preoperative	277 ± 118	289 ± 112
3-mo follow-up	391 ± 118	398 ± 107
12-mo follow-up	389 ± 118	396 ± 112
P value*	<.001	<.001

*Each time point versus preoperative. Test of interaction: $P = .317$ (partial pressure of oxygen in arterial blood); $P = .205$ (modified Bruce exercise test); $P = .962$ (6-min walking distance).

TREATED BRANCHES



75 TREATED BRANCHES

59 TREATED BRANCHES

63 TREATED BRANCHES

IT IS IMPORTANT THE NUMBER OF TREATED BRANCHES?

International Journal of CARDIOLOGY

PULMONARY ENDARTERECTOMY IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RELATIONSHIP BETWEEN TREATED BRANCHES AND OUTCOME

Andrea M. D'Armini[1 2], Maurizio Pin[1], Anna Celentano[1], Leslie Joyce Te Masiglat[1], Ermelinda Borrelli[1], Benedetta Vanini[1], Catherine Klersy[3], Giuseppe Silvaggio[4], Cristian Monterosso[4], Alessia Alloni[4], Carlo Pellegrini[1 4], Stefano Ghio[5].

Int J Cardiol 2023 Apr 15;377:124-130

UNIVERSITY OF PAVIA SCHOOL OF MEDICINE - SAN MATTEO HOSPITAL - PAVIA - ITALY

BACKGROUND

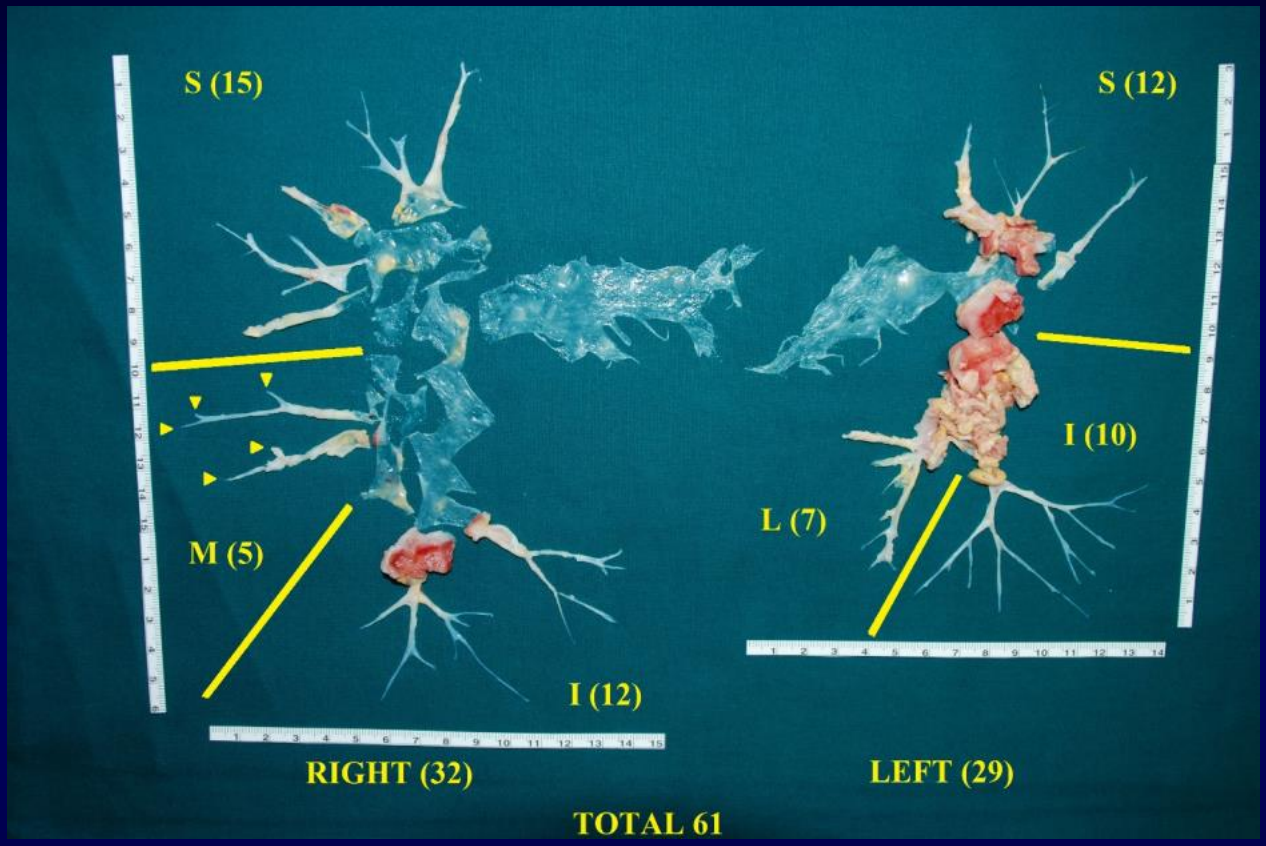
In patients with CTEPH undergoing PEA it is important to minimize residual obstructions, in order to achieve low postoperative pulmonary vascular resistances and better clinical results

METHODS

In 564 consecutive CTEPH pts undergoing PEA the count of the number of treated branches was performed directly on the surgical specimens

Post-operative FUP visits were scheduled at 3 months and 12 months after surgery including right heart catheterization and modified Bruce test

ANALYSIS OF PEA SPECIMENT



Surgical specimen of a patient with a Jamieson 1 disease. A total of 32 branches were treated in the right lung and 29 in the left lung. The arrows indicate how 5 treated branches were counted in the middle lobe of the right lung

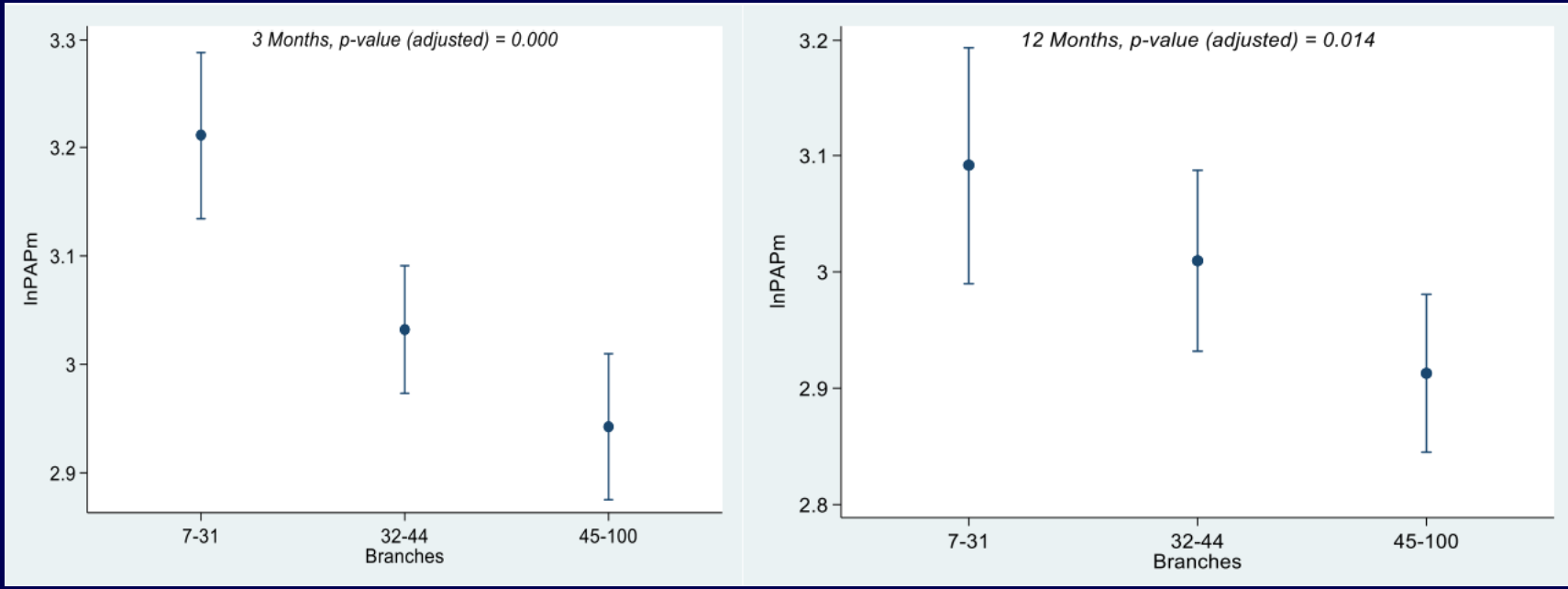
S= superior lobe, M = middle lobe, I = inferior lobe, L = lingula

POPULATON

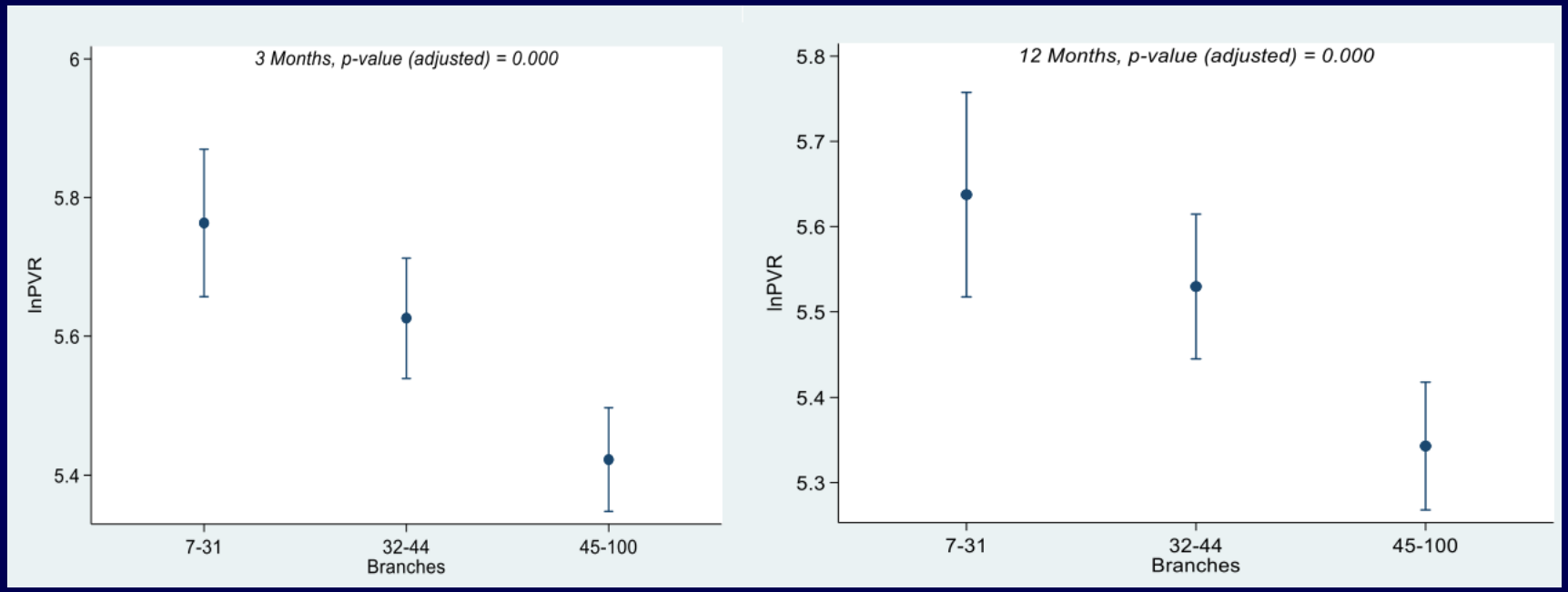
The population was divided into tertiles based on the number of treated branches

Group 1	from 4 to 30 treated branches	194 patients
Group 2	from 31 to 43 treated branches	190 patients
Group 3	from 44 to 100 treated branches	180 patients

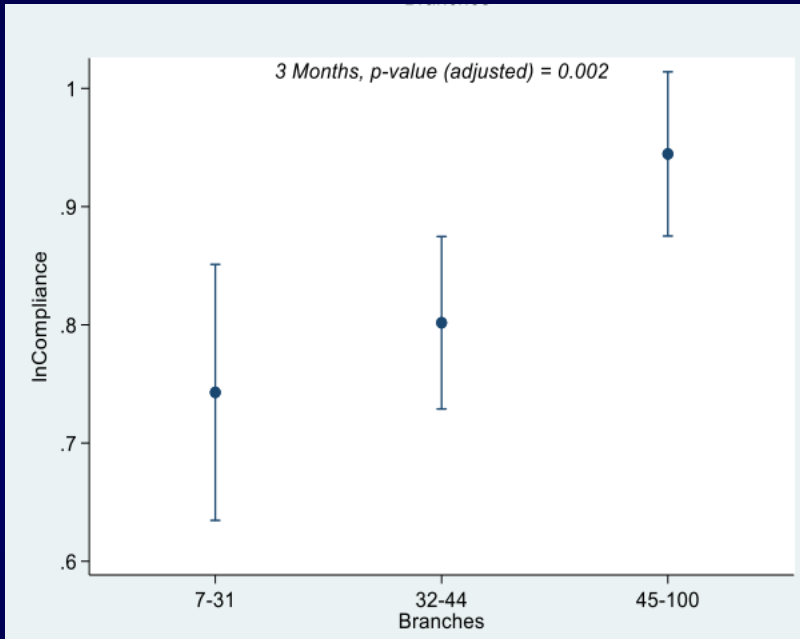
MPAP AT 3 AND 12 MONTHS AFTER PEA

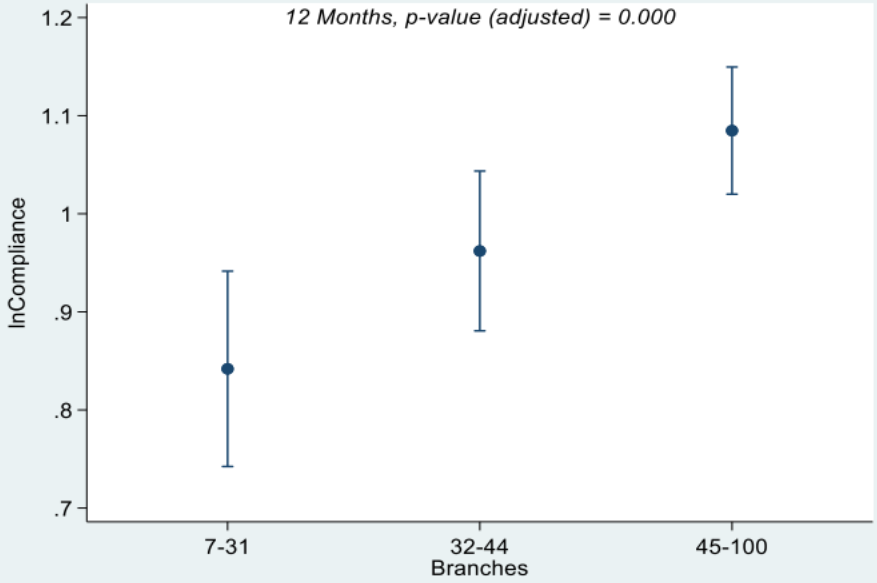


PVR AT 3 AND 12 MONTHS AFTER PEA

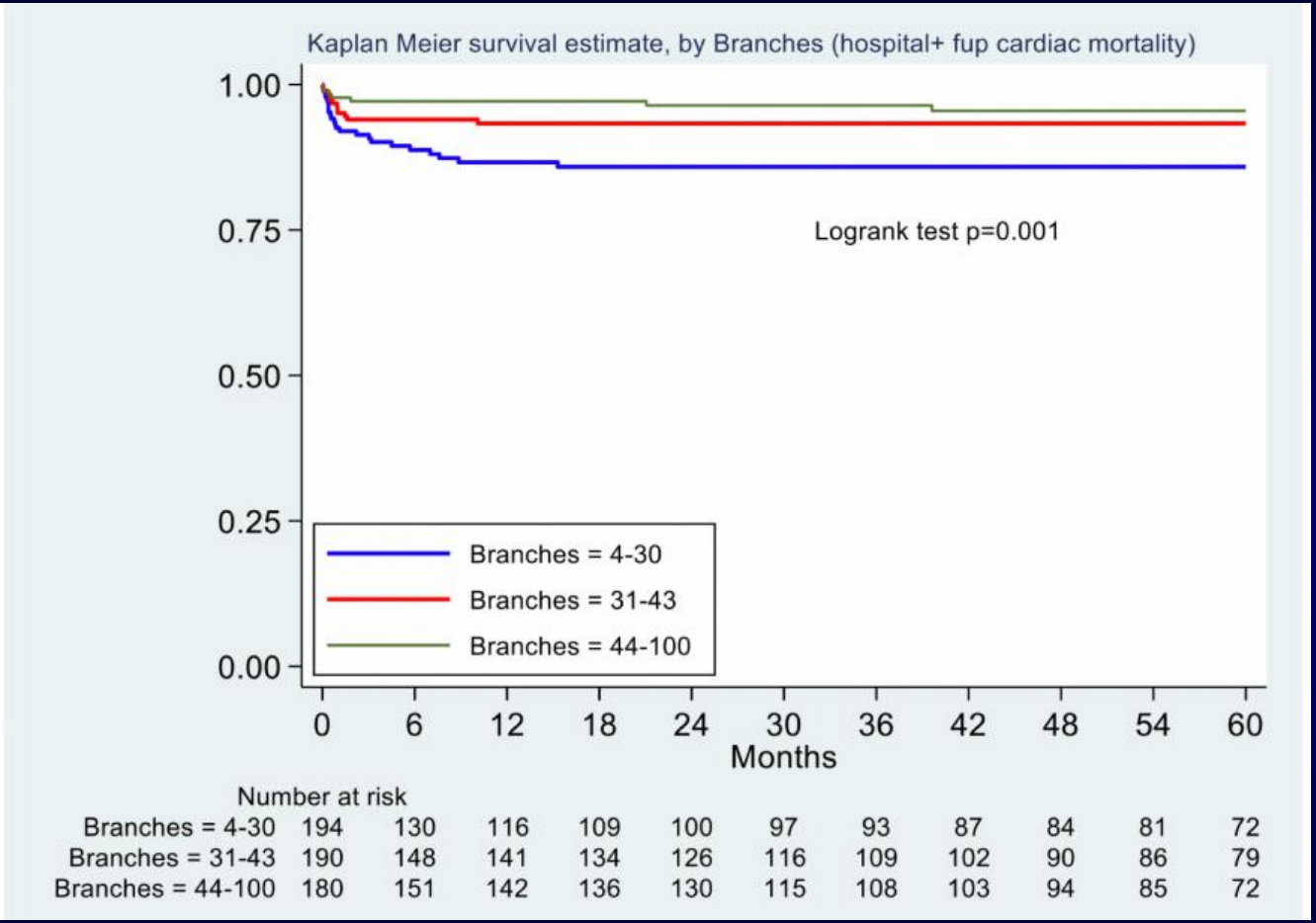


PCa AT 3 AND 12 MONTHS AFTER PEA





LONG-TERM SURVIVAL



EVOLVING SURGICAL TECHIQUE

Editorial

Surgical management of pulmonary endarterectomy avoiding deep hypothermia: the Pavia experience

Andrea M. D'Armini[1,2], Anna Celentano[1], Alessia Alloni[3], Giuseppe Silvaggio[3], Cristian Monterosso[3], Carlo Pellegrini[1,3], Stefano Ghio[4]

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ANNALS OF CARDIOTHORACIC SURGERY

UNIVERSITY OF PAVIA SCHOOL OF MEDICINE - SAN MATTEO HOSPITAL - PAVIA - ITALY

SURGICAL PROTOCOL

TAILORED AND LESS INVASIVE SURGERY

Since 15-10-2009 (#245)

	Original San Diego protocol	Actual Pavia protocol
Aortic clamp	Yes	No
Cardioplegia	Yes	No
Hypothermia	Deep (18°C)	Moderate (24°C)
Circulatory arrest	A single (20 minutes) period of circulatory arrest for each side (with a maximum of a third)	Intermittent short periods of circulatory arrest (≈7-10 minutes) followed by short re-perfusion periods (≈5-7 minutes)
Total arrest time	Maximum 60 minutes	Maximum 180 minutes

SURGICAL PROTOCOL

Up today more than 1100 PEAs were performed with this technique

CONCLUSIONS

In our hands a longer total circulatory arrest time (average 95 minutes) allows the surgeon to explore all the pulmonary vascular bed, find unexpected chronic thromboembolic material and clean more branches (average 45) even in more complex clinical conditions as well as in distal vassels

NEUROPSYCHOLOGICAL OUTCOMES

ELSEVIER

The Journal of Heart and Lung Transplantation
http://www.jhltonline.org

INNOVATION

Neuropsychological outcomes after pulmonary endarterectomy using moderate hypothermia and periodic circulatory arrest

Benedetta Vanini, PsyD, PhD,[a,b] Valentina Grazioli, MD,[a] Antonio Sciortino, MD,[a] Maurizio Pin, MD,[a] Vera N. Merli, MD,[a] Anna Celentano, MD,[a] Ilaria Parisi,[a] Catherine Klersy, MD,[c] Lucia Petrucci, MD,[d] Maurizio Salati, MD,[a] Pierluigi Politi, MD, PhD,[b] and Andrea M. D'Armini, MD[a]

J Heart Lung Transplant 2018 Jul;37(7):860-4

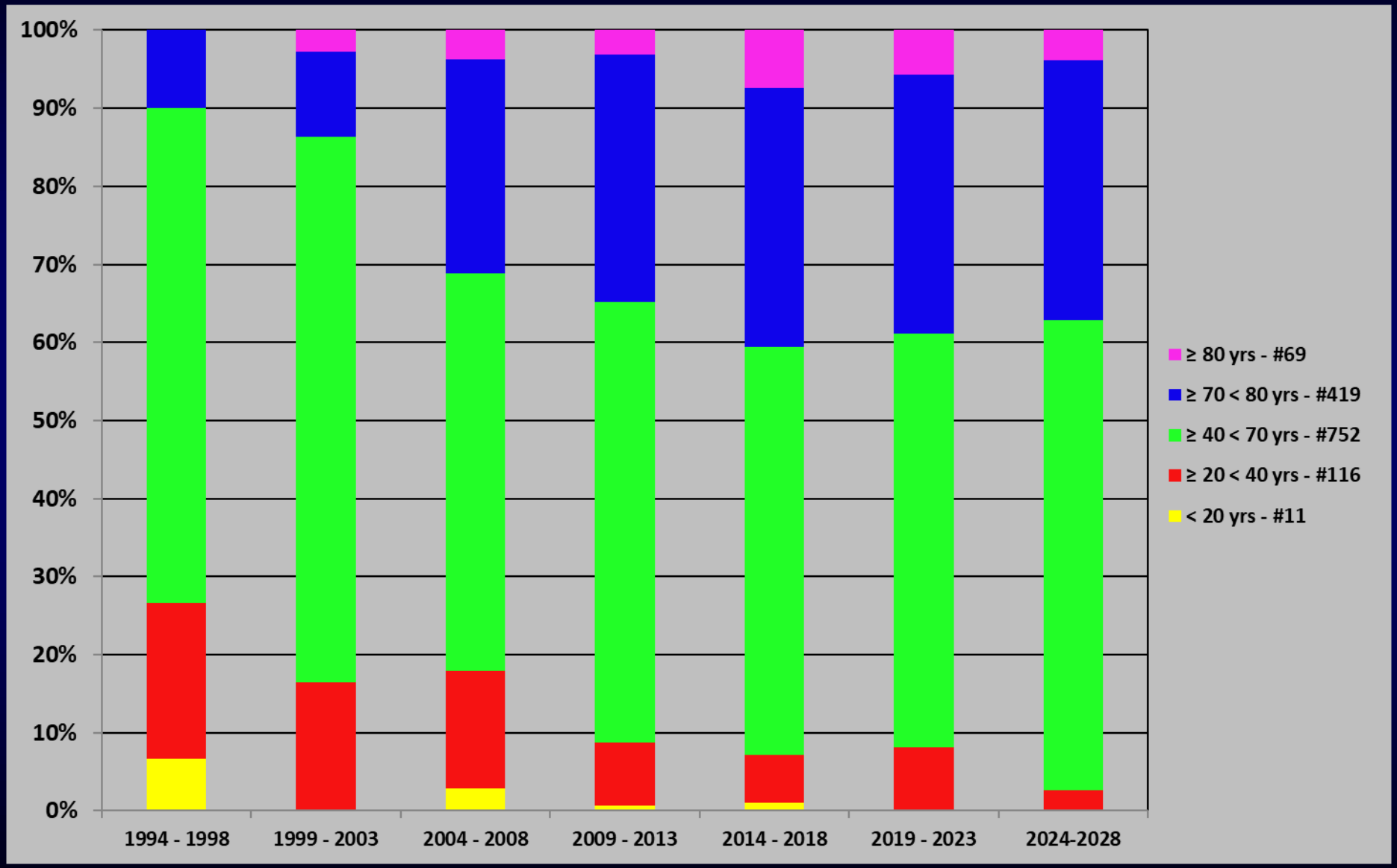
NEUROPSYCHOLOGICAL OUTCOMES

The main findings of our study show that repeated short periods of MHCA during PEA in patients with CTEPH did not result in any neuropsychological complications

More remarkably, we found a statistically significant postsurgical improvement in motor speed, which was accompanied by a better quality of life and reduced symptoms of depression and anxiety

AGE

OF 1367 PEAs



PAVIA CTEPH PROGRAM

January, 1st – December, 31st 2024 → 564

NewEvaluations (244 pts)

CTEPH-PEA FUP (320 pts)

CONFIRMED (155 pts - 64%)

- PROXIMAL LESIONS (118 pts)
  - 70 PEAs
  - 11 waiting for PEA
  - 14 refused evaluation for PEA
  - 5 refused PEA
  - 4 with severe co-morbidities
  - 5 died before evaluation or before PEA
  - 6 CTEPD
  - 2 bridge to candidacy
  - 1 too frigile

- DISTAL LESIONS / COMORBIDITIES / REFUSED / AFTER PEA (37 pts)
  - Medical therapy (15 pts)
    - 11 Mono Rio
    - 4 Combination Rio+Bos
  - BPA program (22 pts)
    - 12 BPAs
    - 10 waiting for BPA

OTHER DIAGNOSIS (89 pts - 36%)

- RECENT APE (10 pts)
  - 9 medical therapy
  - 1 surgical embolectomy
- PREVIOUS APE WITHOUT SIGNS (12 pts)
  - 12 medical therapy
- APE OVER CTE LESIONS (6 pts)
  - 6 3-month medical therapy → new evaluation
- MINIMAL CTE LESIONS WITHOUT/LOW PH (14 pts)
  - 14 medical therapy
- PULMONARY ARTERY SARCOMA (9 pts)
  - 7 PEAs
  - 1 inoperable
  - 1 died before evaluation or PEA
- PULMONARY ARTERY ANEURYSM (1 pt)
  - 1 medical therapy
- MISCELLANEOUS (37 pts)

OPERABILITY RATE

thecnically	***76%***
+ refused	***64%***
+ other	***52%***

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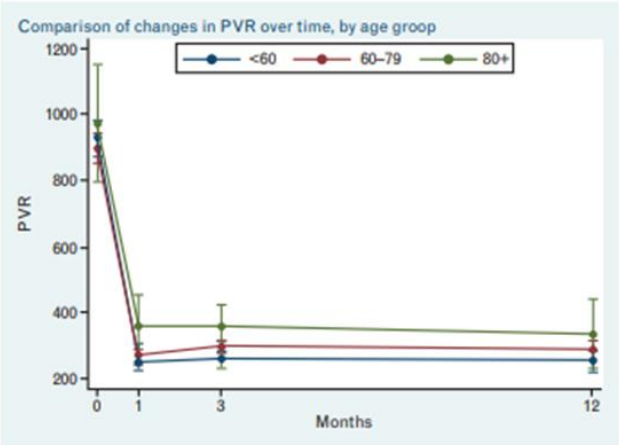
AGE

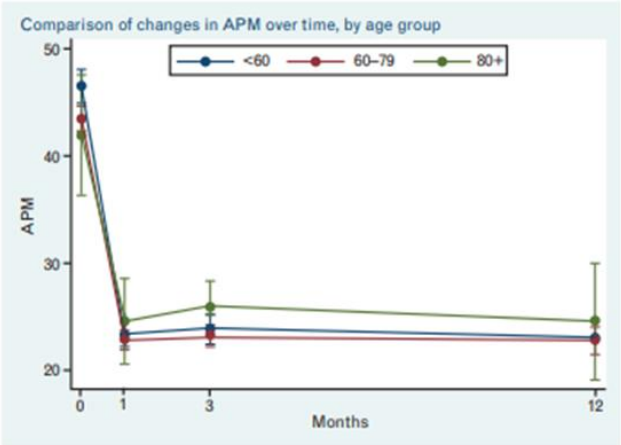
Pulmonary endarterectomy in the octogenarian population: safety and outcomes

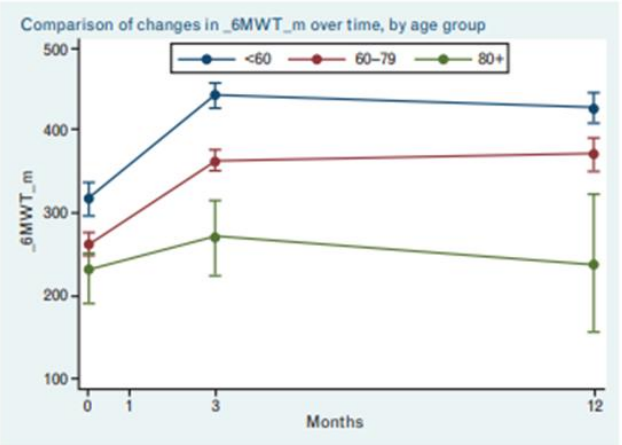
Valentina Grazioli[a], Stefano Ghio[b], Maurizio Pin[a], Antonio Sciortino[a], Anna Celentano[a], Giuseppe Silvaggio[a], Cristian Monterosso[a], Annalisa Turco[b], Catherine Klersy[c], Vera N. Merli[a], Benedetta Vanini[a,d] and Andrea M. D'Armini[a]

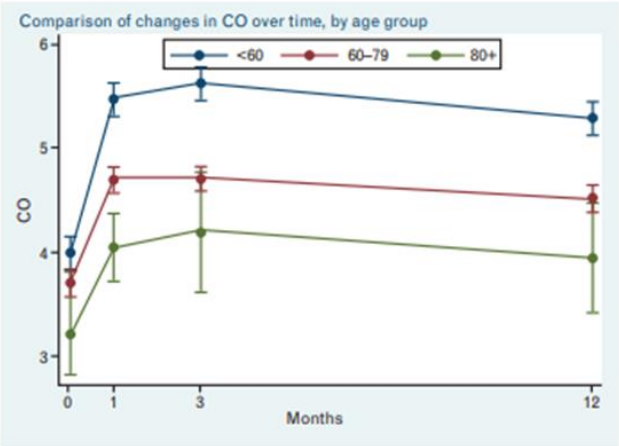
J Cardiovasc Med (Hagerstown)
2021 Jul 1;22(7):567-571.

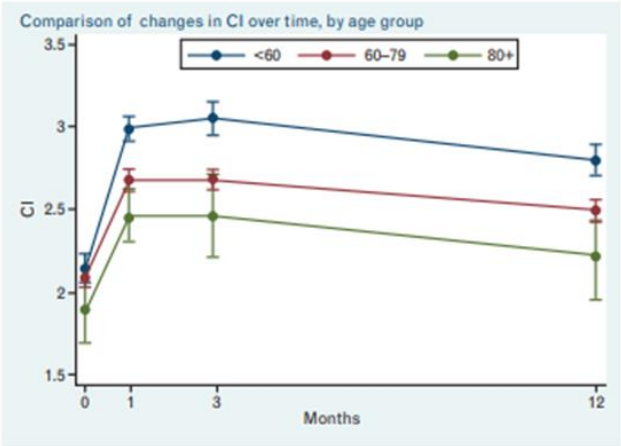
AGE

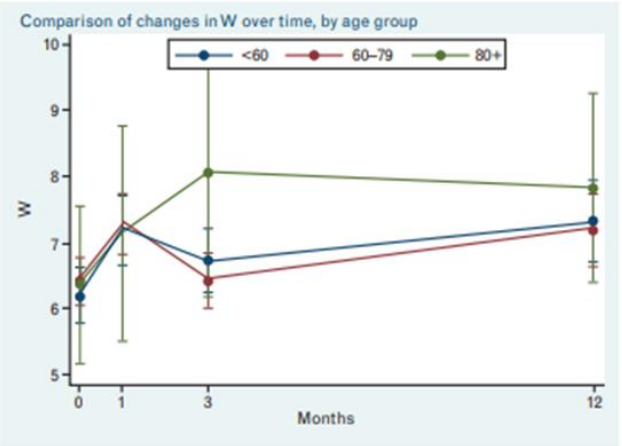












J Cardiovasc Med (Hagerstown). 2021 Jul 1;22(7):567-571.

PATIENTS SUITABLE FOR BPA - CLASSIFICATION

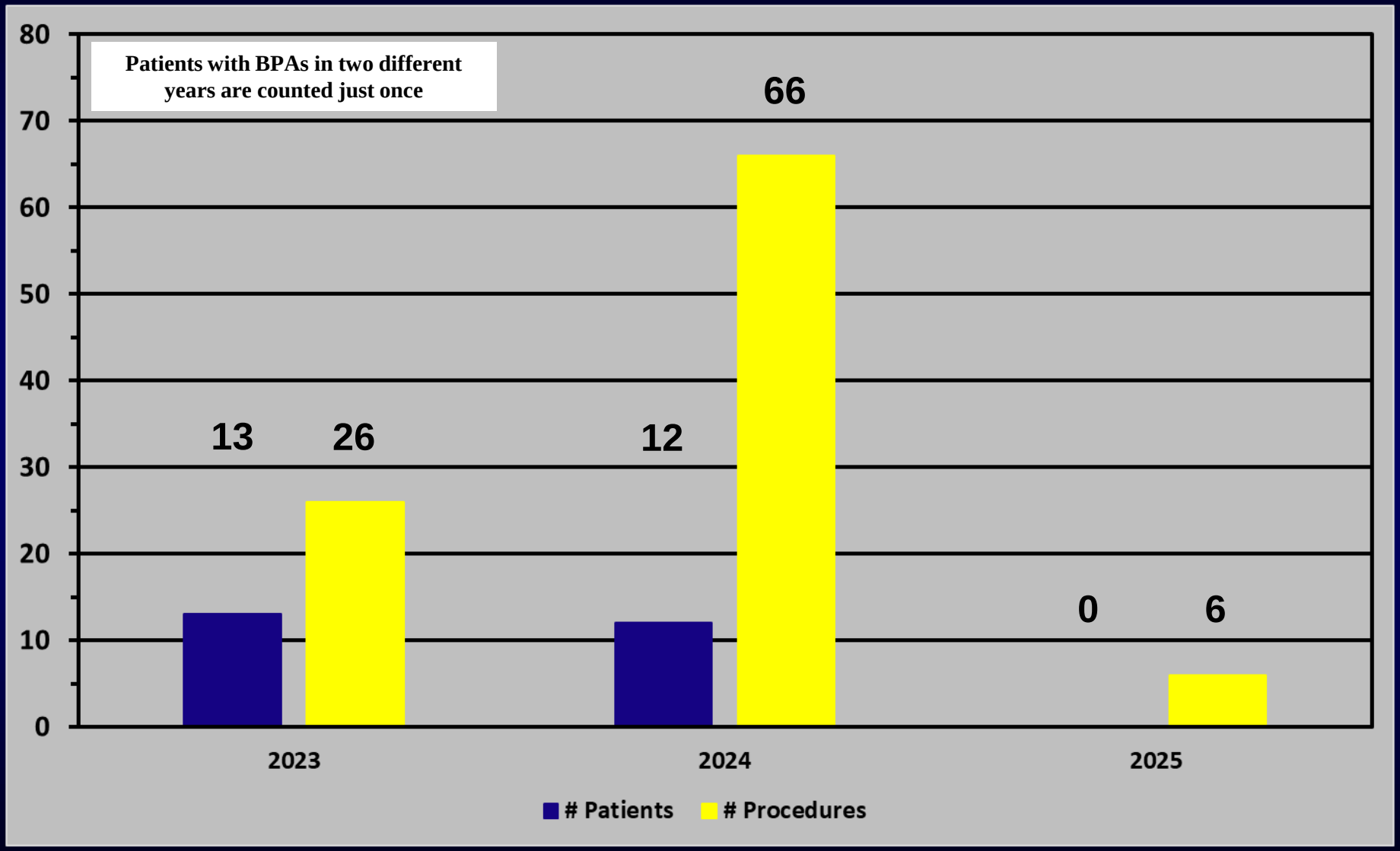
- Technically inoperable
- Excessive comorbidities
- Refuse PEA
- Post-PEA non suitable of redo-PEA

OUR NEW BPA's PROGRAM

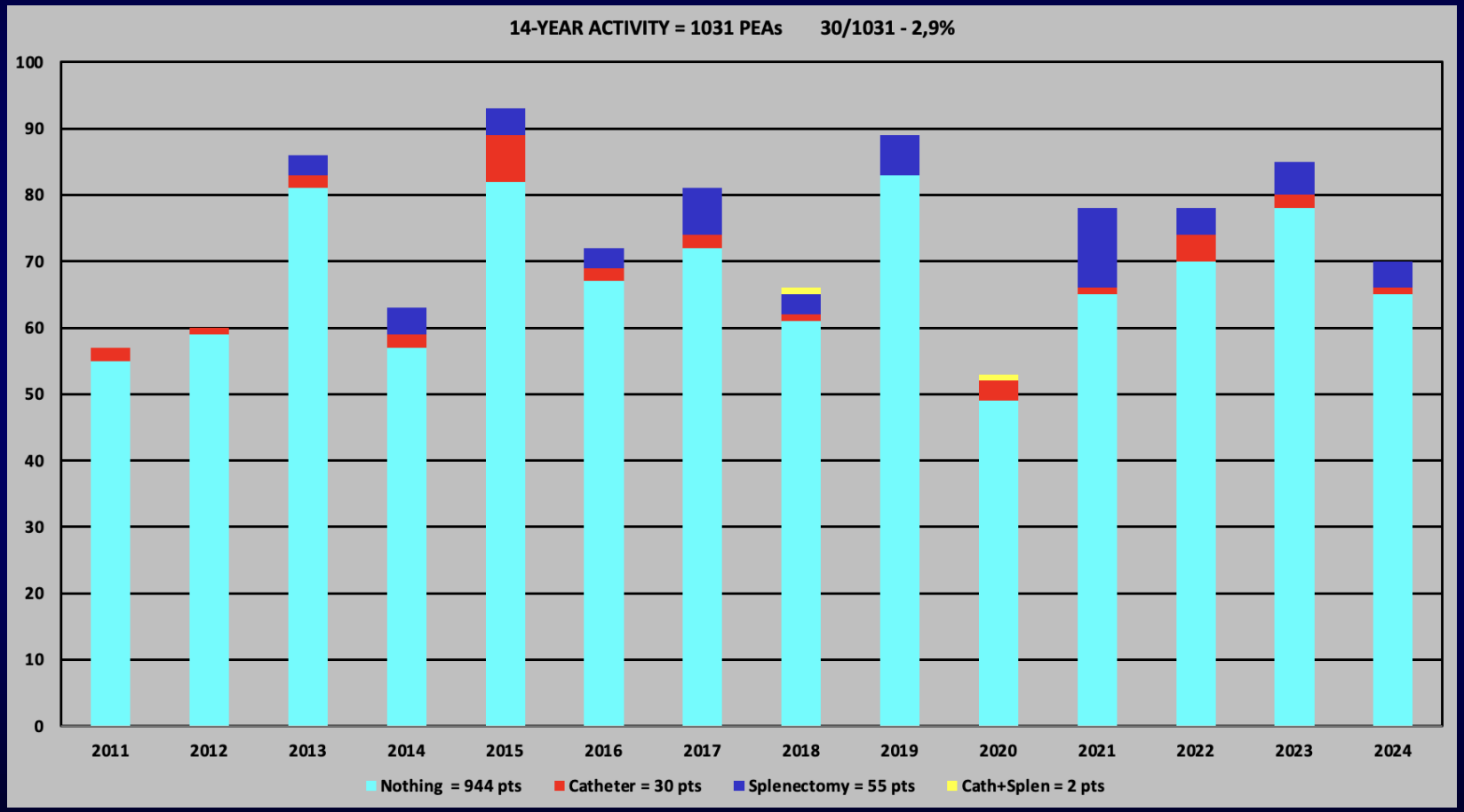
- National referral program
- Begin: March 2023
- January 2025: 25 pts with 98 BPAs performed

BPA: PATIENTS & PROCEDURES

25 & 98



CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT



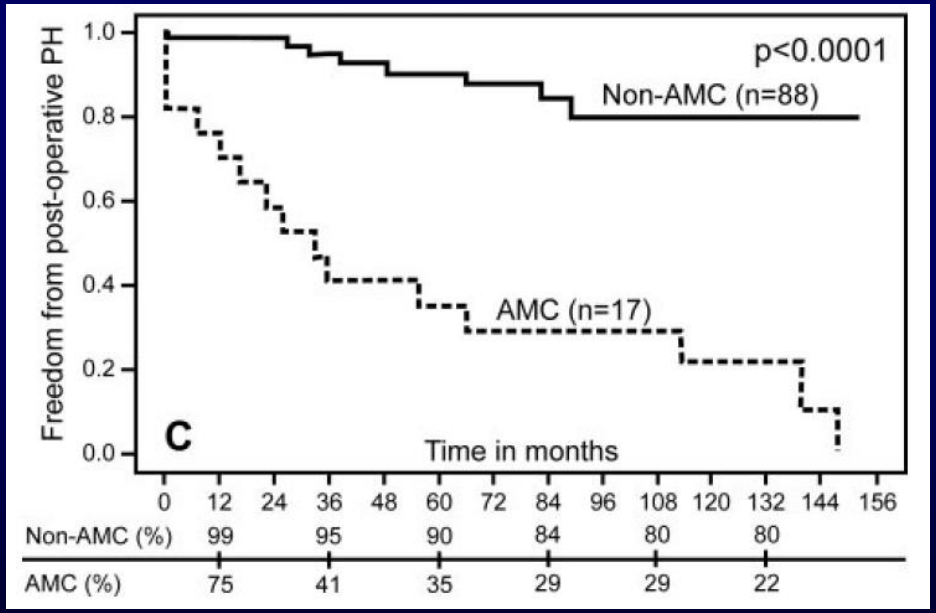
Predictors of Outcome in Chronic Thromboembolic Pulmonary Hypertension

Diana Bonderman, MD; Nika Skoro-Sajer, MD; Johannes Jakowitsch, PhD; Christopher Adlbrecht, MD; Daniela Dunkler, MSc; Sharokh Taghavi, MD; Walter Klepetko, MD; Meinhard Kneussl, MD; Irene M. Lang, MD

(*Circulation*. 2007;115:2153-2158.)

Associated Medical Conditions:
- splenectomy
- ventriculoatrial shunt
- chronic central venous line
- inflamatory bowel disease
- osteomyelitis

AMC hospital mortality 24%
Non-AMC hospital mortality 9%



CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

- M.N. 52-year-old man
- 1979 (12 yrs old) ventriculo-atrial shunt due to a non-communicating hydrocephalus
- 1989 (22 yrs old) post-traumatic splenectomy
- 19-month-hystory of worsening dyspnea before admission to our Center

CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

- September 2020 admission to our Center
- Thrombophilic conditions:
- MTHFR gene → C677T mutation
- ANA+

ONE RISK FACTOR FOR CTEPH:
- TROMBOPHILIA

DISTAL DISEASE:
- CATHETER
- SPLENECTOMY

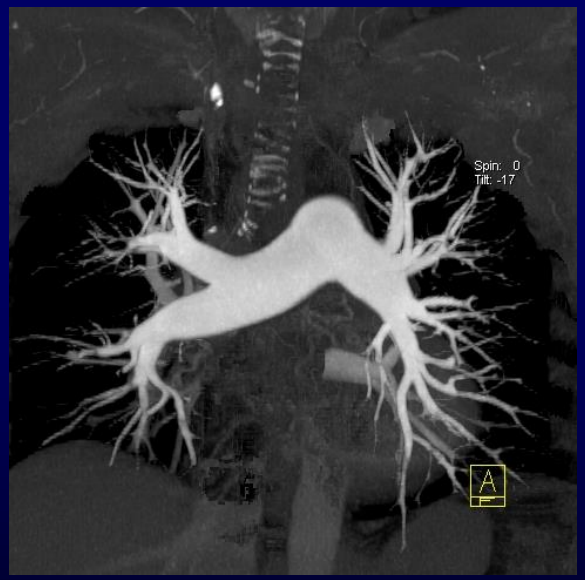
	BEFORE PEA
O_2 (L/min)	2
WHO	IV
PaO2 (mmHg)	65.3
6MWT (m)	70; interruption due to desaturation (O2-sat 83%)
sPAP (mmHg)	66
dPAP (mmHg)	27
mPAP (mmHg)	44
PVR (dyne*sec*cm^{-5})	655
CO (L/min)	3.3
CI (L/min/m^2)	1.8

CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

MIP RECONSTRUCTIONS BEFORE PEA

LATERAL VIEW RIGHT PA

FRONTAL VIEW R&L PA



LATERAL VIEW LEFT PA

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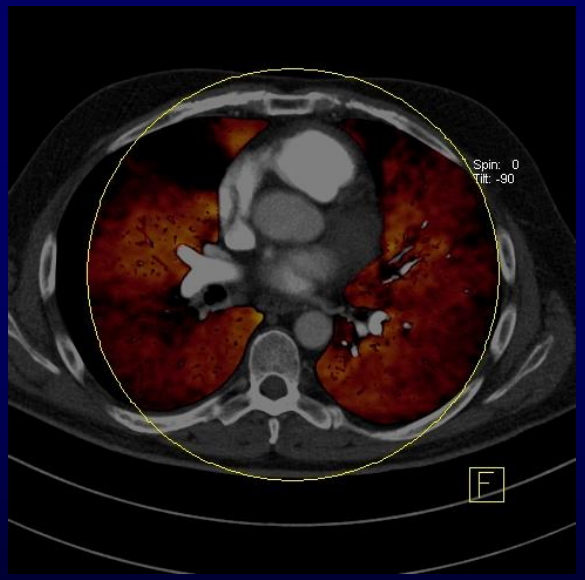
CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

DUAL SCAN BEFORE PEA

RIGHT LUNG - SAGITTAL

R&L LUNGS - CORONAL

R&L LUNGS - AXIAL



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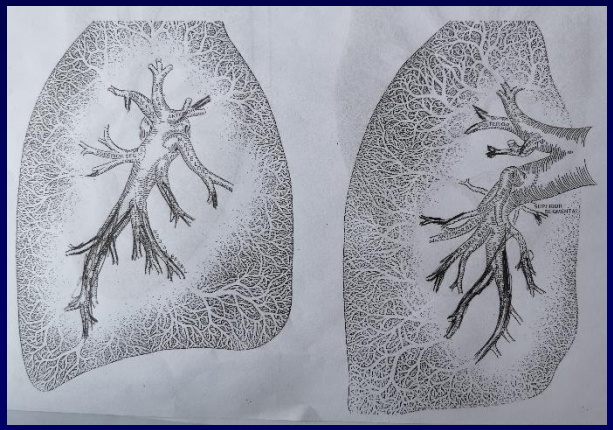
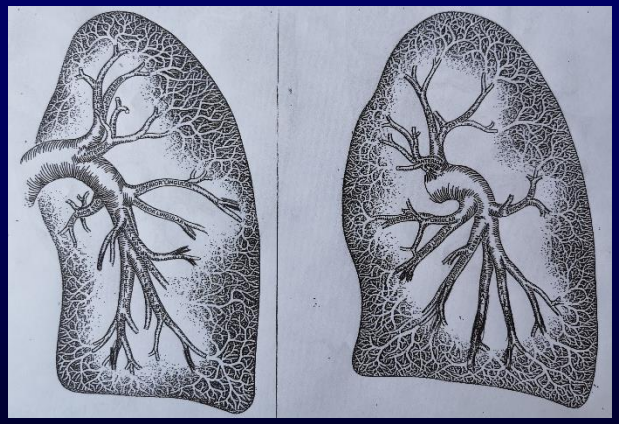
CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

- Confirmed CTEPH with distal disease (J3 on both lungs with very few lesions in the left lung)
- Presence of PFO
- Scheduled procedures: closure of PFO, removal of V-A catheter, right monolateral PEA
- 6-month FUP with check of anatomical, hemodynamic and functional results
- Consideration for potential BPA in the left lung if 6-month FUP results will be not optimal

RIGHT LUNG





LEFT LUNG

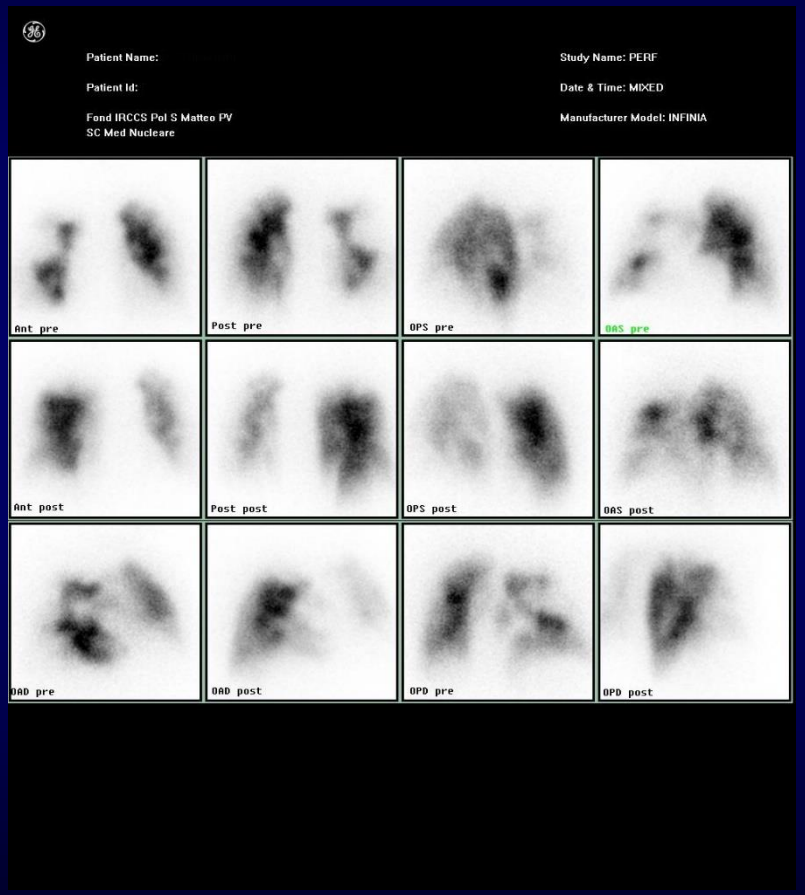
CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

- Small neck incision, VA catheter resection and connection with external drainage system
- RA incision, PFO closure, removal SVC-RA portion of VA catheter
- Right PEA with confirmed distal lesions (RUL J3; RML J4; RLL J3)
- Moderate hypotermia (23° C) with total circulator arrest 67 min and no aortic clamp / no use of cardioplegia (Pavia's protocol)
- Total treated / reopened branches 21 (6 RUL; 15 RLL) up to fifth grade of bifurcation (principal, lobar, segmental, first subsegmental and second subsegmental)
- Sternum left open with Teflon patch for right ventricle disfunction (closed on 2nd POD)
- Complete removal of VA shunt on 15th POD (brain CT before surgical removal shows no evidence of enlargement of the ventricular system with VA shunt closed)
- Discharged on 21st POD

CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

Perfusion scintigraphy is a morphological and functional exam

Before PEA	**Right Lung**	**38%**
	Left Lung	**62%**
6-month FUP	**Right Lung**	**74%**
	Left Lung	**26%**



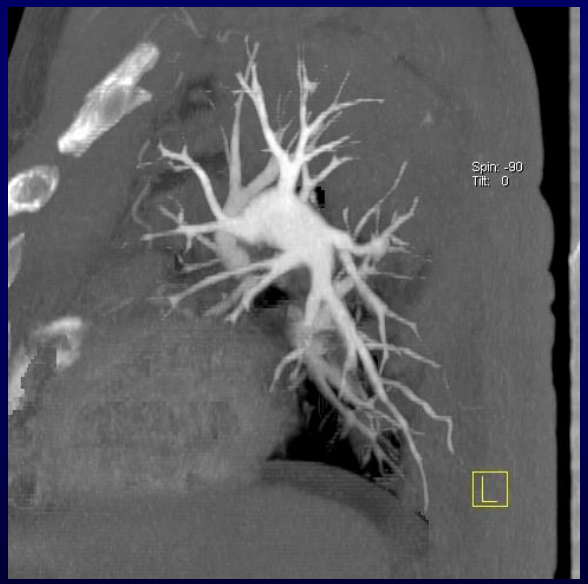
CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

MIP RECONSTRUCTIONS AT 6-MONTH FUP

LATERAL VIEW RIGHT PA

FRONTAL VIEW R&L PA

LATERAL VIEW LEFT PA



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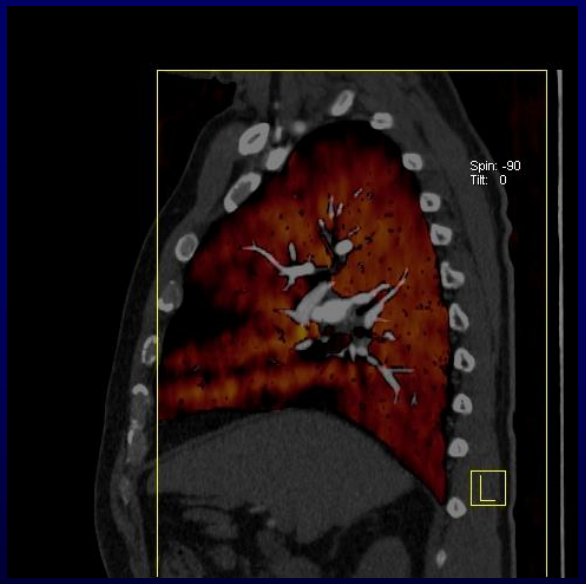
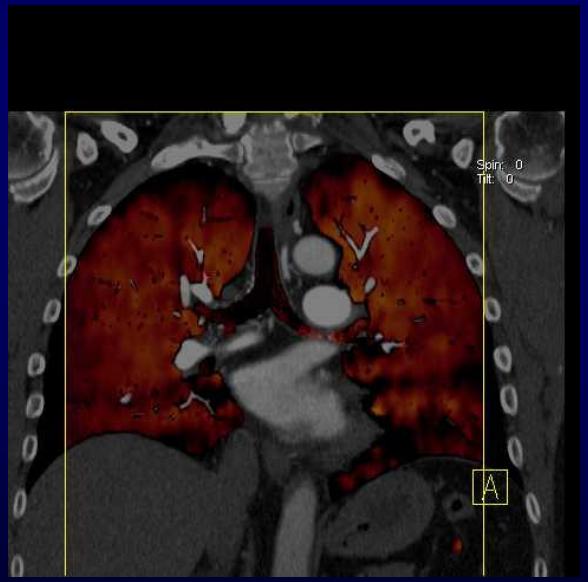
CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

DUAL SCAN AT 6-MONTH FUP

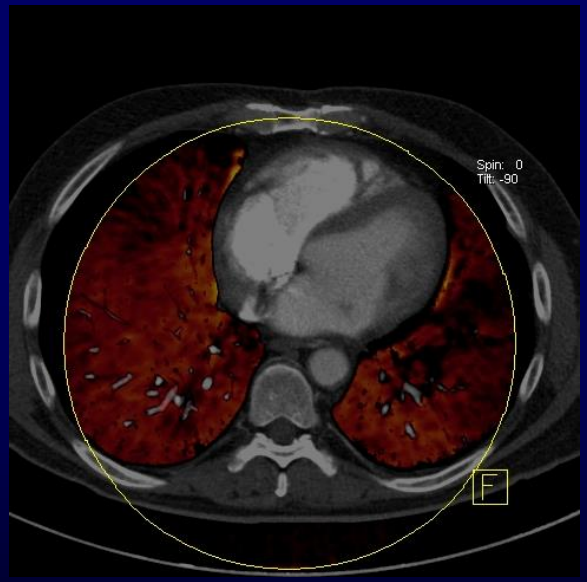
RIGHT LUNG - SAGITTAL



R&L LUNGS - CORONAL



R&L LUNGS - AXIAL



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CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

	BEFORE PEA	6-MONTH FUP	2-YEAR FOLLOW-UP
O_2 (L/min)	2	No	No
WHO	IV	I	I
PaO2 (mmHg)	65	89 +37%	89 +37%
6MWT (m)	70	450 +542%	490 +600%

CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

	BEFORE PEA	6-MONTH FUP	2 YEARS FUP
sPAP (mmHg)	66	36	27
dPAP (mmHg)	27	15	10
mPAP (mmHg)	42	24 -43%	18 -57%
PVR (dyne*sec*cm^{-5})	655	245 -63%	160 -76%
CO (L/min)	3.3	5.6	5
CI (L/min/m^2)	1.8	3.1 +70%	3 -43%

CASE REPORT
PEA IN CATHETER-INDUCED CTEPH PATIENT

- At 6-month FUP we already have WHO functional class I, normalization of pO2 (stop O2-therapy), adequate 6mwt distance and normal hemodynamic data → ***no need to consider BPA on the left lung***
- At 2-year FUP we confirm WHO functional class I, normalization of pO2, adequate 6mwt distance (490 m) and normal hemodynamic data