University of Pavia - School of Medicine
Foundation I.R.C.C.S. Policlinico "San Matteo"
Cardiac Surgery - Intrathoracic Transplantation - Pulmonary Hypertension
Pavia, Italy

Prof. Andrea M. D'Armini, M.D.

TECHNIQUE AND OUTCOMES OF PULMONARY ENDARTERECTOMY SURGERY. HOW TO SELECT THE RIGHT PATIENT?

FINANCIAL DISCLOSURE

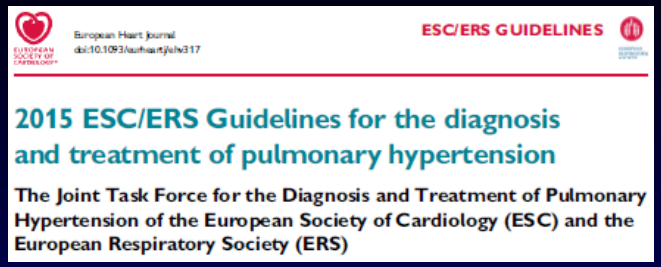
Last three years

Actelion Pharmaceuticals Ltd

Bayer HealthCare

Merk Sharp Dohme

PULMONARY ENDARTERECTOMY: THE PAVIA EXPERIENCE


European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

1. Pulmonary arterial hypertension

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2 mutation
  - 1.2.2 Other mutations
- 1.3 Drugs and toxins induced
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 Human immunodeficiency virus (HIV) infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease (Table 6)
  - 1.4.5 Schistosomiasis

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

- 1'.1 Idiopathic
- 1'.2 Heritable
  - 1'.2.1 EIF2AK4 mutation
  - 1'.2.2 Other mutations
- 1'.3 Drugs, toxins and radiation induced
- 1'.4 Associated with:
  - 1'.4.1 Connective tissue disease
  - 1'.4.2 HIV infection

1''. Persistent pulmonary hypertension of the newborn

2. Pulmonary hypertension due to left heart disease

- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital / acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
- 2.5 Congenital /acquired pulmonary veins stenosis

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 lung diseases (Web Table 11)

4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions

- 4.1 Chronic thromboembolic pulmonary hypertension
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Angiosarcoma
  - 4.2.2 Other intravascular tumors
  - 4.2.3 Arteritis
  - 4.2.4 Congenital pulmonary arteries stenoses
  - 4.2.5 Parasites (hydatidosis)

5. Pulmonary hypertension with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders, sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: pulmonary tumoral thrombotic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension

4%

INTRODUCTION

- *Chronic thromboembolic pulmonary hypertension (CTEPH)* represents the *only* type of pulmonary hypertension surgically treatable, in the majority of cases, without transplant

- This life-saving conservative surgery is called *pulmonary endarterectomy (PEA)*

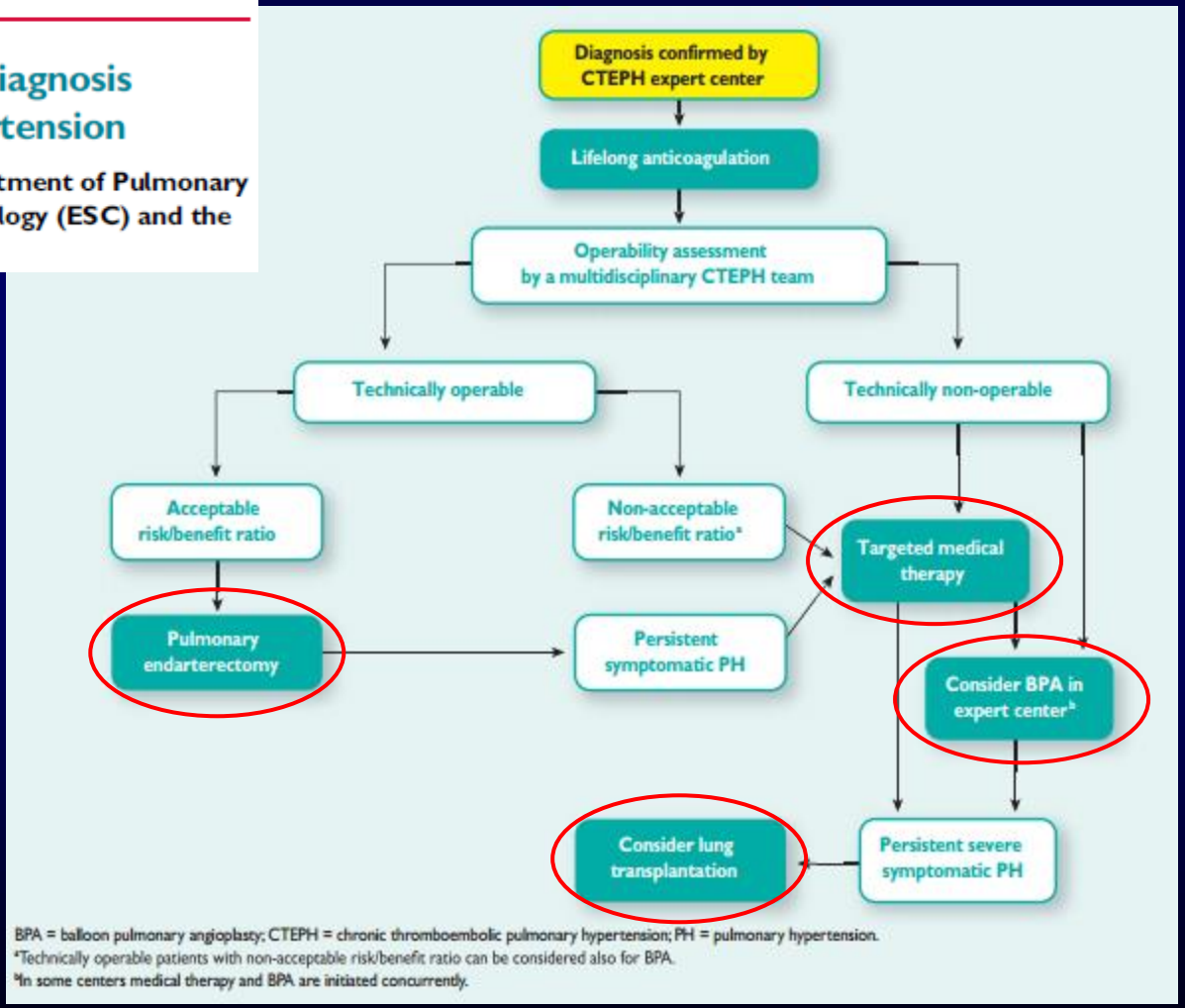
LTx IN THE GUIDELINES FOR CTEPH

European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)



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.

SURGICAL TREATMENT OF CTEPH

First HLTx for CTEPH

First PEA

First DLTx for CTEPH

Reverse right ventricular remodeling after pulmonary endarterectomy

Andrea M. D'Armini, MD,[a] Giorgio Zanotti, MD,[a] Stefano Ghio, MD,[b] Giulia Magrini, MD,[b] Matteo Pozzi, MD,[a] Laura Scelsi, MD,[b] Giulia Meloni, MD,[c] Catherine Klersy, MD,[d] and Mario Viganò, MD[a]

J Thorac Cardiovasc Surg 2007;133:162-8

First PEA in patient listed for DLTx

First PEA in patient previously enrolled in RCT for inoperability

08-MAR-1991

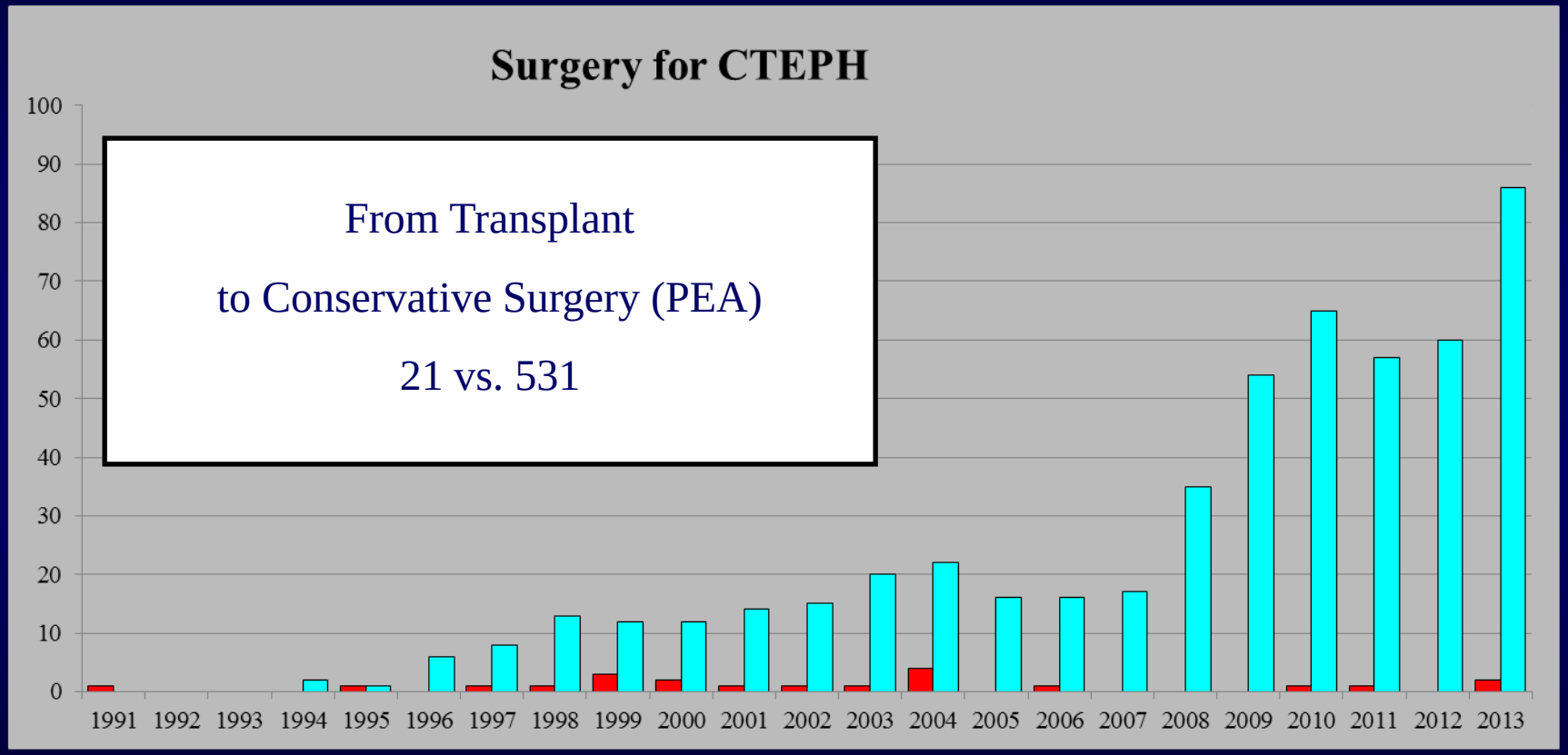
11-APR-1994

25-DEC-1995

28-JUL-2003

30-NOV-2009

SURGICAL TREATMENT OF CTEPH



1991 — 22 yrs — 2013

SURGICAL TREATMENT OF CTEPH

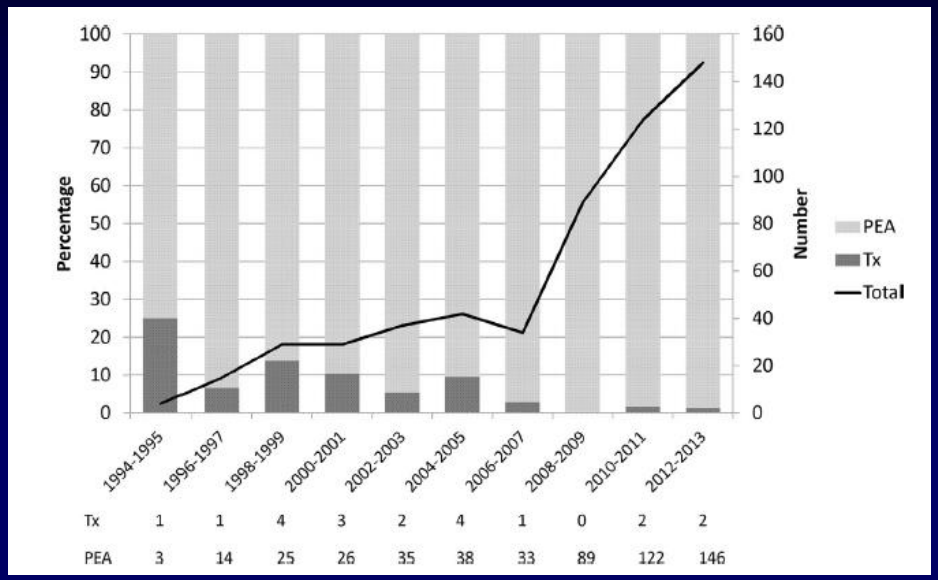
Chronic thromboembolic pulmonary hypertension: From transplantation to distal pulmonary endarterectomy

Andrea M. D'Armini, MD,[a]
Marco Morsolini, MD, PhD,[b]
Gabriella Mattiucci, MD,[b]
Valentina Grazioli, MD,[a]
Maurizio Pin, MD,[a] Antonio Sciortino, MD,[a]
Eloisa Arbustini, MD,[c] Claudio Goggi, MD,[a] and
Mario Viganò, MD[a]

The Journal of Heart and Lung Transplantation

J Heart Lung Transplant. 2016 Jan 6. pii: S1053-2498(16)00024-3

SURGICAL TREATMENT OF CTEPH



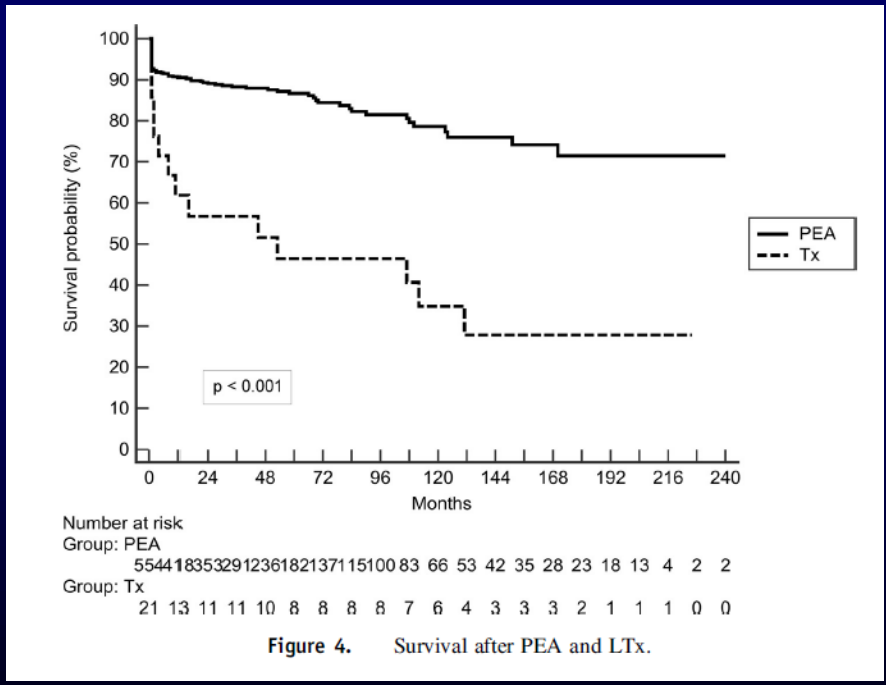
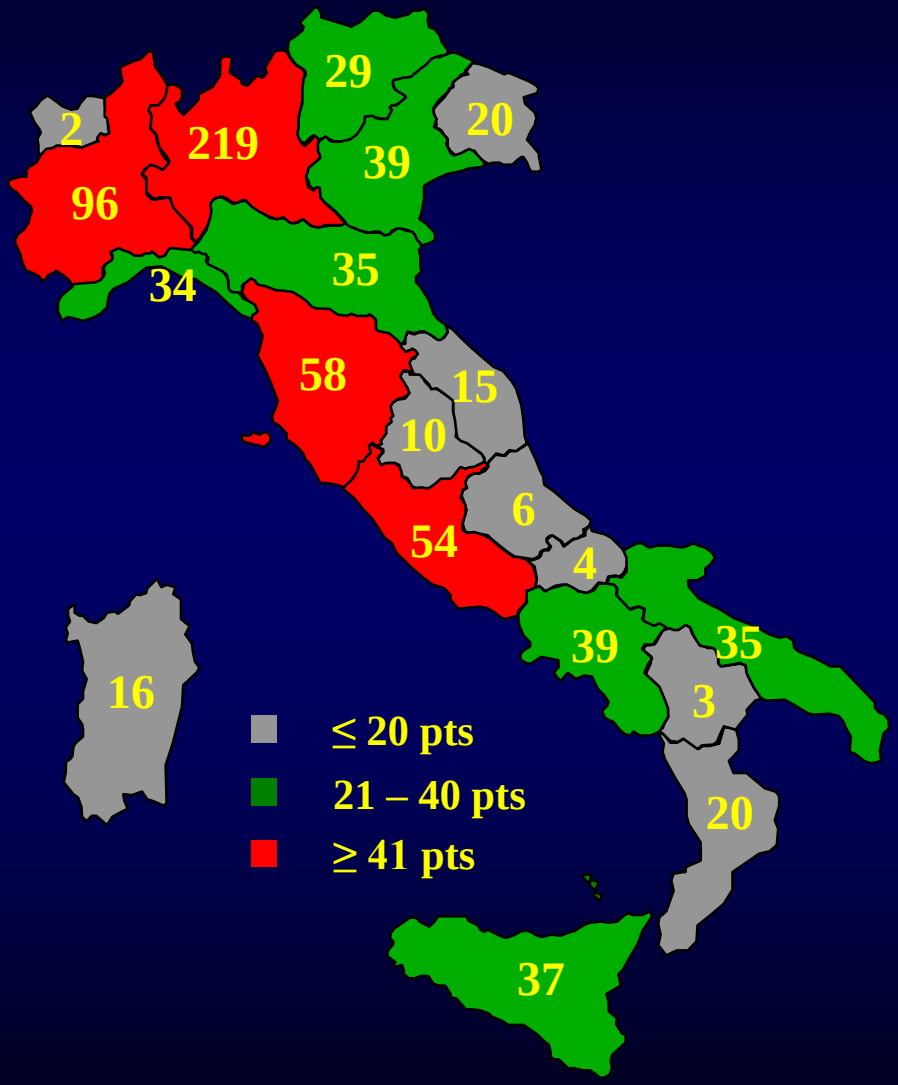


Figure 4. Survival after PEA and LTx.

PATIENTS' REFERRAL



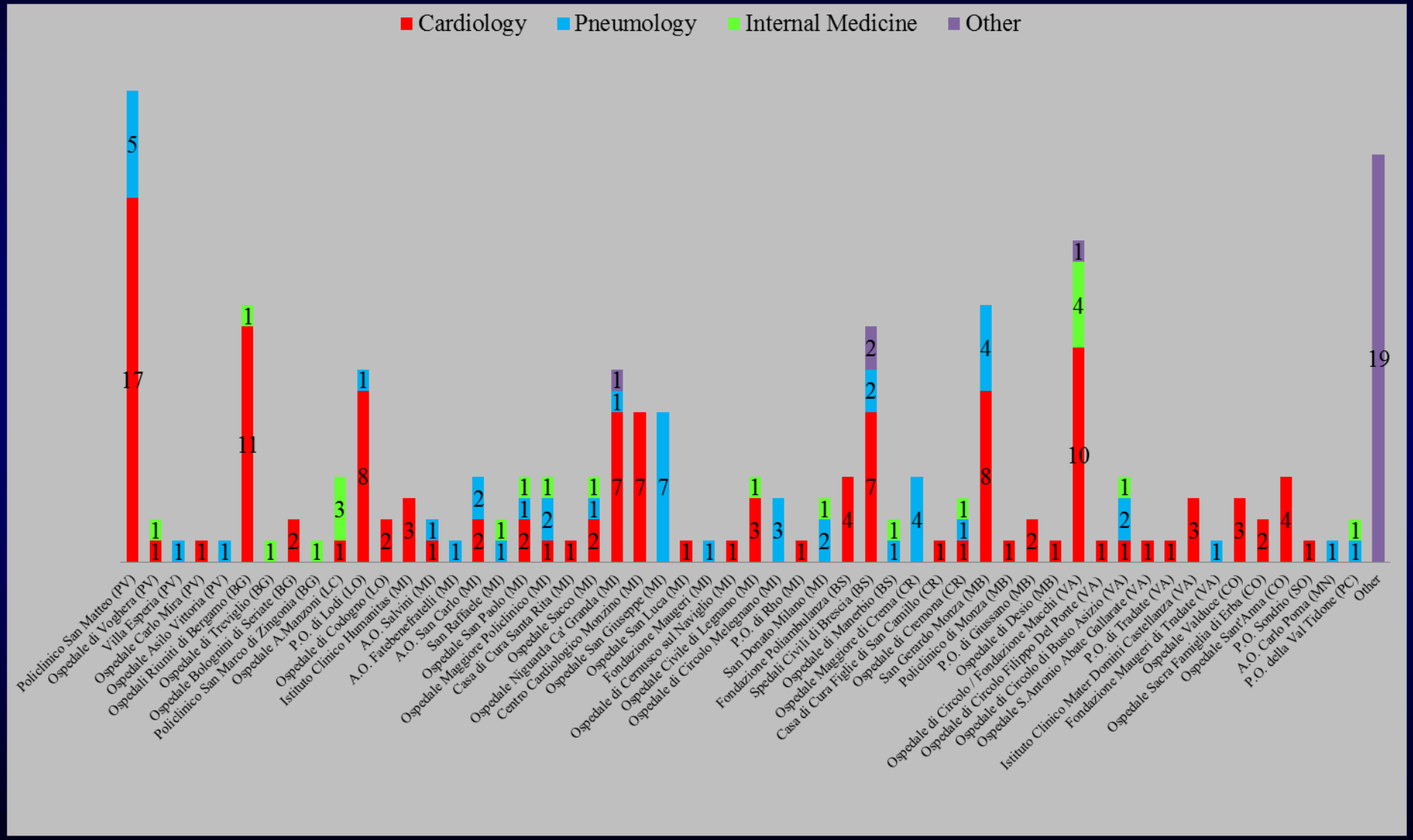
FROM 4/1994 TO 3/2017
780 PEAs

Pts coming from outside Italy	
- Albania	1
- Greece	1
- Israel	1
- Kosovo	1
- Romania	2
- Russia	1
- Uganda	1
- U.S.A.	1

PULMONARY ENDARTERECTOMY: THE PAVIA EXPERIENCE

PATIENTS' REFERRAL FROM LOMBARDIA

FROM 4/1994 TO 3/2017 - 219 PEAs



INDICATIONS FOR SURGERY

- The indications for the *surgical treatment* of these patients are based on

CLINIC

HEMODYNAMIC

- The indications for the *type of surgery* are based on

ANATOMY

CLINIC

- *CTEPH* patients *must be* in *NYHA functional class III or IV* before being *evaluated for PEA!*
- *Since 2003* we have performed *PEA* in *NYHA functional class II* patients, given the *natural history of CTEPH* …and the *good results of PEA…*

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-

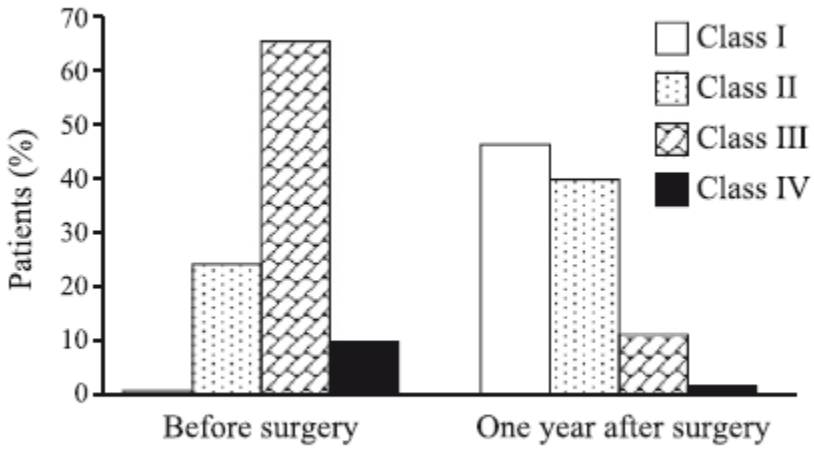
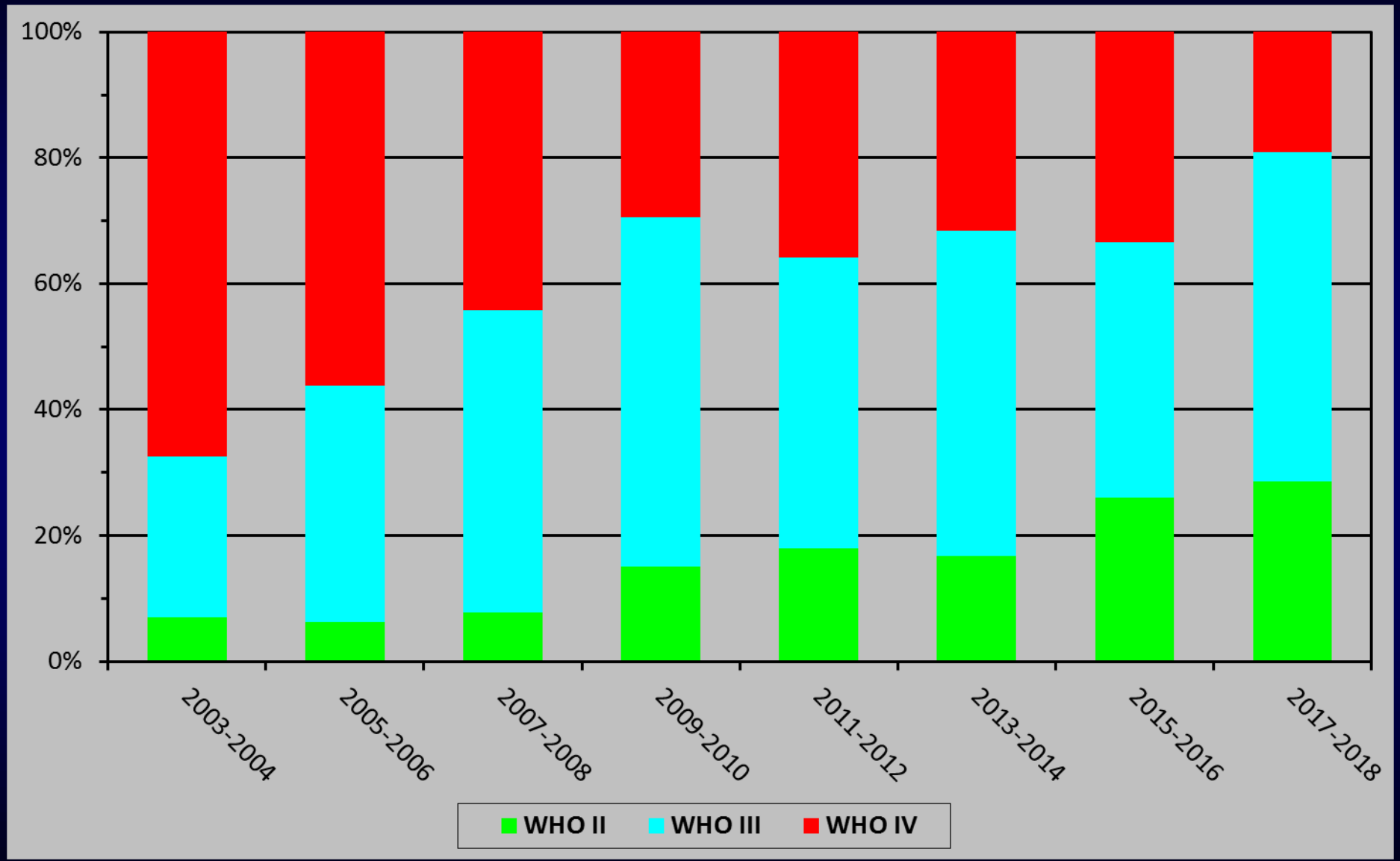


FIGURE 1. NYHA functional class before surgery and within 1 year after surgery (n = 221).

NYHA CLASS DISTRIBUTION 2007-2008

	INTERNATIONAL REGISTRY	PAVIA
II	25%	5%
III	65%	50%
IV	10%	45%

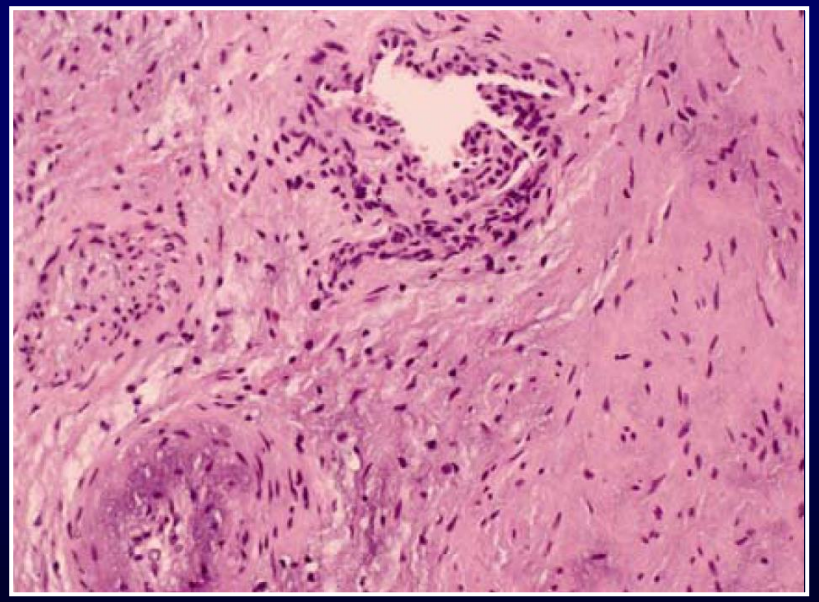
WHO CLASS DISTRIBUTION



PATHOPHYSIOLOGY

ACCORDING TO THE LENGTH OF THE DISEASE

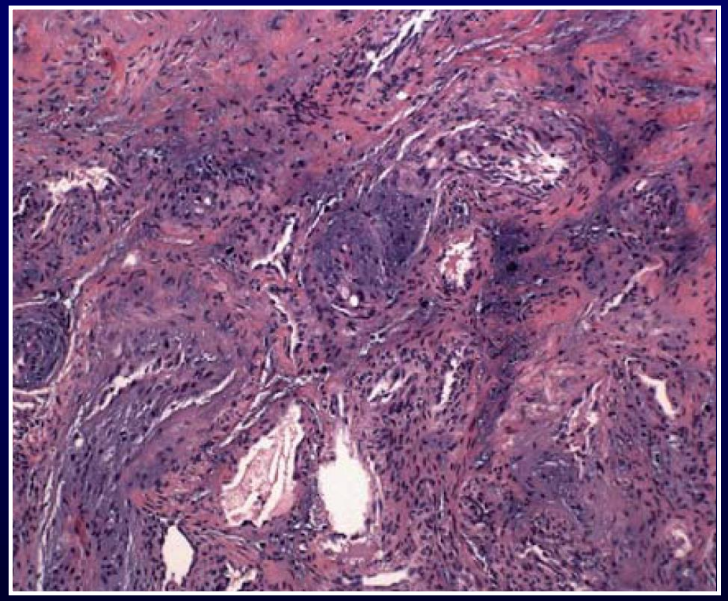
1. Hypertensive remodeling of the patent pulmonary vascular bed *(Eisenmenger-like)* due to volume and pressure overload



PATHOPHYSIOLOGY

ACCORDING TO THE LENGTH OF THE DISEASE

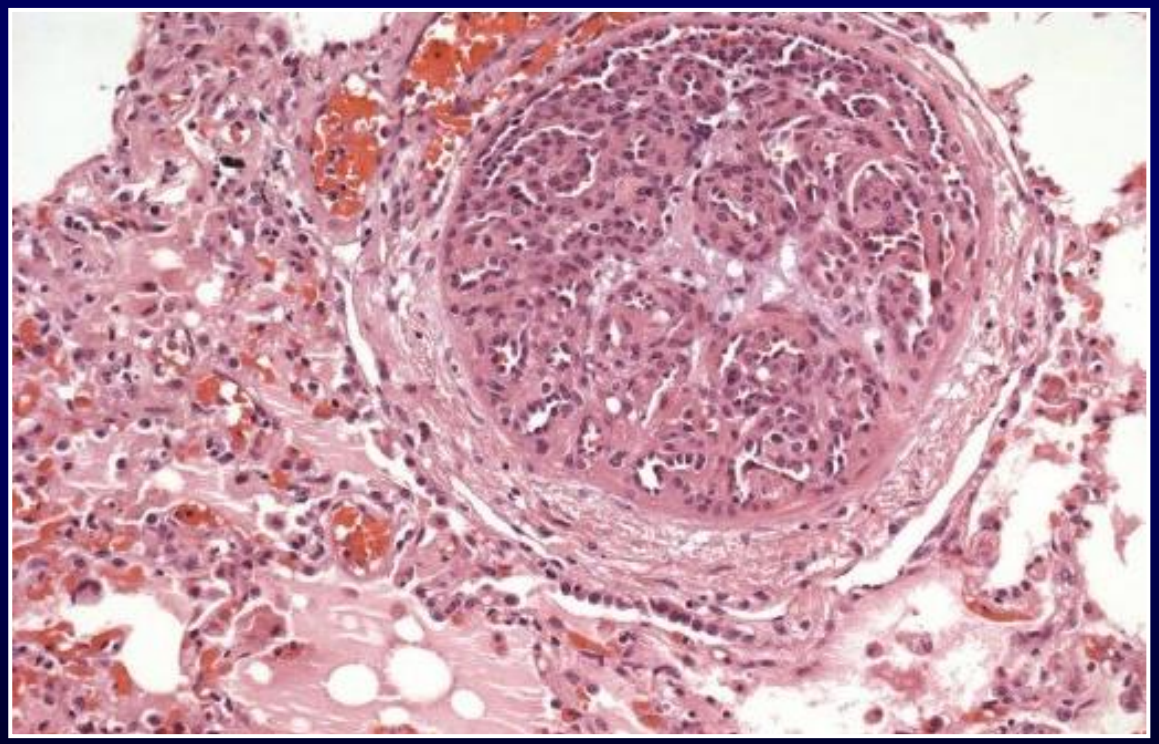
2. Chronic arteriopathy of the obstructed branches with *calcifications* and possible *retraction* of the distal vessels



PATHOPHYSIOLOGY

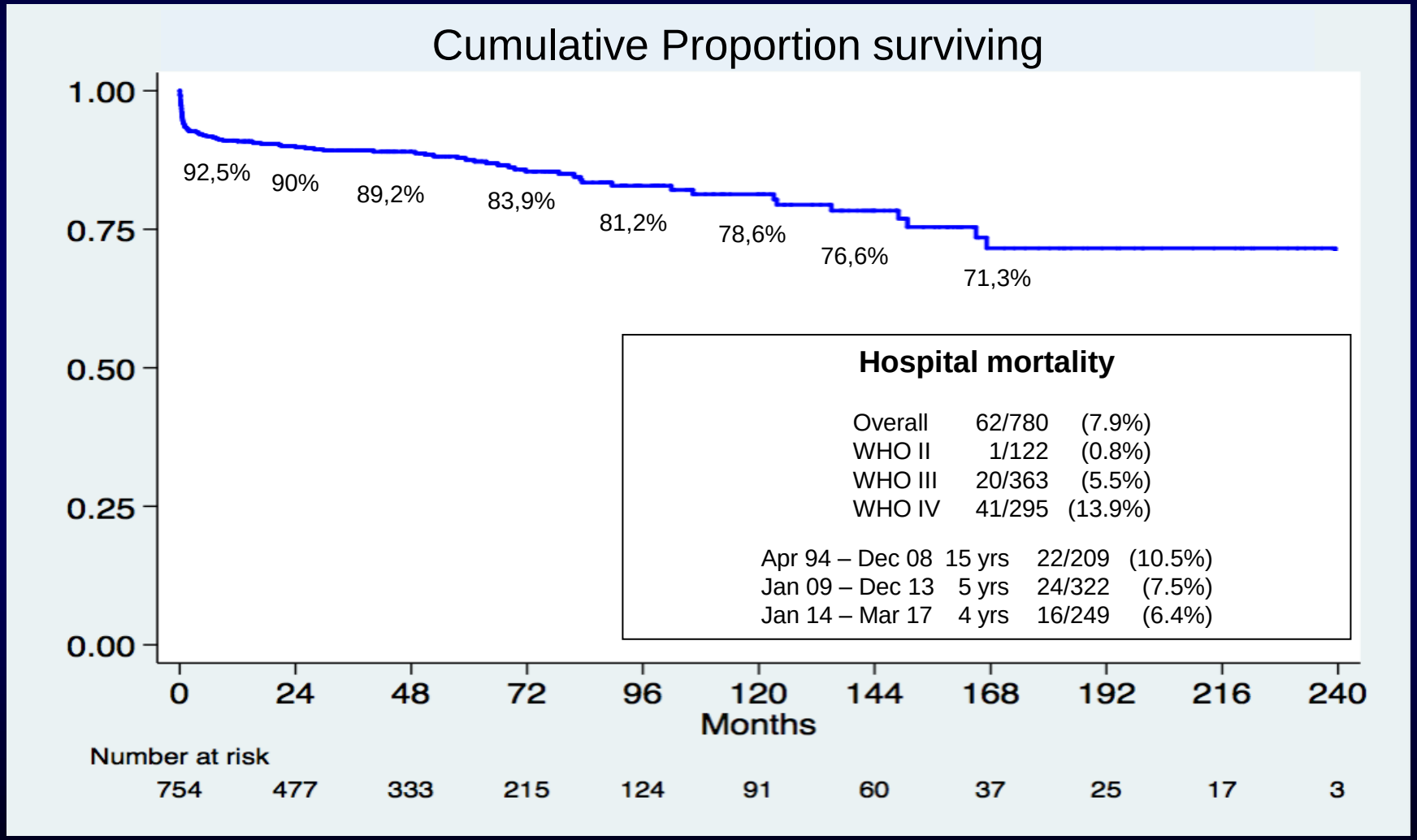
ACCORDING TO THE LENGTH OF THE DISEASE

3. *Plexiform lesions* stemming from the capillary bed



CUMULATIVE PROPORTION SURVIVING

OF 780 PEAs



CLINIC

- Full anticoagulation for *at least 3 months*

INDICATIONS FOR SURGERY

THREE MONTHS OF ANTICOAGULATION THERAPY

ACQUIRED CARDIOVASCULAR DISEASE

Surgical management and outcome of patients with chronic thromboembolic pulmonary hypertension: Results from an international prospective registry

Eckhard Mayer, MD,[a] David Jenkins, FRCS,[b] Jaroslav Lindner, MD,[c] Andrea D'Armini, MD,[d] Jaap Kloek, MD,[e] Bart Meyns, MD,[f] Lars Bo Ilkjaer, MD,[g] Walter Klepetko, MD,[h] Marion Delcroix, MD,[f] Irene Lang, MD,[h] Joanna Pepke-Zaba, MD,[b] Gerald Simonneau, MD,[i] and Philippe Dartevelle, MD[j]

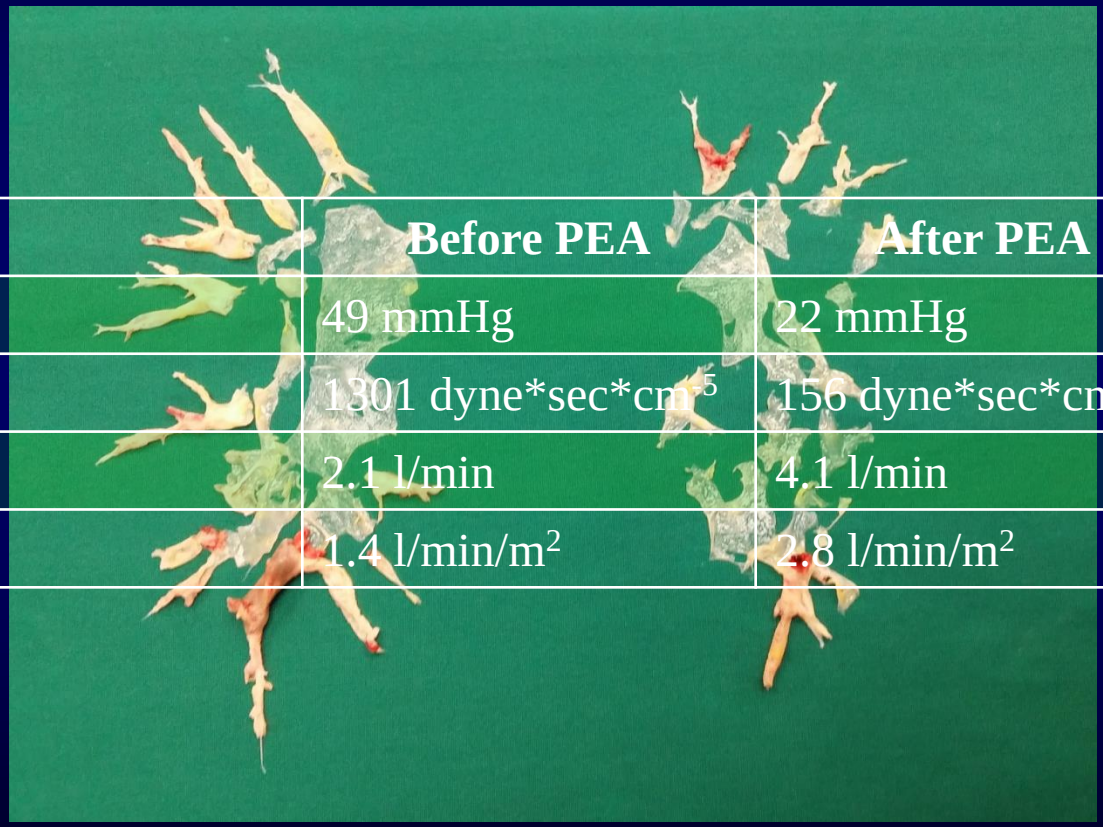
(J Thorac Cardiovasc Surg 2011;141:702-10)

Inclusion Criteria

At all participating institutions, the diagnosis of CTEPH was established according to clinical guidelines valid at study initiation[11] and within 6 months of inclusion in the registry. To qualify for inclusion, patients were to be 18 years or older and to have established pulmonary hypertension as confirmed by mean pulmonary artery pressure (mPAP) 25 mm Hg or greater at rest or 30 mm Hg or greater after exercise and pulmonary capillary wedge pressure 15 mm Hg or less during a right heart catheterization. CTEPH was to be confirmed as the cause of pulmonary hypertension by abnormalities in ventilation/perfusion scan (including at least 1 mismatched segmental perfusion defect), computed tomography (CT) scan, or pulmonary angiography. Abnormal CT scan/pulmonary angiography demonstrated proximal lesions (webs, bands, and narrowed vessels). Before diagnosis, patients were required to have at least 3 months of anticoagulation therapy and no PAH-specific treatment.

INDICATIONS FOR SURGERY

THREE MONTHS OF ANTICOAGULATION THERAPY



	Before PEA	After PEA
mPAP	49 mmHg	22 mmHg
PVR	1301 dyne*sec*cm^{-5}	156 dyne*sec*cm^{-5}
CO	2.1 l/min	4.1 l/min
CI	1.4 l/min/m^2	2.8 l/min/m^2

INDICATIONS FOR SURGERY

- The indications for the *surgical treatment* of these patients are based on

CLINIC

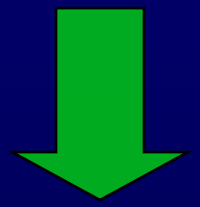
HEMODYNAMIC

- The indications for the *type of surgery* are based on

ANATOMY

HEMODYNAMIC

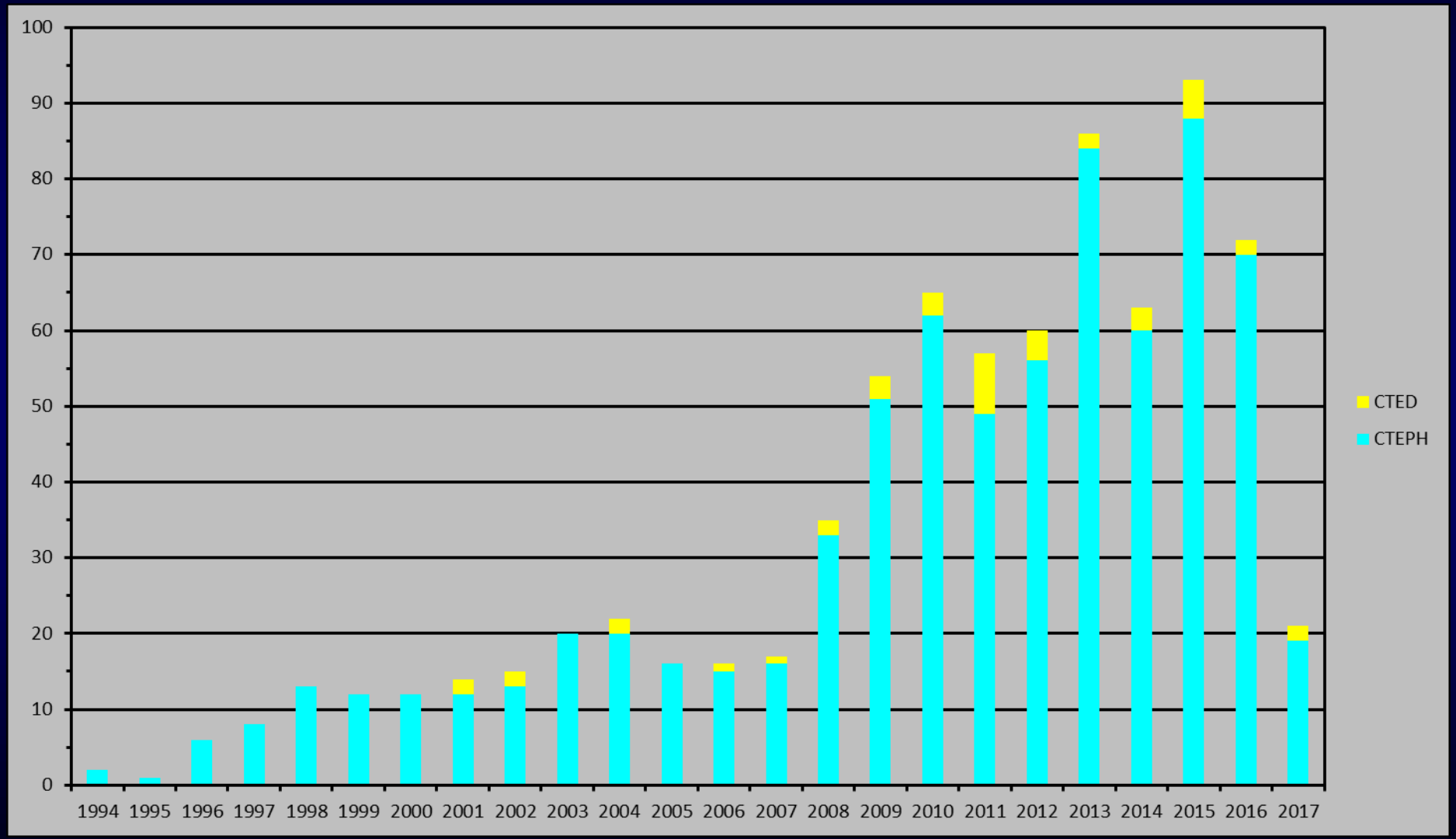
- Pulmonary hypertension (mPAP ≥ 25 mmHg)
- Causing low cardiac output



- Resulting in calculated pulmonary vascular resistances (PVR) > *300 dyne*sec*cm^{-5}*

CTEPH & CTED PATIENTS

738 & 42 → 780 PEAs



INDICATIONS FOR SURGERY

- The indications for the *surgical treatment* of these patients are based on

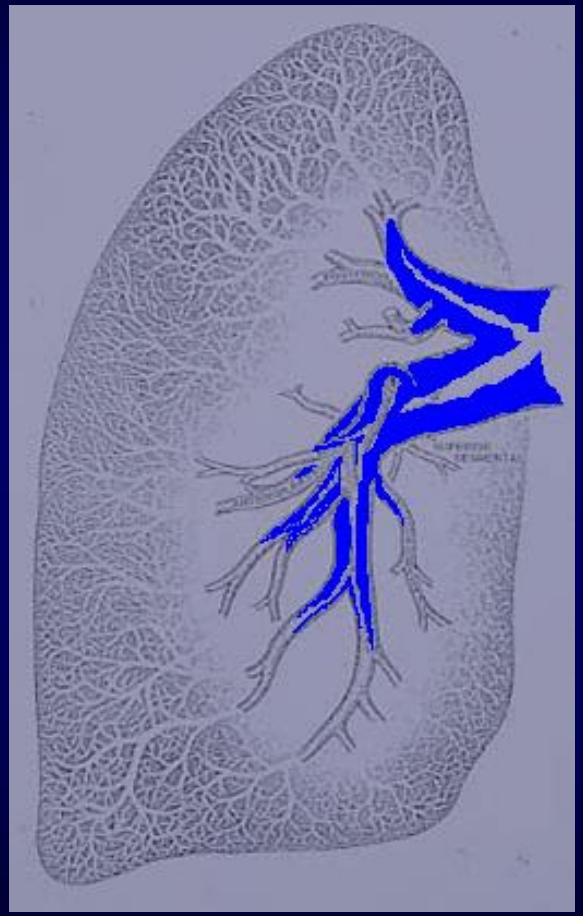
CLINIC

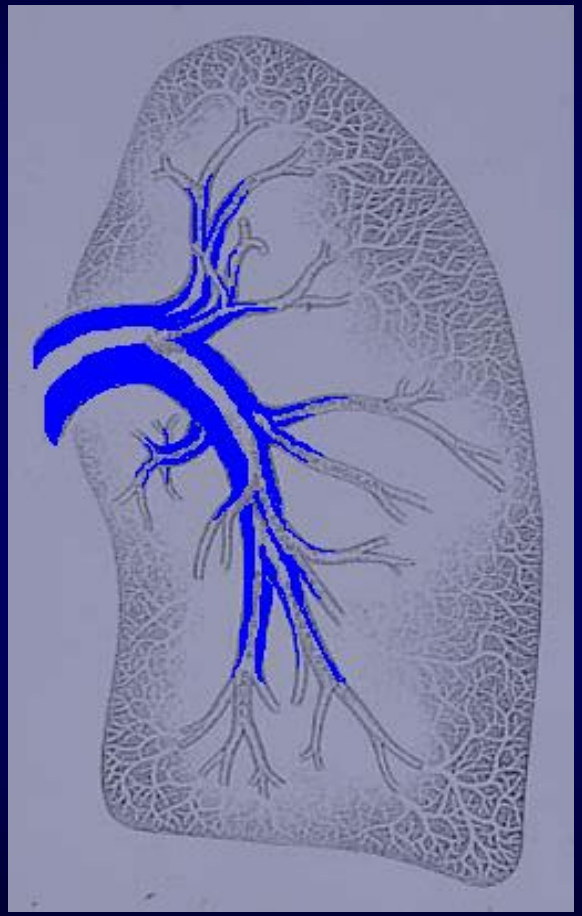
HEMODYNAMIC

- The indications for the *type of surgery* are based on

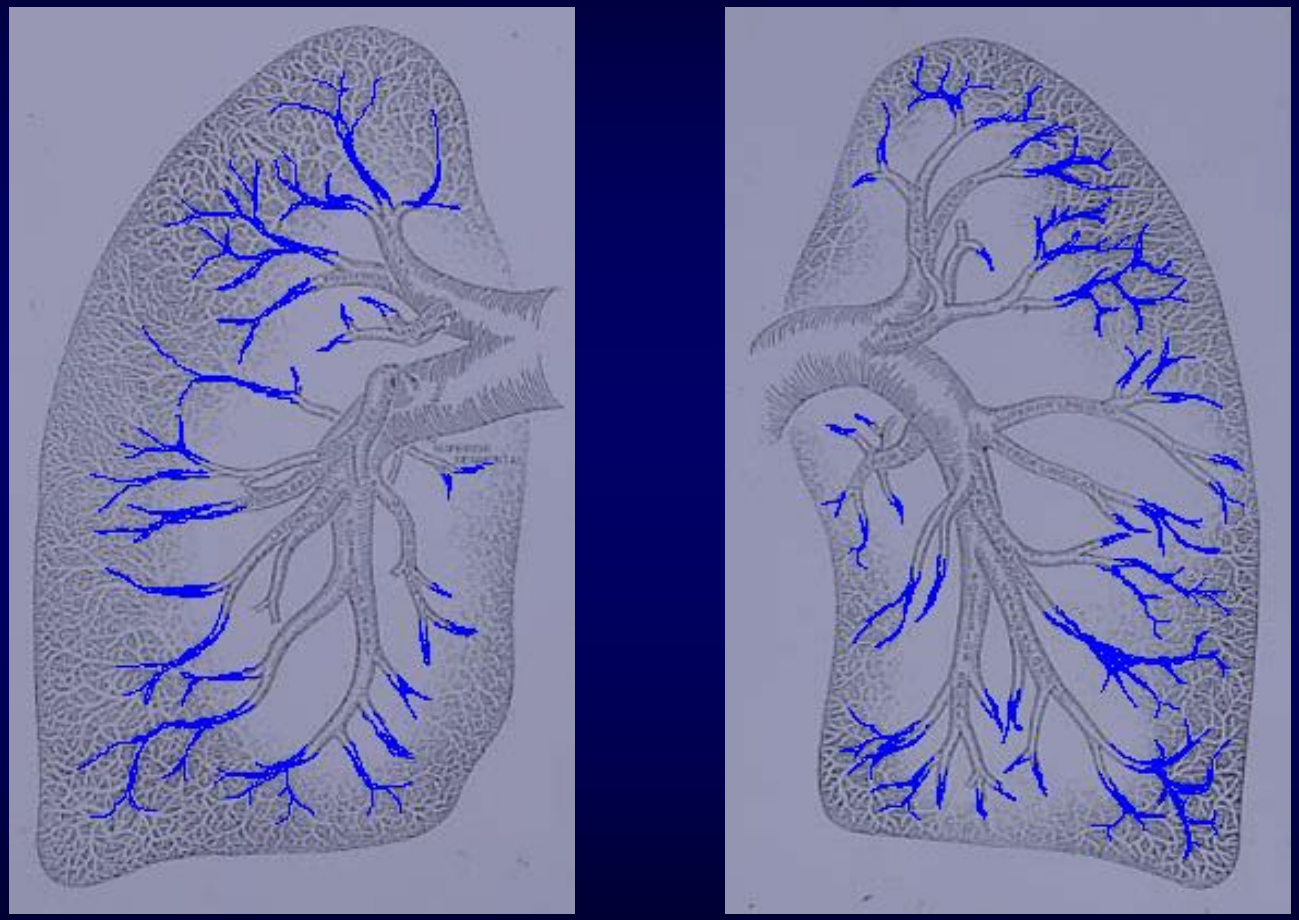
ANATOMY

PROXIMAL LESIONS





DISTAL LESIONS



EVOLVING SURGICAL TECHIQUE

Evolving surgical techniques for pulmonary endarterectomy according to the changing features of chronic thromboembolic pulmonary hypertension patients during 17-year single-center experience

Marco Morsolini, MD,[a,b] Salvatore Nicolardi, MD,[a,b] Elisa Milanesi, MD,[c] Eleonora Sarchi, MD,[d] Gabriella Mattiucci, MD,[a] Catherine Klersy, MD, MSc,[e] and Andrea Maria D'Armini, MD[a]

(J Thorac Cardiovasc Surg 2012;144:100-7)

SURGICAL TREATMENT

	Original San Diego protocol	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

LESS INVASIVE SURGERY

CEREBRAL PROTECTION STRATEGY

NIRS MONITORING

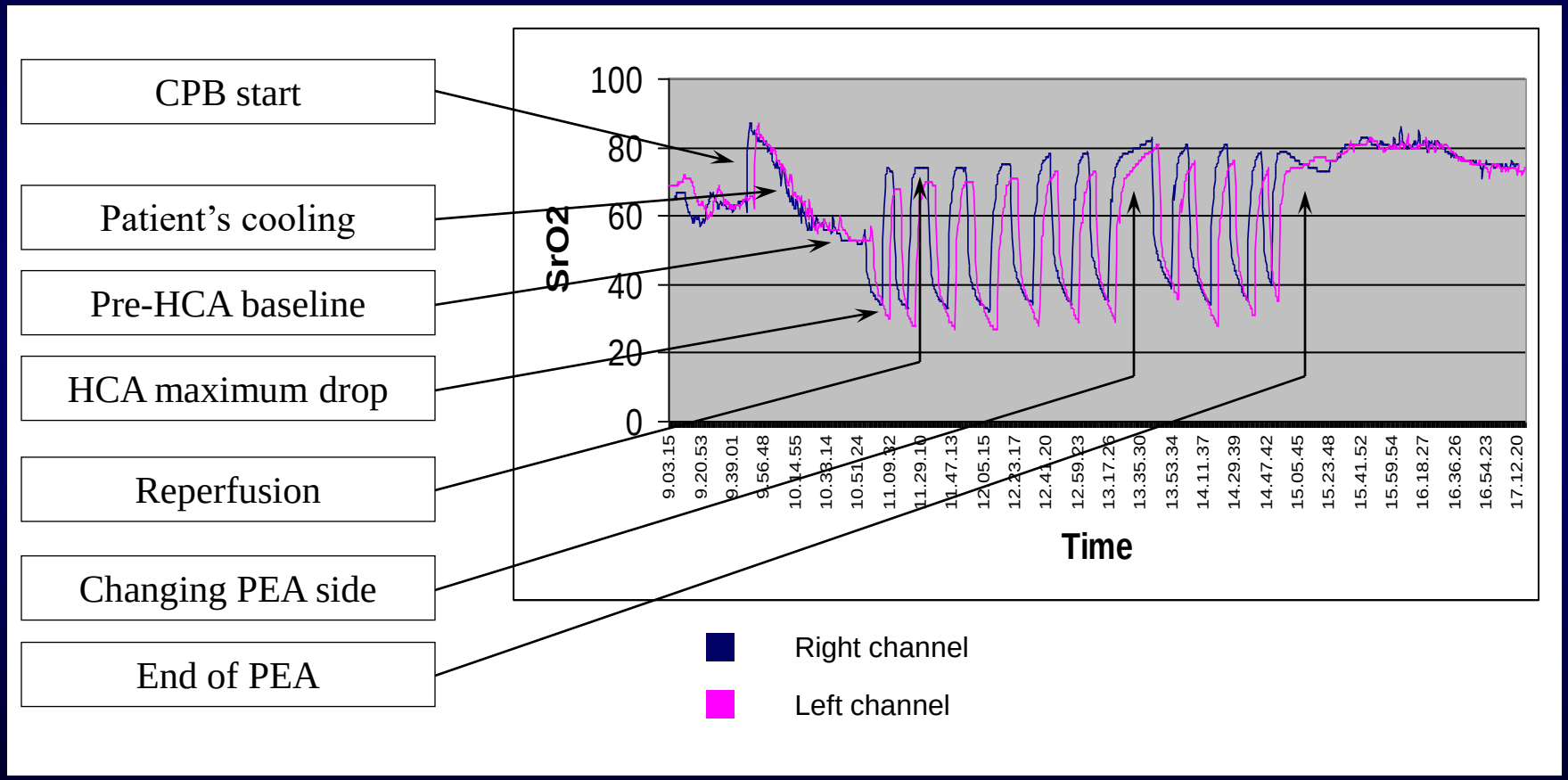
Near-InfraRed Spectroscopy

Clinical application

CEREBRAL PROTECTION STRATEGY

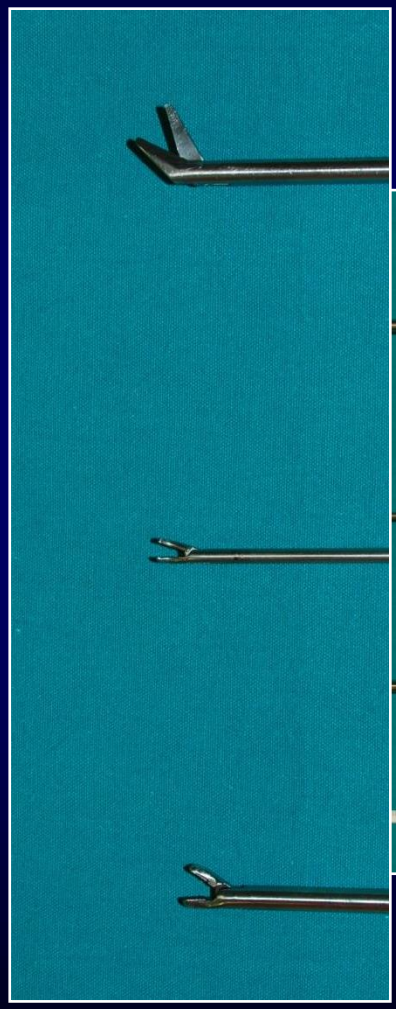
NIRS MONITORING

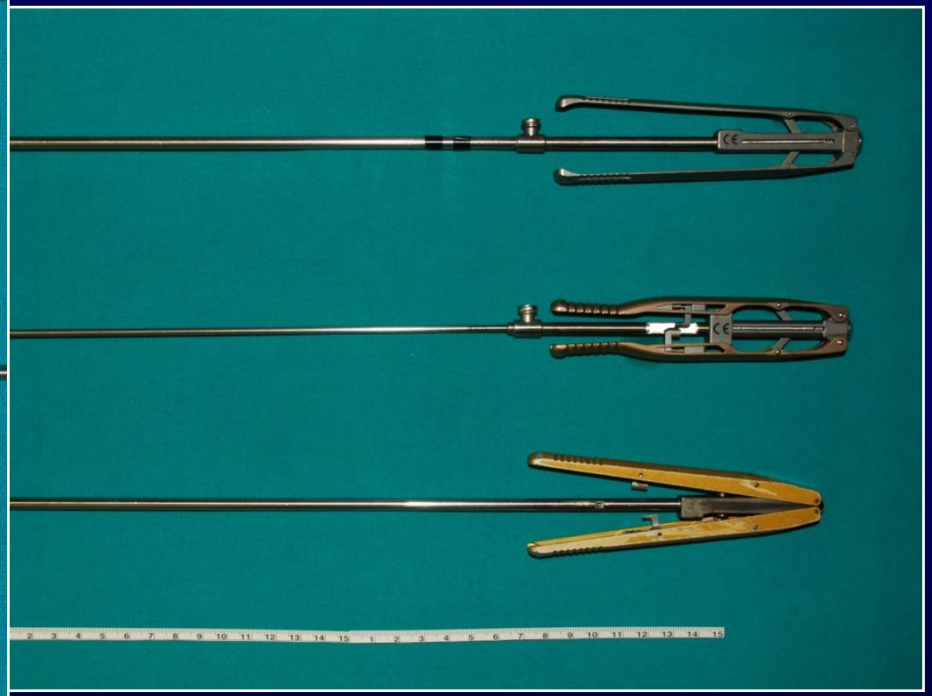
Near-InfraRed Spectroscopy



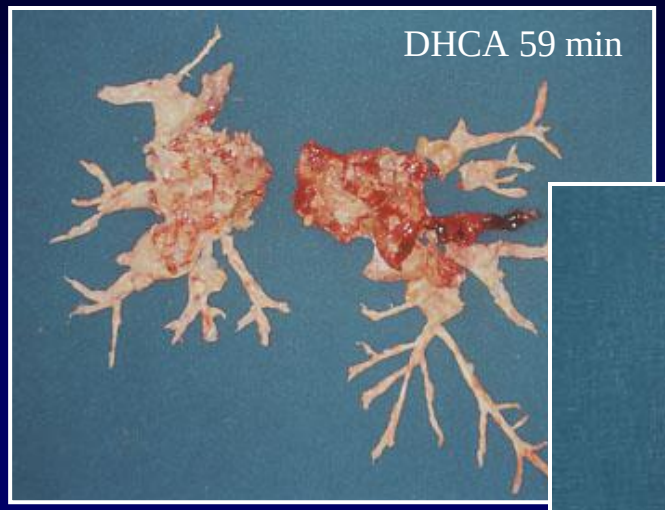
SURGICAL INSTRUMENTS

Derived from minimally-invasive cardiac surgery





JAMIESON TYPE I vs. TYPE II vs. TYPE III

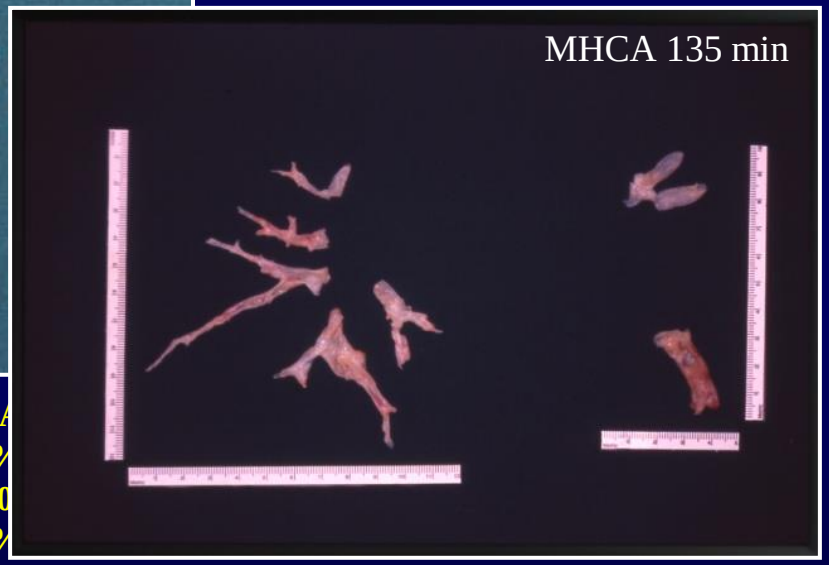


L.M.E.L. - 65 yrs M - Oct 2004 - PE

mPAP	**39**	→	**19**	**(-51%)**
CO	**4.4**	→	**5.4**	**(+23%)**
PVR	**665**	→	**222**	**(-66%)**

G.A.C. - 52 yrs F - Jul 2003 - PE

mPAP	**48**	→	**27**	**(-44%**
CO	**2.1**	→	**4.2**	**(+100**
PVR	**1638**	→	**381**	**(-77%**



B.A. - 43 yrs F - May 2009 - PEA #233

mPAP	**49**	→	**19**	**(-61%)**
CO	**3.3**	→	**5.0**	**(+52%)**
PVR	**1067**	→	**224**	**(-79%)**

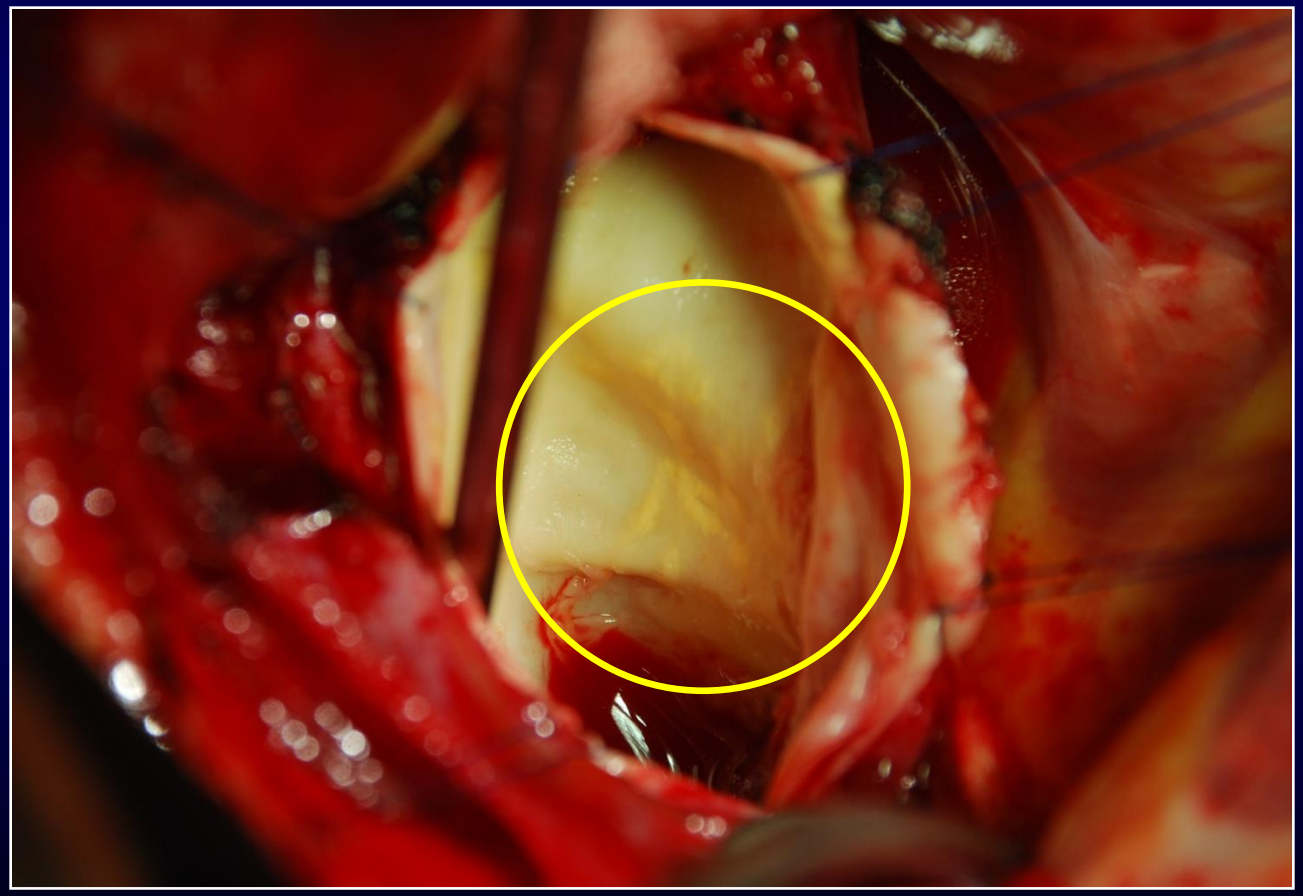
DHCA, deep hypothermic circulatory arrest;

MHCA, moderate hypothermic circulatory arrest.

TRICKS AND TIPS

The correct arterial dissection plane

Yellow-fibro-lipid plaques included into the removed cast



TRICKS AND TIPS

The correct arterial dissection plane

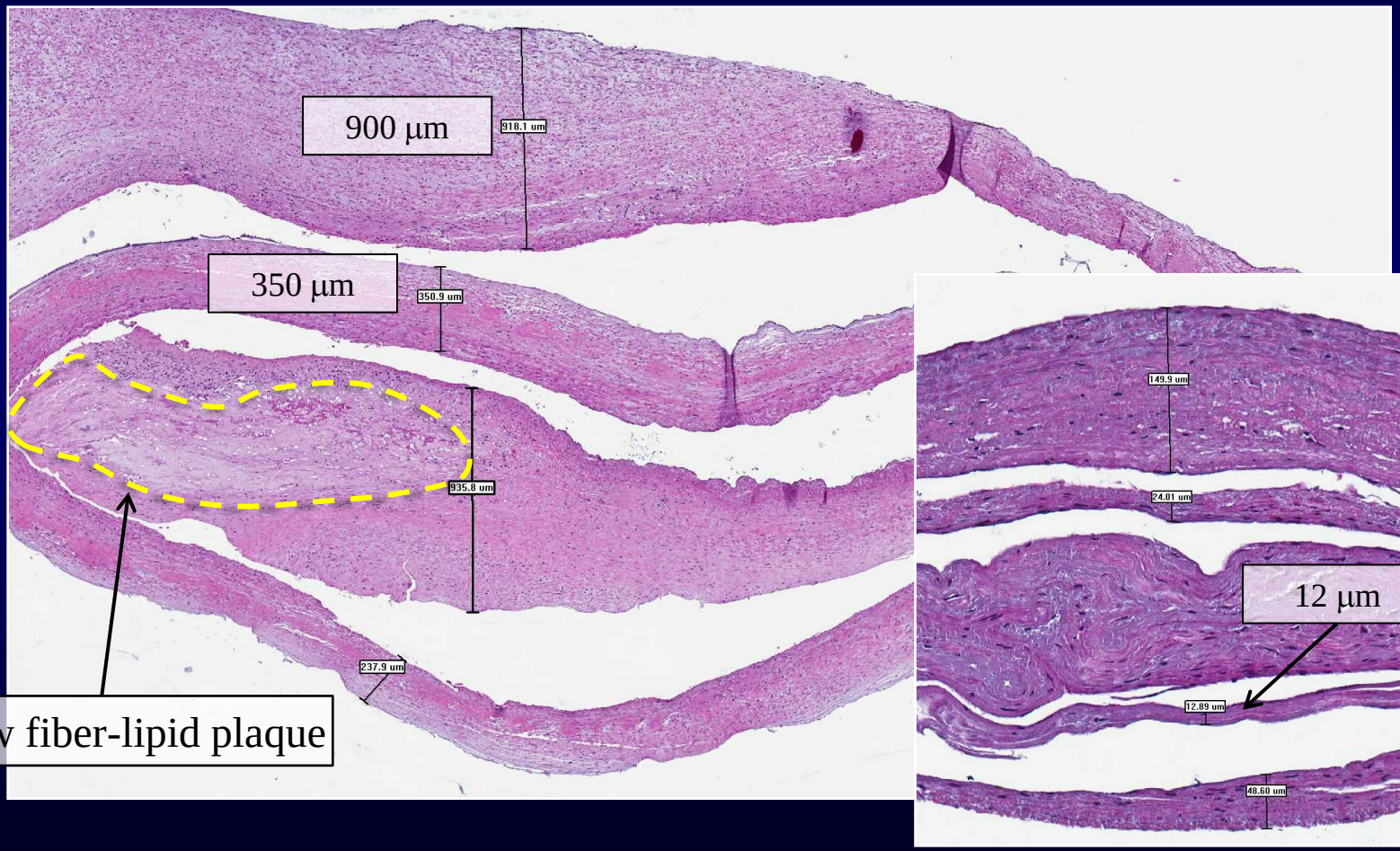
Yellow-fibro-lipid plaques included into the removed cast

TRICKS AND TIPS

Proximal dissection for the clearance of distal obstructions

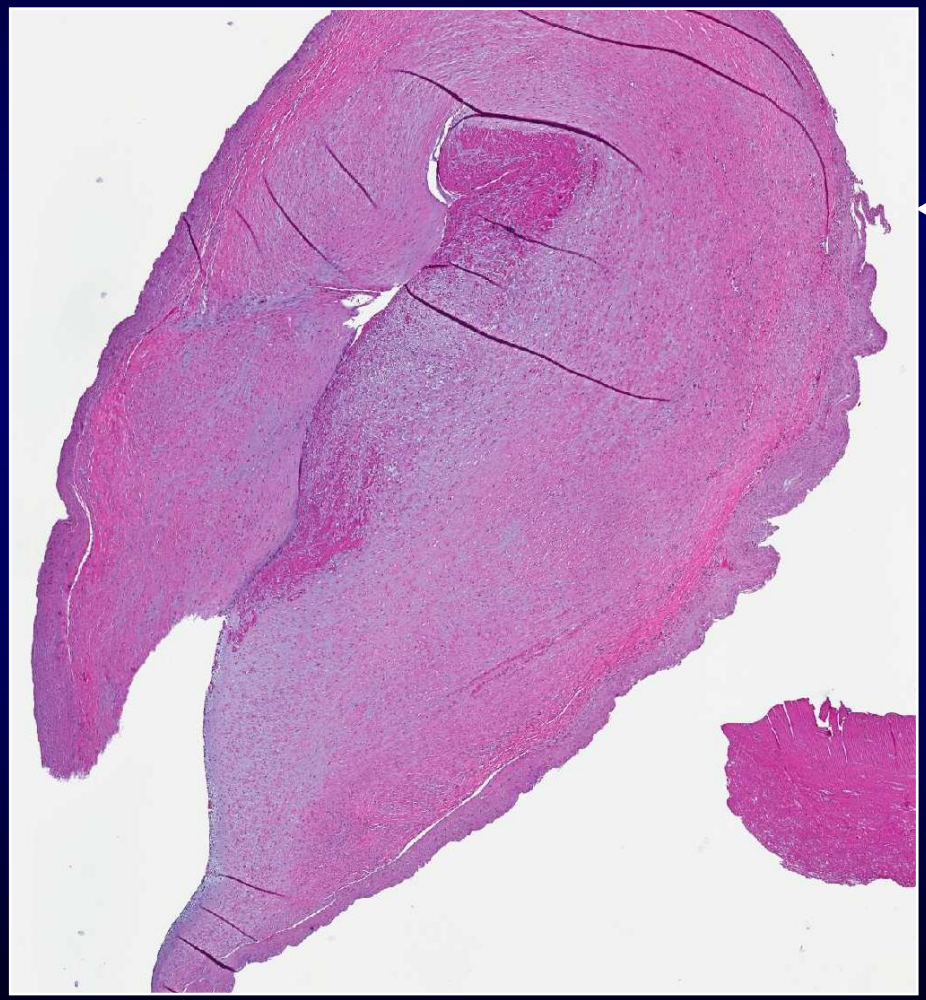
TRICKS AND TIPS

Proximal dissection for the clearance of distal obstructions

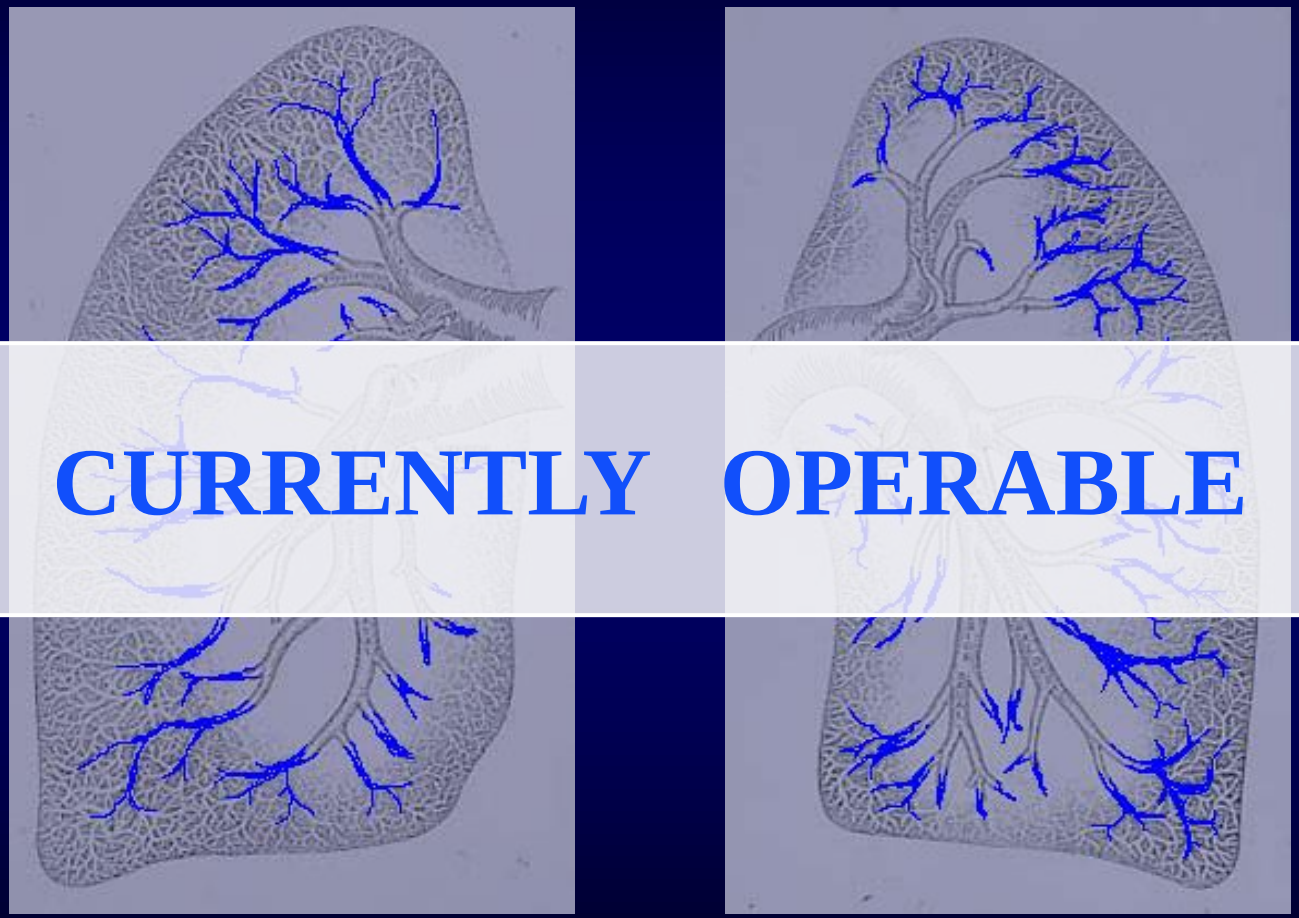


TRICKS AND TIPS

Proximal dissection for the clearance of distal obstructions

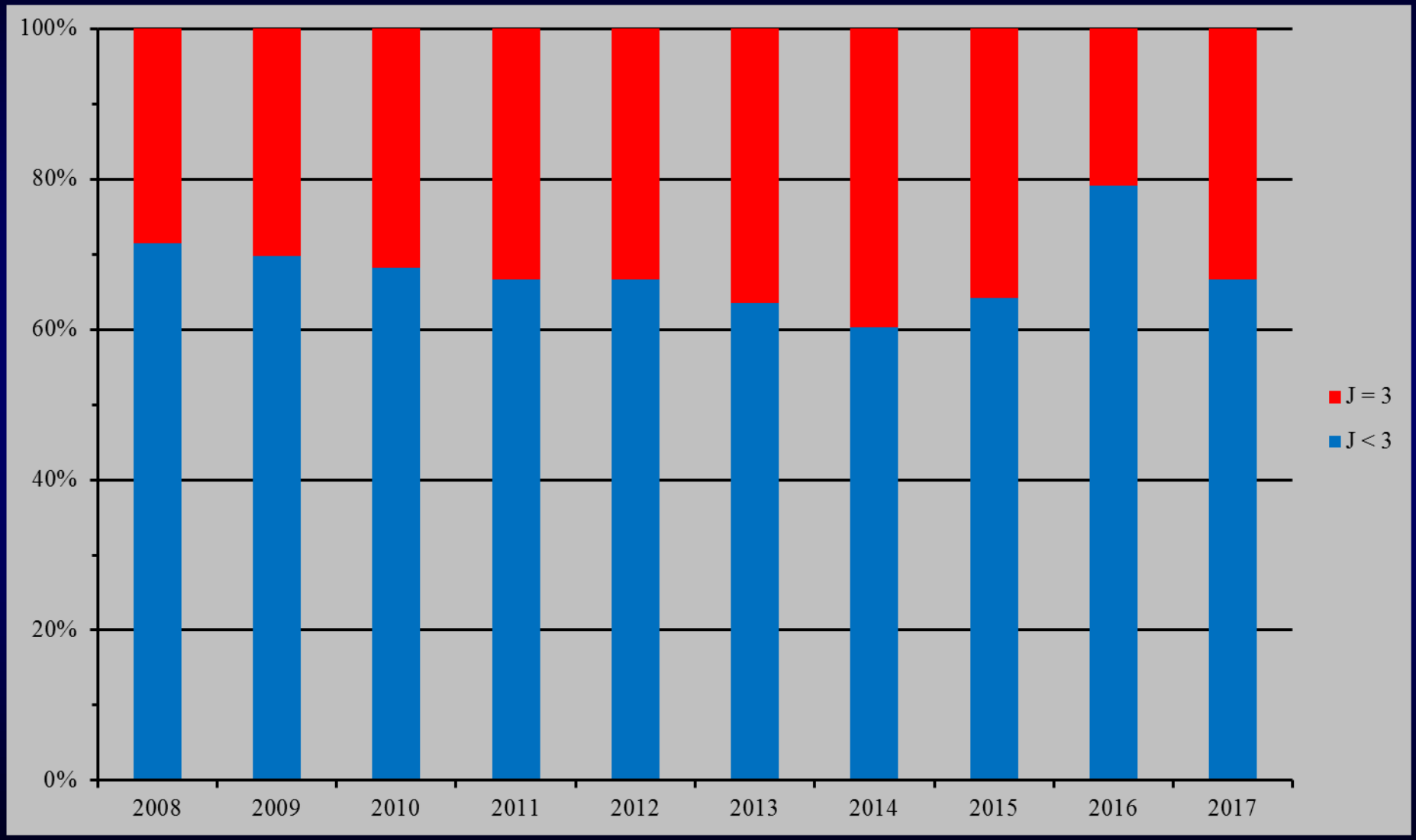


DISTAL LESIONS



CURRENTLY OPERABLE

JAMIESON TYPE III



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

TABLE 2. Intraoperative comparison and early postoperative outcome

	Proximal	Distal	*P* value
Bilateral PEA (n)	192 (86.9)	95 (86.4)	1.000
Associated procedures (n)	38 (17.2)	19 (17.3)	1.000
Total CPB time (min)	338 ± 81 (327-348)	361 ± 64 (349-373)	**.005**
Hypothermia (°C)	24.0 ± 0.9 (23.9-24.1)	23.7 ± 1.0 (23.5-23.8)	**.003**
Total HCA time (min)	84 ± 32 (80-89)	102 ± 28 (97-107)	**<.001**
Pa_{O_2}/Fi_{O_2} 6 h	284 ± 91 (271-296)	280 ± 112 (259-301)	.758
MV duration (d)	2 (1-3)	2 (1-4)	.565
ICU stay (d)	4 (3-7)	4 (3-8)	.962
Postoperative hospital stay (d)	13 (10-16)	13 (11-17)	.541

	Risk	Risk difference (95% CI)	*P* value
Univariate analysis			
Hospital mortality		1.8 (−4.2 to 7.9)	.647
Proximal	6.3%		
Distal	8.1%		
Lung reperfusion edema		−0.5 (−4.4 to 3.4)	1.000
Proximal	3.2%		
Distal	2.7%		
Tracheostomy		−1.9 (−7.8 to 3.9)	.662
Proximal	8.3%		
Distal	6.4%		
Neurologic event		−4.7 (−10.6 to 1.1)	.209
Proximal	10.2%		
transient 13/22			
permanent 9/22			
Distal	5.5%		
transient 5/6			
permanent 1/6			

Bold values indicate significance ($P < .05$). *CI*, Confidence interval; *CPB*, cardiopulmonary bypass; *HCA*, hypothermic circulatory arrest; *ICU*, intensive care unit; *MV*, mechanical ventilation; Pa_{O_2}/Fi_{O_2} *6 h*, partial pressure of oxygen in arterial blood/fraction of inspired oxygen ratio 6 hours after admission to ICU; *PEA*, pulmonary endarterectomy.

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).

DISTAL LESIONS

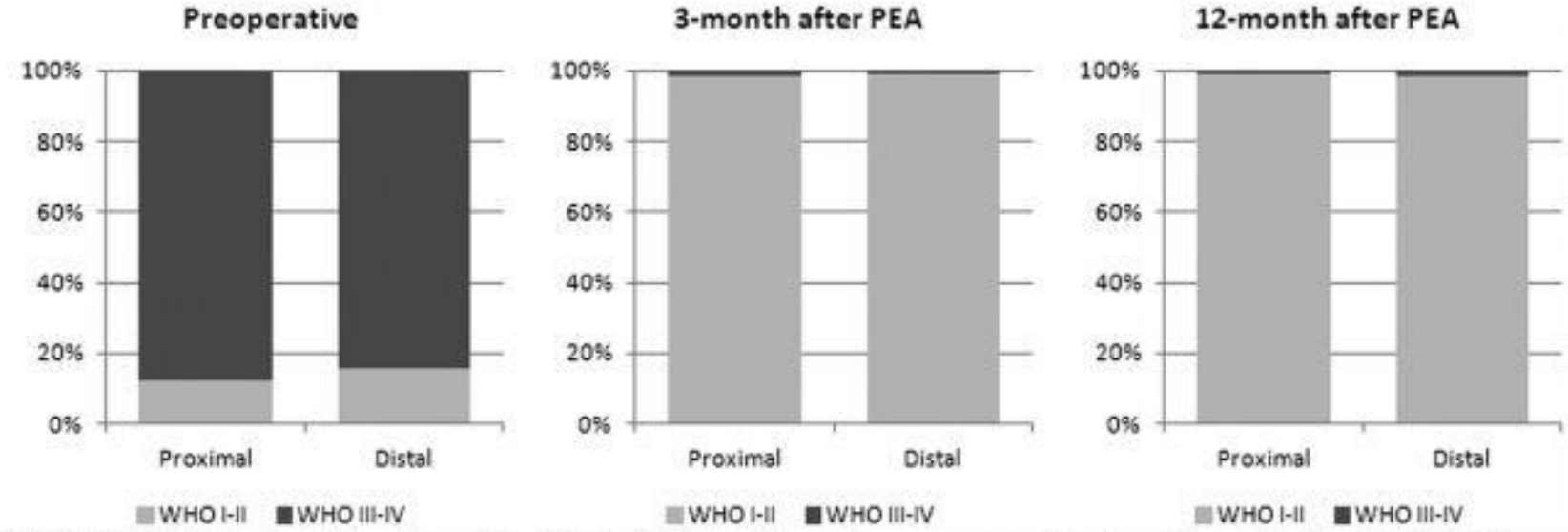


FIGURE E2. WHO functional class changes after PEA. $P < .001$ at each time point versus preoperative. Test of interaction: $P = .327$. *PEA*, Pulmonary endarterectomy; *WHO*, World Health Organization.

DISTAL LESIONS

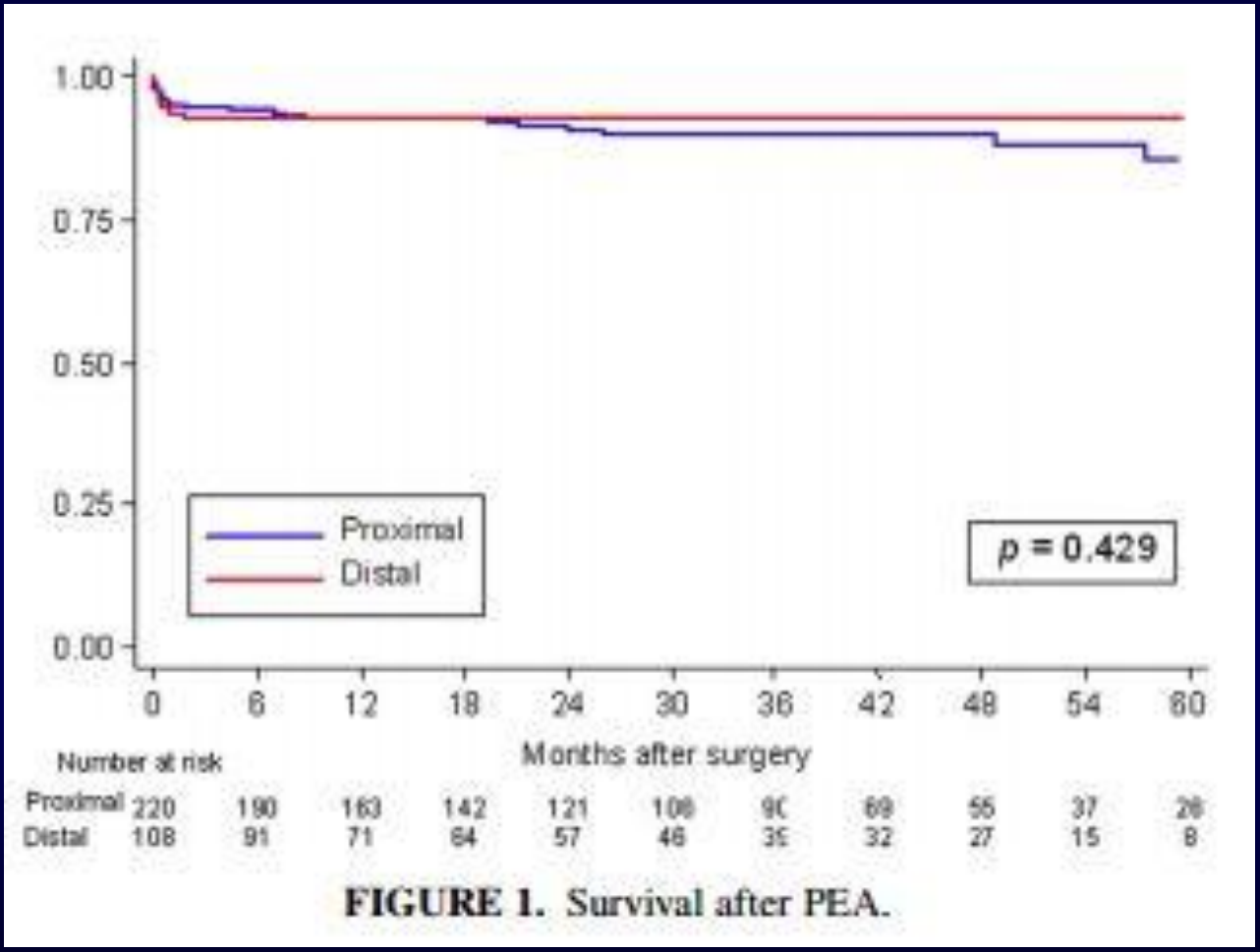
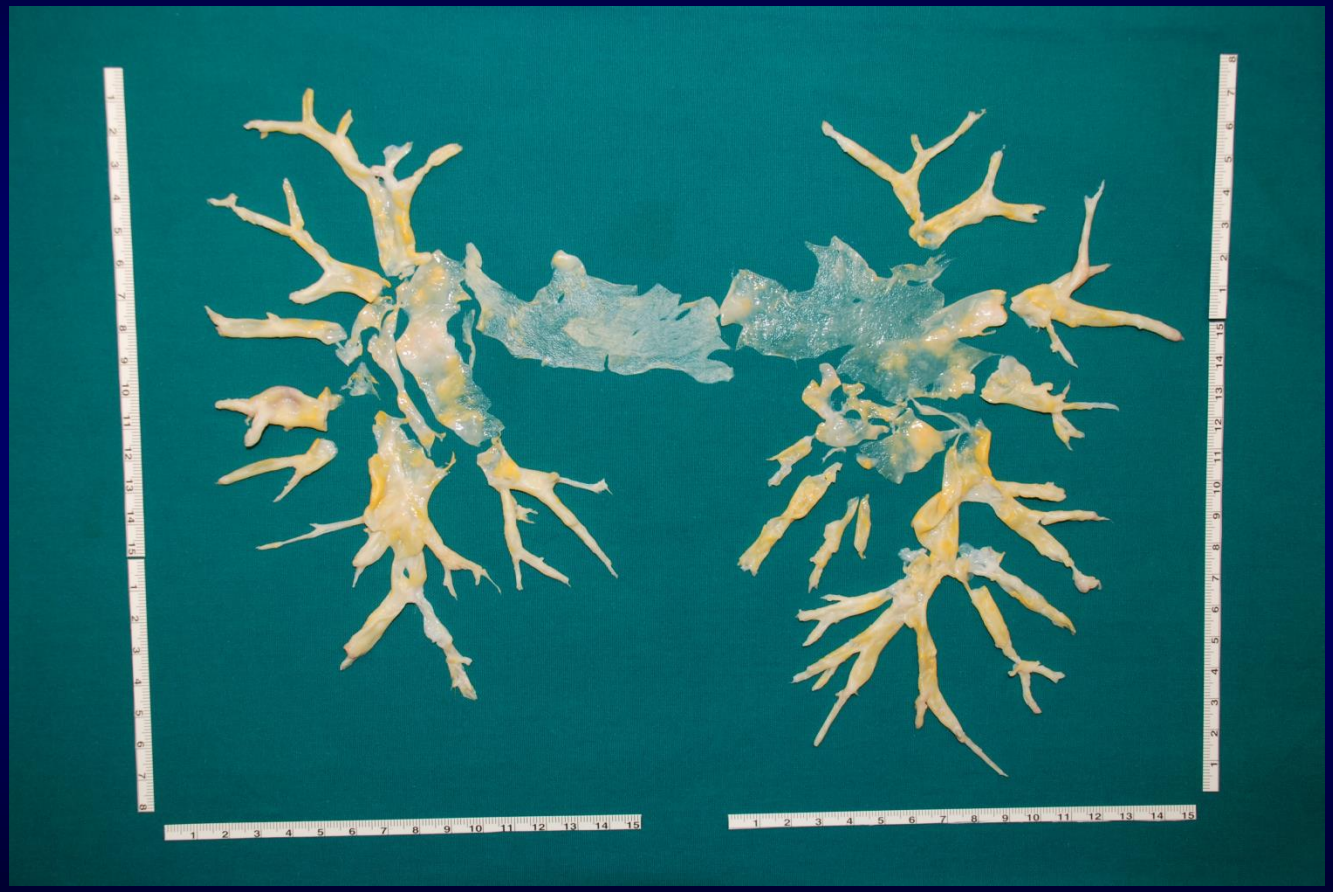


FIGURE 1. Survival after PEA.

PEA SPECIMEN



PEA EXPERT CENTER

EUROPEAN SOCIETY OF CARDIOLOGY®

European Heart Journal (2009) **30**, 2493–2537
doi:10.1093/eurheartj/ehp297

ESC/ERS GUIDELINES

Guidelines for the diagnosis and treatment of pulmonary hypertension

The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT)

A centre can be considered to have sufficient expertise in this field if it performs at least 20 PEA operations per year with a mortality rate $<10\%$.

PEA CENTER EXPERT

BETTER DEFINITION

Surgical management and outcome of patients with chronic thromboembolic pulmonary hypertension: Results from an international prospective registry

Eckhard Mayer, MD,[a] David Jenkins, FRCS,[b] Jaroslav Lindner, MD,[c] Andrea D'Armini, MD,[d] Jaap Kloek, MD,[e] Bart Meyns, MD,[f] Lars Bo Ilkjaer, MD,[g] Walter Klepetko, MD,[h] Marion Delcroix, MD,[f] Irene Lang, MD,[h] Joanna Pepke-Zaba, MD,[b] Gerald Simonneau, MD,[i] and Philippe Dartevelle, MD[j]

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

TABLE 4. Center expertise and surgical management

	Average no. of PEAs per year			
	1–10	11–50	>50	Combined
No. of centers performing PEA, n (%)	6 (35.3%)	8 (47.1%)	3 (17.6%)	17*
No. of patients, n (%)	54 (14.0%)	191 (49.5%)	141 (36.5%)	386*
Change in PVR from diagnosis to end of intensive care, dyn.s.cm^{-5}, median (range), n	–476 NS (–1760 to 80) n = 43	–476† (–2256 to 22) n = 144	–400‡ (–2261 to 554) n = 65	–457 (–2261 to 554) n = 252
Death, n (%)§				
In-hospital	4 (7.4%) NS	9 (4.7%) NS	5 (3.5%)§	18 (4.7%)
1 y after PEA	6 (11.1%) NS	14 (7.3%) NS	7 (5.0%)§	27 (7.0%)

NS, Not significant; *PEA,* pulmonary endarterectomy; *PVR,* pulmonary vascular resistance. *Two patients underwent operations in 2 nonparticipating centers performing > 50 PEAs per year and < 10 PEAs per year, respectively. NS compared with ‡ (Wilcoxon 2-sample test) or § (Fisher's exact test). †$P < .05$ compared with ‡ (Wilcoxon 2-sample test).

Chronic Thromboembolic Pulmonary Hypertension

Nick H. Kim, MD,* Marion Delcroix, MD,† David P. Jenkins, MB BS,‡ Richard Channick, MD,§ Philippe Dartevelle, MD,‖ Pavel Jansa, MD,¶ Irene Lang, MD,# Michael M. Madani, MD,* Hitoshi Ogino, MD, PHD,** Vittorio Pengo, MD,†† Eckhard Mayer, MD‡‡

(J Am Coll Cardiol 2013;62:D92–9) © 2013

bility assessment by a CTEPH team (Fig. 2). Because the operability assessment remains complex, we recommend that only an experienced CTEPH team should determine that a case of CTEPH is inoperable. Furthermore, recognizing the subjective nature of the operability assessment process, we encourage a re-evaluation of operability by a second experienced CTEPH center, whenever feasible, in cases initially deemed inoperable. In cases of operable CTEPH, medical

PEA EXPERT CENTER

PROPOSED DEFINITION

- In most recent guidelines

  no definition of PEA expert center

  no suggestion of second opinion

REFERENCE

Pulmonary endarterectomy in the management of chronic thromboembolic pulmonary hypertension

David Jenkins[1], Michael Madani[2], Elie Fadel[3], Andrea Maria D'Armini[4] and Eckhard Mayer[5]

Eur Respir Rev 2017; 26: 160111

REFERENCE

TABLE 2 Characteristics of an expert centre

Extensive experience with cardiothoracic surgery, including procedures requiring DHCA
Excellent pulmonary and cardiac services
Emphasis on pulmonary hypertension
Expert diagnostic imaging
Experienced multidisciplinary team comprising surgeons, radiologists, anaesthetists, intensivists, nurses, perfusionists, respiratory therapists and interventionalists, including specialists experienced in BPA

DHCA: deep hypothermic circulatory arrest; BPA: balloon pulmonary angioplasty. Data from [12].

Eur Respir Rev 2017; 26: 160111

REFERENCE

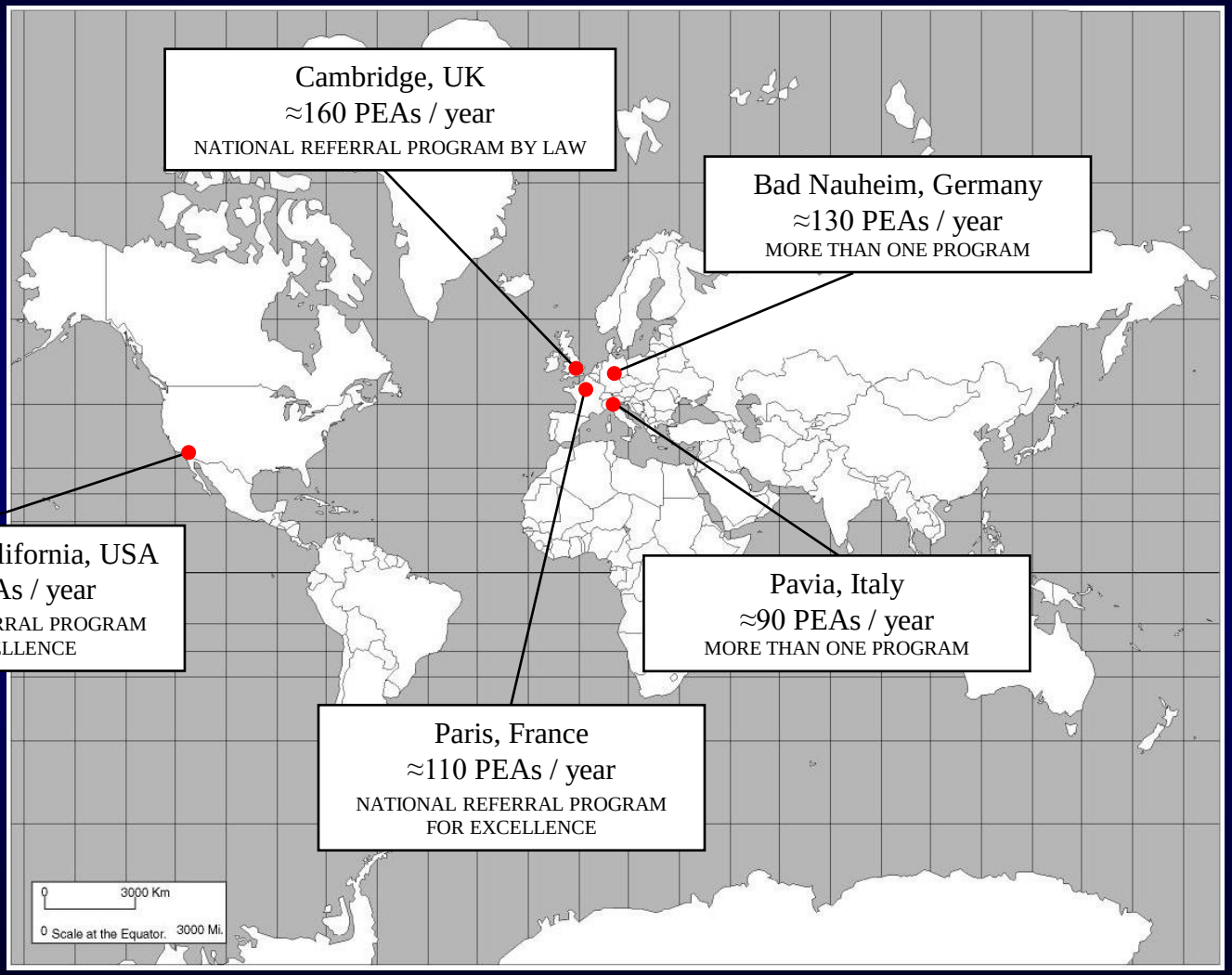
TABLE 3 Proposed identification criteria of expert or high-quality centres

Level of expertise	Criteria		
I	30-day or in-hospital mortality <5%		
II	30-day or in-hospital mortality <5%	**plus** ⩾50 procedures·year^{-1}	
III	30-day or in-hospital mortality <5%	**plus** ⩾50 procedures·year^{-1}	**plus** ability to perform segmental endarterectomy/operate on distal disease **plus** ability to provide PEA, BPA and medical therapy

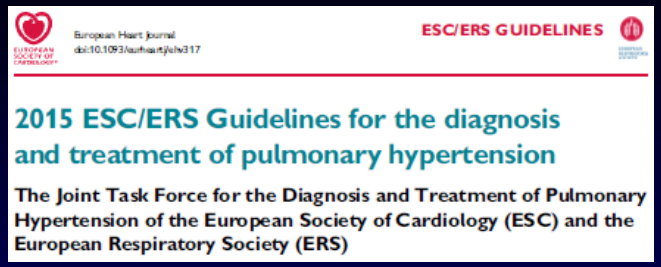
PEA: pulmonary endarterectomy; BPA: balloon pulmonary angioplasty.

Eur Respir Rev 2017; 26: 160111

MAIN WORLD PEA CENTERS



PULMONARY ENDARTERECTOMY: THE PAVIA EXPERIENCE


European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)

1. Pulmonary arterial hypertension

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2 mutation
  - 1.2.2 Other mutations
- 1.3 Drugs and toxins induced
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 Human immunodeficiency virus (HIV) infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease (Table 6)
  - 1.4.5 Schistosomiasis

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

- 1'.1 Idiopathic
- 1'.2 Heritable
  - 1'.2.1 EIF2AK4 mutation
  - 1'.2.2 Other mutations
- 1'.3 Drugs, toxins and radiation induced
- 1'.4 Associated with:
  - 1'.4.1 Connective tissue disease
  - 1'.4.2 HIV infection

1''. Persistent pulmonary hypertension of the newborn

2. Pulmonary hypertension due to left heart disease

- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital / acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
- 2.5 Congenital /acquired pulmonary veins stenosis

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 lung diseases (Web Table 11)

4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions

- 4.1 Chronic thromboembolic pulmonary hypertension
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Angiosarcoma
  - 4.2.2 Other intravascular tumors
  - 4.2.3 Arteritis
  - 4.2.4 Congenital pulmonary arteries stenoses
  - 4.2.5 Parasites (hydatidosis)

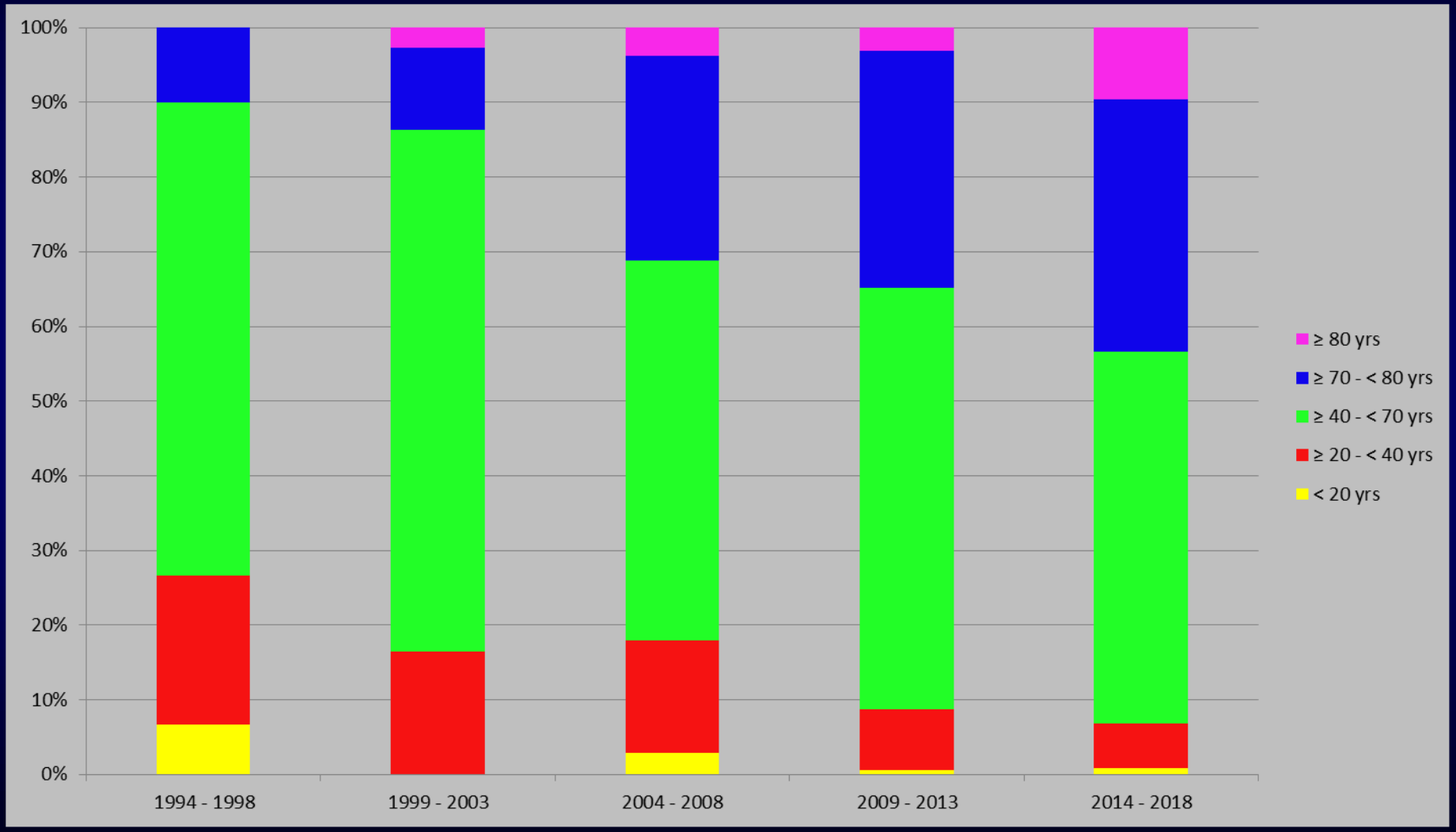
5. Pulmonary hypertension with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders, sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: pulmonary tumoral thrombotic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension

4%

AGE

OF 780 PEAs



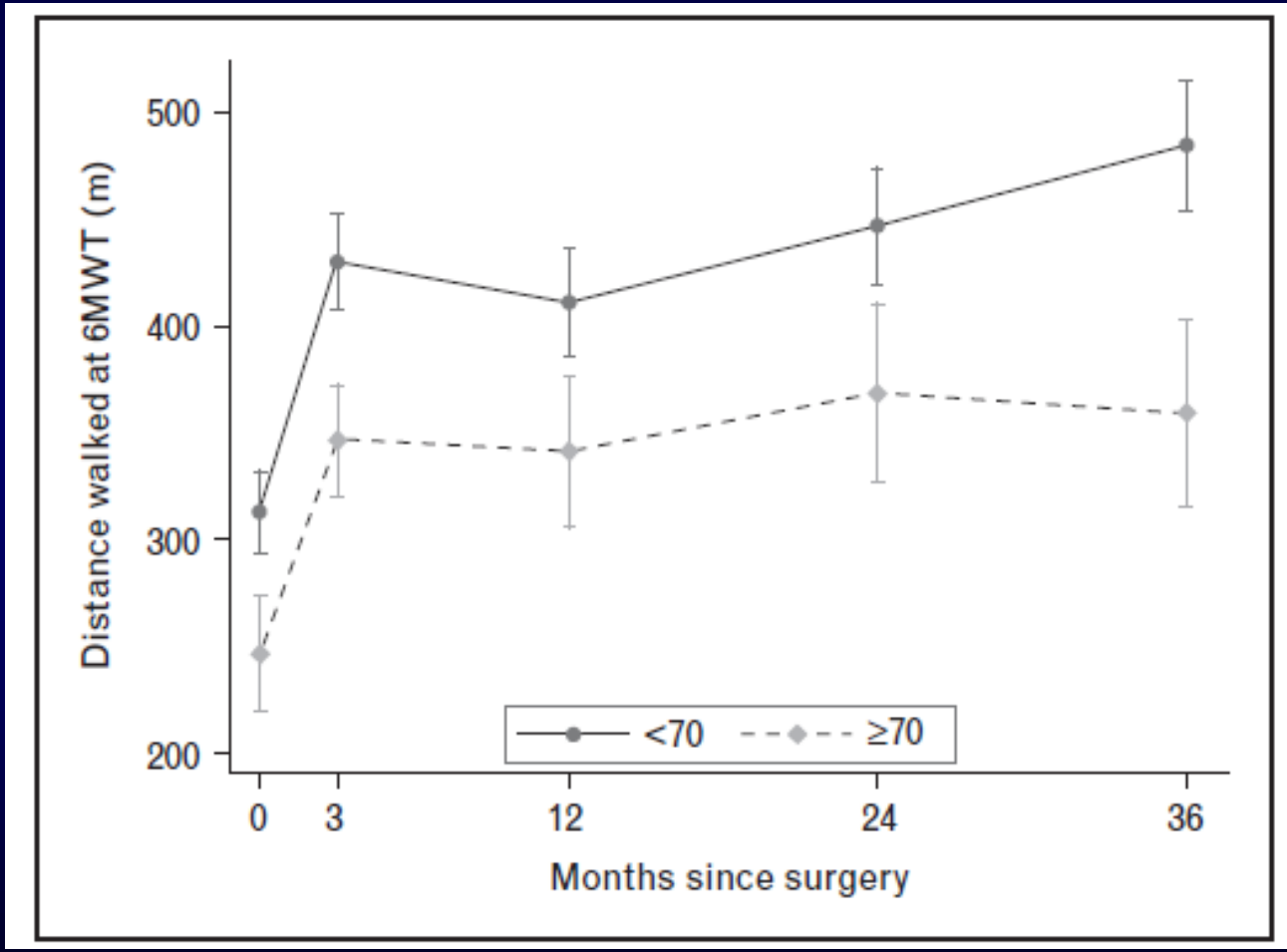
ELDERLY PATIENTS

Pulmonary endarterectomy in the elderly: safety, efficacy and risk factors

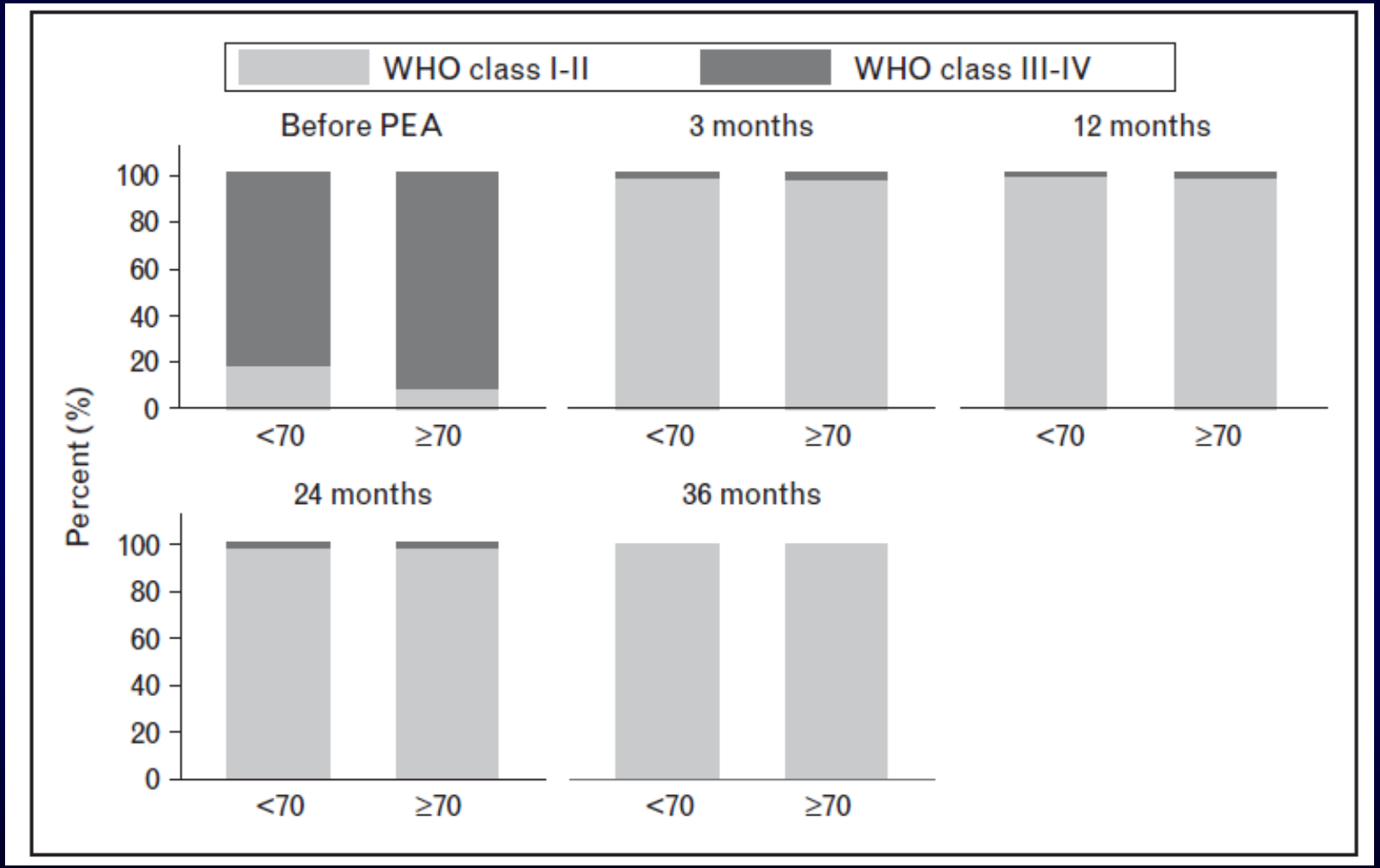
Nicola Vistarini[a], Marco Morsolini[a], Catherine Klersy[b], Gabriella Mattiucci[a], Valentina Grazioli[a], Maurizio Pin[a], Stefano Ghio[c] and Andrea Maria D'Armini[a]

J Cardiovasc Med 2016, 17:144–151

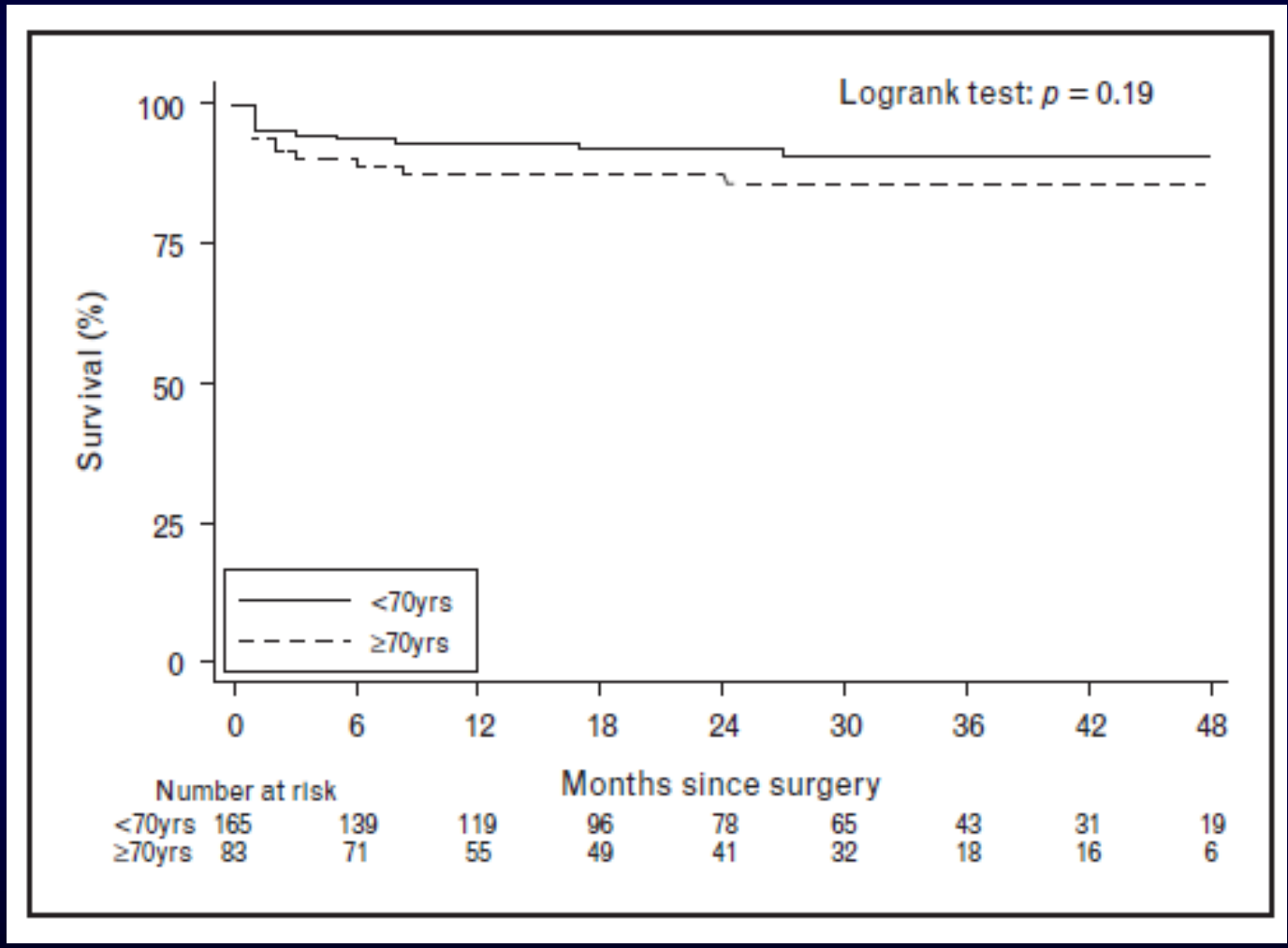
ELDERLY PATIENTS



ELDERLY PATIENTS



ELDERLY PATIENTS



PAVIA CTEPH PROGRAM

January, 1st – December, 31st 2016 → 436

New Evaluations (204 pts) CTEPH-PEA FUP (226 pts) PAS-PEA FUP (6 pts)

CONFIRMED (100 pts)

- PROXIMAL LESIONS (92 pts)
  - 72 PEAs (8 pts evaluated in 2015)
  - 3 waiting for PEA
  - 5 refused evaluation for PEA
  - 2 refused PEA
  - 13 with severe co-morbidities
  - 3 died before evaluation or before PEA
  - 1 association with severe emphysema
  - 1 complete occlusion with vessel retraction

OPERABILITY RATE *thecnically* ***92%***

+ refused ***85%***

+ comorbility ***70%***

- PERSISTENT / RECURRENT PH (1 pt)
  - 1 medical therapy
- DISTAL LESIONS (7 pts)
  - 6 controindication for DLTx
  - 1 too early for DLTx

OTHER DIAGNOSIS (104 pts)

- BPA evaluation (4 pts) – 4 waiting
- RECENT APE (30 pts)
  - 29 mt - 1 mt+ECMO
- PREVIOUS APE WITHOUT SIGNS (12 pts)
  - 12 medcal therapy
- APE OVER CTE LESIONS (3 pts)
  - 3 3-month m.t.→new evaluation
- MINIMAL CTE LESIONS WITHOUT/LOW PH (12 pts)
  - 12 medical therapy
- PULMONARY ARTERY SARCOMA (10 pts)
  - 8 PEAs - 1 inoperable - 1 refused PEA
- PULMONARY ARTERY ANEURYSM (2 pts)
  - 2 medical therapy
- MISCELLANEOUS (30 pts)
  - 10 Eisenmenger
  - 3 Talassemia&splenectomy
  - 1 PH by hydatid cystic embolism
  - 1 Mediastinal fibrosis
  - 1 Systemic sclerosis
  - 9 PAH
  - 2 PVOD
  - 1 HIV
  - 1 1&4
  - 1 1&5

BENEFiT STUDY

CLINICAL RESEARCH Clinical Trials

Bosentan for Treatment of Inoperable Chronic Thromboembolic Pulmonary Hypertension

BENEFiT (Bosentan Effects in iNopErable Forms of chronIc Thromboembolic pulmonary hypertension), a Randomized, Placebo-Controlled Trial

Xavier Jaïs, MD,* Andrea M. D'Armini, MD,† Pavel Jansa, MD,‡ Adam Torbicki, MD,§ Marion Delcroix, MD,‖ Hossein A. Ghofrani, MD,¶ Marius M. Hoeper, MD,# Irene M. Lang, MD,** Eckhard Mayer, MD,†† Joanna Pepke-Zaba, MD,‡‡ Loïc Perchenet, PHD,§§ Adele Morganti, MSC,§§ Gérald Simonneau, MD,* Lewis J. Rubin, MD,‖‖ for the BENEFiT Study Group

Clamart, France; Pavia, Italy; Prague, Czech Republic; Warsaw, Poland; Leuven, Belgium; Giessen, Hannover, and Mainz, Germany; Vienna, Austria; Cambridge, United Kingdom; Allschwil, Switzerland; and La Jolla, California

(J Am Coll Cardiol 2008;52:2127–34)

CHEST STUDY

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Riociguat for the Treatment of Chronic Thromboembolic Pulmonary Hypertension

Hossein-Ardeschir Ghofrani, M.D., Andrea M. D'Armini, M.D., Friedrich Grimminger, M.D., Marius M. Hoeper, M.D., Pavel Jansa, M.D., Nick H. Kim, M.D., Eckhard Mayer, M.D., Gerald Simonneau, M.D., Martin R. Wilkins, M.D., Arno Fritsch, Ph.D., Dieter Neuser, M.D., Gerrit Weimann, M.D., and Chen Wang, M.D., for the CHEST-1 Study Group*

BALLON PULMONARY ANGIOPLASTY

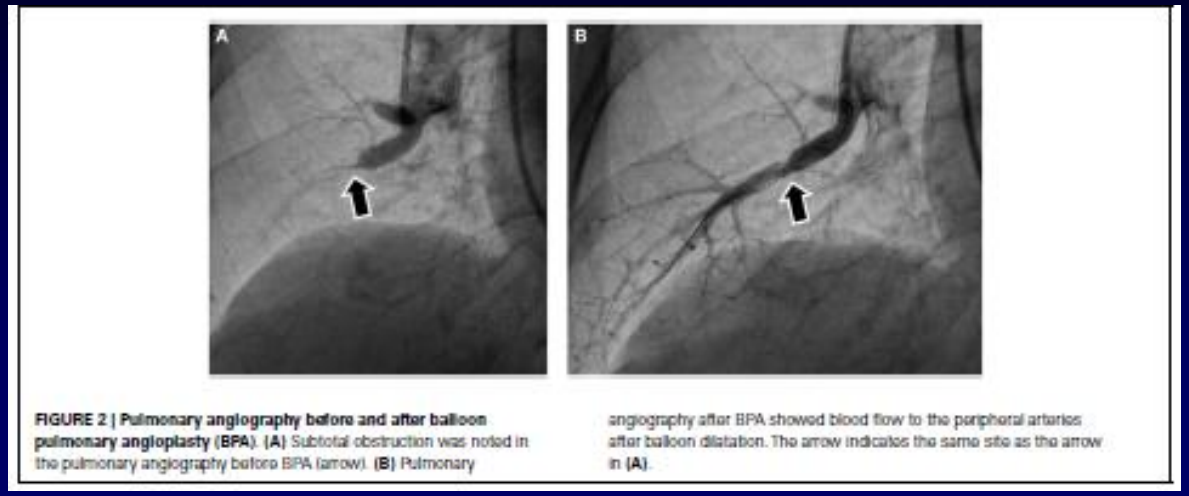
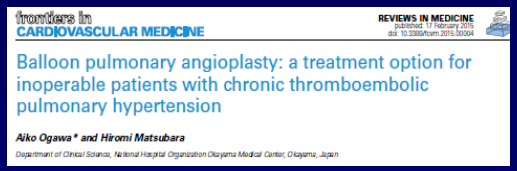


FIGURE 2 | Pulmonary angiography before and after balloon pulmonary angioplasty (BPA). (A) Subtotal obstruction was noted in the pulmonary angiography before BPA (arrow). **(B)** Pulmonary angiography after BPA showed blood flow to the peripheral arteries after balloon dilatation. The arrow indicates the same site as the arrow in **(A)**.


frontiers in CARDIOVASCULAR MEDICINE

REVIEWS IN MEDICINE

published: 17 February 2015
doi: 10.3389/fcvm.2015.00004

Balloon pulmonary angioplasty: a treatment option for inoperable patients with chronic thromboembolic pulmonary hypertension

Aiko Ogawa and Hiromi Matsubara*

Department of Clinical Science, National Hospital Organization Okayama Medical Center, Okayama, Japan

- An option for *INOPERABLE* pts, *HIGH SURGICAL RISK* pts, *REFUSED PEA* pts, *POST-PEA* pts
- Important to performed BPA in expert Centre able to offer all options to the pts and with a high volume pts
- *BPA program* started in *Pavia* on *February 2017*: to date *3 pts treated* out of a total of *8 pts evaluated and ready* for BPA

PRIMARY PULMONARY ARTERY SARCOMA

Surgical treatment of primary pulmonary artery sarcoma

Valentina Grazioli, MD,[a] Nicola Vistarini, MD,[a] Marco Morsolini, MD, PhD,[a] Catherine Klersy, MD, MSc,[b] Giulio Orlandoni, MD,[a] Roberto Dore, MD,[c] and Andrea Maria D'Armini, MD[a]

Cardiovasc Surg 2014;148:113-8)

OTHER TYPE OF OBSTRUCTIONS: primary pulmonary artery sarcoma

Bilateral PEA + pulmonary valve replacement
(Carpentier Edwards 21)

CTEPH & PAS PATIENTS

780 & 28 → 808 PEAs