Surgical management of CTEPH

Aleem Siddique
Division of Cardiothoracic Surgery

University of Nebraska Medical Center
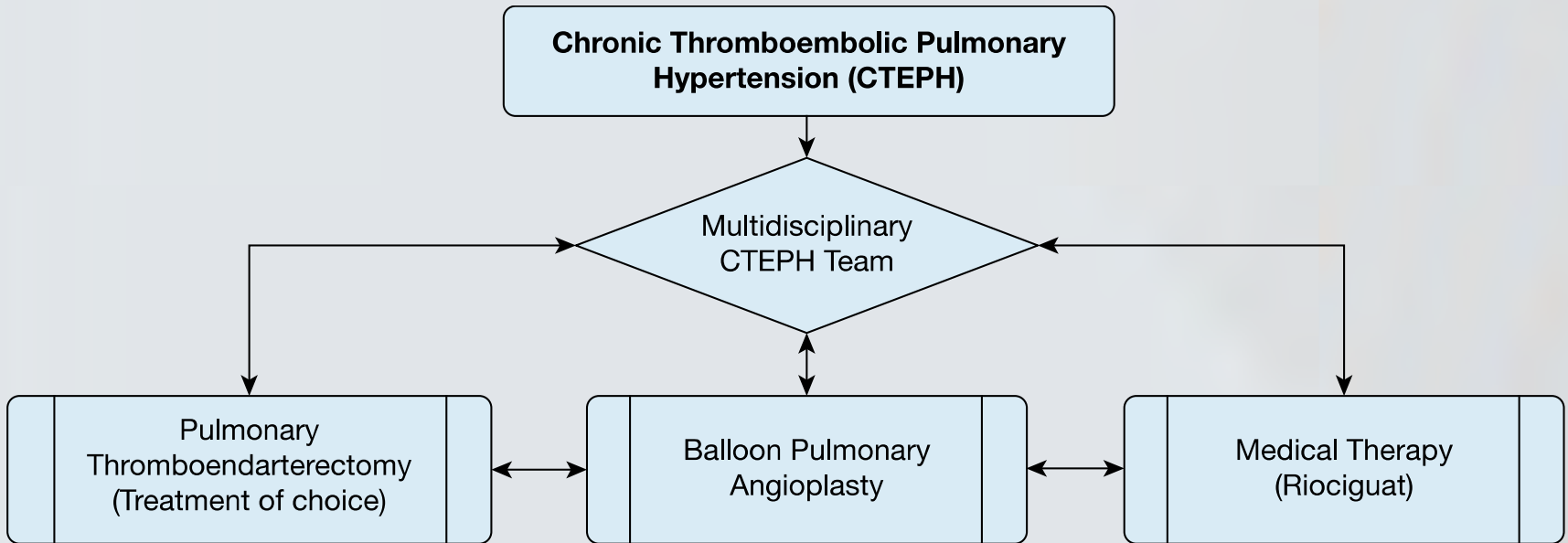
No relevant disclosures

Objectives

Recognize appropriate patients for surgical management

Understand pre-operative optimization

Appreciate key steps in the surgery

Describe surgical outcomes

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│  Chronic Thromboembolic Pulmonary    │
│       Hypertension (CTEPH)           │
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                  │
                  ▼
            Multidisciplinary
              CTEPH Team

┌──────────────────┐   ┌──────────────────┐   ┌──────────────────┐
│    Pulmonary     │   │ Balloon Pulmonary│   │  Medical Therapy │
│Thromboendarterectomy│ │   Angioplasty    │   │   (Riociguat)    │
│(Treatment of choice)│ │                  │   │                  │
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Studies for surgical assessment

V/Q scan

TTE

Pulmonary angiogram

Right heart catheterization
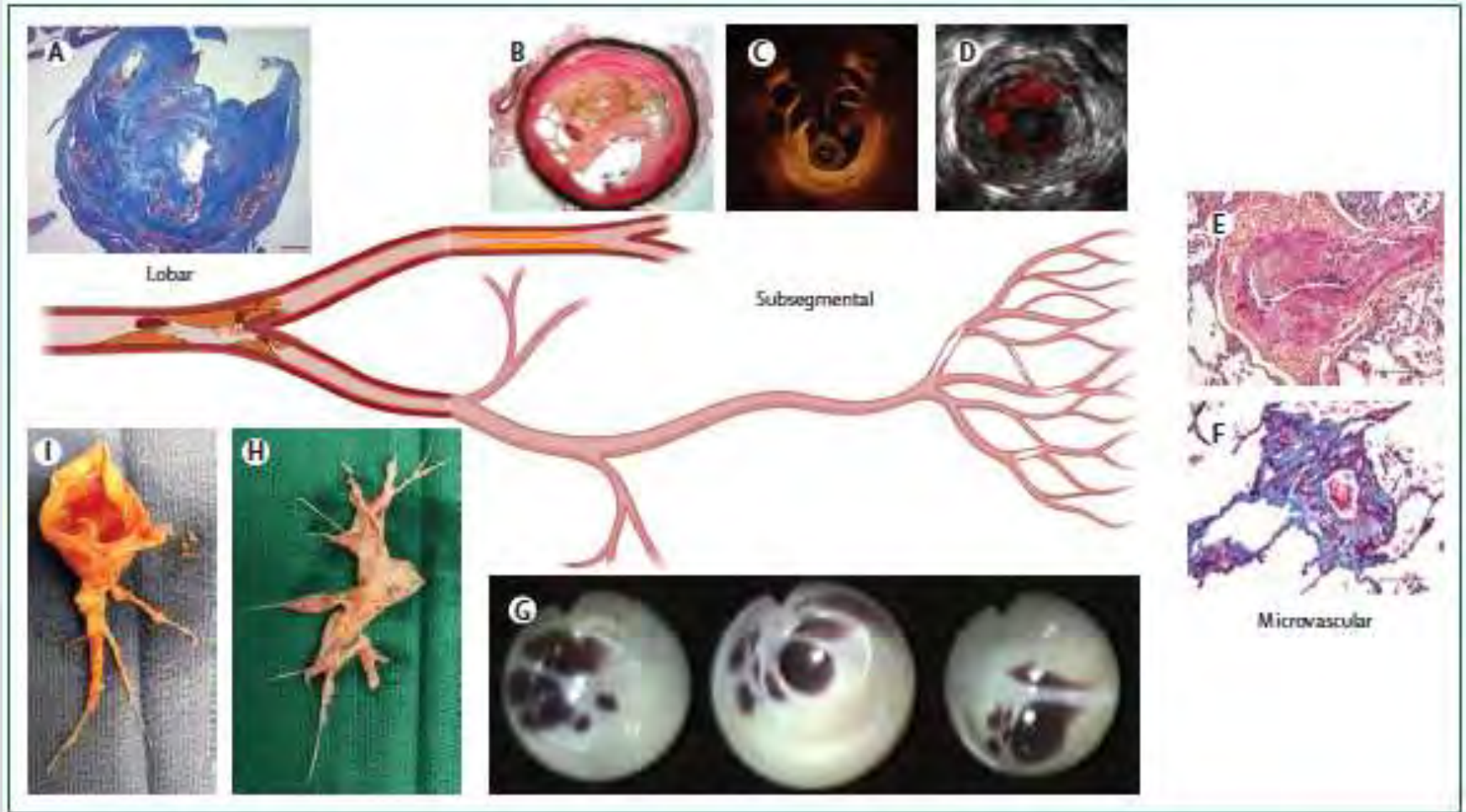
CT- PA protocol

Assessment of operability

Is the disease technically operable?

Location of disease (Level 1 and 2 best)

Severity of vascular resistance (PVR) correlates with surgically accessible obstructive disease

Surgeon/center expertise

Disease Classification by Location

Disease Classification by Location

Level 1- involves main PAs

Level 2- involves lobar branches

Level 3- involves segmental branches

Level 4- subsegmental disease

Assessment of operability

Is the disease technically operable?

Location of disease (Level 1 and 2 best)

Severity of vascular resistance (PVR) correlates with surgically accessible obstructive disease

Surgeon/center expertise

Assessment of operability

Is the patient reasonable for surgery?

Physiologic impact of the disease- RV failure

Co-morbidities

Debility, malnutrition

High risk features

Advanced symptoms (NYHA 4, severely impaired walk distance)

Severely elevated vascular resistance (PVR)

RV failure

COPD

CAD

LV dysfunction

Severely elevated Pulmonary Vascular Resistance (PVR)

Table 1 Patient characteristics

Characteristics	PVR >1,000 dynes.s.cm^{-5} (n=100)	PVR <1,000 dynes.s.cm^{-5} (n=301)	P value
Age (years)	58±15	58±15	0.99
Sex			0.21
Male	44 (44%)	154 (51%)	
Female	56 (56%)	147 (49%)	
NYHA functional class			<0.001
I–II	7 (7%)	87 (29%)	
III–IV	93 (93%)	214 (71%)	
Home oxygen			0.02
Yes	29 (29%)	53 (18%)	
No	71 (71%)	248 (82%)	
6-minute walk distance (m)	315±159	387±138	<0.001
Brain natriuretic peptide (pg/mL)	597±571	182±295	<0.001
Right heart catheterization			
RAP	12±6	10±6	0.004
PAP systolic	90±10	66±21	<0.001
PAP diastolic	34±9	24±8	<0.001
PAP mean	55±10	40±12	<0.001
PCWP	11±5	12±5	0.08
Cardiac index	1.6±0.4	2.3±0.6	<0.001
PVR	1374±378	544±248	<0.001
TPR	1575±438	724±278	<0.001
Targeted PH medical therapy			<0.001
Yes	38 (38%)	55 (18%)	
No	62 (62%)	246 (82%)	
Hospital admission for RHF	26 (26%)	14 (5%)	<0.001

NYHA, New York Heart Association; RAP, right atrial pressure; PAP, pulmonary artery pressure; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance; TPR, total pulmonary resistance; PH, pulmonary hypertension; RHF, right heart failure.
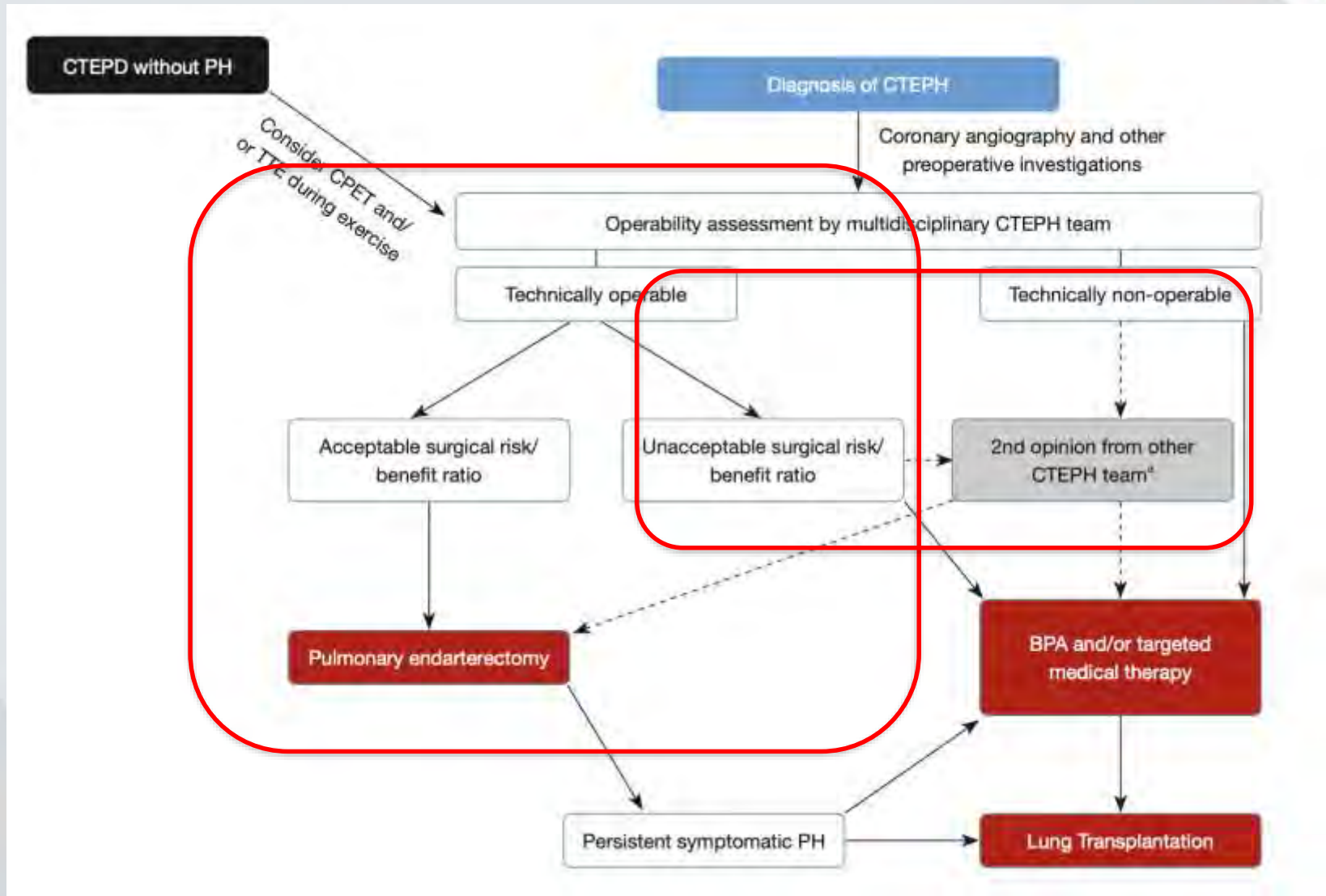
Medical optimization

For high risk features, technically operable

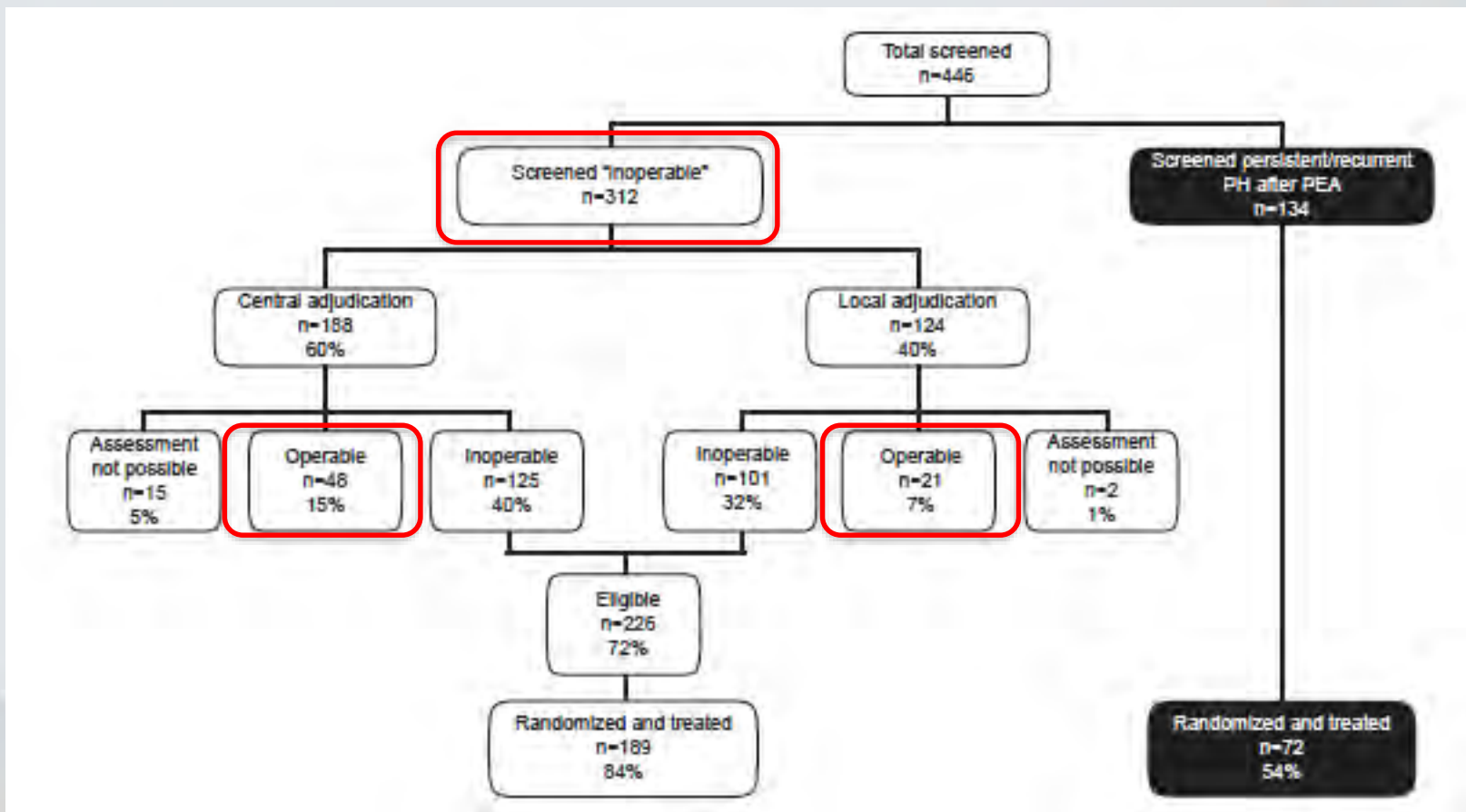Lower PVR- riociguat

Diuresis

Optimize co-morbidities

Concomitant CABG for amenable CAD

Reassess 'inoperable' patients

20+% may be operable

Surgical approach

Sternotomy

Cardiopulmonary bypass

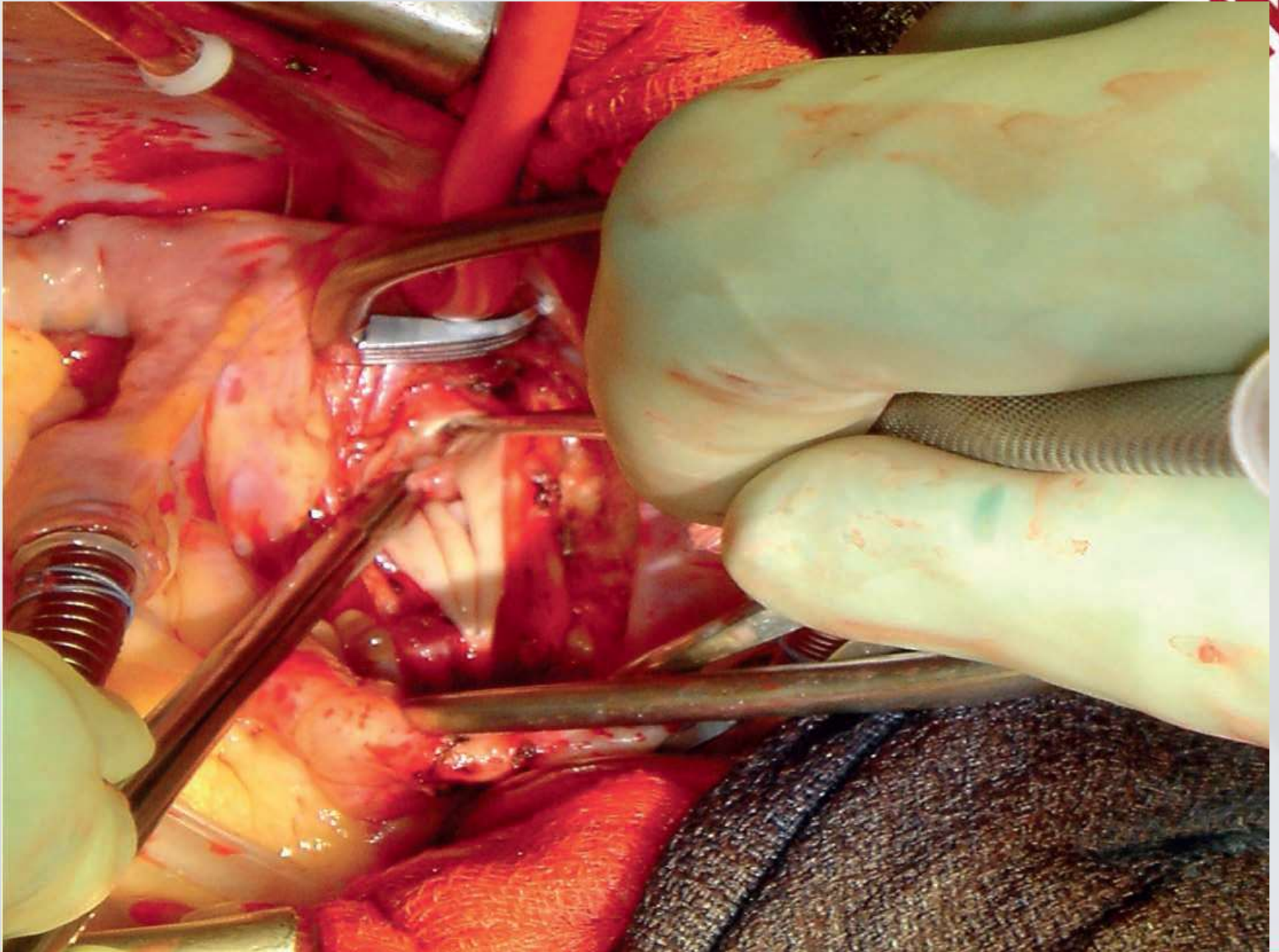Systemic cooling- deep hypothermia

Cardiac arrest

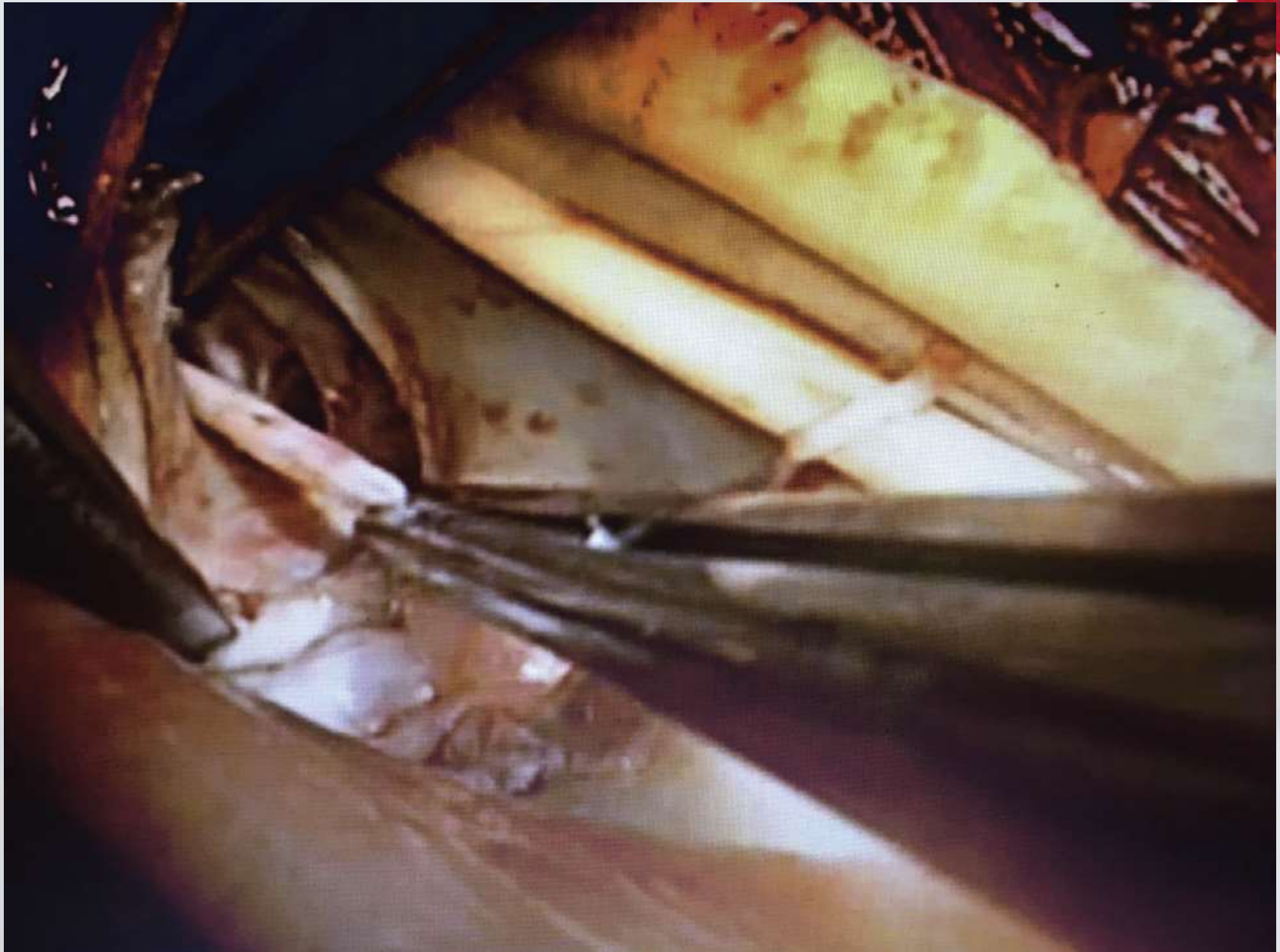R and L pulmonary arteries opened sequentially

Plane of dissection is developed

Circulation ceased to assist endarterectomy

Rewarming

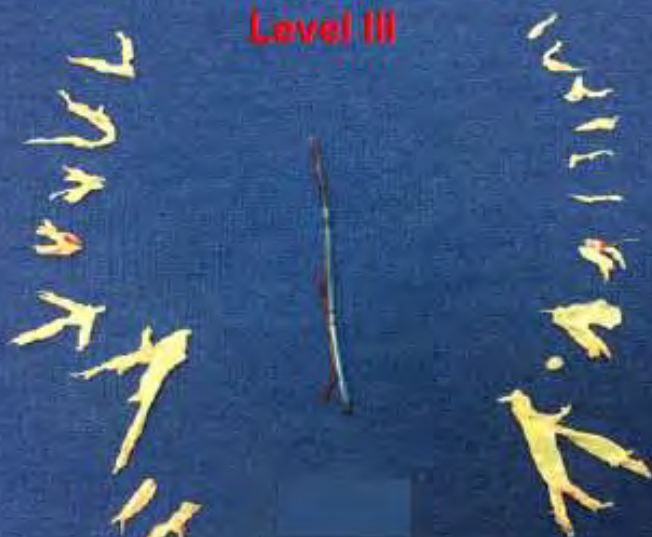Weaning from cardiopulmonary bypass with optimization of ventilation, volume status, systemic vascular tone.
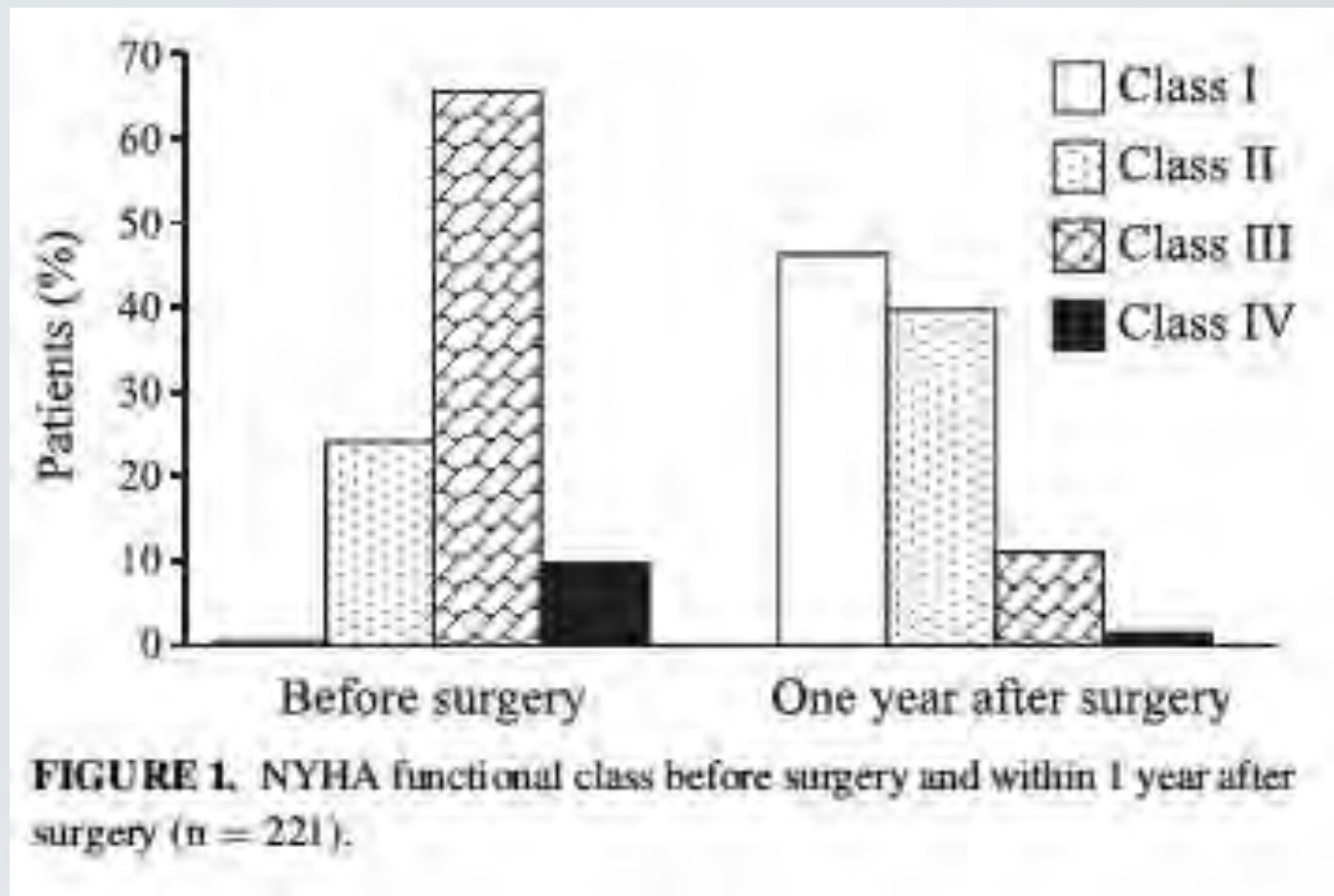
Complications

Reperfusion edema

Airway hemorrhage

Residual PH

RV failure

Functional outcome with PEA

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

Functional outcome with PEA
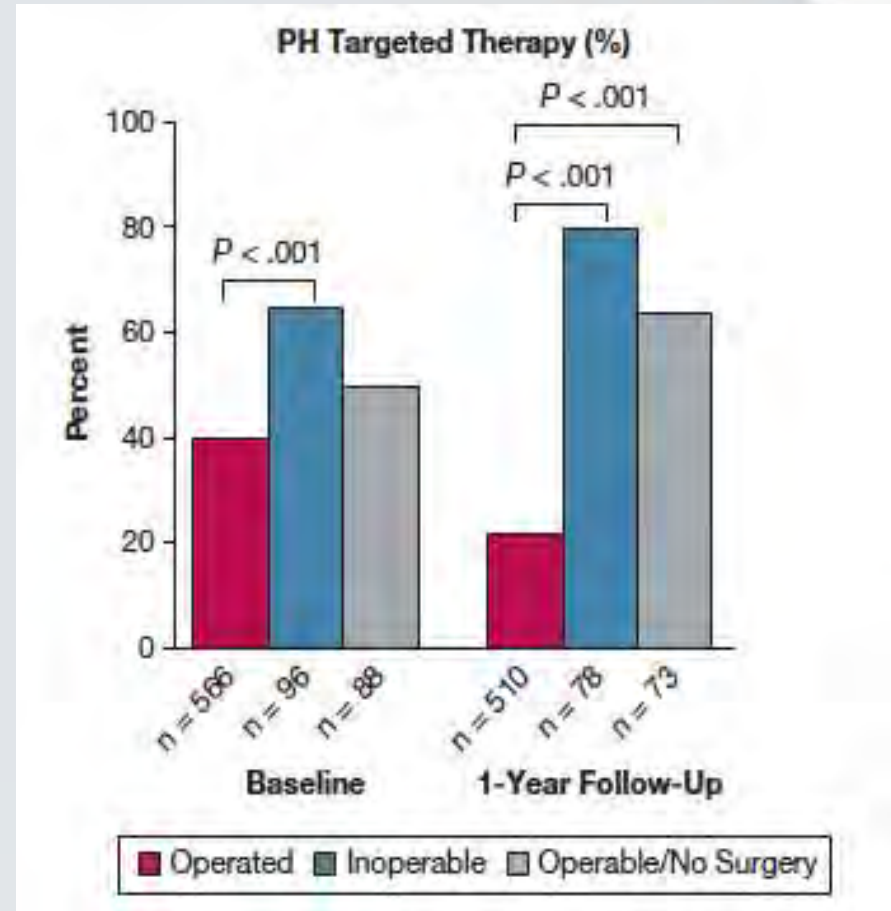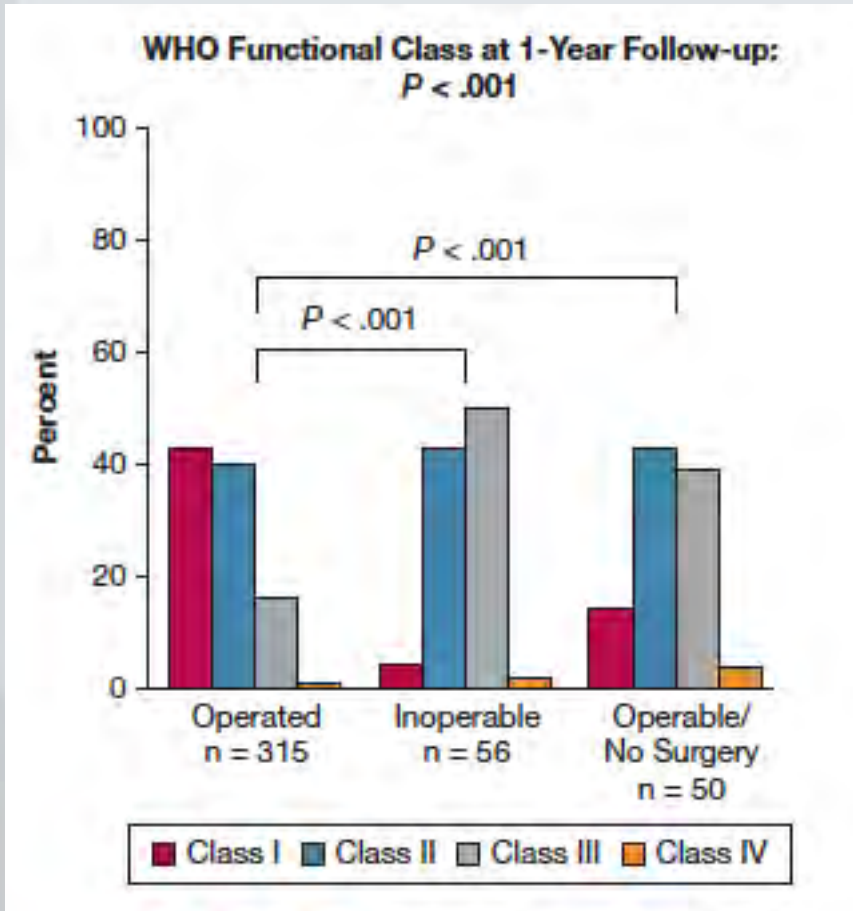
Long term outcome with PEA

Long term outcomes

Cumulative Survival vs Months from Diagnosis

NOT operated, N = 275
Operated, N = 404

p<0.0001 (log-rank test)
patients at risk at the end of the time period

404	383	374	366	361	355	335	244	158
275	246	228	214	200	188	164	120	58
0	6	12	18	24	30	36	42	48
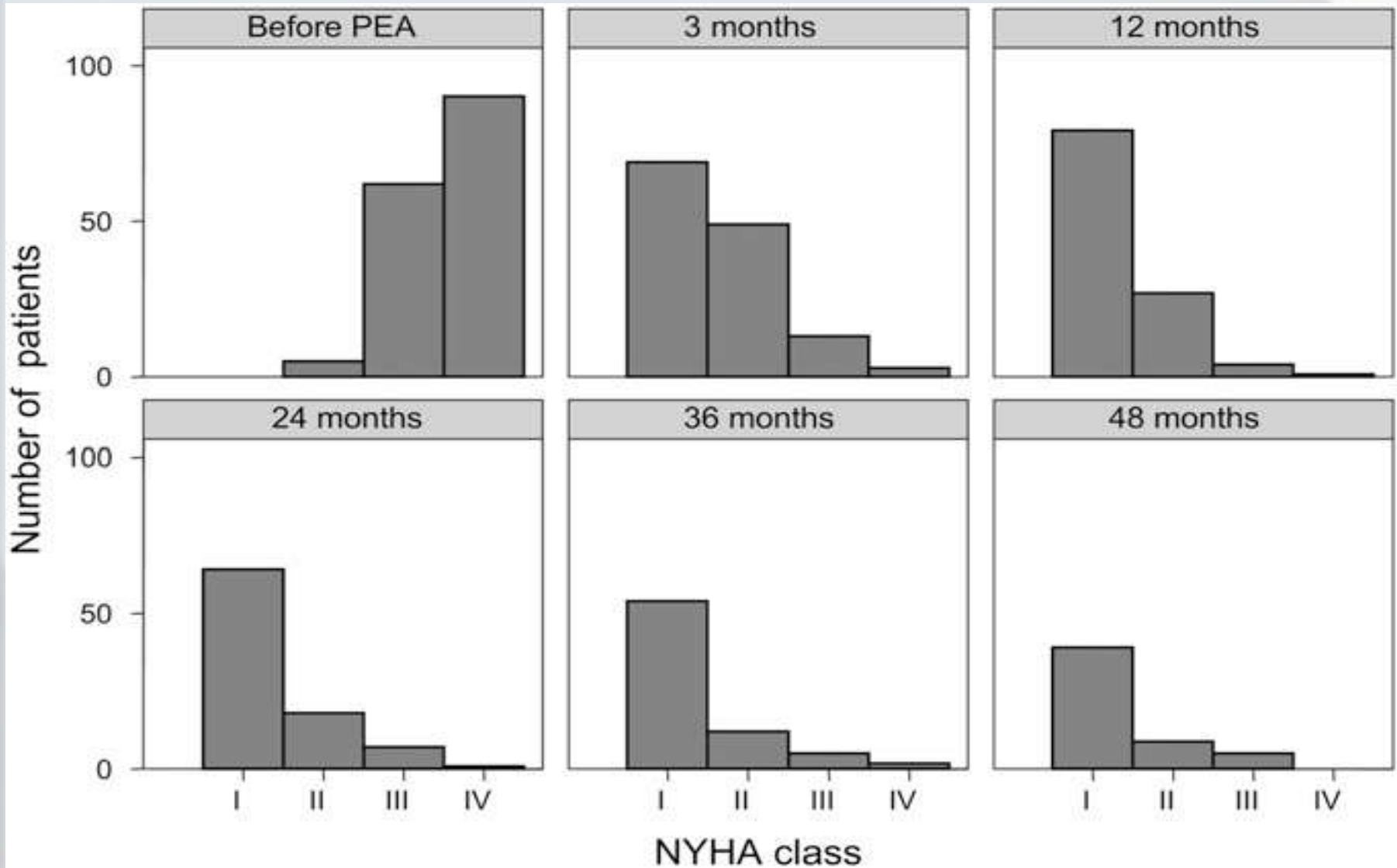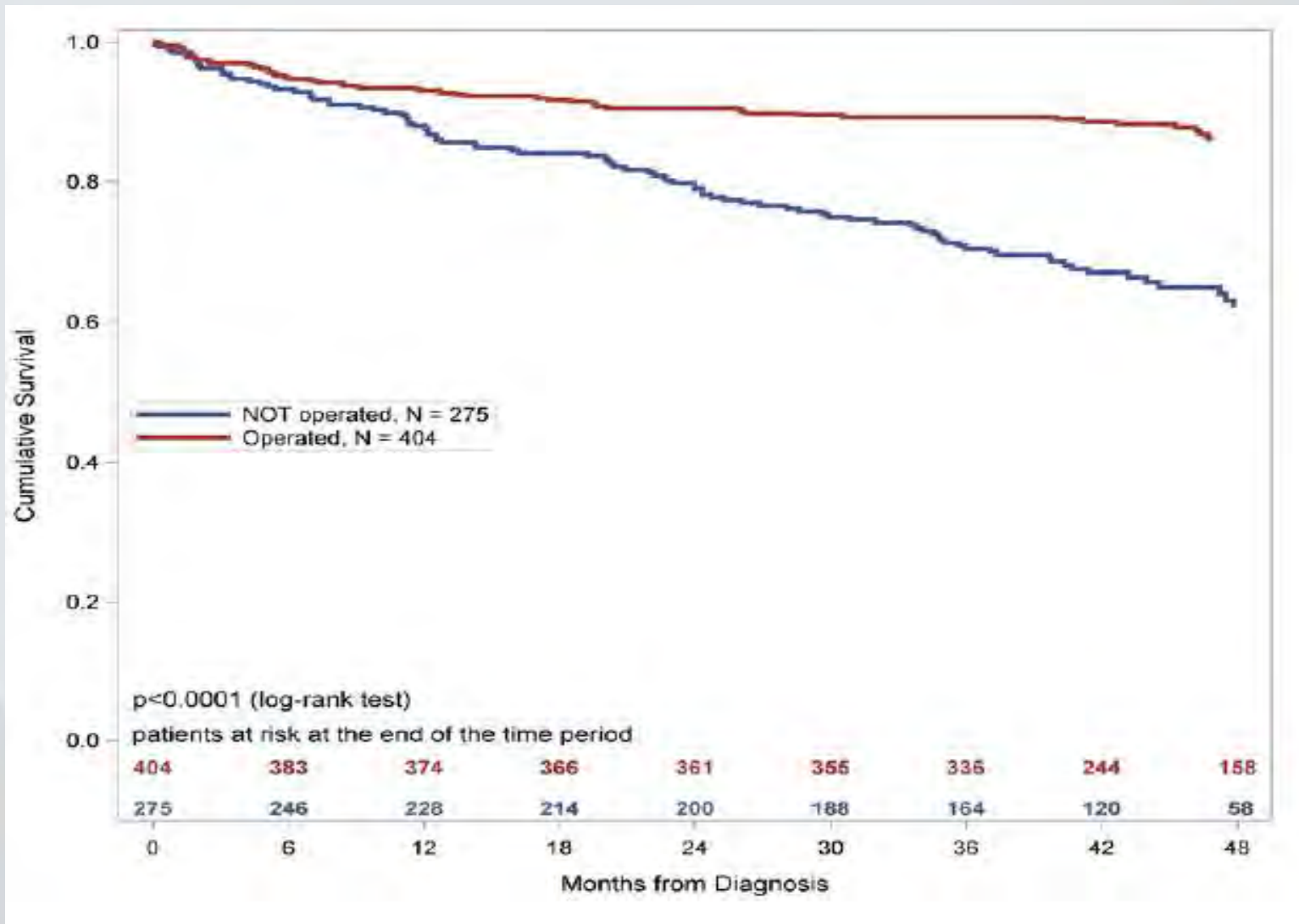
Long term outcomes
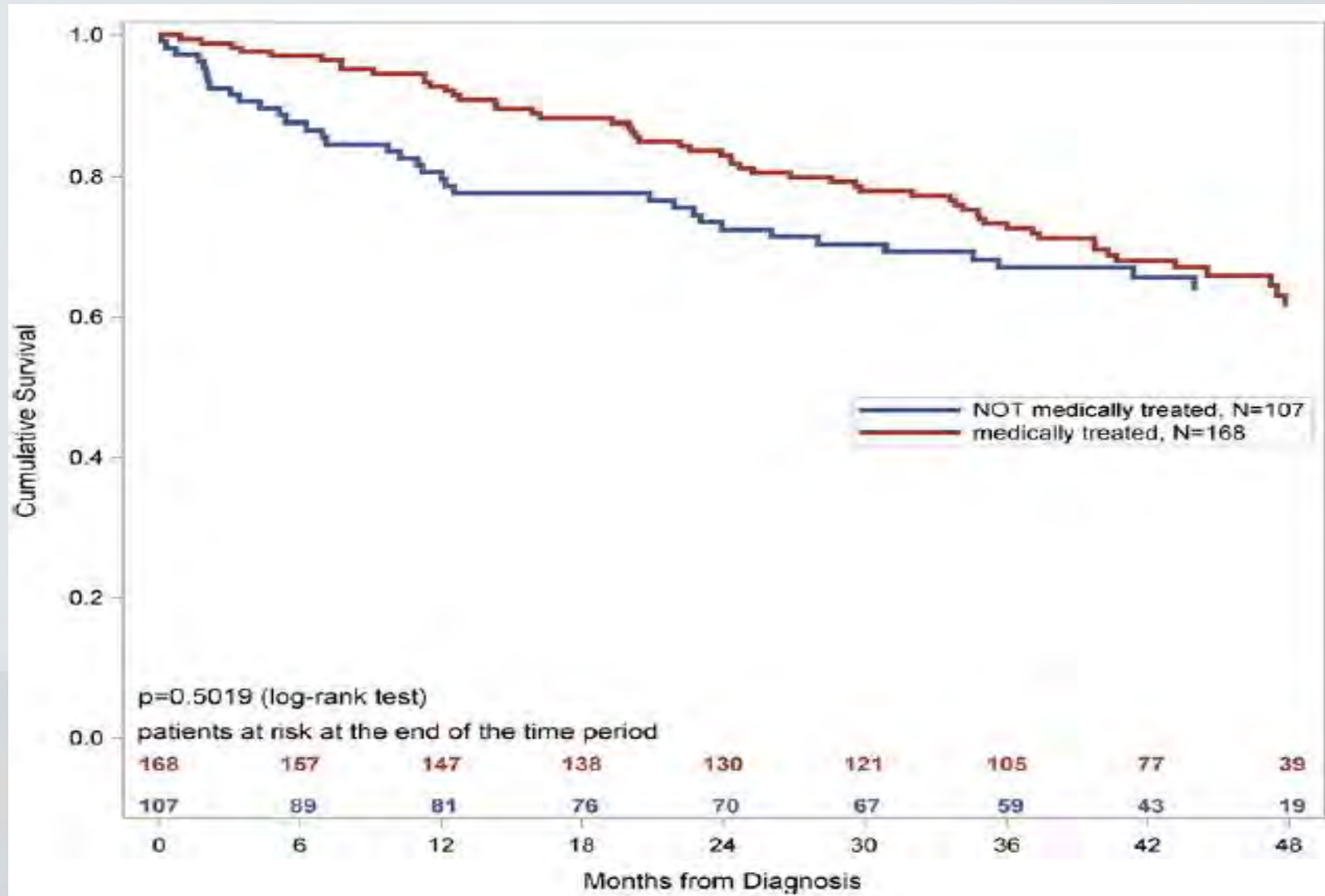
Survival with/without surgery

Table 5. Multivariable Correlates of Mortality for All Patients

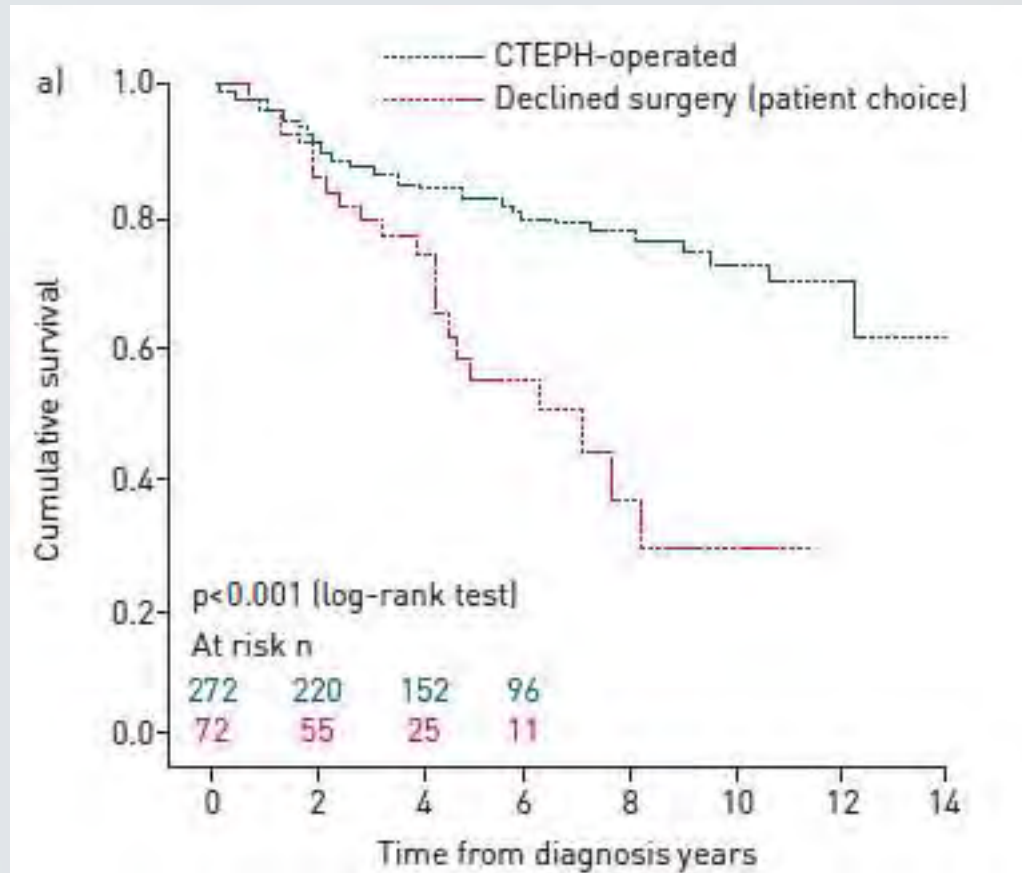|	HR Risk factor		
	HR	95% CI	P Value
PEA performed, yes vs no	0.37	0.24–0.58	<0.0001
Age, /10 y	1.27	1.05–1.53	0.0142
NYHA class			
III vs I/II	1.24	0.61–2.56	0.55
IV vs I/II	2.81	1.25–6.28	0.0118
RAP, /6 mm Hg	1.38	1.14–1.67	0.0011
History of cancer, yes vs no	2.04	1.23–3.39	0.0059
CHF or LV systolic/diastolic dysfunction, yes vs no	2.16	1.20–3.87	0.0097
Dialysis-dependent renal failure, yes vs no	13.32	1.79–99.17	0.0115

Multivariable analysis including PEA in all CTEPH patients. CHF indicates congestive heart failure; CI, confidence interval; CTEPH, chronic thromboembolic pulmonary hypertension; HR, hazard ratio; LV, left ventricle; NYHA, New York Heart Association; PEA, pulmonary endarterectomy; and RAP, right atrial pressure.
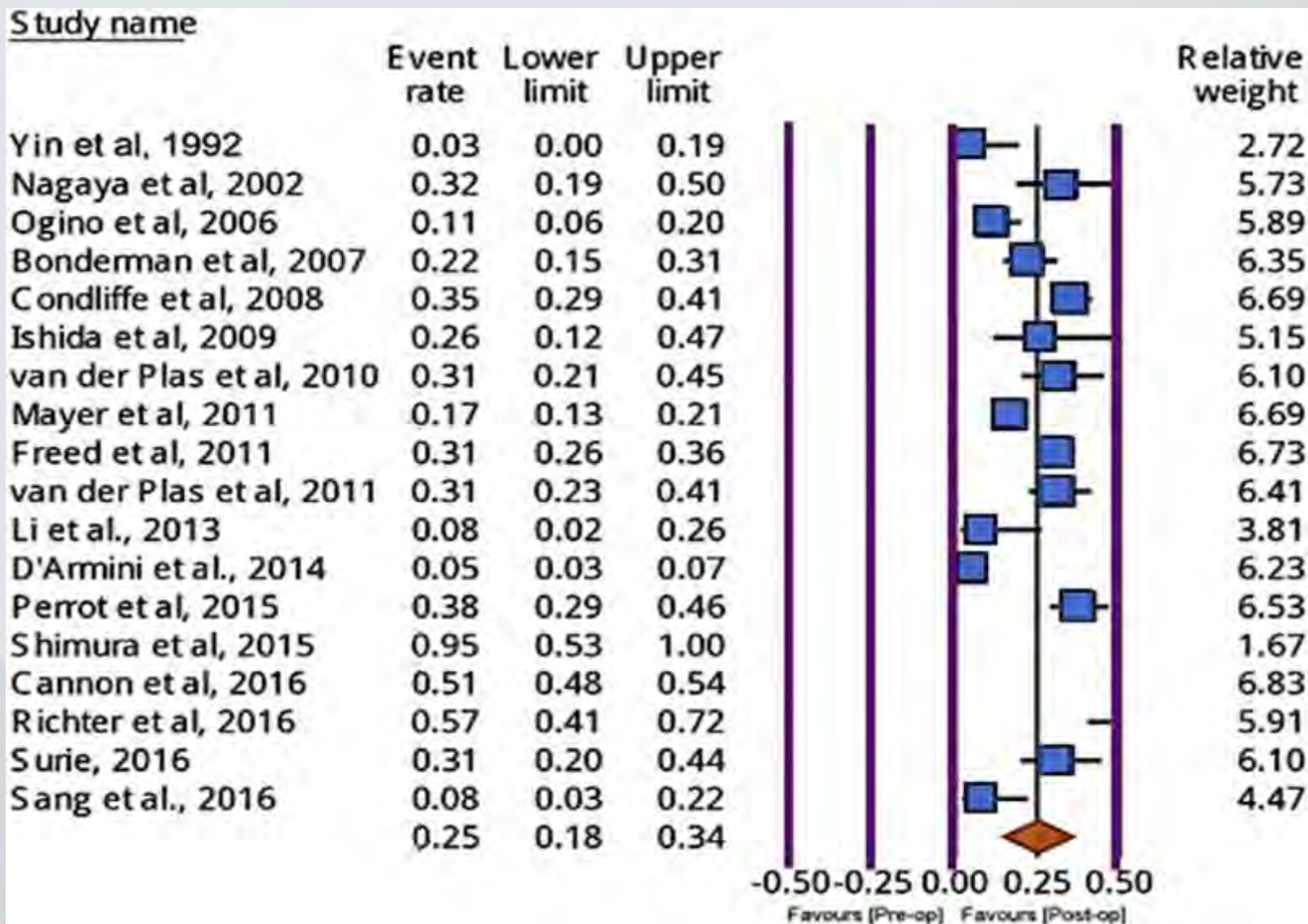
Severely elevated Pulmonary Vascular Resistance (PVR)

Table 2 Surgical and outcome characteristics

Characteristics	PVR >1,000 dynes.s.cm^{-5} (n=100)	PVR <1,000 dynes.s.cm^{-5} (n=301)	P value
Duration			
CBP (min)	252±32	246±41	0.18
Aortic cross clamp (min)	138±26	130±29	0.01
Circulatory arrest (min)	46±15	40±13	0.001
Combined procedure			
CABG, valve replacement	8 (8%)	13 (4%)	0.15
Jamieson classification			0.24
Type 1–2	69 (69%)	190 (63%)	
Type 3–4	31 (31%)	111 (37%)	
Improvement in TPR	1,088±473	373±273	<0.001
Duration of intubation (days)			
Mean ± SD	5.8±8.3	3.6±5.5	0.003
Median [IQR]	3 [2–6]	2 [1–4]	
Duration of ICU (days)			
Mean ± SD	9.2±10.6	5.8±6.3	0.001
Median [IQR]	5 [4–9]	4 [2–6]	
Duration of hospital stay (days)			
Mean ± SD	21.9±17.9	17.1±18.3	0.02
Median [IQR]	15 [11–27]	12 [9–17]	
Post-operative ECMO	12 (12%)	8 (3%)	0.002
30-day mortality	4 (4%)	6 (2%)	0.26

CPB, cardiopulmonary bypass; CABG, coronary artery bypass grafting; TPR, total pulmonary resistance; SD, standard deviation; IQR, interquartile range; ECMO, extracorporeal membrane oxygenation.

Residual PH after surgery

Study name	Event rate	Lower limit	Upper limit		Relative weight
Yin et al, 1992	0.03	0.00	0.19		2.72
Nagaya et al, 2002	0.32	0.19	0.50		5.73
Ogino et al, 2006	0.11	0.06	0.20		5.89
Bonderman et al, 2007	0.22	0.15	0.31		6.35
Condliffe et al, 2008	0.35	0.29	0.41		6.69
Ishida et al, 2009	0.26	0.12	0.47		5.15
van der Plas et al, 2010	0.31	0.21	0.45		6.10
Mayer et al, 2011	0.17	0.13	0.21		6.69
Freed et al, 2011	0.31	0.26	0.36		6.73
van der Plas et al, 2011	0.31	0.23	0.41		6.41
Li et al., 2013	0.08	0.02	0.26		3.81
D'Armini et al., 2014	0.05	0.03	0.07		6.23
Perrot et al, 2015	0.38	0.29	0.46		6.53
Shimura et al, 2015	0.95	0.53	1.00		1.67
Cannon et al, 2016	0.51	0.48	0.54		6.83
Richter et al, 2016	0.57	0.41	0.72		5.91
Surie, 2016	0.31	0.20	0.44		6.10
Sang et al., 2016	0.08	0.03	0.22		4.47
	0.25	0.18	0.34		

-0.50 -0.25 0.00 0.25 0.50

Favours [Pre-op]   Favours [Post-op]

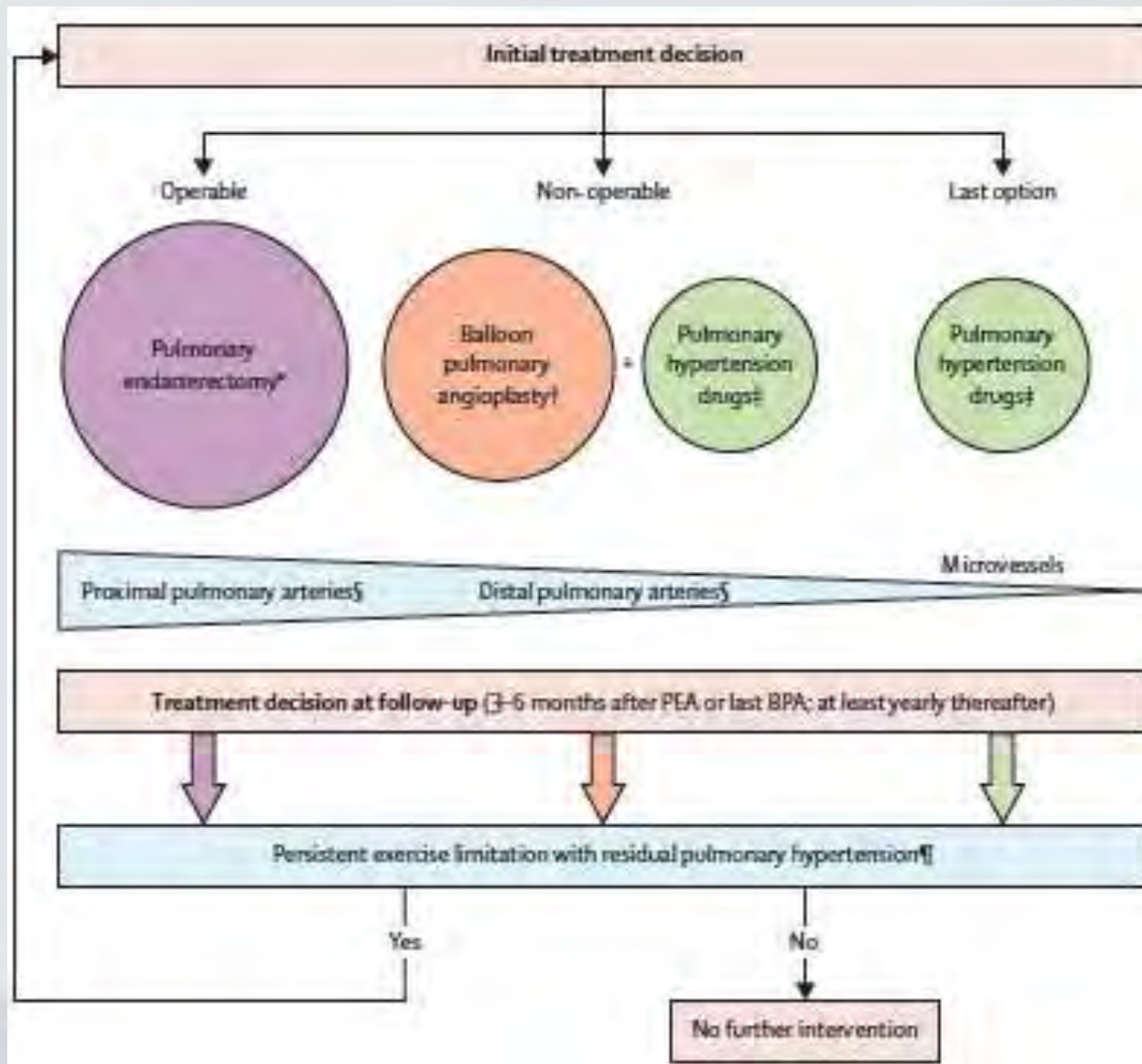# Riociguat for the Treatment of Chronic Thromboembolic Pulmonary Hypertension

Important trial proving the efficacy of Riociguat

Subjective (NYHA class) and objective measures (walk distance- 6MWD, vascular resistance- PVR)

Included patients with PH after surgery

These patients also benefited from Riociguat

Persistent exercise limitation with residual PH

Symptoms plus mean PAP > 30

Combination of PH targeted therapy and BPA

BPA to treat residual distal lesions

Summary

Surgery associated with significant benefit-strongest predictor

Surgery is difficult and complex

Success depends on selection, optimization, execution

Team of experts