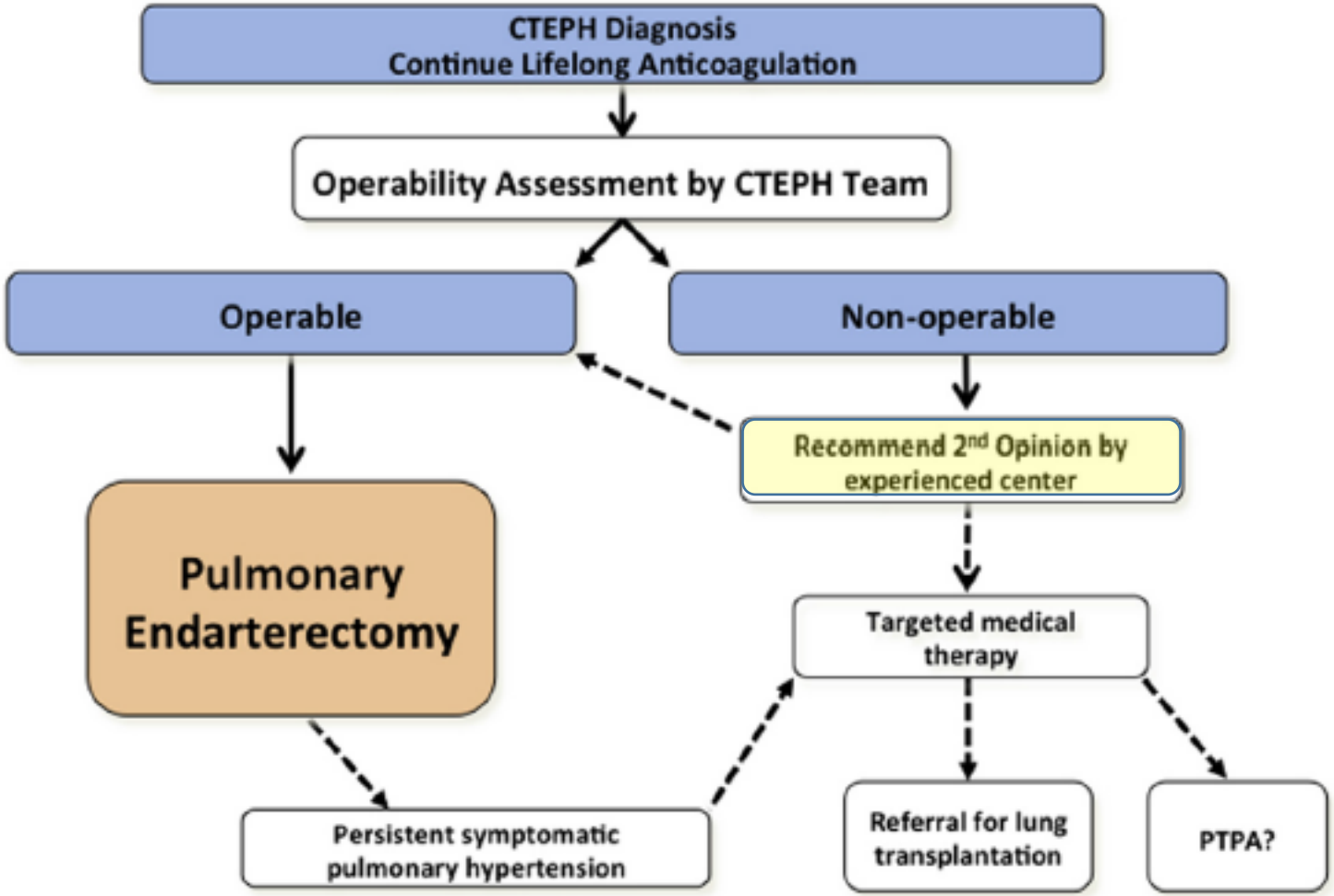


JACC Vol. 62, No. 25, Suppl D, 2013
December 24, 2013:D92–9

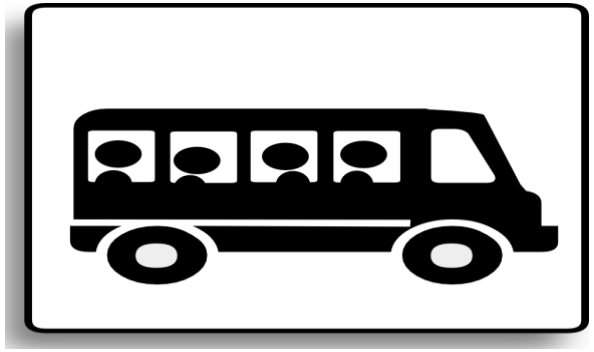
Second opinion of an experienced PEA center is suggested in the CTEPH management algorithm. Would you ask for such an opinion:

1. Only in case your local surgeon would ask for a second opinion?
2. In all pts disqualified from PEA by your local surgeon?
3. In all pts qualified or disqualified from PEA by your local surgeon?

Clinical Case (early Dec, 2012)

- 32 year old lawyer,
- since 2001 DDD pacemaker implanted for severe vasovagal syndrome

- Overworked
- decided to take a short relax and went skiing...
- took a bus ride from Warsaw to Grenoble



- Day 1st – somewhat short of breath on the slopes
- Day 2nd – difficulty to reach the ski lift..
- Addmitted to a local municipal hospital
- Clinical and CT-angio diagnosis: acute PE
- Standard AC treatment started

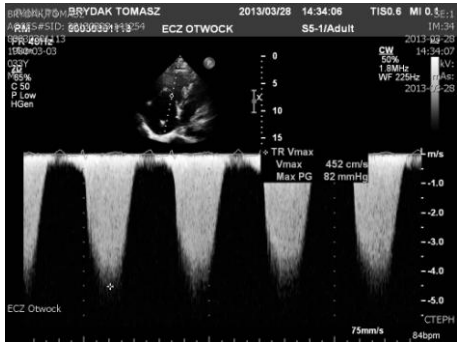
- Transferred to University Hospital of Grenoble
  - Dr Helen Bouvaist, referral PH unit
- Suspicion of distal CTEPH + an acute PE episode?

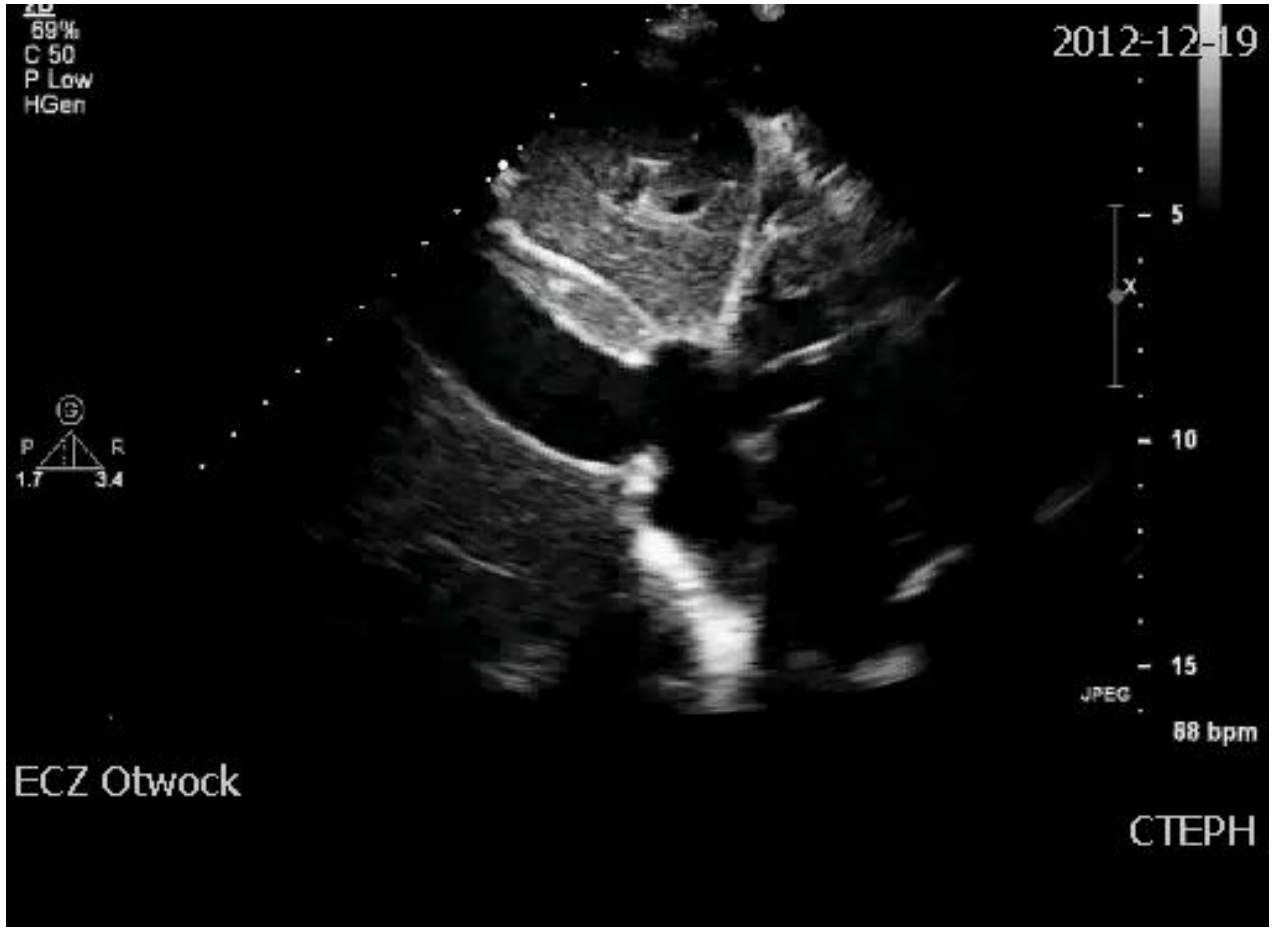
- Transfer to our center: ECZ-Otwock
  - to continue AC treatment
  - to select optimal management strategy

- Arrived in III WHO FC

Dept. Pulmonary Circulation & Th-E diseases
Center of Postgraduate Medical Education
ECZ-Otwock, Poland

Echocardiography

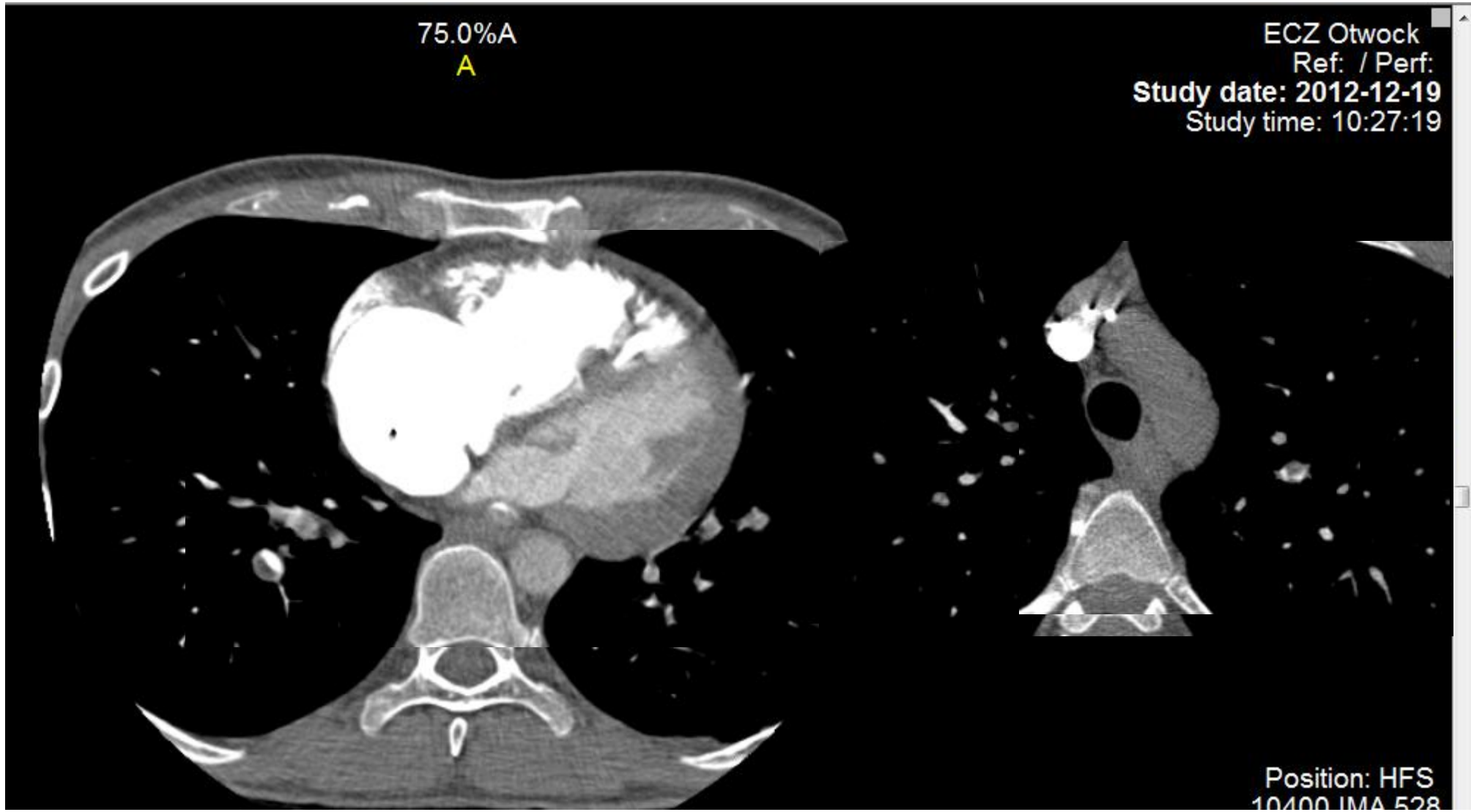




- TVPG = 82 mm Hg
- IVC= 27 mm (exp=insp)
- TAPSE 13 mm
- RVEDd 58 mm (LVEDd 35 mm)
- RAA 29 cm2 (LAA 15 cm2)
- suspected lead vegetations on TTE - not confirmed on TEE

CT - angiography

CT - angiography

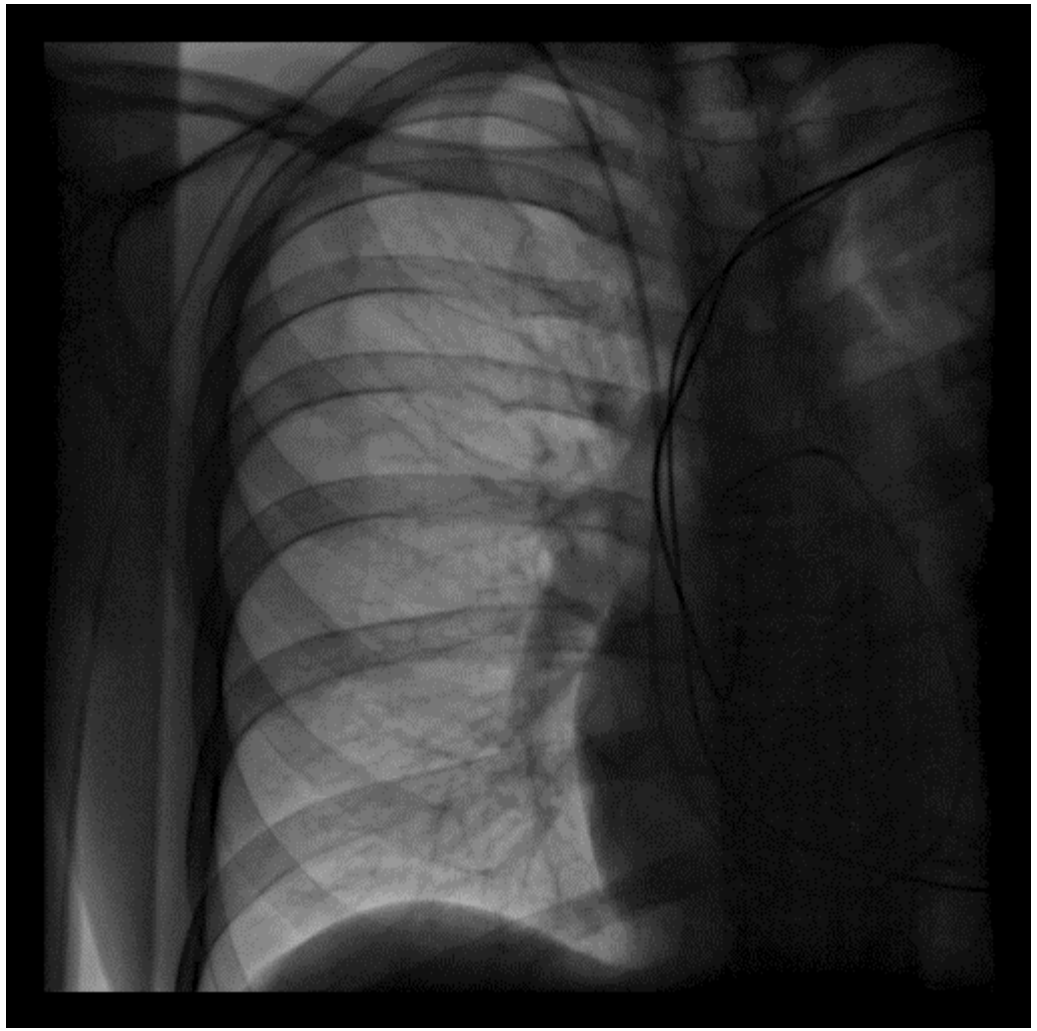


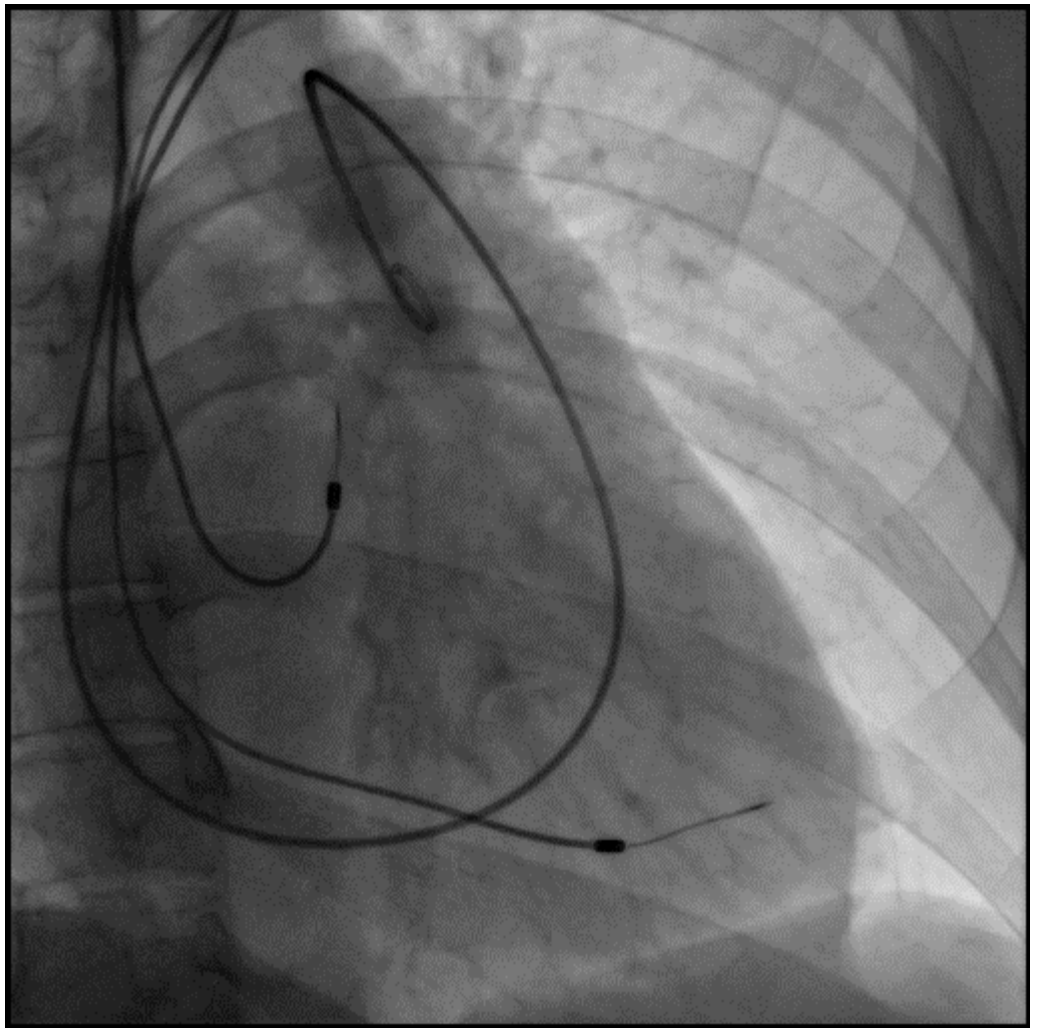
RHC (15th Feb 2013)

HR	101			
RAPa/v/m	23	12	18	mmHg
RVPs/d/-ed	90	9	21	mmHg
PAPs/d/m	89	40	53	mmHg
PCWP a/v/m			9	mmHg
SAPs/d/m	116	69	80	mmHg
CO		4,76		l min-1
SV		47		ml
SVI		24		ml/m2
SatO2art		95		%
SatO2mv		65		%

CI	2,46	l min m-2
TPG	44	mmHg
TPR	11,13	mmHg l-1 min
	890	dyn sek cm-5
PVR	9,24	mmHg l-1 min
	739	dyn sek cm-5
SVR	13,03	mmHg l-1 min
	1 041	dyn sek cm-5
PVR/SVR	0,71	

Pulmonary angiography

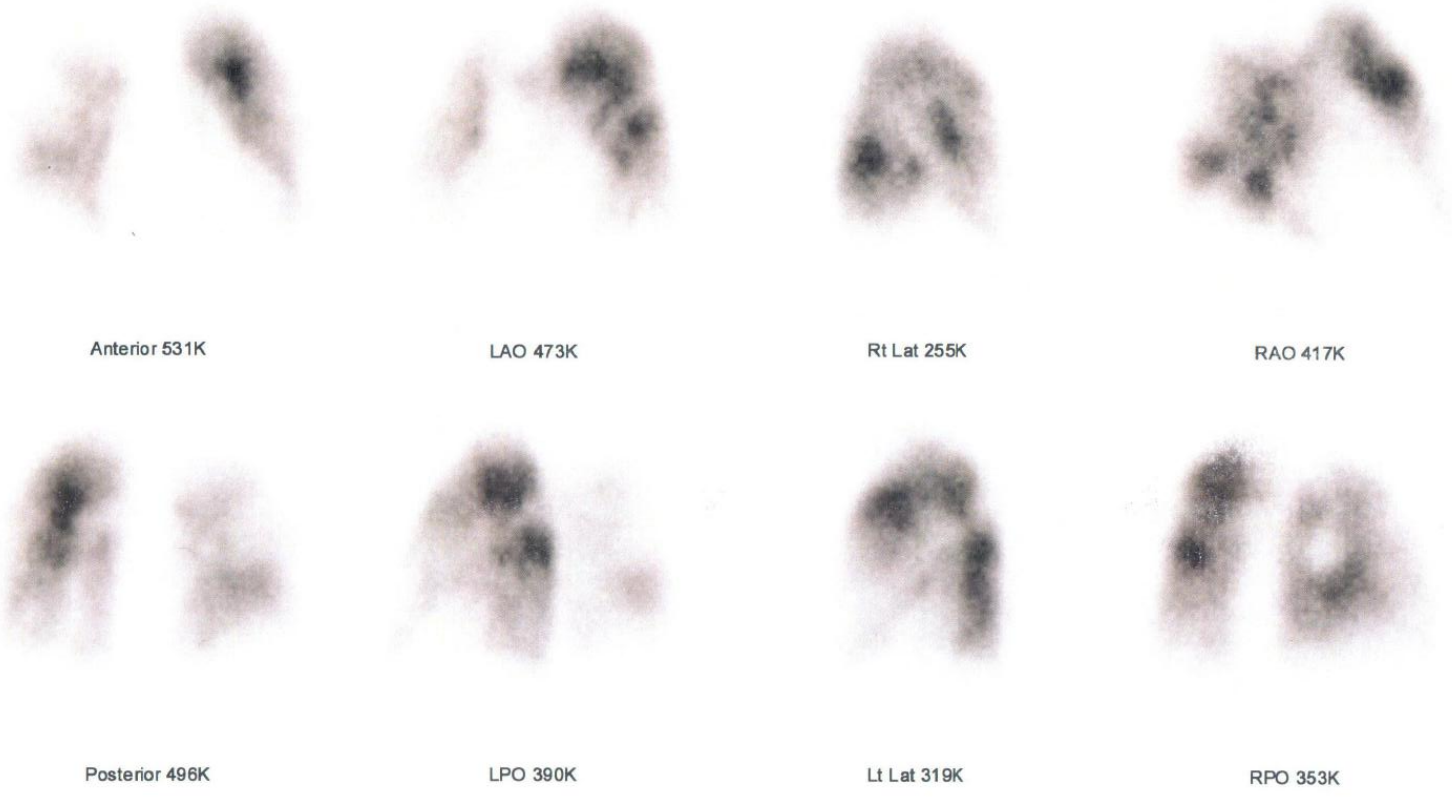




Lung Perfusion Scintigraphy (21st Feb 2013)

Mazowiecki Szpital Bródnowski, Zakład Diagnostyki Obrazowej, Warszawa tel.22-326-54-06
Brydak Tomasz DOB: 1980-03-03 ID: 8003030113
STUDY: Lung Perfusion Scan STUDY DATE: 2013-02-21 ACCESSION #: 306/13

99m Technetium
185,0 MBq (5,00 mCi) MAA



Your next step would be to:

1. Remove the pacemaker leads; after > 10 years of chronic infection is the most likely source of recurrent pulmonary emboli and pulmonary hypertension
2. Refer to PEA-surgeon despite distal location of CTEPH changes originating from the intracardiac leads
3. Try medical therapy and/or BPA before attempting PEA because its risk/benefit ratio is highly questionable .

- Normal CRP/WBC/body temperature/blood cultures
- Consulted at referral electrophysiologic laboratory (National Institute of Cardiology) where the pacemaker was implanted/followed

  ✓still pacemaker dependent,
  ✓TEE: no indications to remove the leads

- Started on sildenafil waiting for decision on PEA

Papworth Hospital NHS

NHS Foundation Trust

Thank you very much for asking us to review the above patient's case on our MDT meeting. We discussed the case in the presence of Mr Jenkins, Pulmonary Endarterectomy Surgeon, Dr Screaton and Dr Gopalan, Vascular Radiologists. For the benefit of our records I will briefly summarise the case. This is 33 year old male with the implanted DDR pacemaker in 2001. More recently the patient has complained of a dry cough and peripheral oedema. He is in functional WHO Class III.

The opinion of the group was that the patient has got chronic thromboembolic pulmonary hypertension with mixed distribution of the disease, more proximal on the right side. We believe that the patient should benefit from the pulmonary endarterectomy however he is of a higher risk to be left with residual pulmonary hypertension as there is distal distribution on the left side of the circulation. Additionally a high diastolic PA pressure on right heart catheter and the risks associated with pacemaker leads could suggest component of the distal distribution. However bearing in mind the very young age of the patient, the current functional status and PVR of 739 he should overall benefit from the pulmonary endarterectomy.

IV. EPIKRYZA

33 - letni pacjent z ciężkim nadciśnieniem płucnym o etiologii zakrzepowo zatorowej, po wszczepieniu ukł
stymulujacego typu DDD został przyjęty do Oddziału Kardiochirurgii Szpitala Allenort celem operacyjnego
leczenia CTEPH. Zabieg wykonano 06.06.2013. Z zastosowaniem krążenia pozaustrojowego, w całkowity
zatrzymaniu krążenia i głębokiej hypotermii wykonano endarterectomię obu tętnic płucnych. Zmiany o
charakterze obwodowym, niewielkie zmiany w głównych tętnicach płucnych bardziej nasilone po stronie le
nie do końca korelowało ze zmianami
utrzymujace się na
odłączeniem od kra
powodu nasilonego
konkretnego miejsc
krwawienia. W drug
kaniule od ECMO.
okresowo wlewem
neurologicznych- pr
tlenoterapia bierna
zachowaniem diure
Zastosowano leczer
worek osierdziowy u
Po przeniesieniu na
stwierdzono znaczn
około 100 ml do dnia następne. W kolejnych dobach bardzo powolna poprawa z intensywną rehabilitacją
oddechową i w niewielkim stopniu ruchową.
Od 25.06 znaczące pogo
do 80%. W kontrolnym e
szmer oskrzelowy w nad
środkowym i dolnym płuc
od operacji wydawało sie
oddechową, nieinwazyjn
niedodmowym. 27.06 wy
segmentarnych. Pobranc
kolonie Candida spp. Uz
dwukrotnie bronchofibero
segmentarne. W kontroln
zagęszczenia śródmiąższ
Pacjent intensywnie reha
płucnymi. Pacjent w dalsz
wykonywanych bronchos
Clexanem w dawce 1 x 6
Rana pooperacyjna zagoj
ECZ celem kontynuacji leczenia i dalsze kontrole.

ednio po operacji stwierdzono w
du niewydolności krążenia i trud
ECMO tetniczo-żylnego. 07.06
z rewizją śródpiersia, nie stwier
CT uzyskując zmniejszenie
spomagania krążenia i usunętc
niewielkim wlewem Dobutrexu i
cjent wybudzony, bez deficytów
względnie wydolny, wysoka
. Powrót funkcji nerer z

ku osierdziowym do 2,5 cm, nal
a blaszek osierdzia po nakluciu
cji 88 - 92%. W wykonanym e
uzyskujac 300 ml krwistego płyn

nkową i spadkiem saturacji w spoc
wolnego płynu. Osłuchowo słyszaln
rtg klp zmiany zapalne w płacie
perfuzujnym, co z uwagi na odległy
ykoterapię, intensywną rehabilitacj
rdzono zmiany o charakterze zapa
zielinę z odetkaniem licznych oskrz
no wzrostu bakterii, a jedynie poje
nu i 95% na tlenie. Wykonano jesz
ponownie udrażniając liczne oskrz
dalszym ciągu utrzymauja się
). Zalecono intensywna nebulizacj
a się zmian osłuchowych nad pola
Pulmicortu i Berodualu, Z powodu
lantem, pacjent leczony przewlekl
mie osierdzia,
ynchozrost, mostek stabilny. Pacjent w dniu dzisiejszym przeniesiony

V. ZALECENIA DOTYCZACE DALSZEGO

6th June 2013 - PEA

Distal changes at PEA (Prof. A. Biederman)

Persistent PH/RVF post-op

ECMO a-v 48h

Intrathoracic bleeding; re-op 2nd day

Extubated 3rd day

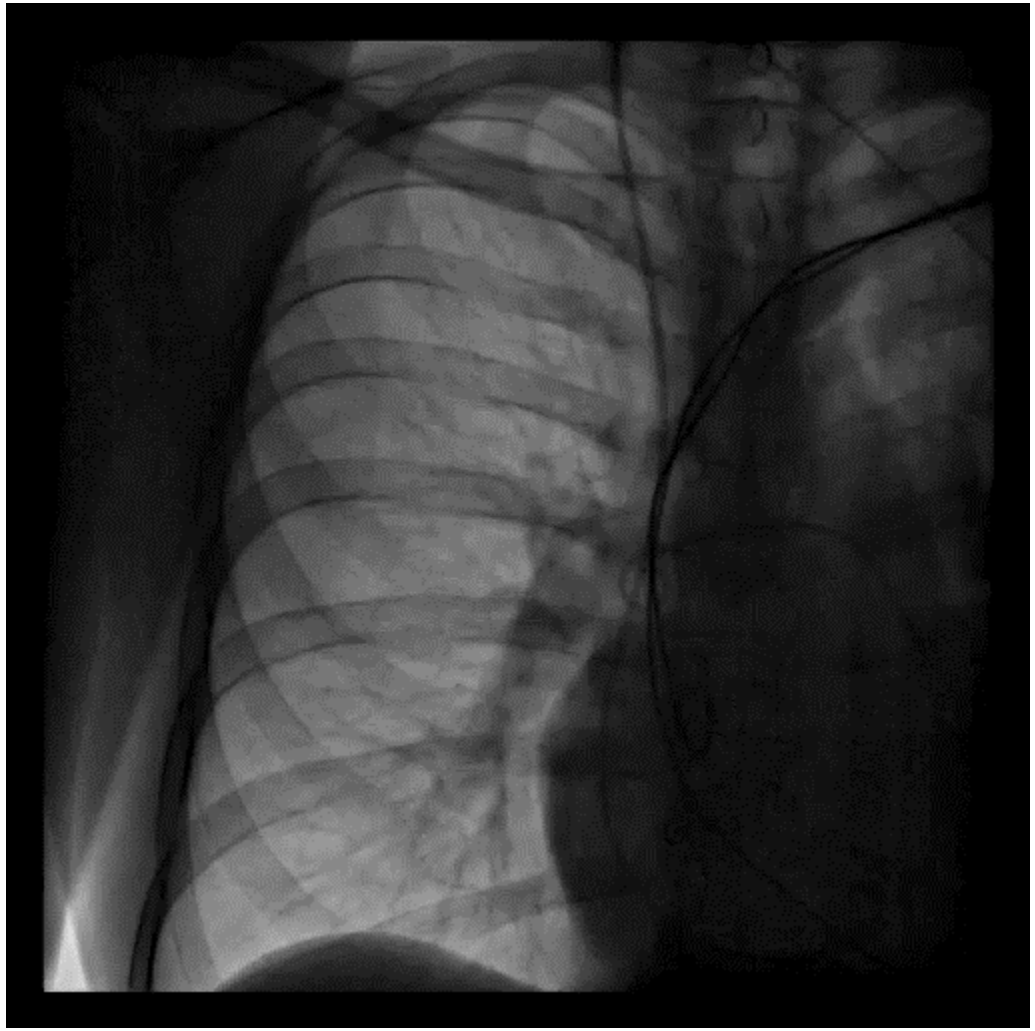
Pericardiocentesis

CRRT terminated on 7th day

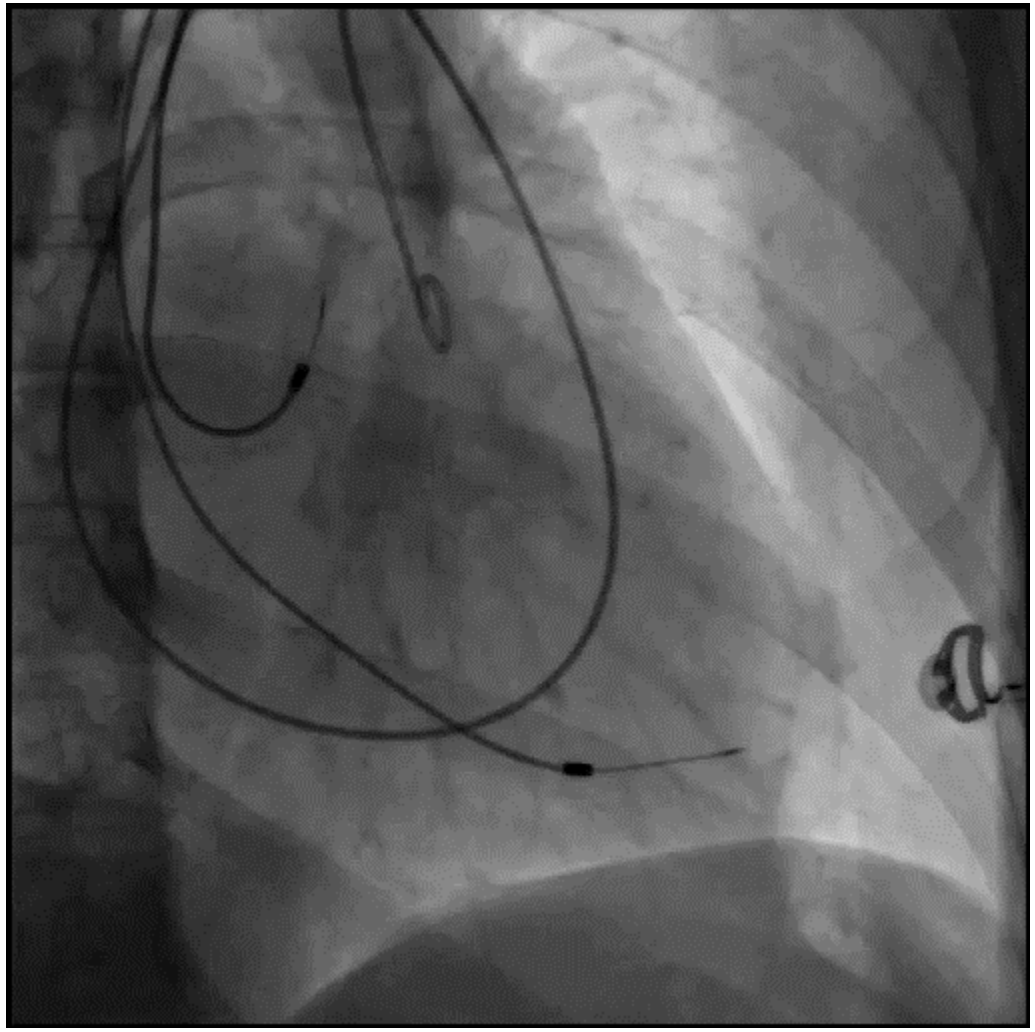
Pulmonary infection/respiratory failure

Overall 4 weeks at Cardiosurgical Dept.

Angiography 6 months post PEA:

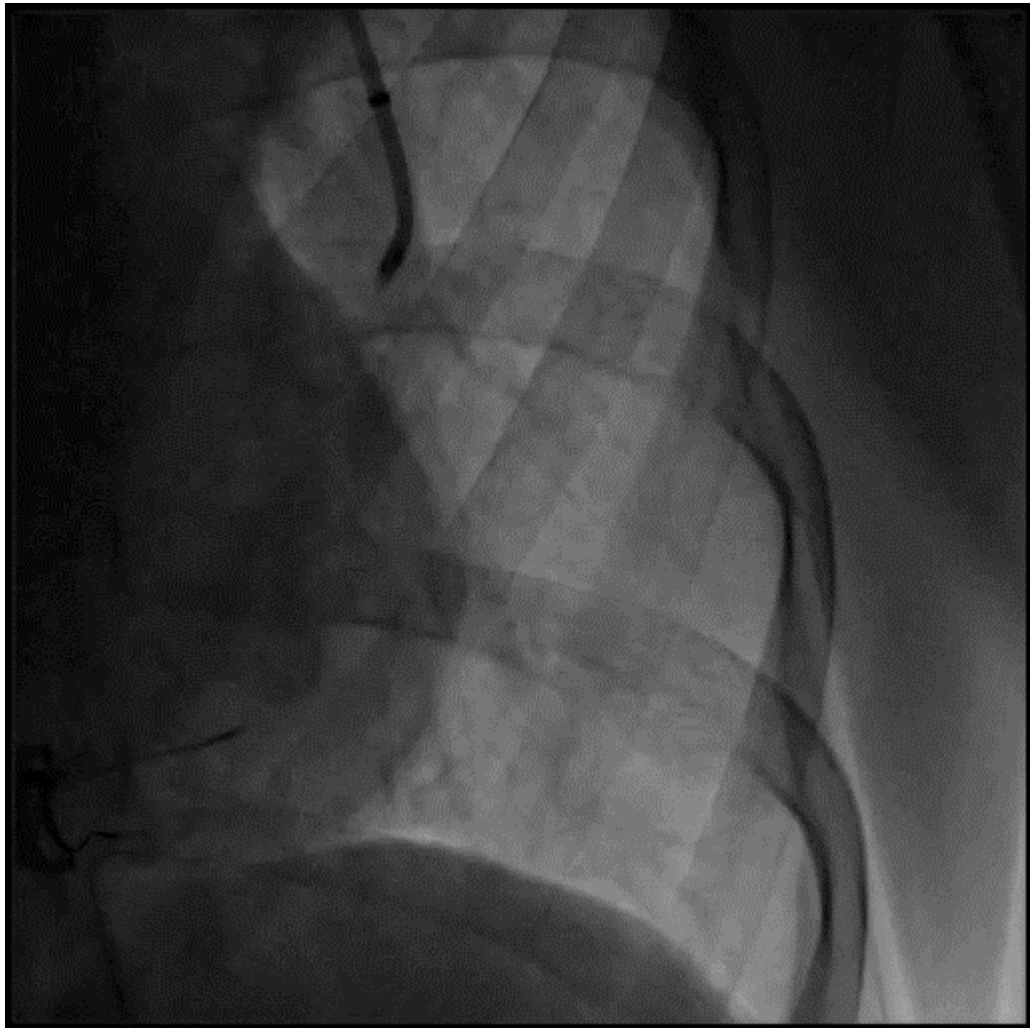


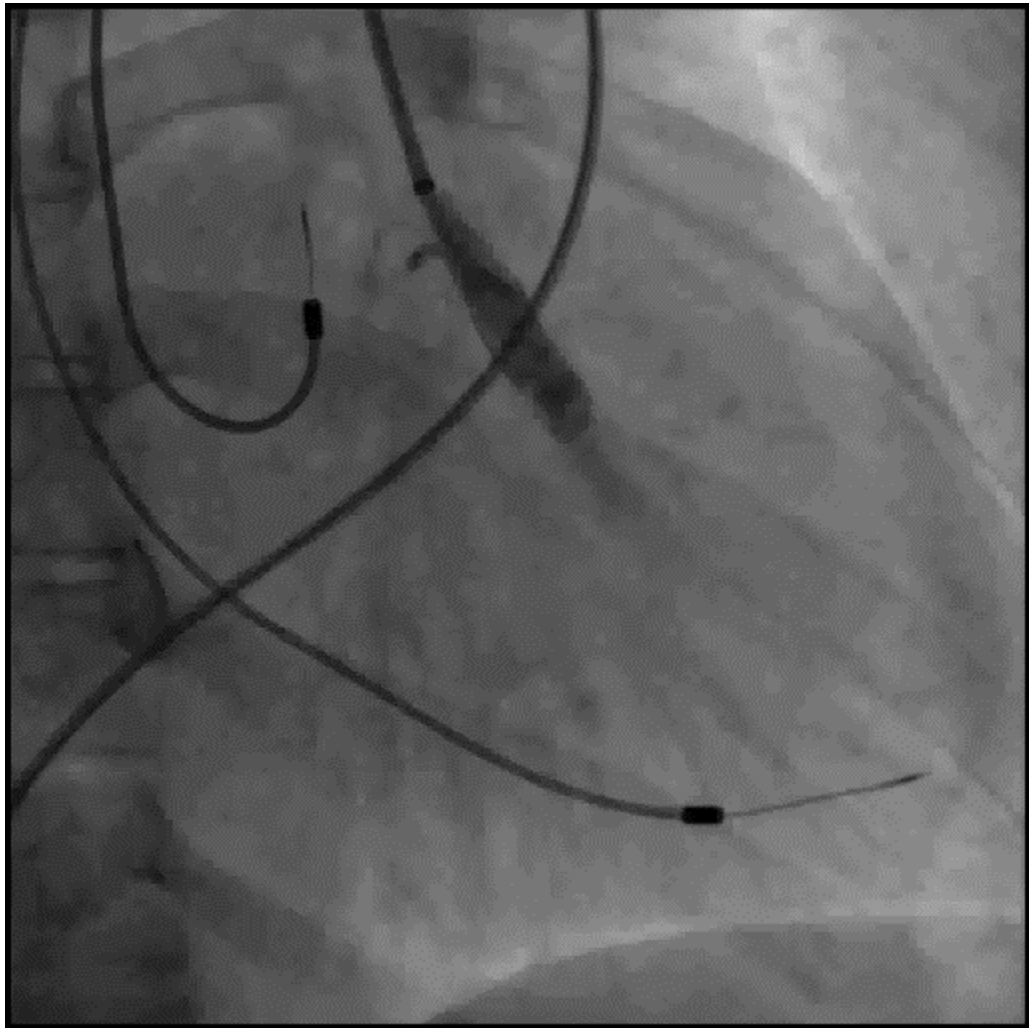
Angiography 6 months post post PEA:



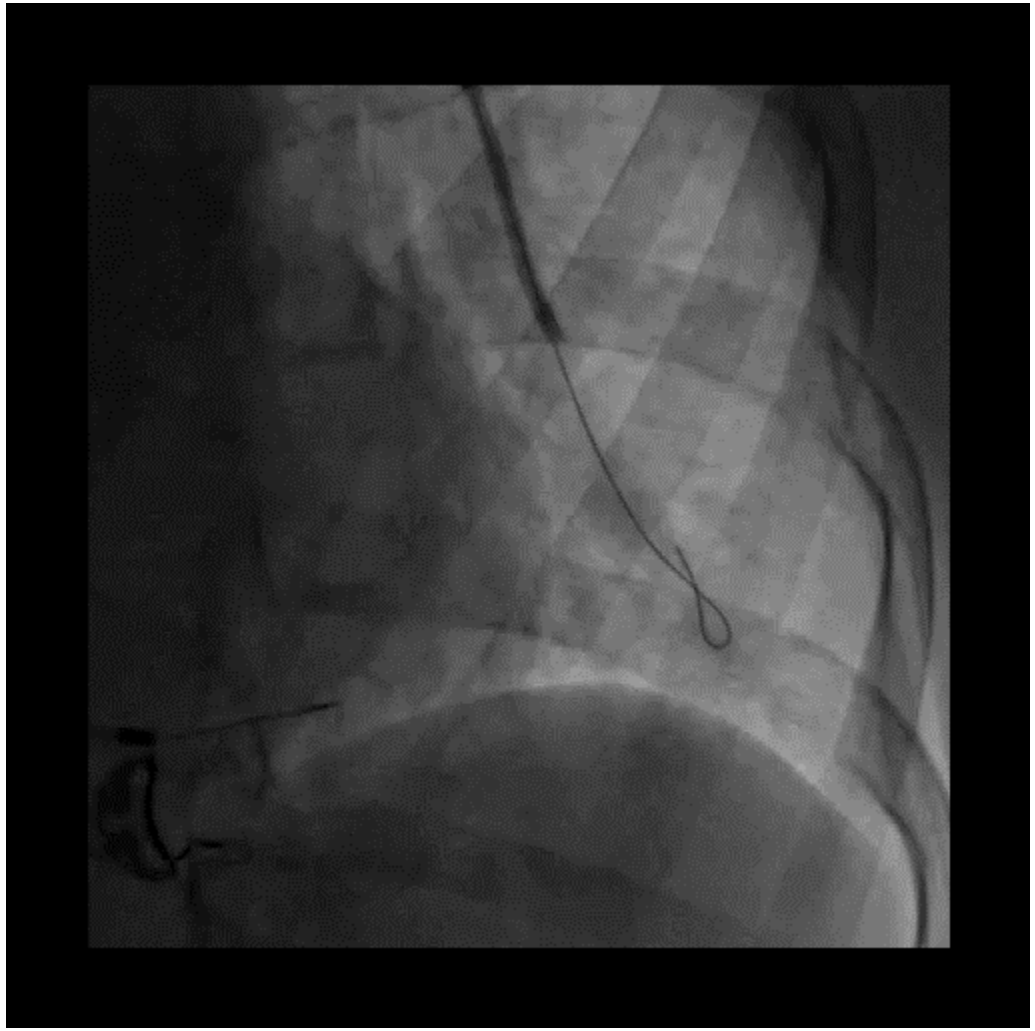
	Before PEA	After PEA
Functional class (WHO)	III	III
6MWT [m]	-	-
NT-proBNP [pg/ml]	4539	2377
PVR [Wood units]	9,24	8,28
mPAP [mmHg]	53	51
mRAP [mmHg]	18	12
PCWP [mmHg]	9	12
CO [l/min]	4,76	4,71
CI [l/min*m^2]	2,46	2,48

Balloon pulmonary angioplasty (Apr 22nd 2014)

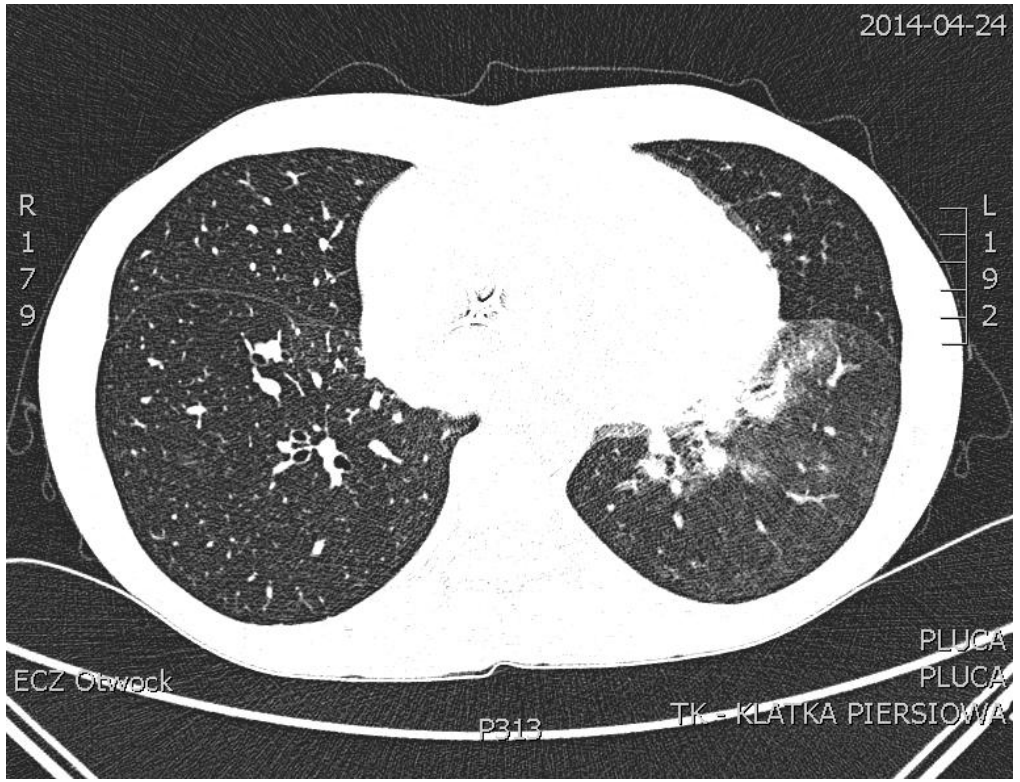




Angiography post PEA: reperfused segment



Local reperfusion edema and alveolar bleeding



Mild transient hemoptysis during BPA

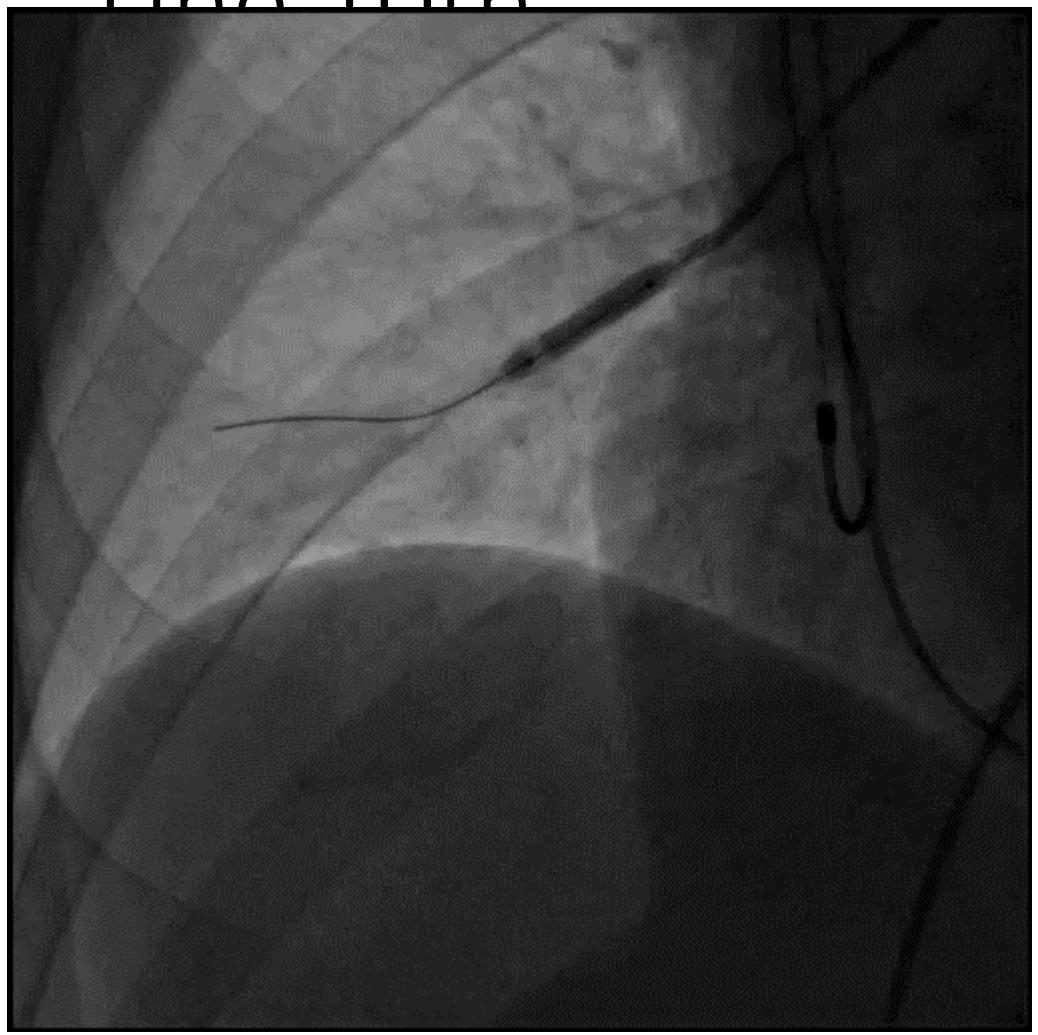
desaturation 97 – 93% over next 24 h

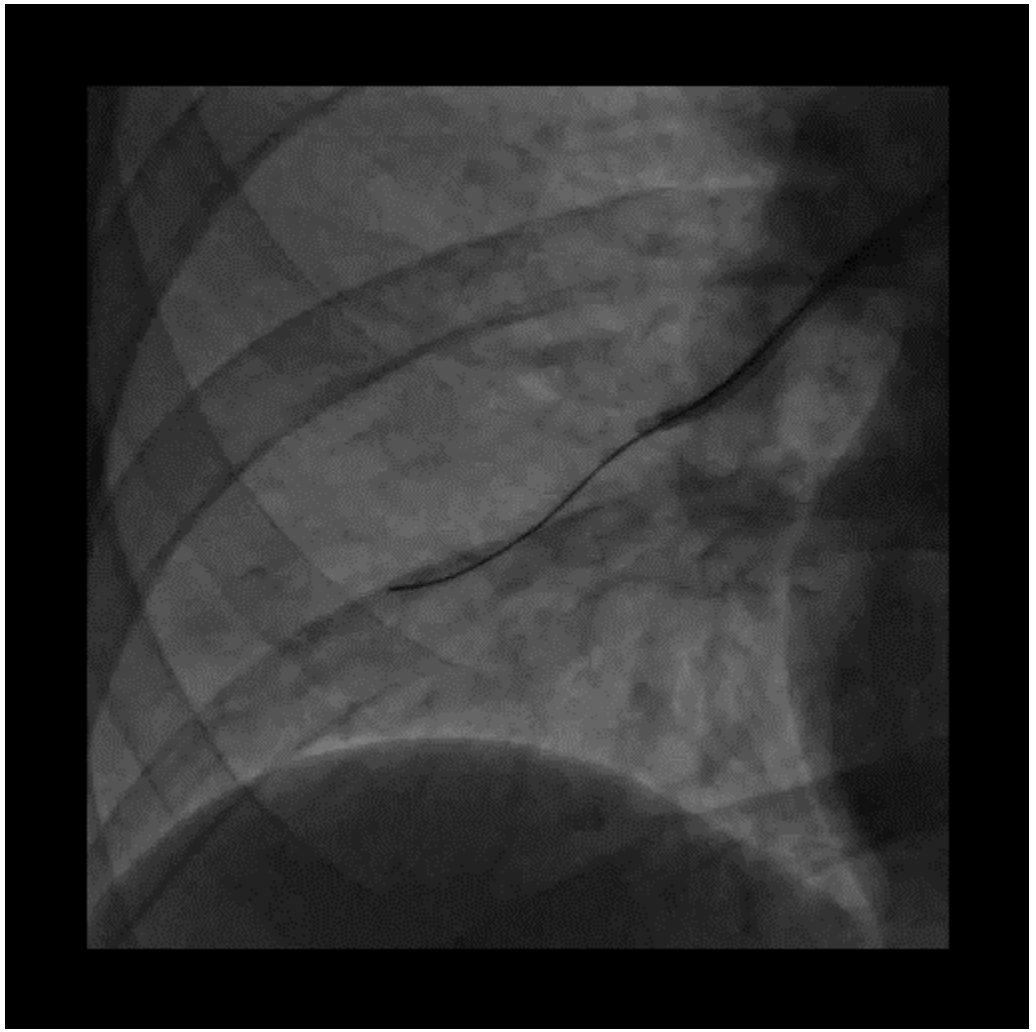
	Before PEA	After PEA	Before BPA_1	After BPA_1
Functional class (WHO)	III	III	III	-
6MWT [m]	-	-	480	-
NT-proBNP [pg/ml]	4539	2377	2756	-
PVR [Wood units]	9,24	8,28	7,68	8,02
mPAP [mmHg]	53	51	48	49
mRAP [mmHg]	18	12	12	17
PCWP [mmHg]	9	12	10	10
CO [l/min]	4,76	4,71	4,95	4,86
CI [$l/min*m^2$]	2,46	2,48	2,59	2,55

What would you do next

- Refrain from further BPA sessions (due to no-effect/high bleeding risk) and start riociguat therapy
- Refrain from further BPA sessions and also from riociguat therapy (contraindicated in pts with hemoptysis) and list for LTx
- Perform 2nd session of BPA, as planned

June 10th, 2014



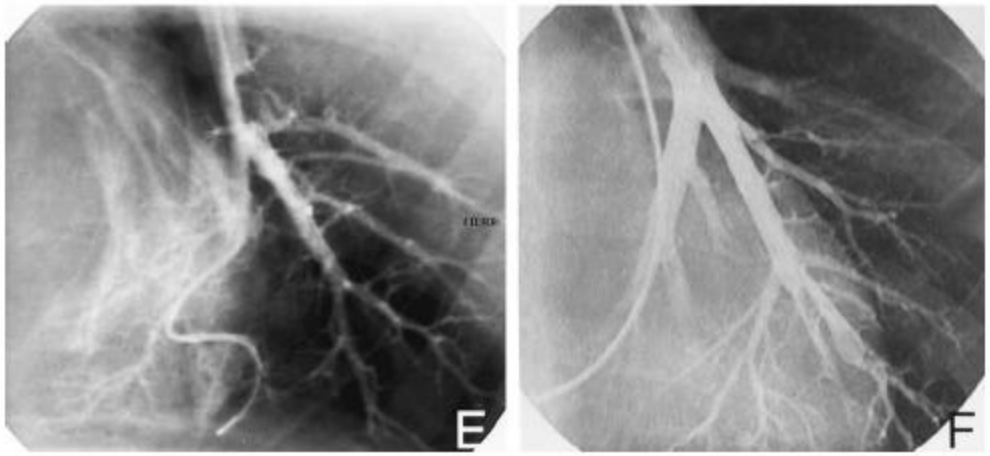


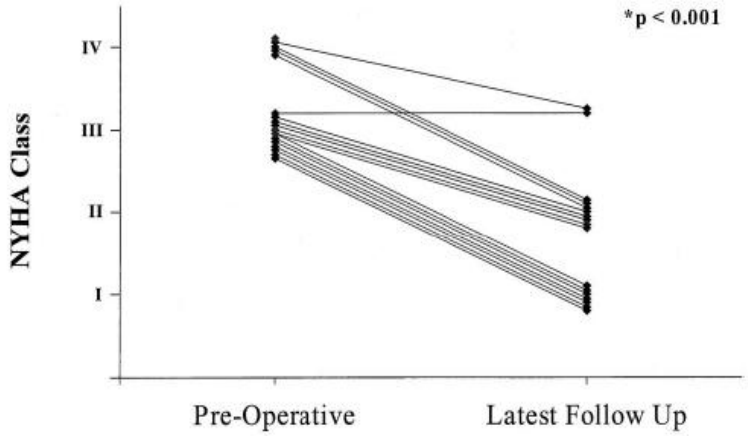
	Before PEA	After PEA	Before BPA_1	After BPA_1	Before BPA_2	After BPA_2
Functional class (WHO)	III	III	III	-	II	-
6MWT [m]	-	-	480	-	495	-
NT-proBNP [pg/ml]	4539	2377	2756	-	1530	-
PVR [Wood units]	9,24	8,28	7,68	8,02		
mPAP [mmHg]	53	51	48	49	43	42
mRAP [mmHg]	18	12	12	17	6	6
PCWP [mmHg]	9	12	10	10	10	10
CO [l/min]	4,76	4,71	4,95	4,86		
CI [l/min*m^2]	2,46	2,48	2,59	2,55		

Gaps in Evidence	Angioplasty in CTEPH
Main controversy:	**Friend Or Foe?**

Balloon Pulmonary Angioplasty for Treatment of Chronic Thromboembolic Pulmonary Hypertension

Jeffrey A. Feinstein, MD, MPH; Samuel Z. Goldhaber, MD; James E. Lock, MD;
Susan M. Ferndandes, PA-C; Michael J. Landzberg, MD





Circulation 2001

Gaps in Evidence	Angioplasty in CTEPH
Main controversy:	Friend Or Foe?

A171.E1609
JACC Maarch 9, 2010
Volume 55, issue 10A

VASCULAR DISEASE

THE EFFICACY AND THERAPEUTIC ROLE OF BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

ACC Poster Contributions
Georgia World Congress Center, Hall B5
Monday, March 15, 2010, 3:30 p.m.-4:30 p.m.

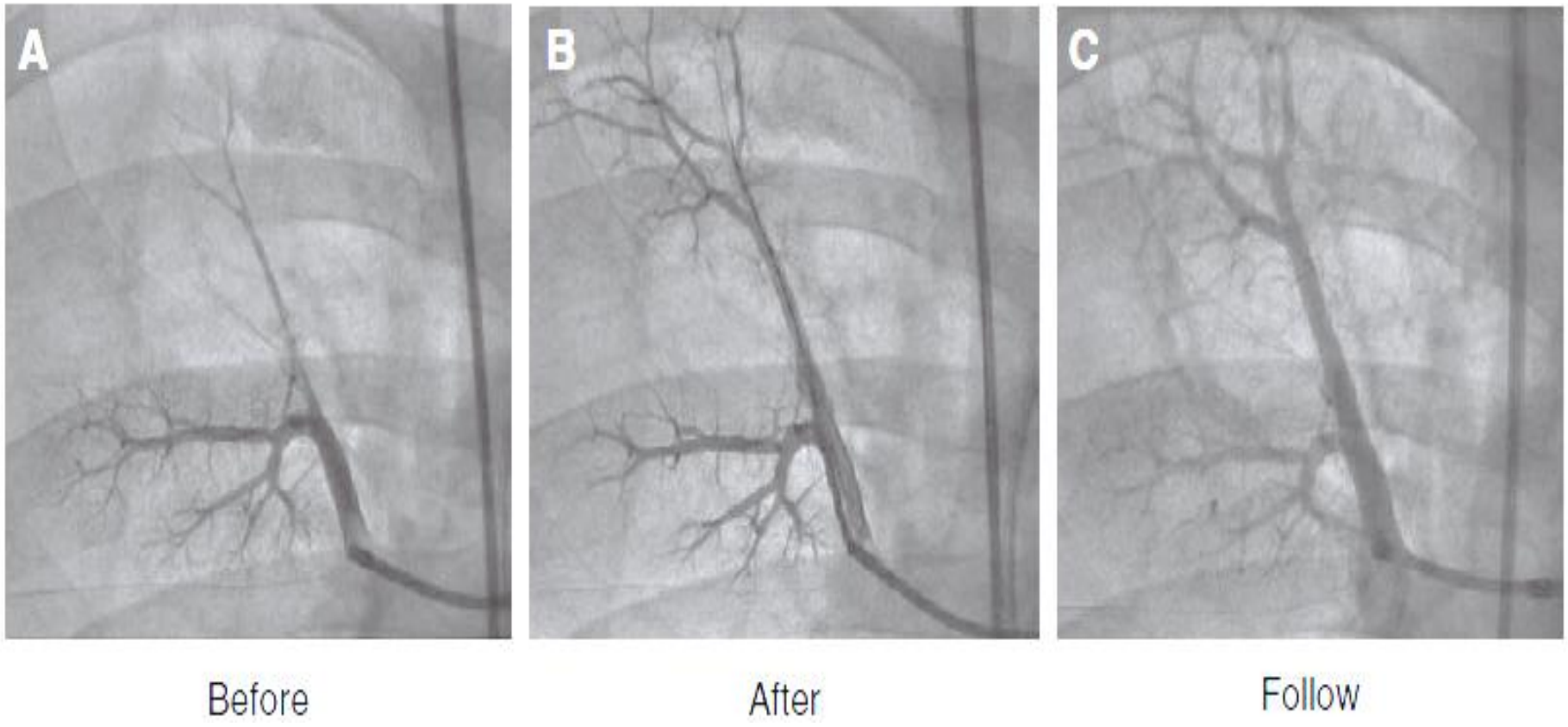
N = 20 pts

Results: After BPA, systolic pulmonary artery pressures (PAP) decreased from 89.1±16.4 to 51.2±12.2 mmHg (P<0.005), mean PAP decreased from 49.8±8.6 to 29.1±6.7 mmHg (P<0.005). Decrease of systolic PAP was independent of the patients' preoperative factors, such as duration of disease, age and medications. None of the preoperative hemodynamic parameters were also correlated with decrease of systolic PAP.

Authors: *Hiroki Mizoguchi, Hiromi Matsubara, Mitsuru Munemasa, National Hospital Organization, Okayama Medical center, Okayama, Japan*

Refined Balloon Pulmonary Angioplasty for Inoperable Patients with Chronic Thromboembolic Pulmonary Hypertension

Hiroki Mizoguchi, MD; Aiko Ogawa, MD, PhD; Mitsuru Munemasa, MD, PhD; Hiroshi Mikouchi, MD, PhD; Hiroshi Ito, MD, PhD; Hiromi Matsubara, MD, PhD

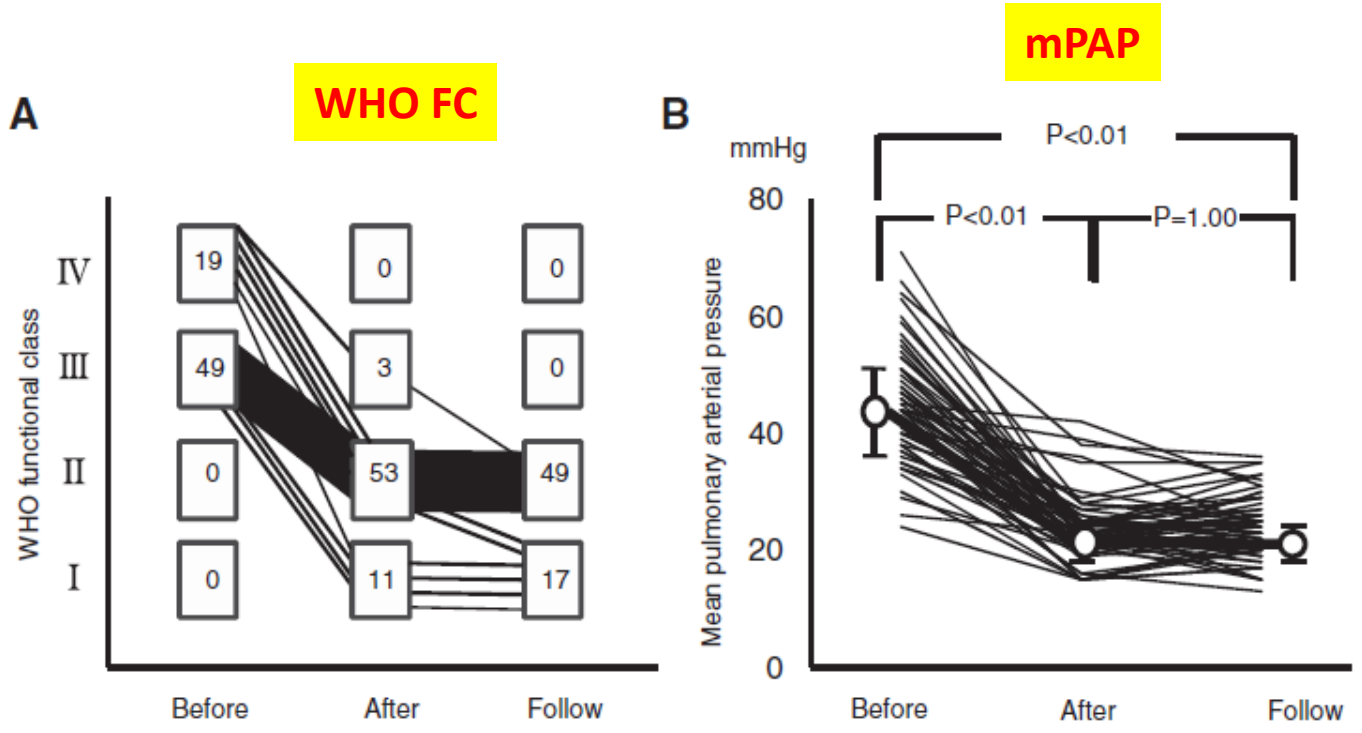


Circulation Cardiovasc. Intervent. 2012,5;748-755

Refined Balloon Pulmonary Angioplasty for Inoperable Patients with Chronic Thromboembolic Pulmonary Hypertension

Hiroki Mizoguchi, MD; Aiko Ogawa, MD, PhD; Mitsuru Munemasa, MD, PhD; Hiroshi Mikouchi, MD, PhD; Hiroshi Ito, MD, PhD; Hiromi Matsubara, MD, PhD

- N=68, CTEPH not suitable for surgery
- 255 BPA interventions, no peri-procedural deaths
- 1 death within 28 days - RVF

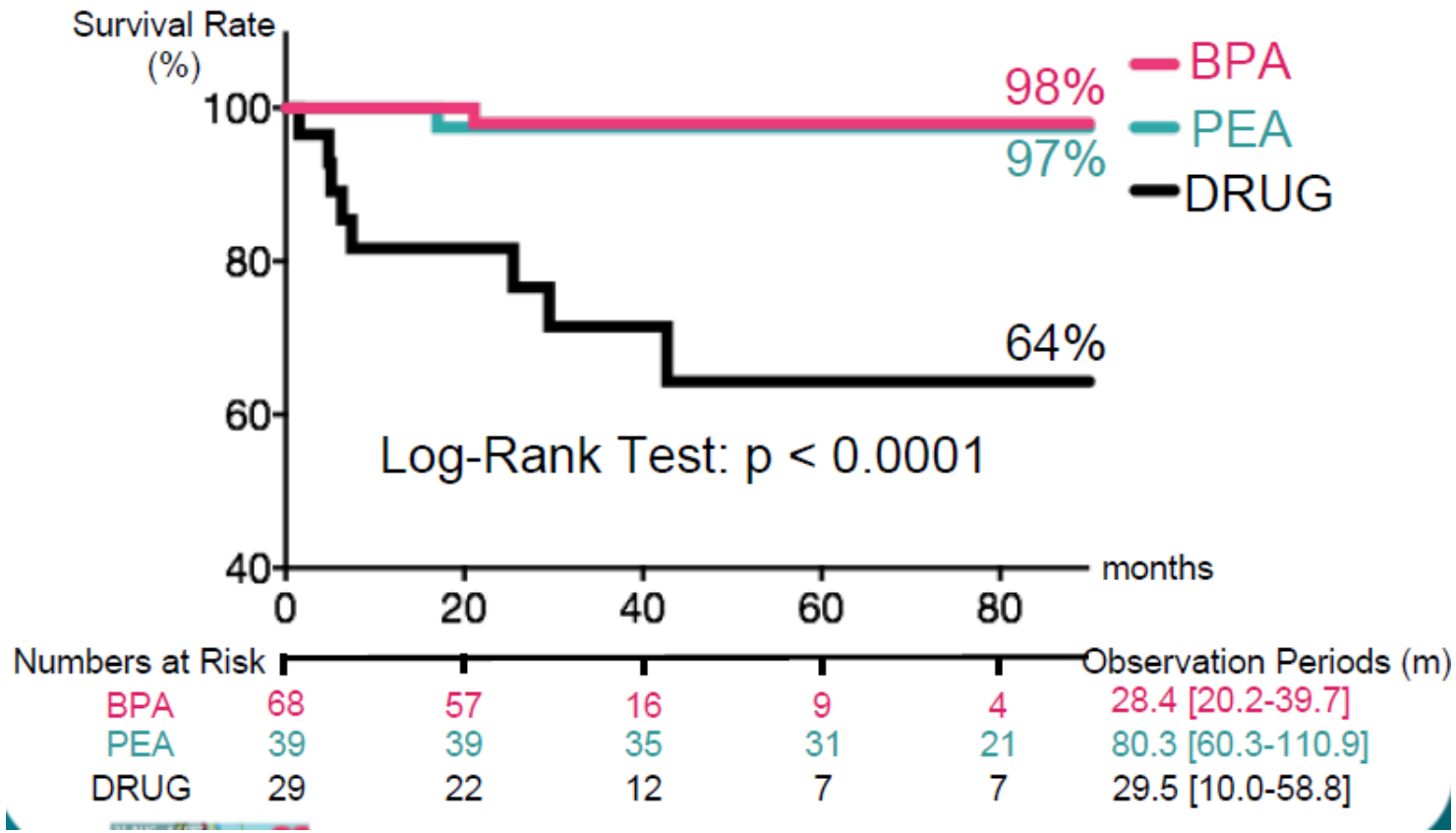


Circulation Cardiovasc. Intervent. 2012,5;748-755

Influence of Balloon Pulmonary Angioplasty on Prognosis in Patients with Chronic Thrombo-Embolic Pulmonary Hypertension

Kyorin University School of Medicine
Second Department of Internal Medicine

Inami T, Kataoka M, Shimura N, Yanagisawa R
Ishiguro H, Kohshoh H, Satoh T, Yoshino H

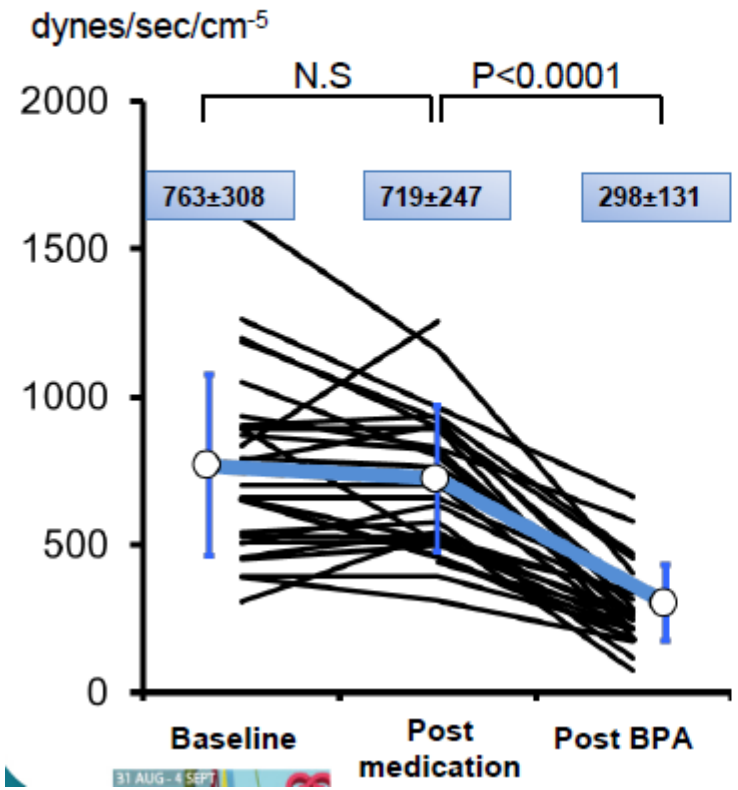


ESC 2013

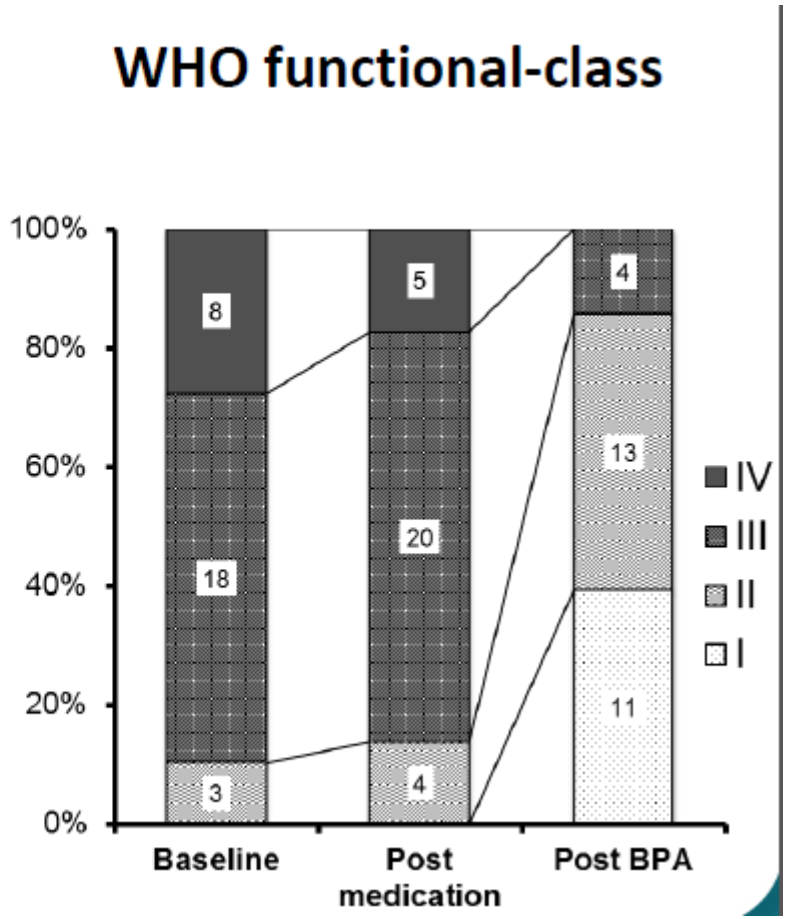
Efficacy and Safety of Balloon Pulmonary Angioplasty for Peripheral Type Chronic Thromboembolic Pulmonary Hypertension.

Yu Taniguchi, Toshiro Shinke, Hiroto Kinutani, Hiromasa Otake
Noriaki Emoto, Ken-ichi Hirata

Pulmonary Vascular Resistance



WHO functional-class



Heart

Balloon pulmonary angioplasty in patients with inoperable chronic thromboembolic pulmonary hypertension

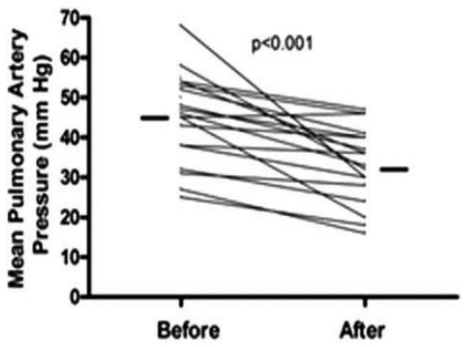
Arne K Andreassen, Asgrimur Ragnarsson, Einar Gude, et al.

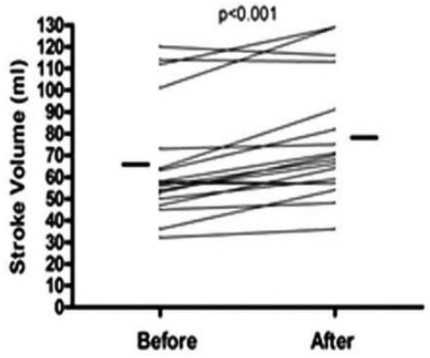
n = 20

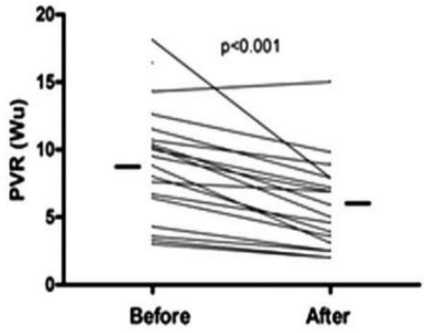
73 sessions

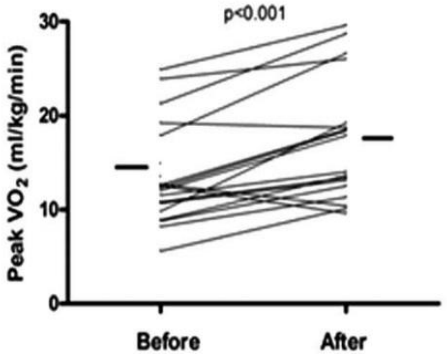
3,7 sessions/pt

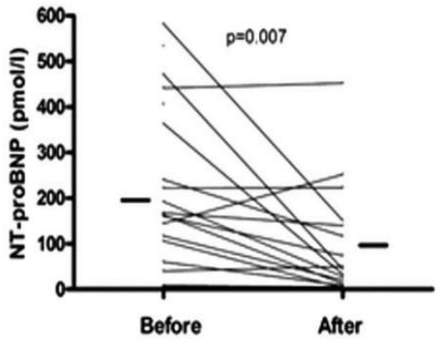
18,6 BPA`s/pat.





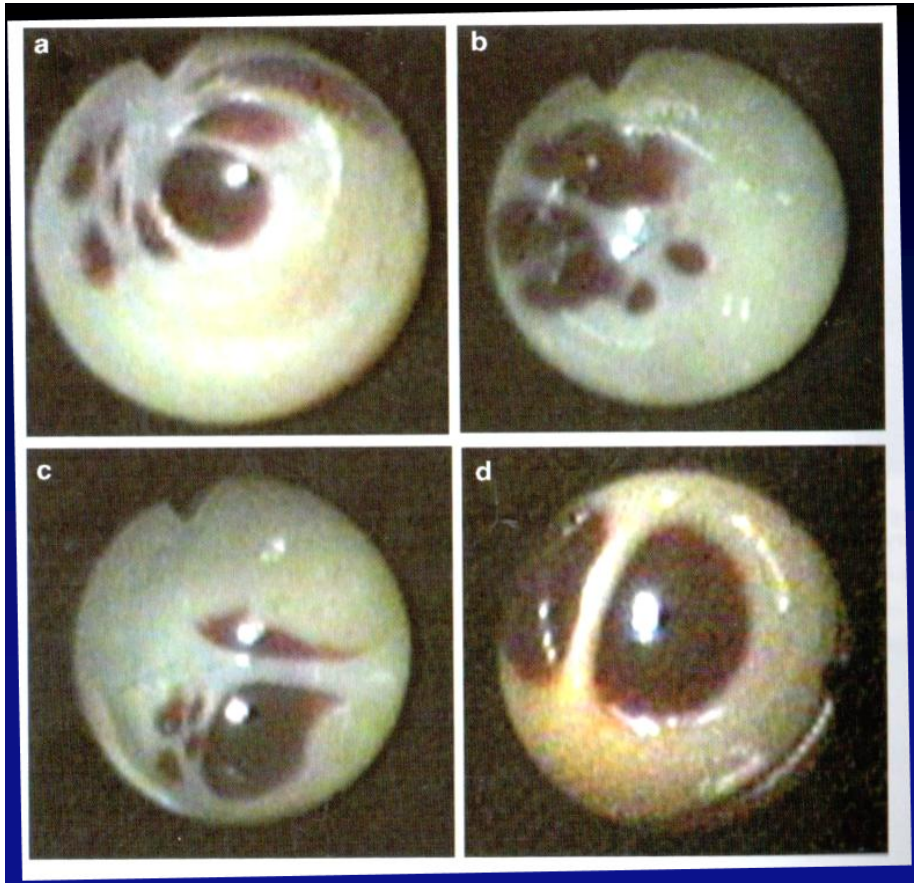






Andreassen AK, Ragnarsson A, Gude E, et al. Heart 2013;99:1415– 1420

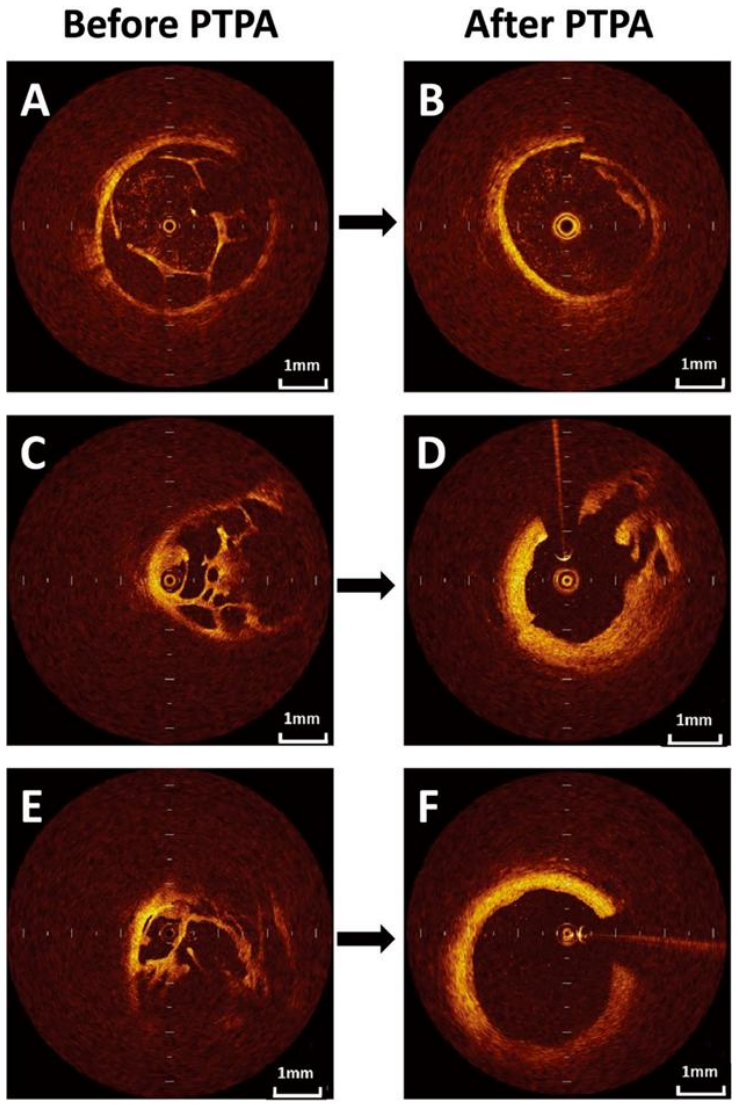
ANGIOSCOPY



www.springerimages.com

Images obtained during angioscopy in chronic thromboembolic pulmonary hypertension (**a**–**c**) and normal bifurcation of pulmonary artery (**d**).

OCT



Sugimura K. 2011

Efficacy and Safety of Balloon Pulmonary Angioplasty for Peripheral Type Chronic Thromboembolic Pulmonary Hypertension.

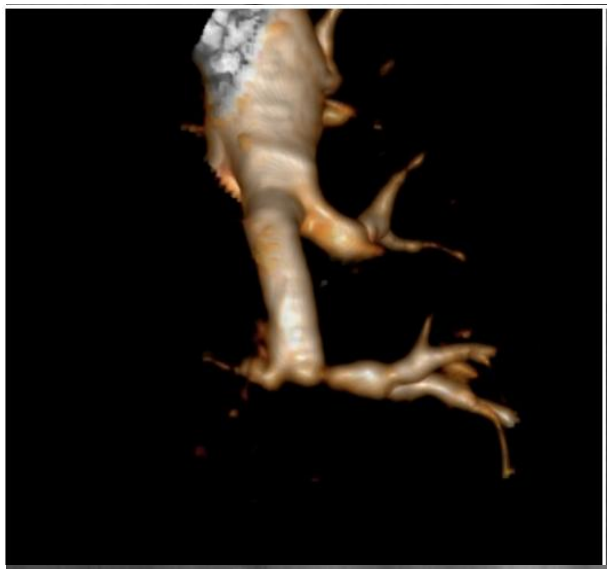
Yu Taniguchi, Toshiro Shinke, Hiroto Kinutani, Hiromasa Otake
Noriaki Emoto, Ken-ichi Hirata

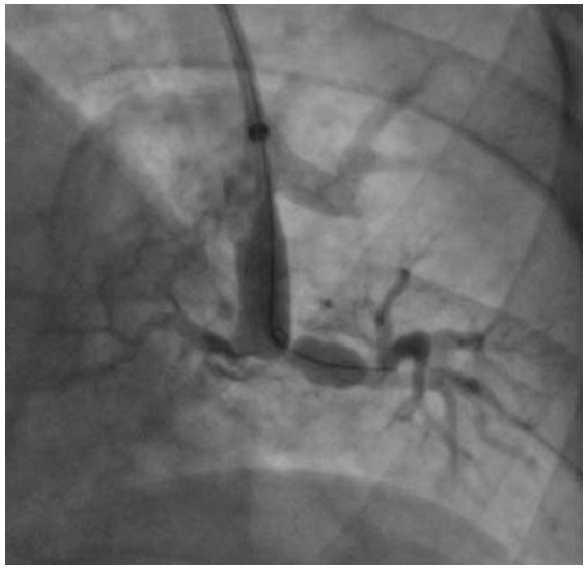
Complications of BPA

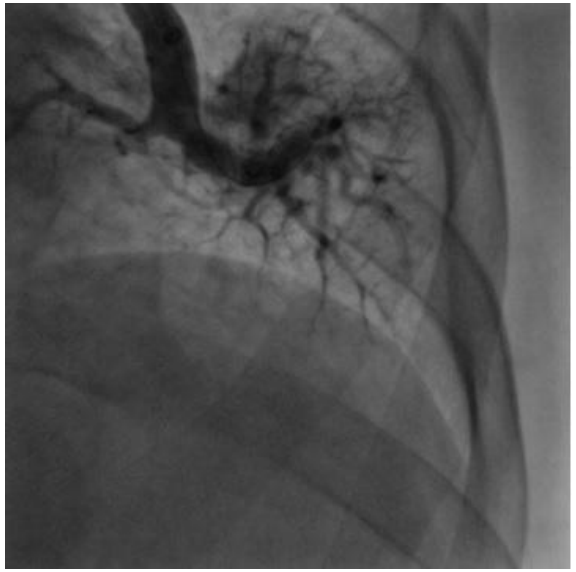
Complications	All sessions (n=79)
Reperfusion pulmonary injury	
・Symptom	
Hemo-sputum	22 (27.8%)
Dyspnea or desaturation	26 (32.9%)
・CT	
hemorrhagic pulmonary edema	12 (15.2%)
mild pulmonary edema only	38 (48.1%)
no sign of pulmonary edema	29 (36.7%)
Wire perforation	4 (5.1%)
Death	1 (1.3%)
Intubation	3 (3.8%)
NIPPV	54 (68.4%)

Our first experience with BPA

- **A 43-year-old female with CTEPH**
- **disqualified from pulmonary endarterectomy.**
- **treated with sildenafil (*off-label*) for one year without improvement.**
- **BPA of two subsegmental arteries was performed**







mPAP 56 mmHg,
CO 6,03 l/min,
PVR 7,96 Wood units

mPAP 47mmHg,
CO 5,99 l/min,
PVR 6,51 Wood units

At 3 months

mPAP 36mmHg,
PVR 4,5 Wood units

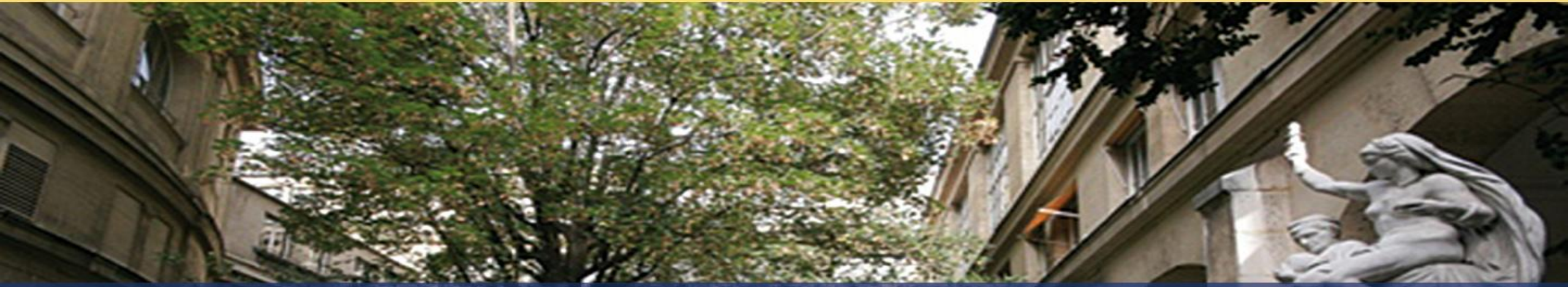
Early own experience

n	Age	Sex	N of sessions	N of segments	Complications
1.	43	F	2	4	-
2.	23	F	1	3	SVT
3.	71	F	1	2	Reperfusion oedema
4.	66	M	1	2	Hemoptysis

(5) (11)

INTERNATIONAL CTEPH CONFERENCE 2014

ICA International CTEPH Association



Summary of Abstracts

A Torbicki and M Delcroix

June 2-3, 2014 - Institut des Cordeliers - Paris, France

Abstracts submitted

	2011 Cambridge	2014 Paris
Participants	250	400
Abstracts	43	77
Topics		
Pathophysiology/biology	8	**21**
Epidemiology and RF	2	4
Imaging/hemodynamics	7	7
PEA	20	22
CTEPH outcome	4	2
BPA	2	**14**
Medical treatment	0	**7**

Restenosis post BPA?

38/103 pts

baseline – final - 6 – **12 months RHC**

PAP mmHg:

42.7 - 27.3 - 21.8 - **19.9**

PVR WU:

10.2 - 5.0 - 3.1 - **2.9**

No signs of restenosis at 12 month…

Takumi Inami , Tokyo/JAP

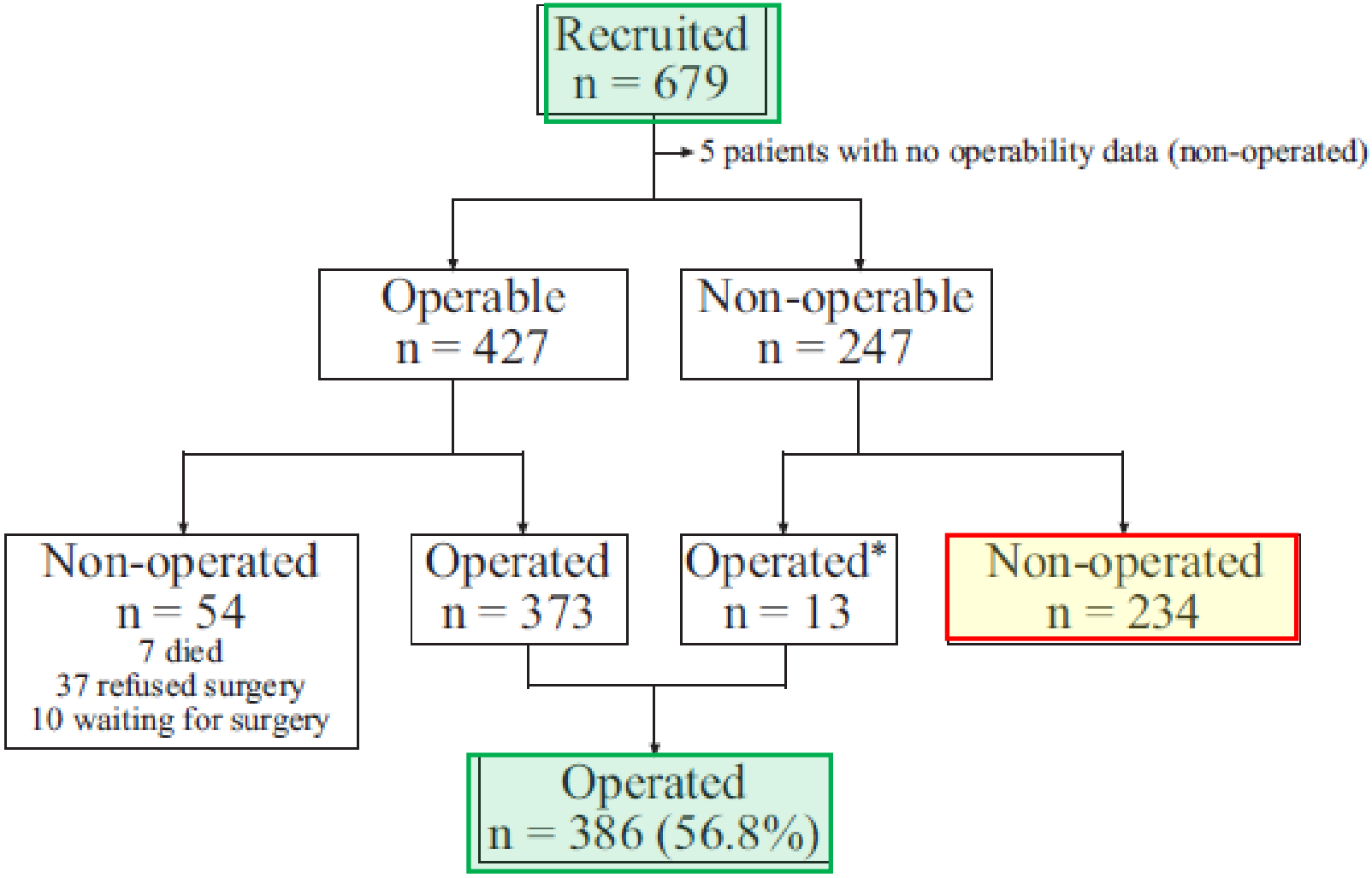
Guided BPA (PTPA?)

Safety and effectiveness of pressure-wire-guided BPA
145/350 sessions

- 0% vs 6.9% critical reperfusion edema
- same hemodynamic improvements
- fewer sessions/lesions treated necessary

Takumi Inami , Tokyo/JAP

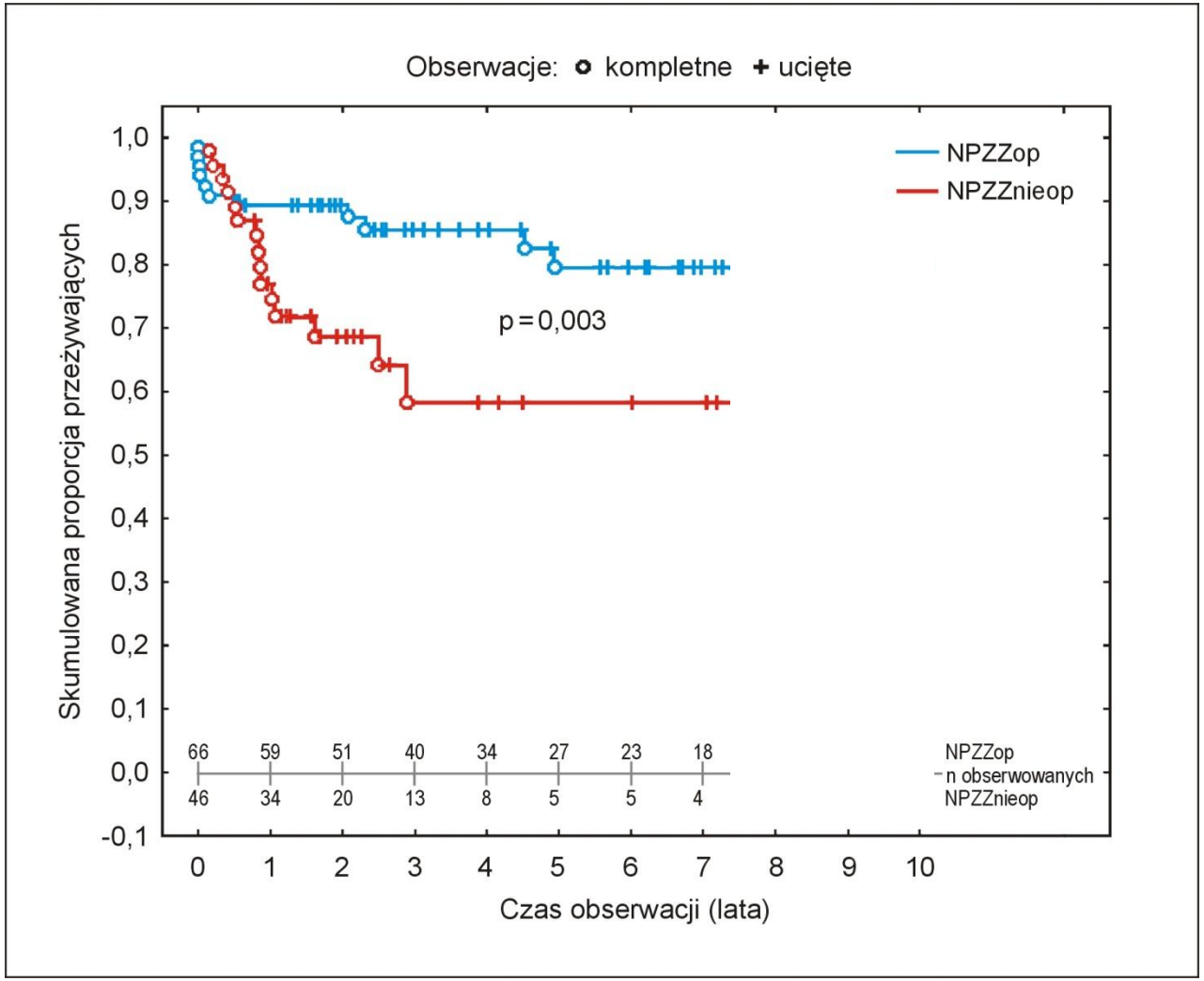
Chronic Thromboembolic Pulmonary Hypertension (CTEPH) : Results From an International Prospective Registry

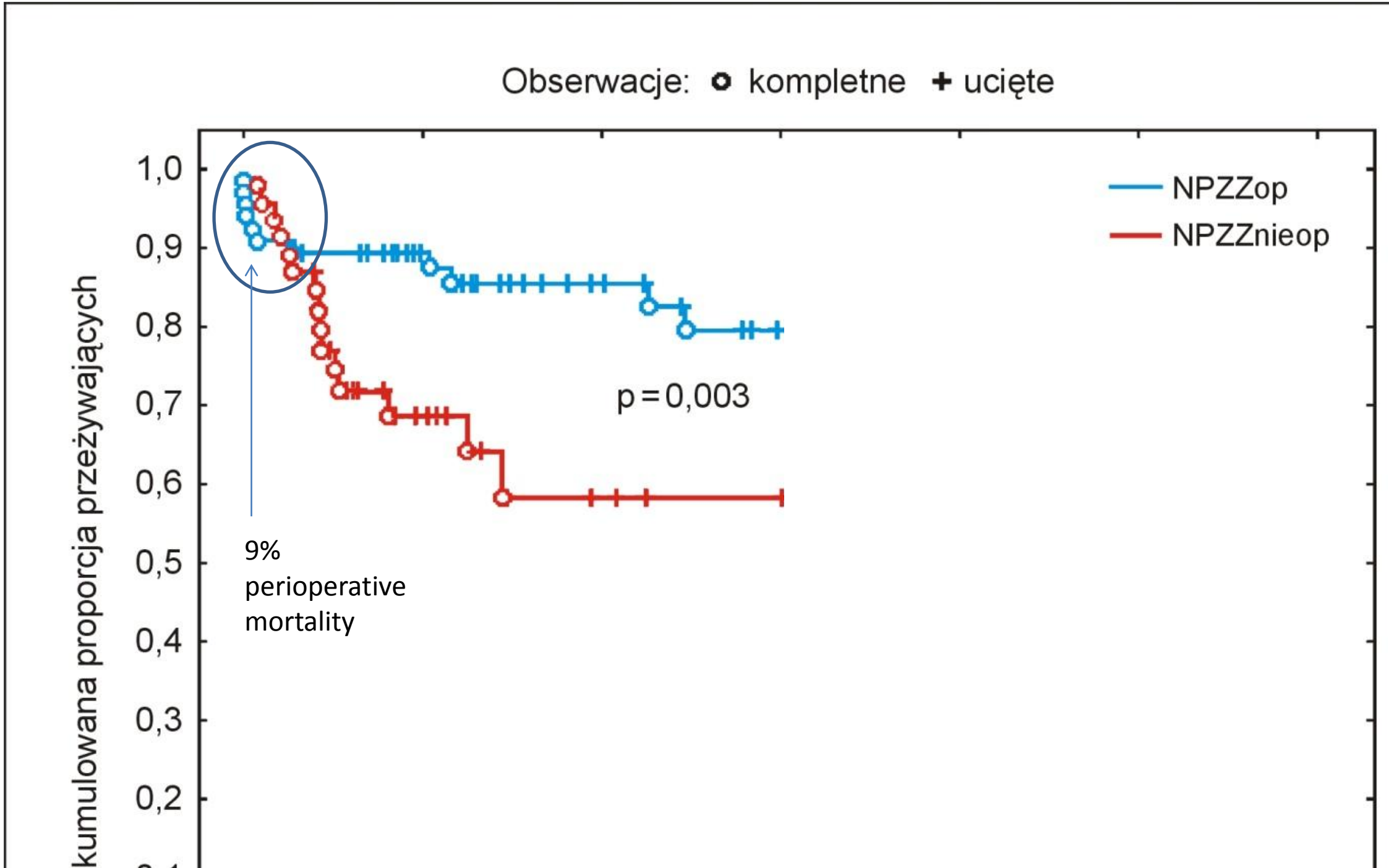


Circulation published online October 3, 2011

CTEPH survival at our Centre

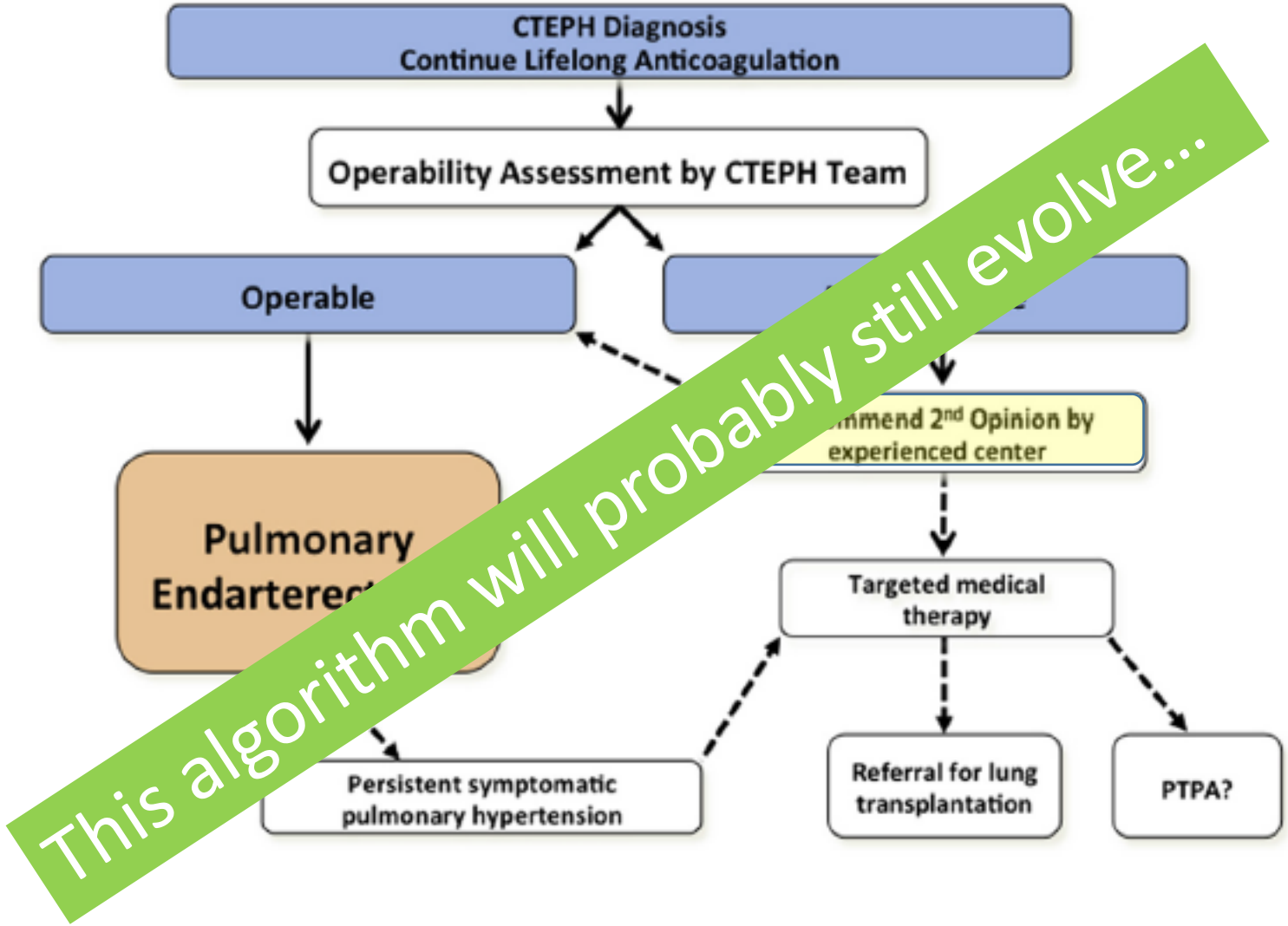
(PEA vs medical Tx)




Obserwacje: kompletne ucięte
1,0
0,9
0,8
0,7
0,6
0,5
0,4
0,3
0,2
kumulowana proporcja przeżywających
NPZZop
NPZZnieop
p = 0,003
9%
perioperative
mortality

Therapeutic opportunities in CTEPH

- PEA = …
- BPA = Balloon pulmonary angioplasty
- MTx = Medical targeted Tx for CTEPH
- LTx = …



JACC Vol. 62, No. 25, Suppl D, 2013
December 24, 2013:D92–9

- CRP n <0.5

2013-02-15
0.28 [mg/dl]

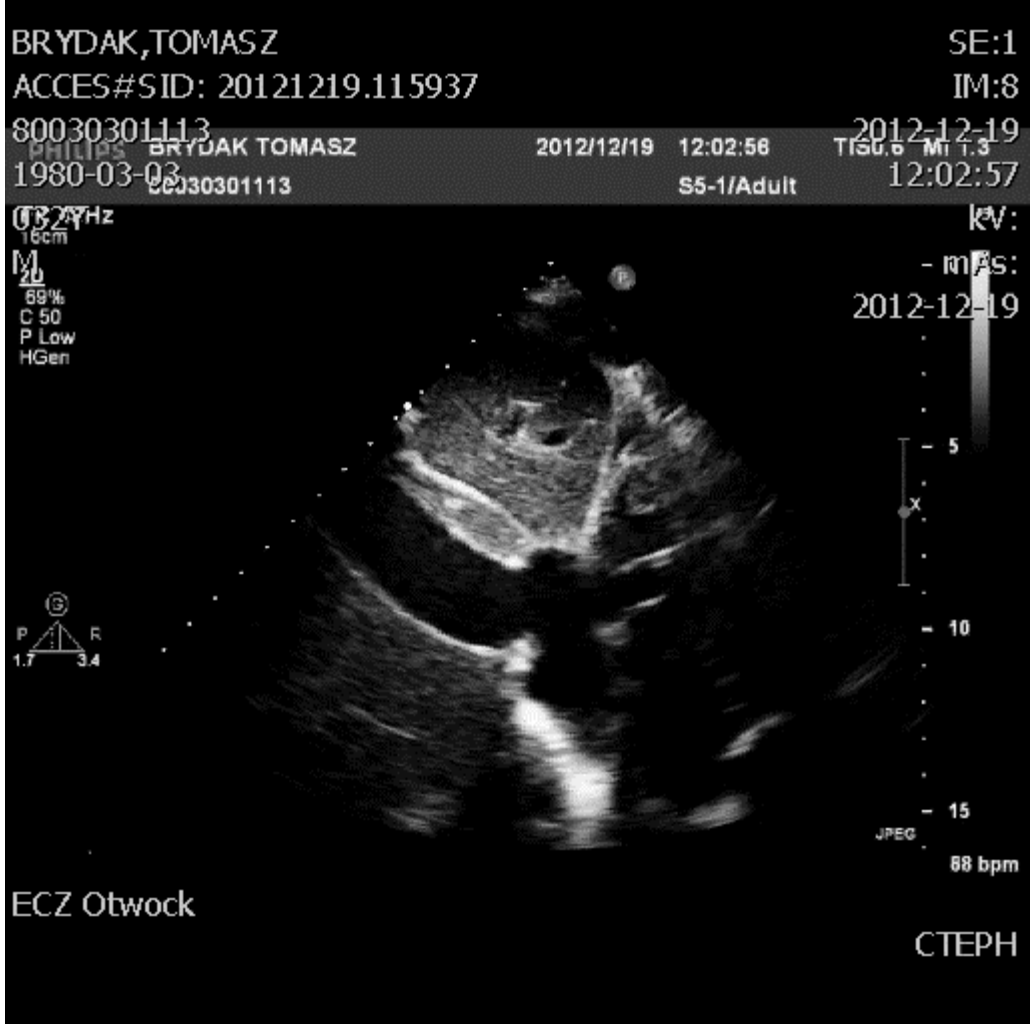
WBC 6.6 [x10^3/µl] Norma: 4,0 - 10,0, NEU% 37.8 [%] Norma: 34,0 - 67,9,

Nazwa badania: **NT-PROBNP (N-TERMINALNY PROPEPT. NATRIURET. T.B)**

2013-02-15
2891.0 [pg/ml]

Thrombofilia testing

Badanie	Wynik	Norma	Opis
Czas protrombinowy data wyk: 2013-02-26 10:53, os. 1)	**12,40 s**	8,7-13,0 s	N
INR data wyk: 2013-02-26 10:53, os. 1)	**1,18**	-	
Wskaźnik protrombinowy data wyk: 2013-02-26 10:53, os. 1)	**84,44 %**	80-120%	N
Czas trombinowy data wyk: 2013-02-26 09:50, os. 2)	**16,9 s**	<21 s	N
Fibrynogen (met. Claussa) data wyk: 2013-02-26 09:50, os. 2)	**4,5 g/l**	2-5 g/l	N
APTT - czas częściowej tromboplastyny po aktywacji data wyk: 2013-02-26 10:53, os. 1)	**27 s**	25-33 s	N
Antykoagulant toczniowy (w oparciu o dRVVT) data wyk: 2013-02-26 10:53, os. 3)	**test ujemny**		N
Czynnik VIII data wyk: 2013-02-26 09:50, os. 1)	**137 %**	50-150 %	N
Aktywność antytrombiny (metodą chromogenną, anty IIa) data wyk: 2013-02-26 10:53, os. 3)	**107 %**	79-112 %	N
Aktywność białka C (met. chromogenną) data wyk: 2013-02-26 10:53, os. 3)	**114 %**	70-140 %	N
Antygen wolnego białka S (met. ELISA) data wyk: 2013-02-26 10:53, os. 3)	**115 %**	50-150 %	N
Test APCR data wyk: 2013-02-26 10:53, os. 3)	**0,92**	0,7-1,2	N
Miano przeciwciał antykardiolipinowych w klasie IgG data wyk: 2013-02-26 10:53, os. 1)	**2,3**	<10	N
Miano przeciwciał antykardiolipinowych w klasie IgM data wyk: 2013-02-26 10:53, os. 1)	**0,6**	<7	N
Miano przeciwciał przeciw Beta-2-Glikoproteinie I w klasie IgG data wyk: 2013-02-26 10:53, os. 1)	**2,1**	<5	N
Miano przeciwciał przeciw Beta-2-Glikoproteinie I w klasie IgM data wyk: 2013-02-26 10:53, os. 1)	**0,6**	<5	N


BRYDAK,TOMASZ
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2012-12-19
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1980-03-03 80030301113 S5-1/Adult 12:02:57
kV:
- mAs:
2012-12-19
5
10
15
JPEG
68 bpm
ECZ Otwock
CTEPH


PHILIPS BRYDAK TOMASZ 2012/12/19 12:18:16 TIS0.6 MI 1.4
1980-03-03 80030301113 S5-1/Adult 12:18:18
kV:
- mAs:
2012-12-19
5
10
15
JPEG
66 bpm
ECZ Otwock
CTEPH