Pulmonary Arterial Hypertension and Congenital Heart Disease: Role of Interventional Cardiology

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TCTAP 2014

Diagnosis of Pulmonary Hypertension in the Congenital Heart Disease Adult Population

Impact on Outcomes

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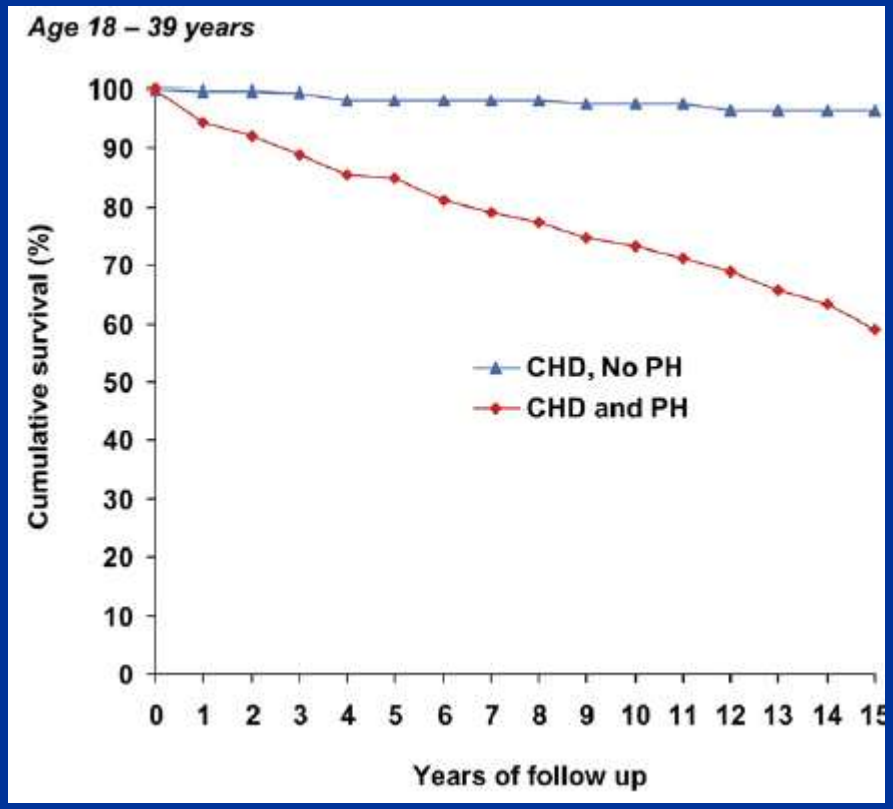
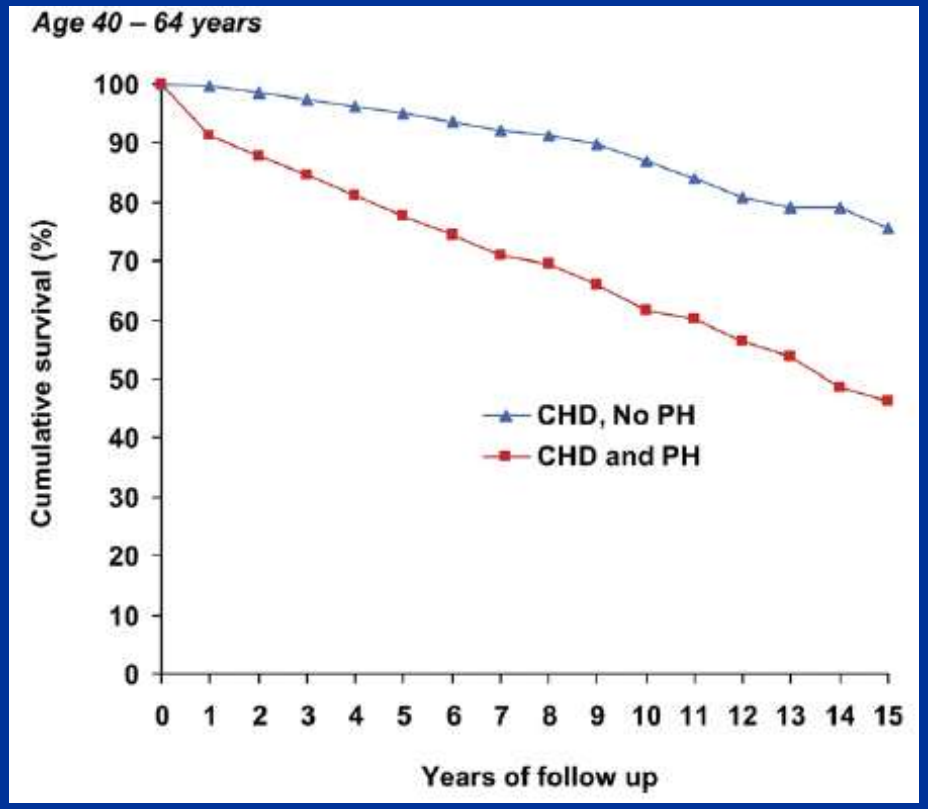
Louise Pilote, MD, MPH, PhD,‡§ Giuseppe Martucci, MD,* Ariane J. Marelli, MD, MPH*





J Am Coll Cardiol 2011;58:538–46

Various Conditions of ACHD with PH

- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

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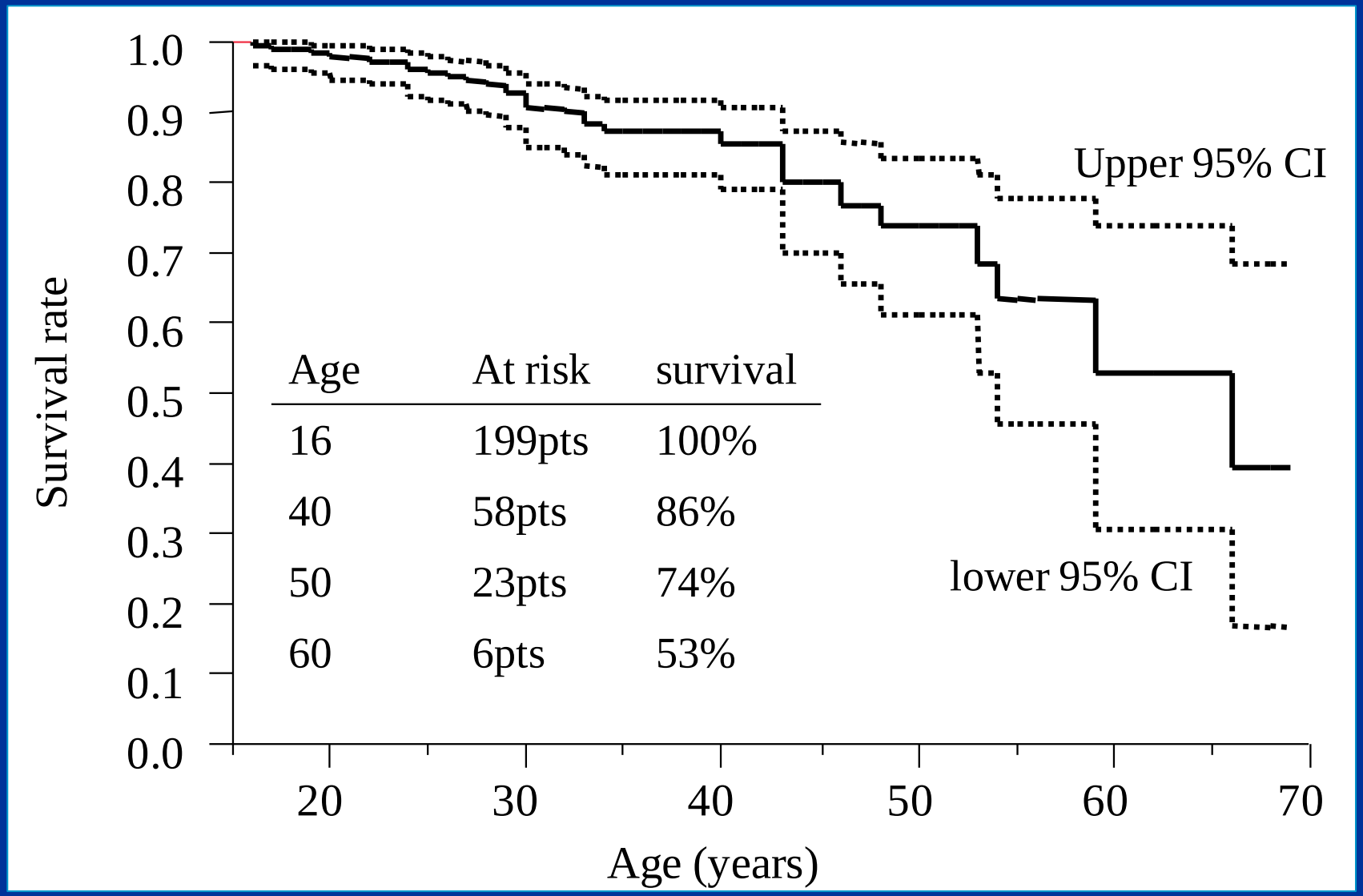
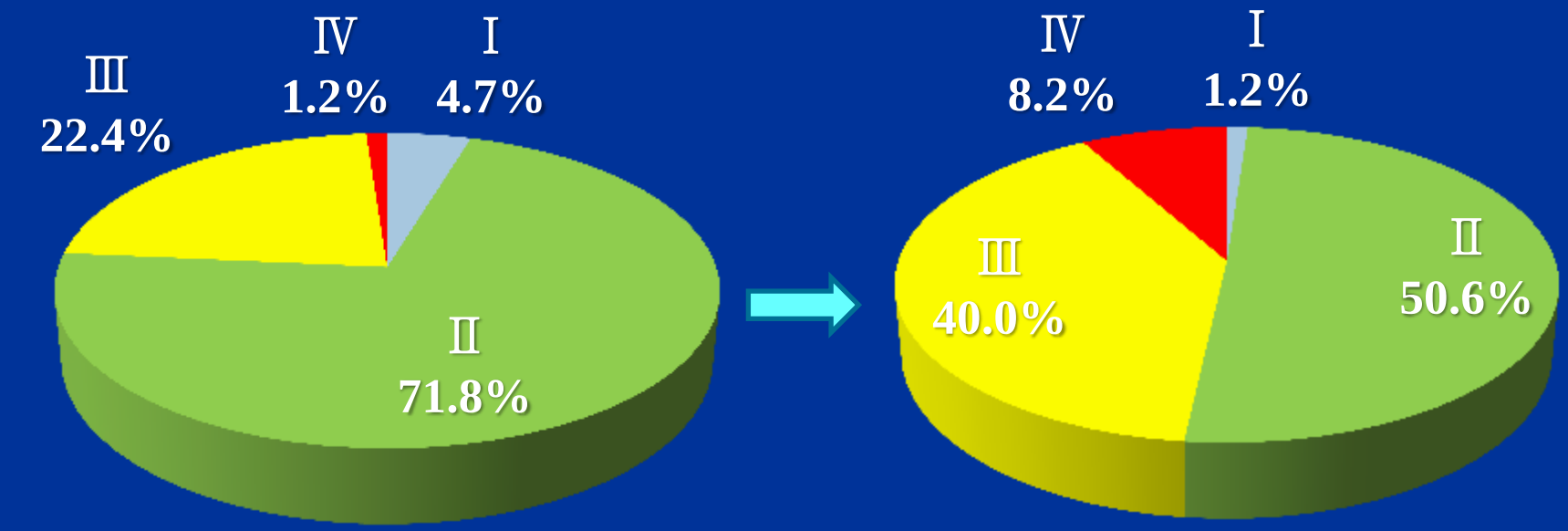
Natural course of Eisenmenger syndrome



Transition of NYHA functional class in Eisenmenger syndrome



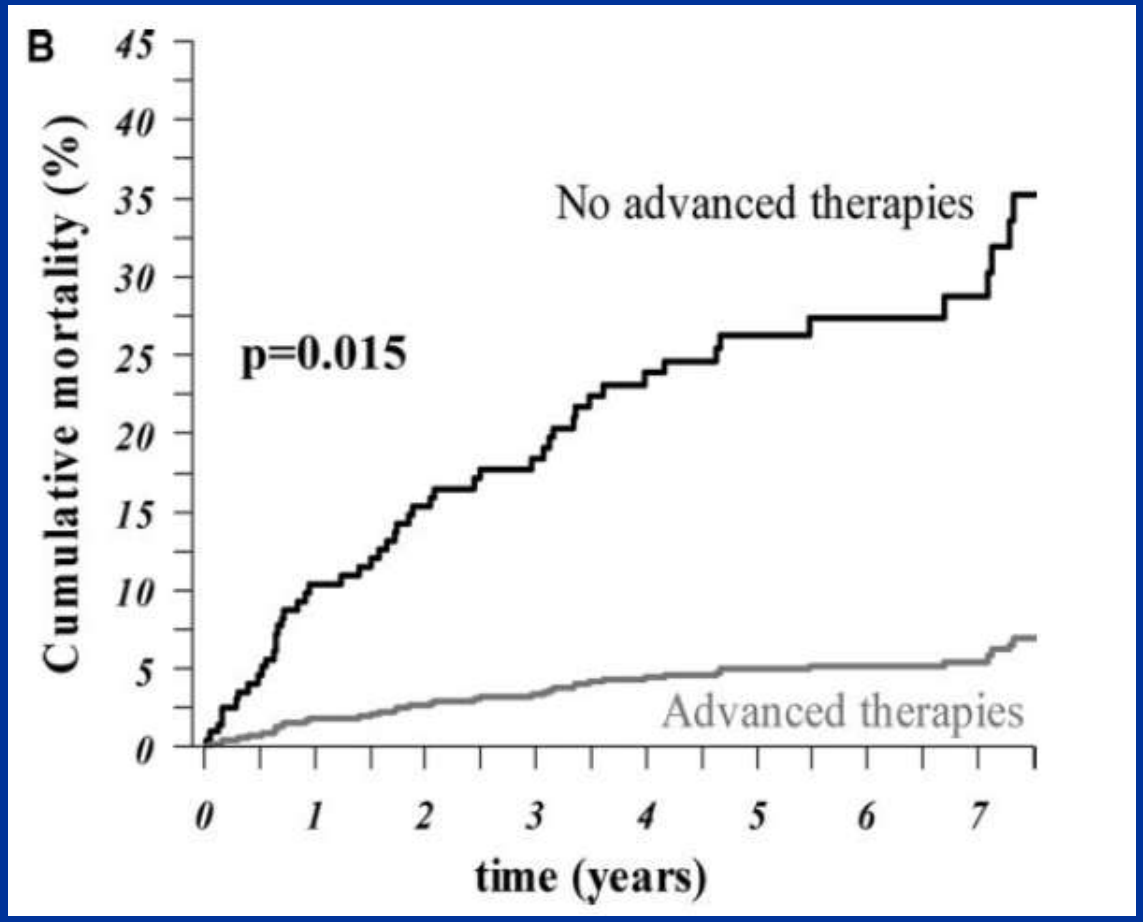
Age at enrollment (30±12 years)

Age at the latest study (38±11 years)

Inohara T, Niwa K, et al. J Cardiol 2014

Improved Survival Among Patients With Eisenmenger Syndrome Receiving Advanced Therapy for Pulmonary Arterial Hypertension

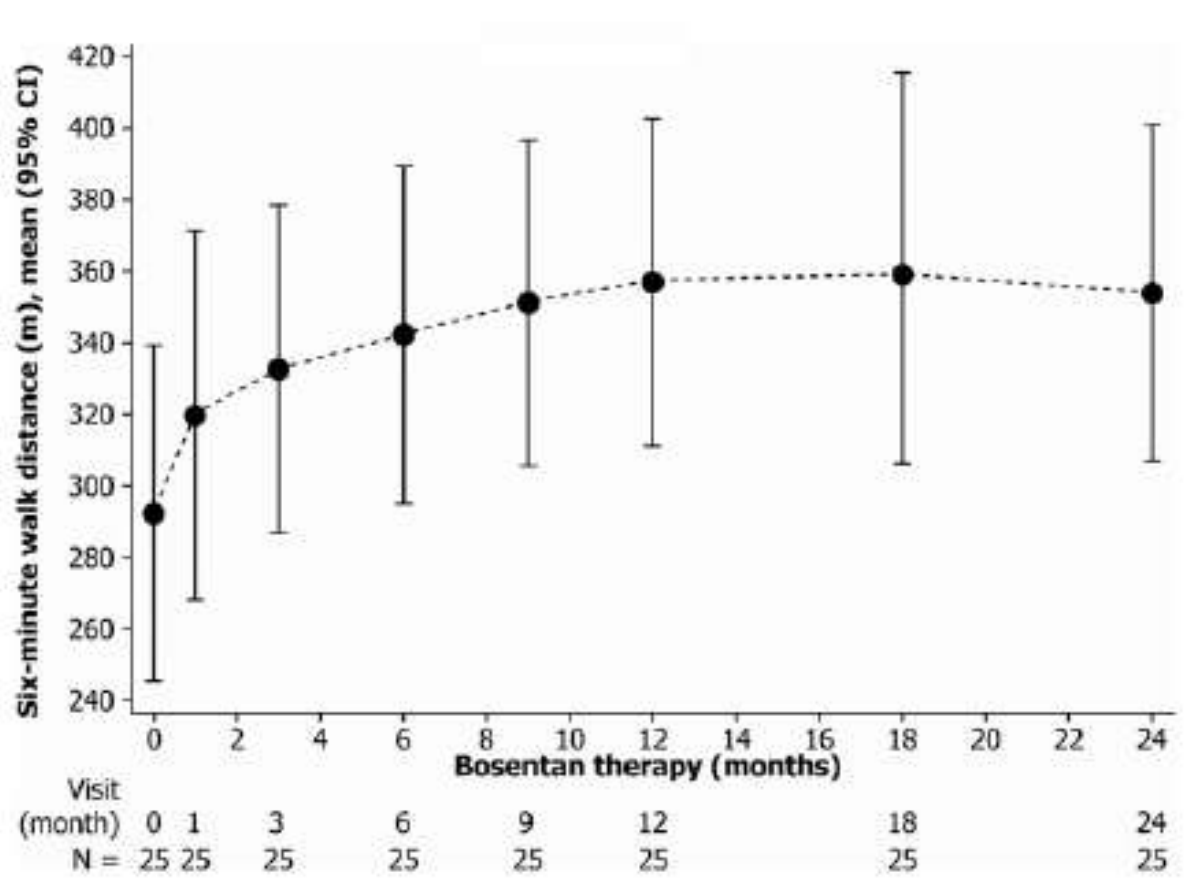
Konstantinos Dimopoulos, MD, MSc, PhD, FESC*; Ryo Inuzuka, MD*; Sara Goletto, MD; Georgios Giannakoulas, MD, PhD, FESC; Lorna Swan, MD, MRCP; Stephen J. Wort, BA, MBBS, MRCP, PhD; Michael A. Gatzoulis, MD, PhD, FESC



Circulation 2010;121:20-25.

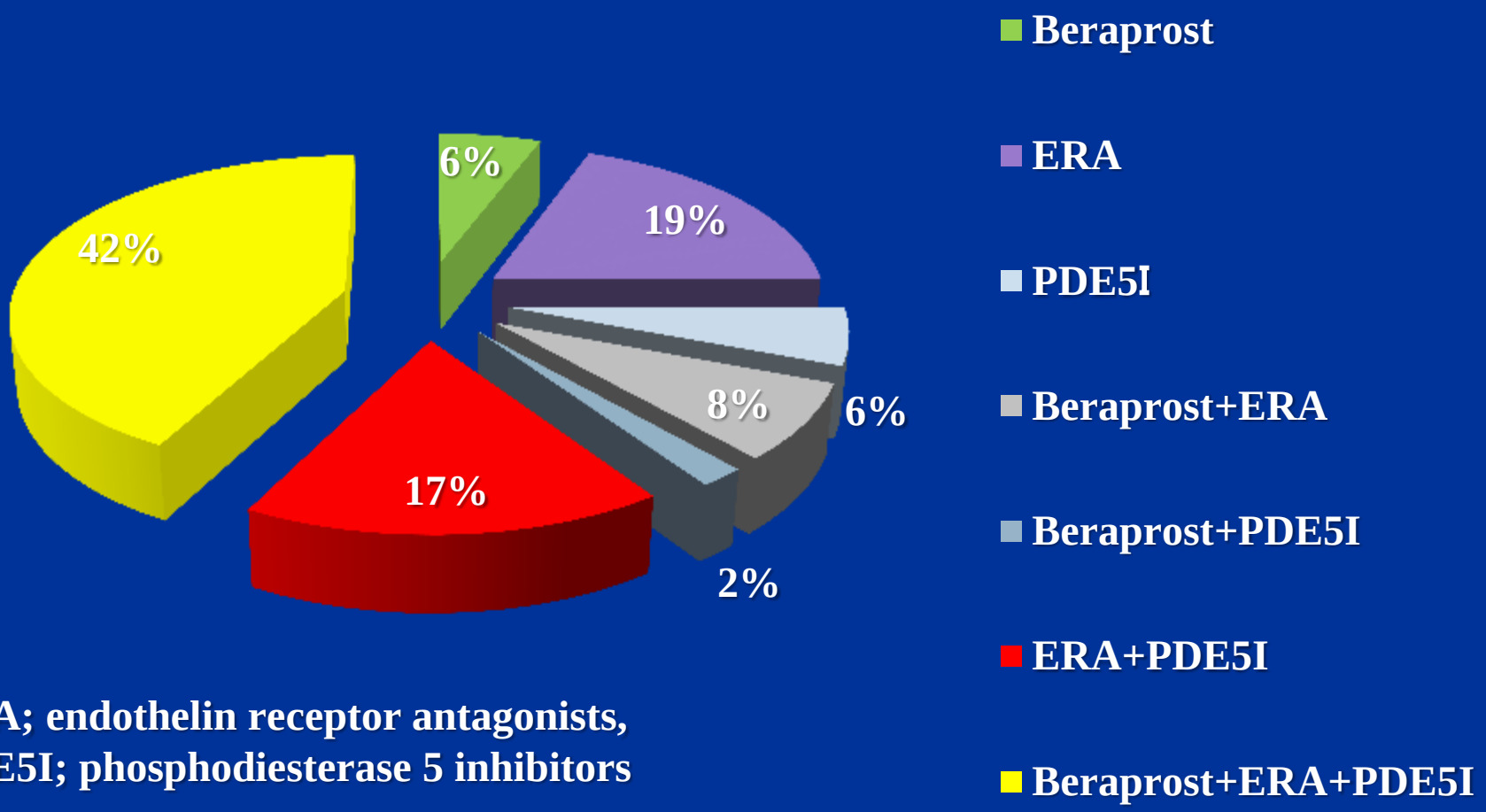
Efficacy and Safety of *Bosentan* for Pulmonary Arterial Hypertension in Adults With Congenital Heart Disease

Oliver Monfredi, MBChB, MRCP[a], Linda Griffiths, RGN, RSCN[b], Bernard Clarke, MD[a,b], and Vaikom S. Mahadevan, MD[a,b,*]



Am J Cardiol 2011

Type of Medication of Disease Targeting Therapy



ERA; endothelin receptor antagonists,
PDE5I; phosphodiesterase 5 inhibitors

Inohara T, Niwa K, et al. J Cardiol 2014

Various Conditions of ACHD with PH

- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

52 years-old female

Atrial septal defect was diagnosed when she was 20 years-old. Recently, she became aware of dyspnea on exertion.

NYHA class Ⅱ

Height 149 cm, Body weight 56 kg

No medication

Transthortahic echocardiogram (initial examination)

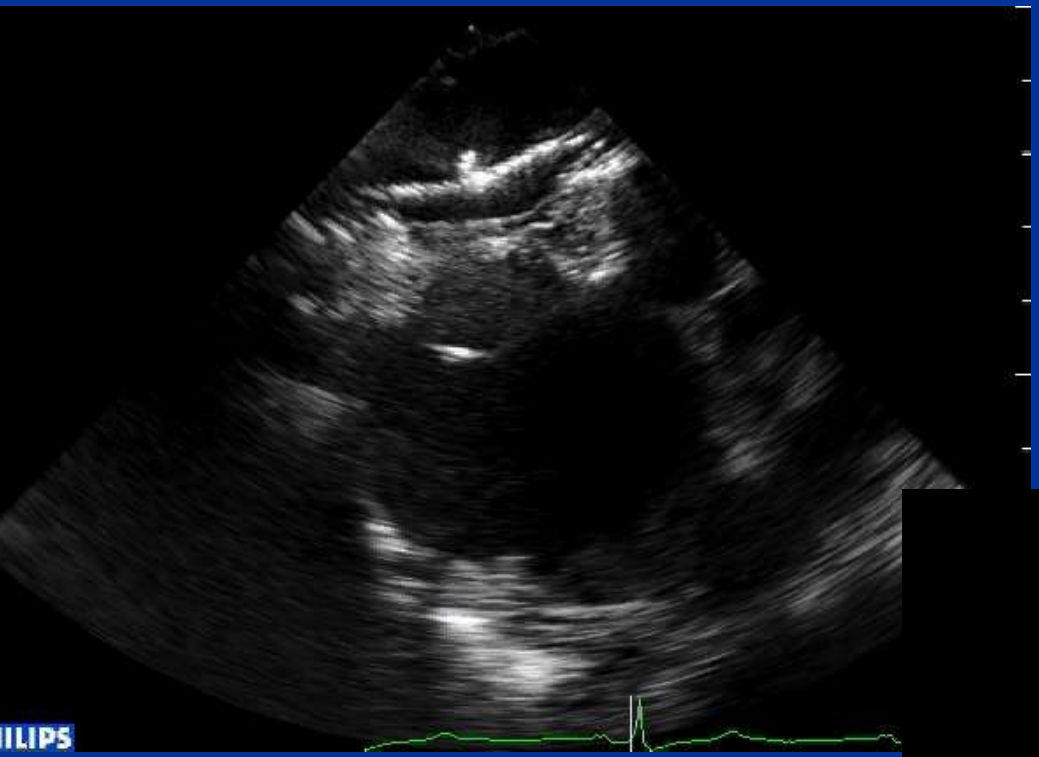
TR mild
TRPG=95mmHg
PR mild
PREDP=15mmHg

IVC 13/5 mm

Preserved LV contraction
LVDd/s 36/22 mm
EF=70% FS=39%
e’ 3 cm, E/e’ 15

ASD (secundum) L ⇒R shunt
Estimated Qp/Qs=2.6, maximal defect dimater 28 mm

0 degree



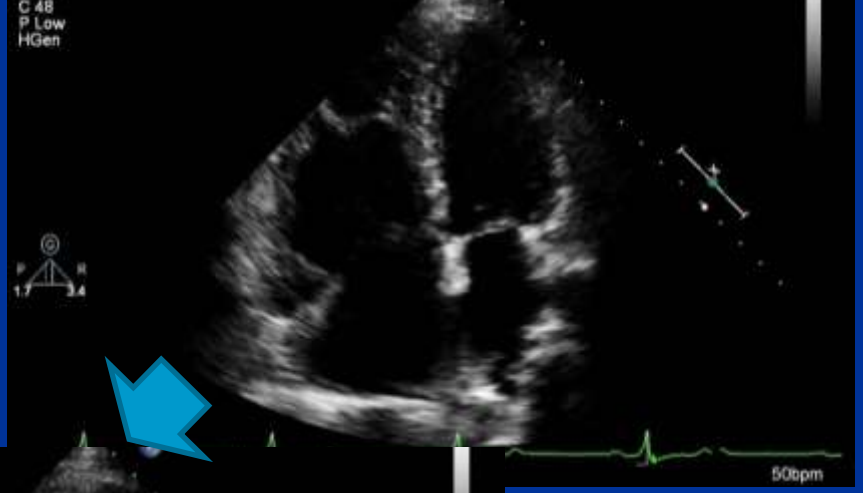
Amplatzer Septal Occluder
30 mm

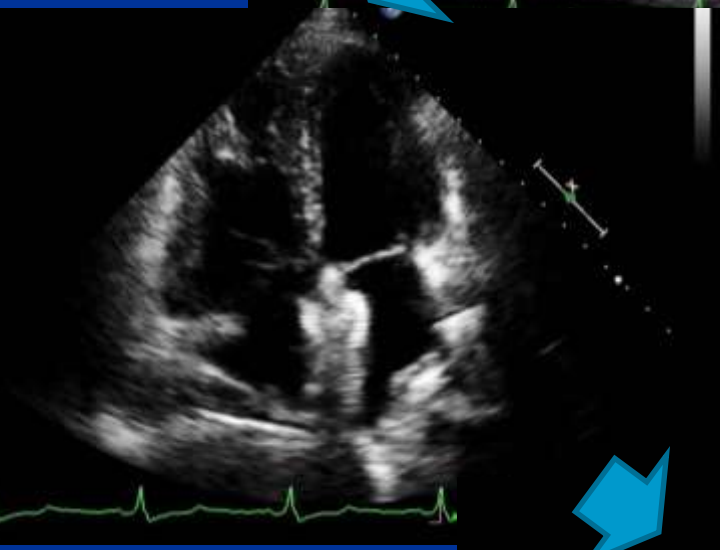
90 degree

Mean PAP
45 → 33 mmHG

Clinical course

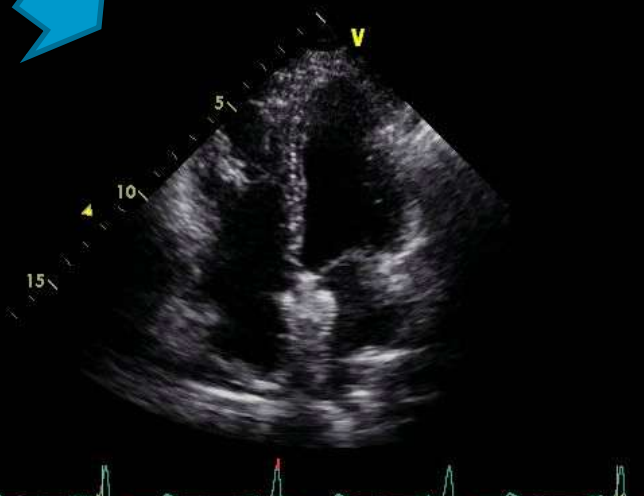
after diuretics treatment
TRPG=60mmHg

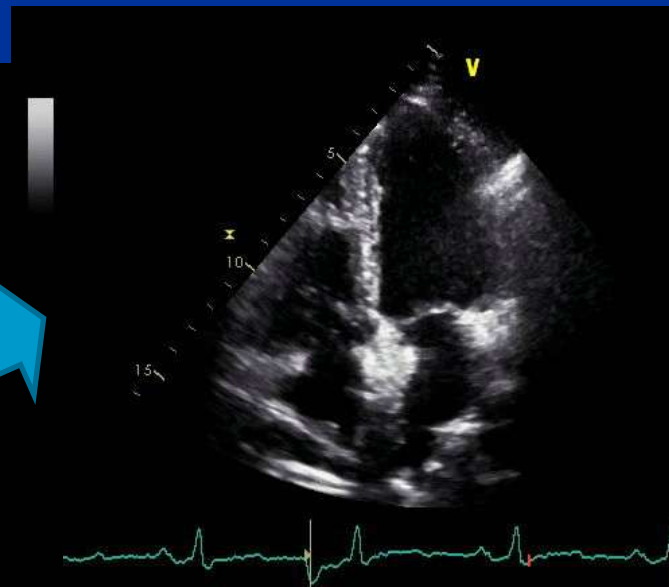




after ASD closure
TRPG=54mmHg

1 year after
TRPG=39mmHg



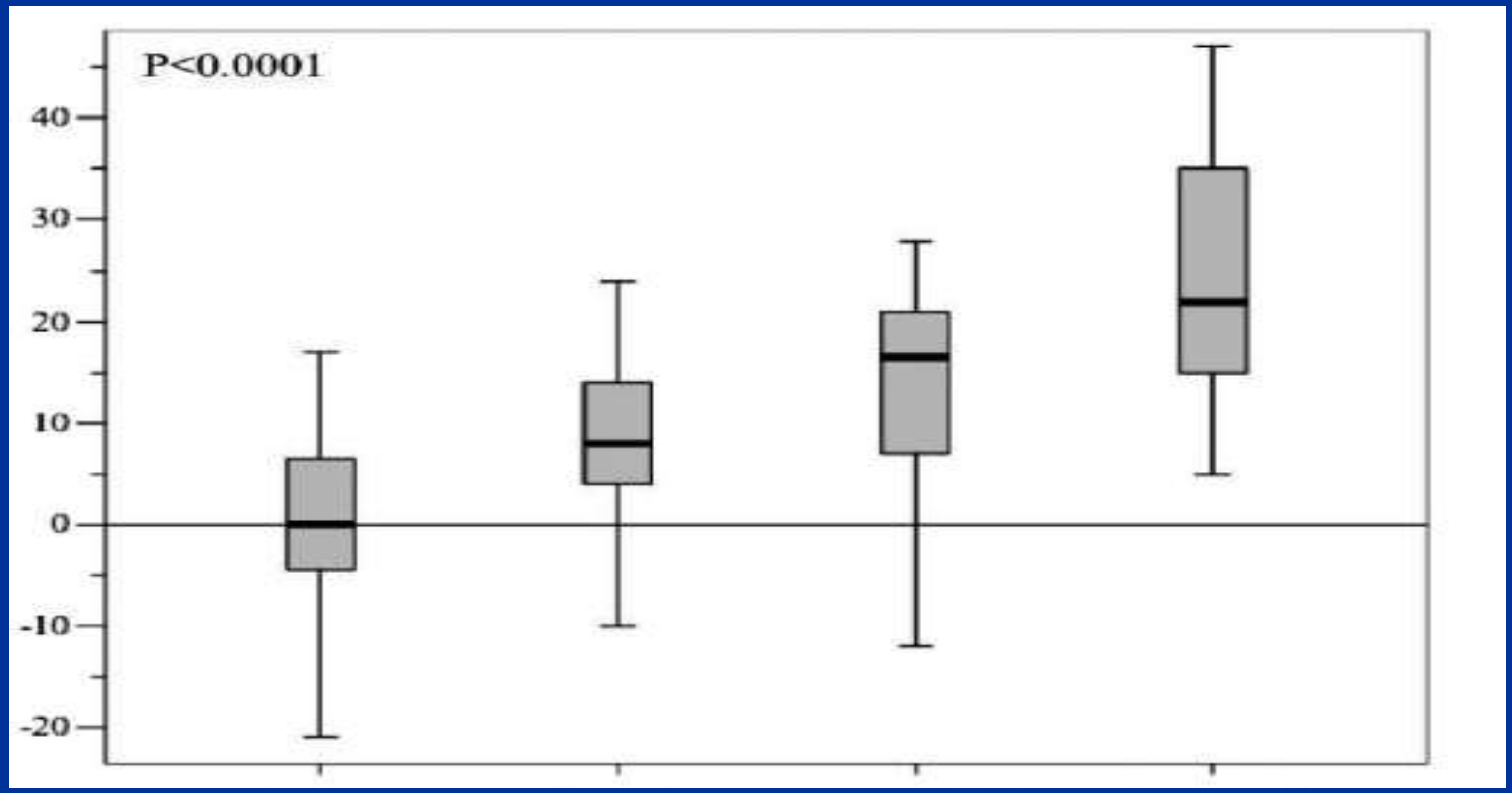


3 mo. after
TRPG=45mmHg

Pulmonary Arterial Hypertension in Patients With Transcatheter Closure of Secundum Atrial Septal Defects

A Longitudinal Study

Gerald Yong, MBBS; Paul Khairy, MD, PhD; Pierre De Guise, MD; Annie Dore, MD; Francois Marcotte, MD; Lise-Andree Mercier, MD; Stephane Noble, MD; Reda Ibrahim, MD



(Circ Cardiovasc Intervent 2009)

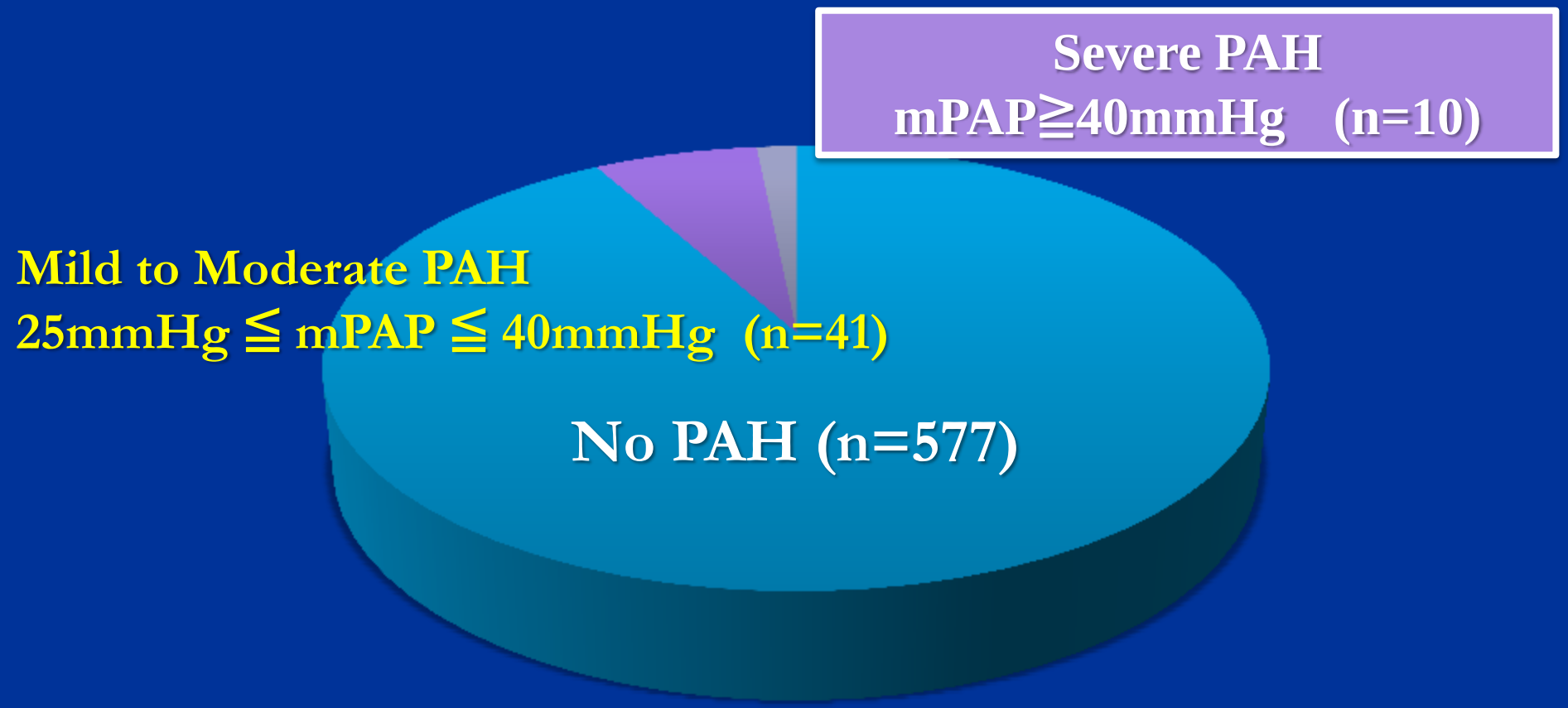
ASD complicated with PH

- PH due to large ASD or increased PA flow
- PH due to Primary Pulmonary Lesions

ESC guidelines for the management of ASD

Indications	Class[a]	Level[b]
Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptoms	I	B[26]
Device closure is the method of choice for secundum ASD closure when applicable	I	C
All ASDs regardless of size in patients with suspicion of paradoxical embolism (exclusion of other causes) should be considered for intervention	IIa	C
Patients with PVR ≥5 WU but <2/3 SVR or PAP <2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L–R shunt (Qp:Qs >1.5) may be considered for intervention	IIb	C
ASD closure must be avoided in patients with Eisenmenger physiology	III	C

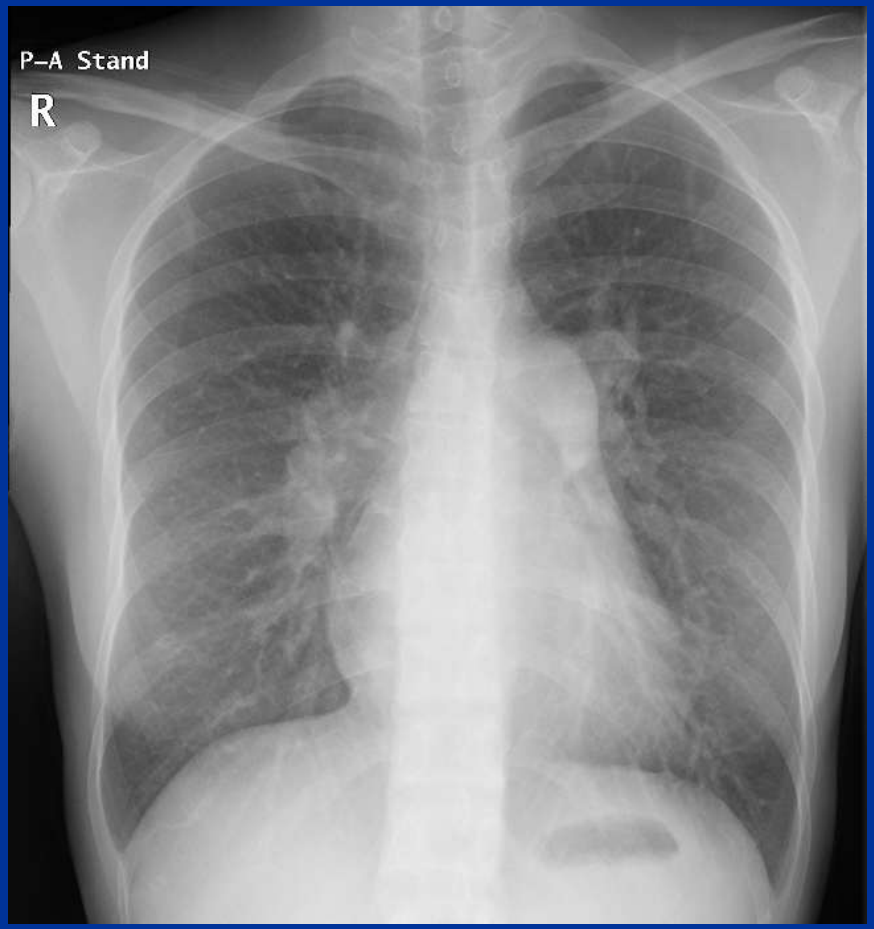
Incidence of PAH in candidates for catheter ASD closure

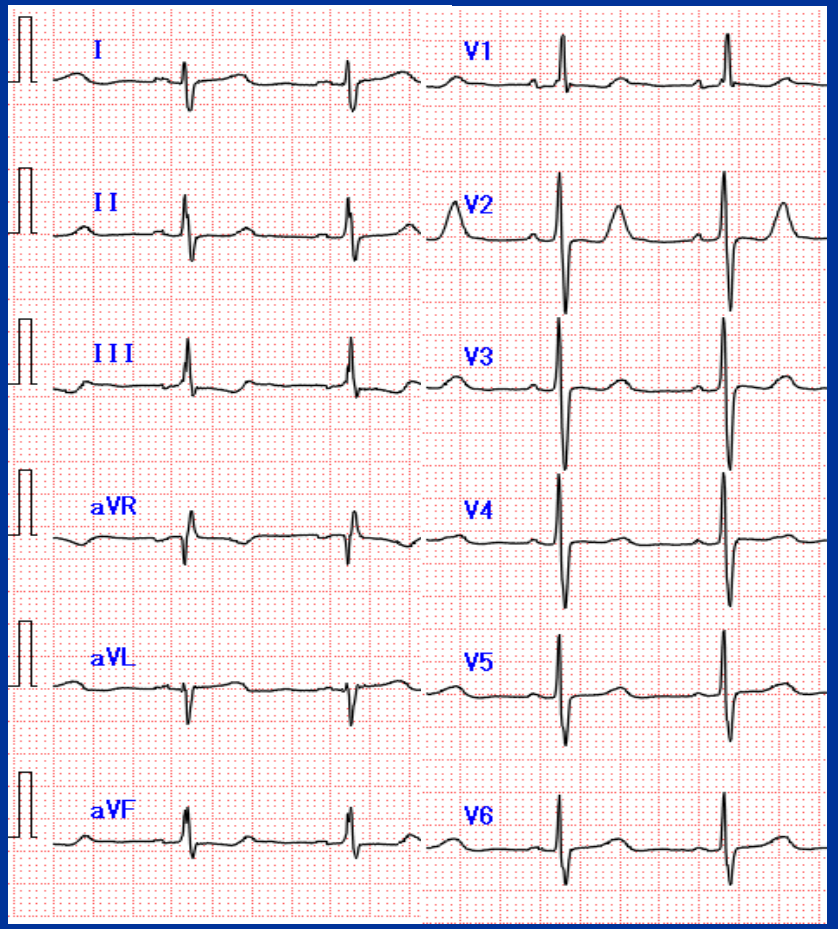


PAH; mean PAP ≧ 25mmHg

(n =628)

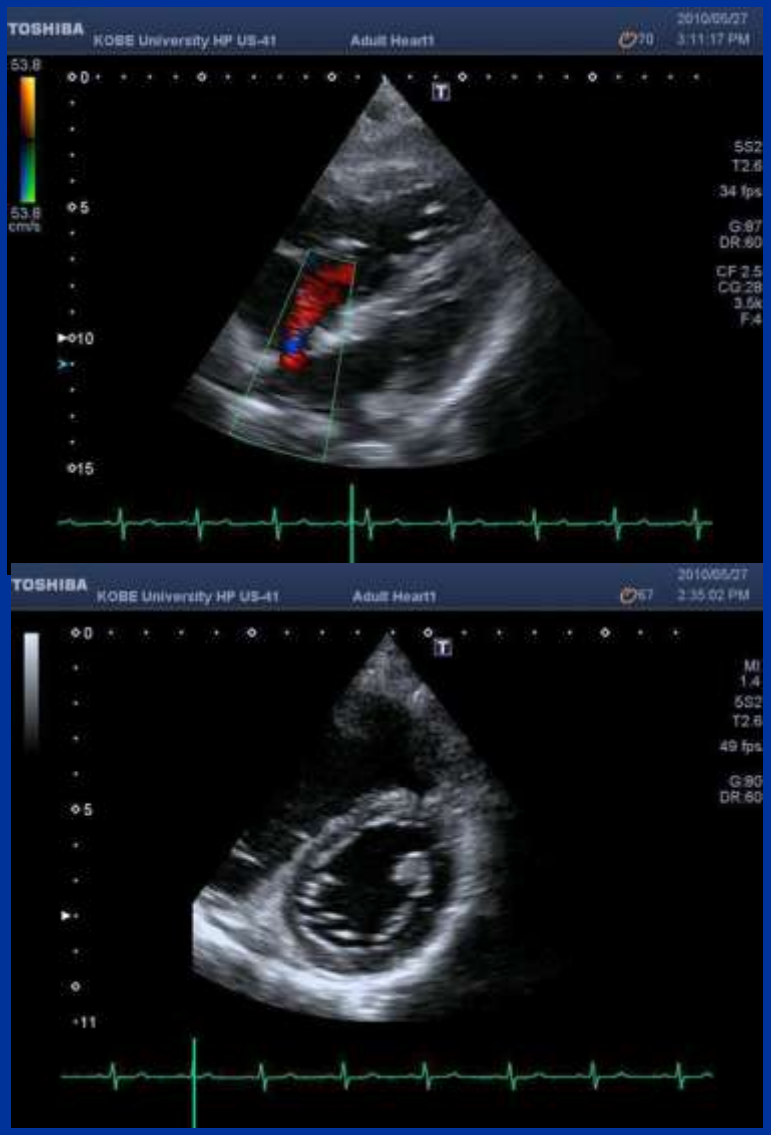
ASD with Severe Pulmonary Hypertension





34 years old female, Dyspnea, Syncope after exercise

ASD + severe PH



Estimated PAP **113/32 mmHg**

RA/RV dilatation

ASD

defect=15 mm x 13.5 mm
L→R shunt (++)
R→L shunt (+)
Qp/Qs = 1.1

ASD with severe PH

Before medication

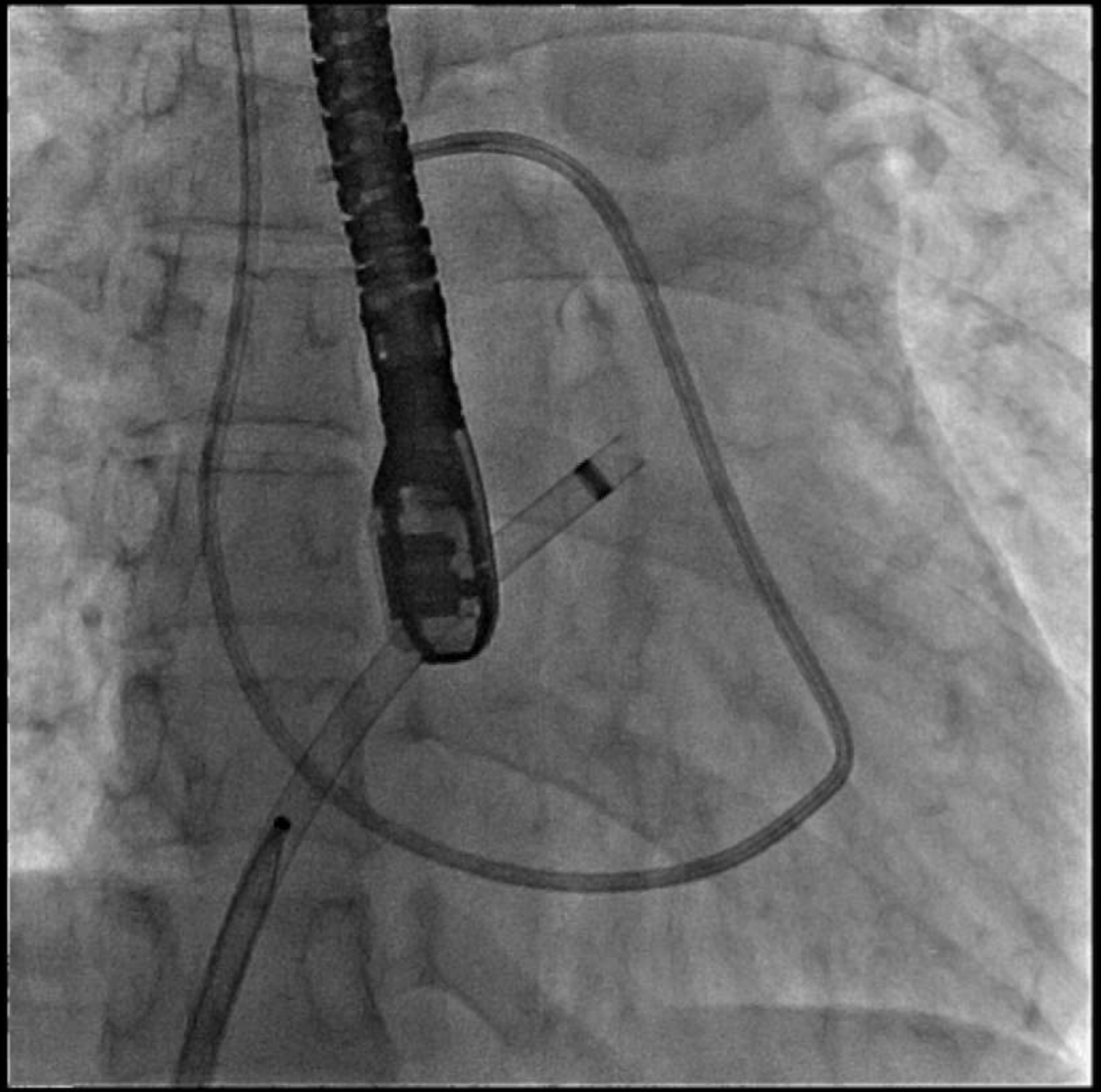
6MWD 20m, PAP 87/30 (57) mmHg,
PVR 8.7 wood unit (697 dyne・sec/cm^{-5})
Qp/Qs=1.32

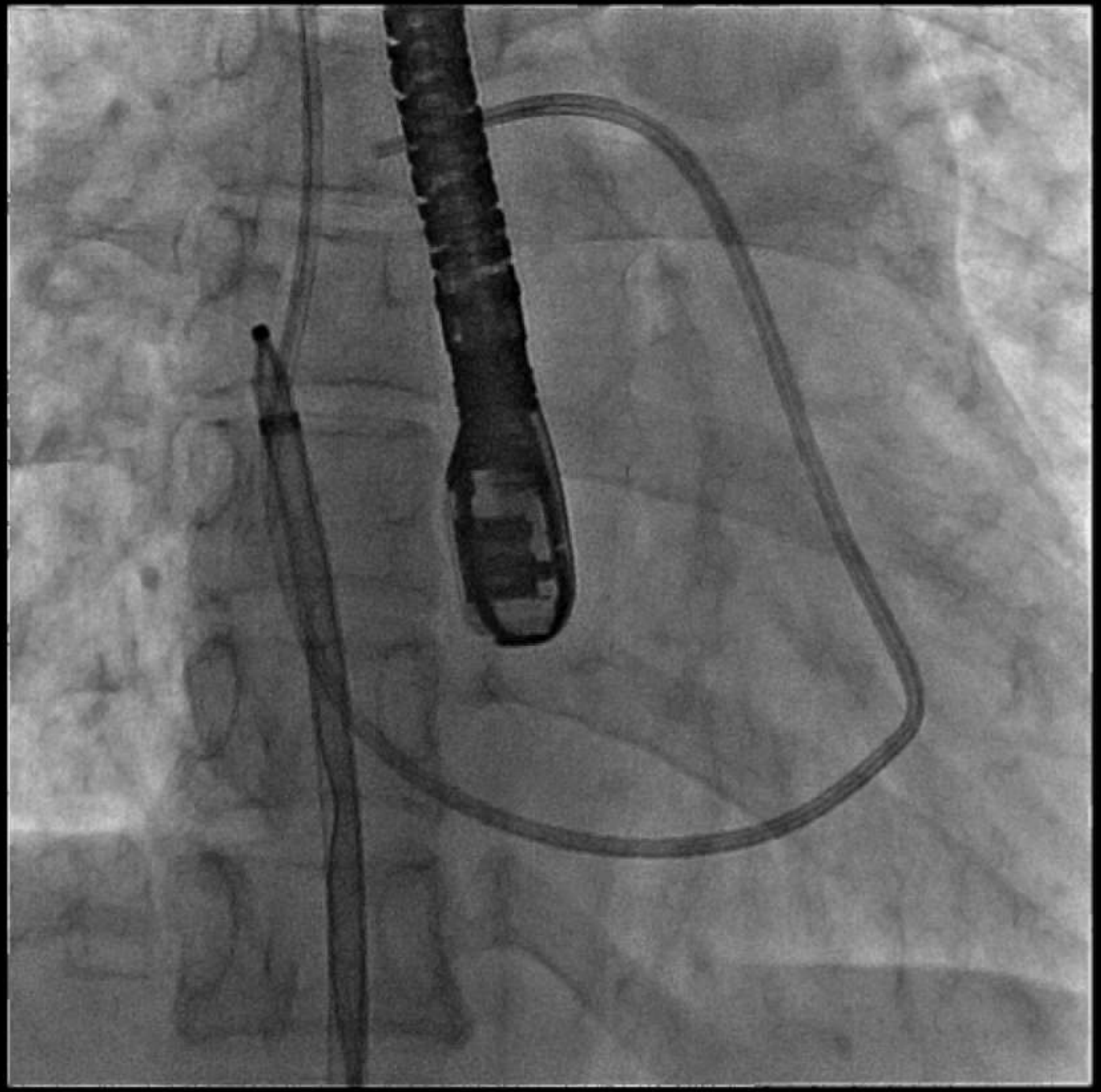
bosentan	250mg
sildenafil	60mg

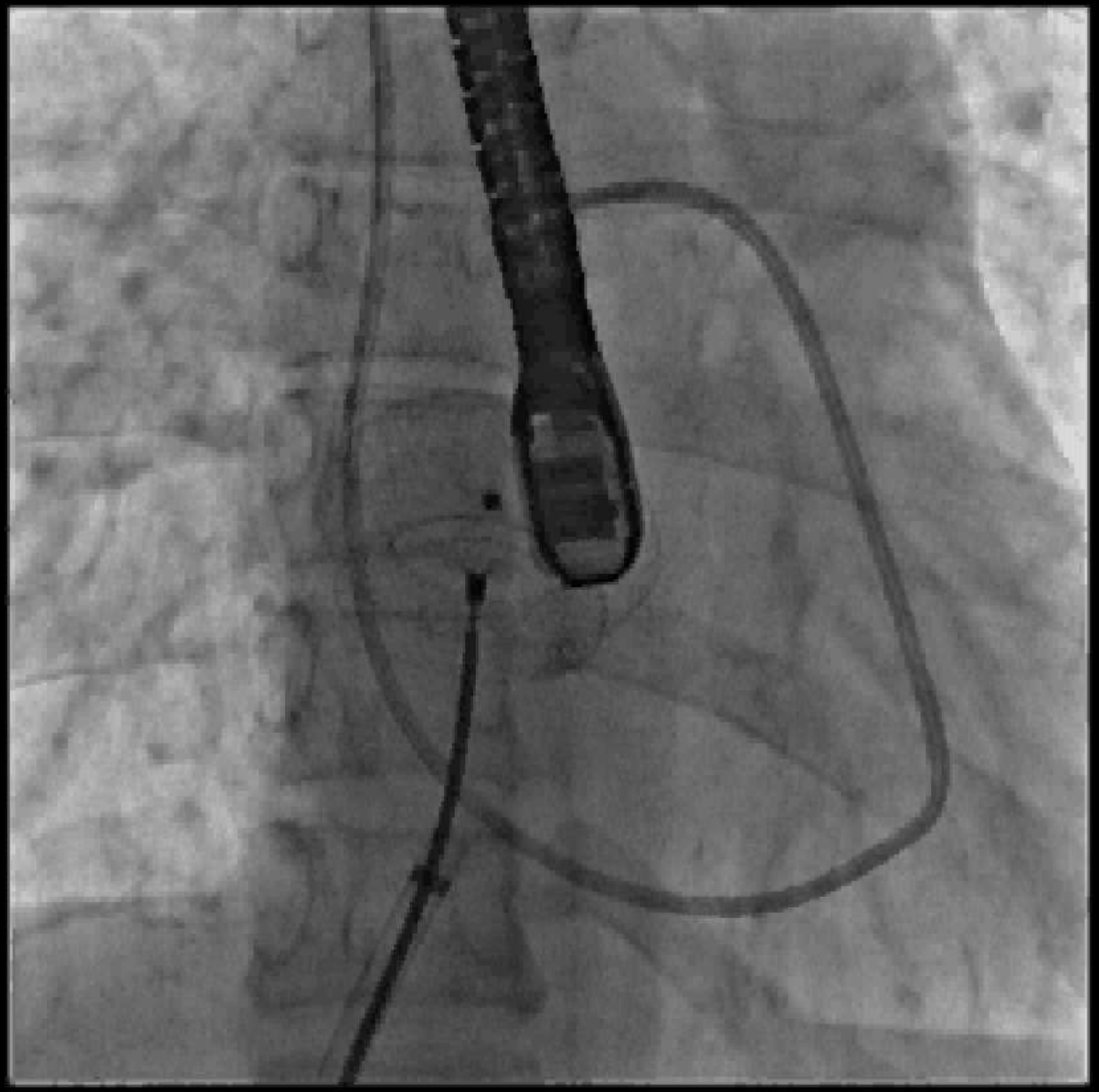
After medication

6MWD 350m, PAP 60/21 (35) mmHg,
PVR 3.6 wood unit (291 dyne・sec/cm^{-5})
Qp/Qs=2.19

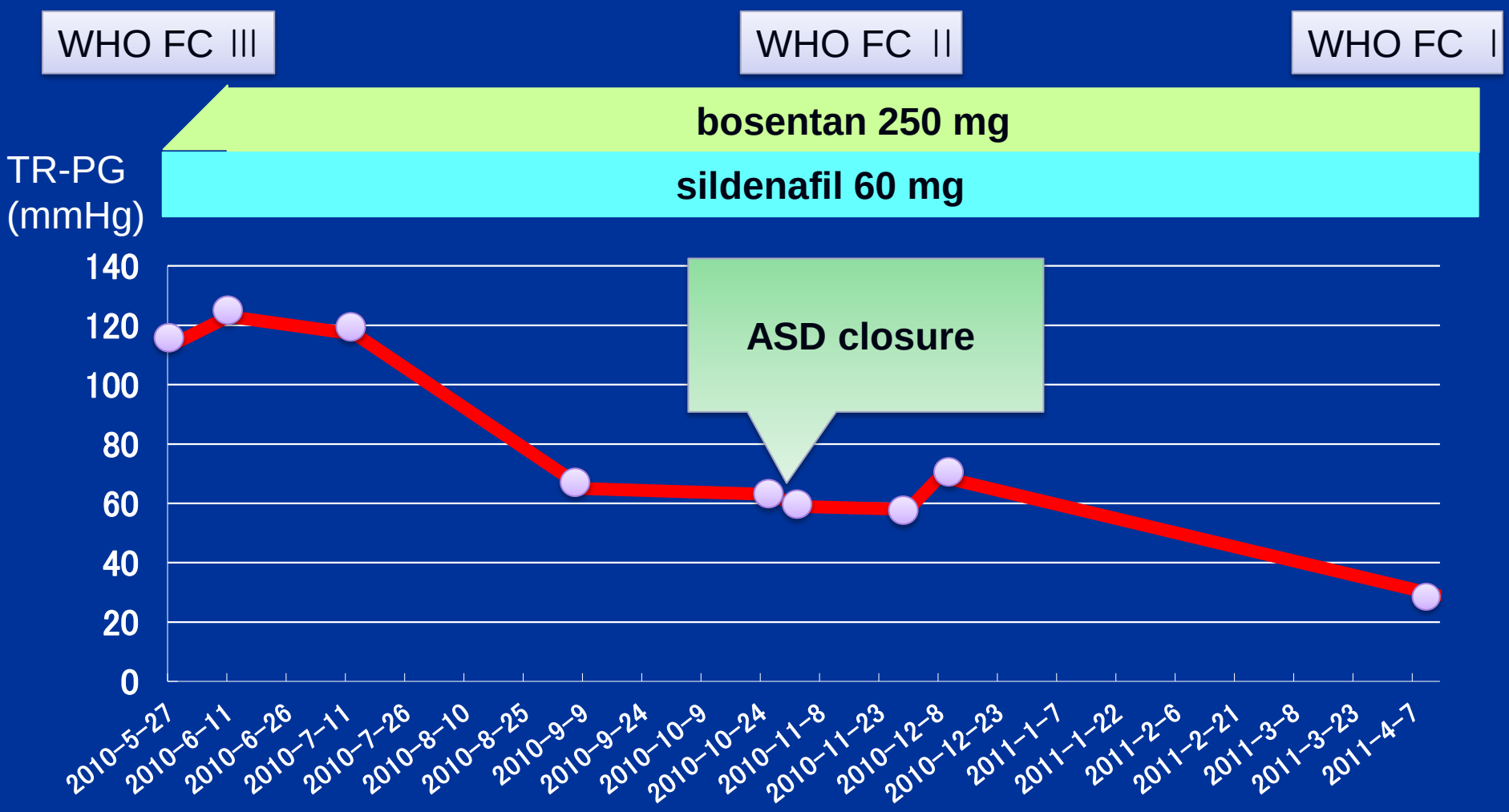
catheter closure of ASD was performed







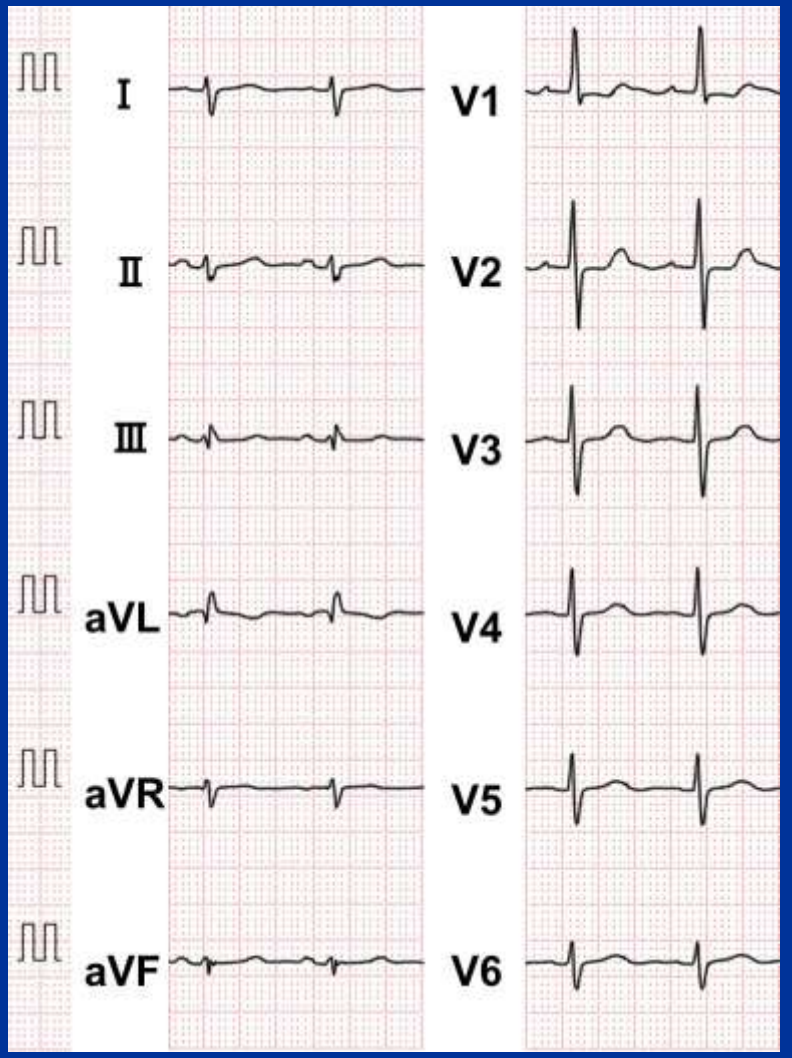
Clinical Course



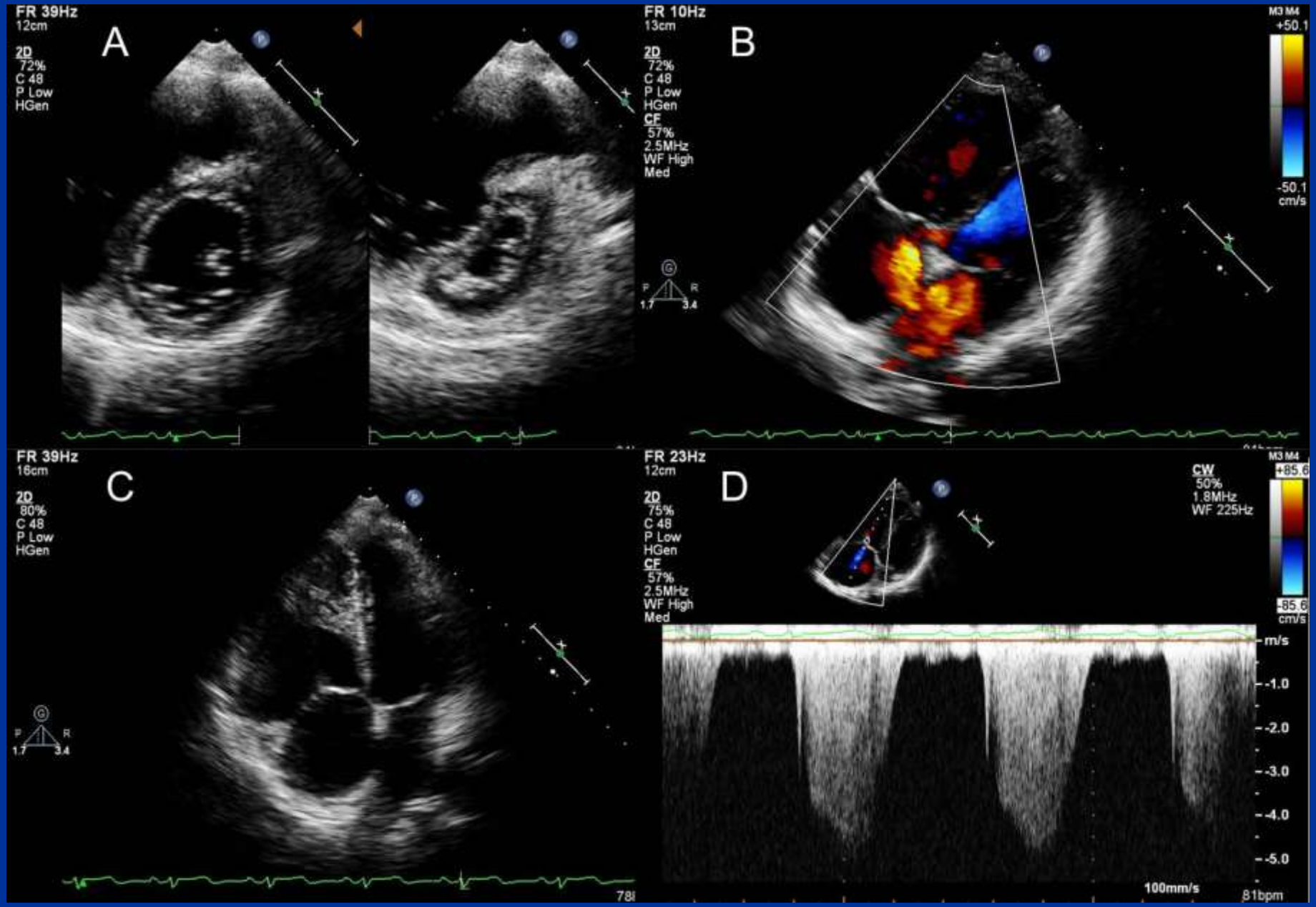
Qp/Qs=1.32
PVR 697
mPAP=57

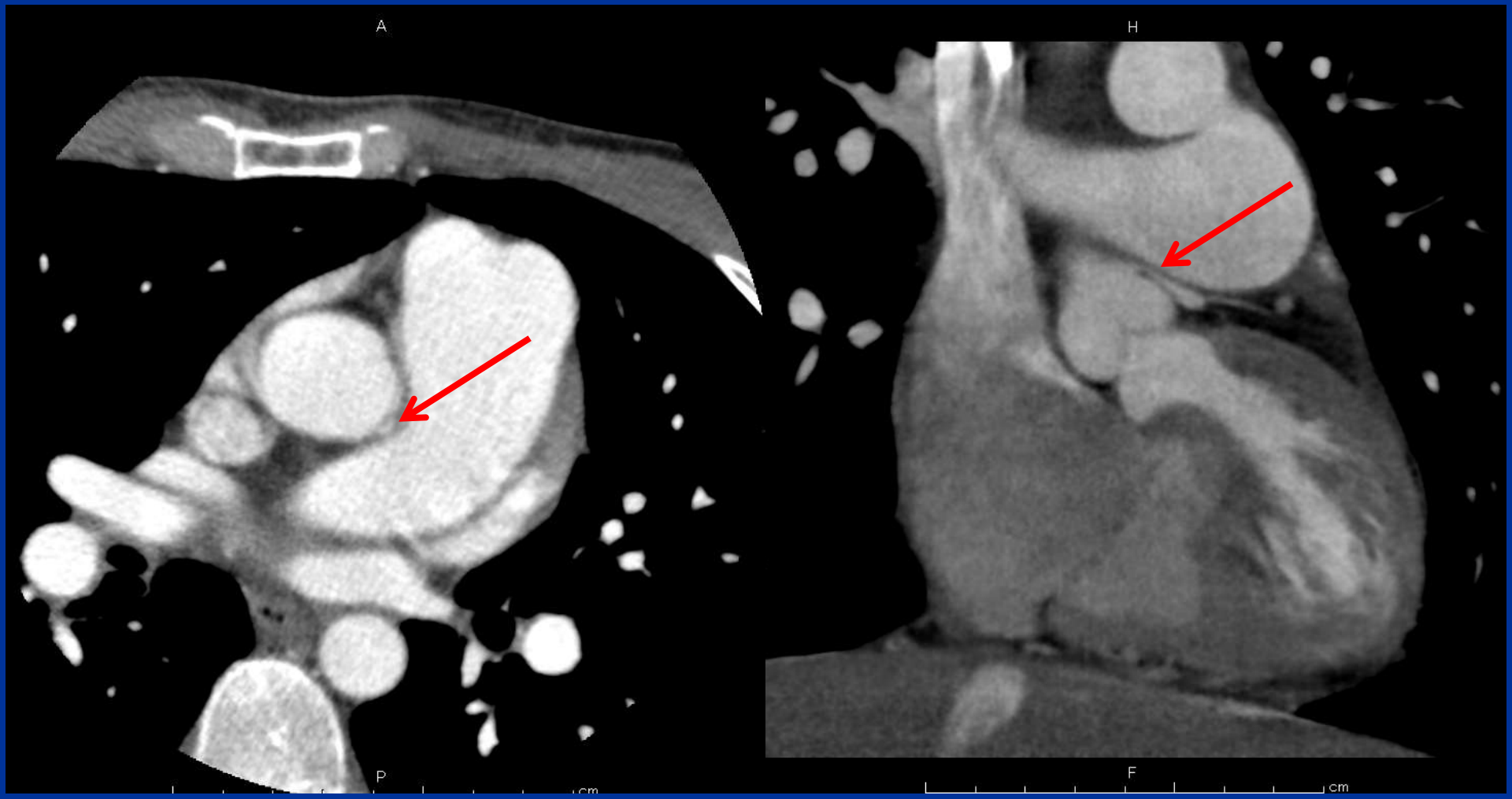
Qp/Qs=2.19
PVR 291
mPAP=35

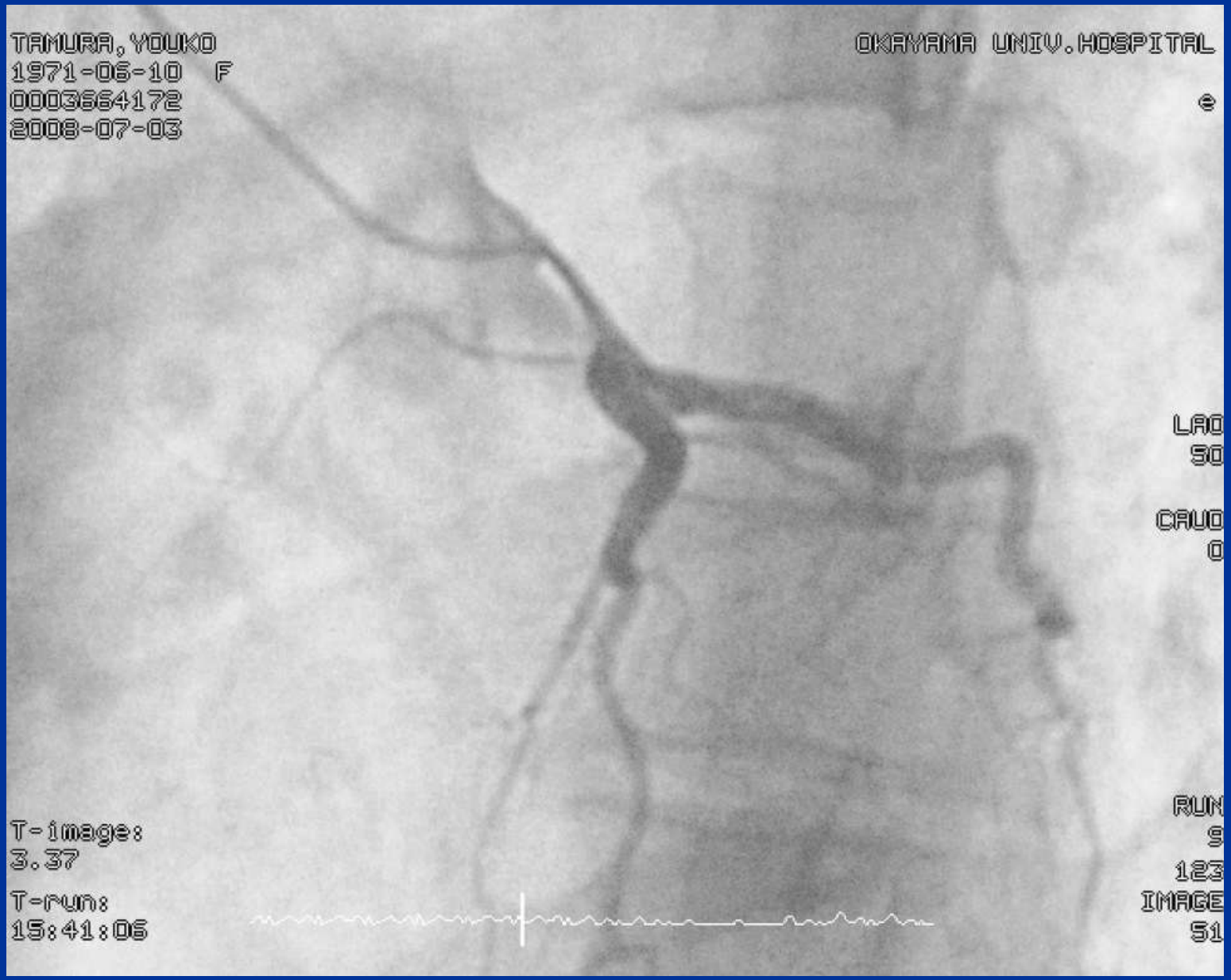
37 years old female, Eisenmenger syndrome ?



PAP 113/50(73) mmHg, PVR 12.8 wood unit
Qp/Qs = 1.0


A
FR 39Hz
12cm
2D
72%
C 48
P Low
HGen
B
FR 10Hz
13cm
2D
72%
C 48
P Low
HGen
CF
57%
2.5MHz
WF High
Med
M3 M4
+50.1
-50.1
cm/s
C
FR 39Hz
16cm
2D
80%
C 48
P Low
HGen
D
FR 23Hz
12cm
2D
75%
C 48
P Low
HGen
CF
57%
2.5MHz
WF High
Med
CW
50%
1.8MHz
WF 225Hz
M3 M4
+85.6
-85.6
cm/s
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
100mm/s
81bpm


A
P
cm
H
F
cm


TAMURA,YOUKO
1971-06-10 F
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e
LAO
50
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T-image:
3.37
T-run:
15:41:06
RUN
9
123
IMAGE
51

37 years-old

PAP 113/50(73) mmHg, PVR 12.8 wood unit

Qp/Qs: 1.0

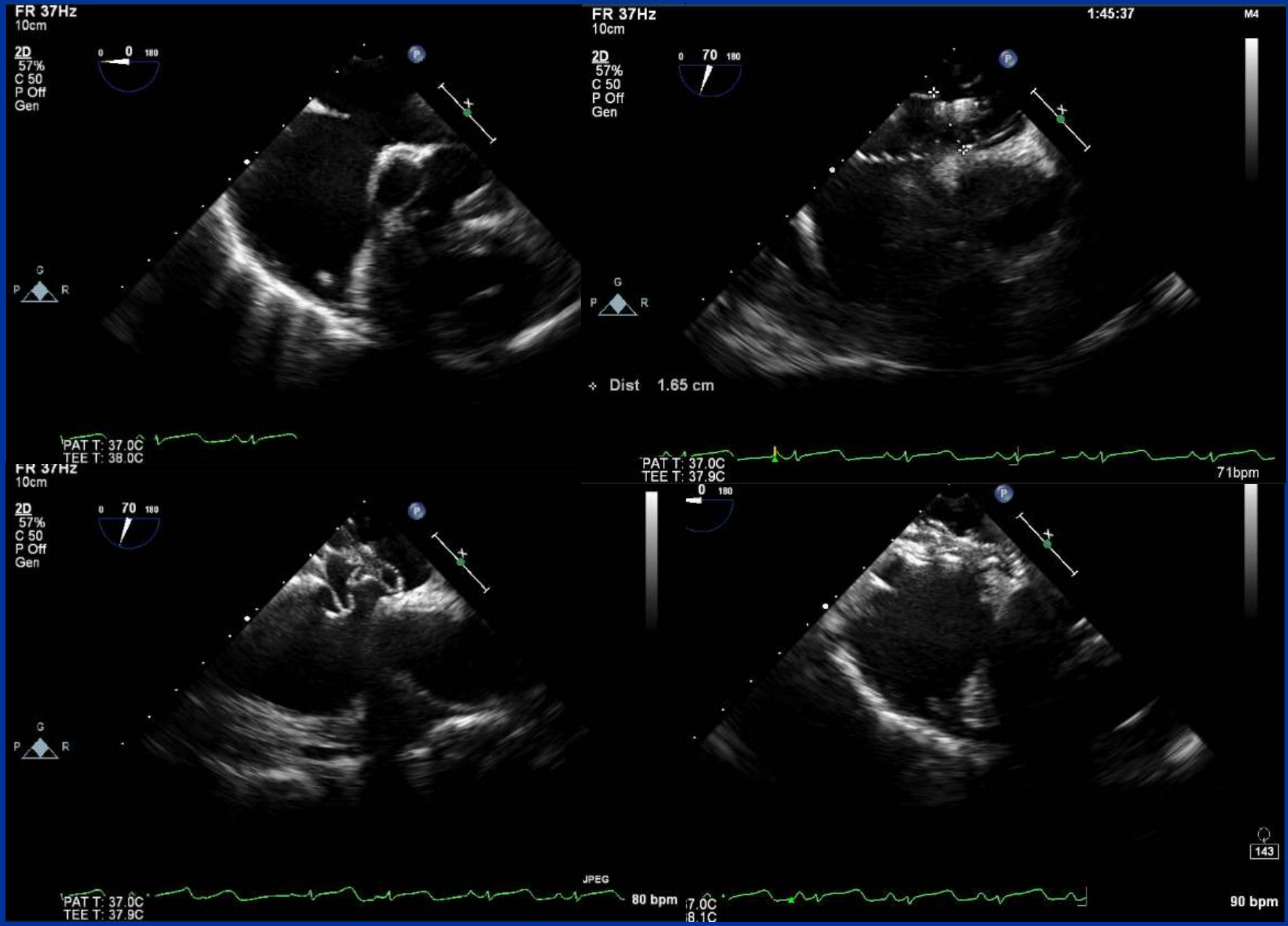
Epoprostenol 110ng/kg/min

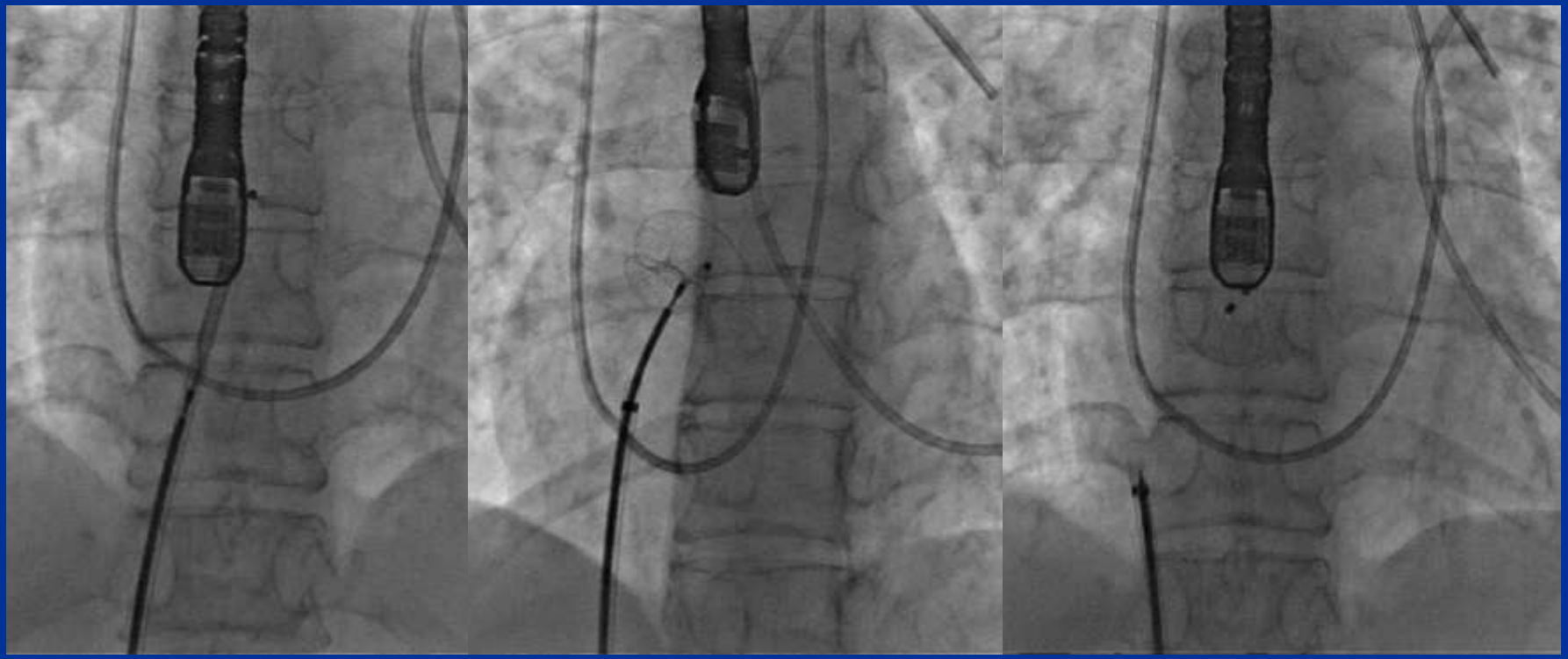
Bosentan 125mg/day

41 years

PAP 53/22(38) mmHg, PVR 4.3 wood unit

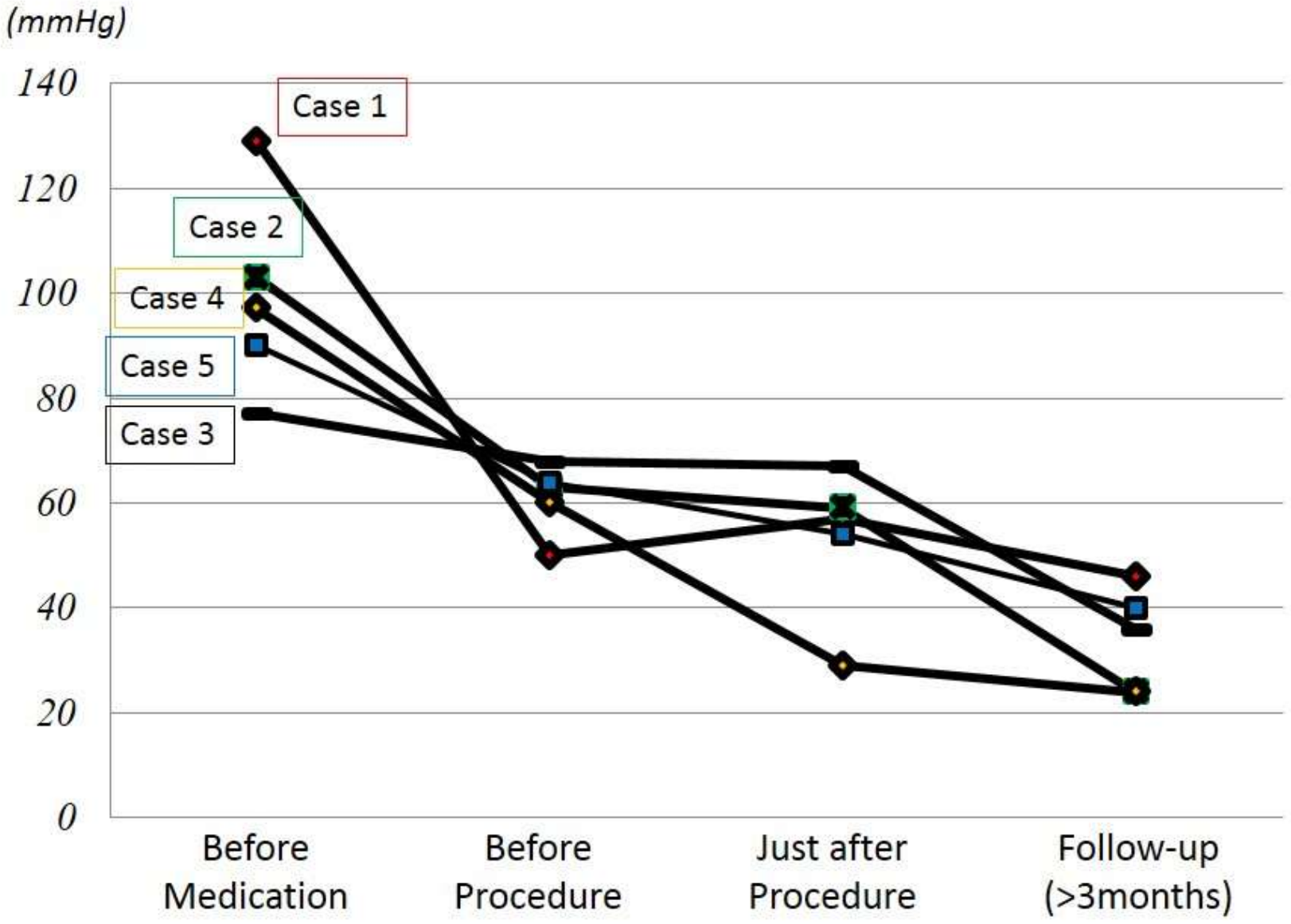
Qp/Qs: 1.6


FR 37Hz
10cm
2D
57%
C 50
P Off
Gen
0 0 180
PAT T: 37.0C
TEE T: 38.0C
FR 37Hz
10cm
1:45:37
M4
2D
57%
C 50
P Off
Gen
0 70 180
Dist 1.65 cm
PAT T: 37.0C
TEE T: 37.9C
71bpm
FR 37Hz
10cm
2D
57%
C 50
P Off
Gen
0 70 180
JPEG
PAT T: 37.0C
TEE T: 37.9C
80 bpm
0 180
143
90 bpm

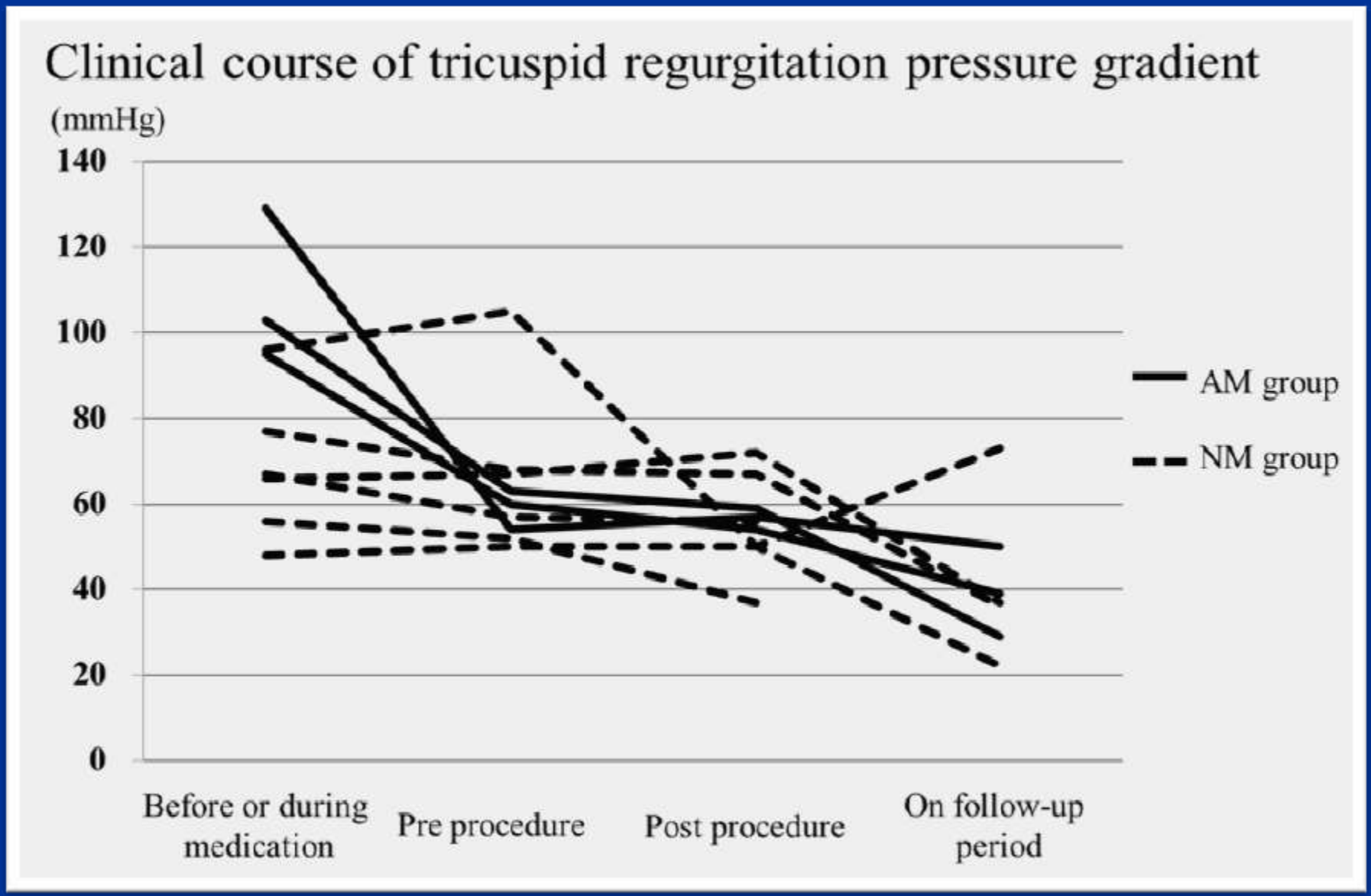


PAH Specific Medical Treatment

Case 1	**epoprostnol 65 ng/kg/min, bosentan 125mg/day**
Case 2	**sildenafil 60 mg/day, bosentan 250 mg/day**
Case 3	**beraprost 360 µg/day, sidenafil 60 mg/day, bosentan 250 mg/day**
Case 4	**bosentan 125 mg/day**
Case 5	**epoprostnol 120 ng/kg/min, bosentan 125 mg/day**


(mmHg)
140
120
100
80
60
40
20
0
Case 1
Case 2
Case 4
Case 5
Case 3
Before Medication
Before Procedure
Just after Procedure
Follow-up (>3months)

Catheter closure of ASD complicated with severe PH (mean PAP >40 mmHg)



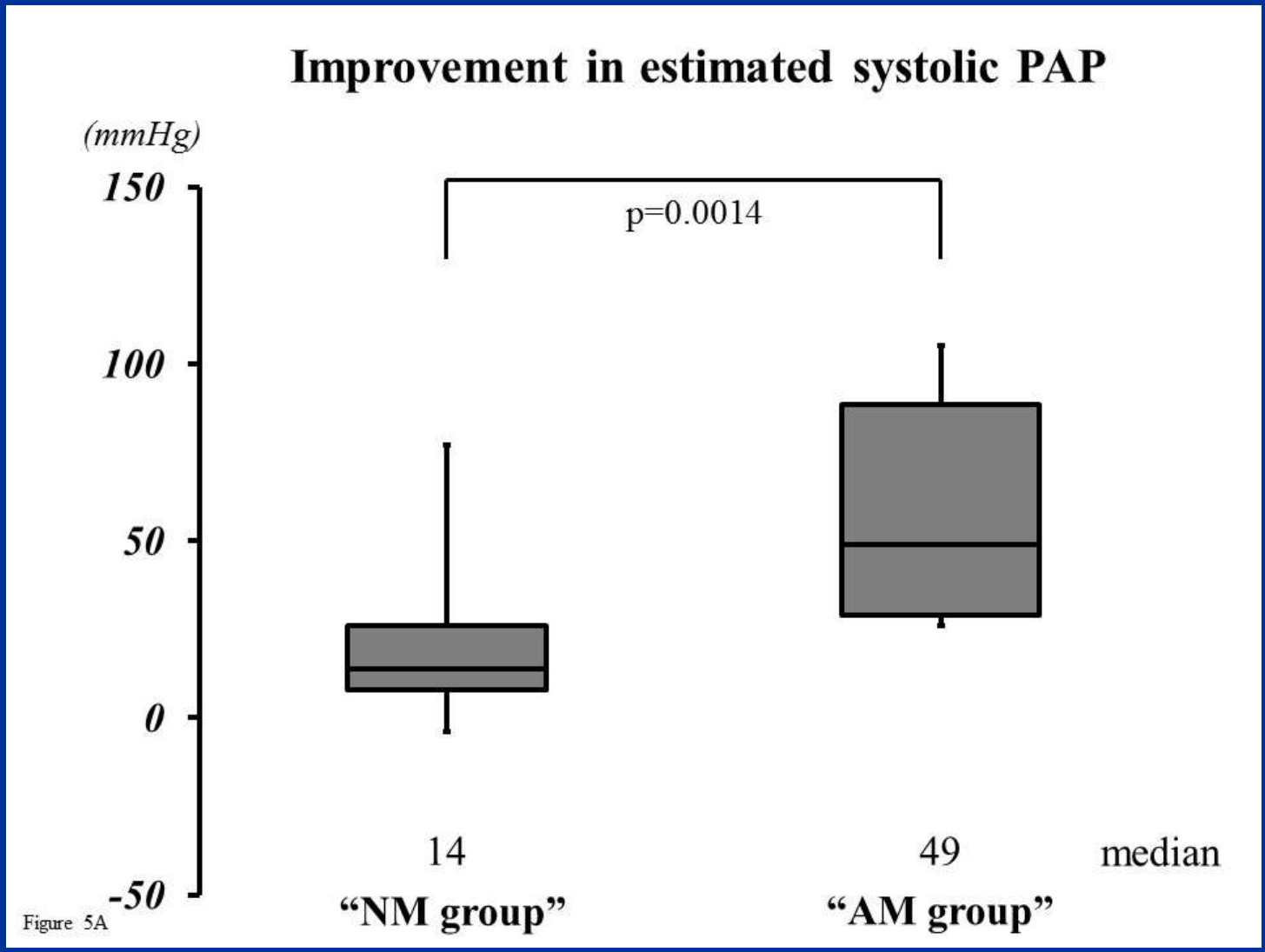
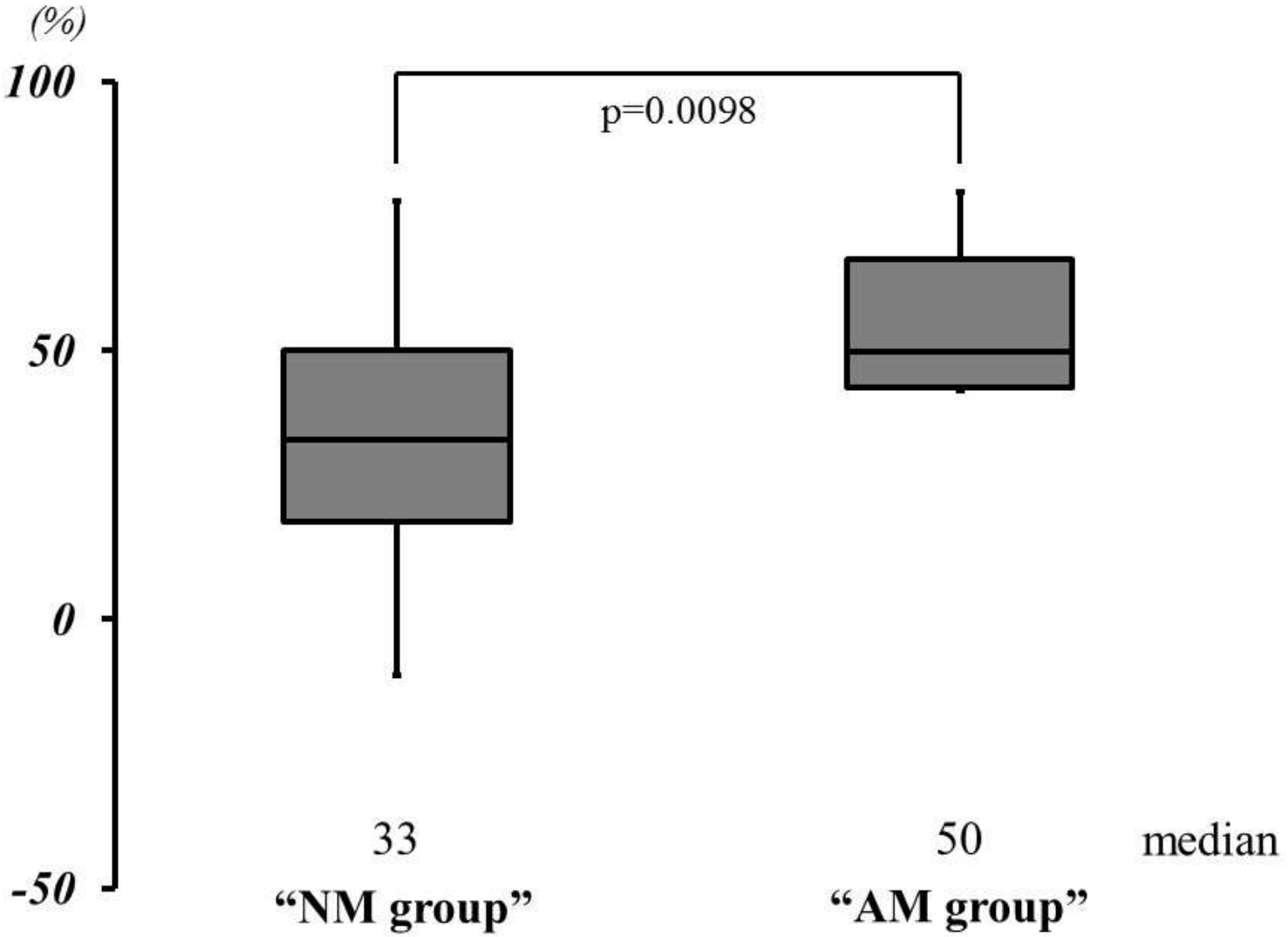

Improvement in estimated systolic PAP
(mmHg)
150
100
50
0
-50
p=0.0014
14
49
median
"NM group"
"AM group"

Figure 5A

Degree of improvement in estimated systolic PAP



Role of Interventional Cardiology for ASD with Pulmonary Hypertension

- Expands the therapeutic indication
- Combination of catheter intervention and PAH specific medication contributes the improvement of long-term outcome.