'Treat-and-Repair' vs. 'Repair-and-Treat' : What's the Evidence?

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SMC TS

Closure of septal defects in patients with severe PAH

Pros	Cons
May prevent Eisenmenger	May convert to iPAH
Advanced medical therapies	Perioperative risk

- Operable?

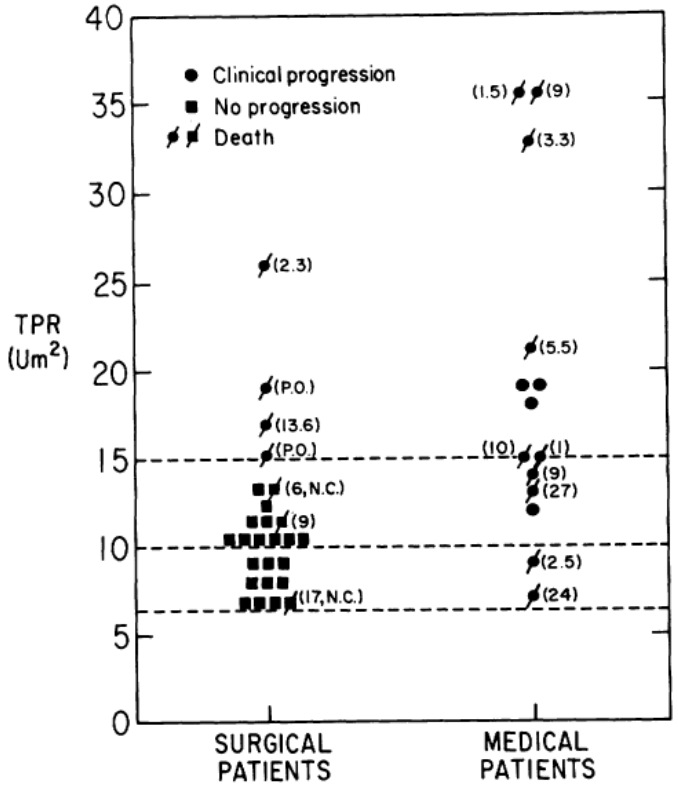
- Reversible? vs. Irreversible?
- Pulmonary vasoreactivity

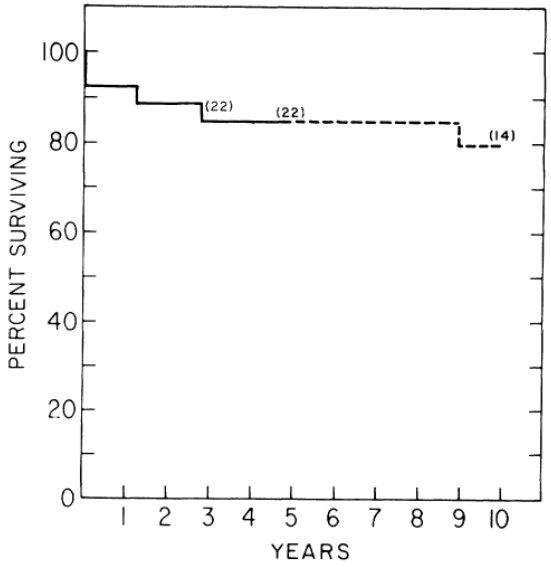
Isolated atrial septal defect with pulmonary vascular obstructive disease — long-term follow-up and prediction of outcome after surgical correction

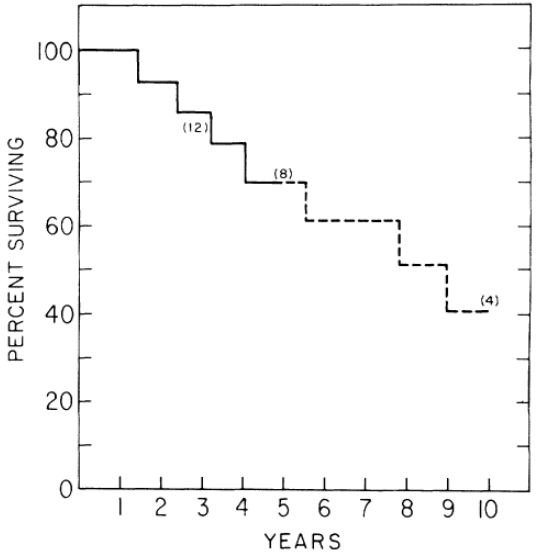
PETER M. STEELE, M.B.B.S. (HONS), VALENTIN FUSTER, M.D., MARC COHEN, M.D., DONALD G. RITTER, M.D., AND DWIGHT C. MCGOON, M.D.

Circulation 1987;76:1037-42

- 40 pts > PVR 7U/m2 out of 702 ASD pts







Isolated atrial septal defect with pulmonary vascular obstructive disease — long-term follow-up and prediction of outcome after surgical correction

PETER M. STEELE, M.B.B.S. (HONS), VALENTIN FUSTER, M.D., MARC COHEN, M.D., DONALD G. RITTER, M.D., AND DWIGHT C. MCGOON, M.D.

Circulation 1987;76:1037-42

- For patients with an ASD and PVOD with a predominant L → R shunt,
  - All patients with a total pulmonary resistance less than 10 U/M^2 should proceed to operation
  - if the total pulmonary resistance is 15 U/M^2 or greater than, operation is not advised
- In patients with borderline total pulmonary resistance, the systemic arterial oxygen saturation provides a good prediction of surgical outcome.

Atrial septal defects versus ventricular septal defects in BREATHE-5, a placebo-controlled study of pulmonary arterial hypertension related to Eisenmenger's syndrome: A subgroup analysis ☆

Rolf M.F. Berger [a,*], Maurice Beghetti [b], Nazzareno Galiè [c], Michael A. Gatzoulis [d], John Granton [e], Andrea Lauer [f], Eleonora Chiossi [f], Michael Landzberg [g]

Int J Cardiol 2010;144:373-8

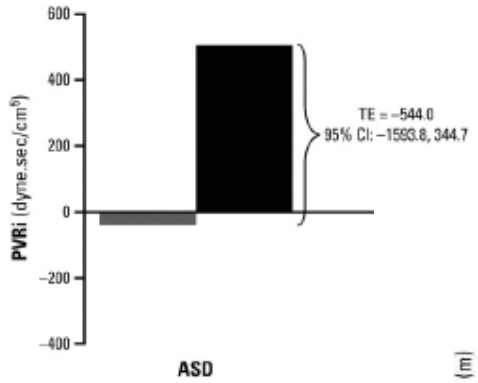


Fig. 1. Indexed pulmonary vascular resistance (PVRi): median cha

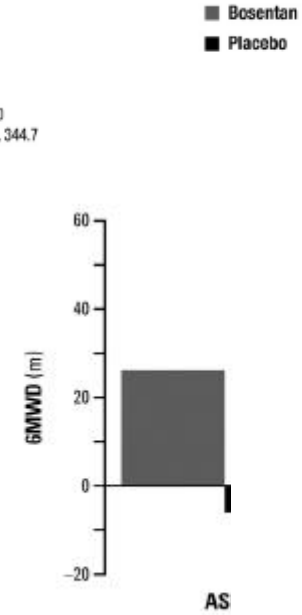


Fig. 2. 6-minute walk distance (6M

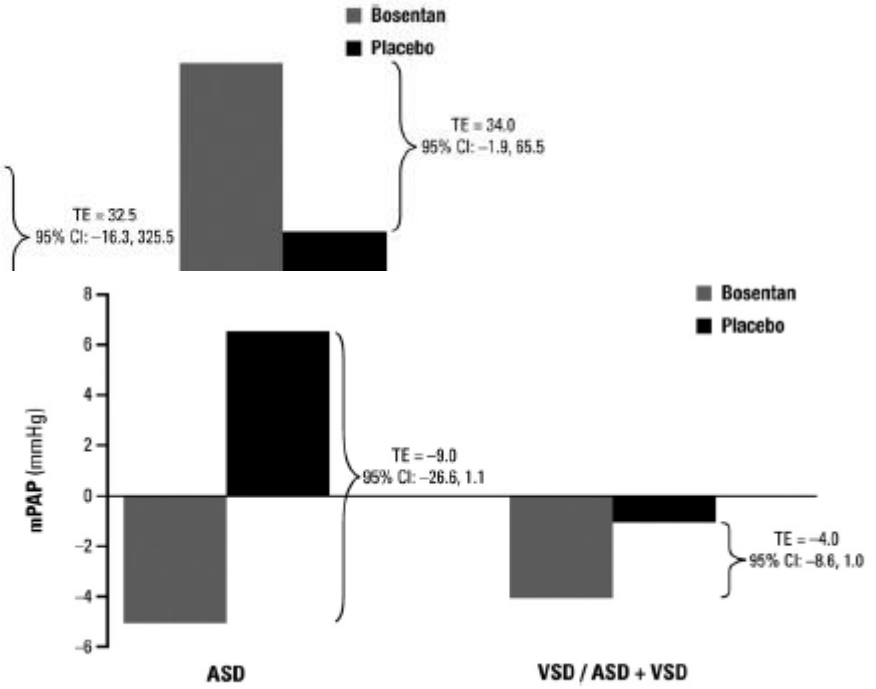


Fig. 3. Mean pulmonary artery pressure (mPAP): median change from baseline to week 16, (TE: treatment effect; CI: confidence interval).

ESC Guidelines for the Management of ACHD 2010

Indications for intervention in ASD	Class	Level
Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of s ymptoms	I	B
All ASDs regardless of size in patients with suspicion of para doxical embolism (exclusion of other causes) should be consi dered 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

ESC Guidelines for the Management of ACHD 2010

Indications for intervention in VSD	Class	Level
Patients with symptoms that can be attributed to L–R shunting through the (residual) VSD and who have no severe pulmonary vascular disease (see below) should undergo surgical VSD closure	I	C
Patients with VSD and PAH should be considered for surgery when there is still net L–R shunt (Qp:Qs >1.5) present and PAP or PVR are <2/3 of systemic values (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy)	IIa	C
Surgery must be avoided in Eisenmenger VSD and when exercise-induced desaturation is present	III	C

ESC Guidelines for the Management of ACHD 2010

Indications for intervention in PDA	Class	Level
PDA should be closed in patients with PAH but PAP <2/3 of systemic pressure or PVR <2/3 of SVR	I	C
PDA closure should be considered in patients with PAH and PAP >2/3 of systemic pressure or PVR >2/3 of SVR but still n et L–R shunt (Qp:Qs >1.5) or when testing (preferably with ni tric oxide) or treatment demonstrates pulmonary vascular re activity	IIa	C
PDA closure must be avoided in PDA Eisenmenger and patients with exercise-induced lower limb desaturation	III	C

"Treat-and-Repair"

Reversal of Pulmonary Hypertension and Subsequent Repair of Atrial Septal Defect After Treatment With Continuous Intravenous Epoprostenol

Adaani E. Frost, MD, Miguel A. Quiñones, MD, William A. Zoghbi, MD, and George P. Noon, MD

- 29 years old
- PAP 86/35 , systemic 110/70, SaO_2 93%
- 4 year therapy of prostacyclin 90 mg/kg/min
- ASD closure , 8 years follow up
- Amlodipine, coumadin, and cessation of prostacyclin

J Heart Lung Transplant 2005;24:501-3

Continuous Epoprostenol Therapy and Septal Defect Closure in a Patient With Severe Pulmonary Hypertension

Aki Hirabayashi,[1] MD, Katsumasa Miyaji,[1]* MD, and Teiji Akagi,[2] MD, FSCAI, FACC

A 31-year-old woman with exertional dyspnea diagnosed as having atrial septal defect (ASD) with severe pulmonary hypertension (PH). Intravenous epoprostenol therapy was started to improve PH. Although pulmonary arterial pressure decreased, her symptoms remained in class III of WHO functional class, probably because of exacerbation of the left-to-right shunt caused by the reduction of pulmonary vascular resistance (PVR). Transcatheter atrial septal closure was therefore performed. Soon after the procedure, additional reduction in pulmonary arterial pressure was achieved. Her symptoms improved and oxygen inhalation was discontinued. One year after the procedure, although intravenous epoprostenol was still required, her symptoms had improved to class I of WHO functional class without exacerbation of PH. Transcatheter atrial septal closure after lowering PVR by intravenous epoprostenol would be a novel therapy for patients with ASD accompanied by PH.

Catheter Cardiovasc Interv 2009;73:688-91

Atrial septal defect repair after a 10-month treatment with bosentan in a patient with severe pulmonary arterial hypertension: A case report

Konrad Hoetzenecker, MD,[a] Hendrik J. Ankersmit, MD,[a] Diana Bonderman, MD,[b] Wolfram Hoetzenecker, MD, PhD,[c] Reinald Seitelberger, MD,[a] Walter Klepetko, MD,[a] and Irene M. Lang, MD,[b] Vienna, Austria, and Tübingen, Germany

J Thorac Cardiovasc Surg 2009;137:760-1

TABLE 1. Time course of clinical and hemodynamic parameters

Parameter	Before bosentan initiation		Before ASD repair	8 months after ASD repair
	Baseline	NO (20 ppm)		
mLAP (mm Hg)	17	17	11	13
mRAP (mm Hg)	16	16	10	7
mPAP (mm Hg)	54	42	30	35
Pulmonary/systemic flow (L/min)	9.5/3.5	14.0/4.3	7.4/3.3	5.2/5.2
Arterial saturation (%)	91	95	88	95
Pulmonary arterial saturation (%)	75	84.5	77.7	62.0
Total pulmonary resistance (dynes · s · cm^{-5})	460	240	325	538
B-type natriuretic peptide (pg/mL)	4675		1104	420
6-min walking distance (m)	152.2		241.0	247.0

ASD, Atrial septal defect; *NO*, nitric oxide; *mLAP*, mean left atrial pressure; *mRAP*, mean right atrial pressure; *ND*, not done; *mPAP*, mean pulmonary arterial pressure.

Repair of Atrial Septal Defect With Eisenmenger Syndrome After Long-Term Sildenafil Therapy

Young-Hwue Kim, MD, PhD, Jeong Jin Yu, MD, PhD, Tae-Jin Yun, MD, PhD, Yonghee Lee, MD, PhD, Yong Beom Kim, MD, Hyung Soon Choi, MD, Won Kyoung Jhang, MD, Hong Ju Shin, MD, Jeong-Jun Park, MD, PhD, Dong-Man Seo, MD, PhD, Jae-Kon Ko, MD, PhD, and In-Sook Park, MD, PhD

Table 1. Hemodynamic Data

	Initial Study		2 Years of Sildenafil Therapy		
Variable	Baseline	O_2 (10 L/min)	Baseline	O_2 (10 L/min)	Balloon Occlusion Test
mRAP, mm Hg	1	2	10	8	8
mLAP, mm Hg	1	2	10	8	
mPAP, mm Hg	87/20, 55	85/20, 55	128/32, 75	110/34, 65	99/26, 56
mSAP, mm Hg	140/80, 100	140/83, 105	137/71, 98	131/75, 95	133/68, 95
$Svco_2$, %	69	74	76	68	79
Pao_2, %	75	80	86	89	
Sao_2, %	89	92	94	99	99
Qp (L/min/m^2)	2.16	2.24	5.15	4.7	
Qs (L/min/m^2)	2.49	2.72	2.98	1.75	2.83
Qp/Qs ratio	0.87	0.82	1.73	2.68	
PVRI (WU · m^2)	25.0	23.7	12.63	12.1	
SVRI (WU · m^2)	39.8	37.8	29.53	49.7	30.73
PVRI/SVRI	0.63	0.63	0.43	0.24	

mLAP = mean left atrial pressure; mPAP = mean pulmonary arterial pressure; mRAP = mean right atrial pressure; mSAP = mean systemic arterial pressure; PVRI = body surface area indexed pulmonary vascular resistance; Pao_2 = partial pressure of arterial oxygen; Qp = body surface area indexed pulmonary blood flow; Qs = body surface area indexed systemic blood flow (cardiac index); Sao_2 = arterial oxygen saturation; $Svco_2$ = oxygen saturation in the superior vena cava; SVRI = body surface area indexed systemic vascular resistance; WU = Wood unit.

‘Repair-and-Treat’

- Advanced medical therapies without correction of the underlying anatomical defect may lead to further insult on the pulmonary circulation
  - ↓ PVR → ↑ pulmonary flow, shear stress
  - Deterioration after the initial short-term improvement
  - Less advanced case → further progression

Staged repair : PAB

Successful Reversal of Pulmonary Hypertension in Eisenmenger Complex

Randas J. V. Batista, José L. V. Santos, Noriaki Takeshita, Lise Eocchino, Paulo N. Lima, Marilu Goehr, Marco A. Cunha, Akira T. Kawaguchi, Tomas A. Salerno

Campina Grande do Sul, P

Arq Bras Cardiol 1997; 68: 279-280

- 19 year old female, VSD + ASD with PAH
- Lung biopsy – grade IV (HE)
- PA banding

- One year later,
  - No cyanosis, lung biopsy – regression of pulmonary vascular change
- VSD, ASD closure was done successfully

Successful Reversal of Pulmonary Hypertension in Eisenmenger Complex

Randas J. V. Batista, José L. V. Santos, Noriaki Takeshita, Lise Eocchino, Paulo N. Lima, Marilu Goehr, Marco A. Cunha, Akira T. Kawaguchi, Tomas A. Salerno

Campina Grande do Sul, PR - Brazil

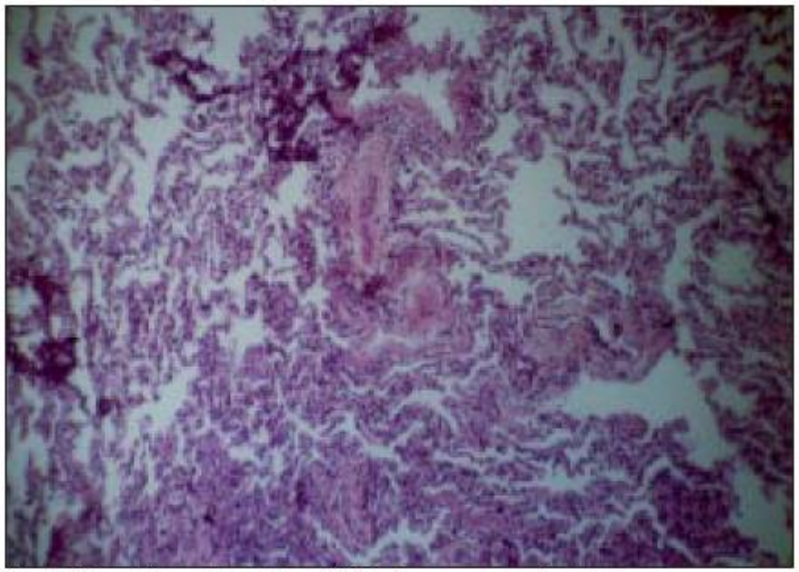


Fig. 1 - Lung biopsy showed grade IV pulmonary vascular changes.

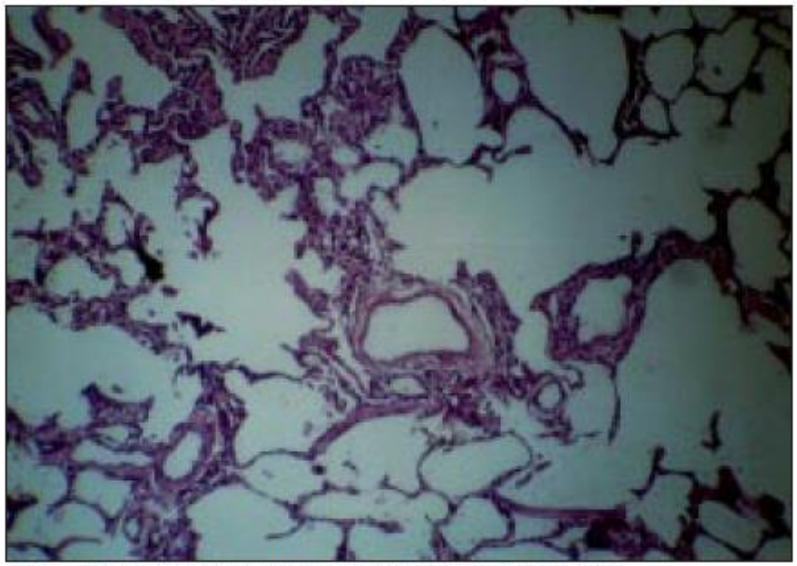


Fig. 2 - Lung biopsy showed total regression of previous lesions.

Arq Bras Cardiol 1997; 68: 279-280

Successful Reversal of Pulmonary Hypertension in Eisenmenger Complex

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Campina Grande do Sul, PR - Brazil

- Lower PA saturation → dilatation of the pulmonary vascular bed and a decrease in PVR → regression of fixed pulmonary lesions
- High oxygen content → increase PVR

- In the course of PA banding, systemic Sao2 was reduced from 97% to 60% or 75% and pulmonary Sao2 from 87% to about 20% in patients with right-to-left shunt without compromising the heart (norespiratory acidosis developed, per a personal communication with Dr. Randas Batista: Chanda. J Thorac Cardiovas Surg 1998;115;484-5)

Arq Bras Cardiol 1997; 68: 279-280

Staged repair : PAB

Evaluation of Pulmonary Artery Banding in the Setting of Ventricular Septal Defects and Severely Elevated Pulmonary Vascular Resistance

Sadaf A. Khan, MD,* Bruce D. Gelb, MD,* Khanh H. Nguyen, MD†

*Division of Pediatric Cardiology and †Department of Cardiothoracic Surgery, Mount Sinai School of Medicine, New York, NY, USA

Table 1. Hemodynamic Data Pre-PAB and Post-PAB for Study Patients

		Mean Right Atrial Pressure (mm Hg)		PCW Pressure (mm Hg)		Indexed PVR (W.u.*m^2)		Systolic PAP (mm Hg)		Cardiac Index (L/min/m^2)		Qp/Qs	
Patient	Status	Room Air	100% O_2	Room Air	100% O_2	Room Air	100% O_2	Room Air	100% O_2	Room Air	100% O_2	Room Air	100% O_2
1	Pre-PAB	1	1	5	8	14.3	9.7	84	100	2.4	4.7	1.7	1.5
	Post-PAB†	5	5	—‡ (LA = 7)	—‡ (LA = 7)	24.4	21.4	72	80	2.7	2.1	0.67	0.84
2	Pre-PAB	2	4	8	14	17.8	5.5	100	90	3.9	4.7	1	2
	Post-PAB	4	4	5	7	3.9	2	24	24	4	5	0.8	1
3	Pre-PAB	1	2	6	8	10	6.2	84	84	4.4	5.1	1.3	1.8
	Post-PAB	2	3	2	3	11.1	8.1	59	58	3.6	4.1	0.84	0.92
4	Pre-PAB	12	14	—§ (LA = 12)	—§ (LA = 14)	10.5	3.1	106	68	2.6	1.9	2.3	6.2
	Post-PAB	10	6	—§ (LA = 10)	—§ (LA = 6)	0.83	2.2	55	63	2.1	1.9	16.5	9.4

†,‡,§Unavailable data. Left atrial pressures listed as reference.
PAB indicates pulmonary artery band; PCW, pulmonary capillary wedge; PVR, pulmonary vascular resistance; PAP, pulmonary artery pressure; LA, left atrial pressure.

Congenit Heart Dis. 2006;1:244–250

Alternative approach for selected severe pulmonary hypertension of congenital heart defect without initial correction — Palliative surgical treatment ☆

Ming-Tai Lin, Yih-Sharng Chen *, Shu-Chien Huang, Hsin-Hui Chiu, Shuenn-Nan Chiu, Chun-An Chen, En-Ting Wu, Ing-Sh Chiu, Chung-I Chang, Mei-Hwan Wu, Jou-Kou Wang

Pediatric Cardiovascular Surgery, Pediatric Cardiology, National Taiwan University Hospital, Taipei, Taiwan

- 2000 - 2009
- 15 patients with severe pulmonary HT with defects
  One patients – HL transplantation – mortality
  3 refuse operation, 3 observation
- 8 patients (age 26±9 years)- Pulmonary artery banding

- Additional PAB in 4
- 3 – corrective surgery

International Journal of Cardiology 2011;151: 313–317

Alternative approach for selected severe pulmonary hypertension of congenital heart defect without initial correction — Palliative surgical treatment ☆

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Pediatric Cardiovascular Surgery, Pediatric Cardiology, National Taiwan University Hospital, Taipei, Taiwan

	Age	Dx	AA	sPA	mPA	sPA/ sBP	mPA/ mBP	Estimated PVR (W)	Estimated Rp/Rs	SaO2
1	33	VSD		76	51	0.7	0.59	7.6	0.16	94
2	24	VSD		109	70	1.01	0.80	8.75	0.17	94
3	34	VSD		129	79	0.99	0.79	26.9	0.67	88
4	19	VSD		134	92	1.15	1.06	23.8	0.7	91
5	17	VSD		119	87	1.00	0.92	14.8	0.8	90
6	15	VSD	CoA*	117	68	1.00	0.89	11.7	N/A	93
7	37	VSD	IAA	121	83	1.19	1.10	37.7	N/A	87
8	33	VSD	TGA	115	71	1.15	0.86	16	0.38	75

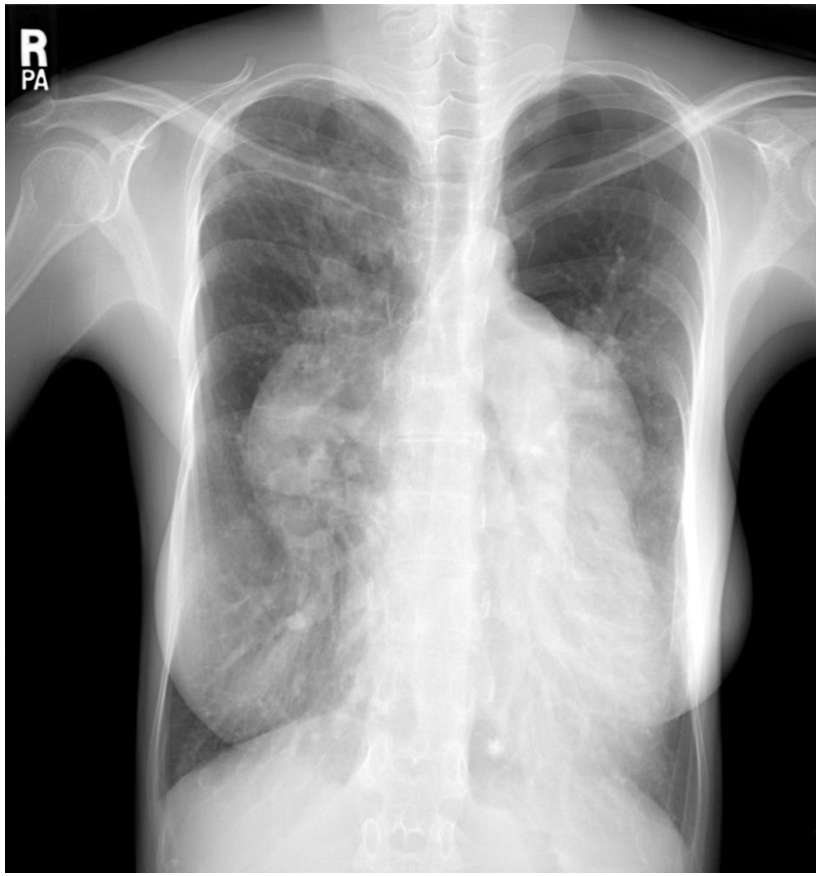
International Journal of Cardiology 2011;151: 313–317

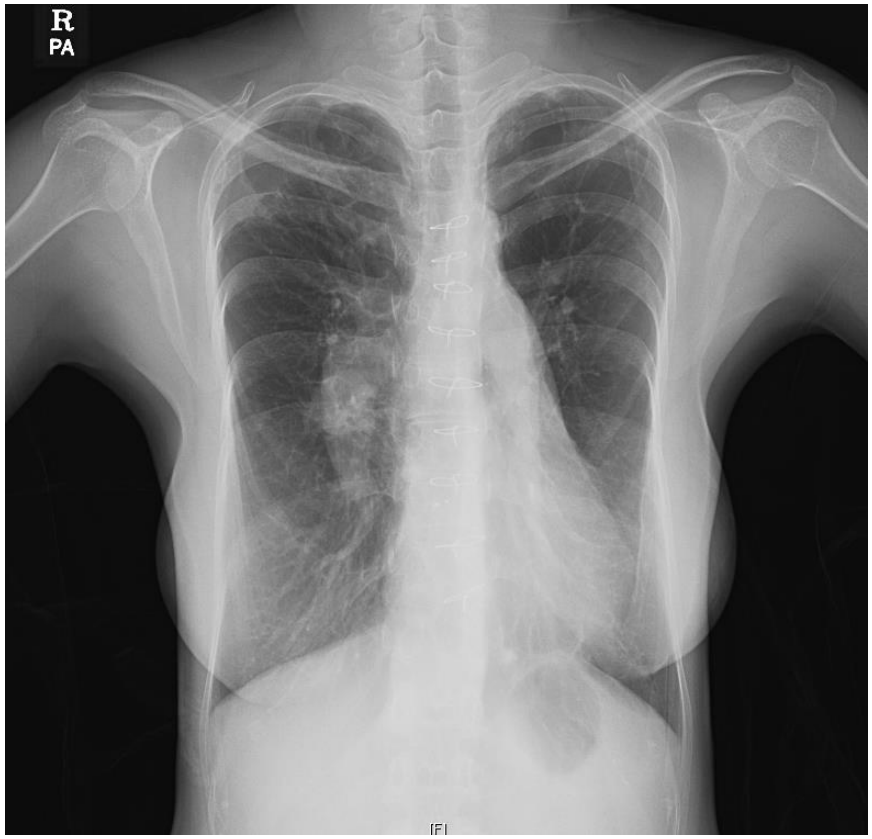
Staged repair : SMC experience

- F/38, VSD, severe PAH
  - NYHA class II, Chest pain after exercise
  - SaO2 90% at rest, SaO2 87% after exercise
- Op. (5Mo after PAB)
  - VSD closure, ASD creation (6㎜)
  - PA debanding, TR repair

Cath data	Pre-PAB	After PAB (6 Mo)	After VSD closure (16Mo after PAB)	After VSD closure (6 yrs after PAB)
Pr PA/Ao	117/118 → 101/117 (O2)	98/96 → 83/83 (O2)	44/138	33/118
mean PAP	96 → 65	57 → 48	26	18
Rp	6.9 →2.4	7.6 → 2.2	3.93	
Qp/Qs	2.9 → 5.7	2.6 → 5.7	1.93	1.2

Staged repair : SMC experience





Staged repair - two shunt lesions

Pediatr Cardiol
DOI 10.1007/s00246-009-9534-y

CASE REPORT

Successful Two-Stage Correction of Ventricular Septal Defect and Patent Ductus Arteriosus in a Patient with Fixed Pulmonary Hypertension

Aysenur Pac · Tugcin Bora Polat · Kerem Vural · Mustafa Pac

Received: 26 June 2009 / Accepted: 9 September 2009

- 6 yrs-old boy with VSD, PDA, NYHA Fc III
  - Initial Cath
    - PAP 92/69 (77), Ao 94/62(80)
    - Qp/Qs 1.2
    - Rp 10.8 Wood units

→ 6Mo of inhaled prostacyclin → Rp 7.2

→ Percutaneous closure of PDA → Rp 5.4

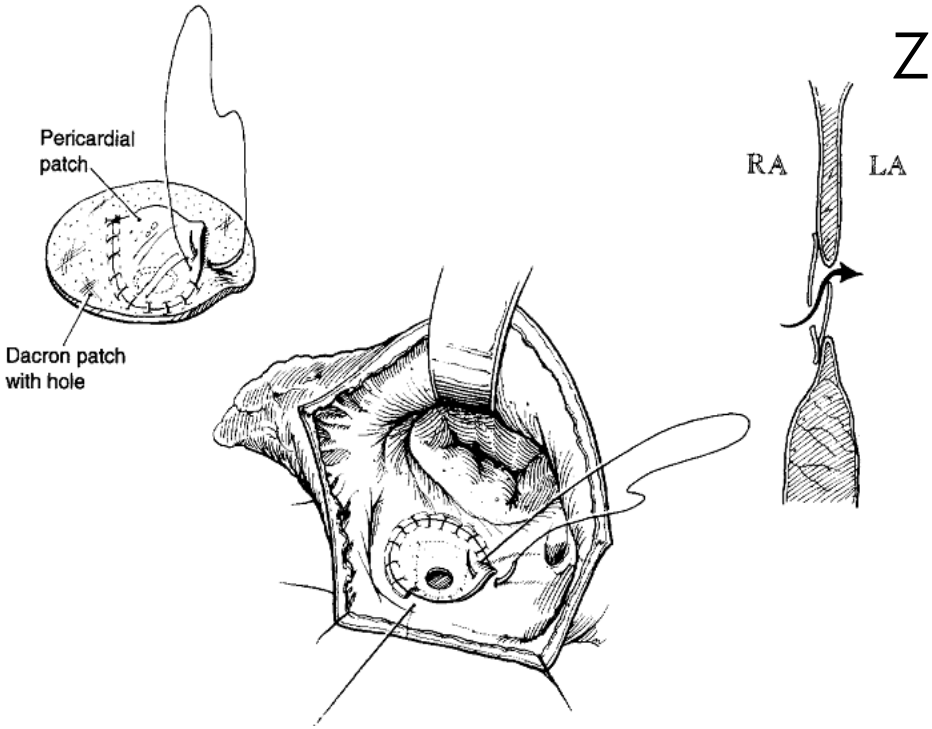
- 1 wk of iv prostacyclin

→ VSD closure → PAP 61㎜Hg

- 4Mo of iv prostacyclin
- NYHA Fc I at 1 yr F/U

One-way flap valve patch

- Unloading of the RV during severe PAH
- Relief of left heart hypovolemia
- First reported in 1959 (Bailey)

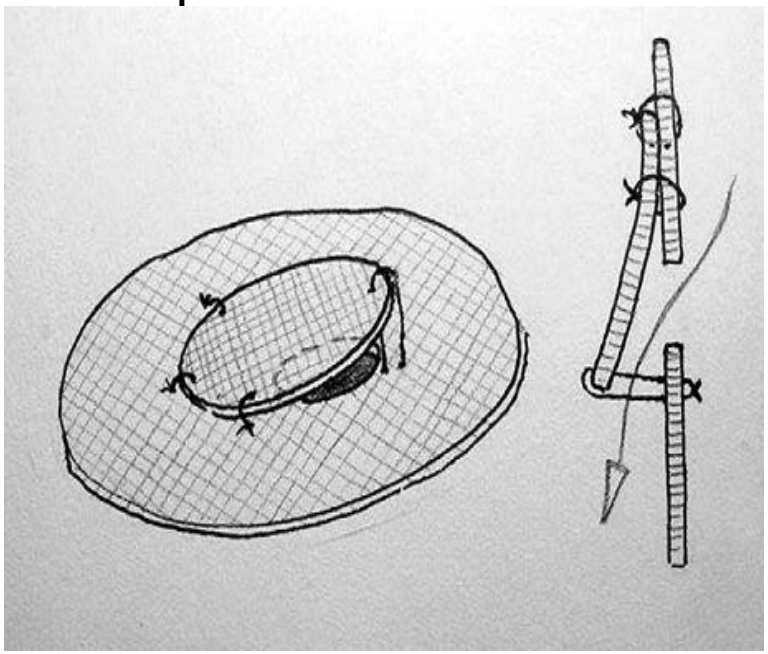


Zhou et al. Ann Thorac Surg 1995

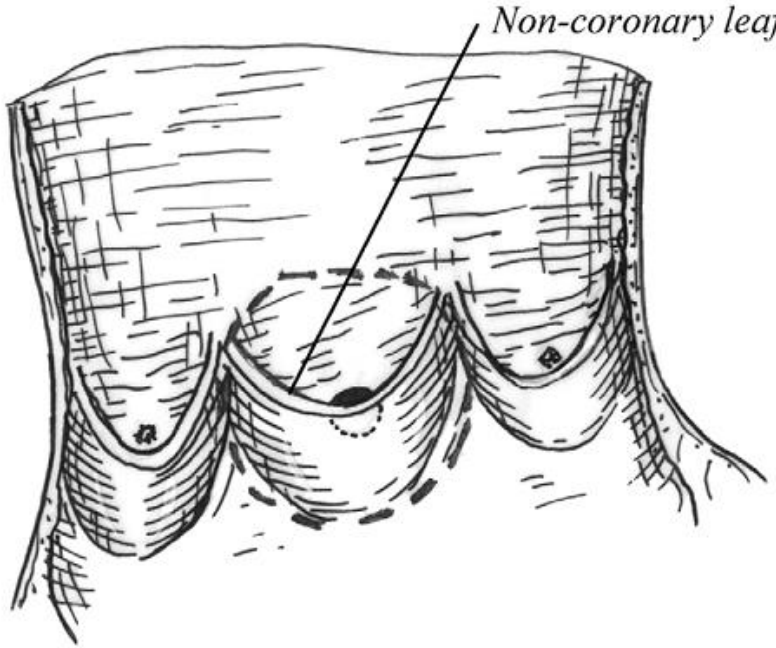
- 24 pts
  - mean age 15.8 yrs
  - 15 VSDs
  - Qp/Qs 1.2±0.5
  - Rp 16±7 Wood unit
- 2/24 early deaths (8.3%)
- Mean 1.1 yr F/U

One-way flap valve patch

- Novick et al. Ann Thorac Surg 2005
  - 91 pts
    - median age 4.0 yrs
    - PVR 10.5 ± 4.9 Wood unit
  - Fenestration (4~8 ㎜) : ½ of the expected aortic annulus
  - Overall early mortality : 7.7%
    - Simple VSD : 3.6%



One-way flap valve patch



- Zhang et al. Ann Thorac Surg 2007
- 27 pts
  - mean age 15.0 ± 5.6
  - PVR 15.2±3.8 Wood unit
- 2/27 early deaths

Table 2. The Standard for the Diameter of Fenestration

BSA	Preoperative Sao_2	Size of the Fenestration
<1 m^2	>91%	4 mm
<1 m^2	<91%	6 mm
>1 m^2	>91%	6 mm
>1 m^2	<91%	8 mm

BSA = body surface area; Sao_2 = arterial oxygen saturation.

Table 3. Preoperative and Postoperative Hemodynamic Status

Characteristics	Before Repair	After Repair
Mean pulmonary artery pressure (mm Hg)	81 ± 12	68 ± 15[a]
Pulmonary to systemic pressure ratio	1.05 ± 0.10	0.78 ± 0.20[a]
Arterial oxygen saturation (%)	89 ± 1	95 ± 2[a]

[a] $p < 0.01$, versus preoperation.

Table 4. Postoperative Echocardiographic Assessment

Time	No Right to Left Shunt (Cases)	Right to Left Shunt (Cases)
Day 1	17	10
Day 7	23	3
Month 3	23	2

The unidirectional valve patch provides no benefits to early and long-term survival in patients with ventricular septal defect and severe pulmonary artery hypertension

Hui-Li Gan, MD, PhD, Jian-Qun Zhang, MD, Zhao-Guang Zhang, MD, Yi Luo, MD, Qi-Wen Zhou, MD, and Ping Bo, MD

J Thorac Cardiovasc Surg 2009

- 876 pts underwent VSD closure with severe PH
- 195 Unidirectional valve patch vs. 681 NVP

- Indication
  - ↓6.5 WU from baseline after O2 or NO
  - Unidirectional valve patch
    - Bidirectional shunt on EchoCG
    - PVR > 10 WU
    - PVRI > 6WU/m2
    - Qp/Qs < 1.5
    - Hb > 16.0

- Mean F/U period : 102.2 ±61.6 Mo

UVP vs. NVP

- No difference btw UVP and NVP with propensity score matching
  - Early deaths : 7(5.1%) vs. 9(6.5%)
  - sPAP : 49.7±29.6 vs. 52.2±28.1㎜Hg (P = 0.4724)
  - 6MWD, NYHA Fc

- Status of UVP
  - open and R-L shunt in all patients by intraoperative TEE
  - Closed in 118 patients by the 3rd POD
  - Closed in 121/125 survivors

- UVP provides no benefits to early and long-term survival when it is used to repair a VSD with severe PAH

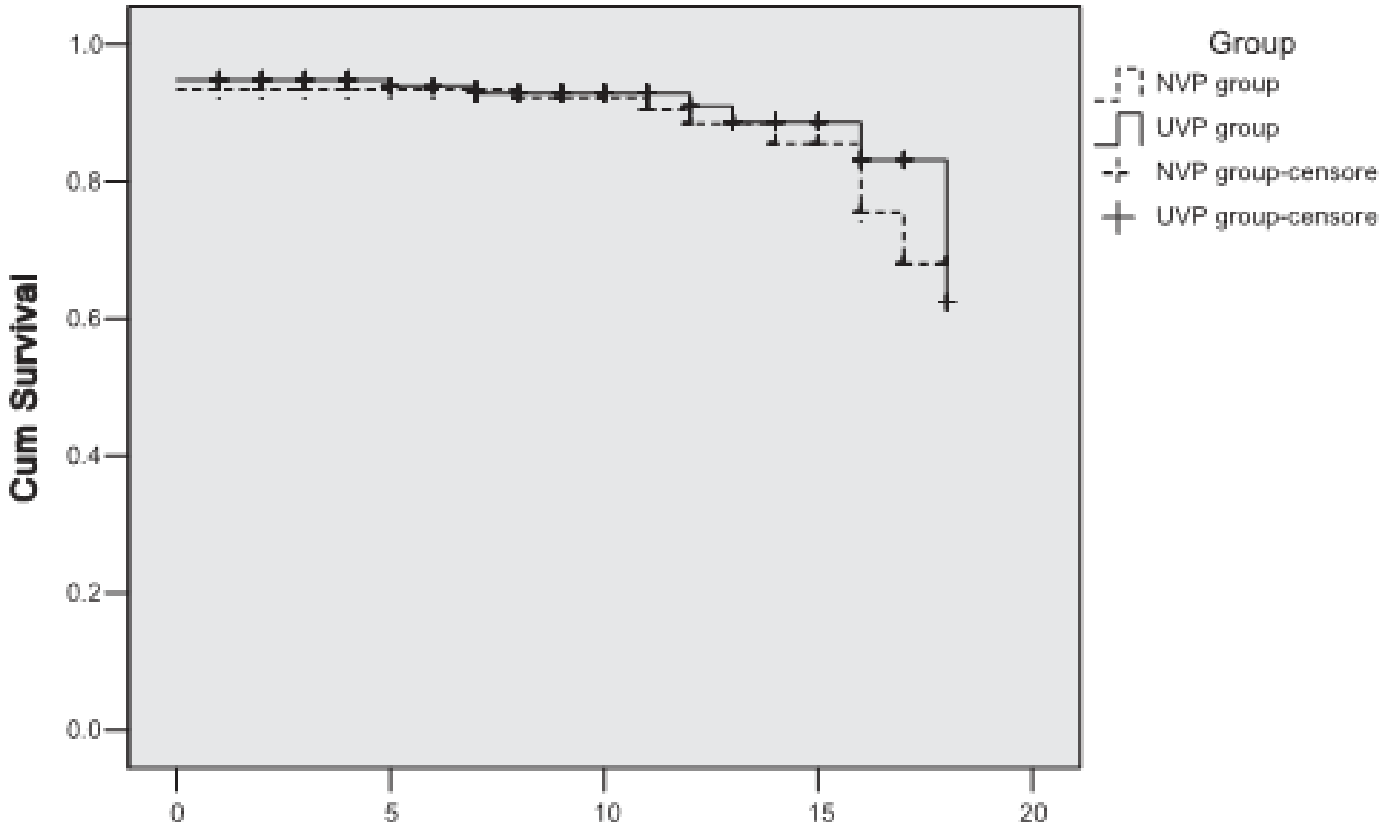
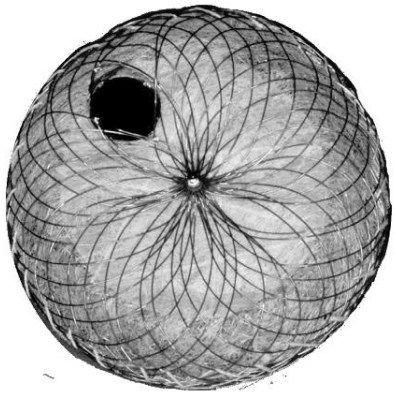


FIGURE 1. The Kaplan–Meier survival curve for the unidirectional valve patch *(UVP)* and nonvalve patch *(NVP)* groups (survival years).

Closure with a fenestration

- Bruch et al. J Interv Cardiol 2008
- Fenestration to allow for a minor to moderate shunt in both directions, depending on the compliance failure of the ventricles



- Entry Criteria.
  - ASD with L-R shunt of at least 50%
  - Mean PAP > 25㎜Hg
    or enlargement of the RV or ↑ RVEDP
- Exclusion Criteria
  - Hx of pulmonary or paradoxical embolism.
  - Fully developed Eisenmenger's syndrome with pressure equalization between right and left heart (Qp/Qs <1.2)

Closure with a fenestration

- 15 pts : mean age 66 (48-77)
- 5~8㎜ fenestration

- NYHA Fc improved
- All fenestrations stay open
- RVEDD ↓ , sPAP ↓ (55±13 to 43±12㎜Hg)
- LVEDD ↑
- Qp/Qs ↓ (2.59±1.19 to 1.38±0.42)
- PVR unchanged

Fenestrated patch closure of ASD
- SMC experience -

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Surgical Strategy in Patients with Atrial Septal Defect and Severe Pulmonary Hypertension

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Fenestrated patch closure of ASD
- SMC experience -

- 2004 ~ 2013
- No. of pts : 20
  - M : F = 4 : 16
  - Age : median 34 (23 ~ 58) yrs
  - Hb 14.1 (8.0~22.9) g/dL, SaO_2 95.4 (87.8~97.8) %

- Prior use of pulmonary vasodilator (n=6)
  - Sildenafil : 3
  - Bosentan : 4
    - +Beraprost : 2

- No preop. Medication (n=19)

Fenestrated patch closure of ASD - SMC experience -

- Hemodynamic data (baseline)
  - Systolic PAP : 82 (60~119) mmHg
  - P(PA/Ao) : 0.67 (0.46 ~ 0.94)
  - Qp/Qs : 2.1 (1.3 ~ 3.0)
  - Rp : 9.3 (3.9 ~ 16.7) WU

Associated diseases	No. of Pts
A. fib	3
MR (mild to moderate)	2
Mild Ebstein anomaly	1
Anomalous origin of single coronary artery	1
PR (mild to moderate)	1

Criteria for fenestrated closure of ASD - SMC experiences -

- Severe pulmonary HT with
  - Response to O_2, NO, test occlusion (↓ 20% of baseline)
  - Response to O_2, NO, test occlusion (↓ 10% of baseline) AND < mean PAP 40㎜Hg

- Dimopoulos K, Peset A, Gatzoulis A (Int J Cardiol 2008)
  - ↓ Mean PAP of > 10㎜Hg + resultant mean PAP ≤ 40㎜Hg
  - No evidence of ↓ cardiac output

Fenestrated patch closure of ASD - SMC experience -

- Operative findings
  - ASD size : 26 (20~46) ㎜
  - Fenestration size : 6 (5~8) ㎜

Concomitant procedures		No. of pts
TR repair	Ring annuloplasty	11
	DeVega	6
Maze op.		3
MAP (Wooler type)		2
PV commissuroplasty		1
MPA translocation		1

Fenestrated patch closure of ASD – SMC experience -

- Extubation at postop. 12 (6~45) hrs
  - Postop. iNO use : 3 pts
- No early deaths
- Complication
  - Bleeding control : 2
  - Pericardiocentesis d/t PE : 1
- Discharge at postop. 10 (6~18) days
  - Medication on discharge
    - All patients had pulmonary vasodilator except 3
      - Sildenafil 17/+bosentan 4/+beraprost 1
    - All patients had anticoagulant
      - Warfarin 15 / ASA 9

Fenestrated patch closure of ASD – SMC experience -

- Follow up duration
  - 51 (7 ~ 98) Months
- No late death
- Long-term Cx
  - Pericardiectomy for constrictive pericarditis 1
  - Window formation for pericardial effusion 1
  - Device closure of fenestration btw Ao and RA 1

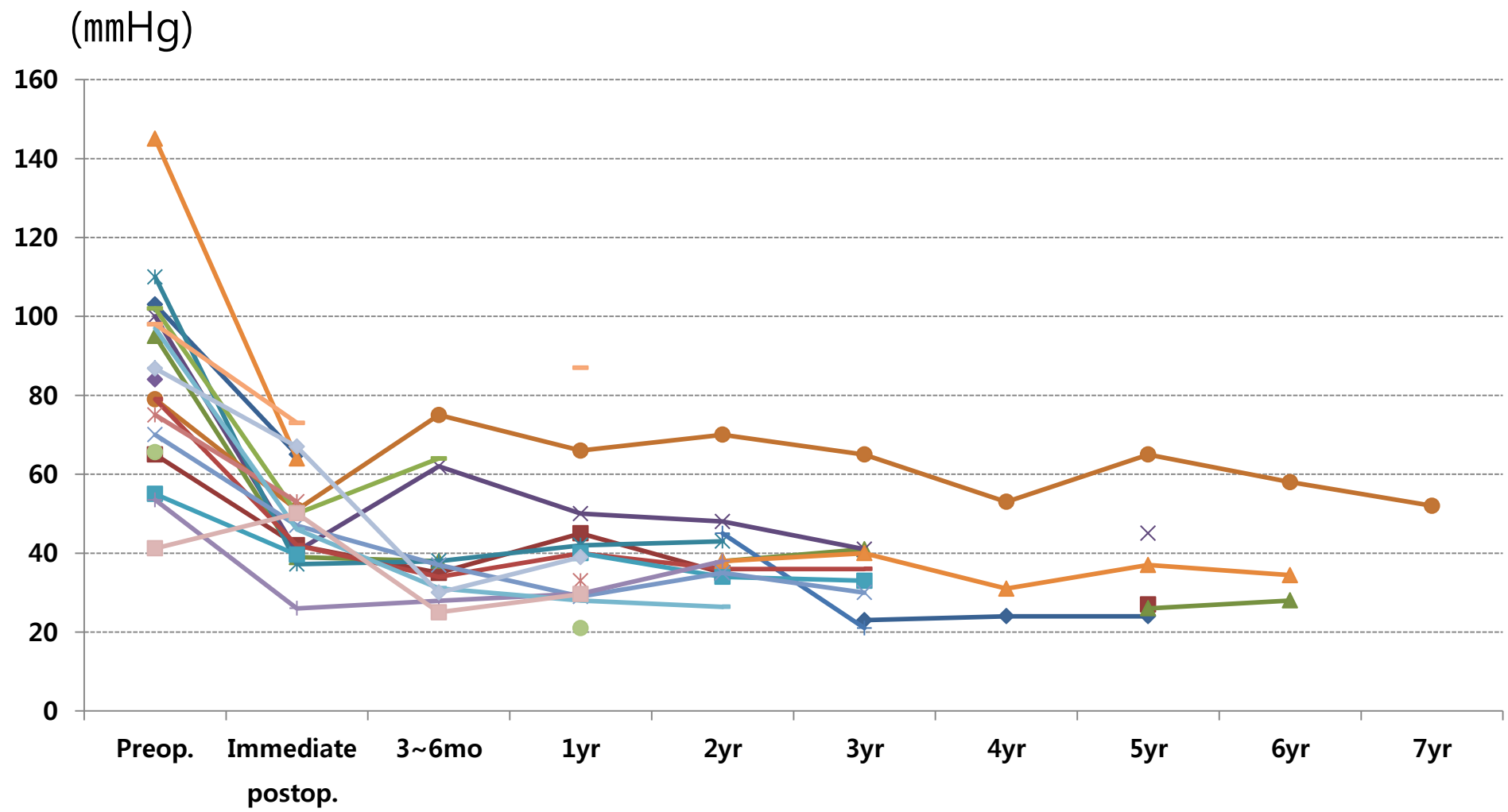
Fenestrated patch closure of ASD – SMC experience -

- Status of fenestration (by EchoCG)

Before discharge (# of pts)		Last F/U (# of pts)	
L → R	15	L → R	14
L ← R	1	L ← R	0
L ↔ R	3	L ↔ R	2
Invisible	1	Invisible*	4

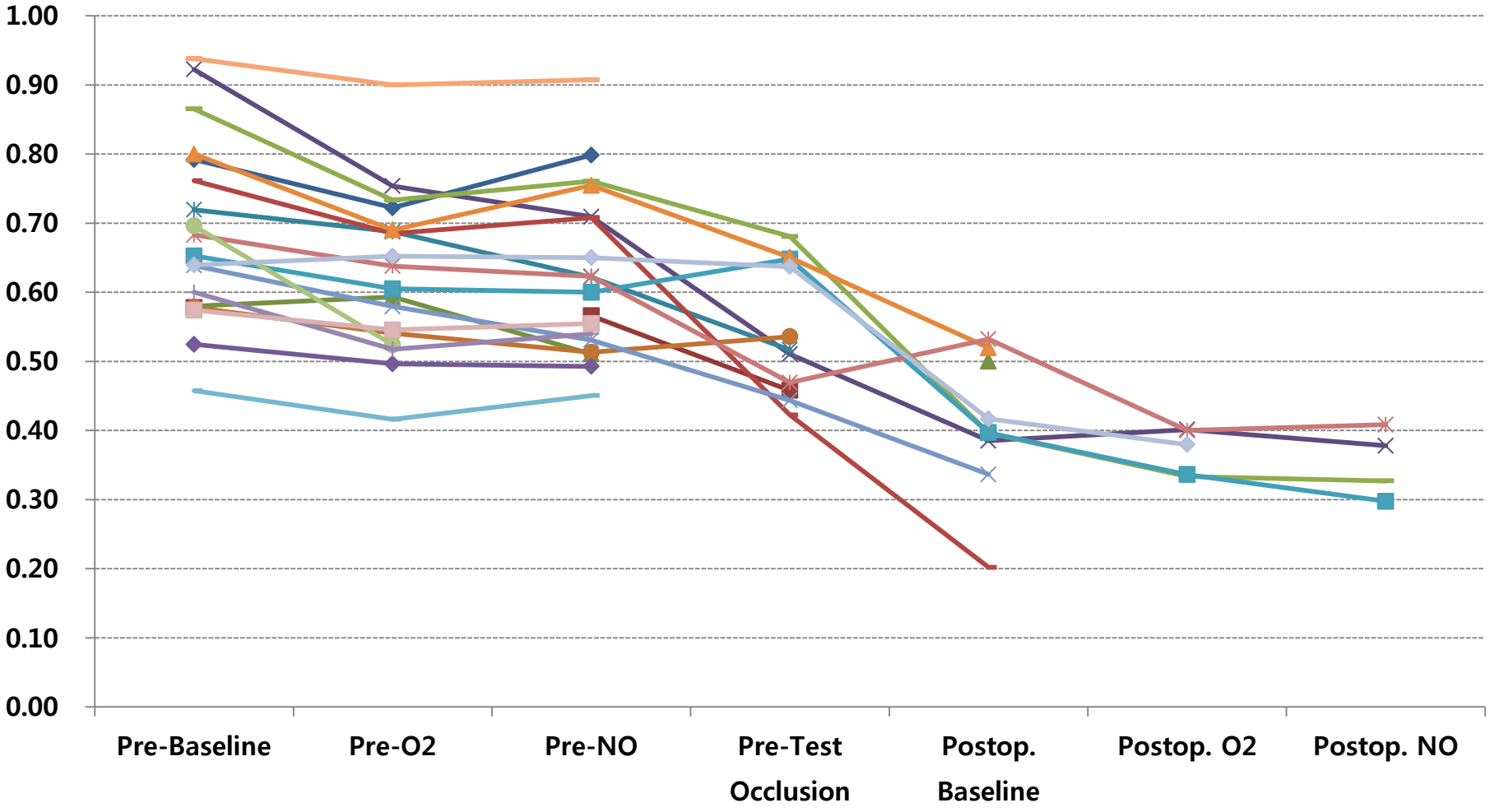
- Current medication
  - Pulmonary vasodilator 8
    - Sildenafil 5
    - Beraprost 1
    - Bosentan 1
    - Sildenafil + Bosentan + beraprost 1
  - ASA 6/Warfarin 2

Change of peak TR gradient by EchoCG

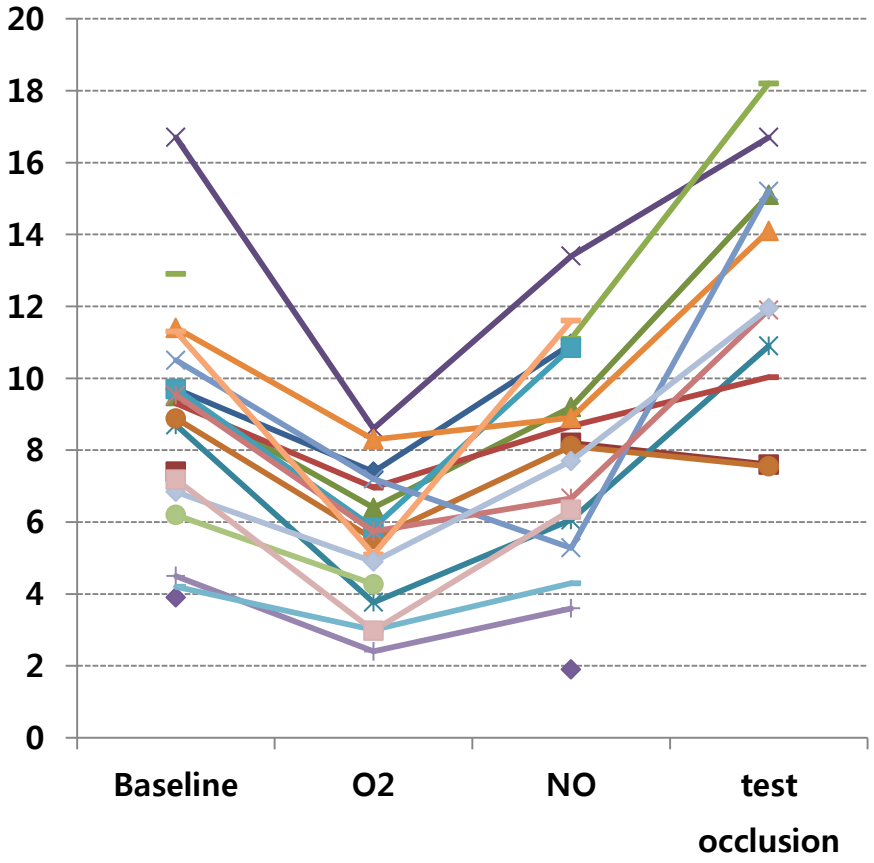


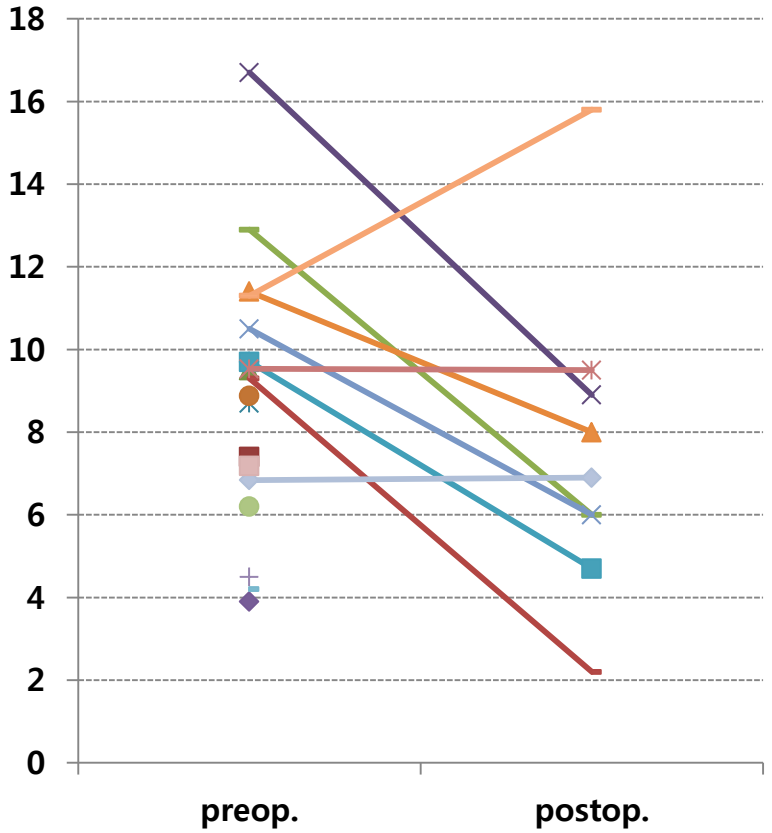
TR gradient could not be measured in 3 patients postoperatively, because of no TR flow

Change of Pressure ratio (PA/Ao)

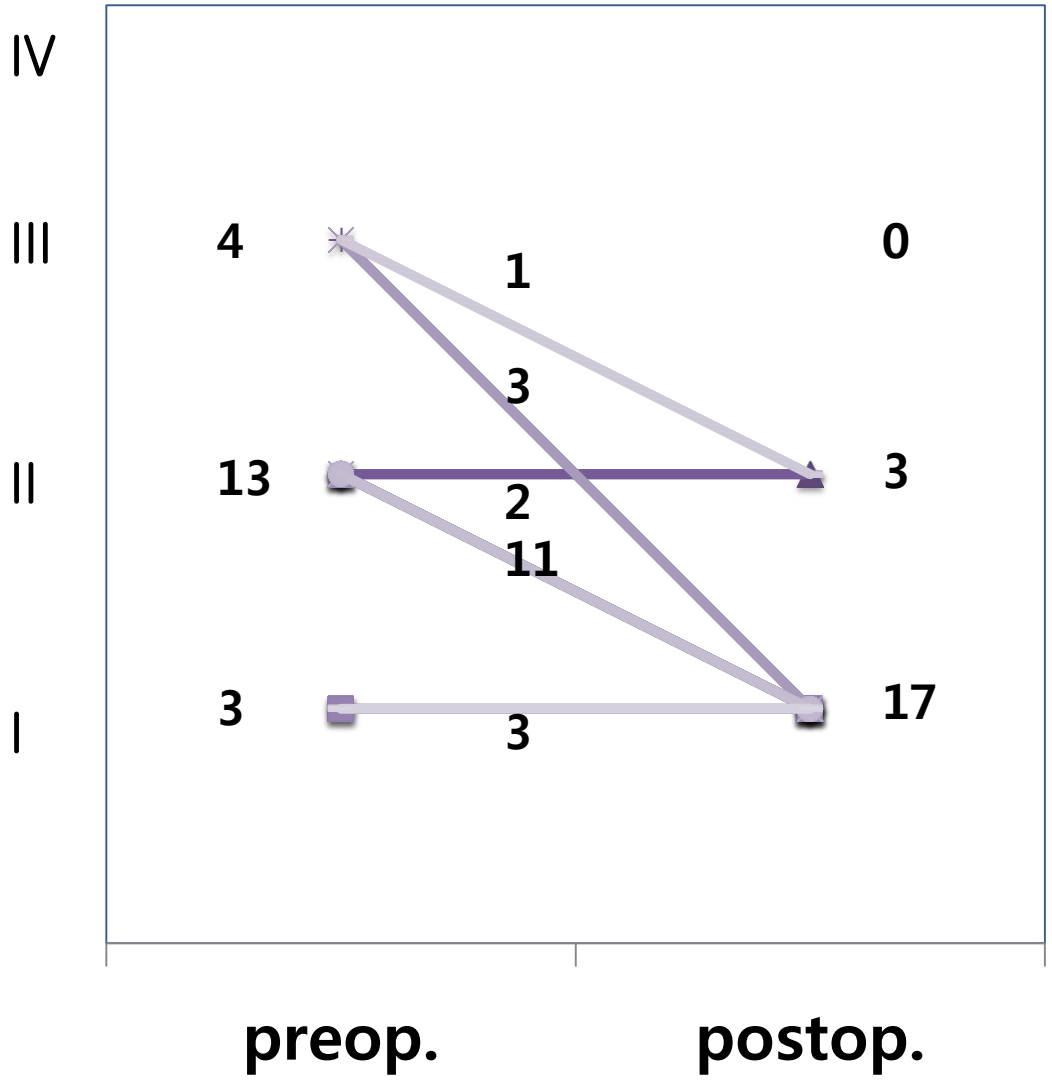


Change of PVR

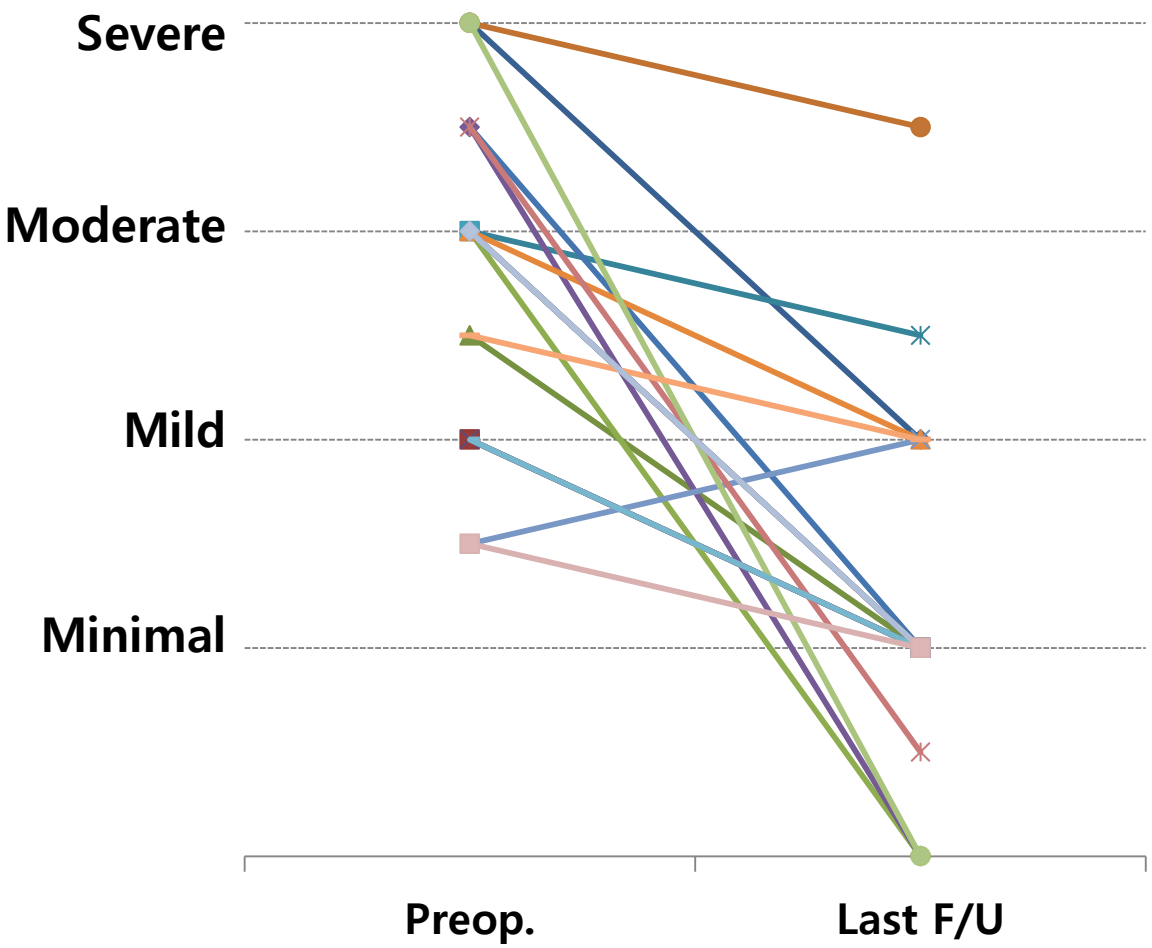




Change of Functional Class (NYHA)

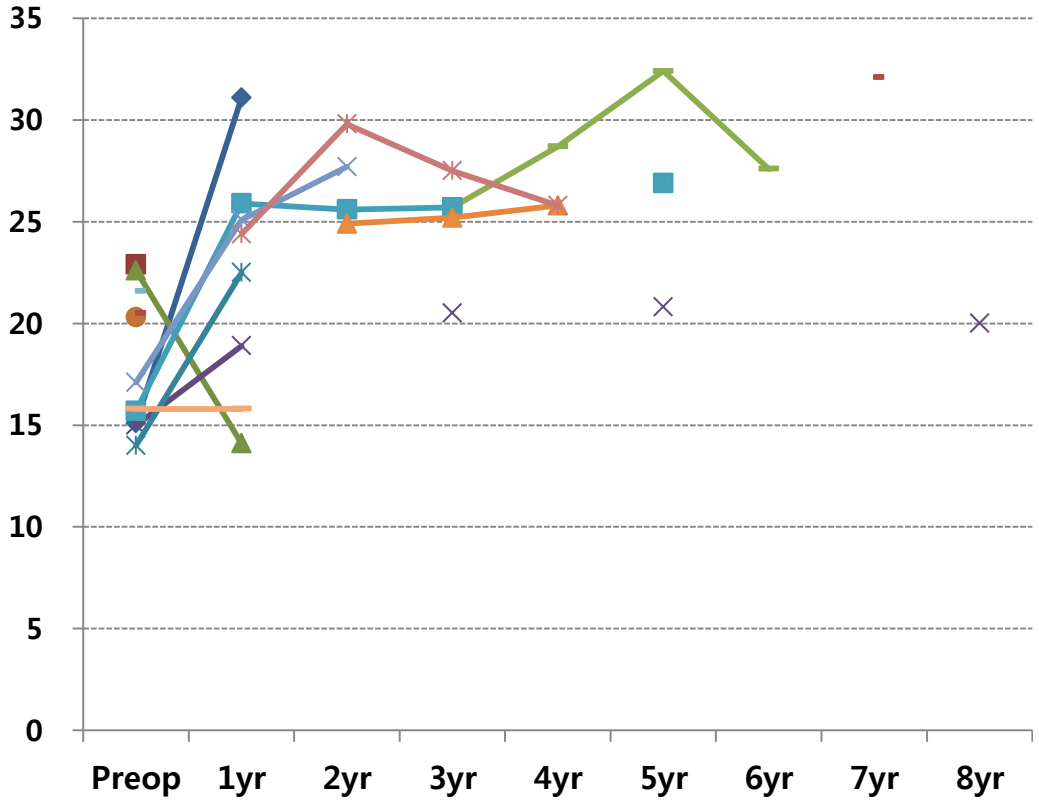


Change of TR grade

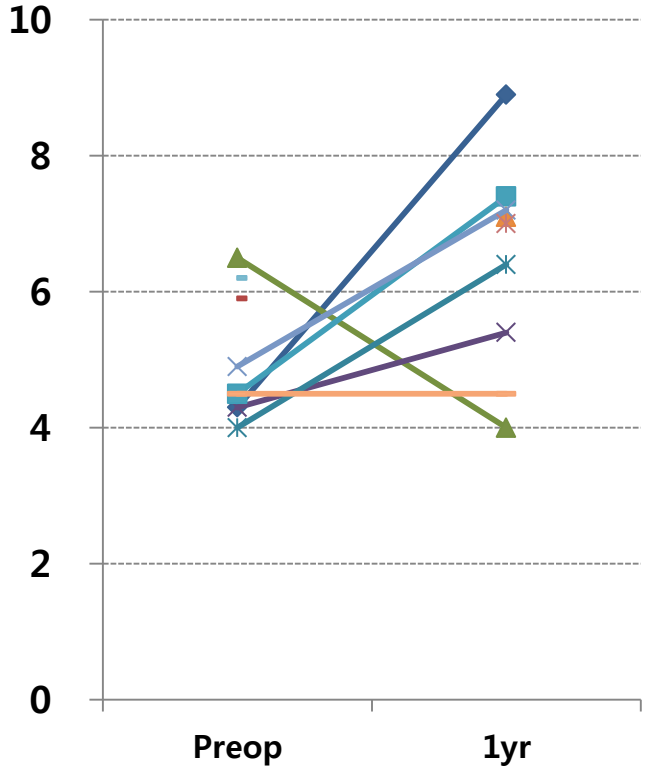


Change of Exercise Capacity

VO2 max (mL/kg/min)



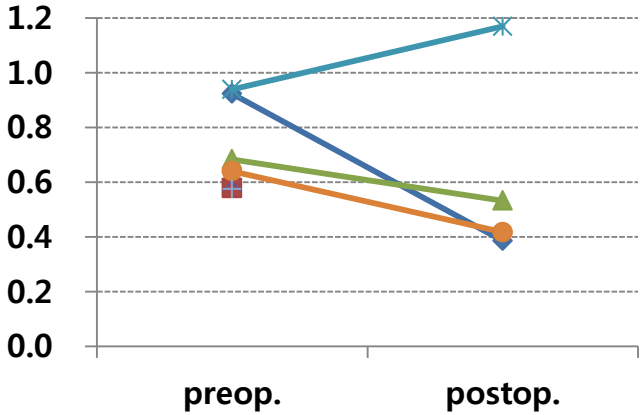
METs

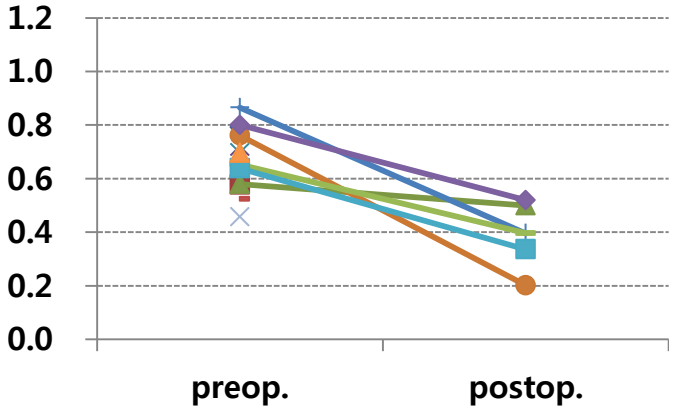


Treat-and-Repair Group (n=7)

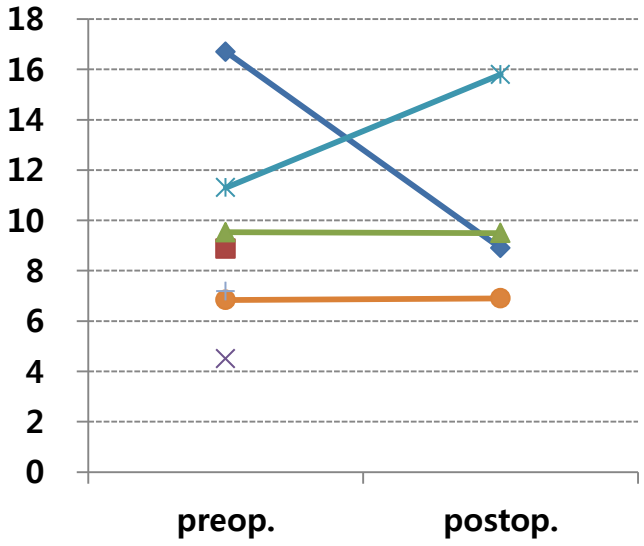
Repair-and-Treat Group (n=13)

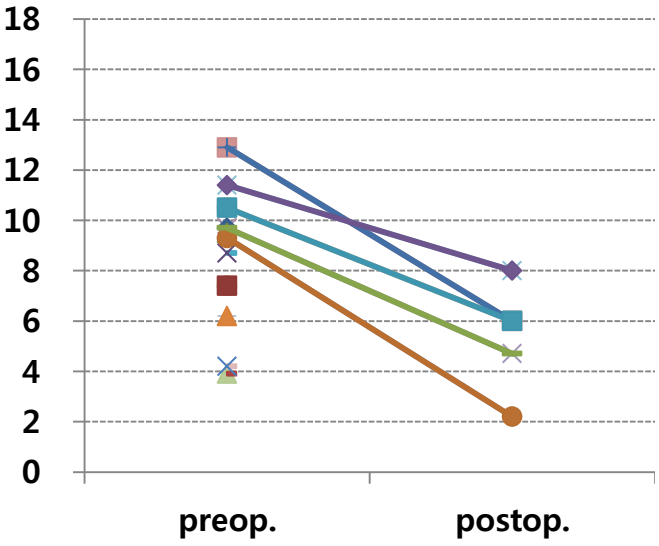
Pr(PA/Ao)





Rp (WU)





Summary

- No solid evidence of which one is better
  - T & R vs. R & T
- Indication of surgery in ACHD patients with severe PAH is still controversial
- Careful preop. evaluation is very important
- Indications for surgery can be extended with the aid of advanced medical therapies
- Fenestrated patch closure for ASD, one-way valve patch closure of VSD may be helpful in borderline patients, although longer term follow-up is needed