ASD with Pulmonary Hypertension Debate

강이석 삼성서울병원 소아청소년과

- 32 Year-old Male
- Large (=28 mm) central secundum ASD with severe PH
- Aorta 120/70(90) mmHg, MPA 95/50(70) mmHg, Rp= 8.2 WU/m2, Rp/Rs=0.33, normal sinus rhythm

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1	PAP with O ₂	PAP Test occ.	Qp/Qs	Rp basal	Rp with O ₂	Rp Test occ	Rp/Rs
28	88/20 (38)	NA	1.1	12.2	2.6	NA	0.76
2 6	88/26 (48)	81/30 (47)	2.0	12.9	8.7	16	0.29
26	75/25 (43)	81/27 (46)	Usu	 Usually much higher PVR in patient with large ASD and PAP of 95/50 mmHg & normal BP 			
6	69/33 (48)	67/34 (47)					
26	73/24 (44)	59/23 (36)	• Nee	ed for	compre	hensive e	evaluat
<u>.</u> 9	81/28 (48)	61/23 (39)	≻No du	No data for CXR, PaO2 or SpO2 at during exercise, Op/Os. vasodilator			
			te	st occlu	usion		

- Early symptomatic improvement does not guarantee long term outcome. (O'Donnell C, 2010)
- Risk of late rise of PAP or high PVR after ASD closure
- Long-term prognosis, not just immediate improvement

Closure guidelines ?

- Old study, incomplete or inadequate preoperative evaluation data
- Kirklin and Barratt-Boyes (Textbook, 1993) noted PVR > 6 U*M2 as major risk factor for surgical closure and >12 U*M2 predicted pulmonary vascular disease to be 'irreversible'.
- * "Modest elevation of PVR" and the presence of L-to-R shunt (Qp:Qs
 > 1.5:1) are reassuring. (O'Donnell C,2010)
- New guidelines in the era of advanced PH therapy; not yet
- Only a few reports of successful closure of ASD, surgically or non-surgically, in patients with PH, but no long term data
- D'als verse la su afita

"Treatment & repair"

- Surgical or trans-catheter closure of ASD without fenestration
- Surgical or trans-catheter closure of ASD with fenestration

cauncin & nepan

- latively safe; at least no bid aggravation
- pect remodelling of Imonary vasculature & crease of PVR
- Only a few anecdotal reports

Cons

- If the patient is in operation
 - May delay repair
 - May increase L-R shunt & aggravate PH

ASD (pre-TV) with PH vs VSD/PDA with PH

Less risk of defect closure, esp. with fenestration

Those who underwent surgical correction and had a preoperative PVR between 9 and 14 Wood units/M² showed no signs of disease progression and those between 7 and 9 Wood units/M² improved. (Steele et al. 1987)

- s invasive
- pid recovery
- s atrial arrhythmia
- s expensive
- orter hospital stay

Cons

- Unable to correct associated problem such as significant TR
- Difficult to adjust fenestration size
- Uncertain for long-term patency or keeping the initial size
 - Only a few case reports about long-tern patency
 - Balloon-dilated fenestrations in the Amp device tend to close spontaneously duri (Kretschmar O, 2010)
- Unknown risk for thrombo-embolise
 - Patch with punch hole vs device with fenestration

Sical Clobale with I chestianon

- repair associated blems
- adjust fenestration
- ter patency of the estration?

Cons

- Surgical risk
- Higher risk of atrial arrhyth
- More expensive
- Longer hospital stay

- July 2004 ~ June 2009
- M : F = 5 : 13 (total 18 patients)
- Age : median 31.3 yrs (3.5 Mo ~ 57.6 yrs)
- Hemodynamic data (baseline)
 - Systolic PAP : 82 (58~119) mmHg
- P(PA/Ao) : 0.70 (0.52 ~ 1.02)
- Qp/Qs : 2.1 (1.1 ~ 2.7)
- Rp : 9.5 (3.9 ~ 16.7) Wood unit / M²
- **Op criteria**
 - PAP decrease ≥ 20% or PAP decrease ≥ 10% & Qp/Qs ≥ 1.5

- eration
- ASD size : 25 (14~35) mm
- enestration size : 6 (4~8) mm

Concomitant procedures		No. of pts (21/18)	
TR repair	Ring annuloplasty	10	
	DeVega or Kay	4	
Maze op.		2	
PDA ligation		1	
MR repair		1	
RPA angioplasty		1	
PV commissuroplasty		1	
MDA translastion		1	

- No early or late deaths
- No immediately problems
- **Complications during FU in 2 patients**
 - Constrictive pericarditis
 - Pericardial effusion
- No significant arrhythmia

Follow up duration

- 32.5 (9.0 ~ 59.7) Mo
- Status of the fenestration on echocardiogram

15

1

1

1

17

Before discharge

- ✓ L-R ✓ R-L
- ✓ Bidirectional
- ✓ Invisible
- Last F/U: patent in all
 ✓ L-R
 - Closed during pericardiectomy

SMC experience – NYHAFC



NYHA FO $III \rightarrow I$ $III \rightarrow I$ $II \rightarrow I$ $II \rightarrow I$ $I \rightarrow I$





0

P (mmHg)





ΟΓ

/u/M²)



- Surgical closure of ASD with fenestration in patients with ASD accompanied by borderline PH
 - Very low surgical risk even with concomitant procedures
 - Functional improvement in most cases
 - Decrease of PAP & PVR in most cases
 - Long-term patency of fenestration in all
 - No thrombo-embolic events during FU
 - No significant atrial arrhythmia
 - ⇒if carefully selected
 - Long-term FU is essential.

- Surgical closure of ASD with fenestration for carefully selected cases
- Transcatheter closure of ASD with fenestration
 - For carefully selected cases without any associated problems
 - need FU for long-term patency and size change of the fenestration
- Treatment & repair for really irreversible cases

If carefully selected"

- ASD closure with fenestration if
 - Not too high PVR < 15 (?) WU and</p>
 - 1) or 2)
 - 1) Positive response to pulmonary vasodilator or test occlusion
 - ✓ PAP decrease ≥ 20% or
 PAP decrease ≥ 10% & Qp/Qs ≥ 1.5
 - 2) Baseline Qp/Qs ≥ 2.0 without systemic desaturation
 - If not: "treat and follow the patients"

After Long-Term Sildenafil Therapy (III) (Kim YH, et al. 2010)

. Hemodynamic Data

	Initia	l Study	2 Years of Sildenafil Therapy			
e	Baseline	O ₂ (10 L/min)	Baseline	O ₂ (10 L/min)	Balloon Occlusi	
mm Hg	1	2	10	8	8	
mm Hg	1	2	10	8		
mm Hg	87/20, 55	85/20, 55	128/32, 75	110/34, 65	99/26, 5	
mm Hg	140/80, 100	140/83, 105	137/71, 98	131/75, 95	133/68, 9	
%	69	74	76	68	79	
,	75	80	86	89		
,	89	92	94	99	99	
nin/m²)	2.16	2.24	5.15	4.7		
nin/m ²)	2.49	2.72	2.98	1.75	2.83	
ratio	0.87	0.82	1.73	2.68		
VU · m²)	25.0	23.7	12.63	12.1		
VU · m²)	39.8	37.8	29.53	49.7	30.73	
VRI	0.63	0.63	0.43	0.24		