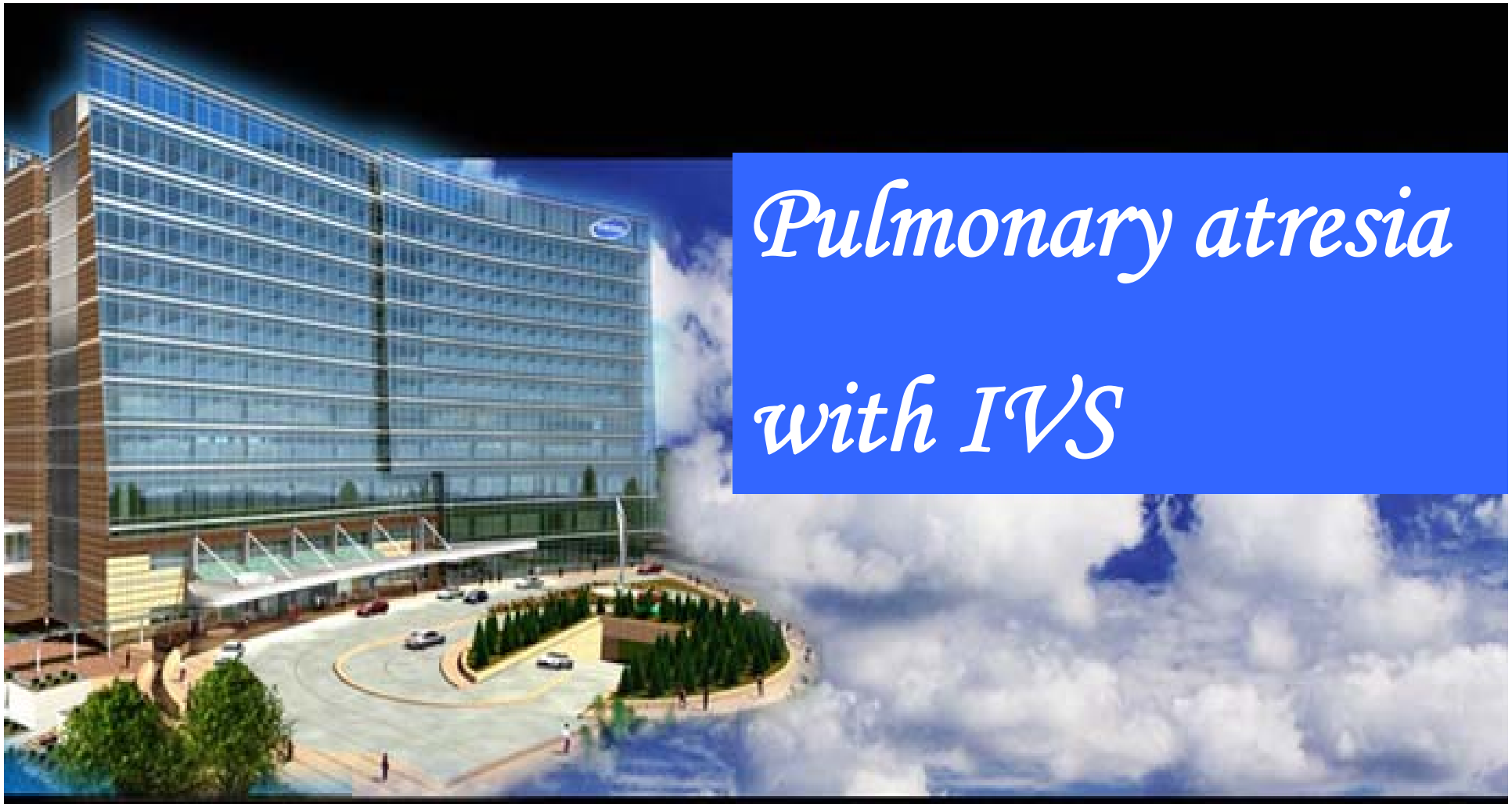


Pulmonary atresia with IVS

Tae-Gook Jun

Samsung Seoul Hospital

Sungkyunkwan University School of Medicine

Pulmonary atresia with intact ventricular septum

- Absence of communication between RV – PA
- Intact septum

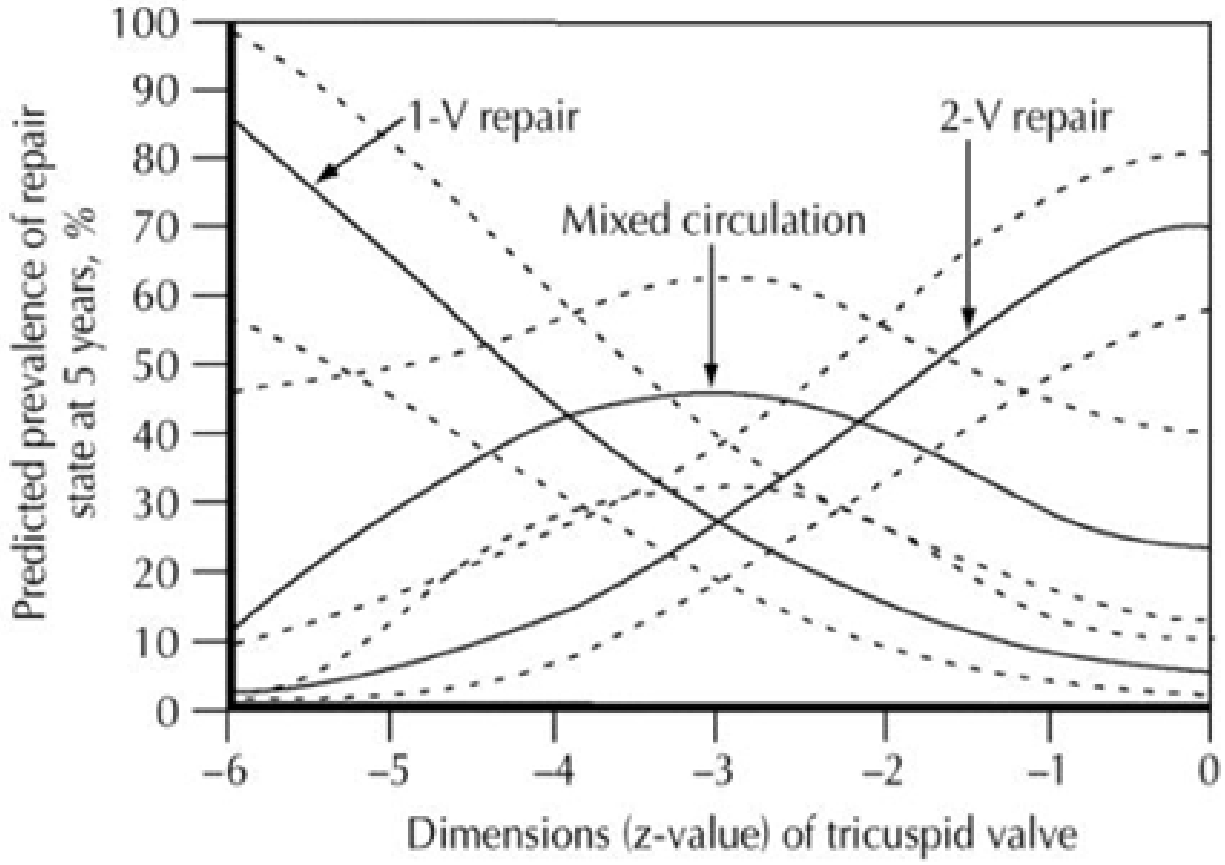
- Morphologically heterogeneous

- Varying degrees of RV and TV hypoplasia
- Aberrations of coronary circulation are common

Treatment strategy for PA IVS

- Initial palliation
- Additional procedure
- Definite repair
- Individualized plan for surgical management, based on the special morphology, is necessary.

Predicted prevalence 5 years after entry (CHSS : 31 institute)



J Thorac Cardiovasc Surg 1993;105:406-27

Determinants of mortality and type of repair in neonates with pulmonary atresia and intact ventricular septum

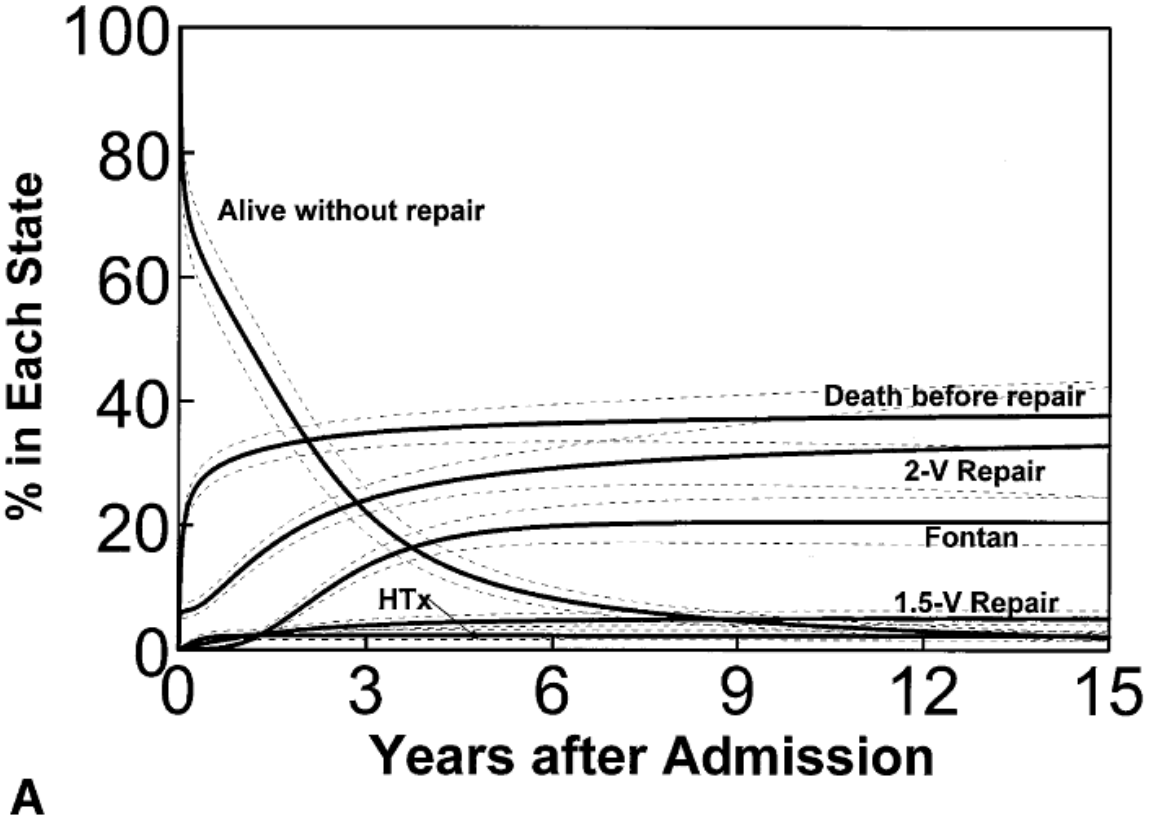
David A. Ashburn, MD[a,b]
Eugene H. Blackstone, MD[c]
Winfield J. Wells, MD[d]
Richard A. Jonas, MD[e]
Frank A. Pigula, MD[f]
Peter B. Manning, MD[g]
Gary K. Lofland, MD[h]
William G. Williams, MD[a]
Brian W. McCrindle, MD, MPH[i]
Members of the Congenital Heart Surgeons Society

- 1987 – 1997
- 408 neonates , 33 institutes
- Overall survival : 77% at 1months, 70% at 6 months
  60% at 5 years, 58% at 15 years
- In the current era, 85% of neonates are likely to reach a definitive surgical end point, with 2-ventricle repair achieved in an estimated 50%

J Thorac Cardiovasc Surg 2004;127:1000-8

Determinants of mortality and type of repair in neonates with pulmonary atresia and intact ventricular septum

Members of the Congenital Heart Surgeons Society



A

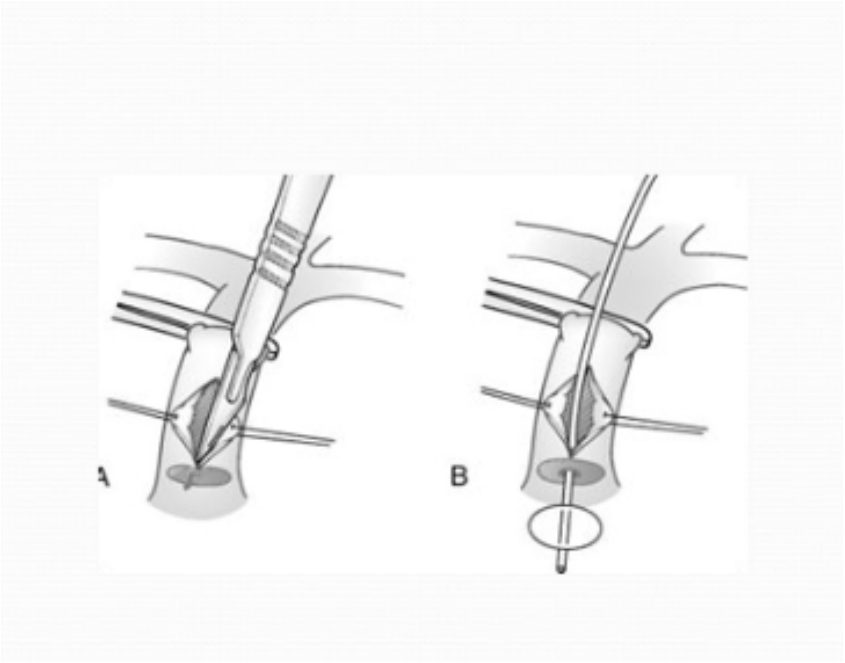
J Thorac Cardiovasc Surg 2004;127:1000-8

Initial palliation

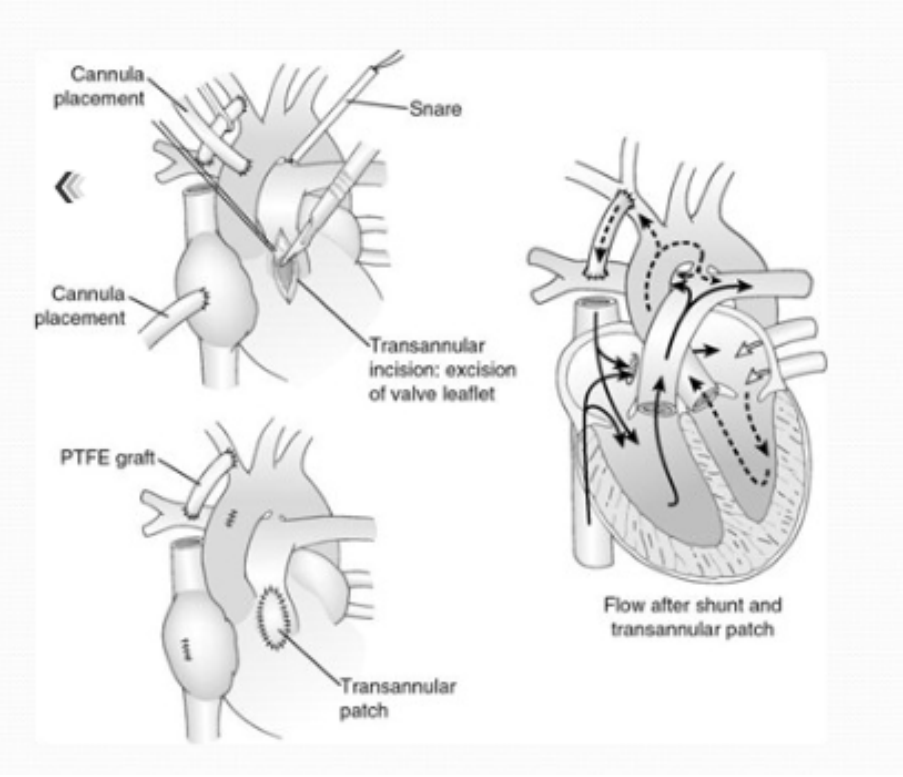
- Adequate RV decompression – 2 ventricle strategy
  - Transpulmonary valvotomy ± shunt
  - Outflow tract patch reconstruction ± shunt
  - Balloon valvuloplasty

- Systemic pulmonary shunt only - 1 ventricle strategy
  - RV dependent coronary circulation
  - Small RV with muscular atresia

Initial palliation



Transpulmonary valvotomy

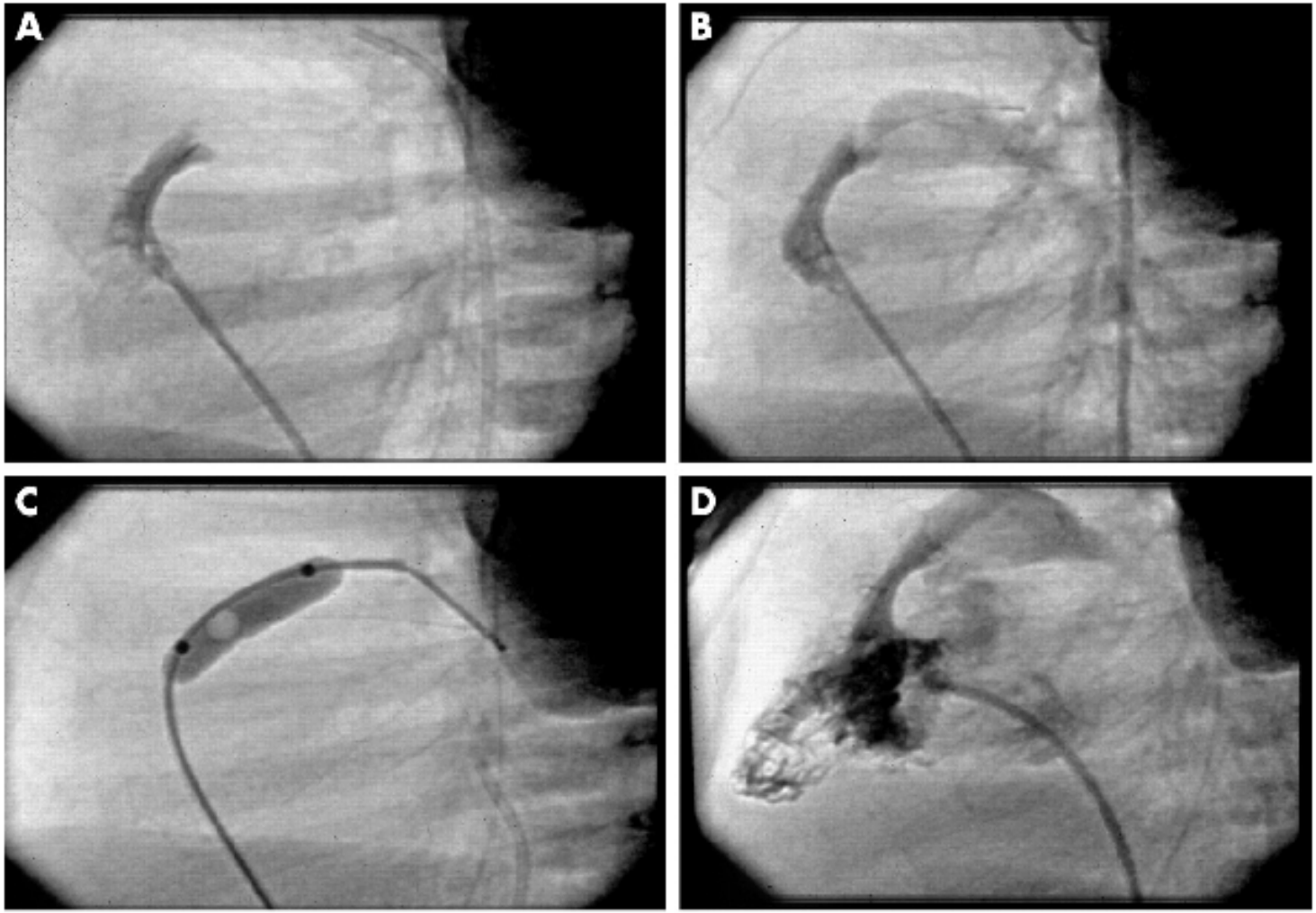


RVOT patch

RV decompression

Transpulmonary valvotomy ± shunt	**RVOT patch widening ± shunt**
Avoid CPB in neonate Less ventricular damage Less pulmonary regurgitation	No residual RVOTO Pulmonary regurgitation can augment RV growth
Residual RVOT obstruction Limitation of RV growth	Need CPB in neonate Heart failure

Catheter based balloon valvuloplasty



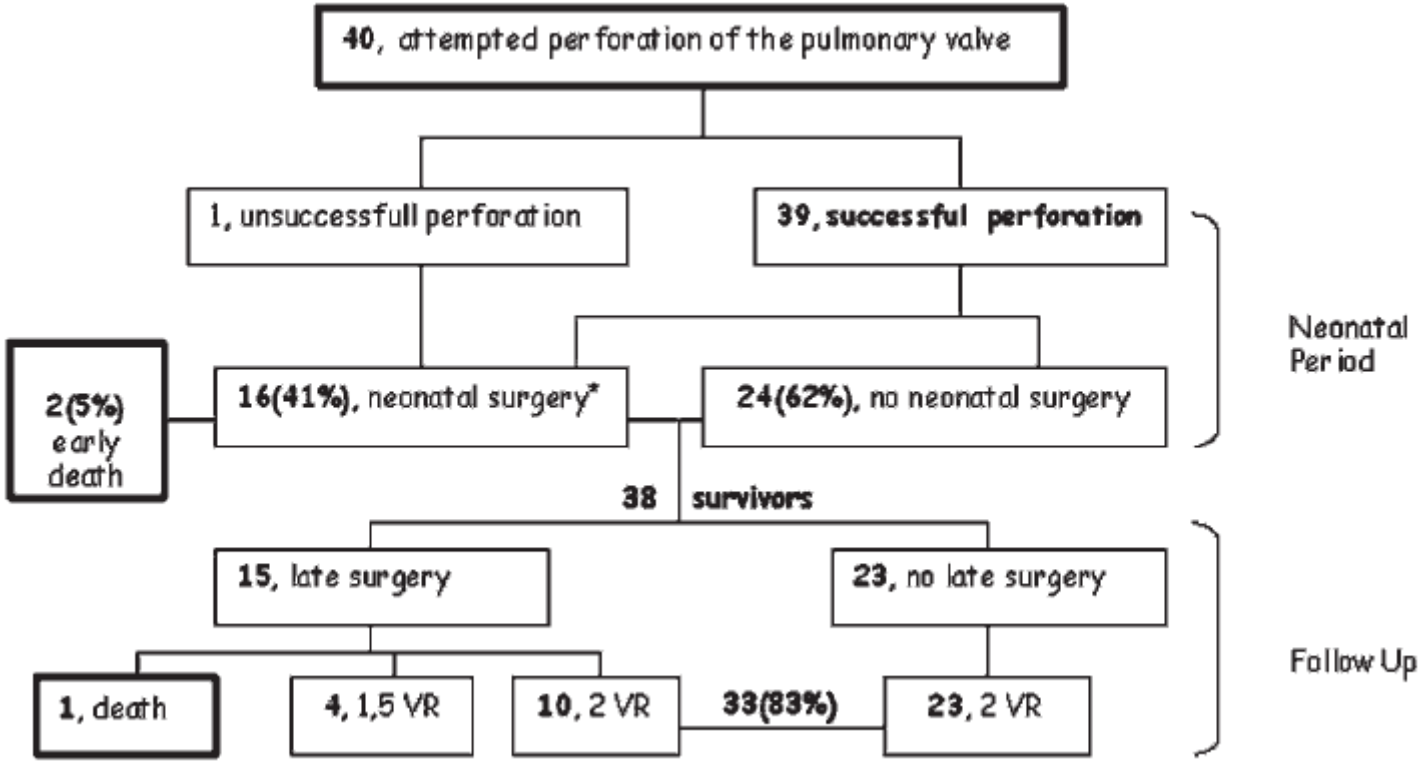
Catheter based balloon valvuloplasty

- Benefits
  - Avoid cardiopulmonary bypass in neonate
  - Delaying surgical intervention

- Limitations
  - Cannot enlarge pulmonary valve annulus
  - Difficult to anticipate pulmonary annulus growth
  - Cannot relieve subpulmonary muscular obstruction

Long-term results of catheter-based treatment of pulmonary atresia and intact ventricular septum

M Marasini,[1] P F Gorrieri,[1] G Tuo,[1,2] L Zannini,[1] P Guido,[1,3] M Pellegrini,[1] S Bondanza,[1] M G Calevo,[4] G Pongiglione[1]

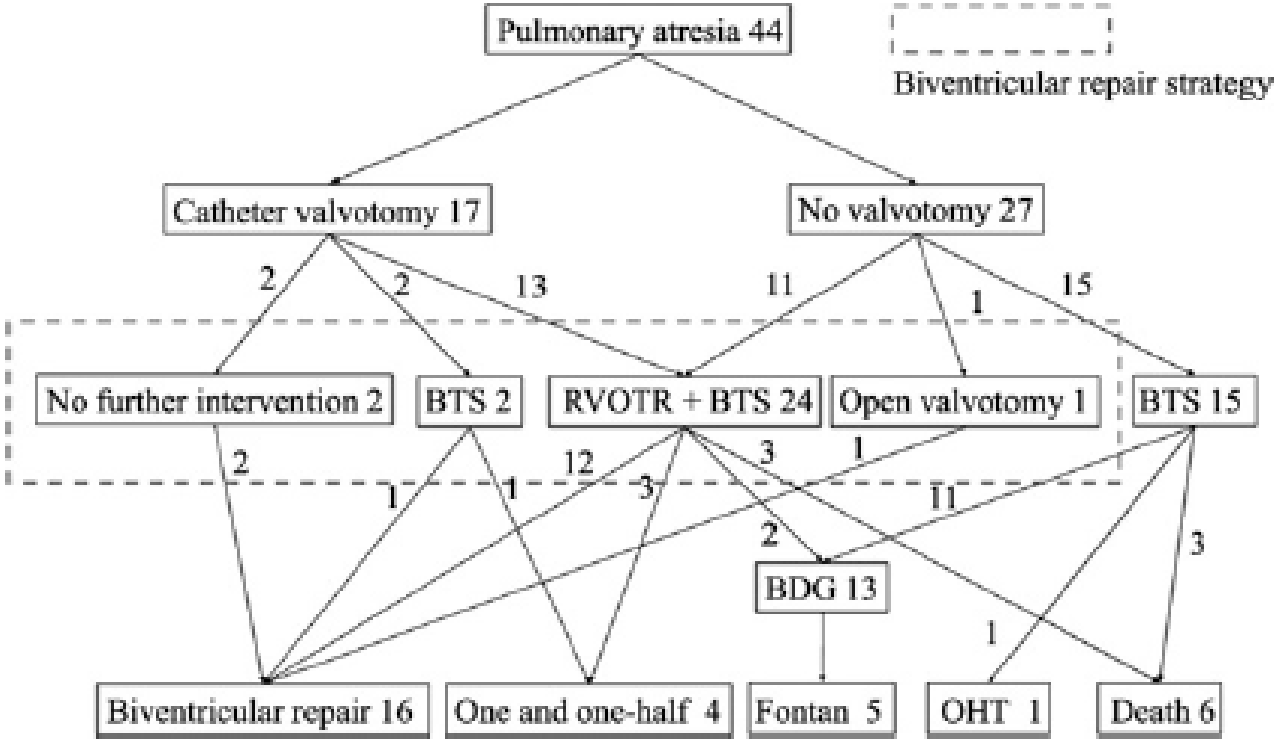


Heart 2009;95:1520–1524

Pulmonary Atresia With Intact Ventricular Septum: Limitations of Catheter-Based Intervention

Yasutaka Hirata, MD, Jonathan M. Chen, MD, Jan M. Quaegebeur, MD, William E. Hellenbrand, MD, and Ralph S. Mosca, MD

The Divisions of Pediatric Cardiac Surgery and Pediatric Cardiology, Columbia University College of Physicians and Surgeons, New York, New York



Ann Thorac Surg 2007;84:574–80

Effect of residual RVOT obstruction

Residual RV obstruction

- Hindrance to RV growth
- Decreased RV dimension and TV size
- Persistent right ventricular sinusoids

- Micro and macro coronary abnormalities

Early re-evaluation after the RV decompression and adequate additional palliative procedures are essential for the preparation for the definite repair

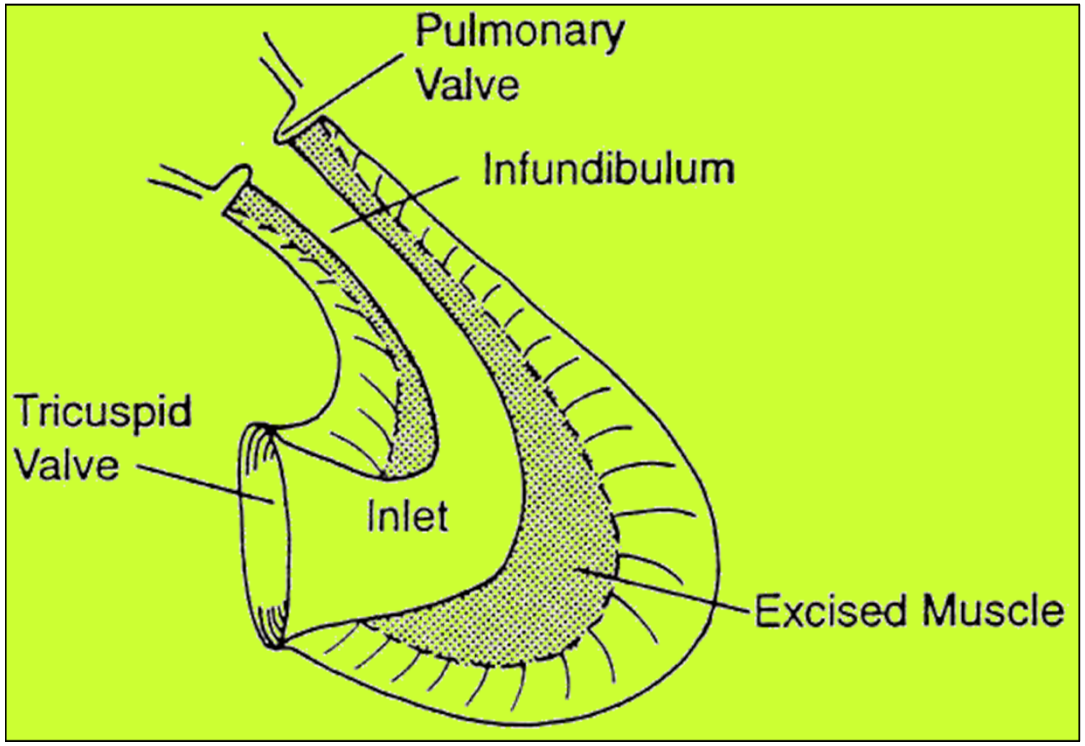
Secondary palliative surgery

- Repeated balloon vavuloplasty

- Preparation for bi-ventricular repair
  - RVOT reconstruction
  - RV overhaul procedure
  - Closure or adjustment of atrial communication

- Preparation for uni-ventricular repair
  - BCPS

Right ventricular overhaul



- TV valvotomy
- RV muscle resection
- PV valvotomy
- RVOT patch

Pawade A et al. J Cardiac Surg 1993;8:371-383

Staged Biventricular Repair of Pulmonary Atresia or Stenosis With Intact Ventricular Septum

Shunji Sano, MD, Kozo Ishino, MD, Masaaki Kawada MD, Emi Fujisawa, MD, Masahiro Kamada, MD, and Shin-ichi Ohtsuki, MD

Departments of Cardiovascular Surgery and Pediatrics, Okayama University Medical School, Okayama, Japan

- 1991 – 1999
- 25 patients (critical PS in 6)
- BAS in 21 patients

- Z value of T valve : - 1.4 (range, -4.4 – 2.3)
- Initial pulmonary valvotomy
- Repeated valvotomy or surgical palliation

Ann Thorac Surg 2000:70:1501-6

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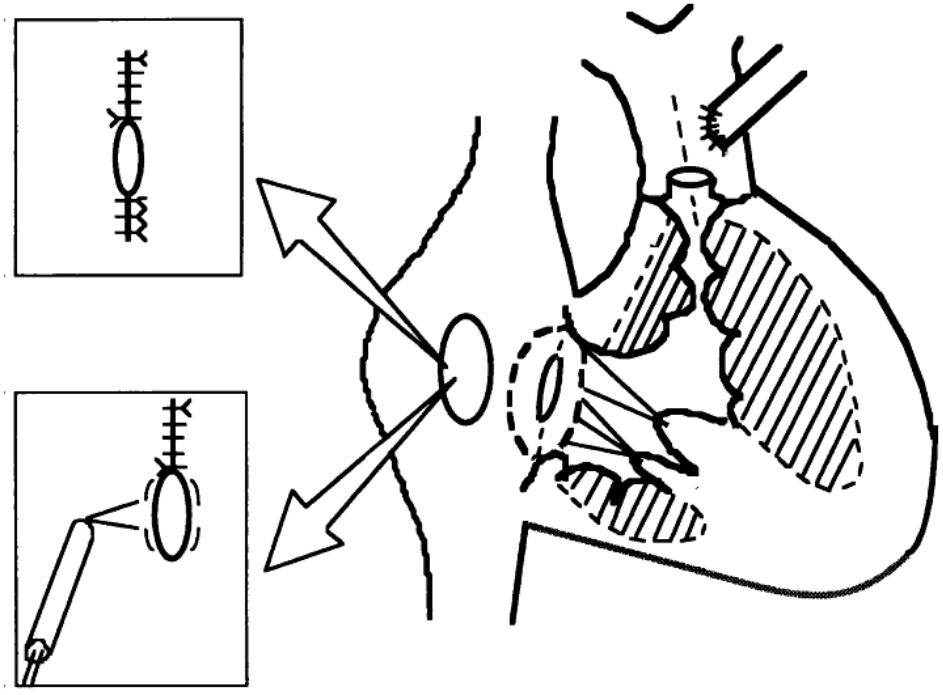

- Initial pulmonary valvotomy
- Overhaul
- Adjustment of interatrial communication

Ann Thorac Surg 2000:70:1501-6

Staged Biventricular Repair of Pulmonary Atresia or Stenosis With Intact Ventricular Septum

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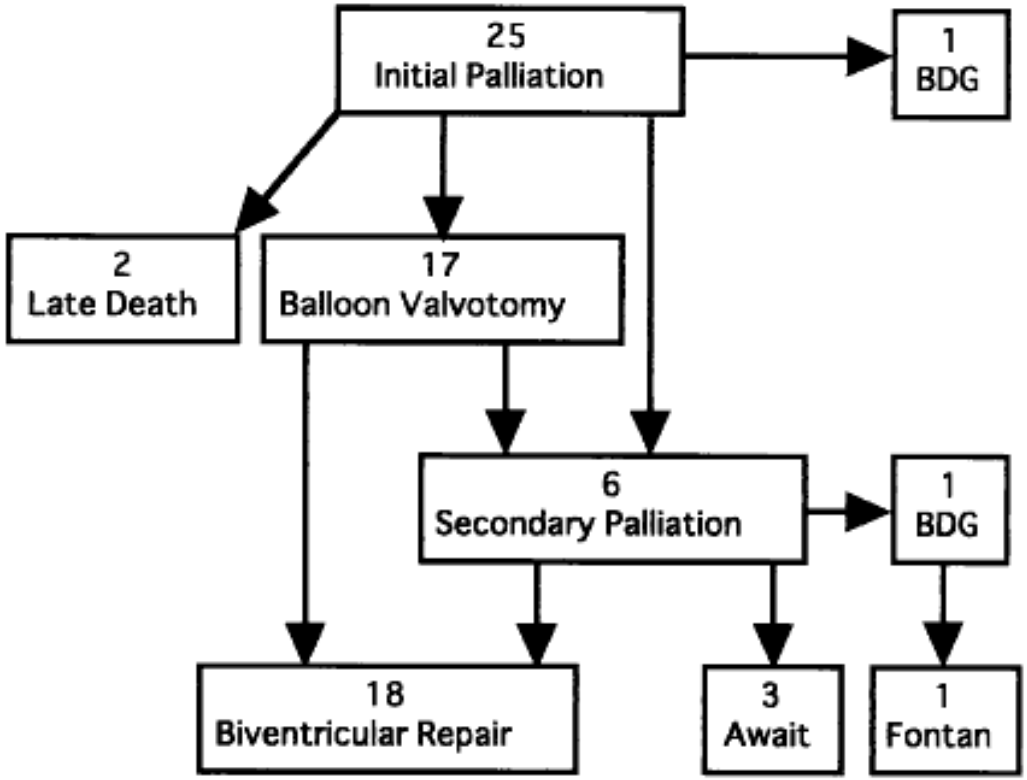

Ann Thorac Surg 2000:70:1501-6

Success and limitations of right ventricular sinus myectomy for pulmonary atresia with intact ventricular septum

Roosevelt Bryant, III, MD,[a] Edward R. Nowicki, MD,[a] Roger B. B. Mee, MB, ChB, FRACS,[b] Jeevanantham Rajeswaran, MSc,[c] Brian W. Duncan, MD,[d] Geoffrey L. Rosenthal, MD, PhD,[b] Uthara Mohan, MD,[d] Muhammad Mumtaz, MD,[b] and Eugene H. Blackstone, MD[a,c]

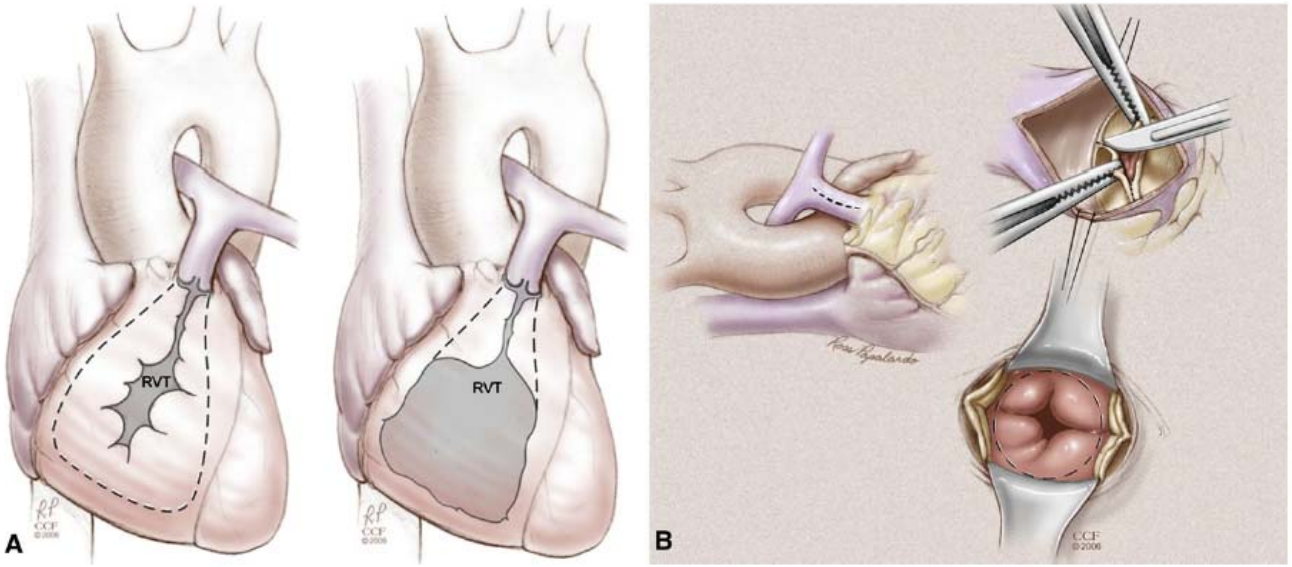

Figure 1. Right ventricular myectomy is performed by means of a combined transatrial/transpulmonary approach. A, On the *left*, the *dashed line* indicates the intended extent of right ventricular sinus resection. On the *right*, the figure represents the results of sinus resection through the tricuspid valve. B, On the *left*, an incision is made in the pulmonary trunk. At *top right*, pulmonary valvotomy is shown. This reveals infundibular muscle *(lower right)* that will be resected *(dashed line)* to complete the right ventricular myectomy. *RVT*, Right ventricular trabecular sinus.

J Thorac Cardiovasc Surg 2008;136:735-42

Success and limitations of right ventricular sinus myectomy for pulmonary atresia with intact ventricular septum

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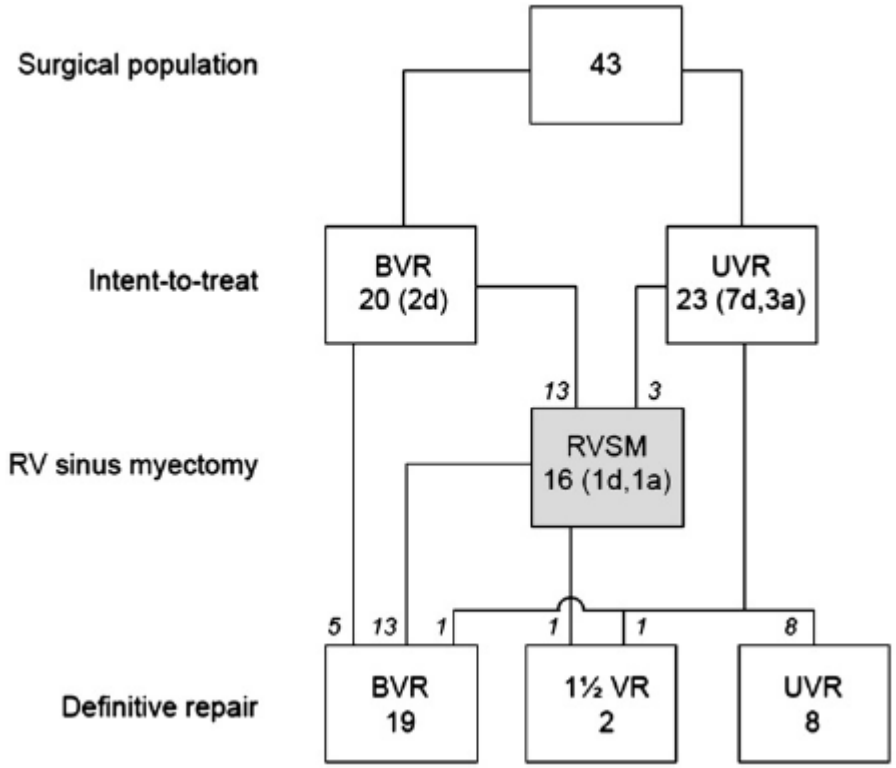

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- 13/16 biventricular repair
- Death: 3 (19%, 3/16)
- Survival rate: 94%, 85%(5year), 64%(10 year)

- Small tricuspid valve - poor result

J Thorac Cardiovasc Surg 2008;136:735-42

Definite repair

- Biventricular repair
- One and half repair
- Single ventricle repair
- Transplantation

Bi- ventricular repair

Bi-ventricular repair

- RVOT reconstruction
  - Pericardial patch, monocuspid patch,
  - homograft, tissue valve
- RV muscle resection and enlargement
- TV repair
- Atrial communication closure or adjustment

- Removal of additional pulmonary blood source.

Late problems after bi-venticular repair

- RV dysfunction
- Tricuspid valve regurgitation
- Pulmonary valve regurgitation
- Pulmonary artery deformity
- Repeated reoperation or interventions

Risk factors for early death and reoperation following biventricular repair of pulmonary atresia with intact ventricular septum

Jonah Odim*, Hillel Laks, Thomas Tung

Division of Cardiothoracic Surgery, Department of Surgery, David Geffen School of Medicine at UCLA, 10833 Le Conte Avenue, CHS 62-226B Los Angeles, CA 90095, USA

- 1982 – 2001: 106 patients
- Biventricular repair (n=30), one and half repair (n=26)
- Single ventricle repair (n=23), transplantation (n=2)
- death before definite repair (n=9), follow up loss (n=8), waiting definite repair (n=8)

- 5 – year survival after definite repair : 91.5%
- Risk factor for early death:
  - Omission of palliative RVOT relief
  - Non Causasian race

Eur J Cardio-thorac Surg 2006;29:659—665

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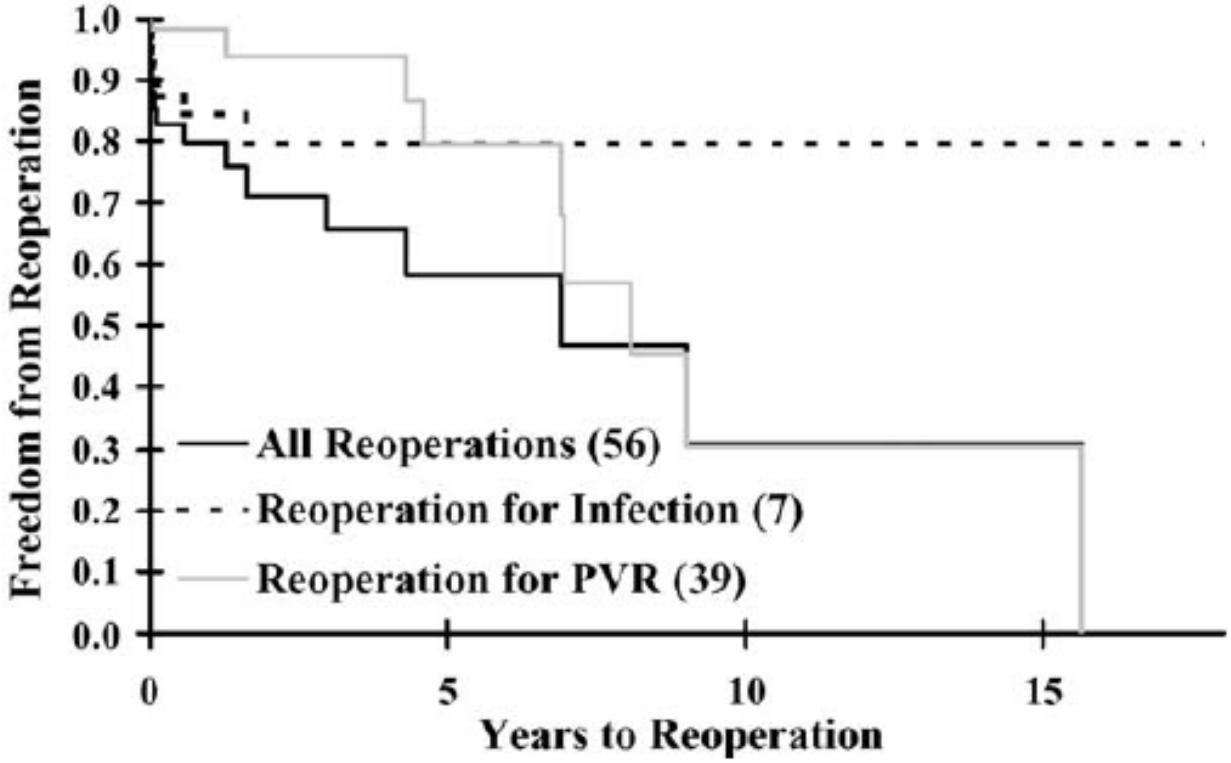

Eur J Cardio-thorac Surg 2006;29:659—665

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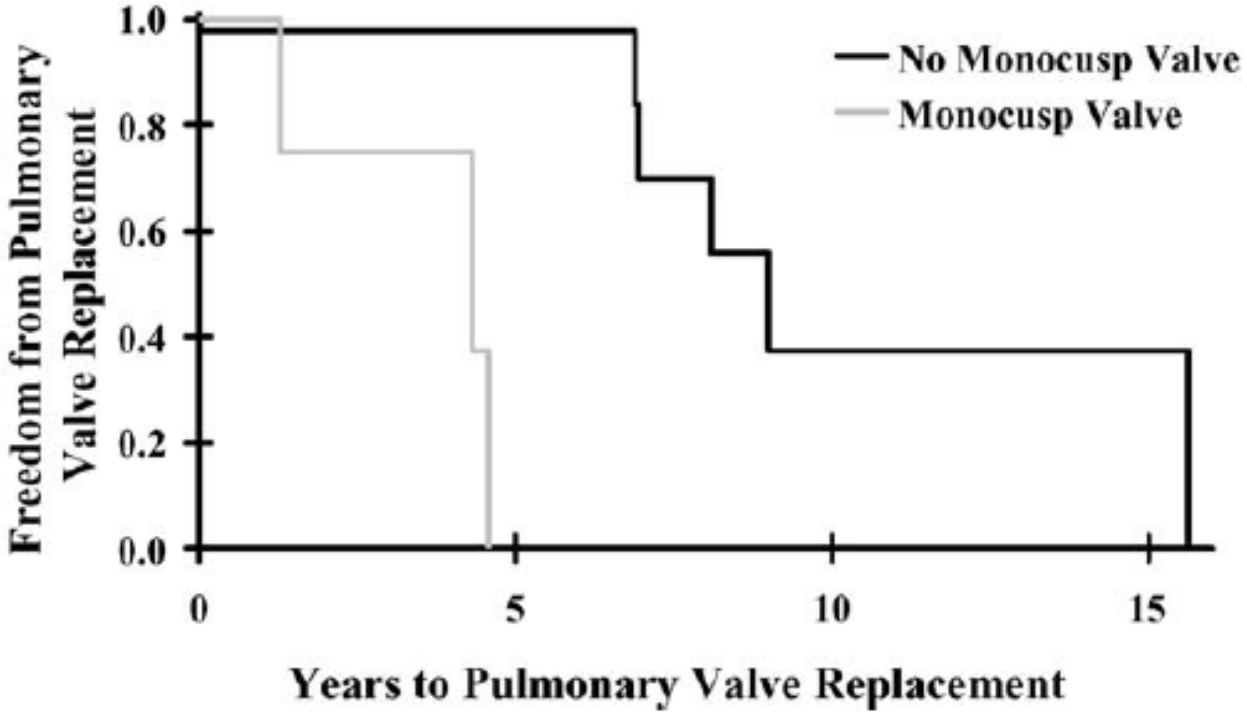


Eur J Cardio-thorac Surg 2006;29:659—665

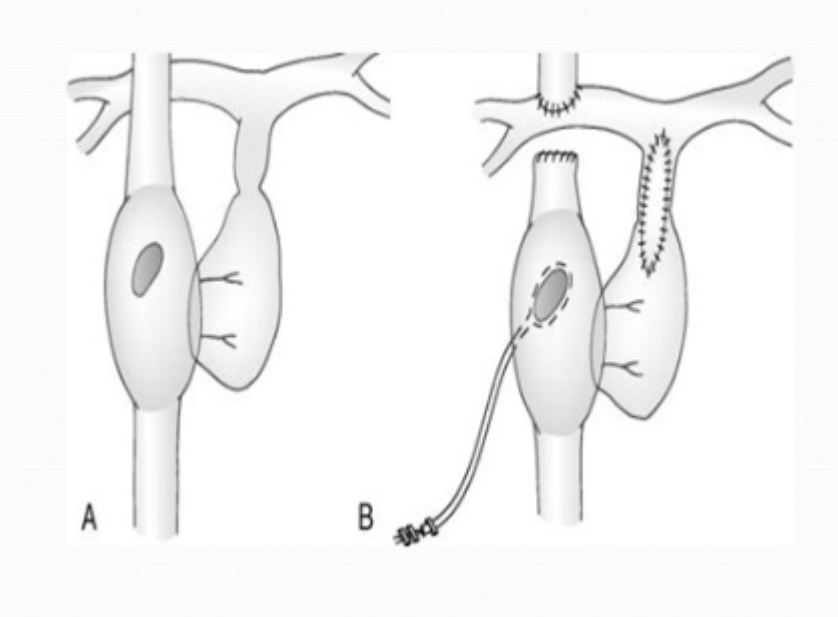
Partial bi-ventricular repair

One and Half ventricle repair

- BCPS
- ASD closure or adjustment

- RVOT patch or homograft
- RV muscle resection

- Shunt take down



Advantage of 1+ 1/2 ventricle repair

- Physiologic benefit of lower systemic venous pressure (corresponding potential salutatory effects on cerebral, coronary, hepatic and splanchnic circulatory beds)
- Prevention of right heart blood stasis and subsequent thrombo-embolism
- Improved exercise tolerance
- Naturally pulsatile pulmonary blood flow.
- Avoid RV volume loading
- RV growth and TV growth

Eur J Cardio-thorac Surg 2006;29:659—665

Fate of one and half repair

- We don’t know the long term result

- Possible problems
  - TR
  - Pulmonary regurgitation
  - RV dysfunction
  - Arrhythmias

- Consier conversion from one-half ventricular repair to Single ventricle type repair just in case

Pulmonary Atresia With Intact Ventricular Septum: Long-Term Results of "One and a Half Ventricular Repair"

Kagami Miyaji, MD, Munehiro Shimada, MD, Akihiko Sekiguchi, MD, Akira Ishizawa, MD, Takayoshi Isoda, MD, and Minoru Tsunemoto, MD

Departments of Cardiovascular Surgery and Pediatric Cardiology, National Children's Hospital, Tokyo, Japan

Background. Between 1982 and 1984, we successfully performed "one and a half ventricular repair" using a Glenn shunt for 3 patients with pulmonary atresia with intact ventricular septum. Here we review the 10-year follow-up results.

Methods. In these patients, the preoperative Z scores of the tricuspid valve diameters ranged from −5.2 to −6.5. Right ventricular outflow tract reconstruction combined with a Glenn shunt were performed in all patients. Cardiac catheterization was done at least 10 years postoperatively.

Results. All 3 patients have maintained New York Heart Association functional class I status for more than 10 years. Angiography in 2 patients confirms sufficient left pulmonary artery pressure with pulsatile blood flow and good right ventricular contraction. A pulmonary arteriovenous fistula has developed in 1 patient.

Conclusions. Although the lower limits of the tricuspid valve diameter for "one and a half ventricular repair" using a cavopulmonary shunt have not yet been determined, we successfully performed this procedure in 3 patients with severely hypoplastic right ventricles and tricuspid valve diameter Z scores of less than −5.0. The results up to 10 years postoperatively are acceptable.

(Ann Thorac Surg 1995;60:1762–4)

Ann Torac Surg 1995;60:1762-4

Updated in 2001 by Hiroo Takayama, MD, Akihiko Sekiguchi, MD, and Masahide Chikada, MD

Division of Cardiovascular Surgery, National Children's Hospital, Tokyo, Japan

- Follow-up : 17 – 19 years
- Functional class : I
- Exercise ECG : supraventricular arrythmia in 2

  ST deression in 1
- Echo
  - Increased RV EDV
  - TR : mild in 1
- Pulmonary regurgitation
- Pulmonary AV fistular in 2

Ann Thorac Surg 2001;72:2179

Long-term functional results of the one and one half ventricular repair for the spectrum of patients with pulmonary atresia/stenosis with intact ventricular septum☆

Satoshi Numata, Hideki Uemura*, Toshikatsu Yagihara, Koji Kagisaki, Masashi Takahashi, Hideo Ohuchi

- National cardiovascular center , Japan
- From 1987 – 1999
- 7 PA IVS, 6 PS RV hypoplasia (3 Ebstein's)
- One half ventricle repair

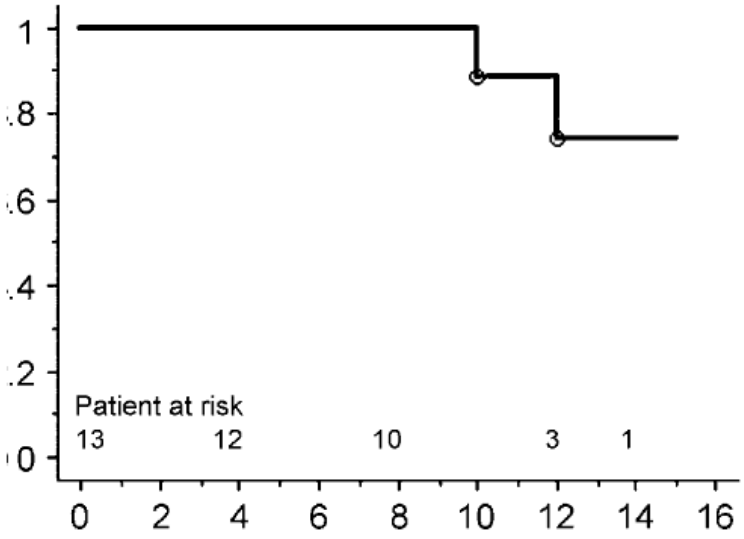
- Age at 1.5 repair: 4 years (10 months – 9years)
- Follow up : 3 – 15 years (10 +_ 4 years)

Eur J Cardio-thorac Surg 24 (2003) 516–520

Long-term functional results of the one and one half ventricular repair for the spectrum of patients with pulmonary atresia/stenosis with intact ventricular septum☆

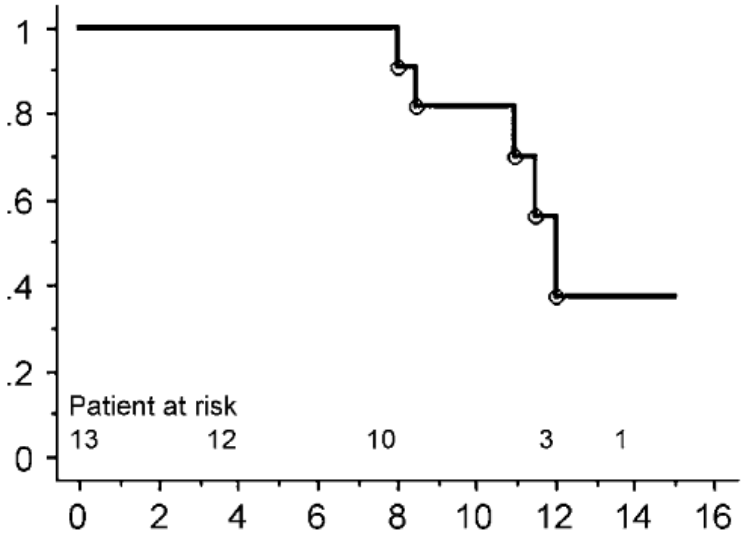
Satoshi Numata, Hideki Uemura*, Toshikatsu Yagihara, Koji Kagisaki, Masashi Takahashi, Hideo Ohuchi

Survival curve



years after operation

Arrhythmia free curve



years after operation

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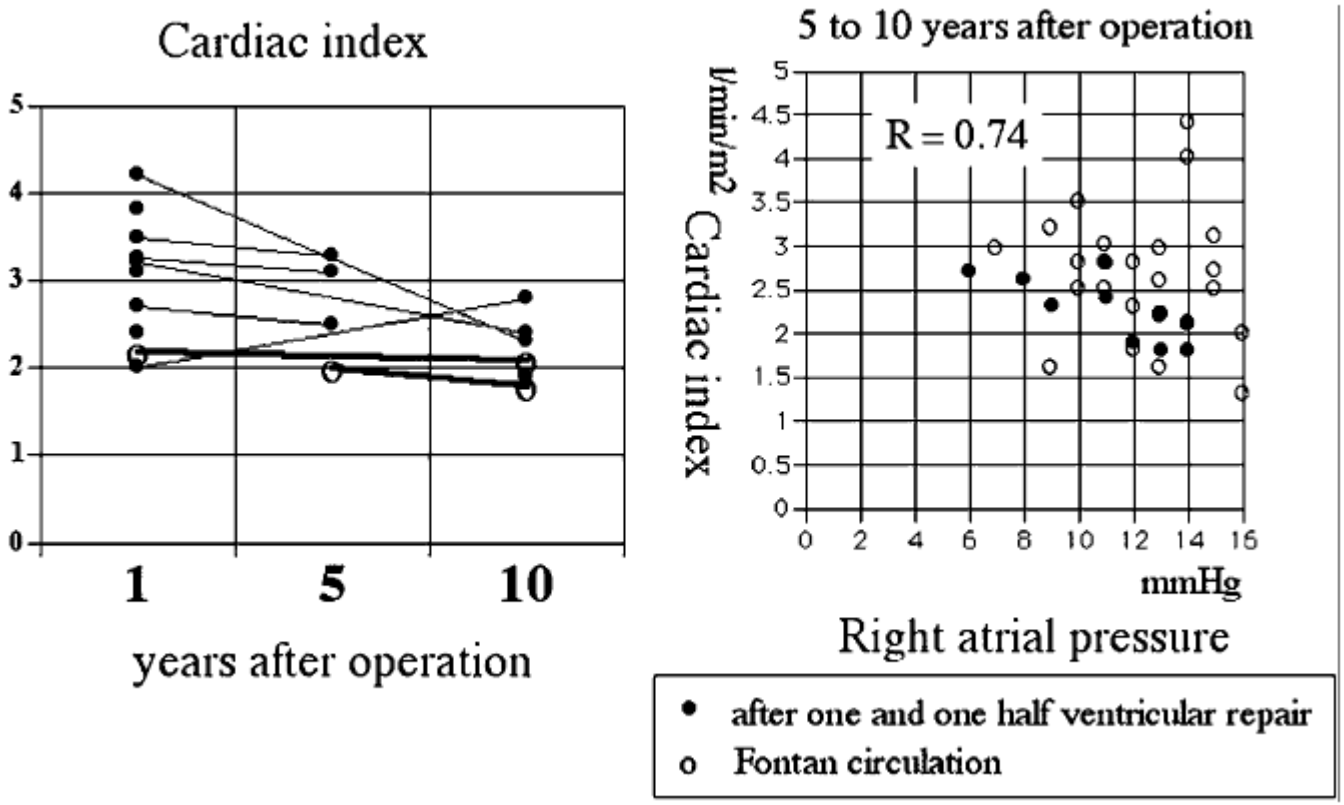


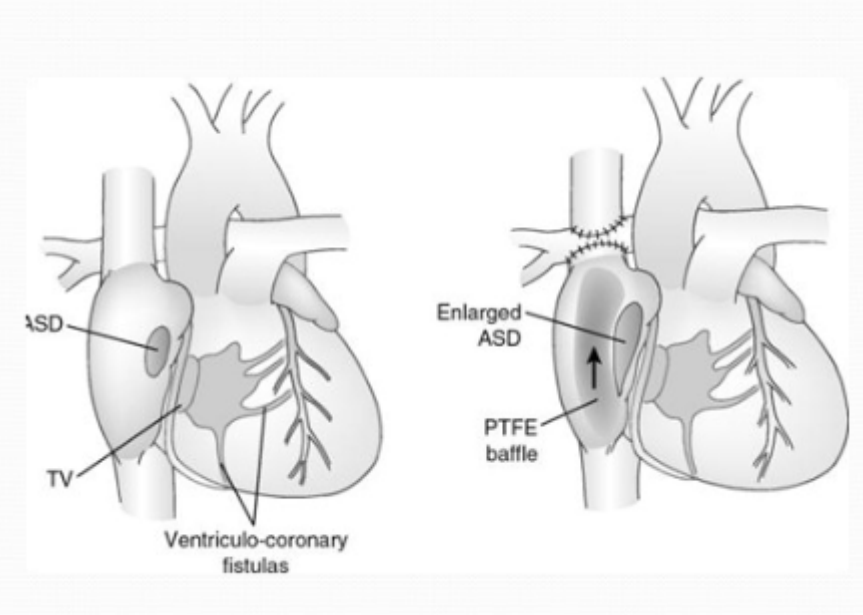
Eur J Cardio-thorac Surg 24 (2003) 516–520

Uni-ventricular repair

Univentricular repair

- BDG after taking down of shunt at 4-6 months of age
- Fontan operation at 2-4 year of age

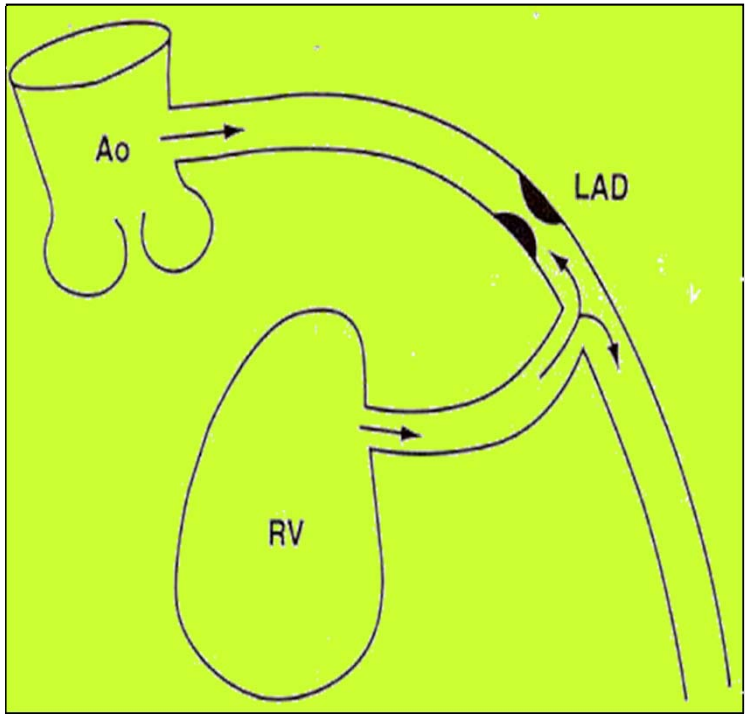
- Cardiac transplantation
- Heart lung transplantation



RV-coronary artery circulation

- Frequency : 17 - 75% (Coles, 1989; Hanley, 1993)
- Fistula alone
- Fistula + stenosis or interruption to coronary arteries
  (RV dependent coronary circulation)

Mechanism of proximal coronary artery stenosis



- Turbulent flow
- Intimal and adventitial fibrosis
- Medial muscular hypertrophy

Najm HK, et al. Ann Thorac Surg 1997;63:669-75

RV-coronary artery circulation

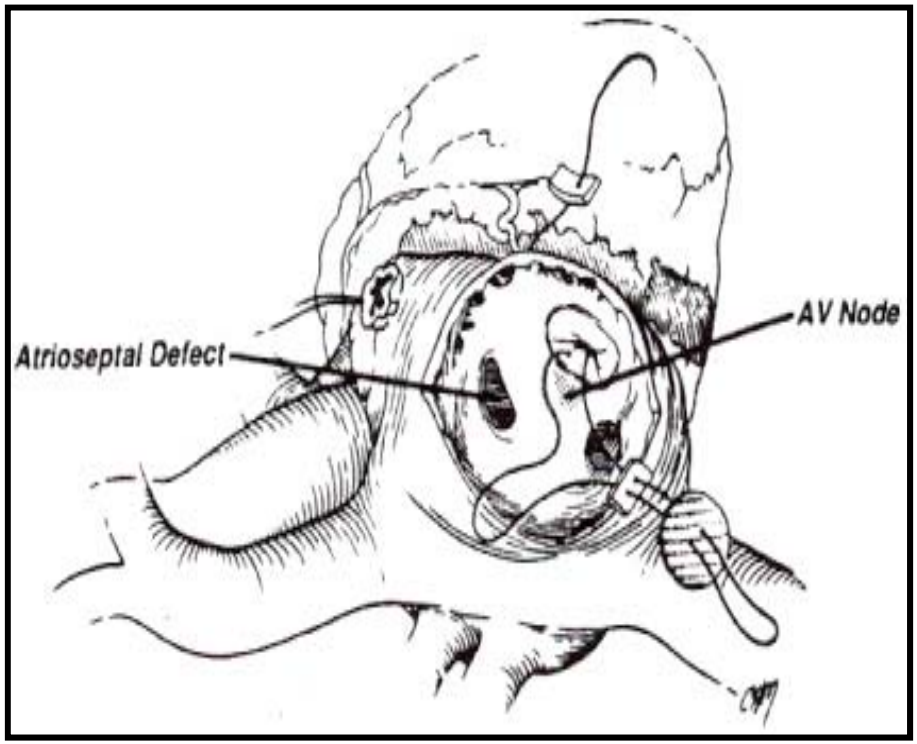
- Deoxygenated blood supply to myocardium (systole)
- Coronary steal (diastole)
- Left ventricular dysfunction

- Decompression of RV -- increase ischemia (Giglia, 1992)

- Tricuspid valve excision or avulsion (Hawkins, 1990)
- TV closure (Waldman, 1995)
- Thromboexclusion of RV (Najm, 1997; William, 1991)
- Aortic - RV shunt (Laks, 1995; Freeman 1993)

- Transplantation

TV closure in RV-CAC



- Assessing hemodynamic stability -- temporary balloon closure
- not indicated RV dependent coronary circulation
- Closure by 6 months of age
- Patch of Dacron or Gore-Tex
- Base of septal leaflet

Waldman JD, et al. Ann Thorac Surg 1995;59:933-41

Thromboexclusion of RV

- Not indicated with RV dependent coronary circulation
- Recommended before 1 year of age at the time of BCPS
- TV closure and Coils and Gelfoam injection to RV

Williams WG. Et al. J Thorac Cardiovasc Surg 1991;101:222-9

LV function after Fontan in PA-IVS

- Poor LV function
  - RVDCC : myocardial ischemia
  - High pressure RV

- Abnormalities in myocardial perfusion

Left ventricular performance of pulmonary atresia with intact ventricular septum after right heart bypass surgery

Yoshihisa Tanoue, MD
Hideaki Kado, MD
Taketoshi Maeda, MD

- Fukuoka children's hospital
- 20 PA IVS vs. 21 TA

- Contractility
- Afterload
- Ventricular efficiency
- Stroke work, pressure – volume area
- Before and after BCPS

J Thorac Cardiovasc Surg 2004;128:710-7

Left ventricular performance of pulmonary atresia with intact ventricular septum after right heart bypass surgery

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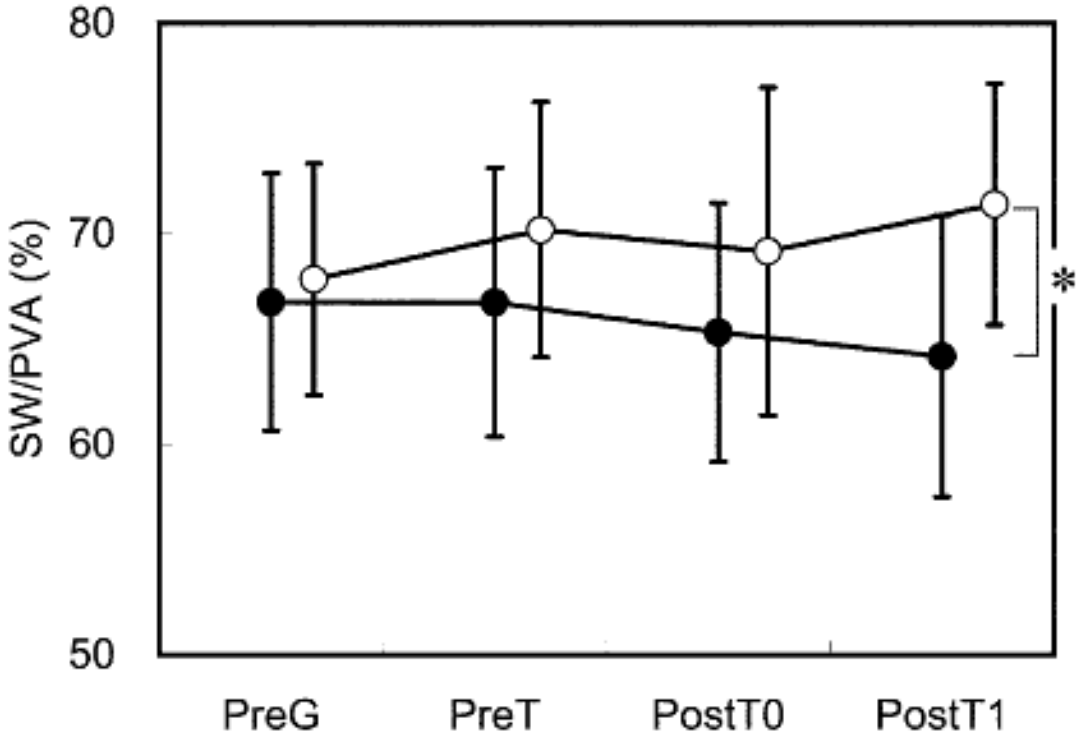


J Thorac Cardiovasc Surg 2004;128:710-7

Exclusion of the non-functioning right ventricle in children with pulmonary atresia and intact ventricular septum☆

Ji-Hyuk Yang[a], Tae-Gook Jun[a,*], Pyo Won Park[a], Kiick Sung[a], Wook Sung Kim[a], Young Tak Lee[a], June Huh[b], I-Seok Kang[b]

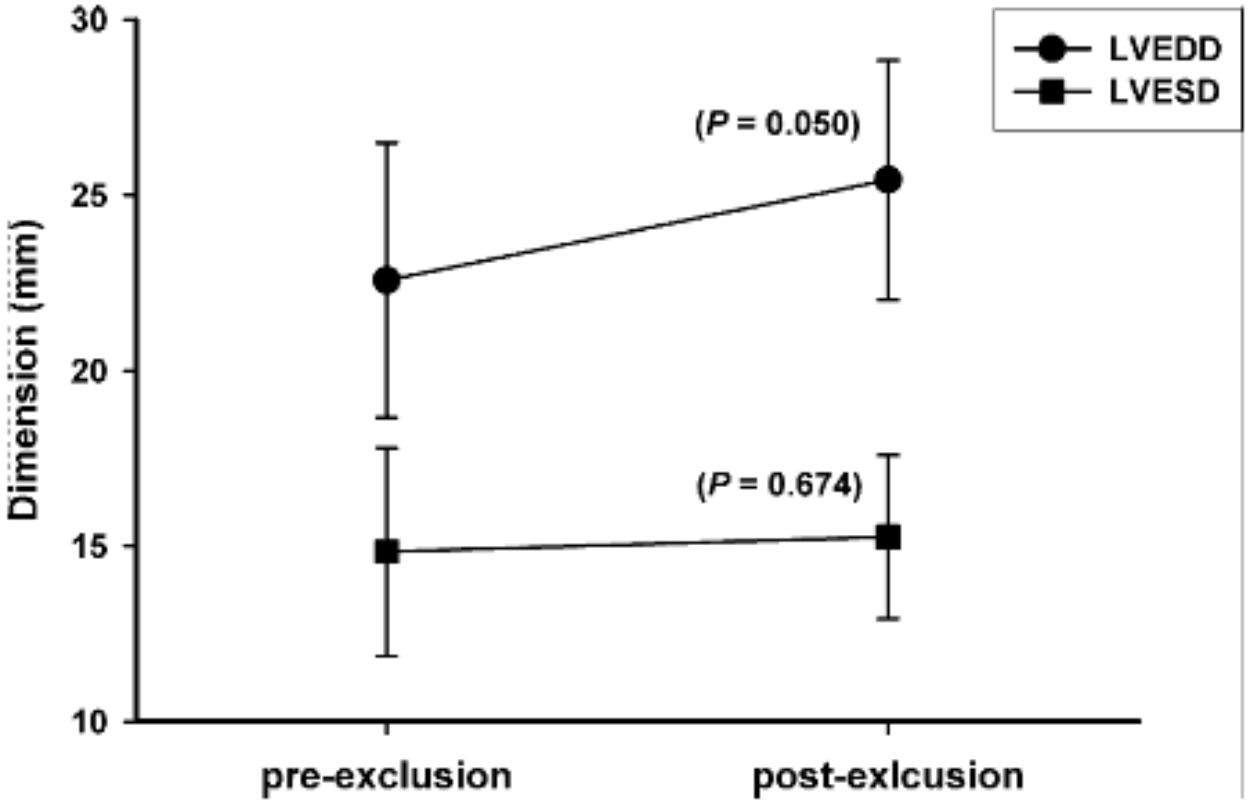
Table 1
Preoperative patients' characteristics and operative procedures

Patient no.	Age (months)	Previous procedure	z-Value	RV-to- coronary sinusoids	Operation				
					Palliative procedure	Tricuspid valve closure	Thrombotic material insertion	Atrial septectomy	Other procedures
1	13.8	RVOT + BT shunt at 3.7 months	−3.6	Yes	BCPS	Patch	–	Yes	–
2	6.9	RVOT + BT shunt at 6 days	−1.3	–	BCPS	Direct	–	Yes	RPA angioplasty
3	4.8	BVP at 8 days	−4	Yes	BCPS	Direct	Yes	Yes	–
4	10.0	BT shunt at 6 days	−3.3	–	BCPS	Direct	Yes	Yes	–
5	2.1	–	−5	–	BT shunt	Direct	Yes	Yes	–
6	6.2	BT shunt at 7 days	−3.2	–	BCPS	Direct	Yes	Yes	–
7	5.2	BT shunt at 23 days	−6.5	Yes	BCPS	Patch	Yes	Yes	–
8	0.2	–	Atretic	–	BT shunt	Direct	Yes	Yes	LPA angioplasty
9	2.6	–	−5.4	Yes	BT shunt	Direct	Yes	Yes	LPA angioplasty
10	0.4	–	−3.3	Yes	BT shunt	Direct	Yes	Yes	Coronary fistula ligation

Eur J Cardio-thorac Surg 2008;33:251-256

Exclusion of the non-functioning right ventricle in children with pulmonary atresia and intact ventricular septum☆

Ji-Hyuk Yang[a], Tae-Gook Jun[a,*], Pyo Won Park[a], Kiick Sung[a], Wook Sung Kim[a], Young Tak Lee[a], June Huh[b], I-Seok Kang[b]



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Clinical outcomes of adult survivors of pulmonary atresia with intact ventricular septum

Anitha S. John [a,b,*], Carole A. Warnes [a]

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Characteristics of PA-IVS cohort, surgical subgroups.

Characteristics[a]	Univentricular, palliated (n = 5)	Univentricular, Fontan (n = 7)	Biventricular repair (n = 8)	Total number (n = 20)
Average age, years[b]	29 (23–35)	28 (23–32)	30 (18–39)	29 (18–39)
Number of patients alive	4	5	6	15
NYHA class I–II[b]	4	4	6	14
Arrhythmias	3	7	6	16
Pulmonary hypertension	4	0[c]	1	5
Endocarditis	3	1	0	4
Intracardiac thrombosis	0	4	2	6
Valvular dysfunction	1	2	8	15
Protein losing enteropathy	0	2	0	2
Echocardiogram parameters				
Left ventricular EF	55% (50–65%)	51% (45–58%)	58% (50–65%)	55% (45–65%)

Inter J Cardiology 2011:XXX

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Types of surgical re-interventions in adulthood (≥18 years).

Characteristics	Number of patients	Number of procedures	NYHA class I–II pts pre/pts post[a]	Average age, years
Univentricular, palliated (n = 5)			2/4	22 (19–24)
Patients with re-interventions, total	3/5	7 (total)		
Shunt revision, surgical	1	4		
Shunt dilation, transcatheter	2	2		
Coil embolization of collaterals	1	1		
Univentricular, Fontan (n = 7)			2/5	21 (17–32)
Patients with re-interventions, total	7/7	14 (total)		
Fontan conversion	4	4		
MAZE procedure	3	3		
Mitral valve repair/replacement	2	2		
Fontan fenestration, transcatheter	1	1		
Ascending aorta replacement	1	1		
Fontan revision	1	1		
Biventricular repair (n = 8)			3/6	27 (19–38)
Patients with re-interventions, total	7/8	31 (total)		
Tricuspid valve repair/replacement	6	7		
Pulmonary valve replacement	5	7		
RV to PA conduit replacement	1	1		
RVOT reconstruction/augmentation	6	7		
Pulmonary artery intervention, transcatheter	3	3		
Mitral valve repair/replacement	2	4		
MAZE procedure	2	2		

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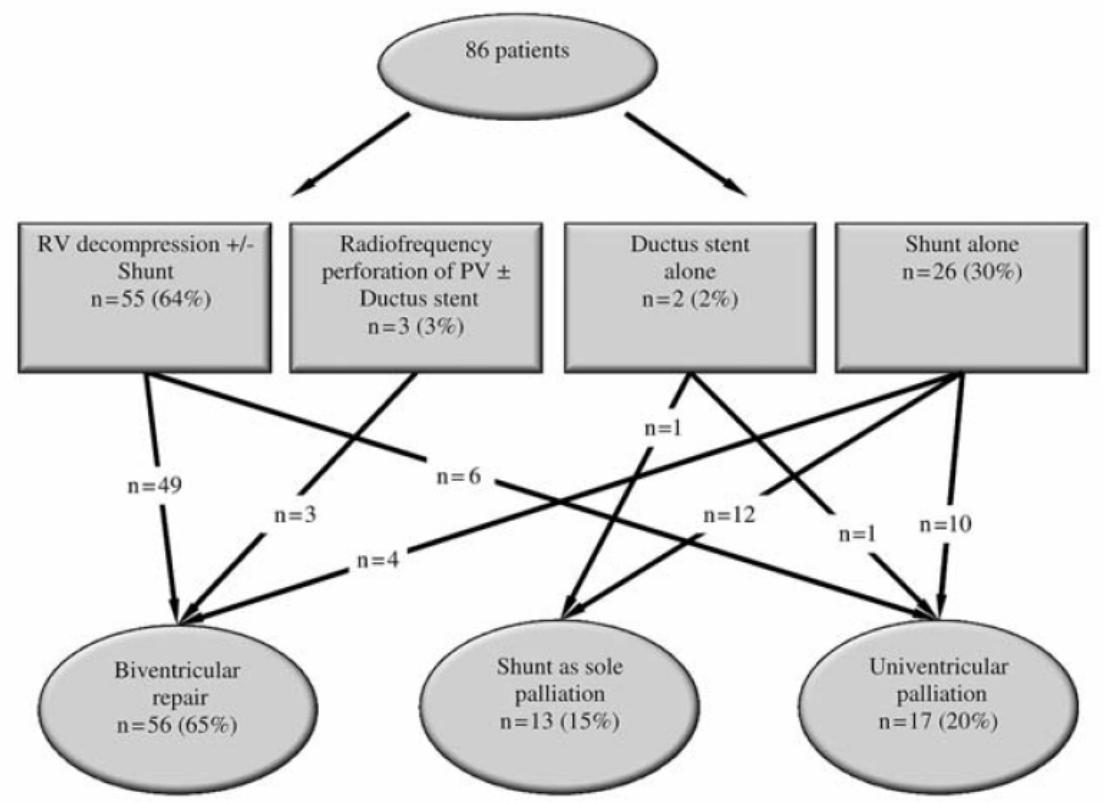
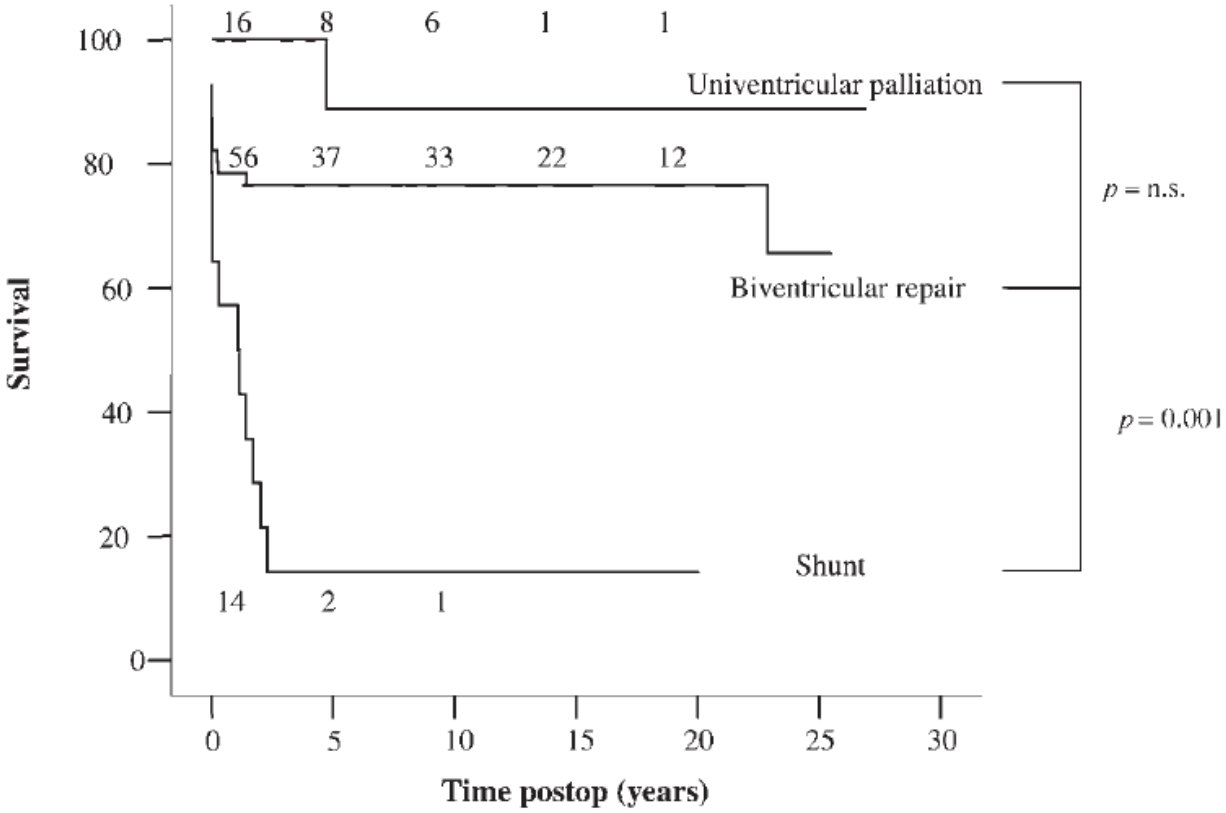
Predictors for Biventricular Repair in Pulmonary Atresia with Intact Ventricular Septum



Thorac Cardiov Surg 2010; 58: 339–344

Predictors for Biventricular Repair in Pulmonary Atresia with Intact Ventricular Septum



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Predictors for Biventricular Repair in Pulmonary Atresia with Intact Ventricular Septum

Table 1 Surgical and morphological factors predicting biventricular repair in 48 patients.

Variable	Patients with biventricular repair (n = 48)	*p*
RV decompression	36	
Systemic-to-pulmonary artery shunt	2	
RV decompression + shunt	10	< 0.001
Right ventricular morphology		
Unipartite RV	0	
Bipartite RV	6	
Tripartite RV	42	< 0.001
TV z-score ≤ −6	13	
TV z-score > −6	35	< 0.001
TR 0-I	2	
TR II–III	46	< 0.001
Coronary fistulae		
► yes	7	
► no	41	< 0.001
Coronary stenoses	0	
No coronary stenoses	48	0.004
Right ventricular dependent coronary circulation	1	
No RV dependent coronary circulation	47	0.008

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Changing trends in the management of pulmonary atresia with intact ventricular septum: the Melbourne experience

Matthew Liava'a [a,c], Paul Brooks [b], Igor Konstantinov [a], Christian Brizard [a], Yves d'Udekem [a,c,1,*]

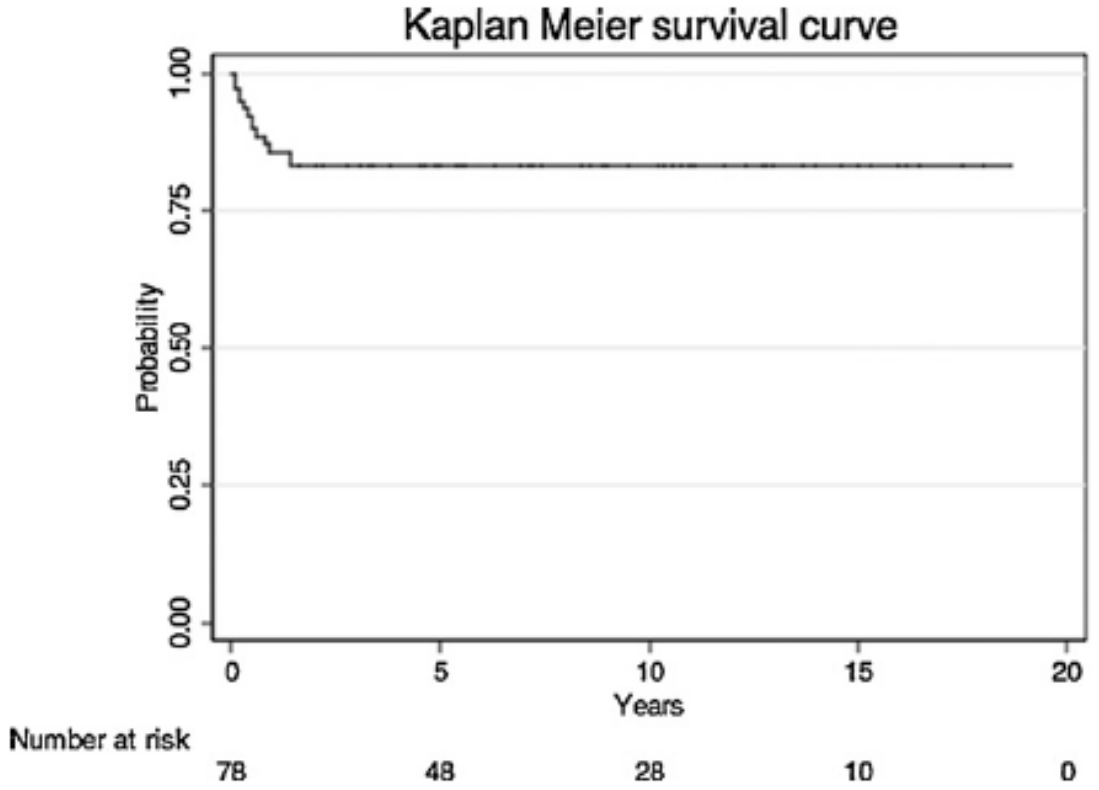

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Table 3. Outcomes based on right-ventricle size.

RV size	Outcome						
	Bi-V	1.5-V	Awaiting	Uni-V	HTx	Dead	Total (%)
Normal	10	1	0	0	0	0	11 (13.5%)
Moderate hypoplasia	19	8	3	9	0	6	45 (55.5%)
Severe hypoplasia	2	1	6	5	1	10	25 (31.0%)
Total	31	10	9	14	1	16	81 (100%)

Bi-V: biventricular; 1.5-V: 11/2 ventricle; Uni-V: univentricular; and HTx: heart transplant.

Eur J Cardio-thoracic Surgery 40 (2011) 1406—1411

SMC Experience

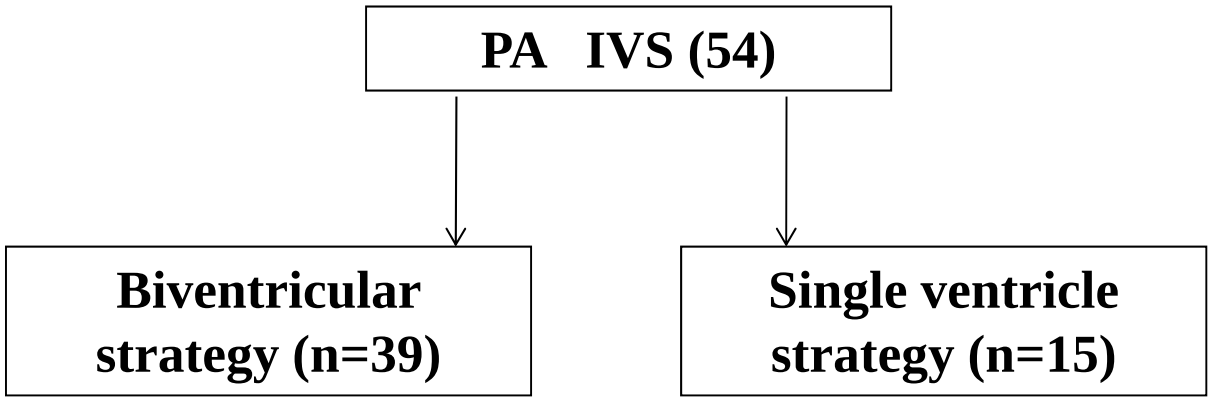
- 1995 – 2009

- 54 patients
- PA IVS

- Exclusion : palliation procedure in other hospital

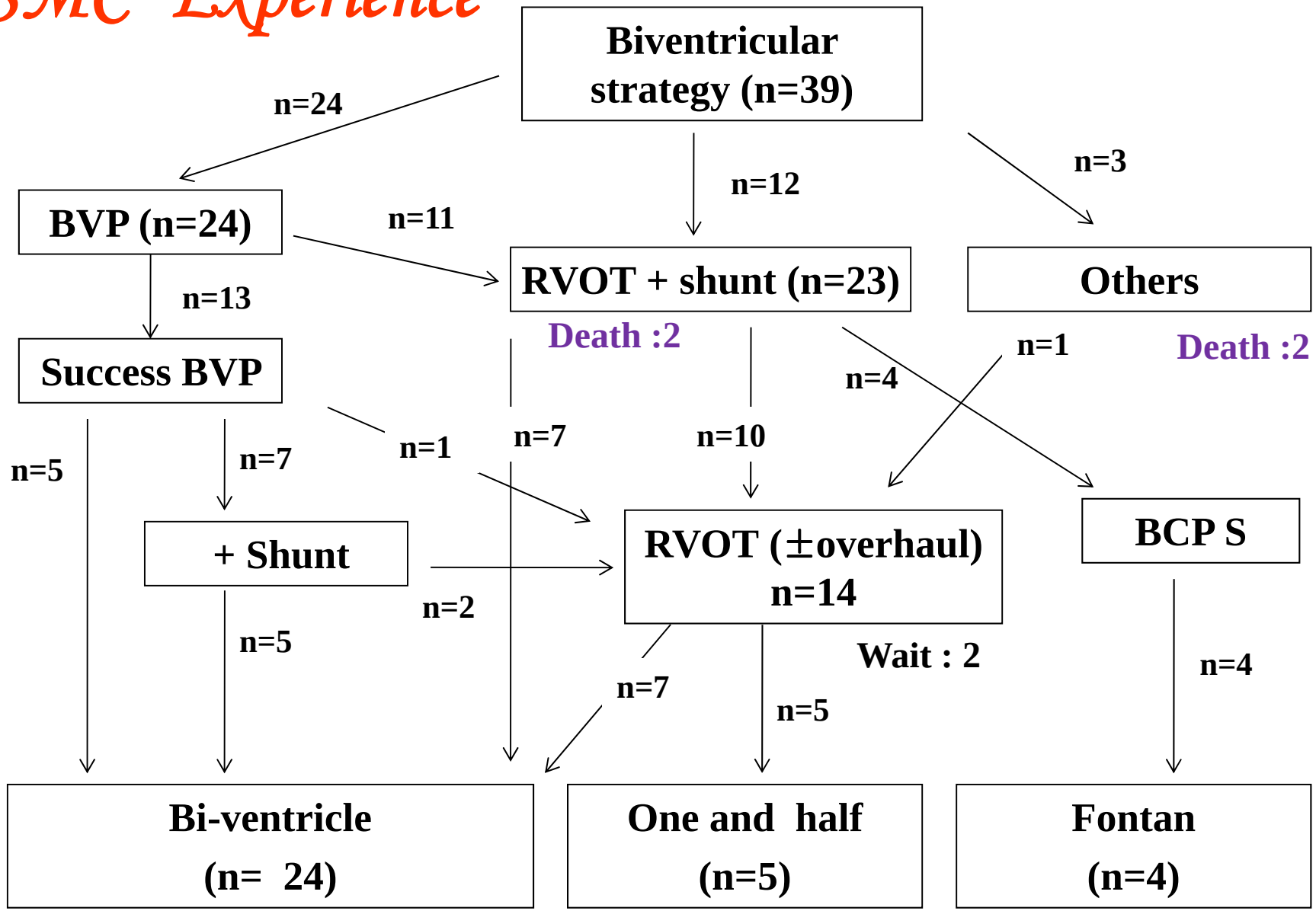
SMC Experience

Initial palliation

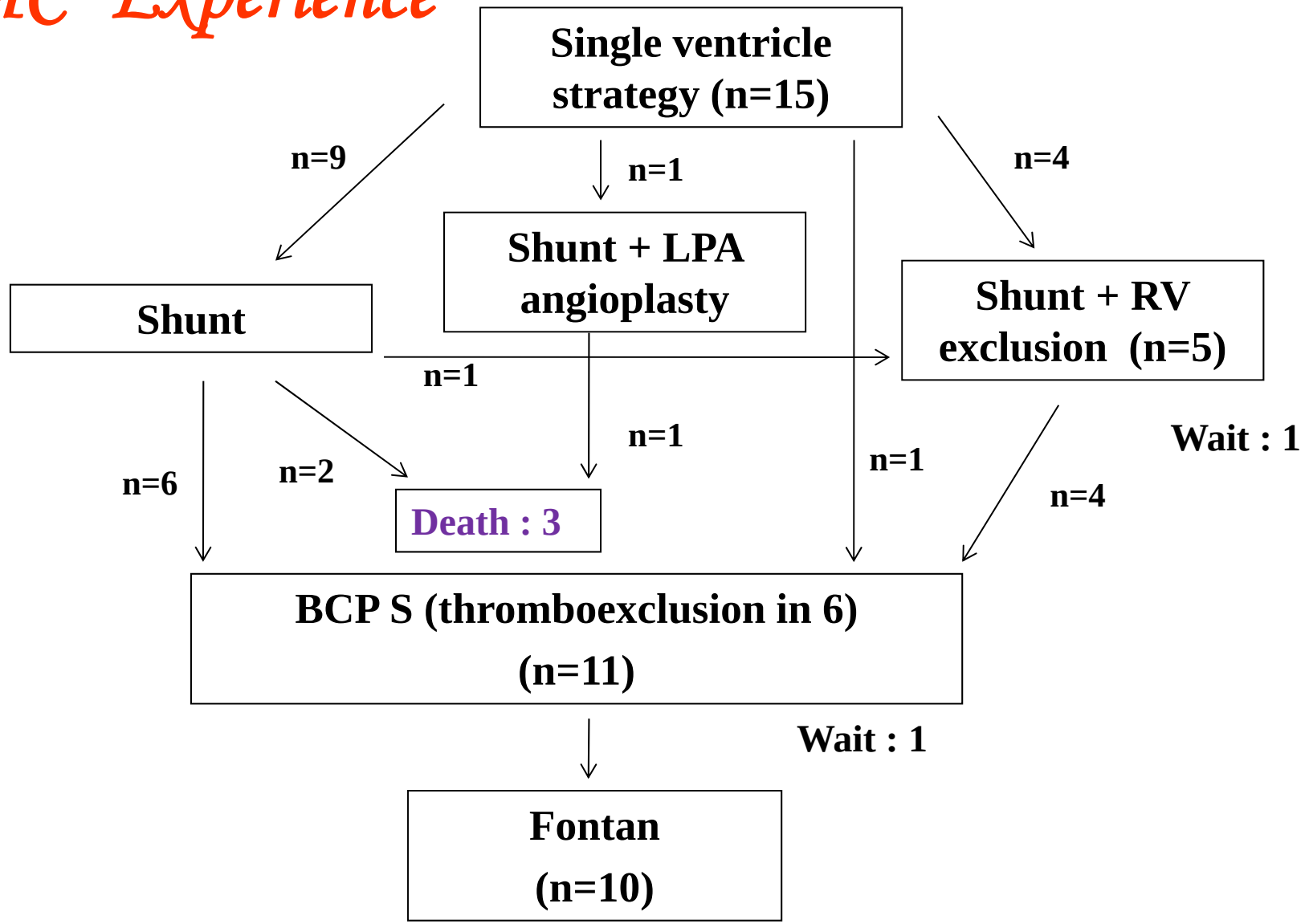


Definite Repair

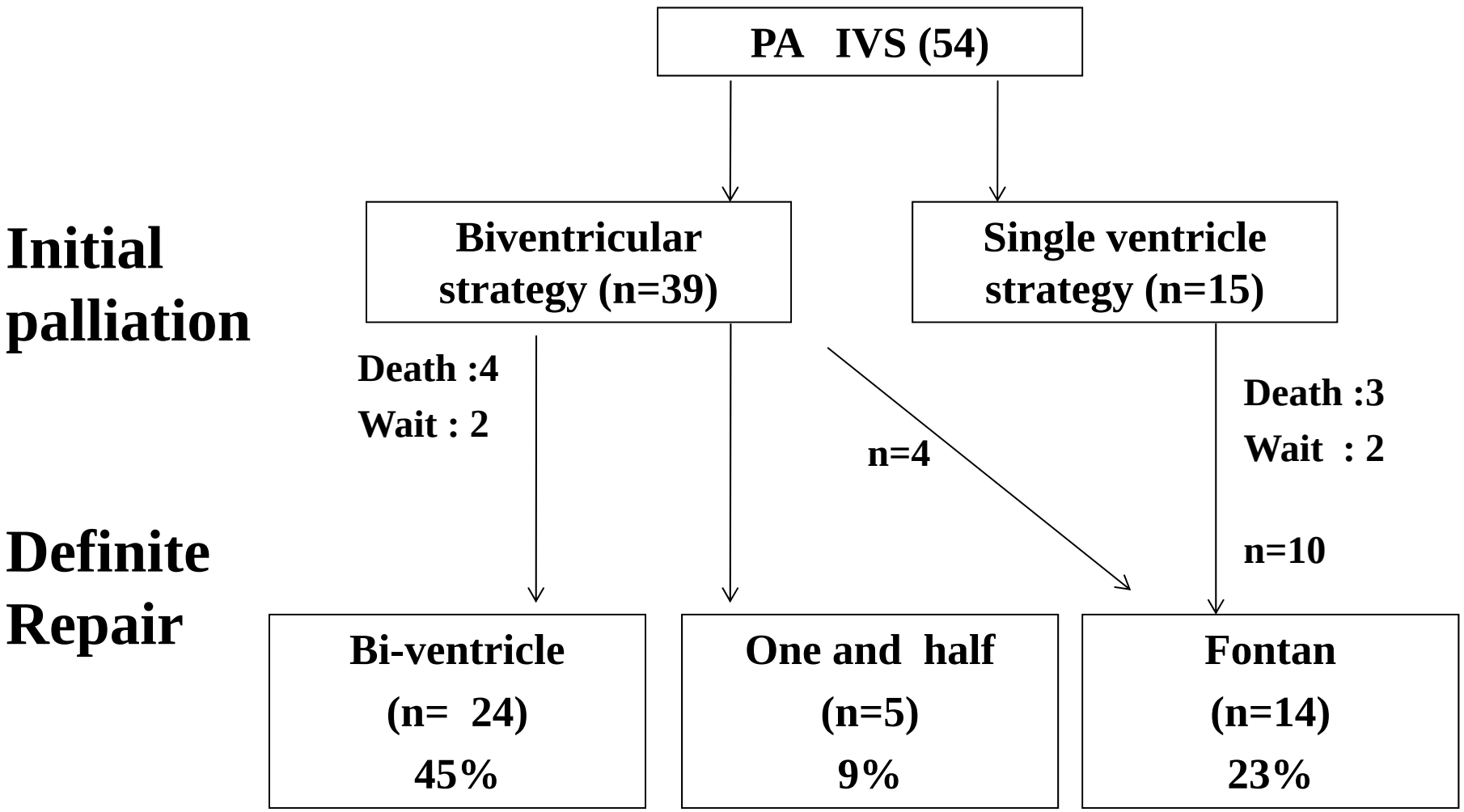
SMC Experience



SMC Experience



SMC Experience



SMC experiences

- Reoperation after definite repair
  - PVR in 3
  - RV exclusion after Fontan in 1

- Last Echo of biventricular + one and half group (29)
  - PR > moderate in 14
  - TR > moderate in 19

Summary

- Catheter based BVP can be a best option for initial palliation, however additional surgical procedure should be considered when it is needed.

- Early re-evaluation after the RV decompression and adequate additional palliative procedures are essential for the preparation for the definite repair

- When the TV size(valve orifice) is too small, it seems that decompression of RV can not guarantee biventricular repair.

Summary

- Angiographic evaluation may be essential to find the RV-coronary fistula (or RVDCC) even in the single ventricle group.

- We cannot insist which protocol is superior to other strategies especially in the point of long term results.

- Every procedure should be considered precisely in the aspect of reducing repeated re-operations and interventions after definite repairs.

Critical Pulmonary Valve Stenosis and Pulmonary Atresia/Intact Ventricular Septum: To Lump or Split? Examining Procedural Success and Risk for Reintervention

Indications for reintervention included failure to tolerate PGE1 withdrawal with excess cyanosis (<70%) and metabolic acidosis. PGE1 was continued for up to 14 days following cardiac catheterization. The need for reintervention within the study period was 15/43 (36%). Reintervention was more commonly needed for PA/IVS (52%) compared with critical pulmonary valve stenosis (17%). Patients with PA/IVS were more likely to require surgery as the second intervention (18%) compared with critical pulmonary valve stenosis (0%). One surgical mortality was encountered.

Catheterization and Cardiovascular Interventions 79:665 (2012)