

PA/IVS
RVOTR + BTS

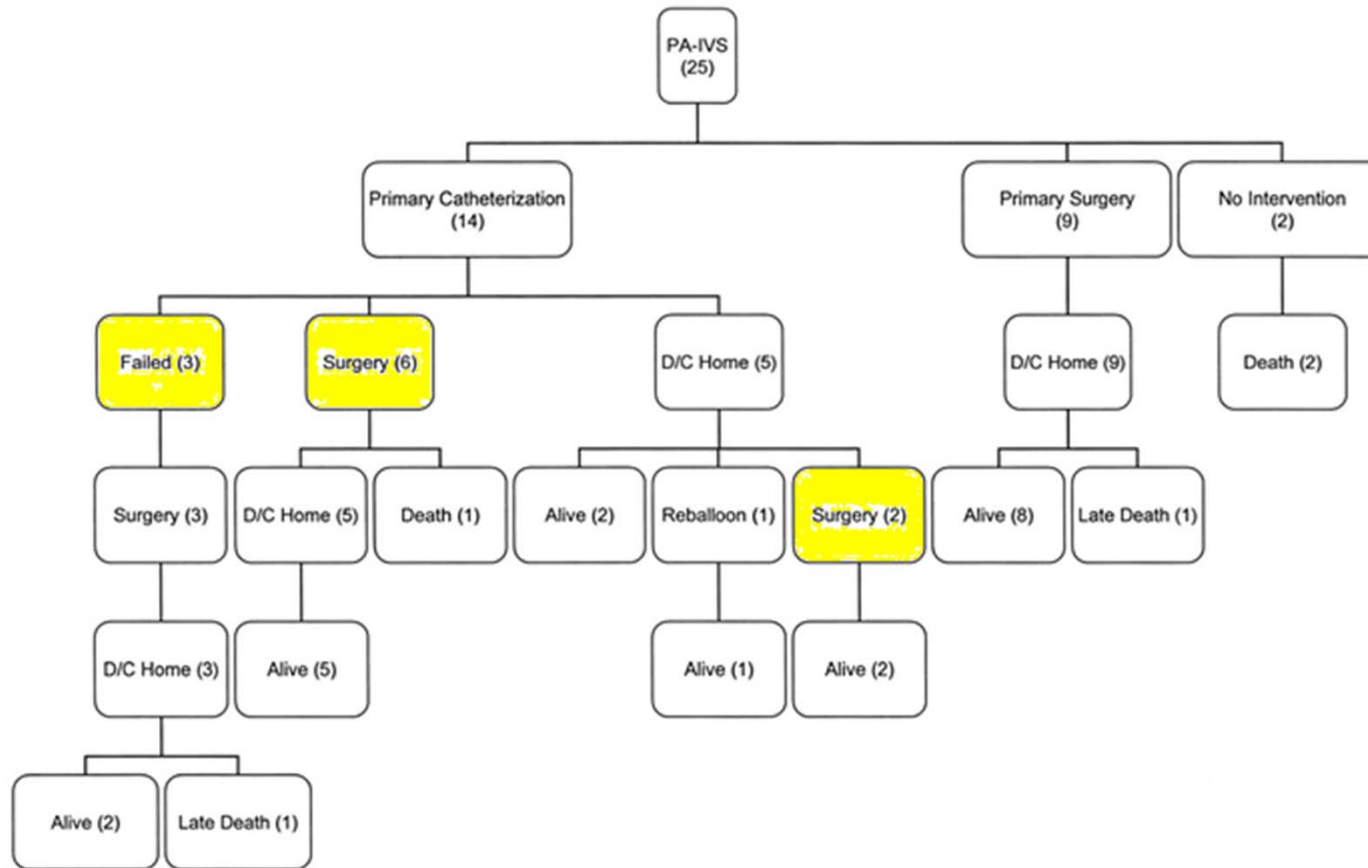
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PA With IVS : Initial Management

- Division of Cardiothoracic Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio
- 25 patients, Mar 1999 – Jun 2005
- Mean age 3.2days
- Mean weight 3.3kg
- Mean T-valve z-score : - 2.1

McLean K. M. et al.; Ann Thorac Surg 2006;82:2214-2220

Treatment and outcomes for all 25 patients admitted with a diagnosis of pulmonary atresia with intact ventricular septum (PA-IVS)



McLean K. M. et al.; Ann Thorac Surg 2006;82:2214-2220

Primary catheter based therapy

- 3 serious complication of 2/14 patients
- Patient 1 ; 10d, 2.3kg, z-score 1.6
- Initial unsuccessful antegrade attempts
- a retrograde attempt, which created a perivalvar channel complicated by tamponade and cardiac arrest

Primary catheter based therapy

- Interventricular hemorrhage, necrotizing enterocolitis, and ischemia of the left lower extremity developed, resulting in left above-knee amputation
- 14 days after her initial intervention for RVOT patch, RV muscle resection, right atrial reduction
- Discharged on 115days

Primary catheter based therapy

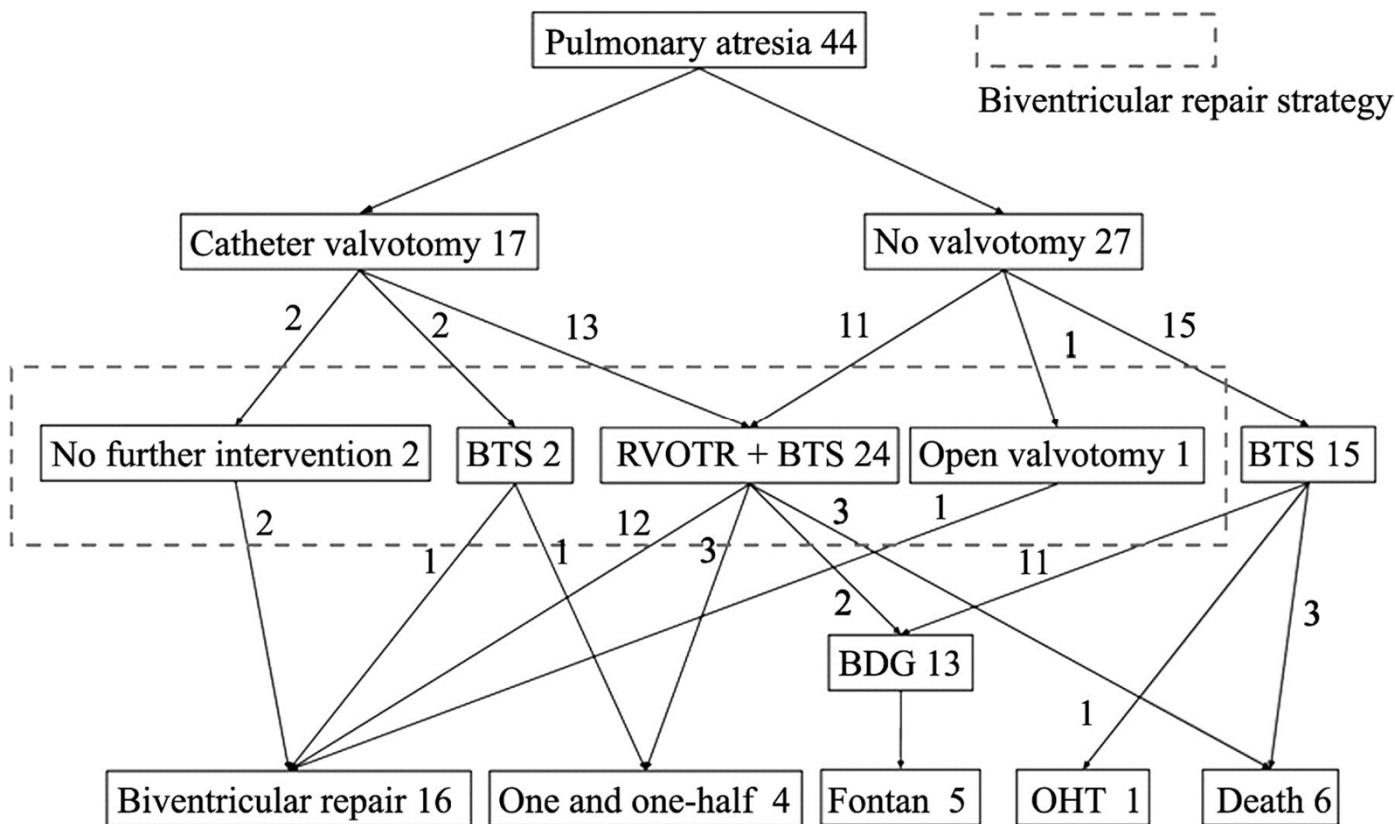
Table 1. Comparison of Duration of Prostaglandin Infusion, Intensive Care Unit Stay, and Hospital Stay Between Group 1 and Group 2

	Group 2	Group 1
Duration of prostaglandins (days)	8.1 ± 6.7	3.0 ± 1.4 ^a
Intensive care unit stay (days)	19.4 ± 20.3	6.6 ± 3.3 ^a
Hospital stay (days)	27.1 ± 26.4	12.2 ± 2.6 ^a

^a $p < 0.05$ versus primary catheter-based intervention (group 2).

Values are expressed as mean ± SD.

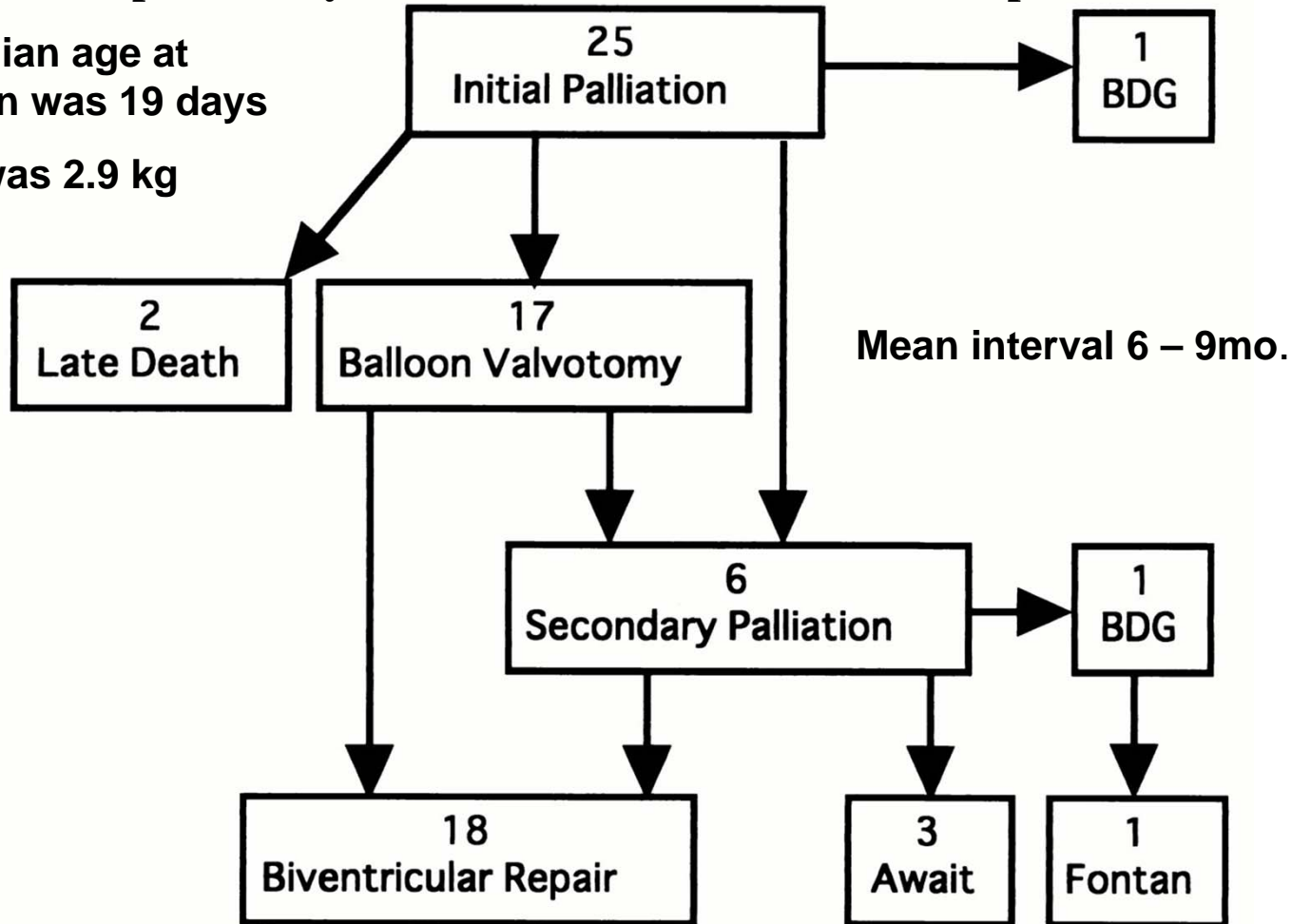
Flow chart of outcome for patients with pulmonary atresia-intact ventricular septum



Hirata Y. et al.; Ann Thorac Surg 2007;84:574-580

Intermediate outcome of 25 infants who underwent the combined pulmonary valvotomy and systemic-pulmonary shunt as a initial palliation for pulmonary atresia or critical pulmonary stenosis with intact ventricular septum

The median age at operation was 19 days
weight was 2.9 kg

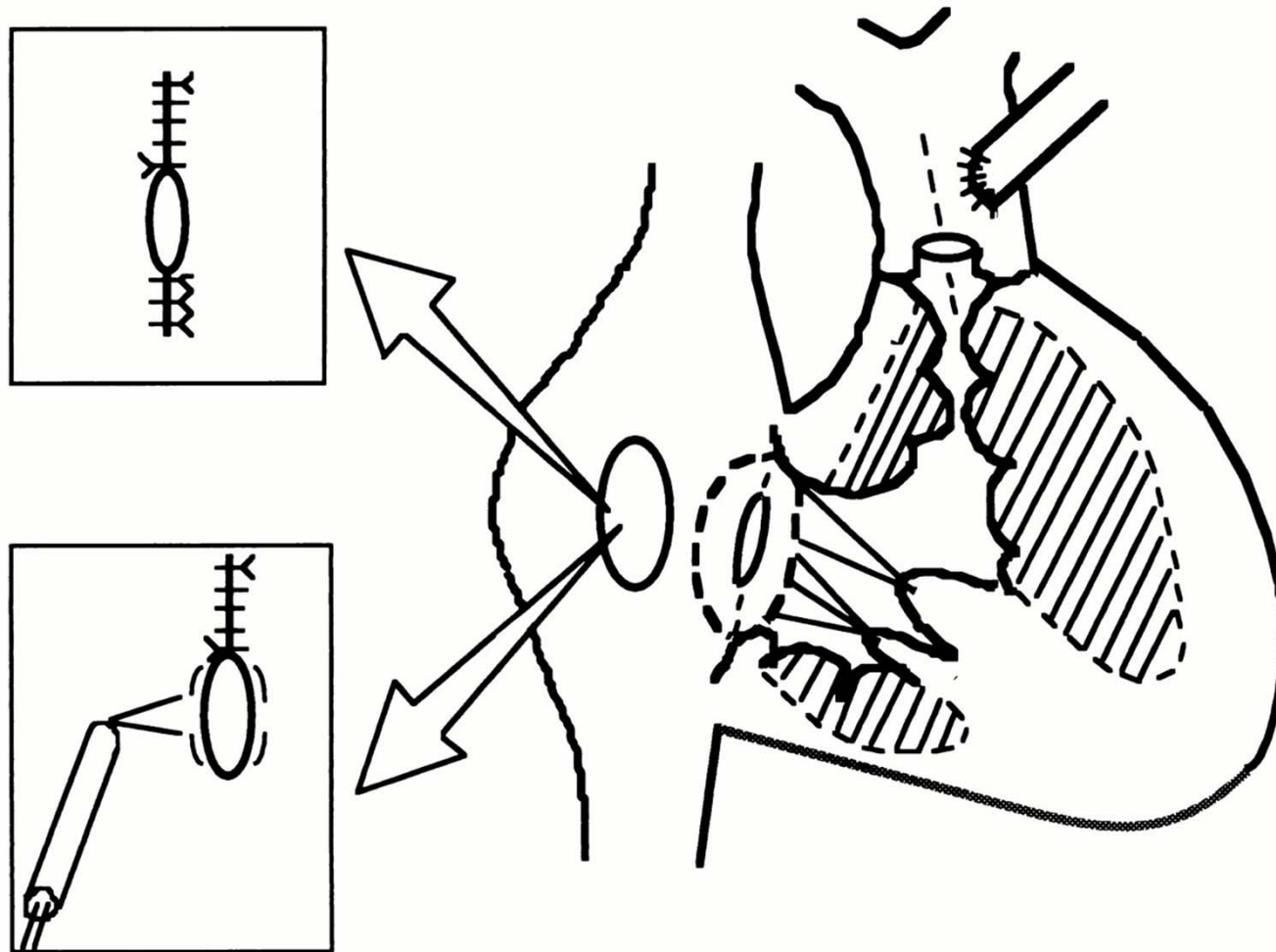


Sano S. et al.; Ann Thorac Surg 2000;70:1501-1506

Secondary surgical procedure

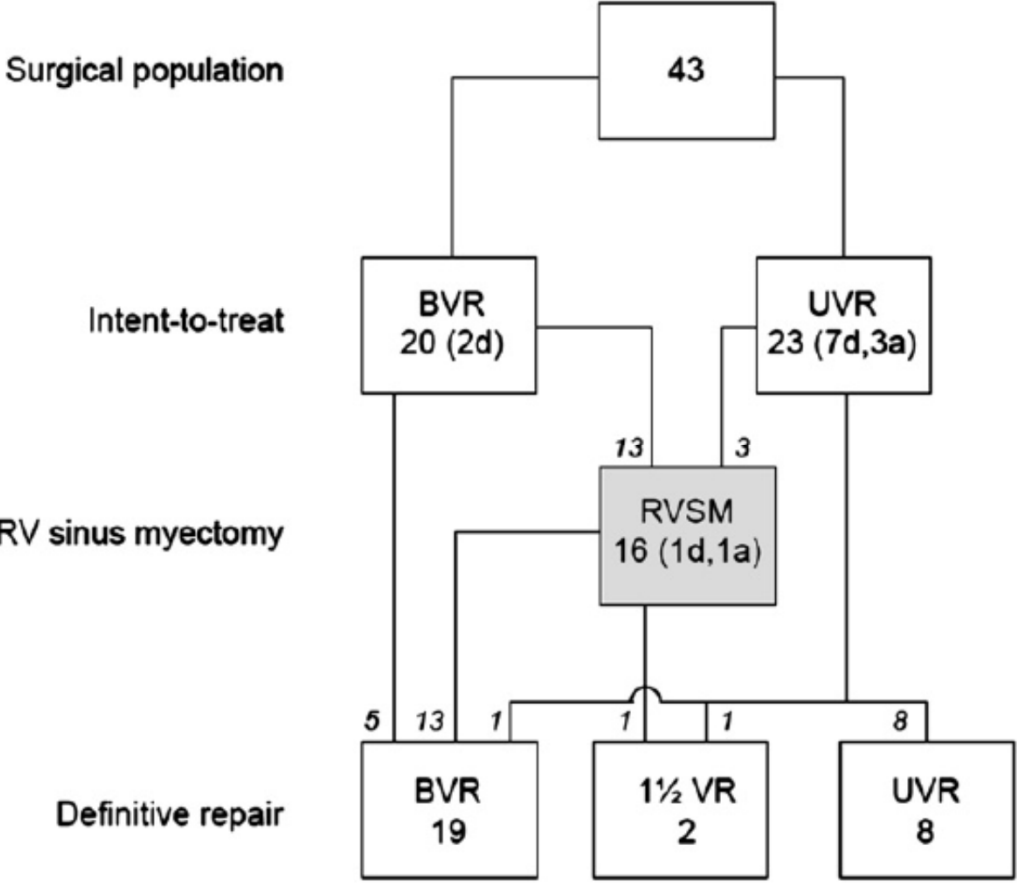
- Indicated when a RVEDV was less than 50% of predicted normal value estimated
- 6 patients at a median age of 13 months (range, 5 to 24 months)

The second palliative operation (right ventricular overhaul [14]) included repeat pulmonary valvotomy, enlargement of right ventricular cavity, and adjustment of an interatrial communication



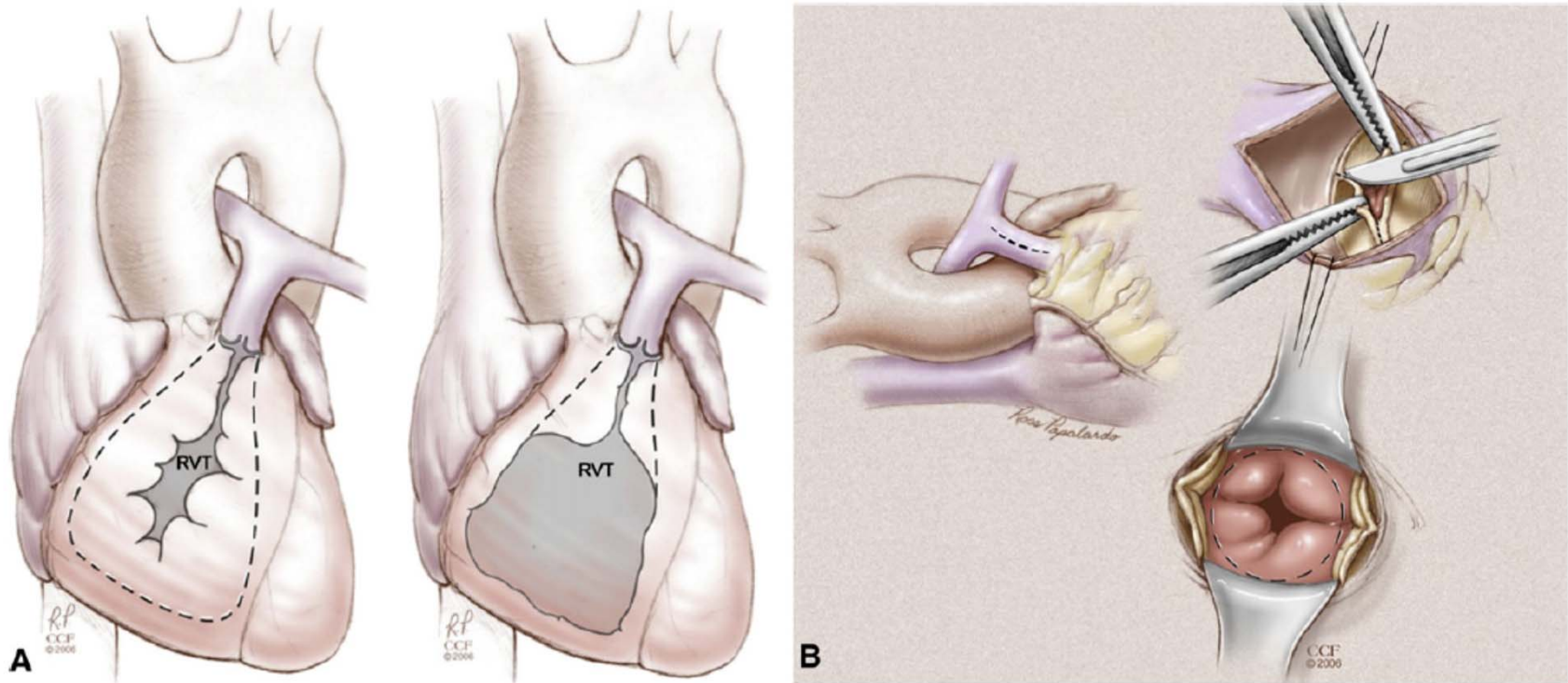
Sano S. et al.; Ann Thorac Surg 2000;70:1501-1506

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



a median age : 1.25 years
(15% at,0.4 years and 15% at.2.9 years)

Right ventricular myectomy is performed by means of a combined transatrial/transpulmonary approach



Briant III et al. J Thorac Cardiovasc Surg 2008;136:735-42

CONCLUSIONS

- **Catheter-based interventions rarely avoid surgical repair.**
- **The multistage palliation procedure to promote RV growth makes a definitive biventricular repair of PA with IVS**
- **Determination of approach should be based on scrutiny of the morbidity, mortality, and short- and long-term outcomes of the different approaches**