PA IVS with good RV anatomy
Transcatheter valvotomy

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- Good RV
  - TV z-score > - 2.5
  - TV/MV ratio ≥ 0.75
  - Tripartite
- Intermediate RV
  - TV z-score - 2.5 ~ - 5.0
  - TV/MV ratio 0.50 ~ 0.75
  - Bipartite RV : apical trabecular component is absent or attenuated
- Severely diminutive RV
  - TV z-score < - 5.0
  - TV/MV ratio < 0.5
  - Unipartite RV : only the inlet component

Perforation of the Atretic Pulmonary Valve

Long-Term Follow-Up

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OBJECTIVES We evaluated the long-term results of perforation of the pulmonary valve in patients with pulmonary atresia with an intact ventricular septum (PA-IVS).

BACKGROUND Interventional perforation of the pulmonary valve is considered the elective first stage treatment for PA-IVS, particularly in patients with a tripartite right ventricle (RV) and normal coronary circulation. However, the long-term results of this procedure are lacking.

METHODS Between January 1991 and December 2001, 39 newborns with a favorable form of PA-IVS underwent attempted perforation of the pulmonary valve. We evaluated the early and long-term outcomes.

RESULTS Median tricuspid and pulmonary *z* values were −1.2 and −2.4, respectively. Perforation was successful in 33 patients. Among them, 17 needed neonatal surgery, 13 did not need any surgery, and 3 had elective surgery after the first month of life. There were two procedure-related deaths, seven nonfatal procedural complications, and four postsurgical deaths. Compared with patients needing neonatal surgery, those having no or elective surgery had a higher incidence of a tripartite RV and a higher median tricuspid *z* value (92% vs. 53%, $p = 0.04$ and −1.7 vs. −0.5, $p = 0.03$). At a median follow-up of 5.5 years (range 0.5 to 11.5), survival was 85% and freedom from surgery was 35%. Five patients, four of whom had neonatal surgery, underwent a partial cavo-pulmonary connection.

CONCLUSIONS Our results show that this technique, although burdened by non-negligible mortality and morbidity, is effective in selected patients with a normal-sized RV. Preselection of patients allows interventional or surgical biventricular correction in the majority of cases. (J Am Coll Cardiol 2003;41:1399–403)

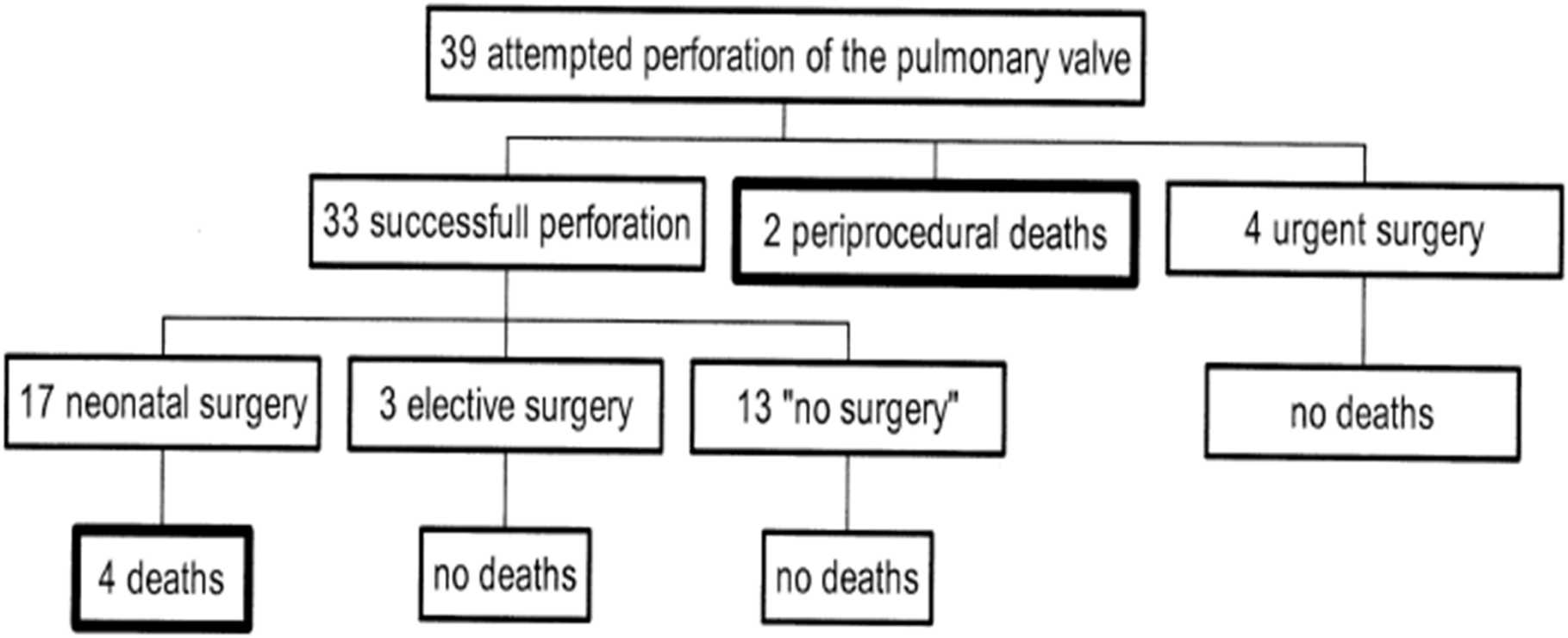

Figure 1. Flow chart summarizing the procedures and outcomes (death or no death) of patients.

- Perforation by guide wire in 20, by radiofrequency in 19
- 2 death due to infundibular perforation, 1 PE, 3 AF, 3 NEC
- 21 neonatal surgery:12 BTS, 8 RVOT patch+BTS, 1 RVOT patch after transcatheter intervention; 4 post-surgical death
- 2 more RVOT patch after neonatal period

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

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Background. Pulmonary atresia with intact ventricular septum (PAIVS) has a wide spectrum of anatomic heterogeneity and invokes a wide variety of treatment strategies. We reviewed the outcome of our patients with PAIVS in order to delineate strategies for the optimal management of PAIVS. In particular, the possibility of avoiding neonatal surgical intervention with catheter-based technology was assessed.

Methods. The study cohort was composed of all patients presented with PAIVS from January 1999 through December 2005. Demographic and anatomic variables were analyzed to determine association with in-hospital mortality.

Results. Forty-four infants with PAIVS underwent catheter valvuloplasty (n = 17) and (or) surgical intervention (n = 42). The mean age and weight of the infants was six days and 3.1 kg, and the average follow-up was 40 ± 29.5 months. Five (11%) had right ventricle dependent coronary circulation (RVDCC) and six (14%) had Ebstein's anomaly. Five (11%) patients died. Of those who underwent catheter valvotomy, three (18%) underwent shunt placement, 12 (71%) underwent right ventricular outflow tract reconstruction with shunt placement, and only two (12%) did not require a further surgical intervention in the newborn period. Multivariable analyses demonstrated RVDCC (odds ratio 21.3, $p = 0.025$) and Ebstein's anomaly (odds ratio 16.0, $p = 0.038$) to be risk factors for in-hospital mortality. Of those patients with Ebstein's anomaly, a single ventricle approach had a better outcome.

Conclusions. We demonstrated excellent recent outcomes for patients with PAIVS. Catheter-based interventions rarely avoid surgical repair. The RVDCC and Ebstein's anomaly were associated with high mortality. In patients with Ebstein's anomaly, single ventricular pathway may be the better strategy for this specific patient population.

(Ann Thorac Surg 2007;84:574–80)

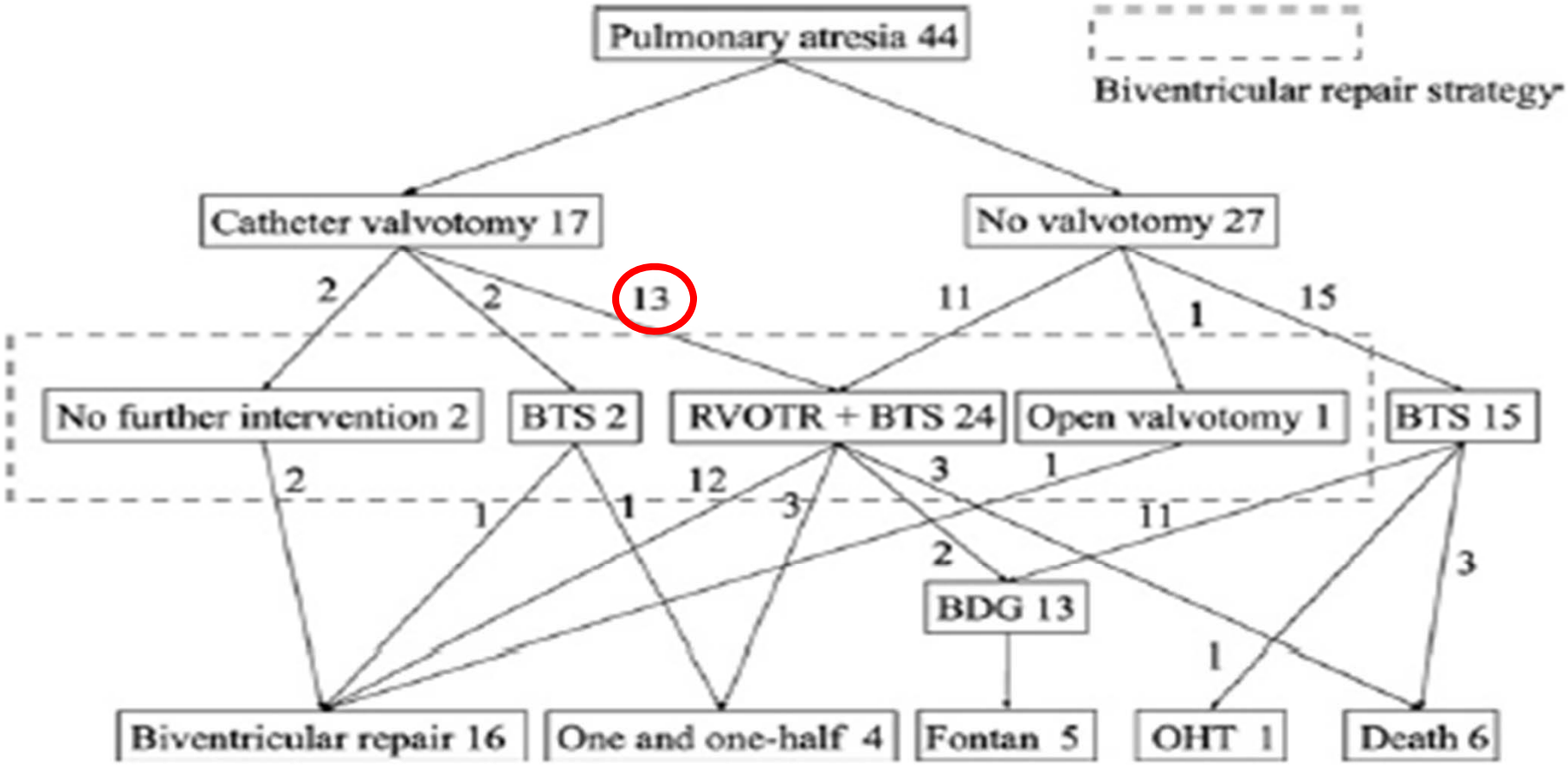

Fig 1. Flow chart of outcome for patients with pulmonary atresia-intact ventricular septum. Box with broken line shows the patients with biventricular repair strategy. (BDG = bidirectional Glenn operation; BTS = modified Blalock-Taussig shunt; OHT = orthotopic heart transplantation; RVOTR = right ventricular outflow tract reconstruction.)

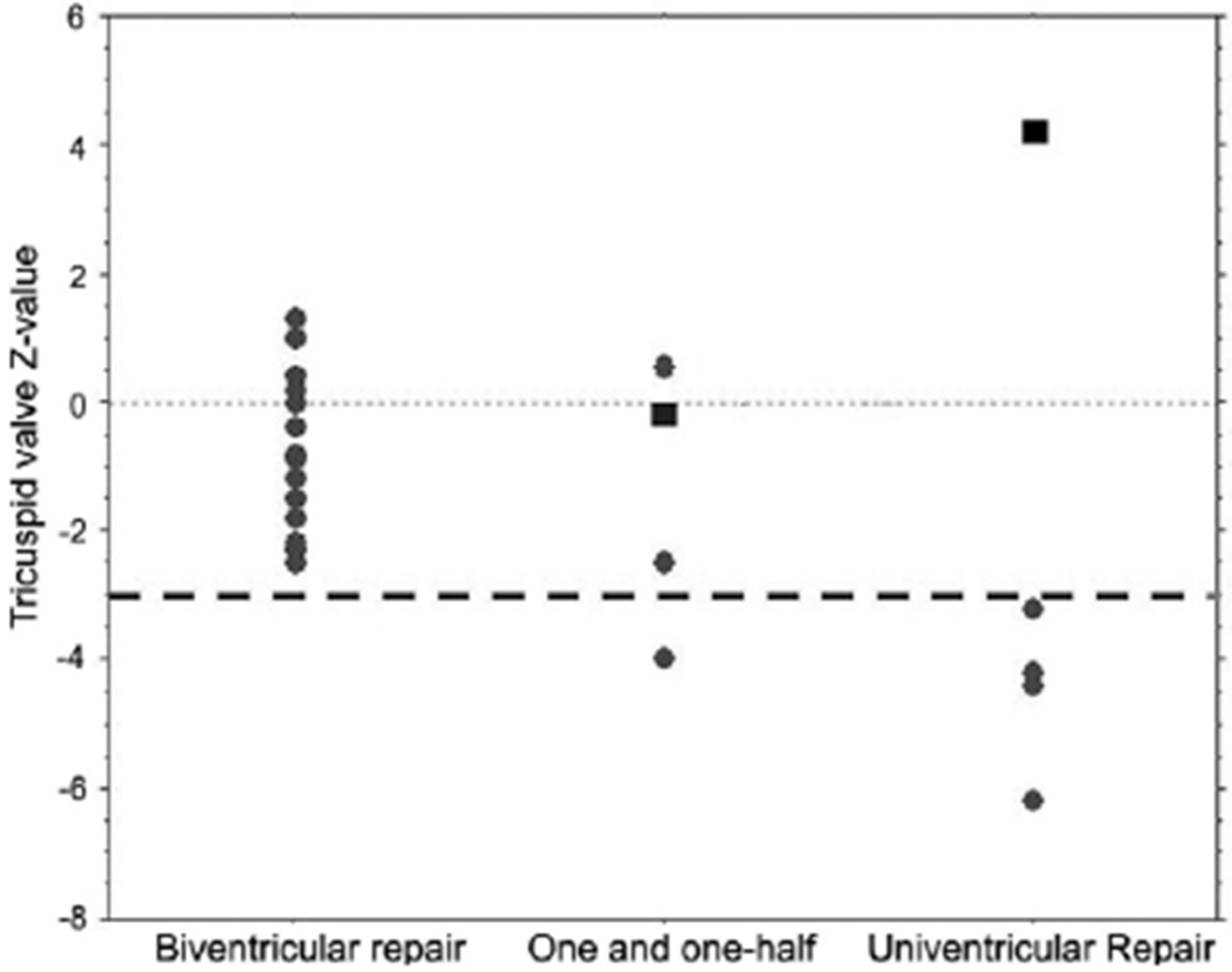

Fig 4. Definitive repair status and the tricuspid valve Z value. (● = pulmonary atresia with intact ventricular septum [PAIVS]; ■ = PAIVS-Ebstein.)

Comment

dies suggest that increasing numbers of undergoing transcatheter pulmonary valvuloplasty for PAIVS and advocate this as the initial procedure [10, 18]. Others have advocated stenting of the ductus arteriosus as well as transcatheter pulmonary valvuloplasty [19]. We believe that these procedures are largely ineffective because they cannot adequately address the intricacies of this disease (pulmonary valve annular hypoplasia, subpulmonary obstruction, and RV noncompliance). Although pulmonary valvuloplasty can relieve obstruction caused primarily by abnormal pulmonary valve leaflets, it cannot enlarge the pulmonary valve annulus. Therefore, these patients with significant pulmonary valve annular obstruction will ultimately require surgical intervention. Indeed, we were not able to evaluate the growth potential of the pulmonary valve annulus in this cohort after catheter valvuloplasty because most of the patients required surgical intervention within the first month. Furthermore, catheter intervention cannot relieve subpulmonary muscular obstruction, a critical

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Core Curriculum

Management Algorithm in Pulmonary Atresia With Intact Ventricular Septum

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Pulmonary atresia with intact ventricular septum (PAIVS) is a disease with remarkable morphologic variability, affecting not only the pulmonary valve but also the tricuspid valve, the RV cavity and coronary arteries. With advances in interventional techniques and congenital heart surgery, the management of PAIVS continues to evolve. This review is an attempt at providing a practical approach to the management of this disease. The basis of our approach is morphologic classification as derived from echocardiography and angiography. Group A, patients with good sized RV and membranous atresia, the primary procedure at presentation is radiofrequency (RF) valvotomy. Often it is the only procedure required in this group with the most favourable outcome. Patients with severely hypoplastic RV (Group C) are managed along the lines of hearts with single ventricle physiology. The treatment at presentation is patent ductus arteriosus (PDA) stenting with balloon atrial septostomy or conventional modified Blalock Taussig (BT) shunt. Bidirectional Glenn shunt may be done 6–12 months later followed by Fontan completion after a suitable interval. Patients in Group B, the intermediate group, are those with borderline RV size, usually with attenuated trabecular component but well developed infundibulum. The treatment at presentation is RF valvotomy and PDA stenting ± balloon atrial septostomy. Surgical re-interventions are not uncommonly required viz. bidirectional Glenn shunt when the RV fails to grow adequately (1½ – ventricle repair) and right ventricular outflow tract (RVOT) reconstruction for subvalvar obstruction or small pulmonary annulus. Catheter based interventions viz. repeat balloon dilatation or device closure of patent foramen ovale (PFO) may also be required in some patients.

Key words: membranous atresia; RV hypoplasia; coronary

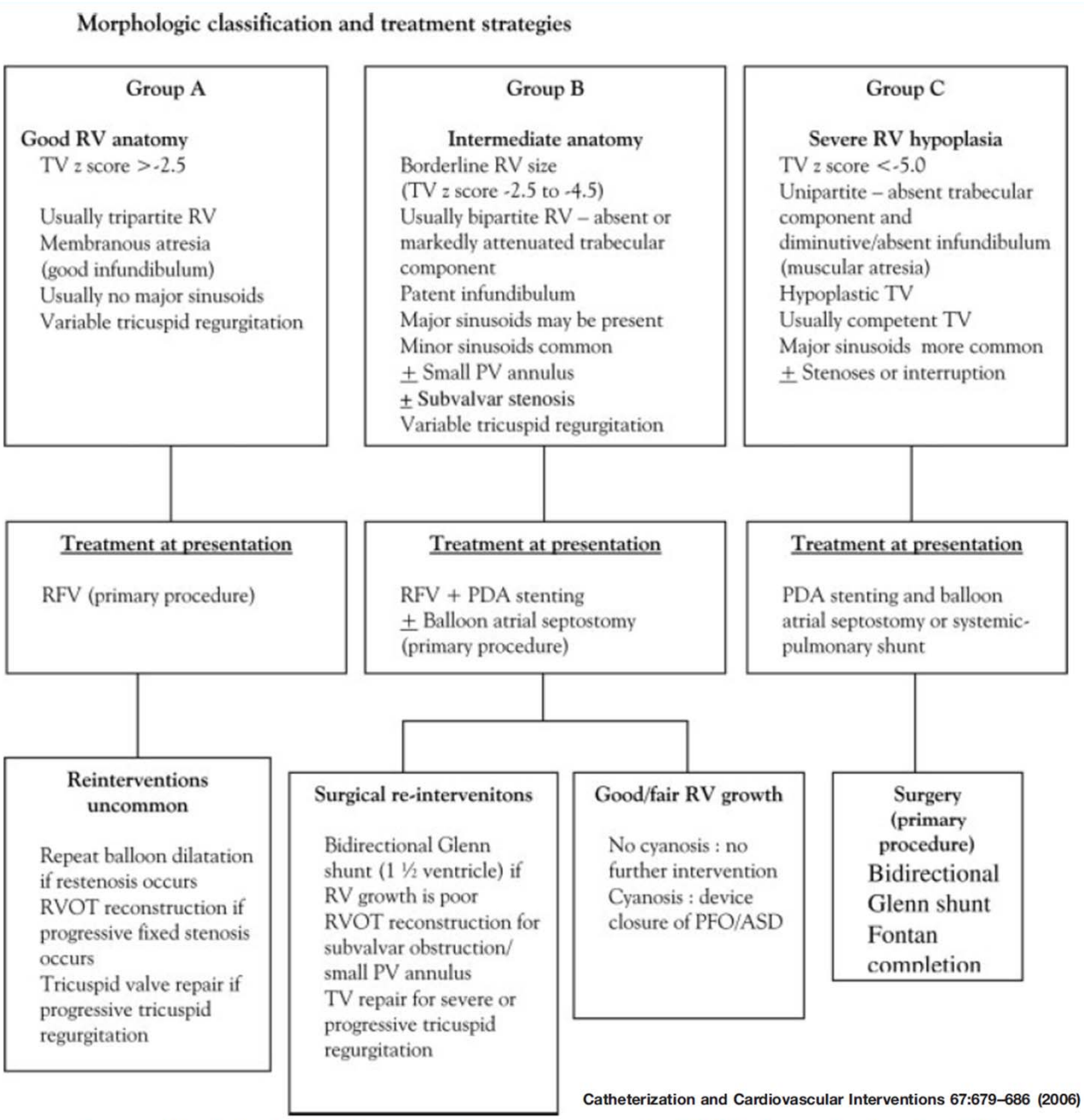

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Fig. 5. Morphologic classification and treatment strategies. PAIVS with markedly dilated RV, RVOT, and RA with dysplastic/Ebstein's malformation of the tricuspid valve and severe tricuspid regurgitation is a rare morphologic subtype which is excluded from this classification scheme.

Concomitant stenting of the patent ductus arteriosus and radiofrequency valvotomy in pulmonary atresia with intact ventricular septum and intermediate right ventricle: Early in-hospital and medium-term outcomes

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Objectives: Our objective was to determine the feasibility and early to medium-term outcome of stenting the patent ductus arteriosus at the time of radiofrequency valvotomy in the subgroup of patients with pulmonary atresia with intact ventricular septum and intermediate right ventricle.

Background: Stenting of the patent ductus arteriosus and radiofrequency valvotomy have been proposed as the initial intervention for patients with intermediate right ventricle inasmuch as the sustainability for biventricular circulation or 1½-ventricle repair is unclear in the early period.

Methods: Between January 2001 and April 2009, of 143 patients with pulmonary atresia and intact ventricular septum, 37 who had bipartite right ventricle underwent radiofrequency valvotomy and stenting of the patent ductus arteriosus as the initial procedure. The mean tricuspid valve z-score was –3.8 ± 2.2 and the mean tricuspid valve/mitral valve ratio was 0.62 ± 0.16.

Results: Median age was 10 days (3–65 days) and median weight 3.1 kg (2.4–4.9 kg). There was no procedural mortality. Acute stent thrombosis developed in 1 patient and necessitated emergency systemic–pulmonary shunt. There were 2 early in-hospital deaths owing to low cardiac output syndrome. One late death occurred owing to right ventricular failure after the operation. Survival after the initial procedure was 94% at 6 months and 91% at 5 years. At a median follow-up of 4 years (6 months to 8 years), 17 (48%) attained biventricular circulation with or without other interventions and 9 (26%) achieved 1½-ventricle repair. The freedom from reintervention was 80%, 68%, 58%, and 40% at 1, 2, 3, and 4 years, respectively.

Conclusions: Concomitant stenting of the patent ductus arteriosus at the time of radiofrequency valvotomy in patients with pulmonary atresia with intact ventricular septum and intermediate right ventricle is feasible and safe with encouraging medium-term outcome. (J Thorac Cardiovasc Surg 2011;■:1-7)

- 143 PA IVS
  - 45 good RV : RFV only
  - 61 severely diminutive RV : PDA stent only
  - 37 intermediate RV

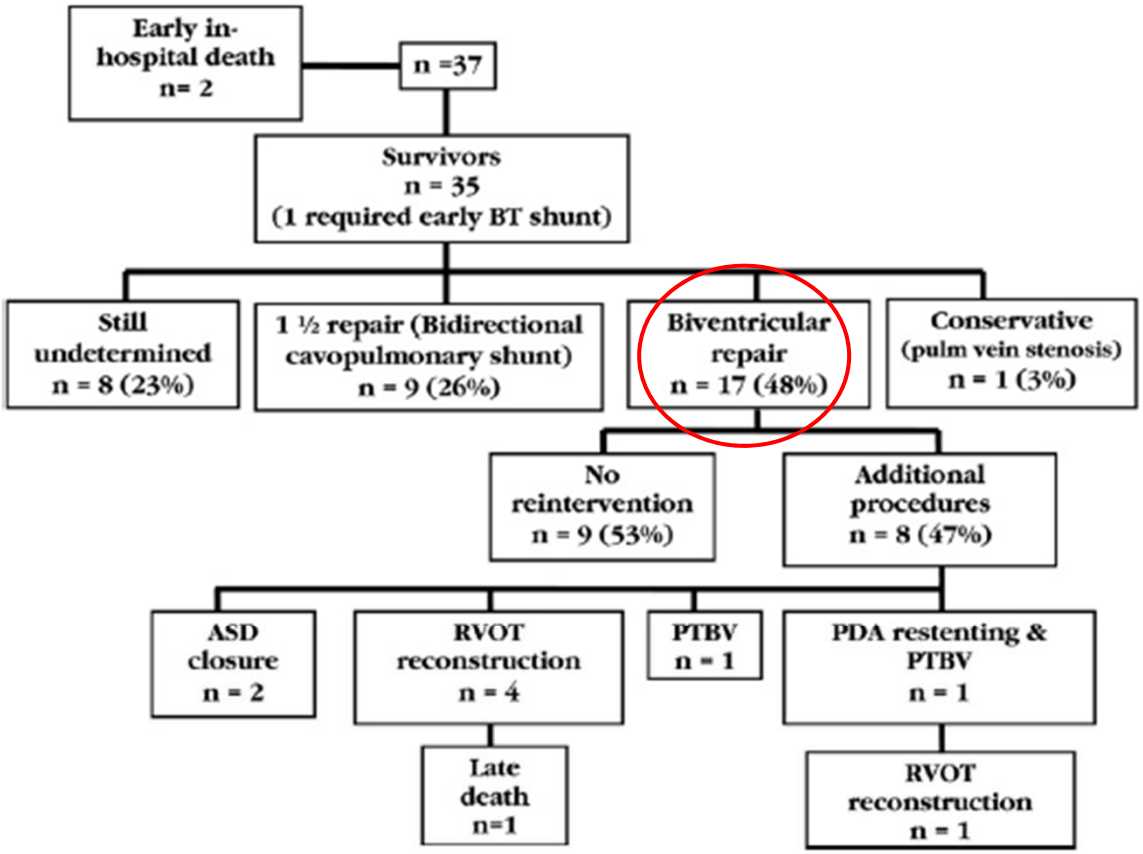


FIGURE 1. Outcome of patients with intermediate right ventricle *(RV). BT,* Blalock–Taussig shunt; *ASD,* atrial septal defect; *RVOT,* right ventricular outflow tract; *PTBV,* Percutaneous transcatheter balloon valvuloplasty; *PDA,* patent ductus arteriosus.

Experience of PA IVS in PNUH/PNUCH

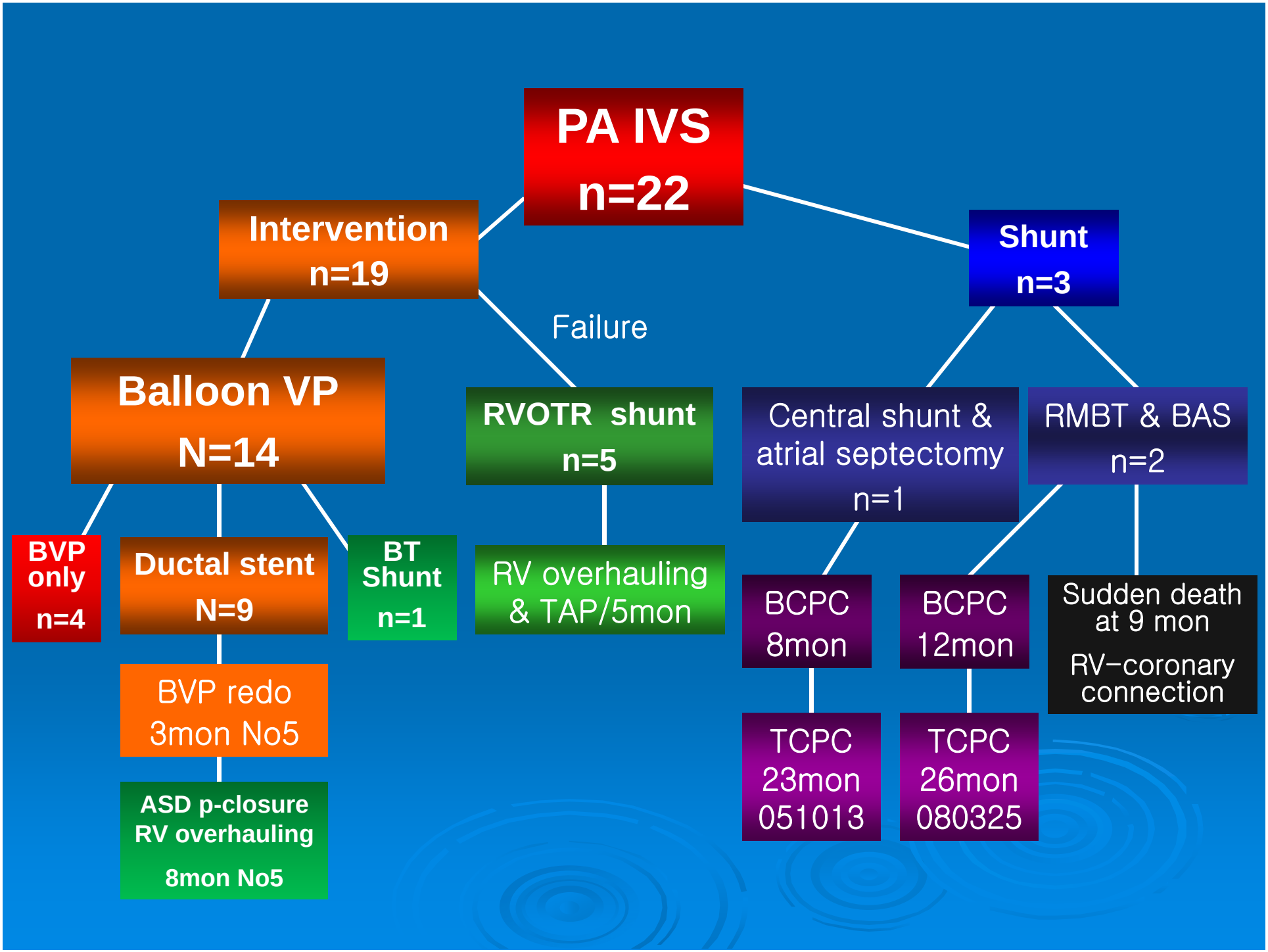
이형두 박지애 조민정 성시찬 김시호 김형태

Patients and Method

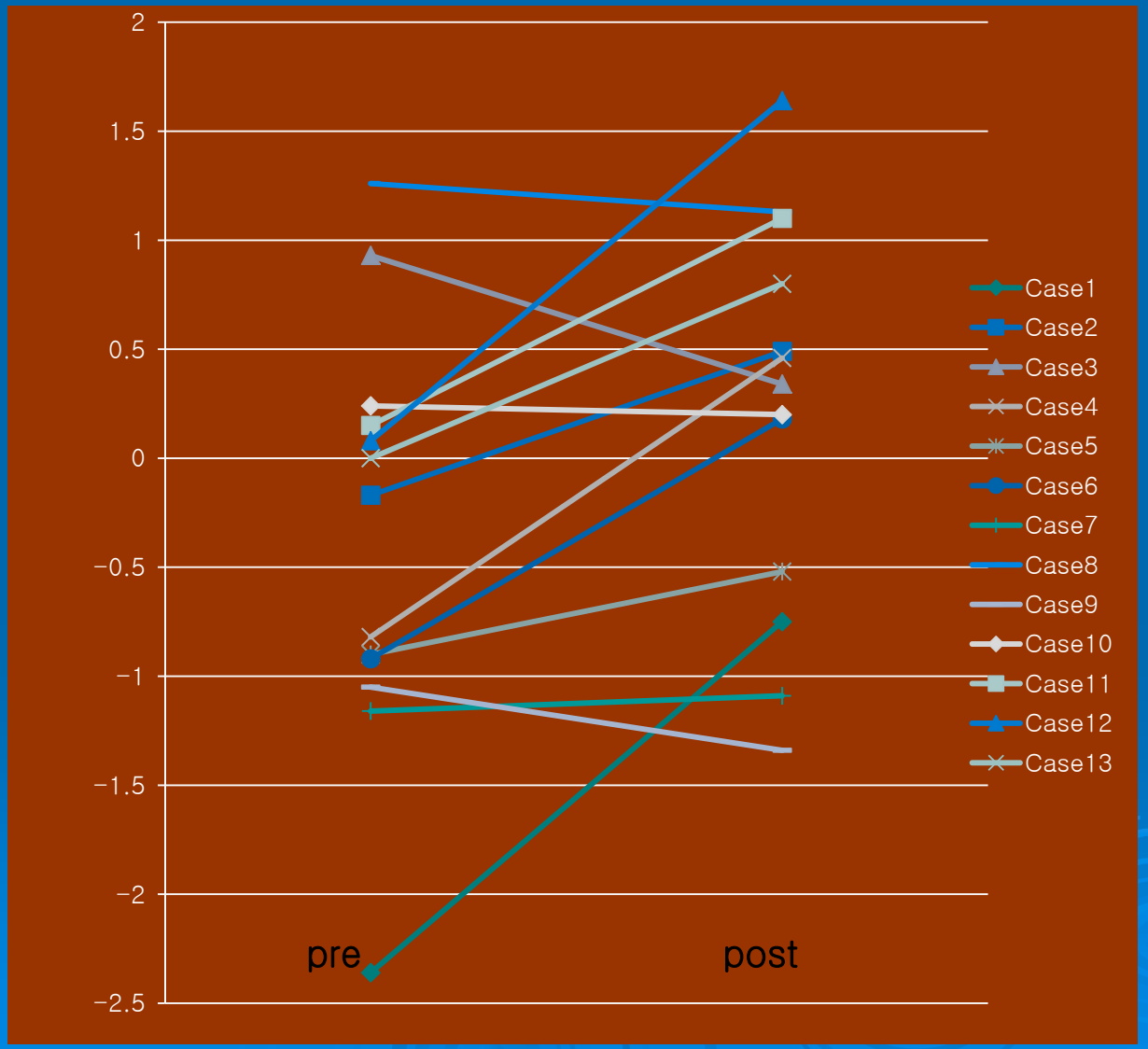
- 22 patient with PA IVS have been treated between Jan. 2004 and Mar. 2011.
- We tried transcatheter guidewire perforation and staged balloon pulmonary valvotomy in 19 patients but successful in 14 cases.
- As an initial procedure, we performed BT shunt in 2 patients and central shunt/atrial septectomy in one infant, who had marked hypoplasia of TV and RV±RV-coronary connection.

Results

- F/U 5 ~ 68 mon (median 28)
- Initial TV z value
  - Valvotomy only(n=4) : 0.00~0.24(m 0.12)
  - Valvotomy + stent(n=9) : -2.36~1.26(m -0.58)
  - Valvotomy + BT shunt(n=1) : -1.05
- Residual RVOT gradient on F/U
  - Valvotomy only(n=4) : 7.3~17.6
  - Valvotomy + stent(n=9) : 10~48 (med 13.7)
  - Valvotomy + BT shunt(n=1) : 19 mmHg
- Fate of stent
  - 3 ductal stent closed spontaneously : confirmed patency at 22, 11, 8 months after stenting
  - Others patent at 5~68 months(median 43) after implantation


PA IVS
n=22
Intervention
n=19
Shunt
n=3
Failure
Balloon VP
N=14
RVOTR shunt
n=5
Central shunt &
atrial septectomy
n=1
RMBT & BAS
n=2
BVP
only
n=4
Ductal stent
N=9
BT
Shunt
n=1
RV overhauling
& TAP/5mon
BCPC
8mon
BCPC
12mon
Sudden death
at 9 mon
RV-coronary
connection
BVP redo
3mon No5
TCPC
23mon
051013
TCPC
26mon
080325
ASD p-closure
RV overhauling
8mon No5

Changes of TV z value



Results : Complications

- Transfusion in 2 cases
- Femoral arterial thrombosis in 3 patients which were resolved completely with systemic/local urokinase infusion.
- Stent migration to main pulmonary artery in No 4 patient, positioned at infrarenal segment of IVC.
- IVC perforation occurred during balloon valvotomy in a neonate, healed with manual compression.

Conclusion

- Transcatheter balloon valvotomy with or without ductal stenting is an attractive alternative to surgical treatment esp in relatively good RV.
- Although balloon pulmonary valvotomy may not prevent surgery, delaying surgical intervention, esp CPB, past the first month maybe beneficial.


Are you a surgeon?
I am a interventional cardiologist!

My Choice

for 2wks old neonates with PA IVS born at full term with 2.8kg of birth weight

tripartite RV with hypertrophy

TV z value of -2.0

Transcatheter valvotomy

+/− PDA Stent