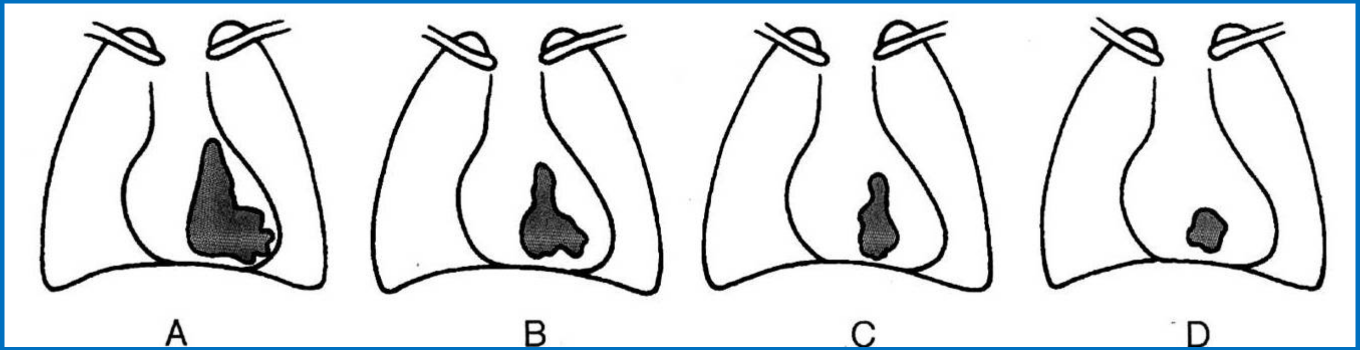
Transcatheter therapy for pulmonary atresia intact ventricular septum:
indication, technique and outcome

부산의대 소아과
이 형 두

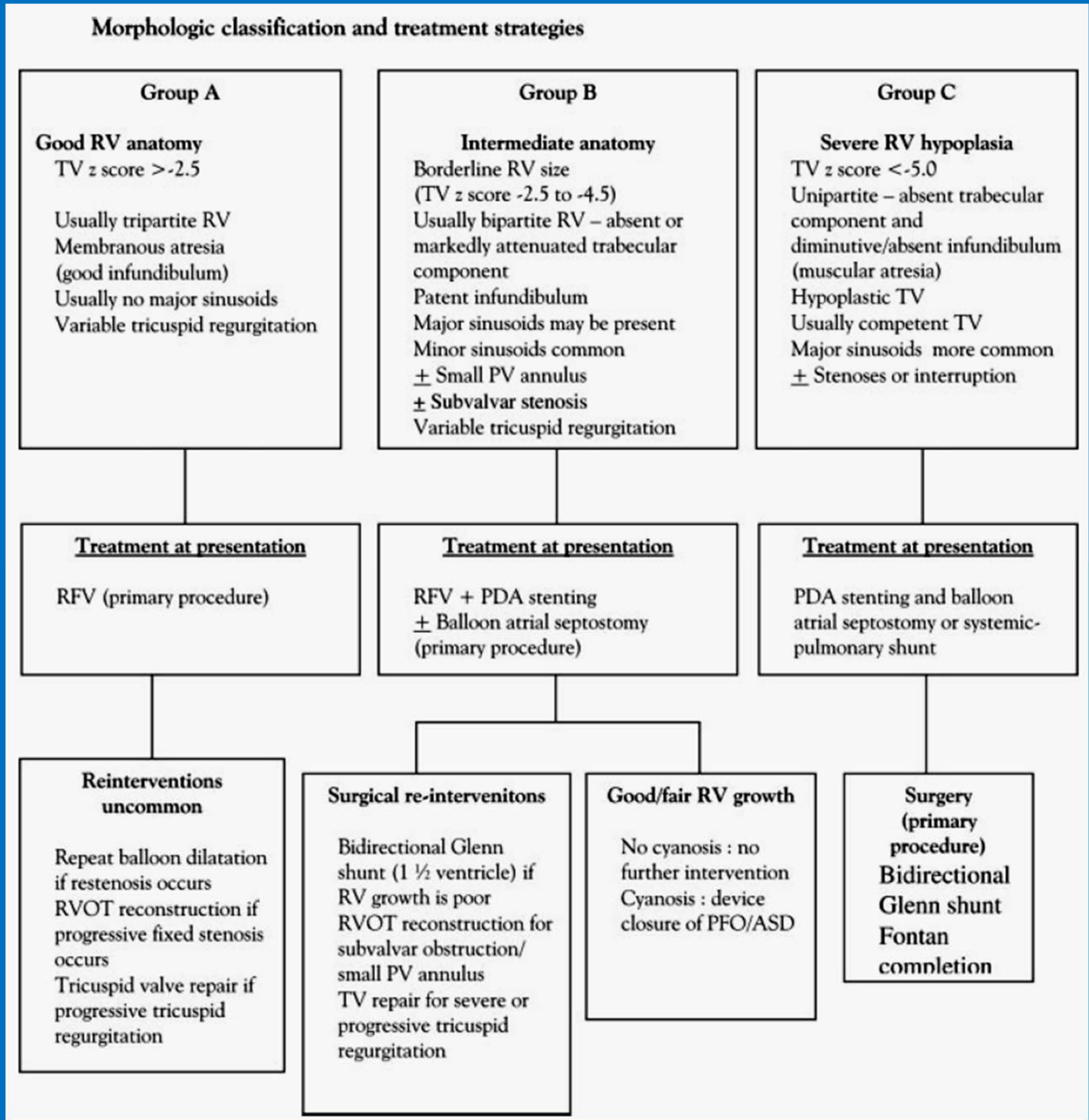
Transcatheter therapy for PA IVS: Indication

- **Patient Selection**
- TV z-score > -2.5, TV/MV ratio ≥ 0.75, tripartite RV : good RV
- TV z-score -2.5 to -5.0, and/or a TV/MV ratio 0.50 to 0.75, bipartite RV : intermediate
- TV z-score < -5.0, TV/MV ratio < 0.5, unipartite RV : severely diminutive

Alwi M JTCS 2011;141:1355-61



Transcatheter therapy for PA IVS: Indication



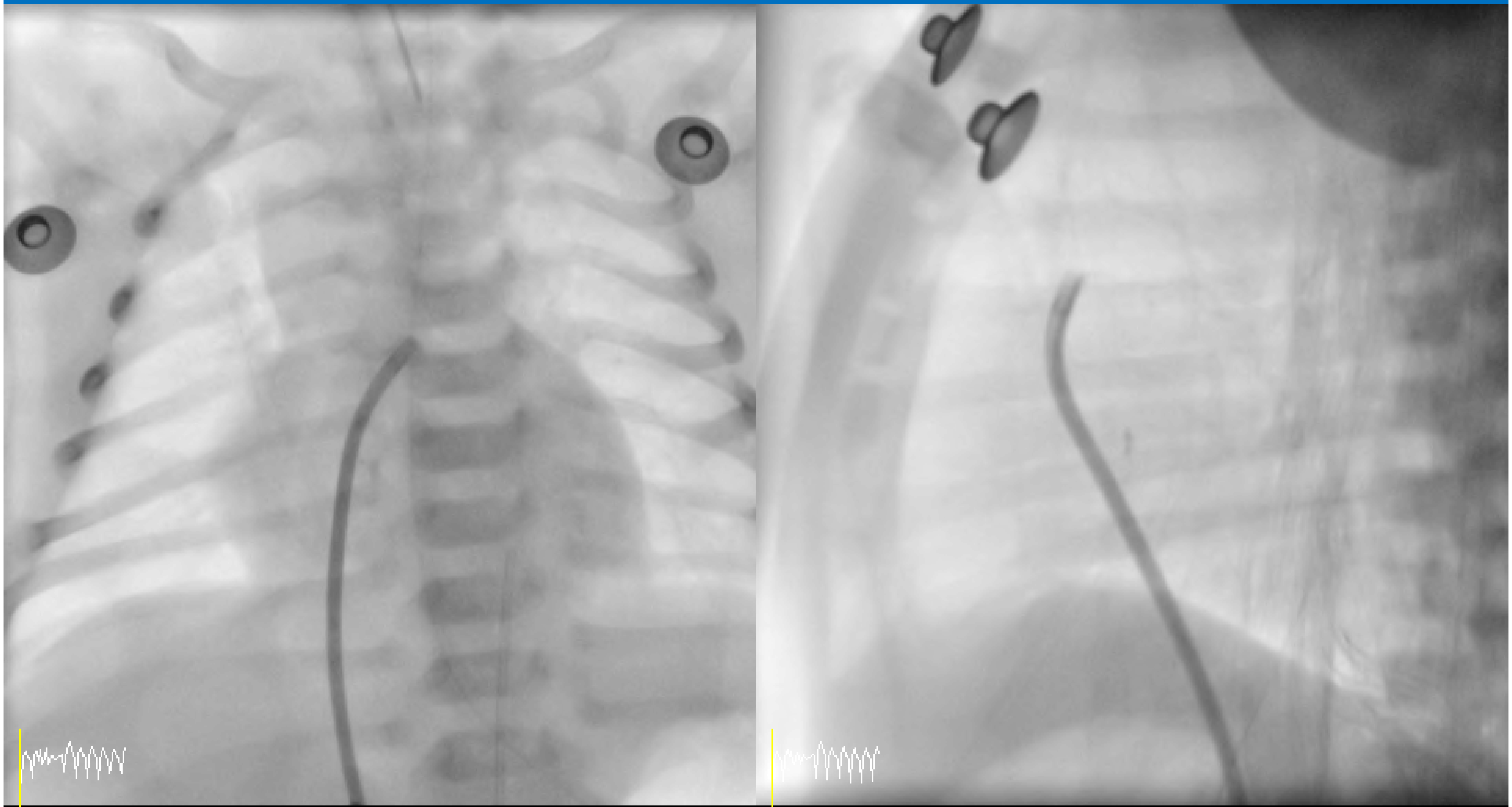
Catheter Cardiovasc Interv 2006;67: 679-86

Transcatheter therapy for PA IVS: Technique

- Sedation or general anesthesia
- Access femoral vessels
  - Vein
  - Artery
- Insertion 4 Fr JR coronary or cobra catheter
- Recording of pressure : RV, aorta, MPA
- Angiography : evaluate suitability
  - 2~3ml nonionic contrast medium
  - AP and lateral projection
  - Measure infundibulum, P valve, MPA

Transcatheter therapy for PA IVS: Technique

- Define landmark of P valve
  - Levo-phase of RV angiogram
  - Simultaneous angiography through V and A catheters
  - Positioning of 5 or 10 mm loop gooseneck snare through the duct
  - Positioning of catheter in aorta side within ant sinus of MPA
- Position catheter appropriately in RVOT
  - 4 Fr JR coronary catheter, 4 Fr cobra catheter or 5F Multipurpose catheter (Cordis) + 0.025 in. tip-deflecting wire with a 5 mm curve (Cook)
  - Catheter in RV : contact with P valve
- Options for catheter valvotomy
  - 0.014" guidewire : floppy or stiff end
  - Radiofrequency wire
  - Hybrid



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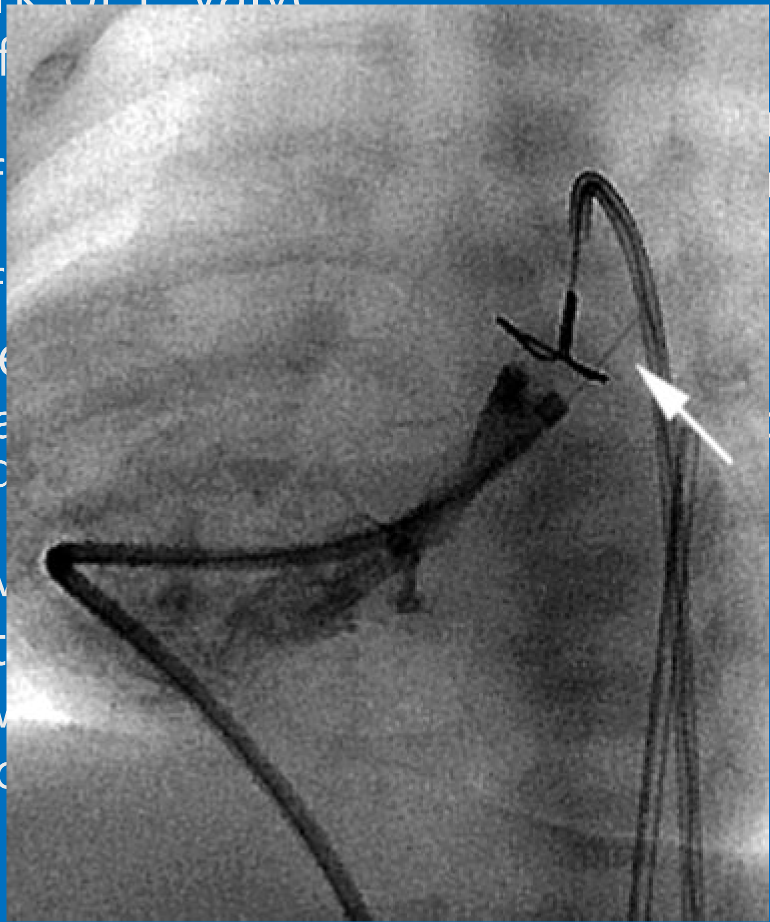

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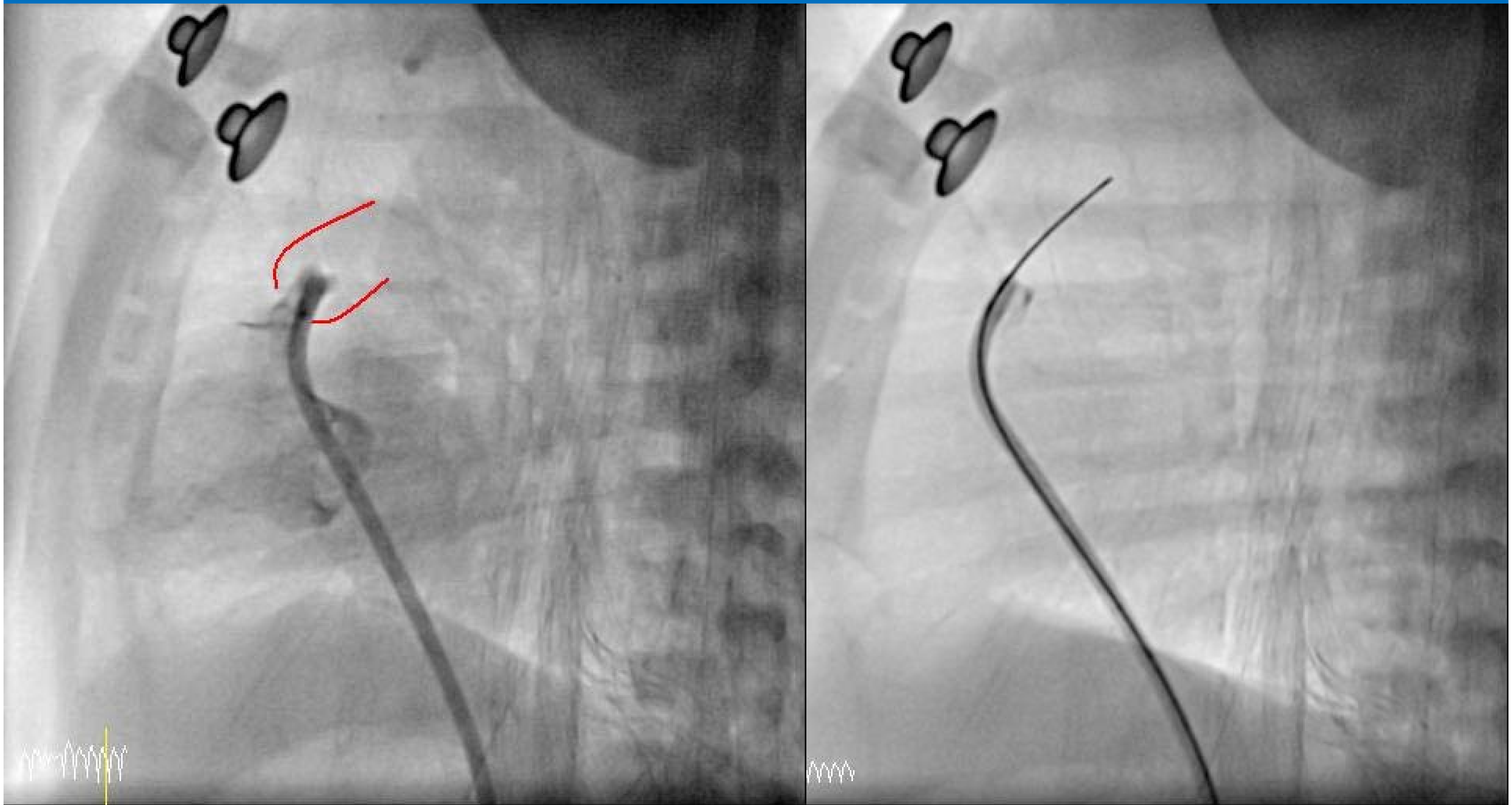
Transcatheter therapy for PA IVS: Technique

- **Catheter valvotomy using guidewire**



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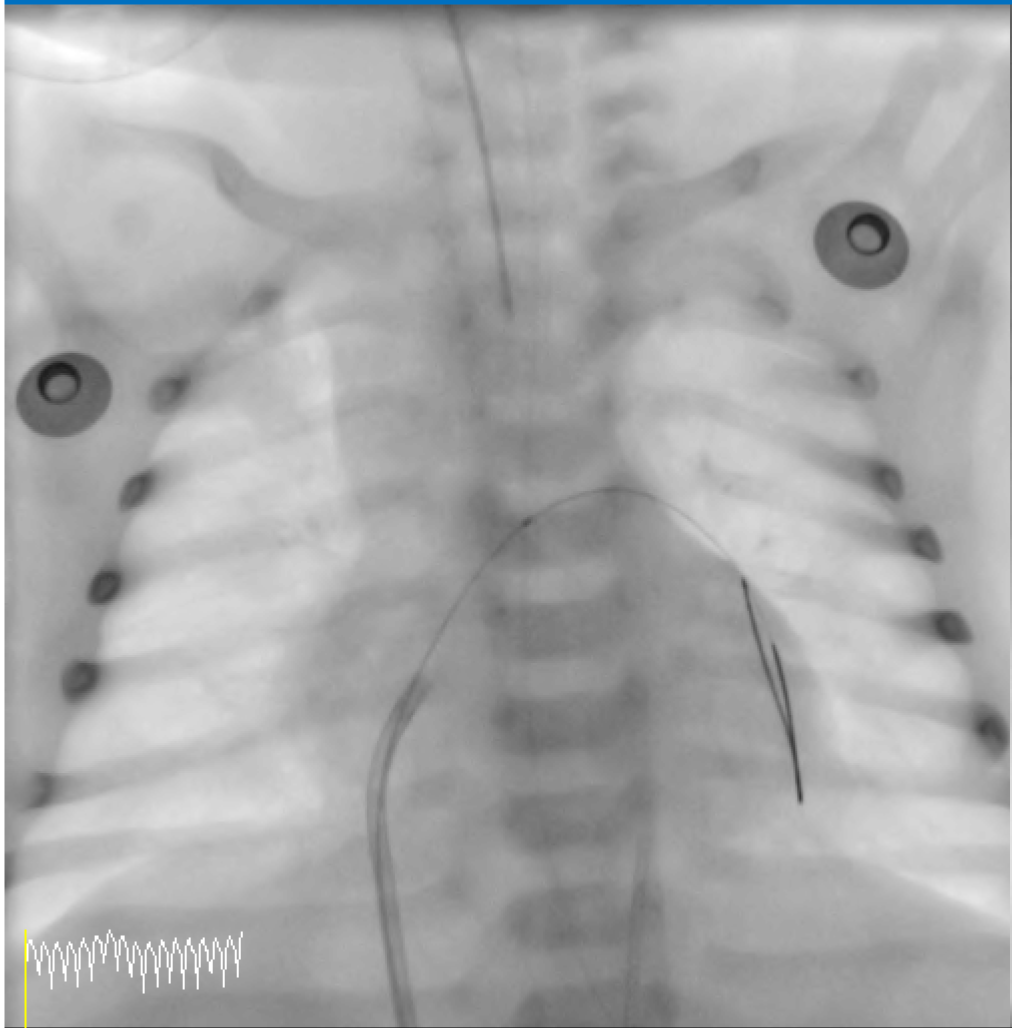

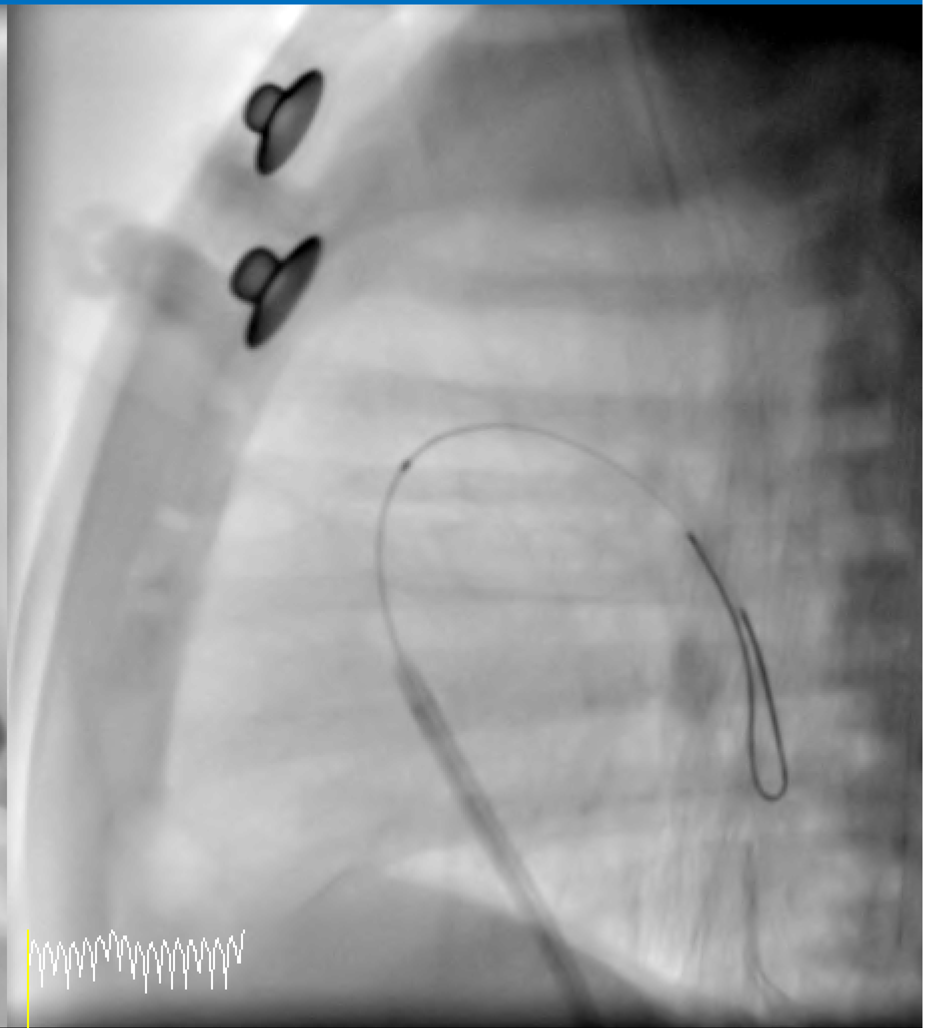

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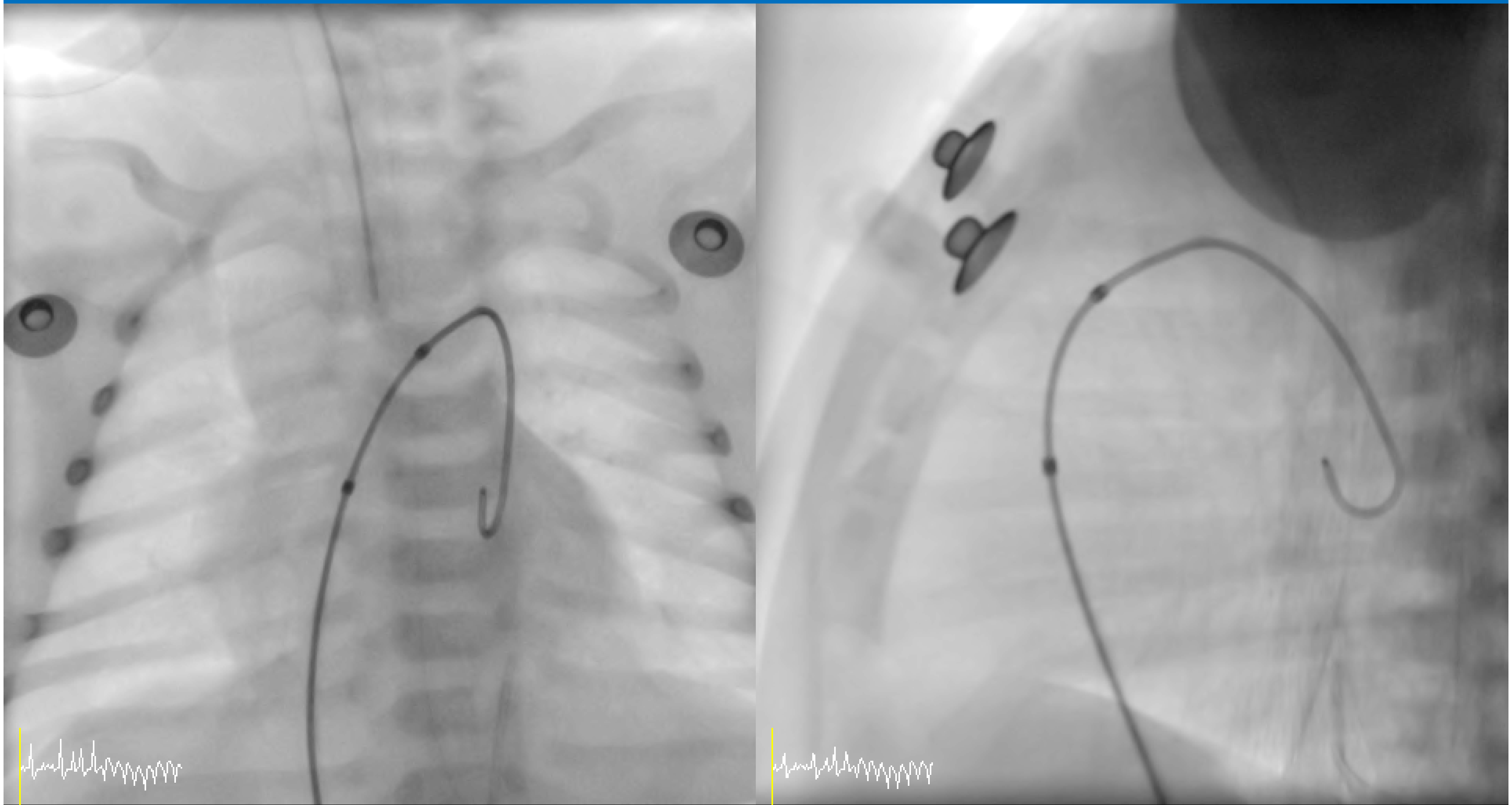

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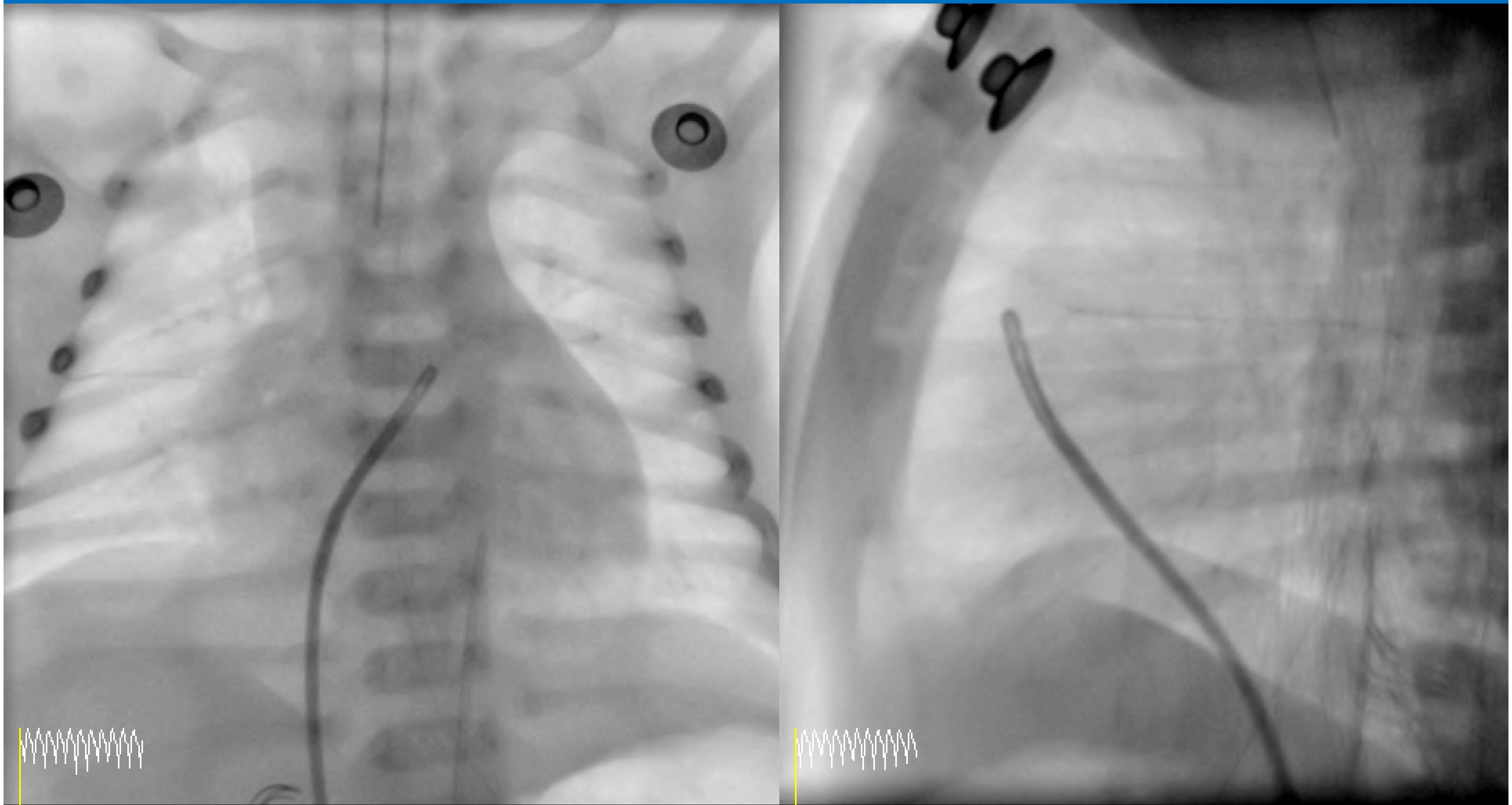
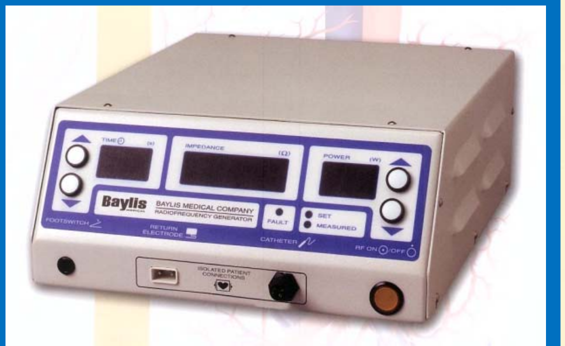
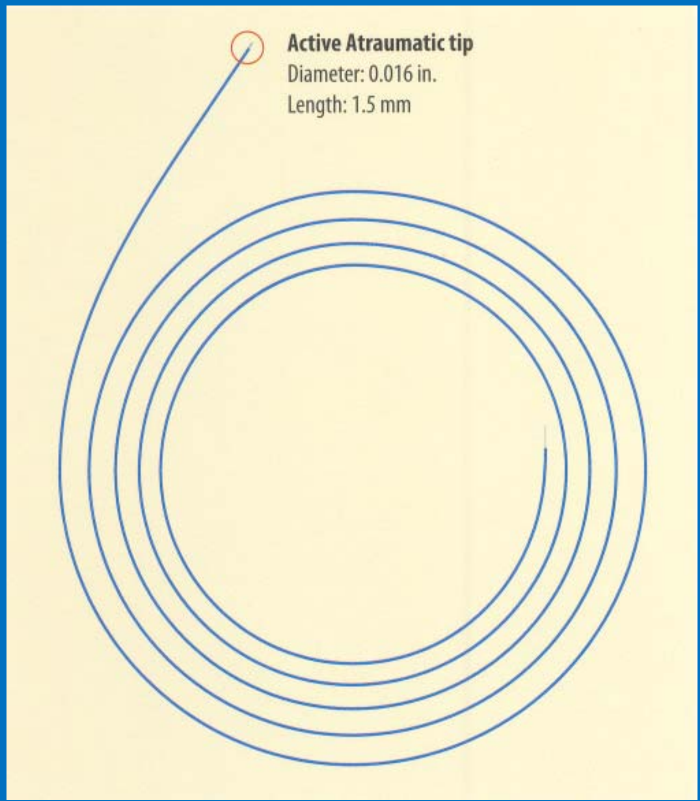
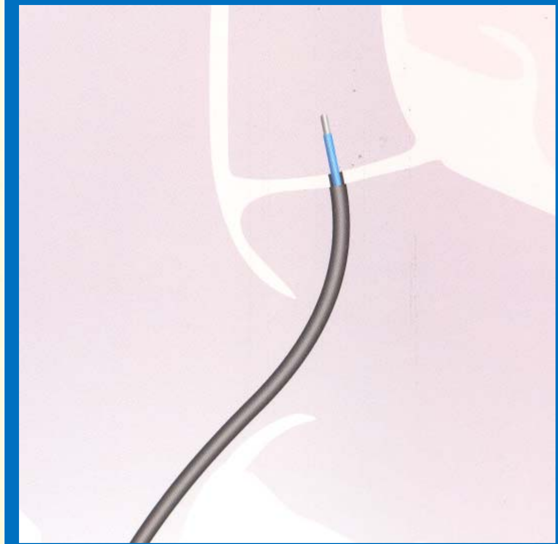


Transcatheter therapy for PA IVS: Technique

- **Catheter valvotomy using radiofrequency catheter**
- Nykanen Radio Frequency Perforation Catheter
  (Baylis Medical Company Inc)

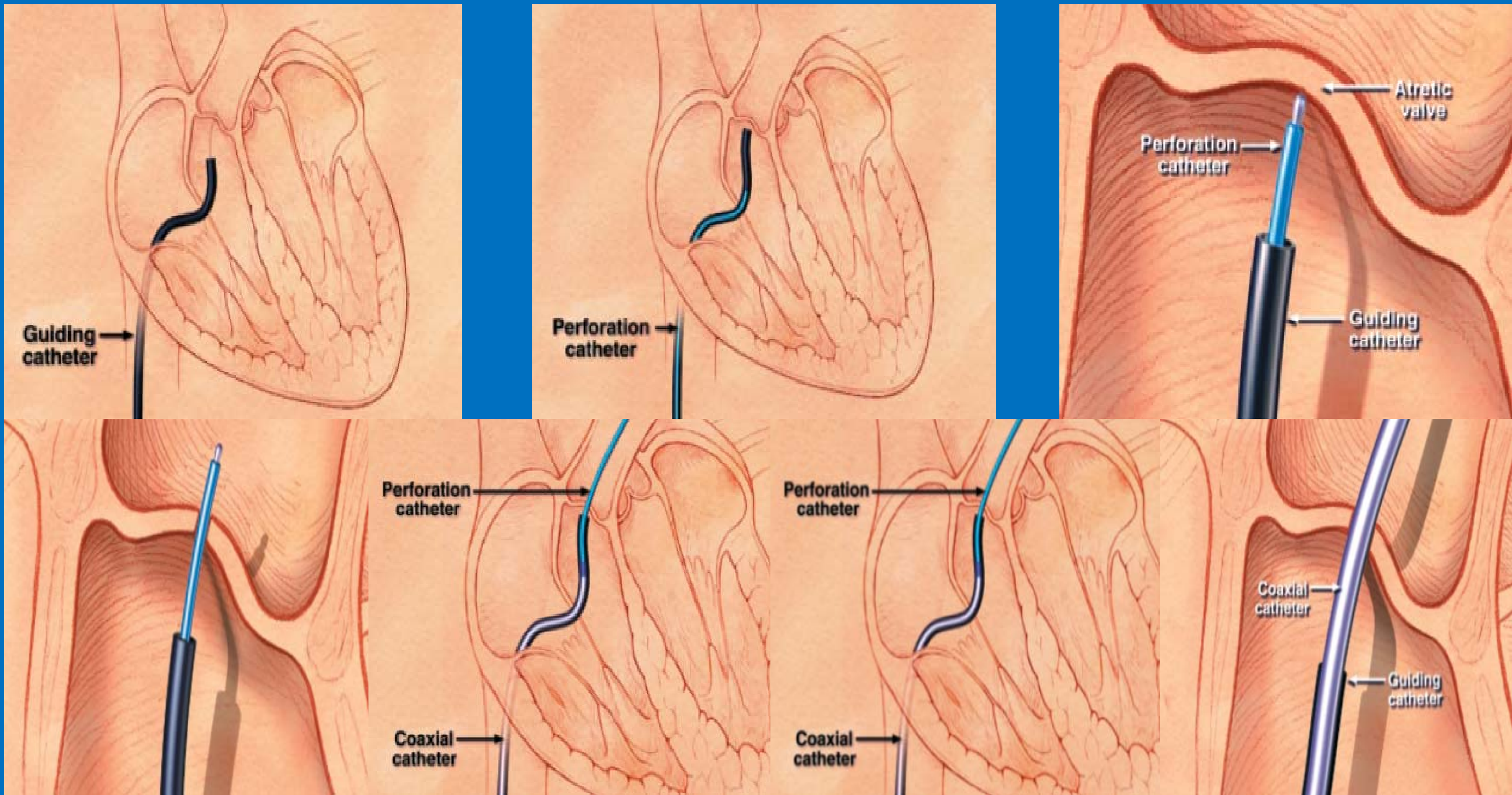


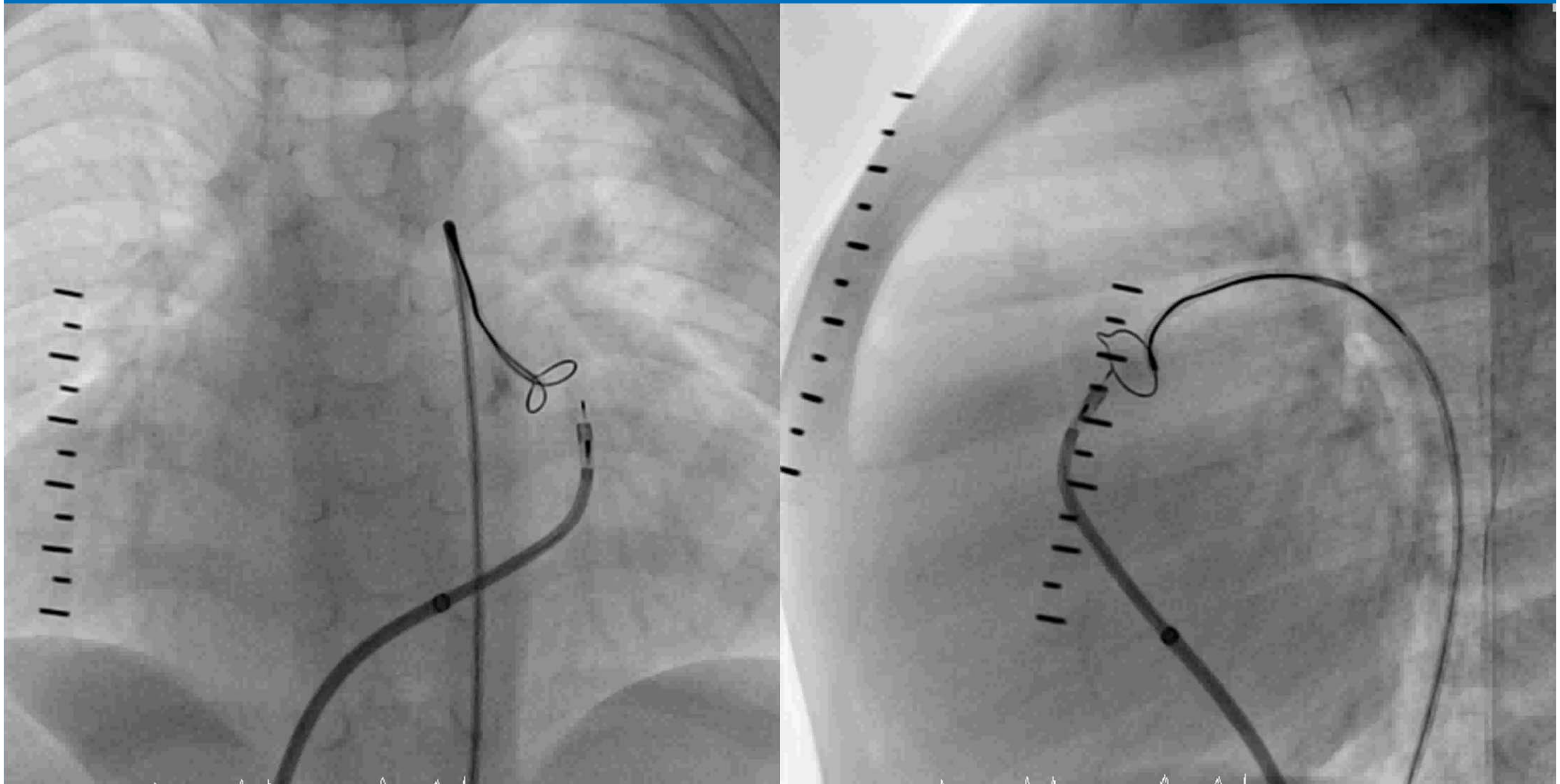




Transcatheter therapy for PA IVS: Technique

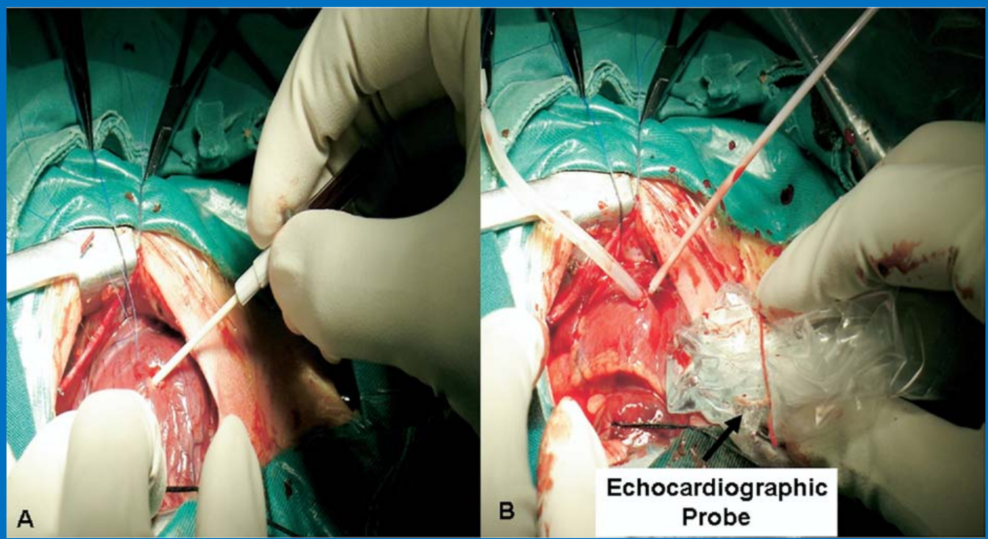
- **Catheter valvotomy using radiofrequency catheter**





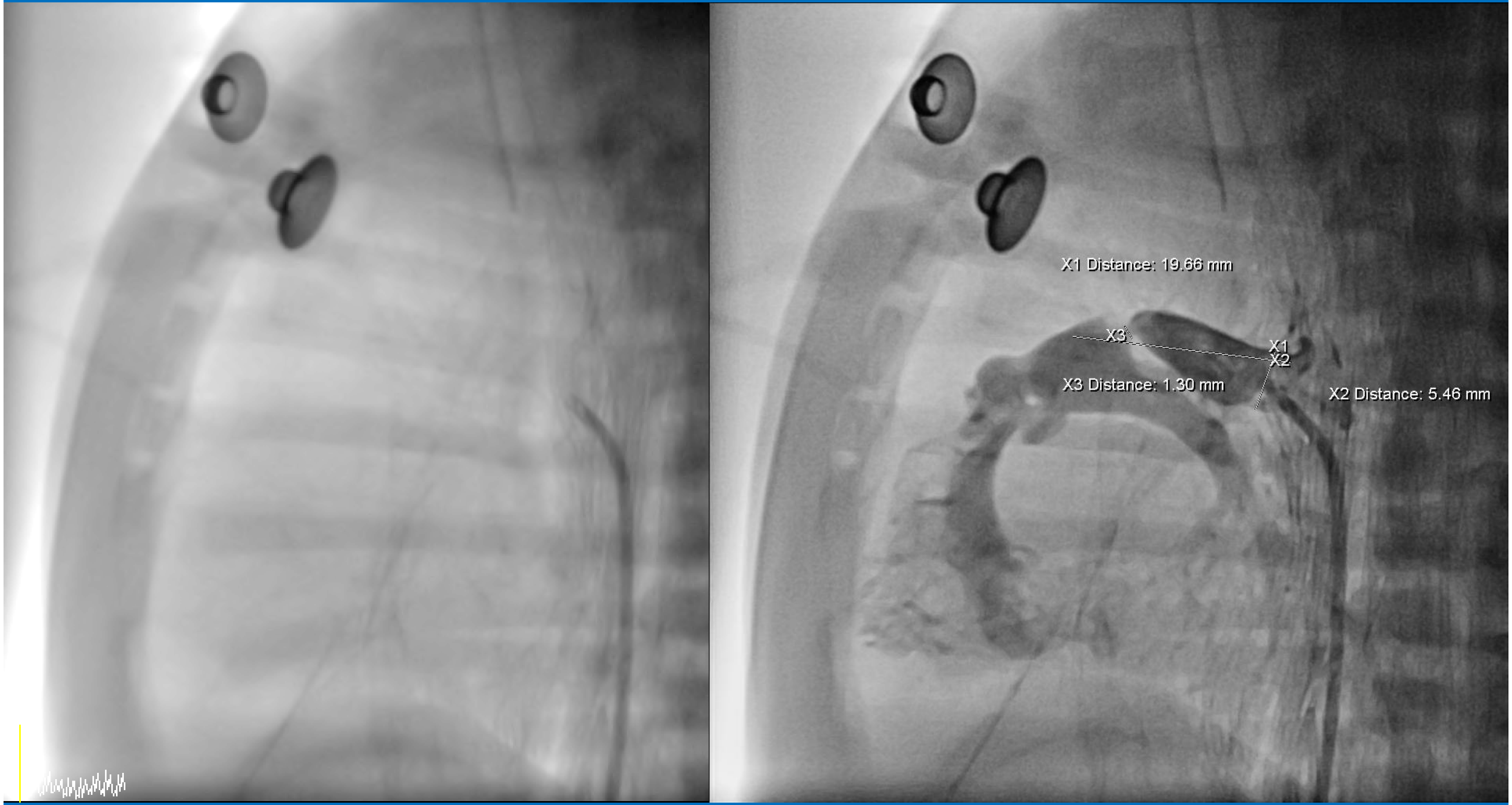
Transcatheter therapy for PA IVS: Technique

- **Catheter valvotomy using hybrid therapy**
  - Midline sternotomy
  - Purse-string suture in RVOT 2 cm away from pulmonary trunk
  - Subxiphoid echo to capture atretic pulmonary valve
  - Perventricular puncture and perforate PV with 16G IV catheter
  - Guidewire insertion
  - Sequential balloon dilations with guidance of epicardial echo
  - 3.5-mm modified Blalock–Taussig (BT) shunt



Transcatheter therapy for PA IVS: Technique

- **Ductus stenting**
  - SpO2 < 80~85%



Transcatheter therapy for PA IVS: Technique

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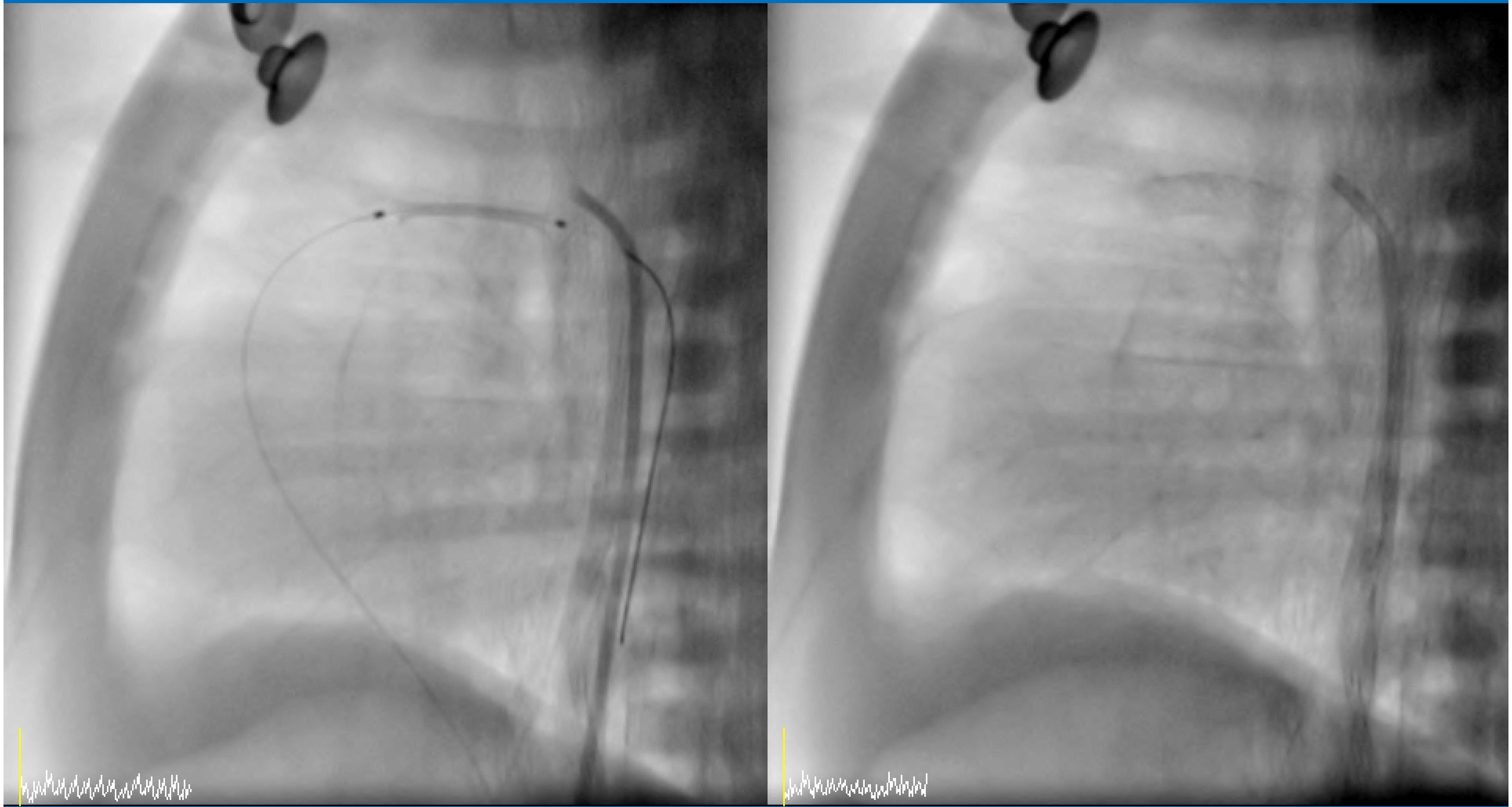

Transcatheter therapy for PA IVS: outcome

Perforation of the Atretic Pulmonary Valve

Long-Term Follow-Up

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Yacine Aggoun, MD,* Tony Abdel-Massih, MD,* Younes Boudjemline, MD,* Christine Le Bihan, MD,*
Damien Bonnet, MD, PhD,* Daniel Sidi, MD, PhD*

Paris, France

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)

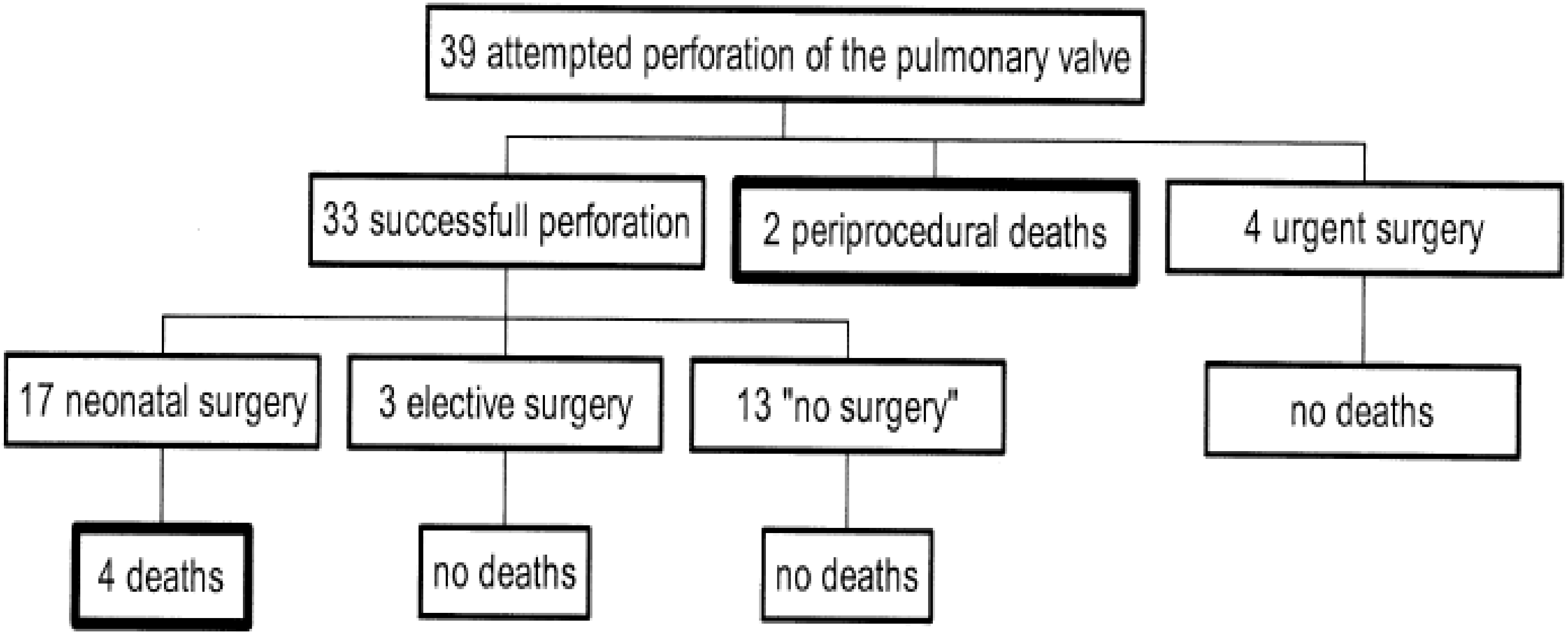

Figure 1. Flow chart summarizing the procedures and outcomes (death or no death) of patients.

- Perforation by guidewire 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)

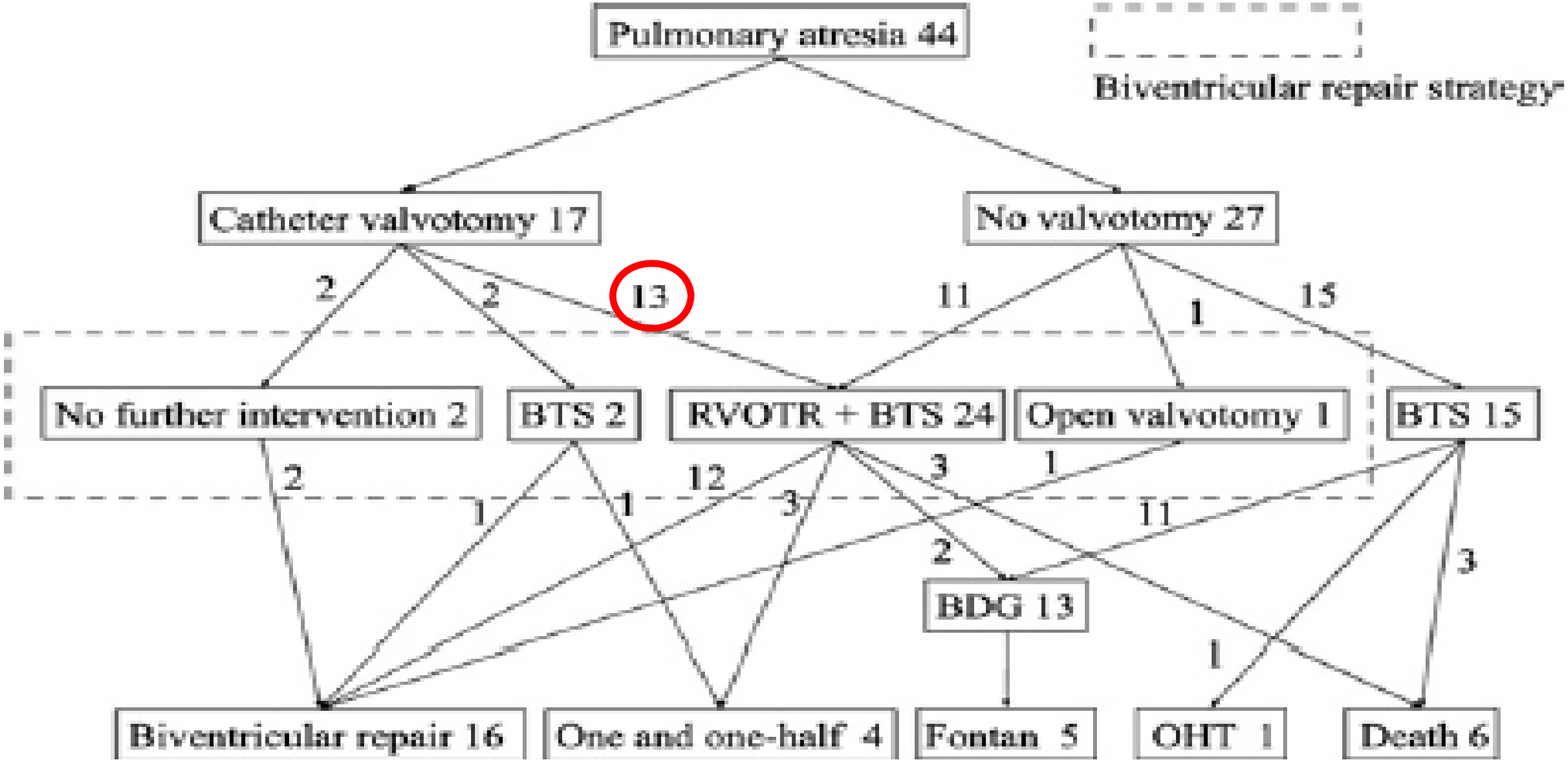

Fig 1. Flow chart of outcome for patients with pulmonary atresia-intact ventricular septum. Box with broken line shows the patients with biventricular repair strategy. (BDG = bidirectional Glenn operation; BTS = modified Blalock-Taussig shunt; OHT = orthotopic heart transplantation; RVOTR = right ventricular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

Comment

dies suggest that increasing numbers of ındergoing transcatheter pulmonary valvu- ıoplasty ıoı ı AIVS 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

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

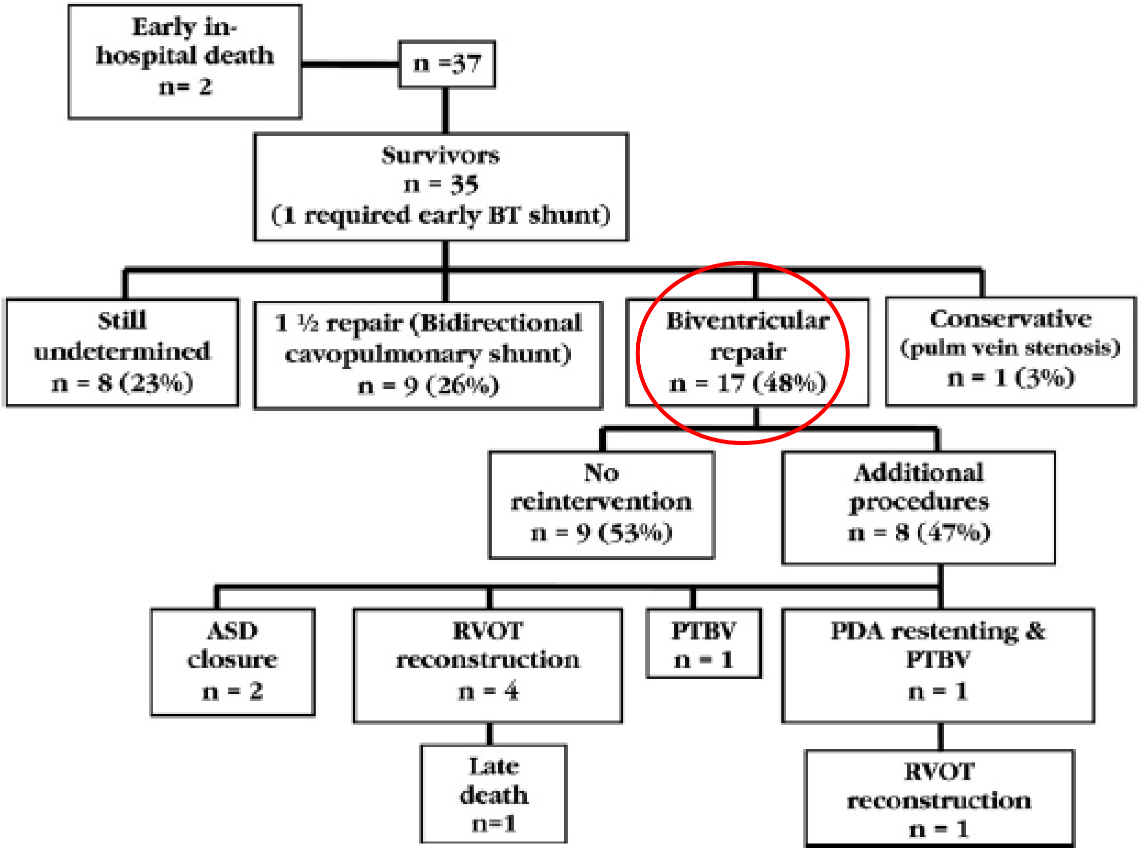
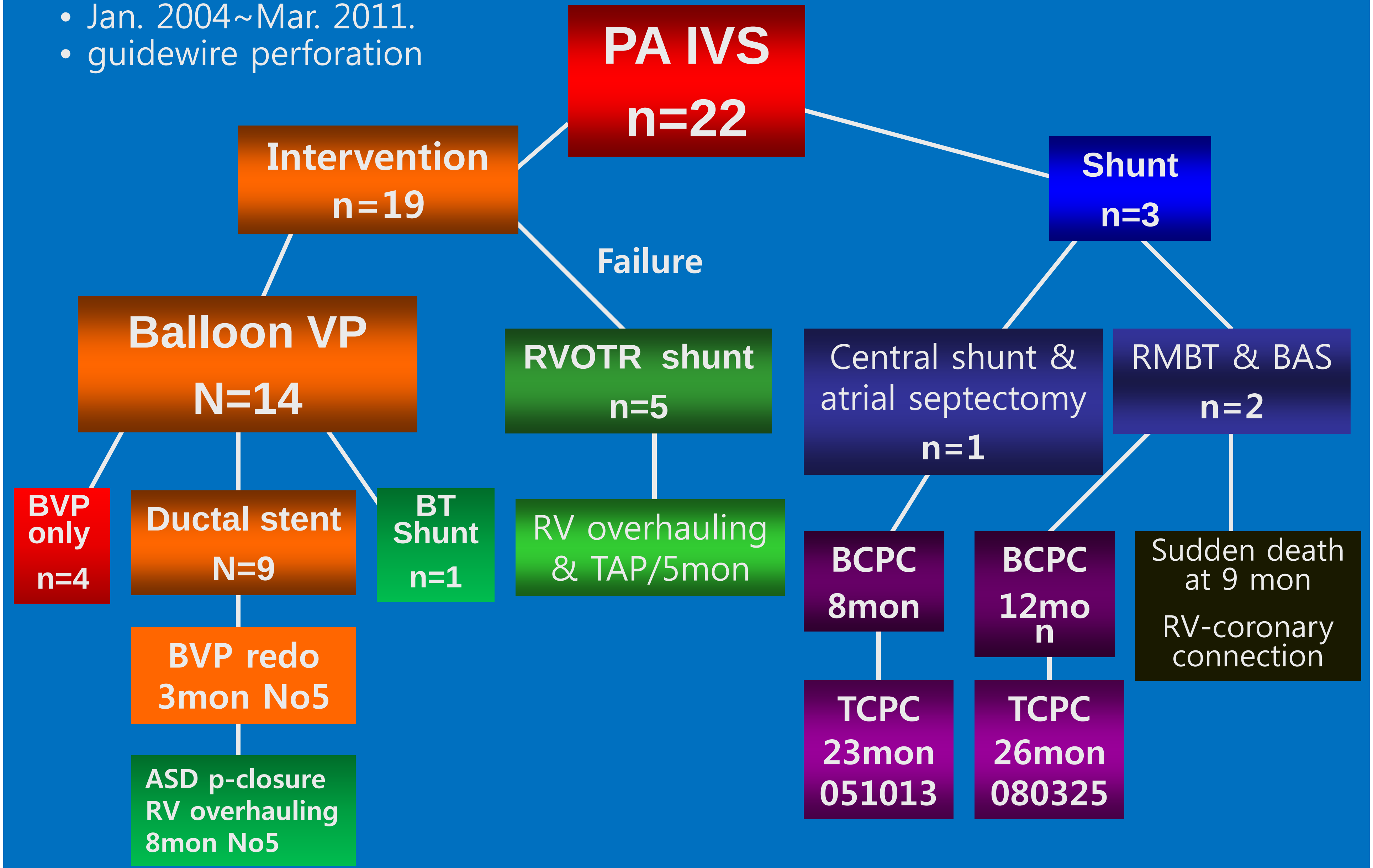


FIGURE 1. Outcome of patients with intermediate right ventricle *(RV)*. *BT*, Blalock–Taussig shunt; *ASD*, atrial septal defect; *RVOT*, right ventricular outflow tract; *PTBV*, Percutaneous transcatheter balloon valvuloplasty; *PDA*, patent ductus arteriosus.

- **PNUCH & PNUH**
- Jan. 2004~Mar. 2011.
- guidewire perforation



Transcatheter therapy for PA IVS: Summary

- Transcatheter balloon valvotomy with or without ductal stenting is an attractive alternative to surgical treatment esp in relatively good RV.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경청해주셔서 감사합니다!