Follow-up results of transcatheter pulmonary valvotomy in patients with pulmonary atresia and intact ventricular septum

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Background :

- Management strategy of pulmonary atresia and intact ventricular septum (PA-IVS) is generally based on size and morphology of right ventricle and presence of major sinusoids
- Transcatheter technique has been applied to treat PA-IVS with great success for 2 decades.
- We report the follow-up results of catheter-based management of PA-IVS

Patients & Methods:

- Between July 1995 & August 2010, 78 neonates with PA-IVS were diagnosed.
- Echocardiography was performed in each patient. Z score of tricuspid valve (TV) annulus was obtained.
- Transcatheter performed was attempted in 55 patients with tricuspid right ventricle (RV) and TV Z score > -3.5 (Z score ranging from -3.3 to 0.5)
- Twenty-three patients with a TV Z score < -3.5 or presence of major sinusoids were excluded for catheter perforation of pulmonary valve.
- An Osypka radiofrequency guide (RF) wire or coronary guide wire was used for perforation in 49 & 6 neonates, respectively. A subsequent balloon dilation was performed.
- Low dose PGE_1 was maintained until stable O_2 saturation above 75 % without use of O_2 supply.
- A concomitant stenting of ductus was performed in patients with TV Z score < -1.5 since July 2008.

Results :

§ Summarized as figure 1

- Successful perforation of pulmonary valve was achieved in 49 patients (34 males, mean TV Z score -1.1 ± 0.9) (Figure 2)
- Failure occurred in 6 patients of whom 3 complicated with temponade requiring emergent drainage.
- Of the 49 patients, the systolic RV pressure decreased from 115 ± 22 to 54 ± 12 mmHg ($p < 0.01$) following valvuloplasty
- PDA stent was deployed in 4. (Figure 3)
- Nine had severe RVOT obstruction or cyanosis requiring PGE_1 > 3 weeks. Seven had a RVOT patch with/ without a shunt and 2 underwent a shunt
- Renal failure requiring dialysis in 2.
- Ligation of ductus in 4.
- 4 early mortalities (1 sepsis & 3 heart failure)

Follow-up :

- Two late mortalities (1 heart failure & 1 sepsis), total mortality n=6
- RVOT patch was required in 3 because of progressive infundibular stenosis
- Two underwent a Glenn shunt
- Catheter closure of interatrial communication in 6.
- In 42 patients available for follow-up, both the mean TV Z score & pulmonary valve Z score were significantly higher in 32 without RVOT patch than 10 with RVOT patch (-0.84 ± 0.85 vs. -1.99 ± 0.34, p=0.042 & -2.23 ± 1 vs. -3.84 ± 1.6, p= 0.04, respectively)
- In 36 patients who received follow-up longer than 12 months, there was significant increase in TV Z score (-1.1 ± 1 vs. 0 ± 1.3 $p < 0.001$)

Discussions :

- A RF guide wire is effective in perforation of atretic pulmonary valve
- A concomitant stenting of ductus in neonates with a TV Z score < -1.5 can eliminate the need of a Blalock-Taussig shunt or prolonged administration of PGE1
- Perforation of infundibulum was not rare using a RF guide wire. Pulmonary overflow from PDA with severe pulmonary regurgitation, where PDA ligation was required was not infrequent.
- Growth of right ventricular size following transcatheter valvotomy was observed in this study

Conclusions :

- Catheter-based therapeutic strategy is effective in selected PA-IVS neonates with a Z score > -3.5.
- Stenting ductus in neonates with a TV Z valve < -1.5 may eliminate the need for a shunt or prolonged PGE_1 administration.

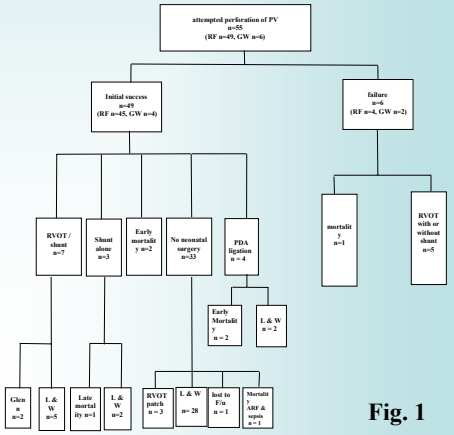

Fig. 1

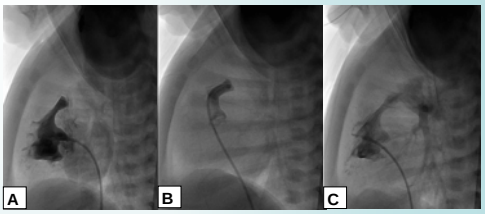

Fig. 2

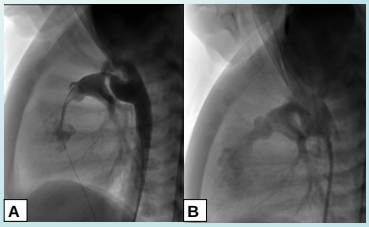

Fig. 3

Figure Legends

Figure 1. A flow chart of results and follow-up in PA-IVS patients undergoing pulmonary valvotomy

Figure 2. angiogram at RV showing atretic pulmonary valve with mild hypoplasia of RV. (A) during (B) and after (C) perforation of pulmonary valve .

Figure 3. A stent was deployed to maintain patency of ductus. (A) before stenting (B) after stenting.