Transcatheter perforation of atretic pulmonary valve by the stiff end of a coronary wire in neonates with pulmonary atresia with intact ventricular septum: A solution in developing countries

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Objectives: To evaluate the safety of using the stiff end of a coronary wire to perforate an atretic pulmonary valve (PV) in patients with pulmonary atresia with intact ventricular septum (PAIVS).

Background: Radiofrequency perforation is an accepted modality to perforate the PV in patients PAIVS. However, the high cost precludes its widespread use.

Patients and methods: This is a single-center experience that spanned from March 2013 to January 2016 and involved 13 neonates who were severely cyanotic with PAIVS and with ductal-dependent pulmonary circulation. The stiff end of a coronary wire was used to perforate the atretic PV anterogradely, followed by balloon pulmonary valvuloplasty.

Results: The mean age of patients was 3.9 ± 2.7 days and their mean weight was 2.8 ± 0.19 kg. The mean oxygen saturation was 77.1 ± 3.2%. All had membranous pulmonary atresia, with patent infundibulum and tripartite right ventricle. The valve was successfully perforated in 11 out of 13 patients. Death occurred in two patients (15.4%) owing to heart failure and sepsis. Patent ductus arteriosus stenting was performed 2 days after the procedure in one patient because of cyanosis followed by one and half ventricle repair at age 5 months. Two patients (15.4%) had one and a half ventricle repair at age of 5 months and 6 months owing to insufficient anterograde pulmonary flow. Two patients (15.4%) underwent second intervention with balloon dilatation of the valve. The remaining seven patients (53.8%) had no further intervention. Two patients (15.4%) had femoral artery thrombosis treated with streptokinase. The mean duration of follow-up was 13.17 ± 7 months. There was significant improvement in the degree of tricuspid incompetence. There was a significant growth in the tricuspid valve annulus during the follow-up (the mean Z score increased from −0.8 ± 0.9 to 0.1 ± 0.9) (p = 0.003). There was also a significant increase in the tricuspid valve annulus/mitral valve annulus ratio as its mean increased from 0.73 ± 0.10 to 0.86 ± 0.11 during follow-up (p < 0.001).
Conclusion: Perforation of the atretic PV in selected cases with membranous atresia and patent infundibulum using the stiff end of a coronary wire is an effective alternative to using radiofrequency perforation.

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Keywords: Coronary wire, Percutaneous balloon dilatation of pulmonary valve, Pulmonary atresia with intact ventricular septum, Pulmonary valve perforation

Introduction

Pulmonary atresia with intact ventricular septum (PAIVS) is a broad-spectrum pathology, depending on the degree of right ventricular hypoplasia varying from mild with patent infundibulum and well-developed tripartite or bipartite ventricle, to a severe form with muscular infundibulum and diminutive right ventricle (RV) [1,2].

Right ventricular assessment can be done echocardiographically by measuring tricuspid valve (TV) annulus Z score, degree of tricuspid incompetence, infundibular patency, pulmonary artery size, and presence or absence of RV-dependent coronary circulation [3–5]. Cheatham suggested that whenever there is a tripartite RV and a patent infundibulum, a TV annulus ≥11 mm, and membranous atretic pulmonary valve (PV; ≥7 mm), transcatheter therapy should be considered, but there are no sufficient data to support these recommendations [6].

Heterogeneity of this pathology classifies the management strategy into two major tracks: two ventricular circulation (or at least one and half ventricle repair) or single ventricle repair [7–10]. If the track is univentricular repair, the interventional role is patent ductus arteriosus (PDA) stenting as an alternative to surgical Blalock–Taussig (BT) shunt, which can be considered with balloon atrial septostomy if needed [1].

However, in patients with membranous atresia and patent infundibulum, the RV morphology usually gives the chance for biventricular circulation. In such cases, the target will be to relieve the RV obstruction and achieve anterograde flow to the pulmonary vascular bed [6,11]. Conventionally, this was achieved by surgical valvotomy in addition to a modified BT shunt [12]. The accepted interventional alternative is the use of radiofrequency-assisted perforation of the atretic valve and subsequent balloon dilation [9,12,13]. However, radiofrequency perforation is expensive and is not available in every center. Meanwhile, the use of coronary wires used for chronic coronary total occlusion has also been reported for PV perforation [12].

We report our experience using the stiff end of a coronary wire to perforate the PV.

Patients and methods

In this single-center experience, which spanned from March 2013 to January 2016, we studied 13 neonates [4 females (30.8%) and 9 males (69.2%)] whose mean age was 3.9 ± 2.7 days (range, 1–10 days). Their mean weight was 2.8 ± 0.19 kg (2.5–3 kg). All patients were severely cyanotic, and the diagnosis of PAIVS with ductal-dependent pulmonary circulation was confirmed by Transthoracic Echocardiography (TTE). The duct patency was preserved by prostaglandin infusion. Echocardiography was used for proper selection of patients. For inclusion, patients had to meet the following criteria (Table 1):

1. Membranous pulmonary atresia
2. Patent infundibulum (mild or no hypoplasia)
3. TV Z score > -2
4. TV annulus/mitral valve (MV) annulus ratio > 0.5
5. Tripartite or bipartite RV
6. Absence of RV-dependent coronary circulation
7. Absence of Ebstein malformation of the TV

Procedure

Our study was performed under the approval of the ethical committee of Tanta University (Medical Science) and complied with the principles of the Declaration of Helsinki in 1964. After explaining
the benefits and risks of the procedure, we obtained informed consent from legal guardians. The procedure was performed under general endotracheal anesthesia. Both femoral vein (5F catheter) and artery (4F catheter) were percutaneously accessed. Patients received heparin at 50–100 international unit/kg. A 4F Judkin’s right coronary or reshaped 4F multipurpose catheters were used to perform right ventricular (RV) angiogram in anteroposterior and lateral projections to delineate the right ventricular inflow, outflow, and to evaluate the degree of right ventricular hypoplasia. Then, an aortogram was performed to rule out the presence of any coronary anomalies or sinusoids, to delineate the ductus morphology and the degree of development of the pulmonary branches.

**Technique of perforation**

- (Fig. 1) The catheter was manipulated from the aorta through the ductus, and an angiogram was performed to delineate the PV and the pulmonary branches.

- Simultaneous injection with one catheter at the end of the ductus and a second catheter facing the PV dimple was essential in all cases, as it demonstrated exactly where to penetrate the membrane.
- A 5-mm goose neck snare (Ev3 Endovascular, Inc., Plymouth, MN, USA) was manipulated through the ductus and kept opened against the valve with exertion of some light pressure on the membranous valve. This stabilized the membrane and provided a clear demarcation of the right ventricular outflow tract (RVOT) to prevent the false passage of the wire.
- The stiff end of the available coronary wires (e.g., Pilot 150, Balance Middle Weight (BMW) wire) was gently manipulated and advanced, keeping the catheter close to the membranous valve (keeping both the wire and the catheter as one assembly perpendicular on the membrane was the key for safety); the wire was required to be pushed very gently to perforate the valve.
- Once the wire perforated the membrane, control hand injection through the ductus was mandatory to confirm the position of the wire within the pulmonary trunk and to detect if there was any RVOT perforation. Then, the catheter was advanced over the wire. Finally, this wire was replaced by another coronary wire by advancing its soft tip.
- The guidewire was further advanced carefully through the ductus, snared into the descending aorta, then exteriorized through the femoral arterial sheath, creating an arteriovenous wire loop and achieving stable position. However, in three cases, the wire passed smoothly into the distal pulmonary branch without a need for an arteriovenous loop; yet, in two of three cases, the balloon did not show good alignment and arteriovenous loop had to be established. The loop provided stability and good alignment of the balloon within the RVOT.
- A coronary balloon, 1.5 mm × 15 mm (Ryujin Plus; Terumo Medical Corporation, Tokyo, Japan), was advanced over the wire to perform the first dilatation. The size of the balloon was serially upgraded from 1.5 mm to a maximum of 10 mm (diameter). The largest final balloon size was chosen to be 120% to no more than 140% of the PV annulus size. At the end of the procedure, pull back gradient from the pulmonary artery to the RV and RV angiogram were performed routinely in all cases.

**Statistical analysis**

Data were fed to the computer and analyzed using the IBM SPSS software package version 20 (IBM Corp., Armonk, NY, USA). For normally distributed quantitative periods, paired *t* test was used to assess the data and Wilcoxon signed ranks test was used for abnormally distributed quantitative periods, whereas chi-square test was used for qualitative periods. Significance of the obtained results was judged at the 5% level.

**Results**

Thirteen patients were eligible for PV perforation. The mean oxygen saturation on presentation...
was $77.1 \pm 3.2\%$ (72–83%) by the effect of prostaglandin infusion (Table 2).

All patients had membranous pulmonary atresia and patent infundibulum. The RV was tripartite in all patients with a mean TV annulus diameter of 10.2 ± 1.7 mm and a mean Z score of $-0.8 \pm 0.9$. The mean TV/MV annulus ratio was 0.73 ± 0.10 (i.e., $>0.5$). The pulmonary artery was well developed in all patients, with a mean diameter of 10.0 ± 0.82 mm and mean Z score of 1.42 ± 0.18. None of the selected cases by echocardiographic study had been excluded after RV angiogram; there was no evidence of coronary sinusoids/RV-dependent coronary circulation.

**Outcome**

The valve was successfully perforated in 11 out of 13 patients (84.6%) using the stiff end of the coronary wire. In 10 cases, the wire was exteriorized through the descending aorta creating an arteriovenous loop, which provided stability for the wire and facilitated the passage of the serial balloons through the RVOT.

In three cases, the wire was advanced directly into the distal pulmonary branches. In two of three patients, the balloon could not be advanced to cross the valve. So, we had to pull it back. Then, we redirected it into the descending aorta through the ductus, and exteriorized through the femoral artery creating an arteriovenous loop.
There was significant decrease in mean RV pressure after intervention (the mean RV pressure prior to intervention was 94.2 ± 4.5 mmHg, which improved to 41.9 ± 12 mmHg after the intervention; \( p < 0.001 \) (Fig. 2). There was a significant increase in mean oxygen saturation, from 77.1 ± 3.2% prior to intervention to 88.0 ± 6.51% after the intervention (\( p < 0.001 \)) (Fig. 3).

After the procedure, all patients were kept on prostaglandin infusion until the forward flow achieved by the balloon dilation was assured. Furthermore, positive inotropic support, e.g., milrinone infusion to improve the function of the RV, was kept for all patients then reduced gradually during the following 4–10 days. Follow-up was done in the outpatient clinic.

**Follow-up**

On follow-up, the following results were noted (Fig. 4):

1. Death occurred in two patients (15.4%)—one patient died 1 week after the procedure owing to sepsis and the other patient died after 3 days because of heart failure.
2. PDA stenting was done at age of 5 days in one patient (7.7%) because of persistent desaturation and duct dependency for 2 days after dilation followed by one and a half ventricle repair at age of 5 months.
3. Glenn shunt (one and half ventricle repair) was performed in two patients (15.4%) at age of 5 months and 6 months because of continued desaturation with a maximum oxygen saturation of around 70%.
4. Two patients (15.4%) underwent second intervention with balloon dilatation of the PV at age 7 months and 8 months because of residual RVOT obstruction with maximum gradient across RVOT 70 mmHg and 60 mmHg, respectively.
5. Seven patients (53.8%) did not require any further intervention.
6. In two patients (15.4%), the femoral artery was found to be thrombosed immediately after intervention. In both cases, streptokinase treatment was initiated and pulse was restored.

The mean duration of follow-up was 13.2 ± 7 months. The definition of success was successful perforation of PV with oxygen saturation ≥ 85%. The follow-up parameters were:

- Oxygen saturation
- Mean Z score value of TV annulus
- Regression of tricuspid incompetence
- TV/MV annulus ratio as an indicator of right ventricular growth

Nine patients out of 13 have achieved successful biventricular circulation; two had received what we call one and half ventricle repair (bidirectional Glenn with inferior vena cava flow to the RV).

Table 2. Data after intervention.

|                  | Before intervention | After intervention | p       |
|------------------|---------------------|--------------------|---------|
| Saturation       | Mean ± SD           | Median (min–max)   |         |
|                  | 77.1 ± 3.2          | 77 (72–83)         |         |
|                  | 88.0 ± 6.51         | 90 (77–95)         | <0.001  |
| RV pressure      | Mean ± SD           | Median (min–max)   |         |
|                  | 94.2 ± 4.5          | 95 (85–100)        |         |
|                  | 41.9 ± 12           | 35 (30–70)         | <0.001  |

RV = right ventricle; SD = standard deviation.
* significant p value.
There was significant decrease in the degree of tricuspid incompetence (\( p = 0.014 \)). There was significant growth of TV annulus as its mean diameter increased from \(10.2 \pm 1.7 \text{ mm} \) prior to perforation to \(17.5 \pm 3.8 \text{ mm} \) during follow-up (\( p = 0.001 \)); meanwhile, the mean Z score improved from \(-0.8 \pm 0.9 \) before to \(0.1 \pm 0.9 \) during follow-up (\( p = 0.003 \)). There was a significant increase in TV/MV annulus ratio as its mean increased from \(0.73 \pm 0.10 \) prior to closure to \(0.86 \pm 0.11 \) during follow-up (\( p < 0.001 \)) (Table 3 and Figs. 5–8).

**Discussion**

The morphological heterogeneity in patients with PAIVS with its hemodynamic implications provides a great challenge in evaluation and clinical decision-making for the interventional cardiologists and surgeons [1,14].

In selected morphological situations in patients with PAIVS where the nature of the obstruction is membranous atresia, and not a muscular one, and patent infundibulum with accepted degree of growing RV, and finally absence of coronary sinusoids, then decompression of RV is the optimal solution [6,8].

Radiofrequency perforation had gained much acceptance compared with surgical valvotomy [8,9,13]. Also, laser valvotomy and balloon dilatation have been used successfully in PAIVS [15]. However, because of high cost, both radiofrequency perforation and laser valvotomy are not
available in all centers. This explains why a trial for mechanical perforation using coronary wires had been reported [13].

Tanıdır et al. [12] reported the successful use of Conquest Pro 12 guidewire in the perforation of an atretic PV after failure of radiofrequency perforation of the valve.

Alcíbar-Villa et al. [13] and Bakhru et al. [16] reported the use of special coronary wires for total occlusion (CTO wires) depending on its power of penetration as they were concerned about the degree of risk using the stiff end of the coronary wire. In addition, controlling the stiff end is not easy. Alcíbar-Villa et al. [13] reported that the
Fig. 5. Change in tricuspid incompetence during follow-up.

Fig. 6. Change in tricuspid annulus size during follow-up.

Fig. 7. Change in tricuspid annulus Z score during follow-up. TV = tricuspid valve.
survival rate was 72% of the children. There was significant reduction in RV pressure, significant increase in the TV diameter, but nonsignificant reduction of tricuspid incompetence degree, which we had reported [13].

Bakhru et al. [16] reported the successful perforation of PV in 16 out of 20 patients. Cardiac tamponade due to RVOT perforation was seen in one patient, who required pericardiocentesis. Six patients required additional procedures in the immediate post-perforation period because of insufficient anterograde pulmonary flow (requiring duct stenting in four patients, RVOT stenting in one patient, and BT shunt in another patient). In that cohort, three patients underwent one and a half ventricular repair [16].

Çelebi et al. [17] reported successful anterograde perforation of the PV in two patients with PAIVS using the stiff end of a coronary guidewire by snaring the guidewire into the pulmonary artery, then performing balloon pulmonary valvuloplasty. Çelebi et al. [17] also reported a successful retrograde perforation in one patient.

In our study, we used the stiff end of a coronary wire to perforate the atretic valve. We could not use special CTO wires because they are not widely available in our country and are considerably costly. However, we did not encounter any perforation leading to pericardial effusion/tamponade as we used the catheter and the stiff end of the coronary wire as one assembly, keeping them firmly in apposition to the atretic membrane. Then, we relied on the pressure exerted by the snare loop positioned against the membrane. This technique enabled us to succeed in penetrating the membrane without any unwanted RVOT perforation. The establishment of the arteriovenous loop provided very good stability to track and position the balloons easily at the desired location. The valve was successfully perforated in 11 out of 13 patients (84.6%). Death occurred in two patients (15.4%). Meanwhile, PDA stenting was required in one patient (7.7%), and Glenn shunt with one and a half ventricular repair was performed in two patients (15.4%). Two patients (15.4%) required a second setting of balloon dilation of the PV. Seven patients (53.8%) did not require any other intervention. Femoral artery thrombosis occurred in two cases (15.4%), treated by streptokinase with full resolution of the problem. There was a significant decrease in the degree of tricuspid incompetence, a significant growth of TV annulus, and a significant increase of TV/MV annulus ratio during the follow-up.

**Conclusion**

Perforation of the atretic PV in selected cases with membranous atresia and patent infundibulum, aiming to decompress the RV, is safe and effective in promoting the right ventricular growth and in achieving biventricular repair. Using the stiff end of a coronary wire with some technical modifications (using the opened snare against the membrane for stability and using the stiff end within the catheter as one assembly) reduced the incidence of unwarranted perforations. Then, creation of the arteriovenous wire loop provided stability to track and position the balloons. We believe that this technique is a good alternative
to radiofrequency perforation, which is not readily available in all centers, and it saves money, which is of importance in developing countries.

Conflict of interest

There are no conflicts of interest.

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Limitations

This is a retrospective observational study from a single center with a small number of highly selected patients with PAIVS. Therefore, our results cannot be applied to all the cases with pulmonary atresia, intact septum.

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