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# Outcomes in Neonates with Pulmonary Atresia and Intact Ventricular Septum Underwent Pulmonary Valvulotomy and Valvuloplasty Using a Flexible 2-French Radiofrequency Catheter

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**Purpose :** Outcomes in 6 neonates with pulmonary atresia and intact ventricular septum (PAIVS) undergoing radiofrequency pulmonary valvulotomy and valvuloplasty (RPVV) were reported to identify the factors favorable for RPVV as the treatment of choice. **Materials and Methods:** From May 2000 to January 2008, 6 patients with PAIVS were included in this retrospective study. They were aged 1 day to 90 days old. Study modalities included review of recordings of presentations and profiles of chest radiography, electrocardiography, echocardiography, and cardiac catheterization with angiography. Hemodynamic profiles from the echocardiography and the cardiac catheterization were analyzed. **Results:** Echocardiography showed severe tricuspid regurgitation, membranous atresia of the pulmonary valve, intact ventricular septum, patent ductus arteriosus, and hypoplastic right ventricle in 6 patients. The pulmonary valve annulus were 4.2 to 6.9 mm in diameters, and those of the tricuspid valve were 7.1 to 10.1 mm. Elevated serum level of cardiac enzymes were found in 1 patient with ventriculocoronary communication (VCC). At cardiac catheterization, the ratio of systolic pressure of the right ventricle to that of the left ventricle ranged from 1.43 to 2.33 before RPVV, and from 0.54 to 1.15 after RPVV ($p$=0.027). The pressure gradients ranged from 76 to 136 mmHg before RPVV, and from 15 to 39 mmHg after RPVV ($p$=0.028). The echocardiographic gradients ranged from 16 to 32 mmHg within 24 hours after RPVV, and from 15 to 50 mmHg at the follow-ups. **Conclusion:** RPVV can be a treatment of choice for neonates with PAIVS, if there is patent infundibulum, no right-ventricular dependent coronary circulation, and adequate tricuspid valve and pulmonary valve.

**Key Words :** Pulmonary atresia, intact ventricular septum, radiofrequency, pulmonary valvulotomy, pulmonary valvuloplasty, unipolar, gradational balloon dilatation, Z values, ventriculocoronary communication

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## INTRODUCTION

Previously, surgical repair for neonates with pulmonary atresia and intact ventricular septum (PAIVS) involved: either 1) a palliative systemic-to-pulmonary shunt, or 2) a corrective pulmonary valvulotomy together with reconstruction of the right ventricular outflow tract.[1] However, the mortality and the morbidity remained high after surgery, especially in those with severe hypoplastic right ventricles with or without infundibulum, non-confluent pulmonary arteries, ventriculocoronary communication (VCC), and right ventricle-dependent coronary circulation (RVDCC).[2] Recently, transcatheter radiofrequency pulmonary valvulotomy and valvuloplasty (RPVV) emerged as a promising alternative to surgical pulmonary valvulotomy for PAIVS in patients with favorable morphology of the right ventricle, the tricuspid valve, and the pulmonary valve.[3]

Up to date, it is a win-win scenario for patients with PAIVS and pediatric cardiologists performing RPVV, provided that there was presence of patent infundibulum and absence of RVDCC.[4]

## MATERIALS AND METHODS

From May 2000 to January 2008, 6 consecutive patients (4 male and 2 female) with PAIVS were included in this retrospective study. They were aged from 1 day to 90 days old. Patients having pulmonary atresia associated with ventricular septal defect, Ebstein's anomaly, or other congenital heart diseases were excluded from the study. Study modalities included review of profiles of recordings of the clinical features, arterial blood gas, chest radiographs, electrocardiograms, two-dimensional echocardiography with Doppler (Acuson 128XP/10c, Mountain View, CA, USA), and cardiac catheterization with angiography. In all 6 patients, diagnostic angiography (right ventriculogram, left ventriculogram, and ascending aortogram) was performed to visualize the morphology of the right ventricle and VCC and define the course and distribution of the coronary arteries as they arose from the aorta. RPVV was performed after obtaining informed consent from their parents. Using a standard Seldinger maneuver, cardiac catheterization was performed through the femoral vein and artery in each patient, to whom intravenous prostaglandin $E_1$ ($PGE_1$), intravenous antibiotics, mechanic ventilation, and local anesthesia were applied beforehand. Prior to performing RPVV, systolic pressures of the right ventricle (sRV) and main pulmonary artery (sPA) were measured simultaneously by 2 catheters. The sRV was measured by a 4-French (or 5-French) Multi-Purpose Catheter (Cordis, Miami, FL, USA), a Right Coronary Judkins Catheter (Cordis, Miami, FL, USA), or a Berman Catheter (Arrow, Mt. Holly, NJ, USA) approaching the right ventricle antegradely from the femoral vein, and the sPA was measured by a 4-French Multi-Purpose Catheter approaching the main pulmonary artery retrogradely from the femoral artery via the patent ductus arteriosus (PDA). The pressure gradient was determined by the equation of sRV-sPA, i.e., subtracting systolic pressure of the main pulmonary artery from that of the right ventricle. A flexible 2-French unipolar radiofrequency catheter (CERABLATE PA 120, Grenzach-Wyhlen, Germany), which was passed through a 5-French Right Coronary Judkins Catheter or a 6-French Coronary Guiding Catheter (TRANSIT, Cordis, Miami Lakes, USA) was used to deliver radiofrequency energy of 2 to 15 watts (W) for 3 to 5 seconds from the generator (HAT 200 S, Grenzach-Wyhlen, Germany) to perforate the atretic pulmonary valve. Initially, a small coronary balloon catheter (2.0 mm in diameter and 20.0 mm in length; HAYATE, Terumo, Tokyo, Japan) was used to dilate the already open pulmonary valve, and subsequently, some larger coronary balloon catheters (3.0 mm and 4.0 mm in diameter; HAYATE, Terumo, Tokyo, Japan; 5.0 mm and 6.0 mm in diameter Maverick, Boston, MN, USA) and even Wanda balloon catheter (8.0 mm in diameter; Business Park, Allybrit, Galway, Ireland) were applied to dilate gradationally. After RPVV, the pressure gradients across the pulmonary valve were obtained, according to the records of the pressure tracings, by pulling back the balloon catheter from the main pulmonary artery to the right ventricle. Hemodynamic data, obtained within 24 hours, of the echocardiography and cardiac catheterization were analyzed to evaluate the initial results of RPVV. Transvalvular pressure gradients by echocardiography and oxygen saturations by peripheral pulse oximeter were recorded for each patient at follow-ups to evaluate the final outcomes of RPVV.

## RESULTS

The clinical profiles and hemodynamics of 6 neonates with PAIVS treated by RPVV are summarized in Table 1 and Table 2, respectively. Echocardiography with Doppler showed dilated right atrium, severe tricuspid regurgitation, small atrial septal defect secundum with restrictive right-to-left shunt, hypoplastic right ventricle with visible right ventricular outflow tract, membranous atresia of pulmonary valve with visible pulmonary annulus and main pulmonary trunk, confluent pulmonary arteries without branch pulmonary artery stenosis, intact ventricular septum, and PDA in 6 all cases. Patient 2 had non-obstructive VCC, but none of these 6 patients had RVDCC, judged by diagnostic angiography. Before RPVV, a 4-French Multi-Purpose Catheter can be situated in the main pulmonary trunk serving as the "landmark catheter" in 2 cases with upstream PDA. However, the process of putting a "landmark catheter" into the main pulmonary artery was demanding in 4 patients with downstream PDA. The sRV/sLV ratios ranged from 1.43 to 2.33 (1.90 ± 0.30, median 1.88) before RPVV, and from 0.54 to 1.15 (0.80 ± 0.27, median 0.76) after RPVV ($p = 0.027$). The pressure gradients of sRV-sPA ranged from 76 to 136 mmHg (104.2 ± 24.8, median 99.5 mmHg) before RPVV, and from 15 to 39 mmHg (27.5 ± 9.8, median 27.5 mmHg) after RPVV ($p = 0.028$). The echocardiographic pressure gradients ranged from 16 to 32 mmHg (25.2 ± 5.7, median 25.5 mmHg) within 24 hours after RPVV.

Five patients, except Patient 1, could be weaned off mechanical ventilation and intravenous $PGE_1$ without surgery. Three of these 5 patients (Patients 3, 5, and 6) were discharged smoothly after RPVV without significant medical events during their hospitalizations. Two of these 5 patients (Patients 2 and 4) suffered from 2 episodes of nosocomial infection in each; with bacterial pneumonia and Rotaviral gastroenteritis in Patient 2 and bacterial pneumonia and septicemia in Patient 4. These episodes of infection were under control by intravenous third-generation cephalosporins and other supportive care. Patient 2 had elevated serum levels of creatine kinase (CK; 2030 U/L >

**Table 1.** Clinical Profiles of 6 Patients with PAIVS Treated by RPVV Using a Flexible 2-French Radiofrequency Catheter Followed by Gradational Balloon Dilatation

| No / Age / Gender | 1/14 d/M | 2/90 d/M | 3/1 d/M | 4/1 d/F | 5/2 d/M | 6/1 d/F |
|---|---|---|---|---|---|---|
| $SpO_2$ (at onset) | 70% | 80 | 65 | 68 | 60 | 80 |
| Liver span (cm) | 3.5 | 4.0 | 3.5 | 3.5 | 3.0 | 3.0 |
| CTR (%) | 73 | 71 | 72 | 72 | 68 | 64 |
| ASD II (mm) | 2.5 | 3.2 | 2.8 | 3.0 | 2.4 | 2.7 |
| VCC | Negative | Positive | Negative | Negative | Negative | Negative |
| $PGE_1$ (ng/kg/min) | 20, 40, 60 | 20, 40, 60 | 20, 40 | 20, 40, 60 | 20 | 20 |
| PDA morphology | Downstream | Downstream | Downstream | Upstream | Downstream | Upstream |
| TGRM | Nil | Nil | Nil | Positive | Nil | Positive |
| RF energy, time | 5 W, 4 sec | 5 W, 5 sec | 4 W, 5 sec | 5 W, 3 sec | 2 W, 3 sec | 15 W, 5 sec |
| Medical events | 1) cyanosis ($SpO_2$ 50%) and PHT due to RPVC s/p effective RPVV ($SpO_2$ 90%) and BTS<br>2) treated with NOi ($SpO_2$ 88%)<br>3) digitalis, furoxemide, captopril, and aspirin | 1) MI (with elevated CK/CK-MB/Tn-I)<br>2) pneumonia<br>3) Rotaviral infection<br>4) regression of MI and VCC s/p RPVV;<br>5) propanolol | Nil | 1) pneumonia<br>2) sepsis<br>3) cyanosis ($SpO_2$ 68%) and PHT due to RPVC s/p effective RPVV ($SpO_2$ > 92%)<br>4) treated with NOi ($SpO_2$ above 95%)<br>5) propanolol | Nil | Nil |
| Surgical events | BTS (right, stenotic; left, patent) | Nil | Nil | Nil | Nil | Nil |
| Sequelae | Nil | Nil | Nil | Nil | Nil | Nil |
| $SpO_2$ (at FU) | 82 - 88% | 82 - 85% | 96 - 100% | 92 - 93% | 94 - 96% | 97 - 99% |

ASD II, atrial septal defect secundum; BTS, Blalock-Taussig shunt; CK, creatine kinase; CK-MB, myocardial fraction of thecreatine kinase; CTR, cardiothoracic ratios calculated on the plain chest films; FU, follow-ups; MI, myocardial ischemia; NOi, nitric oxide inhalation; PAIVS, pulmonary atresia and intact ventricular septum; PDA, patent ductus arteriosus; $PGE_1$, prostaglandin $E_1$; PHT, pulmonary hypertension; RF, radiofrequency; RPVC, reactive pulmonary vasoconstriction; RPVV, radiofrequency pulmonary valvulotomy and valvuloplasty; sec, seconds; $SpO_2$, saturation of oxygen; s/p, *status post*; TGRM, transductal guidewire railing maneuver; Tn-I, Troponin-I; VCC, ventriculocoronary communication that is not right-ventricular dependent; W, watt.

**Table 2.** Hemodynamics of 6 Patients with PAIVS Treated by RPVV Using a Flexible 2-French Radiofrequency Catheter Followed by Gradational Balloon Dilatation

| No / | PV/TV vs Z-values | | | | Balloon modality | Catheter PG before and after RPVV | | | | Echo PG (mmHg) | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Age / | PV | Z | TV | Z | Length (20 mm) | sRV-sPA (mmHg) | | sRV/sLV (%) | | within | F-U at the |
| Sex | mm | | mm | | Diameter in mm | Before | After | Before | After | 24 hrs | age of |
| 1/14 d/M | 6.0 | - 3.7 | 7.1 | - 1.7 | 2.0; 6.0 | 76 | 31 | 1.73 | 0.81 | 25 | 18; (91 m) |
| 2/90 d/M | 5.1 | - 4.5 | 7.5 | - 1.5 | 2.0; 5.0; 6.0 | 136 | 15 | 1.97 | 0.54 | 16 | 15; (55 m) |
| 3/1 d/M | 6.6 | - 3.1 | 10.0 | - 0.9 | 2.0; 4.0; 6.0; 8.0 | 105 | 39 | 2.13 | 0.70 | 32 | 16; (34 m) |
| 4/1 d/F | 4.2 | - 5.5 | 8.8 | - 0.1 | 2.0; 3.0; 6.0 | 131 | 37 | 2.33 | 1.15 | 26 | 21; (25 m) |
| 5/2 d/M | 6.9 | - 2.6 | 9.1 | - 0.8 | 2.0; 4.0; 6.0; 8.0 | 83 | 24 | 1.78 | 1.09 | 22 | 35; (15 m) |
| 6/1 d/F | 6.3 | - 2.2 | 10.1 | - 0.2 | 2.0; 4.0; 6.0; 8.0 | 94 | 19 | 1.48 | 0.53 | 30 | 50; (5 m) |
| Range | | | | | | [76, 136] | [15, 39] | [1.43, 2.33] | [0.54, 1.15] | [16, 32] | [15, 50] |
| Median | | | | | | 99.5 | 27.5 | 1.88 | 0.76 | 25.5 | 19.5 |
| Mean ± SD | | | | | | 104.2 ± 24.8 | 27.5 ± 9.8 | 1.90 ± 0.30 | 0.80 ± 0.27 | 25.2 ± 5.7 | 25.8 ± 13.9 |
| *p* value* | | | | | | 0.028 | | 0.027 | | 0.917 | |

d, day; Echo, echocardiographic; F, female; M, male; m, month; PG, pressure gradient; PV, pulmonary valve; RPVV, radiofrequency pulmonary valvulotomy and valvuloplasty; sRV-sPA, systolic pressure in mmHg of the right ventricle minus that of the main pulmonary artery; sRV/sLV, ratio expressed in percentage (%) of the systolic pressure of the right ventricle to that of the left ventricle; SD, standard deviation; TV, tricuspid valve; $V_{max}$, the maximal velocity in meters per second.
**p* value by Wilcoxon signed rank test.

upper normal limit of 224 U/L), myocardial fraction of CK (CK-MB; 429 U/L > upper normal limit of 6.3 U/L), and Troponin-I (Tn-I; 48.20 ng/mL > upper normal limit of 0.04 ng/mL), which were suggestive of myocardial ischemia due to VCC before RPVV. All parameters regressed to normal levels after RPVV. Patients 1 and 4 suffered from blue spells after RPVV. In both patients, echocardiography with Doppler and bubble-contrast test showed spontaneous closure of the PDA, severe tricuspid regurgitation, unchanged pressure gradient across the pulmonary valve, and significant interatrial right-to-left shunt via the atrial septal defect secundum, which may be due to either inadequate effective pulmonary blood flow from the right ventricle with post-procedure infundibular spasm or pulmonary hypertension with reactive pulmonary vasoconstriction. Patient 1 did not initially respond to inhalation of nitric oxide and construction of a right Blalock-Taussig shunt. He could be weaned off mechanical ventilation, intravenous $PGE_1$, and inhalation of nitric oxide after reconstruction of a patent left Blalock-Taussig shunt and application of beta-blocker (propanolol). He was discharged with an oxygen saturation of 88%, and treated with oral digitalis, furoxemide, captopril, and aspirin at the outpatient clinic. We did not use a larger 8-mm balloon in Patient 1, because infundibular spasm cannot be ruled out. Septicemia was documented in Patient 4 and was the suspected precipitating factor of blue spell or pulmonary hypertension. After administrations of intravenous third-generation cephalosporins and inhalation of nitric oxide, oxygen saturations could maintain above 95%. The PDA and the ASD II remained open in 1 patient (Patient 2) after RPVV and discontinuance of intravenous $PGE_1$.

The echocardiographic pressure gradients ranged from 15 to 50 mmHg at the follow-ups of 5 to 91 months. Fascinatingly, the VCC disappeared in the follow-up echocardiography and right ventriculography in Patient 2. Four of 6 patients had oxygen saturations above 92%, and 2 patients had saturations above 82%. None of them had neurological sequelae on the follow-up evaluations.

## DISCUSSION

RPVV has recently emerged as a promising alternative to surgical valvulotomy in patients with PAIVS, in whom 4 key points merit our attentions, including 1) echocardiographic diagnosis, 2) angiographic diagnosis and characteristics, 3) interventional techniques, and 4) mechanism of cyanosis and adjunctive treatment of pulmonary hypertension following RPVV.

First of all, echocardiography with color Doppler is considered useful to identify the ductus, to determine the intracardiac pathology, and to guide us in making a decision to prescribe intravenous $PGE_1$ to keep the ductus patent as an emergency medical therapy. Absence of patent infundibulum and presence of VCC could be confirmed and alarmed beforehand. Besides, the morphologic heterogeneity of the right ventricle and the coronary arteries can be assessed, and the diameters of the tricuspid valve and the pulmonary valve can be measured and computed to Z values.[3] So far, transcatheter pulmonary valvotomy was considered a definitive treatment in the neonates with PAIVS, if the Z-value of the tricuspid valve ≥ -0.1 and that of the pulmonary valve ≥ -4.1.[5] In the present study, the Z-values of the pulmonary valves ranged from -2.2 to -5.5, and those of the tricuspid valves ranged from -0.1 to -1.7. Blalock-Taussig shunt was needed only in one patient (Patient 1), whose Z-value of the pulmonary valve was -3.7 and that of the tricuspid valve -1.7. However, 2 of 6 patients (Patients 2 and 4) had smaller pulmonary valve Z-values (-4.5 and -5.5) and tricuspid valve Z-values (-1.5 and -0.1), and they went through successful RPVV without any surgical intervention. The Z-values of the pulmonary valve and the tricuspid valve were not the *sine qua non* determinants for successful RPVV in patients with PAIVS. Smaller Z-values of the pulmonary valve and the tricuspid valve are not absolute contraindications for considering RPVV alone as a definitive treatment in patients with PAIVS.

Secondly, the course and distribution of the coronary arteries should be carefully delineated, and the morphology of the right ventricle, the PDA, and the right atrial appendage be clearly defined or visualized in the diagnostic angiography before performing RPVV. Decisions concerning the surgical or transcatheter right ventricular decompression (RVD) in patients with PAIVS have been evolved in four decades and focused initially on the size and the number of partites constituing the right ventricle (inflow, trabecular, and outflow),[1,2] on the cut-off Z-values of the tricuspid valves and the pulmonary valves,[3] and finally on the presence of VCC with RVDCC.[4] RVDCC was defined as the presence of VCC associated with obstruction of more than 1 (>1) major coronary artery so that the major portion of the left ventricular blood supply was derived from the right ventricle.[6-10] A significant portion of PAIVS patients was found to have VCC (30% to 60%),[3,6,10-12] and RVDCC (9% to 26%).[3,4] Theoretically, RVD could result in a right ventricular steal phenomenon causing diastolic runoff (of the coronary blood flow) from the aorta to the right ventricle, lessening myocardial perfusion, jeopadizing the myocardium to infarction,[10] and potentially unmasking RVDCC.[6] RVDCC is the factor that precludes RVD in PAIVS patients by surgery[10] or by RPVV.[4] Potential right ventricular steal caused by VCC alone, but without RVDCC, did not preclude successful RVD.[10] In this report, Patient 2 had PAIVS associated with VCC, but without RVDCC, successfully underwent RVD by RPVV without incurring significant sequalae. Thus, precise definition of coronary arterial anatomy by angiography is warranted in neonates with PAIVS.[4,8,10] Another decisive factor predicting the feasibility of establishing continuity between the right ventricle and the pulmonary artery is the distance between the distal right

ventricular outflow tract and the proximal main pulmonary artery. There are 2 ways to ascertain this distance. Usually, we used a targeting catheter to label its tip on the membranous pulmonary valve from the right ventricular outflow tract, confirmed its preferred lie by contrast injection or blood drawing manually, and applied a landmark catheter in the ductus or near the ductus in the descending aorta for cine contrast injection. Another way to show this distance was described as double catheter technique.[13] Based on clinical implication, PDA in patients with PAIVS could be angiographically classified into two kinds of morphology: 1) upstream PDA from the descending aorta, and 2) downstream PDA from the transverse aortic arch. The clinical implication of ductal morphology cannot be overemphasized, because transaortic approach of PDA downstream from the transverse aortic arch to place a landmark catheter into the pulmonary artery will be demanding. With the aid of a guiding wire, a 4-French Multi-Purpose Catheter could be guided to advance with less difficulty from the downstream PDA to the main pulmonary artery, serving as a landmark catheter.[14,15] The transaortic approach from the descending aorta will be much easier in cases with upstream PDA. Good placement of a landmark catheter, referenced as a "roadmap", would be instructive during transcatheter perforation.[16] Finally, the anatomic lie of the right atrial appendage could be mistaken as that of the right ventricular outflow tract in patients with PAIVS, in whom the right atrium and the right atrial appendage are hugely dilated due to severe tricuspid regurgitation. As we can imagine, a fatal catastrophe will ensue if the right atrial appendage is the mistaken targeted site of radiofrequency perforation.

Thirdly, RPVV was conventionally performed by approaching antegradely from the femoral vein, with only rare exceptions that were unconventionally performed by approaching retrogradely or transductally from the descending aorta,[17] or from the right carotid artery.[18] Rather, the unconventional or transductal approach presented some problems intrinsic to neonates with PAIVS: 1) vascular compromise after cannulation of the artery using a larger catheter, 2) vessel damage or vasoconstriction caused by sawing action of the catheter on the ductus, which is the sole source of pulmonary blood flow, and 3) a more difficult angle to perforate the membrane centrically from a larger main pulmonary trunk above into a smaller right ventricular outflow tract below, which is liable to cause cardiac tamponade or perforation.[16] Application of flexible 2-French radiofrequency catheters offered some advantages to the pediatric cardiologist performing RPVV for the neonates with PAIVS, including: 1) avoidance of vascular compromise by using smaller catheters for cannulation, 2) timesaving without exchanging guidewire for balloon dilatation, and 3) avoidance of cardiac perforation frequently encountered by using a standard, but stiffer, 5-French radiofrequency catheter that will straighten the 5-French guiding catheter and result in cephalad misdirection toward the free wall of the right ventricular outflow tract.[16,18]

After successful radiofrequency valvulotomy, gradational balloon valvuloplasty is recommended. Initially, a small coronary balloon catheter (2.0 mm in diameter and 20.0 mm in length, HAYATE, Terumo, Tokyo, Japan) could be used to predilate the already perforated pulmonary valve. Later on, some larger coronary balloon catheters could be used to dilate the pulmonary valves gradationally.

Another adjunctive role of the ductus for balloon valvuloplasty is exemplified via the creation of a transductal guidewire rail,[19] which involved using a snare catheter to grasp the guidewire from the descending aorta that may facilitate the balloon catheter to pass across the small pulmonary valve opening. However, the prerequisite is that the ductus should be upstream, rather than downstream, in morphology. Prolonged stay or irritation of a guidewire within the downstream ductus during manipulation to create a transductal guidewire rail may cause ductal constriction and can be lethal. If a transductal guidewire rail is created, one should be careful not to pull the guidewire forcefully with the snare catheter to avoid a sawing action on the ductus, which is harmful.[19] Thus, transductal railing maneuver should carefully be used in patients with upstream PDA. However, a transductal railing maneuver can be appallingly demanding in patients with downstream PDA.

Finally, the mechanism of cyanosis or persistent oxygen desaturation should be sought out desperately before application of surgery or inhalation of the nitric oxide. Anatomic obstruction due to decreased pulmonary venous inflow (obstructive total anomalous pulmonary venous connection, congenital mitral stenosis, and congenital pulmonary vein stenosis) or inadequate pulmonary arterial blood flow (stenotic or obstructive Blalock-Taussig shunt or infundibular spasm) should be discriminated from the reactive pulmonary vasoconstriction.[19] In this report, Patients 1 and 4 suffered greatly after RPVV. In both patients, echocardiography with Doppler and bubble-contrast test showed spontaneous closure of the PDA, severe tricuspid regurgitation, unchanged pressure gradient across the pulmonary valve, and significant interatrial right-to-left shunt via the atrial septal defect secundum, which may be due to either inadequate effective pulmonary blood flow from the right ventricle with post-procedure infundibular spasm or pulmonary hypertension with reactive pulmonary vasoconstriction. A clinical trial of inhaled nitric oxide may help differentiate anatomic obstruction from reactive pulmonary vasoconstriction.[20,21] Failure to respond to inhaled nitric oxide in neonates after cardiac surgery should prompt investigations for further surgical correction. However, unexpected cyanosis can be caused by reactive pulmonary vasoconstriction in newborns after placement of a patent systemic-to-pulmonary shunt.[19] In Patient 1, the first episode of cyanosis was unresponsive to inhaled nitric oxide, which might have been related to inadequate pulmonary blood flow due to either a stenotic Blalock-Taussig shunt or infundibular spasm. In spite of placing a

second patent Blalock-Taussig shunt, a second episode of cyanosis due to pulmonary hypertension occurred and responded well to nitric oxide inhalation. There are many possible reasons for cyanosis after the interventional procedure in patients with PAIVS, including infundibular spasm, spontaneous closure of PDA, severe tricuspid regurgitation with restrictive interatrial right-to-left shunt, and severe pulmonary regurgitation due to pulmonary hypertension with reactive pulmonary vasoconstriction. However, persistent pulmonary hypertension with reactive pulmonary vasoconstriction after the procedure may be a rare cause of cyanosis in PAIVS following RPVV. Inhaled nitric oxide has been proved to be a potent selective pulmonary vasodilator, which is effective in the management of postoperative pulmonary hypertension in patients with congenital heart disease.[22-26] However, in spite of routine application in the clinical practice, nitric oxide inhalation has rarely been documented in the English literature in the postoperative care of patients with PAIVS treated by RPVV.

RPVV can be a treatment of choice for neonates with PAIVS, provided that there is patent infundibulum and no RVDCC. The cut-off values of the tricuspid valve and pulmonary valve prerequisite for RPVV remain to be determined, because of limitations of short-and mid-term follow-ups in a small series of only 6 patients in the present study. Long-term follow-up of their biventricular status is warranted to determine the cut-off Z-values while considering RPVV as a definitive treatment for patients with PAIVS. Though with potential right ventricular steal in VCC, VCC alone did not preclude performance of RPVV. Inhalation of nitric oxide can be used to treat pulmonary hypertension and to increase oxygenation, which would be helpful to wean patients off $PGE_1$ and mechnical ventilation before a second try of balloon dilatation or surgical repair for patients with PAIVS.

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