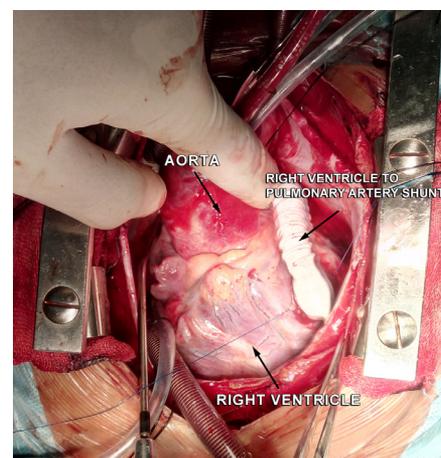# Mid-Term Outcome of Right Ventricle to Pulmonary Artery Shunt for Older Children and Young Adults With Ventricular Septal Defect, Pulmonary Atresia, and Hypoplastic Pulmonary Arteries

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Management strategy for patients of ventricular septal defect and pulmonary atresia (VSD/PA) with hypoplastic pulmonary arteries presenting in late childhood or adolescence is still controversial. We present our experience with the use of right ventricle-pulmonary artery shunt (RV-PA) in management of this entity. Between January 2014 and April 2018, 25 patients of VSD/PA underwent valveless RV-PA shunt at our center. The size of the RV to PA shunt was calculated as half the expected diameter of the main pulmonary artery. We retrospectively reviewed the data from hospital records. Follow-up data were recorded from outpatient records or via telephone. Mean age of the cohort was 12.25 ± 3.18 years. There was 1 early and 1 interstage mortality. None of the patient developed acute renal failure, ventricular dysfunction, and arrhythmias. At interstage follow-up of 8.28 ± 3.7 months, both Nakata index (from 66.23 ± 24.12 to 185.8 ± 58 mm^2/m^2) and McGoon ratio (0.9 ± 0.22 vs 1.84 ± 0.4) increased significantly compared to preoperative value, whereas RPA-LPA ratio was not significantly changed (1.095 ± 0.39 vs 1.01 ± 0.56, $P = 0.63$). Prerepair pulmonary vascular resistance in 17 patients, who underwent complete repair, was 2.9 ± 0.69 woods unit/m^2. Postrepair right

RV-PA shunt in VSD pulmonary atresia.

Central Message

RV-PA shunt is an effective strategy for uniform growth of hypoplastic pulmonary arteries and provides low pulmonary arterial bed resistance for definitive intracardiac repair.

Perspective Statement

Older children/adolescence and young adults presenting with ventricular septal defect/pulmonary atresia with hypoplastic pulmonary arteries still remain a difficult subset to manage. Right ventricle to pulmonary artery (RV-PA) shunt provides an effective alternative option for rapid and uniform growth of central pulmonary arteries in this subset of patients.

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ventricle-left ventricle pressure ratio was 0.5 ± 0.14. There was no early or late mortality and none of the patient required conduit revision or VSD fenestration. On follow-up of 25.75 ± 17.94 months, 16 patients were in NYHA I and 1 patient was in NYHA II. Appropriate-sized RV-PA shunt is an effective strategy for achieving balanced pulmonary artery growth in VSD/PA with hypoplastic pulmonary arteries presenting late without the risk of pulmonary over circulation or systemic malperfusion.

INTRODUCTION

Ventricular septal defect with pulmonary atresia (VSD/PA) is a complex congenital cardiac malformation. It has widely variable spectrum; on one side of the spectrum, there are patients with well-developed central pulmonary arteries without major aorto-pulmonary collaterals (MAPCAs), whereas on the other side, there is completely absent central pulmonary arteries with both of the lungs supplied entirely by MAPCAs only.[1−3] Most of the patients lie somewhere in the middle of the spectrum. Devoid of management in time, >90% of the patients with VSD/PA die in their first decade of life.[1−3] In the developed countries, these patients usually undergo surgical correction in their infancy or early childhood. However, in the developing countries, the first time presentation of these patients is either in their adolescence or in the adulthood. These are naturally selected patients with sufficient pulmonary blood flow either through a patent ductus arteriosus or MAPCAs. These patients become symptomatic due to decrease in pulmonary blood flow because of increasing stenosis of either MAPCAs or the PDA. Some of them have hypoplastic central pulmonary arteries and management of this subset still remains a challenge.

There are 2 schools of thought, for the management of patients with VSD/PA with hypoplastic central pulmonary arteries and MAPCAs presenting in infancy. One prefers unifocalization of all MAPCAs with single-stage primary repair, while the other prefers staged repair where early palliation by unifocalization of MAP-CAs with either systemic to pulmonary artery shunt or a right ventricle (RV) to pulmonary artery (PA) continuity constitutes first stage followed by complete repair preferably before 2 years of age.[4−10] However, there has been paucity of data regarding management of the older children/adolescence (≥8 years) and young adults with VSD/pulmonary atresia and hypoplastic pulmonary arteries. This category deserves a special management challenge due to limited availability of surgical options. Further, there have also been reports of slower growth of PAs if modified BT shunt procedure is performed in the older children,[11,12] while performing central shunt leads to balanced growth of pulmonary arteries at the expense of massive volume overload of the left heart, systemic hypoperfusion, significant coronary artery run-off, and development of pulmonary arterial hypertension.[13]

In the literature review, a good outcome with right ventricle to pulmonary artery (RV-PA) shunt in neonates, especially operated for Norwood procedure and in infants and children operated for VSD/pulmonary atresia has been well documented.[14−16] To the best of our knowledge, there is still a scarcity of data regarding use of RV-PA shunt in older children/adolescence and young adults with VSD/pulmonary atresia having hypoplastic pulmonary arteries. In our institute, we are following the strategy of staged repair in this group of patients. In first stage, valveless RV-PA conduit with or without unifocalization of MAPCAs is performed, while the complete repair is performed in the second stage. In this article, we present our experience of right ventricle to pulmonary artery shunt in management of this entity.

MATERIALS AND METHODS

Patients and Methods

A retrospective study from January 2014 to April 2018 of 25 patients having VSD, PA with hypoplastic branch PAs was done who underwent valveless RV-PA shunt at our center. The study protocol was approved by the institutional ethics committee and written informed consent from the parents has been taken. The perioperative data including echocardiogram, angiographic studies, and computed tomography were reviewed from hospital records. Follow-up data were recorded from outpatient records or via telephonic conversation.

Surgical Technique

All patients were operated through median sternotomy using moderate hypothermic (32°C) cardiopulmonary bypass (CPB). Patients in whom MAPCAs unifocalization was planned, MAPCAs were dissected and looped. Before establishing CPB, distal anastomosis of valveless conduit was performed either end to end at pulmonary confluence or end to side with main pulmonary artery (MPA). CPB was then established with ascending aorta and right atrial cannulation. At this stage, PDA, if present, was ligated. The MAPCAs that were planned for unifocalization were ligated at origin, divided, and anastomosed in the end to side fashion with ipsilateral pulmonary artery. After cross clamping of the aorta, cold blood cardioplegia was delivered and right ventriculotomy was performed at proposed site. Hypertrophied muscle bundles were minimally resected and proximal end of RV-PA shunt was performed in end to side fashion using 6-0 polypropylene sutures (Fig. 1). In the last 6 patients, proximal continuity of the RV-PA shunt was established using "DUNK Technique."[17] (Kindly refer
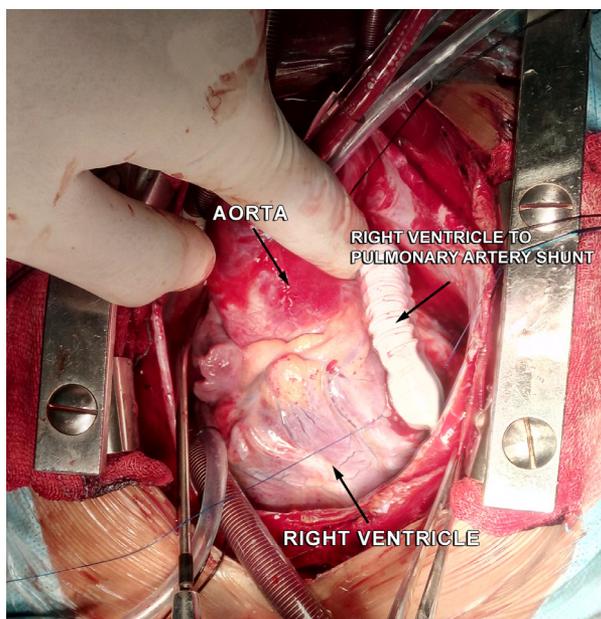
Figure 1. Intraoperative picture of a patient of ventricular septal defect and pulmonary atresia with completed right ventricle to pulmonary arterial shunt using 8 mm PTFE graft.

to the supplementary video for further details.) After completion of proximal anastomosis, heart was deaired, aortic cross clamp was released, and the aortic root was vented. The CPB was gradually weaned off and routine chest closure was performed. The patients were shifted to intensive care unit for further management.

Strategy for MAPCAs

The MAPCAs which are the sole supply to the segment of the lung were unifocalized preferably during the first stage of surgery. While the MAPCAs with stenosis and/or additional source of pulmonary blood flow were left alone for the spontaneous regression. The MAPCAs that were still patent at the time of complete repair were either surgically ligated or coiled.

Post-Op Management

All the patients were managed as per our intuitional ICU protocol. They were put on heparin infusion once there was no risk of bleeding oral antiplatelet, tablet aspirin (5 mg/kg, max 75 mg) was started, and heparin was discontinued once oral feeds were started. All patients were discharged on diuretics and aspirin.

Follow-Up Data

After discharge, the patients were followed up at 1 month and then at 3-month interval. Transthoracic echocardiogram was performed during each visit. After 6 months, CT angiography was performed to assess the adequacy of branch PAs and the status of MAPCAs. Adequate growth of the pulmonary artery was defined as Nakata index of >150 mm^2/m^2. Patients were also catheterized before proceeding for the complete repair.

Statistical Analysis

Statistical analyses were performed with the SPSS 22.0 software (SPSS, Inc., Chicago, IL). Continuous variables are expressed as mean with standard deviations. Categorical data are expressed as percentages. Continuous variables before and after RV-PA shunt were compared using paired Student's t test. P value of <0.05 was considered as statistically significant.

RESULTS

Mean age of the patients was 12.25 ± 3.18 years, with age ranging from 8 to 21 years. Mean preoperative saturation, Nakata index (21 patients)/neopulmonary arterial index (4 patients), and McGoon ratio were 73 ± 7.18%, 66.23 ± 24.12 mm^2/m^2, and 0.9 ± 0.22, respectively. Only 2 patients had Nakata/neopulmonary index of >90 mm^2/m^2. Thus, 23 patients had extreme hypoplastic pulmonary arteries. The sources of the pulmonary blood flow were MAPCAs alone (20 patients), PDA with MAPCAs (3 patients), and PDA alone (2 patients). Other demographic and morphologic details of the patients are presented in Table 1.

Mean CBP time and aortic cross-clamp time were 83.8 ± 27.8 minutes and 24.6 ± 15.5 minutes, respectively. A ringed polytetrafluoroethylene (PTFE, IMPRA, Bard Peripheral Vascular Inc., USA) grafts in 22 patients and a polyester (Dacron) graft in 3 patients. Unifocalization was performed in 4 patients: 2 concomitantly during the shunt surgery and as a staged procedure in 2 patients (in a second stage followed by complete repair). Two patients required aortic valve replacement at the time of shunt surgery for associated severe aortic regurgitation. Other concurrent procedures performed have been described in Table 2.

Table 1. Baseline Characteristics of the Study Cohort

Variables	Value ($n = 25$)
Age (mean ± SD, y)	12.25 ± 3.18
Sex (male, n, %)	18 (72%)
Weight (mean ± SD, kg)	23.74 ± 7.01
Height (mean ± SD, cm)	136.4 ± 11.78
BSA (mean ± SD, m^2)	0.96 ± 0.17
SpO_2 (mean ± SD, %)	73 ± 7.18
Systolic blood pressure (mean ± SD, mm Hg)	94.29 ± 7.28
Diastolic blood pressure (mean ± SD, mm Hg)	59.29 ± 4.57
Hemoglobin (mean ± SD, mg/dL)	16.86 ± 3.18
Nakata index (mean ± SD, mm^2/m^2)	66.23 ± 24.12
McGoon ratio (mean ± SD)	0.9 ± 0.22
RPA size (mean ± SD, mm)	5.8 ± 1.65
Z-score RPA (mean ± SD)	−4.4 ± 1.6
LPA size (mean ± SD, mm)	6.3 ± 1.7
Z-score LPA (mean ± SD)	−3.08 ± 1.8
RPA:LPA ratio (mean ± SD)	1.01 ± 0.56
Associated anomalies	
Aberrant RSCA (n, %)	2 (8%)
Severe AR (n, %)	2 (8%)
mBT shunt blocked (n, %)	3 (12%)

AR, aortic regurgitation; BSA, body surface area; LPA, left pulmonary artery; MAPCA, major aortopulmonary collaterals; mBT, modified Blalock Taussig; PDA, patent ductus arteriosus; RPA, right pulmonary artery; RSCA, right subclavian artery; SpO_2, systemic saturation.

Table 2. Intraoperative Data of the Study Cohort

Variables	Value (n = 25)
Conduit sizes (mm)	
Ringed (PTFE)	
6 mm (n, %)	5 (20%)
8 mm (n, %)	11 (44%)
10 mm (n, %)	6 (24%)
Dacron	
12 mm (n, %)	3 (12%)
CPB time (mean ± SD, min)	83.8 ± 27.8
ACC time (mean ± SD, min)	24.6 ± 15.5
Associated procedure	
PA plasty (n, %)	6 (24%)
MAPCA coiling (n, %)	4 (16%)
MAPCA ligation (n, %)	4 (16%)
Unifocalization (n, %)	4 (16%)
Aortic valve replacement (n, %)	2 (8%)

ACC, aortic cross clamp; CPB, cardiopulmonary bypass; MAPCA, major aortopulmonary collaterals; PA, pulmonary artery; PTFE, polytetrafluroethylene.

Postoperative Course

Mean mechanical ventilation duration was 1.1 ± 0.97 days while the mean intensive care stay was 4.9 ± 1.59 days. None of the patient, in our series, developed acute renal failure, ventricular dysfunction, arrhythmias, early shunt occlusion, pseudoaneurysm formation, or sepsis. Two patients required re-exploration for mediastinal bleeding (in non-DUNK technique). Other postoperative complications have been shown in Table 3. There was 1 early death (4%) due to a sudden cardiac arrest 5 days after the surgery in postoperative ward. The cause of death in this patient could not be ascertained as autopsy of the patient was refused by the parents.

Table 3. Postoperative Parameters of the Study Population

Variables	Value (n = 25)
ICU stay (mean ± SD, d)	4.9 ± 1.59
Ventilation time (mean ± SD, d)	1.1 ± 0.97
Systolic blood pressure (mean ± SD, mm Hg)	90.71 ± 5.93
Diastolic blood pressure (mean ± SD, mm Hg)	56.43 ± 4.43
SpO$_2$ (mean ± SD, %)	89.3 ± 2.98
Complication	
Early mortality (n, %)	1 (4%)
Reintubation (n, %)	2 (8%)
Pleural effusion (n, %)	2 (8%)
Pericardial effusion (n, %)	2 (8%)
Re-exploration (n, %)	2 (8%)
Sepsis (n, %)	0
Acute renal failure (n, %)	0
Ventricular dysfunction (n, %)	0
Early shunt failure (n, %)	0
Shunt inflow stenosis (n, %)	1 (4%)
Pseudo aneurysm (n, %)	0

ICU, intensive care unit; SpO$_2$, systemic saturation.

Follow-Up and Complete Repair

The mean follow-up duration was 25.75 ± 17.94 months (Table 4A). There was one late mortality 7 months post shunt surgery. Actual cause of the death could not be ascertained, but at last follow-up visit prior to death, patient had respiratory tract infection with adequate growth of branch PAs and was awaiting for the complete repair.

As shown in Table 4B, Nakata index increased significantly compared to the preoperative value (66.23 ± 24.12 mm^2/m^2 vs 185.8 ± 58 mm^2/m^2, P < 0.0001). Similarly, McGoon ratio was also significantly increased compared to the preoperative value (0.9 ± 0.22 to 1.84 ± 0.4, P < 0.0001). The RPA Z-score increased from −4.4 ± 1.6 to −0.35 ± 1.3, while the mean LPA Z-score increased from −3.08 ± 1.8 to 0.04 ± 1.19. Increase in size of both branch PAs was also statistically significant (P < 0.0001, Fig. 2). The rate of combined growth of the branch PAs was 1.038 ± 0.62 mm/mo. There was no significant change in the tricuspid valve annular plane systolic excursion (TAPSE, 20.66 ± 4.68 vs 19.8 ± 5.76, P = 0.57) and the RPA:LPA ratio postoperatively (preoperative 1.095 ± 0.39 vs postoperative 1.01 ± 0.56, P = 0.633).

Till the last follow-up, all 22 surviving patients had achieved adequate pulmonary artery growth, and out of them, 17 patients underwent successful complete repair. One patient, who underwent RV-PA shunt 2 months prior to completion of the study, is still in the follow-up. Other 5 patients have achieved adequate growth of pulmonary arteries and are awaiting surgery.

As shown in Table 5, 17 patients who underwent complete repair, systolic, diastolic, and mean pulmonary artery pressures on cardiac catheterization were 58.57 ± 14.64 mm Hg, 7.14 ± 0.98 mm Hg, and 23.57 ± 5.09 mm Hg, respectively. Mean pulmonary vascular resistance index was 2.9 ± 0.69 woods unit/m^2. Post complete repair, right ventricle to left ventricle pressure ratio (pRV/LV) was 0.5 ± 0.14 (ranged from 0.3 to 0.6). There was no early or late mortality following complete repair at the mean follow-up of 21.57 ± 9.54 months. None of the patient required conduit revision or VSD fenestration. Sixteen patients remained in NYHA class I, while 1 patient was in NYHA II due to small residual VSD.

DISCUSSION

Lillehei first repaired VSD and pulmonary atresia in 1955 using the controlled cross circulation.[18] However, the concept of systemic rehabilitation of pulmonary arteries emerged with pioneering work of the Mayo and Melbourne group in the mid 80s[5,7] and Stanford group in the 90s.[10] Since then, there has been frameshift change in the management of these patients. The surgical management has evolved from multistage unifocalization followed by complete repair to single-stage unifocalization and complete repair to no unifocalization and staged repair (growth of native pulmonary arteries by RV-PA connection/shunt followed by complete repair). However, most of the authors emphasize on the maximum recruitment of pulmonary arterial segments in the early infancy and childhood for better early and long-term

Table 4. (A) Follow-Up Data of the Study Population. (B) Comparison of Different Variables Before and After RV-PA Shunt

Variables	Value (n = 24)
Duration of follow-up (mean ± SD, mo)	25.75 ± 17.94
Complete repair n, (%)	17/24 (70.8%)
Awaiting complete repair n (%)	5 (20.8%)
Interval to complete repair (mean ± SD, mo, n = 17)	8.28 ± 3.7
Interstage mortality (n, %)	1/24 (4.2%)
Late mortality (n, %)	0

Variables	Pre RV-PA Shunt (n = 25)	6 Months Post RV-PA Shunt (n = 23)	P Value
Weight (mean ± SD, kg)	23.74 ± 7.01	25.72 ± 8.29	0.42
BSA (mean ± SD, m^2)	0.96 ± 0.17	1.01 ± 0.19	0.47
SpO$_2$ (mean ± SD, %)	73 ± 7.18	90 ± 3.4	<0.0001
Hemoglobin (mean ± SD, mg/dL)	16.86 ± 3.18	11.14 ± 1.05	<0.0001
Systolic blood pressure (mean ± SD, mm Hg)	94.29 ± 7.28	90.71 ± 5.63	0.75
Diastolic blood pressure (mean ± SD, mm Hg)	59.29 ± 4.57	56.43 ± 4.43	0.71
Nakata index (mean ± SD, mm^2/m^2)	66.23 ± 24.12	185.8 ± 58	<0.0001
McGoon ratio (mean ± SD)	0.9 ± 0.22	1.84 ± 0.4	<0.0001
RPA size (mean ± SD, mm)	5.8 ± 1.65	11.05 ± 2.97	<0.0001
Z-score of RPA (mean ± SD)	−4.4 ± 1.6	−0.35 ± 1.3	<0.0001
LPA size (mean ± SD, mm)	6.3 ± 1.7	10.48 ± 2.37	<0.0001
Z-score of LPA (mean ± SD)	−3.08 ± 1.8	0.036 ± 1.19	<0.0001
RPA:LPA ratio (mean ± SD)	1.01 ± 0.56	1.095 ± 0.39	0.63
TAPSE (mean ± SD)	20.66 ± 4.68	19.8 ± 5.76	0.57

BSA, body surface area; LPA, left pulmonary artery; RPA, right pulmonary artery; SpO$_2$, systemic saturation; TAPSE, tricuspid annular plane systolic excursion.
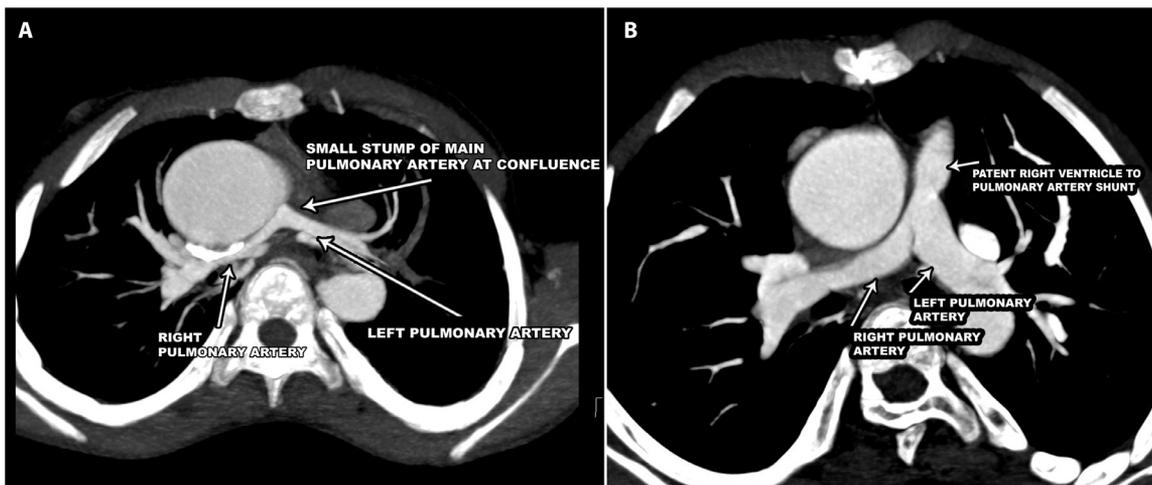

Figure 2. (A) Preoperative contrast computed tomography (CT) images of a 14 years' patient with ventricular septal defect and pulmonary atresia with hypoplastic branch pulmonary arteries (Nakata index 45 mm^2/m^2). (B) Postoperative contrast CT of the same patient 8 months postshunt showing balance growth of pulmonary arteries (Nakata index 290 mm^2/m^2).

results.[5,7,10,19−21] Further, there have also been reports of slower growth of PAs if modified BT shunt procedure is performed in the older children.[11,12] So the management strategy for the patients presenting in late childhood or in adolescence is still uncertain and in evolution. In 2008, Bradley et al[14] introduced RV-PA shunt as a first-stage palliation for the hypoplastic pulmonary arteries in the patients intended for future biventricular repair. However, data regarding its use in older children/adolescence with hypoplastic pulmonary arteries are still scarce.

Why This Approach?

In our study cohort, there was no continuity existed between MPA and RV and mostly a small residual stump of MPA was present near the bifurcation of branch PAs. So anatomy itself is unsuitable for establishment of direct RV-PA connection. Use of an autologous material like left atrial appendage (LAA) for establishment of direct continuity mainly in neonates has been reported.[22,23] But the application of this approach in older children is not feasible due to greater distance of RV infundibulum

Table 5. Prerepair and Intraoperative Parameters of the Patients Who Underwent Complete Repair

Prerepair cath data	Variables (n = 17)	Value (Mean ± SD)
	Systolic PA pressure (mm Hg)	58.57 ± 14.64
	Diastolic PA pressure (mm Hg)	7.143 ± 0.98
	Mean PA pressure (mm Hg)	23.57 ± 5.09
	PVRI (woods unit/m²)	2.9 ± 0.69
Intraoperative data	pRV/LV	0.5 ± 0.14

LV, left ventricle; PA, pulmonary artery; PVRI, pulmonary vascular resistance index; RV, right ventricle.

from the PA bifurcation. This approach (LAA) also has around 10−15% incidence of pseudoaneurysm formation further limiting its use in older children.[21,22] Previous studies also reported that for growth of pulmonary arteries rigid valveless conduit would be better as compared to pericardial tube or bovine jugular vein because of their tendency to form aneurysmal dilatation leading to energy loss in later.[24−26]

Advantages of RV-PA Shunt

Hemodynamic Profile

An important advantage of RV-PA shunt over modified BT or central shunt is the liberty to insert a relatively larger size shunt thereby decreasing the need of shunt revision for the shunt occlusion. It also has an advantage of minimizing the risk of pulmonary over circulation or coronary run-off even in patients with hypoplastic pulmonary arteries.[13,16,27] This is an important prerequisite in grown-up patients with hypoplastic pulmonary arteries. In our cohort, none of the patients developed hemodynamic instability, acute renal failure, or low cardiac output syndrome in the postoperative period suggesting absence of significant coronary run-off, systemic hypoperfusion, pulmonary over circulation, or the left ventricular volume overload denoting good hemodynamics after RV-PA shunt.

Selection of the Shunt Size

Selection of a proper size of shunt is also difficult task in older patients without any clear guidelines. Our criteria for the selection of shunt size were based on the expected size of main pulmonary artery (according to "Z" score) for body surface area (BSA) of the patient. We selected a shunt 50−60% of expected diameter of MPA. For an example, if a patient has a BSA of 1.3 mm², his expected main pulmonary artery size would be 2.1 cm. A shunt of 10 or 12 mm would be a suitable conduit for his RV-PA shunt. Absence of hemodynamic instability, acute renal failure, or the low cardiac output syndrome in our patient cohort in the postoperative period suggests absence of significant coronary run-off, systemic hypoperfusion, pulmonary over circulation, or the left ventricular volume overload due to indicating the proper selection of shunt size.

Growth of Pulmonary Arteries

In our study, the mean Nakata index was increased 3 times of the baseline at mean follow-up of 7.33 ± 3.4 months. Thus, the RV-PA shunt has the advantage of shorter interstage interval as described by others.[13,14] In our study, average growth rate of the branch PA was 0.5 mm/mo. Many authors have reported similar findings in their studies.[24,25,27,28] Further, the RV-PA shunt also leads to an uniform growth of PAs as evident by no significant change in ratio of RPA and LPA during the follow-up.

We believe that inclusion of ventricle as a pump for the shunt flow offers several advantages. The shunt flows only during systole thereby reducing the risk of left ventricular volume overload. Further, our analogy of choosing a valveless conduit is based on out of proportion growth of branch pulmonary arteries in patients with ToF with absent pulmonary valve syndrome and systemic arteries in patients with chronic aortic regurgitation. We believe that wide pulse pressure and not absolute systolic pulmonary arterial pressure allows greater stretching of the elastic lamina of the pulmonary arteries and hence the growth of PAs. The findings of our study confirm our analogy as also stated by others.[24,29] Apart from pulsatile flow, other factors responsible for rapid growth as cited by others are larger diameter of shunt employed and centrally placed distal anastomosis.[29]

Disadvantages of RV-PA Shunt

Despite many advantages, this procedure is not without shortcomings. Complications that may occur after RV-PA shunt include shunt inflow stenosis, pseudoaneurysm formation at anastomotic site, bleeding, perioperative hypoxia, and ventricular arrhythmias.[30] In our study, there was no incidence of early or late shunt failure, anastomotic pseudoaneurysm, or dehiscence. Probable explanation for this finding is use of rigid (ringed) valveless conduit instead of bovine jugular vein or pericardial tube which has the tendency to dilate and cause pseudoaneurysm formation.[24−26]

Conventionally, the proximal end of RV-PA shunt is anastomosed to the right ventriculotomy. This results in a high incidence of bleeding from the needle holes resulting in increased use of blood and blood products. In our series also, 2 patients required re-exploration for bleeding (both operated by conventional technique). We believe that the use of DUNK technique may ameliorates this problem as none of the last 6 patients operated by DUNK technique had bleeding problem in our series. We recommend larger studies to confirm the superiority of DUNK technique of the proximal anastomosis over conventional technique.

Another disadvantage of RV-PA shunt is regurgitation into the RV leading to RV volume overload. But, in our experience, this is a rare finding. The probable reasons are diminutive PAs, somewhat smaller shunt size (50−60% of expected PA size) and the less compliant hypertrophied RV. Further, we believe that as the size of the branch PAs increases, there is also increase in their compliance, reducing the chance of regurgitation.

In our series, we had 1 late mortality. This patient died 7 months after the surgery at home following an episode of respiratory tract infection. The cause of death in this patient was not certain. However, we believe that it may have been due to pulmonary and systemic circulation mismatch following lower respiratory tract infection.

Strategies for MAPCAs

We routinely do not unifocalize all the MAPCAs. Our strategy is to unifocalize the MAPCAs that are nonstenotic and a sole supply to a segment of the lung. From our previous experience, we have learnt that MAPCAs, which are additional supply to any of the lung segment, even after unifocalization tend to stenose/obliterate and they also hamper the growth of the native PAs. Also, in majority of the patients, those MAPCAs that are stenotic prior to the shunt surgery and left alone cannot be detected by angiography or CT angiography after the RV-PA shunt surgery as a result of establishment of the antegrade flow. This finding suggests that MAPCAs tend to obliterate as the blood flow through native pulmonary arteries increase. Similar findings have been reported by the other authors.[26,31]

Complete Repair and Catheterization Data

Following complete repair, pRV/LV ratio was 0.5 ± 0.14 (ranged between 0.3 and 0.6). Low postoperative pRV/LV despite high systolic pulmonary arterial pressure is indicative of absence of pulmonary hypertension. This finding is further substantiated by low diastolic PA pressure and low pulmonary vascular resistance index in all the 17 patients who underwent cardiac catheterization. This is a very important finding for rehabilitation of pulmonary arteries in this group of patients. The repair of VSD/PA is still complicated by higher pRV/LV due to incomplete rehabilitation of pulmonary arteries which might be prevented using this approach. Thus, RV-PA shunt offers adequate progressive growth of the pulmonary arteries while maintaining the systemic oxygen saturation until the time of complete repair, without the risk of developing pulmonary hypertension. This is particularly important for the developing countries where attrition of patients is common and abnegates the need of alternative palliative procedure. In an article by Belli et al, palliation in the form of modified BT/central shunt was the ultimate destination in almost 40% of their cohort of late adolescence and young adults with type B VSD/PA.[32] Though no pressure data are available, we believe that use of RV-PA shunt in these patients instead of modified BT/central shunt may had converted some of these patients into a suitable candidate for the biventricular repair.

Study Limitation

First, the study is retrospective in nature. Second, the number of patients included in study is very small. Third, there was no comparison group as modified BT or central shunt group. Therefore, we recommend randomized study including larger number of patients comparing the outcome of modified BT or central shunt with the RV to PA shunt. Another limitation of our study is limited duration of the follow-up. A longer follow-up would give us a better insight about the longevity of this approach.

CONCLUSION

Older children and young adults presenting with ventricular septal defect and pulmonary atresia with hypoplastic pulmonary arteries still remain a difficult subset to manage. Appropriate-sized RV-PA shunt is an effective strategy for achieving rapid and uniform growth of hypoplastic pulmonary arteries without the danger of pulmonary over circulation and allows for a definitive intracardiac repair.

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:
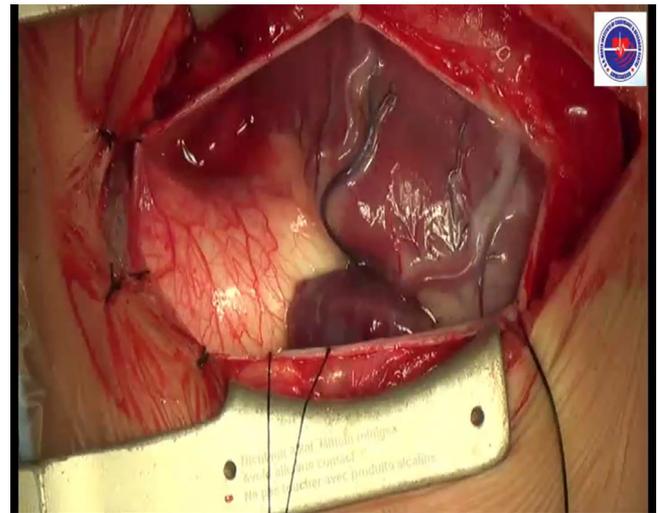
Video 1. Intraoperative video procedure of ventricular septal defect and pulmonary atresia with completed right ventricle to pulmonary arterial shunt using 8 mm PTFE graft.

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