Long-Term Outcome of the Right Ventricular Outflow Tract Palliation Procedure in Children With Cyanotic Congenital Heart Disease: A Case-Series Study

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Abstract

Background: The right ventricular outflow tract (RVOT) palliation has been shown to be a proper interventional procedure for lowering risk of mortality and improving clinical condition in cyanotic congenital heart disease (CHD) patients.

Objectives: The present study aimed to assess the consequences of RVOT palliation in patients with TOF.

Patients and Methods: This prospective case series was performed on 17 children who suffered from cyanotic CHD. The study end-points were assessed by pulse oximetry, echocardiography, and electrocardiography immediately and also 12 months after RVOT palliation procedure.

Results: The mean age of patients was 24.76 (median 10 months). Comparing laboratory and respiratory parameters 12 months after RVOT palliation showed a significant increase in arterial oxygen saturation (from 69.34 ± 13.07 to 86.29 ± 6.64 , $P = 0.001$), RPA index of right pulmonary artery (from 5.49 ± 1.67 mm to 7.59 ± 1.79 mm, $P < 0.001$), Z score of right pulmonary artery (from -1.56 ± 2.34 to 0.53 ± 2.55 , $P < 0.001$), LPA of left pulmonary artery (from 5.64 ± 1.88 mm to 8.06 ± 2.72 mm, $P < 0.001$), and also in Z score of left pulmonary artery (from -1.56 ± 2.33 to 0.78 ± 2.15 , $P = 0.001$). Also, a significant decrease in the spell rate (from 88.2% to 17.6%, $P < 0.001$), and in the rate of tricuspid regurgitation (from 23.5% to 11.8%, $P = 0.023$) was shown. Cardiac arrhythmia occurred in only one patient that was transient. Stent fracture was found in none of the patients. Stent stenosis was also found in one patient. One-year death occurred only in one child.

Conclusions: The right ventricular outflow tract palliation in children with cyanotic CHD leads to long-term favorable outcome regarding improvement in oxygen saturation, increase in Z score of both left and right pulmonary arteries and also considerable decrease in spell. Also, death and complications are rare following RVOT palliation.

Keywords: Palliative Care, Heart Defects, Congenital

1. Background

Congenital heart defects are frequently characterized by anatomical cardiovascular malformation occurring within intrauterine growth and neonatal development. These defects are mainly categorized as cyanotic or non-cyanotic defects based on the clinical condition (1). In cyanotic defects, systemic venous vessels bypass pulmonary circulatory system leading a right-to-left shunt in which higher resistance to right ventricular outflow results in more severe cyanosis symptoms and inappropriate systemic arterial oxygenation (2). Tetralogy of Fallot (TOF) as the most common cyanotic heart malformation is characterized by four structural defects including pulmonary infundibular stenosis, overriding aorta, ventricular septal defect (VSD), and right ventricular hypertrophy. In the lat-

est defect, due to the disarrangement of the external ventricular septum, the right ventricular wall increases in size to deal with the increased obstruction to the right ventricular outflow tract RVOT (right ventricular outflow tract) (3). This feature is now generally agreed to be a secondary anomaly, as the level of hypertrophy tends to increase with age (4).

The main goal for management and treatment of TOF as surgical repairing is considered with the lowest mortality and morbidity (5). This procedure can lead to prevent subclinical bacterial endocarditis, treatment of dehydration, continuous monitoring and treatment of anemia. Despite medical management of some children affected with TOF, repair of TOF defects by operation should be considered immediately (6). There is still controversy concerning

the initial

Management of severely symptomatic or duct-dependent infants with TOF, 'Fallot-type' double outlet right ventricle (DORV) or similar lesions with restriction of antegrade pulmonary blood flow and adverse risk factors for corrective surgery. These risk factors include low body weight, prematurity, young age (neonates), unfavorable pulmonary arterial anatomy, abnormal coronary distribution, significant comorbidities and critical preoperative condition (7). Recently, balloon valvuloplasty has been selected as a proper alternative, especially in those with valvular stenosis as the main cause for RVOT obstruction (8). In surgical repairing procedures, resection of infundibulum and repair of RVOT with a long-term following of surgical prognosis is considered (9). Despite favorable outcome of operation, replacement of pulmonary valve due to pulmonary insufficiency may be essential (10).

In total, RVOT palliation has been shown to be a proper interventional procedure for lowering the risk of mortality and improving clinical condition in cyanotic CHD patients, Figures 1 and 2. According to shortened hospitalization and less frequent in-hospital postoperative complications, this type of procedure can be replaced to palliative shunt surgery (11).

Figure 1. Postinterventional RV Injection at AP View After Transvalvular Implantation of Peripheral Blue Genesis Stent

2. Objectives

The present study aimed to assess the consequences of RVOT palliation in patients with cyanotic CHD.

Figure 2. A, Severe Infundibular Stenosis LAT RV Injection with RVH; B, Angiography After Implantation of Transvalvular Stent Demonstrating Complete Relief of Out-flow Tract Obstruction

3. Patients and Methods

This prospective case series was performed on 17 children who suffered from cyanotic CHD Figure 3. All patients were not possible to candidate for complete surgical repairing because of low age, hypoplastic PA branches or other comorbidities but were scheduled for a palliative procedure including RVOT obstruction removal by stenting or balloon pulmonary valvuloplasty (BPV) because of severe hypoxic spell and hemodynamic instability. The ex-

clusion criteria were the possibility for complete repairing of RVOT. Baseline characteristics included demographic characteristics, growth rate of pulmonary branches, arterial oxygen saturation, the number of hospitalizations before complete repairing, and early and long-term complications after procedure. The study endpoints were assessed by pulse oximetry, echocardiography, and electrocardiography immediately and also 12 months after RVOT palliation procedure. All echocardiography examinations were performed using the VIVID 3GE machine equipped with 3.6 MHz transducer.

Figure 3. Different Patterns Of Cyanotic CHD, Green: 53%, blue: 29%, yellow: 12%, purple: 6%

Results were presented as mean $\pm$ standard deviation (SD) for quantitative variables and were summarized by absolute frequencies and percentages for categorical variables. The normality of the data was assessed using normal probability plots and

Shapiro-Wilk's statistics. The change in quantitative variables was assessed using the paired t-test or nonparametric Wilcoxon signed-rank test. Statistical significance was determined as a P value of ≤ 0.05 . All statistical analyses were performed using SPSS software version 16.0 (SPSS Inc., Chicago, Illinois, USA).

4. Results

The mean age of the patients was 24.76 (median 10 months) and the mean body weight was 10.21 kg (median 7.5 kg). From a total of 17 patients, 41.2% were male. One of the patients had coronary artery anomaly. All patients had

spell on admission with active cyanosis (Table 1). Comparing laboratory and respiratory parameters 12 months after RVOT palliation (Table 2) showed a significant increase in arterial oxygen saturation (from 69.34 ± 13.07 to 86.29 ± 6.64 , $P = 0.001$) Figure 4, increase in RPA index of right pulmonary artery (from 5.49 ± 1.67 mm to 7.59 ± 1.79 mm, $P < 0.001$), increase in Z score of right pulmonary artery (from -1.56 ± 2.34 to 0.53 ± 2.55 , $P < 0.001$) Figure 5, increase in LPA of left pulmonary artery (from 5.64 ± 1.88 mm to 8.06 ± 2.72 mm, $P < 0.001$), increase in Z score of left pulmonary artery (from -1.56 ± 2.33 to 0.78 ± 2.15 , $P = 0.001$) Figure 6, decrease in the spell rate (from 88.2% to 17.6%, $P < 0.001$), and also decrease in the rate of tricuspid regurgitation (from 23.5% to 11.8%, $P = 0.023$). The mean hemoglobin level remained unchanged (from 15.15 ± 2.96 to 14.25 ± 2.20 , $P = 0.667$). Regarding postsurgical outcome of RVOT palliation, cardiac arrhythmia occurred in only one patient (5.9%), and another patient (5.9%) had overcirculation. Stent fracture was found in none of the patients. Stent stenosis was also found in one patient. There was no complication during BPV, but in one patient from 12 patients who underwent BPV, cyanotic spell was returned. Although need to intubation was revealed in 52.9% of children, some of the patients were intubated. One-year death occurred only in one child.

Figure 4. Mean Arterial Oxygen Saturation Before and After Intervention

5. Discussion

To assess one-year outcome of RVOT stenting and BPV as a useful palliative modality for young children with cyanotic CHD, the present case series study described the clinical

Table 1. Baseline Characteristics of Study Children with Tetralogy of Fallot

No	Age, mo	Weight	Gender	Coronary Defect	Spell	O ₂ SAT (Primary)	Active Cyanosis
1	5	7.5	Male	No	Yes	56	Yes
2	12	6.3	Female	No	Yes	55	Yes
3	1.5	4.3	Male	No	Yes	69	Yes
4	213	48	Male	No	Yes	85	Yes
5	19	9.8	Male	No	Yes	60	Yes
6	9	6	Male	No	Yes	76	Yes
7	24	10	Female	No	Yes	80	Yes
8	34	8.5	Female	No	Yes	80	Yes
9	1.5	5.4	Female	No	Yes	70	Yes
10	22	10	Male	Yes	Yes	77	Yes
11	14	10	Male	No	Yes	80	Yes
12	6	7.3	Female	No	Yes	80	Yes
13	10	7.5	Male	No	Yes	50	Yes
14	5	7.3	Male	No	Yes	72	Yes
15	31	13	Female	No	Yes	85	Yes
16	7	6.8	Male	No	Yes	42	Yes
17	7	5.8	Female	No	Yes	60	Yes

Table 2. Laboratory and Respiratory Indices Before and 12 Months After Right Ventricular Outflow Tract Stenting

O ₂ sat1	O ₂ sat2	Spell 1	Spell 2	Hb1	Hb2	RPA1	RPA2	RPAZ1	RPAZ2	LPA1	LPA2	LPAZ1	LPZ2	TR1	TR2	No
56.0	90.0	1	0	19.3	15.2	5.0	8	-2.0	0.5	3.8	9	-3.5	1	0	0	1
55.0	85.0	1	0	17.4	15.0	6.0	9	1.5	4	7.5	9.5	2.0	4	0	0	2
69.0	77.0	1	0	12.1	14.0	5.0	6.2	0.0	1	3.0	5	-3.0	-1	0	0	3
85.0	95.0	0	0	19.9	17.0	6.8	9	-5.0	-3	6.5	9	-5.0	-3	1	0	4
60.0	70.0	1	1	18.9	20.0	5.5	6.3	-2.0	-1	4.8	6.5	-2.5	0	0	0	5
76.0	95.0	1	0	17.5	16.0	6.3	7.3	1.0	1	6.6	7.5	1.0	1	0	1	6
80.0	90.0	1	0	14.7	13.0	4.0	6	-4.5	-1	4.5	5.3	-4.0	2	0	0	7
80.0	87.0	1	1	16.0	15.0	10.0	10	3.0	2	10.0	10	3.0	2	0	0	8
70.0	84.0	1	0	10.4	12.0	4.0	10.5	-2.5	6	4.0	10	-2.5	3	0	0	9
77.0	90.0	1	0	13.8	12.1	8.0	10	0.5	2	8.0	15	0.5	4	0	0	10
80.0	86.0	1	0	14.0	13.0	4.8	5.5	-3.5	-2	4.9	5.6	-3.5	-2	0	0	11
80.0	90.0	1	0	15.0	14.0	5.3	8	-1.0	1	6.5	10	0.0	3	0	0	12
50.0	93.0	1	0	11.7	12.0	5.1	8.8	-1.5	2.5	5.2	7.5	-1.5	1	0	0	13
72.0	80.0	0	0	14.0	14.0	4.0	5.8	-3.0	-2	4.5	6	-2.5	-0.5	0	0	14
85.0	90.0	1	0	17.4	16.0	4.5	5.4	-4.0	-3.5	7.0	9	-1.5	0	1	0	15
42.0	85.0	1	1	14.9	11.9	6.0	6.2	0.5	0.3	6.0	8.0	0.5	0	1	0	16
60.0	80.0	1	0	10.5	12.0	3.0	5.7	-4.0	1	3.0	4	-4.0	-2	1	1	17

Abbreviations: Hb, hemoglobin; LPA, Left pulmonary artery; RPA, Right pulmonary artery; RPA Z or LPA Z, Z score; Spell 1, before intervention; Spell 2, after intervention; TR, tricuspid regurgitation.

cal characteristics and prognosis of the children with cyanotic CHD. Overall, our study could demonstrate favorable consequences of RVOT palliation within 12 months after operation with significant increase in respiratory capacity, improvement in arterial oxygen saturation and compliance of right and left pulmonary arteries. Also, this proce-

dure led to rare mortality and morbidity within a year after operation that only one child suffered post-procedural morbidity including arrhythmias, tricuspid regurgitation and need to readmission. However, the study had two important points. First, the change in serum hemoglobin level remained insignificant. Moreover, more than half

Figure 5. Mean LPA Diameter Before and After Intervention

Figure 6. Mean RPA Diameter Before and After Intervention

of the children needed to intubation and oxygen support; however, this requirement did not lead to increased risk of mortality or life-threatening events.

Reviewing the literature confirms our findings with respect to the outcome of RVOT stenting in TOF children. In a similar study by Castleberry et al. (12), following RVOT stenting led to improvement of oxygen saturation from 71% to 94%, no stent fractures occurred and only one patient had repair 10 days after stent placement secondary to stent malposition and tricuspid valve injury; however, their study covered a short-term follow-up study. In Dohlen study (13), RVOT stenting improved arterial oxygen saturation from a median of 73% to 94%. Median Z-score for the left pulmonary artery increased from -4.9 before stent implantation to -1.5 before surgical repair. Median Z-score for the right pulmonary artery increased from -3.7 to -0.8. There were no procedural complications. Six patients had undergone successful repair. There were no deaths. In a

similar study by Cools et al. (14), the procedure resulted in adequate palliation with good antegrade flow to the pulmonary arteries and near normal saturations in all three patients (> 92%); there was no associated morbidity. Additional transvenous stenting was required in all patients because of progressive muscular infundibular stenosis after a median of three months. Two out of three patients evolved to full repair at the age of 5 months and one patient with multiple hilar stenosis requires additional percutaneous procedures through the stented RV outflow tract. In another study by Barron et al. (15), stenting improved saturations from 72% to 92%. There was no operative mortality after complete repair. There was one late death at three months due to chronic lung disease. The median left PA Z-score increased from a preinterventional value of -1.27 to a presurgical value of +0.11. Also, the median right PA Z-score increased from -2.02 to -0.65 over the preinterventional and presurgical time intervals. Growth was greatest in the right PA. Finally, in a study by Dryzek et al. (16), an increase occurred in blood saturation of up to 89%. There were neither rhythm nor conduction disturbances in the control ECG after the procedure.

5.1. Conclusion

In total, it can be concluded that the procedure of RVOT palliation in children with cyanotic CHD may lead to long-term favorable outcome regarding improvement in oxygen saturation, increase in Z score of both left and right pulmonary arteries and also considerable decrease in SPELL and until the end of the study, one of the RVOT stent that underwent complete surgical repair was extracted successfully without any problem. Also, death and complications are rare following RVOT stenting and BPV; thus, this procedure can be very safe and effective in the treatment of cyanotic CHD children.

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