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Right Ventricular Outflow Tract Stenting as the First Option for Palliation of Neonates with Severe Tetralogy of Fallot

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Abstract

Background: Neonates with Tetralogy of Fallot (TOF) might be prostaglandin dependent and cannot be discharged from the hospital except after complete correction or palliation. Palliation could be achieved by modified Blalock-Taussig Shunt (BTS), ductal stenting, or right ventricular outflow tract (RVOT) stenting.

Objective: The objective of this study was to present our experience of palliating neonates with severe TOF who require augmentation of the pulmonary blood flow by RVOT stenting.

Methods: This study was conducted on a series of patients who had RVOT stenting in PSCC-Qassim, Saudi Arabia, from August 2016 till December 2019. Cases that had RVOT stenting are included. The patient's clinical data, echocardiography, cardiac catheterization, and follow-up data were recorded. Data were analyzed using The Microsoft Excel data analysis tool.

Results: During the study period, four consecutive patients with a diagnosis of TOF with significant cyanosis underwent RVOT stenting. The mean age and weight were 2.8±1.7 days and 2.9±0.1 kg respectively. All were on prostaglandin infusion before the intervention. The mean saturation before and after RVOT stenting was 63% (±11%) and 82% (±4%) respectively ($p = 0.018$). One patient required RVOT stent dilatation after 4 months. All patients are alive and had complete TOF repair at a median age of 6.5 months (range 6 to 12 months). The median follow-up period is 13 months. The median branch PAs diameter and Z-score before RVOT stenting and immediately before complete TOF repair were 2.8 mm (Z score: -2.5) and 5.5 mm (Z score: -0.5 to -1.16) respectively ($p = 0.001$).

Conclusion: RVOT stenting is a reasonable alternative palliation for patients with severe TOF. The technique of stent deployment in the RVOT is challenging and requires pre-intervention planning and discussion. Using soft catheters, coronary wires, and the premounted coronary stents facilitate crossing of the RVOT and pulmonary valve and then stent deployment.

Keywords: Tetralogy of Fallot, Neonatal cyanosis, RVOT stenting, Neonatal TOF repair

Abbreviations / Acronyms:
BTS: Blalock-Taussig shunt, **PA:** Pulmonary Arteries, **CHD:** Congenital Heart Disease, **TOF:** Tetralogy Of Fallot, **PA/IVS:** Pulmonary Atresia with Intact Ventricular Septum, **PA/VSD:** Pulmonary Atresia with Ventricular Septal Defect, **PG infusion:** Prostaglandin infusion, **RVOT:** Right Ventricular Outflow Tract, **LPA:** Left Pulmonary Artery, **MPA:** Main Pulmonary Artery.

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1. Introduction

Cyanotic congenital heart diseases (CHD) with no forward pulmonary blood flow include patients with pulmonary atresia (PA), and any related combination like those with PA with intact ventricular septum (PA/IVS), PA with ventricular septal defect (PA/ VSD), or VSD with severe right ventricular outflow tract (RVOT) obstruction as in patients with severe tetralogy of Fallot (TOF). It includes also patients with single ventricle pathologies associated with significant pulmonary or subpulmonary stenosis or PA at the extreme. Such patients will require Prostaglandin infusion to maintain the patency of the ductus arteriosus as a source of pulmonary blood flow. They cannot be discharged from the hospital except after securing some sort of pulmonary blood flow either by ductus arteriosus stenting, modified Blalock–Taussig shunt (BT shunt), RVOT stent, or even by neonatal complete surgical repair (in certain lesions and selected patients) (1-3). Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease (4). Some neonates with TOF will have severe restriction of the pulmonary blood flow and desaturation. The clinical manifestations are related to the severity of the RVOT obstruction, the anatomy of the main pulmonary artery and pulmonary artery branches, and the patient's general condition. Patients with severe TOF might need to be given prostaglandin infusion, as a bridge to palliation or complete neonatal TOF repair. The decision regarding the type and timing of intervention will depend upon: the patient's weight and maturity, the anatomy of the RVOT, the status and anatomy of the ductus arteriosus, and the anatomy of the main and branch pulmonary arteries and the center experience in neonatal interventions and neonatal cardiac surgery (5).

2. Material and Methods

2.1. Methods

During the period from August 2016 till December 2019, 4 consecutive patients with severe TOF had RVOT stenting in our center as a palliative procedure. The patient's demographic, echocardiographic, catheterization as well as ICU data were reviewed. Follow-up data were reviewed including the saturation, the status of the branch pulmonary arteries, the requirement for unplanned intervention as well as the timing of complete surgical repair. The patient’s saturation, as well as branch pulmonary arteries size before RVOT stenting and immediately before complete TOF repair, were reviewed and compared. The study was approved by the institutional IRB. The Microsoft Excel data analysis tool was used for statistical analysis. Continuous variables were expressed as mean ± Standard deviation or a median and range as appropriate. Categorical variables were expressed as numbers and percentages.

Under general anesthesia, the patient will be prepped and draped as per protocol. One of the femoral veins will be accessed percutaneously using a 4 or 5 Fr sheath. It is better to have a Vaygon cannula in the femoral artery for hemodynamic monitoring as well as for possible requirements for ductal stenting in case RVOT stenting is not feasible. A 4 Fr MPA2 catheter is used to engage the right side to the RV and RVOT. RVOT hand injection angiogram will be performed. To cross to the RVOT, sometimes we have to use a soft hydrophilic catheter like the 4 Fr Glide catheter ((Terumo) (curved tip)) over a hydrophilic guidewire. The wire followed by the catheter might be advanced to the ascending aorta and then pulled gradually to engage the RVOT/sub-pulmonary area. With the help of a curved tip catheter (JR, MPA2, or a Curved tip Glide catheter), a coronary wire will be manipulated to cross the RVOT and advanced to the distal branch PAs. The catheter will be removed. A BARE METAL, premounted, coronary stent will be advanced over the wire to the RVOT. The stent will be deployed in the RVOT making sure to cover the whole infundibulum and cross the pulmonary valve till the mid-portion of the main pulmonary artery or even proximal to PAs bifurcation. It is better to advance the stent in a long sheath or a guiding catheter for RV angiography before stent deployment, to make sure that the stent is in a good position and covering the whole infundibulum and pulmonary valve (but we feel that long sheath and/ or guiding catheter are too much stiff and are difficult to be pushed to the RVOT in small infants. Stiff catheters might also lead to arrhythmia and hemodynamic instability). After inflating the balloon and stent deployment, the coronary balloon will be exchanged with the MPA2 or, the more hydrophilic, Glide catheter, leaving the coronary wire in position (in case we need to put another stent to cover areas of the RVOT /infundibulum uncovered by the previous stent). Using the catheter, RVOT/PA hand injection will be performed. After confirming the proper position of the stent, the catheter and wire will be removed. Prostaglandin infusion will be stopped in the cath lab once we make sure that the whole infundibulum and pulmonary valve are covered by the stent. If not required, sheaths will be removed. The patient will be shifted to the ICU for observation. Patients will be kept on IV heparin infusion 10 to 20 units/kg/hr, fixed dose for 24 hours. Aspirin will be started once the patient is on full PO intake. Follow-up transthoracic echocardiography will be done immediately after the procedure as well as on the second day and before discharge from the hospital. Patients will be seen after 2 to 4 weeks of discharge, and then every 1 to 2 months till the time of complete TOF repair. On follow up we concentrate mainly on the patient's general condition, making sure that the saturation is maintained above 70% with no history suggesting hypercyanotic spells.

2.2. Ethics of case series and Consent for publication

This was approved by Prince Sultan Cardiac Center research Committee. Written consent was obtained from the patients' families for the study and the use of anonymous data for research purposes.

3. Results

During the period from August 2016 and December 2019, four consecutive patients with a diagnosis of severe TOF and significant cyanosis underwent RVOT stenting. (Table 1) Three males and one female. The mean age and weight were 2.8±1.7 days and 2.9±0.1 kg respectively. All were on prostaglandin infusion before the intervention. The mean saturation before RVOT stenting was 63% 63% (±11%) (50 to 75%). RVOT stenting was performed under general anesthesia. The mean procedure time was 85±38 minutes (60-135 minutes). The mean fluoroscopy time was 21±20 minutes (5-51 minutes). The median amount of contrast given was 7 ml (6-25 ml). The mean saturation immediately after RVOT stenting was 82% (±4%) (76% to 86%) which was statistically significant from the pre-intervention saturation ($p = 0.018$). The median hospital stay was 4.5 days (3-6 days). All patients are alive and had complete TOF repair at a median age of 6.5 months (range 6-12 months). One patient required RVOT stent dilatation 4 months after the first intervention (surgery was delayed because of poor weight gain and depressed RV function). The median branch PAs diameter and Z-score before RVOT stenting and immediately before complete TOF repair were 2.8 mm (Z score: -2.5) and 5.5 mm (Z score: -0.5 to -1.16) respectively. Figure 1 shows one of our patients with very severe RVOT obstruction, with RVOT angiogram, prior and post RVOT stenting. No major complications in the catheterization laboratory and ICU, apart from transient arrhythmias and hemodynamic instabilities during catheters and wires manipulation in the RV and RVOT. The median follow up period is 13 months (8-36 months).

Table 1. Descriptive statistics of the patients with data before and after RVOT stenting as well as before complete TOF repair.

Patients Variables	Mean	Median	Standard Deviation	Minimum	Maximum
Age at RVOT stenting (days)	2.75	2.5	1.71	1	5
Weight at diagnosis (Kg)	2.875	2.85	0.1	2.8	3
Fluoroscopy Time minutes	21	14.5	20.54	5	50
Contrast given (ml)	15.25	7.5	16.56	6	40
Procedure time (minutes)	78.75	65	38.38	50	135
Saturation prior to intervention	0.6325	0.64	0.11	0.5	0.75
Saturation immediate after intervention	0.8175	0.825	0.04	0.76	0.86
Brach PAs Before RVOT stent	2.7	2.65	0.24	2.5	3
Brach PAs Before TOF repair	5.5	5.5	0.58	5	6
Age at Complete TOF repair (moths)	7.75	6.5	2.87	6	12

4. Discussion

Blalock-Taussig (BT) shunt is the first reported surgical palliation for TOF (6). Other palliative procedures include surgical RVOT enlargement or a right ventricle to pulmonary artery (RV-PA) conduit implantation leaving the ventricular septal defect open, insertion of an aortopulmonary shunt, stenting of the arterial duct, and balloon pulmonary valvuloplasty (2, 3). Early primary repair of cyanotic neonates including those with TOF carries a significantly increased risk compared with complete repair at an older age. Elective complete TOF repair is generally performed between 6 and 12 months of age (7). Currently, more than 95% of children born with TOF are expected to survive into adulthood (8, 9).

TOF with severe cyanosis reflects poor anatomy and is associated with more clinical instability. Early repair of such cases is associated with longer hospitalization than those who had elective surgery beyond the neonatal period (9). The advantages of primary neonatal TOF repair might include avoidance of shunt-related complications, early relief of hypoxia, promotion of normal lung development, prevention of progressive ventricular hypertrophy and fibrosis, and psychological comfort for the family (8, 10, 11). Surgical reconstruction of certain types of TOF might be challenging, due to several reasons including but not limited to low body weight, intrauterine growth retardation, preterm birth, unfavorable pulmonary arterial anatomy, abnormal coronary distribution, significant co-morbidities, critical preoperative condition, and concomitant malformations. In such patients, it might be wiser to do a staged approach (8, 12). Initial palliation can be achieved by either a surgical shunt or by catheter intervention (ductus

arteriosus or RVOT stenting). (13, 14). Bridging symptomatic neonates and infants with TOF by right ventricular outflow tract stenting (stent) is an option and is a current practice in many cardiac centers (1, 9, 15, 16).

RVOT stenting is suggested as a promising alternative to aortopulmonary shunt in neonates with TOF and significant cyanosis, especially when associated with co-morbid conditions such as low body weight, intrauterine growth retardation, preterm birth, unfavorable pulmonary arterial anatomy, abnormal coronary distribution, significant co-morbidities, critical preoperative condition, and concomitant malformations (17). RVOT stenting offers the advantage of pulsatile blood flow and might have a better stimulus for pulmonary arteries growth, aiming to a better patient's and PAs conditions at the time of elective full surgical repair in the future (18).

The efficacy and safety of RVOT stenting in such cases were demonstrated by multiple investigators and interventional pediatric cardiologists (1, 9, 15, 16). Some investigators report comparable overall outcomes of primary neonatal TOF repair and transcatheter palliation for neonates with severe TOF in terms of survival, growth, and hemodynamic parameters. They suggest that both strategies are a reasonable option for children with severe TOF (19). With the development of catheter-based interventional techniques, alternative methods of establishing and securing a reliable pulmonary blood flow may be achieved at a lower risk. Other investigators report the safety and efficacy of RVOT stenting in cases with severe TOF and complex anatomy (e.g. AVSD-with TOF) (20, 21). In some patients with membranous pulmonary atresia it might be feasible to perforate the pulmonary valve followed by balloon pulmonary valvuloplasty and if required RVOT stenting (22). As the initial palliation of Fallot-type lesions, RVOT stenting promotes better pulmonary arterial growth and oxygen saturations compared with modified BT shunt. This was demonstrated in a single-center nonrandomized study of 67 consecutive patients with TOF, palliated by either modified BT shunt (n=28) or RVOT stent (n=39) and assessed by serial echocardiography. The mixed model analysis showed significantly better branch PAs growth after RVOT stenting (23). In our case series, RVOT stenting provides safe palliation for our patients. All of them had TOF repair at a median age of 6.5 months. Branch PAs size at the time of TOF repair was reasonable with a median branch PAs diameter of 5.5 mm (Z score: -0.5 to -1.16). Cardiac catheterization carries a risk of exposure to radiation as well as the possible complications of contrast injection. One of our patients required re-intervention before complete TOF repair in the form of dilatation of the RVOT stent at the age of 7 months. His weight at that time was 4.5 kg and there was some RV dilatation with RV dysfunction. The surgeon was hesitant to do a complete repair at that time. That patient had a complete repair at the age of 12 months and he is doing well.

5. Conclusions

RVOT stenting is reasonable alternative palliation for patients with severe TOF. The technique of stent deployment in the RVOT is challenging and requires pre-intervention planning and discussion. The type of catheters used to engage the RVOT is important. Using soft catheters and coronary wires might facilitate the crossing of the RVOT and pulmonary valve.

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Conflict of Interest:

There is no conflict of interest to be declared.

Authors' contributions:

Conception or design of the work: All authors; Acquisition of data: All authors; Analysis or interpretation of data: AAA, QA; Drafting the manuscript: AAA, QA; Revising the manuscript: All authors; Accountable for all aspects of the work: All authors. All authors read and approved the final manuscript.

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