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Improved LV Function Following RVOT Stenting in a Late Presenter ToF Patient with Recurrent Cardiac Arrest

Gema Citra Dwijayanti[1*], Alfa Alfin Nursidiq[1], Bagas Ardhimuda Marsudi[1], Radityo Prakoso[1], Oktavia Lilyasari[1]

Departement of Cardiology and Vascular Medicine, National Cardiovascular Centre of Harapan Kita, Universitas Indonesia, Jakarta, Indonesia[1]

Corresponding Author: 1*

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ABSTRACT

Tetralogy of Fallot (ToF) cases with late presentation were more common than previously realized and often undetected despite proper screening. This population initially has a small degree of right ventricular outflow obstruction, allowing them to receive better oxygenation. The incidence of ToF in NCCHK from 2019–2021 was 883 cases. The palliative interventions which aim to increase pulmonary blood flow are still considered the first-choice treatment in patients with critical ToF unsuitable for primary repair. RVOT stent (Right Ventricular Outflow Tract Stent) is frequently used to increase pulmonary artery flow in patients with uncorrected ToF in preparation for a ToF repair. It has been advocated as an intermediary step for high-risk patients. To report a case of late-presentation ToF with LV dysfunction which performed non-surgical palliative approach as bridging procedure due to high risk of surgery. A 7-year-old boy presented to the emergency room (ER) with shortness of breath and cyanosis. The patient has been diagnosed as ToF since 2019 but has lost follow-up due to the COVID-19 pandemic. Echocardiography on admission revealed a decreased left ventricular (LV) ejection fraction (LVEF of 39.5%) with global hypokinetic and thrombus at LV. Two days after admission, a joined cardiology–cardiothoracic surgery meeting was decided to do MSCT Cardiac and right ventricle outflow tract (RVOT) stenting for the patient. The day before being scheduled for MSCT and RVOT stenting, the patient suddenly developed a hypercyanotic spell and fell into cardiac arrest. The Code Blue protocol was activated and the patient was promptly given CPR, intubated, and transported to the ICU. He was having episodes of Arrest-ROSC. The patient was promptly delivered to the Cath lab for emergency right ventricular outflow tract (RVOT) stenting. At the end of the procedure, oxygen saturation reached 92% with stable hemodynamics and the RV-PA gradient was substantially decreased from 101 mmHg to 28 mmHg (evaluated by echocardiography). After RVOT stenting, the patient's clinical condition continued to gradually improve. Echocardiography evaluation showed marked improvement in systolic function (EF 70%). We reported a case with late-presentation ToF patients with LV

dysfunction who were treated with a non-surgical palliative approach as a bridging procedure in high-risk for surgery. The patient's condition gradually improved after the RVOT stent procedure.

1. Introduction

Late presentation of tetralogy of Fallot (TOF) occurs more often than is anticipated and can sometimes be missed despite proper screening. This population initially has less right ventricular outlet obstruction, which allows them to oxygenate more efficiently.1 Incidence of TOF in NCCHK from 2019 – 2021 are 883 cases. When missed by screening, patients with Tetralogy of Fallot typically develop increasing hypoxia as their ductus arteriosus closes and they have gradually decrease of Left Ventricle Ejection Fraction (LVEF), this condition increases the risk of morbidity and mortality for surgical procedure [1], [2]. The palliative intervention which aims to increased pulmonary blood flow is still considered the first-choice treatment in patients with critical tetralogy of Fallot (TOF) unsuitable for primary repair [3]. RVOT stent (Right Ventricular Outflow Tract Stent) is frequently used to increase pulmonary artery flow in patients with uncorrected TOF in preparation for a TOF repair. It has been advocated as an intermediary step for high-risk patients [4], [5]. We report a rare case of Tetralogy of Fallot with severe cyanosis and a low LV ejection fraction in a patient who underwent successful stenting of the right ventricular outflow tract, which unexpectedly increased LVEF after the procedure.

2. Case Illustration

A 7-year-old boy was admitted to our hospital with the chief complaint of dyspnoea. He presented to the ER with shortness of breath with activity or when lying down, fatigue and weakness, with signs of facial oedema and bluish skin. The boy has had a history of frequent episodes of fatigue and dyspnoea since infancy. He is also often seen in the squatting position whenever he feels out of breath. History of worsening cyanosis, malaise, altered consciousness, and seizures was denied by his parents. His mother mentioned that the patient had a history of feeding difficulty and inadequate weight gain. The patient has been diagnosed as ToF since 2019 but has lost follow-up due to the COVID-19 pandemic. He also has poor compliance to medication therapy.

On physical examination, the patient was lethargic and looked very ill. He weighed 15 kilograms and was 100 cm tall. His blood pressure revealed 105/64 (75) mmHg, heart rate 90 bpm, respiratory rate 30 times/minute and peripheral oxygen saturation of 70% in room air. The jugular vein pressure (JVP) was elevated (5 + 4 cm H2O). Heart auscultation was heard as a single, normal, first heart sound, and a single, non-accentuated second heart sound was found with ejection systolic murmur grade 3/6 with maximum intensity at the upper left sternal border. Lung auscultation was relatively unremarkable, with clear, vesicular lung sounds. Hepatic enlargement was palpated 3 cm below the arcus costae. Both extremities showed clubbed fingers with clear signs of cyanosis.

An electrocardiogram (Figure 1) showed sinus rhythm, right axis deviation (RAD) with right ventricular hypertrophy (RVH). Cardiomegaly (CTR 68%), right atrial enlargement, right ventricular hypertrophy (upward apex), and oligemic pulmonary vasculature were all discovered on a chest x-ray (Figure 2). On admission, echocardiography revealed (Figure 3) a decreased left ventricular (LV) ejection fraction (LVEF 39.5%), global hypokinetic, LV thrombus, and severe sub valvar pulmonary stenosis (PG 96 mmHg).

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Several hours after admission, a joined cardiology–cardiothoracic surgery meeting was decided to do MSCT Cardiac and elective right ventricle outflow tract (RVOT) stenting for the patient. Before scheduled for MSCT and RVOT stenting when suddenly he developed a hypercyanotic spell and fell into cardiac arrest. The Code blue protocol was activated and the patient was promptly given cardio pulmonary resuscitation (CPR), intubated, and transported to the pediatric intensive care unit (PICU) where he was put on inotropic support and mechanical ventilation. With 100% FiO2, peripheral saturation only reaches a maximum of SpO2 40-50%. Even though the patient was breathing with 100% FiO2, his oxygen saturation was still low, so he was quickly taken to the cath lab for a right ventricular outflow tract (RVOT) stent.

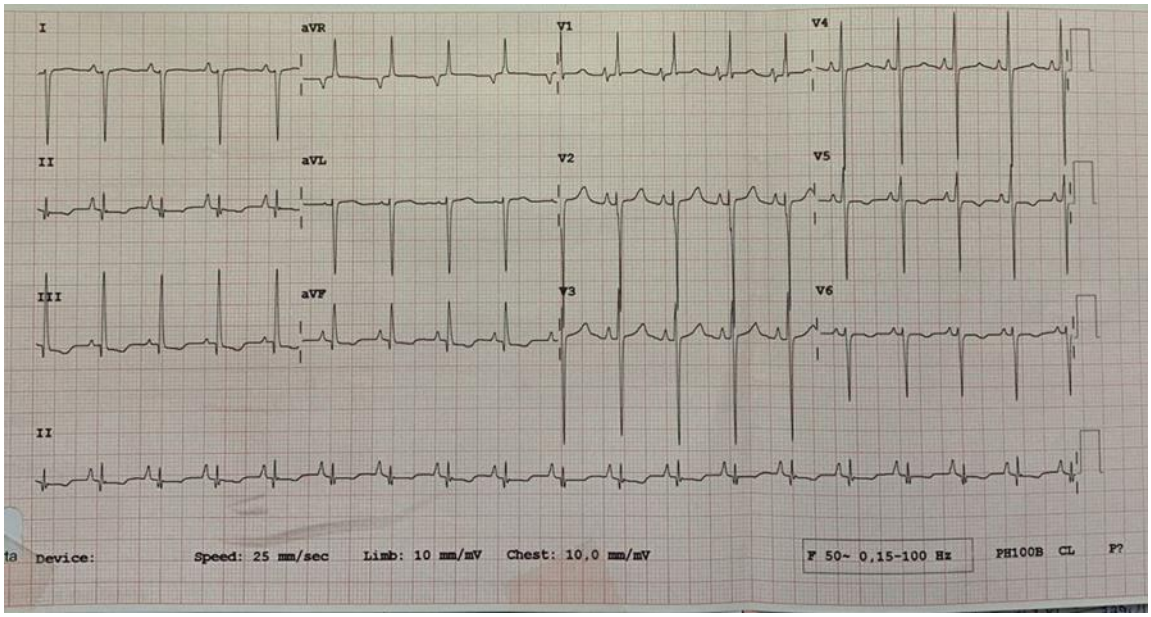

Figure 1. Electrocardiography examination showed sinus rhythm, QRS rate 75 x/m, QRS axis +135, P pulmonale, PR int 160 ms, QRS duration 80 ms, Abrupt R V1-V2, Right Axis Deviation (RAD), Right Ventricular Hypertrophy (RVH).

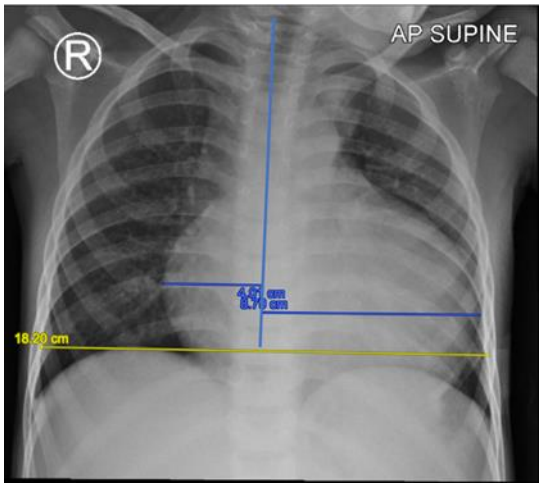

Figure 2. Chest X-ray showed heart enlargement with cardiothoracic ratio 68%, reduced pulmonary segment, flatten cardiac waist, upward apex (RVH)

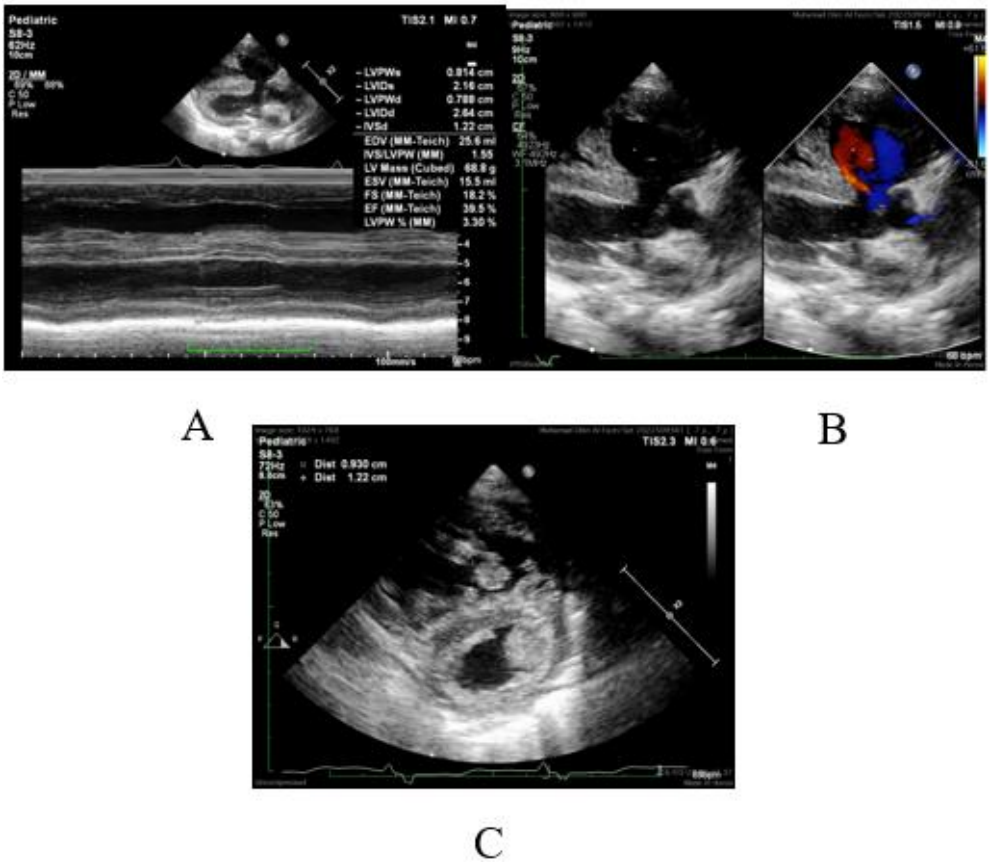

Figure 3. (A). Echocardiography 2D before RVOT stenting showed LV EF 39.5% and (B) Showed >50% Overriding aorta accompanied with anterocephalad deviated septum which caused infundibular and valvar pulmonary stenosis (PS), Large subaortic VSD, R to L shunt with (C) LV thrombus diameter 9x12 mm

In the Cath lab, the patient was prepared using standard protocol. Throughout the procedure, the patient experienced several recurring episodes of asystole. Sustained ROSC was successfully achieved six minutes after stent deployment. At the end of the procedure, oxygen saturation reached 92% with stable hemodynamics and RV-PA gradient was substantially decreased from 101 mmHg to 28 mmHg (evaluated by echocardiography). After RVOT stenting, the patient was given heparin starting at 1 IU/Kg body weight/hour for 3 days with an APTT target of 50-70 second.

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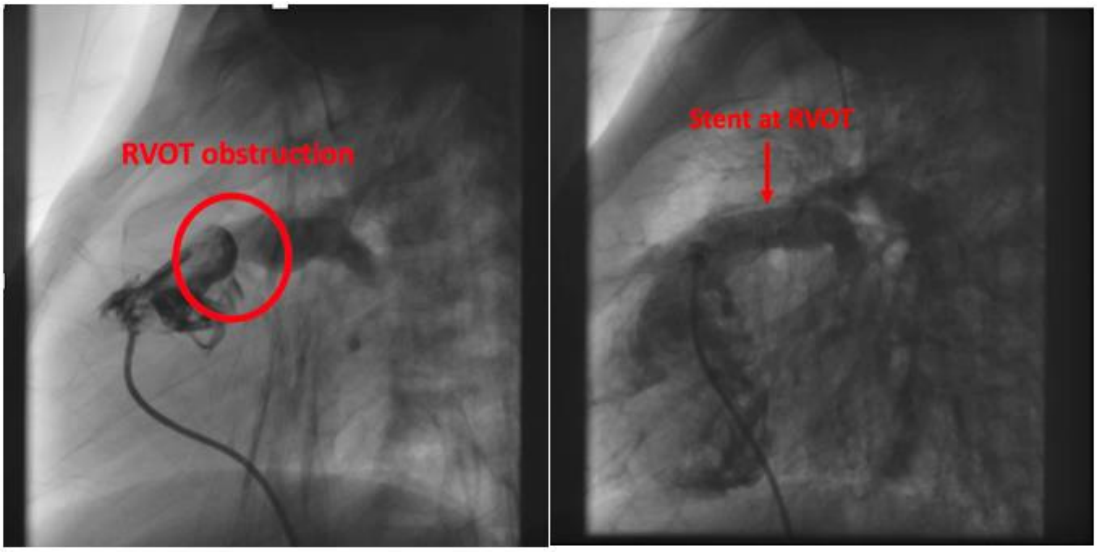

Figure 4. (A). Angiography showed obstruction at right ventricular outflow tract (RVOT) and (B) Flow improvement after RVOT stenting.

His clinical condition continued to gradually improve. Echocardiography evaluation (Figure 4) showed marked improvement in systolic function (EF 70%).

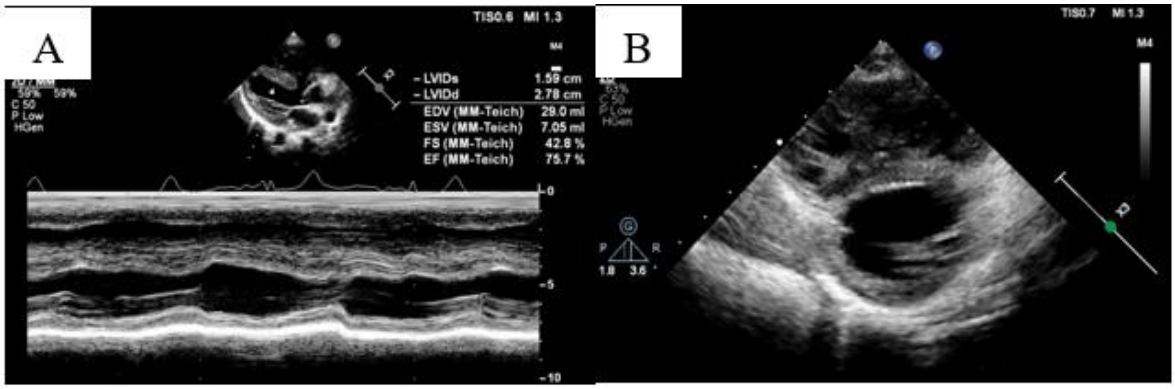

Figure 5. (A). Echocardiography 2D after 8-days of RVOT stenting showed improved LV EF 75.7% and (B) PSAX view no thrombus LV is seen

3. Discussion

ToF is the most common type of cyanotic congenital heart disease (CHD) and accounts for approximately 7% to 10% of congenital morbidities [6]. The presence and severity of cyanosis in this condition are determined by the degree of RVOT obstruction and the development of PAs. Because of its diverse anatomical characteristics, TOF can have a wide range of clinical and hemodynamic consequences. Some patients, such as ToF, do not initially have a significant degree of right ventricular outlet obstruction, allowing them to oxygenate more effectively. Patients with Tetralogy of Fallot who are not identified through screening typically develop increasing hypoxia as their ductus arteriosus closes and have a gradual decrease in Left Ventricle Ejection Fraction [6], [7]. Chronic hypoxia could be one of several explanations for the decreased LV ejection fraction observed in this case. Staged surgical repair of the TOF has been a standard approach for many years, particularly in high-risk infants with low ages and weights, along with complicated anatomy [6], [8]. Consequently, children with TOF who have complications such as critical

RVOT obstruction, Left Ventricular dysfunction, PA hypoplasia, and the presence of a major aortopulmonary collateral artery (MAPCA) are candidates for primary palliation as a first step to prevent hypoxia, improve oxygen saturation, and promote PAs development until the complete repair is performed. in the late forms of TOF, the presence of a severe ventricular dysfunction is a rare finding, and makes the surgical strategy complex and potentially dangerous. In this situation, timing and indications are of particular importance [9]. More recently, catheter interventions have played an alternative role by stenting the arterial duct (PDA) or the right ventricular outflow tract [10]. Here, we report a ToF patients who suffered LV systolic dysfunction and recurrent cardiac arrest which performed RVOT stent as a non-surgical palliative approach as bridging procedure in high risk for surgery.

Left Ventricle Dysfunction in Late Presenter ToF

In any cardiac surgery unit, ventricular dysfunction is a major risk factor for mortality and morbidity [12], [13]. Every child presenting with LV dysfunction must undergo a comprehensive evaluation for causes of ventricular dysfunction, including the search for structural causes such as associated coronary involvement (including aberrant left coronary artery from pulmonary artery), left ventricular outflow tract obstruction, and coarctation of the aorta. In the context of cyanotic CHD, which was prevalent in our patient, these are extremely uncommon [13]. Myocarditis/cardiomyopathy is the most common cause in the general population, but it is difficult to rule out in the absence of ventricular biopsies. Long-term ventricular and atrial arrhythmias can also result in ventricular dysfunction (Tachycardia-induced cardiomyopathy) [13], [14]. This rhythm abnormality does not occur in our patient. While our case, who had untreated hypoxia for 7 years, shows a significant improvement in ventricular function after hypoxia correction by RVOT stenting.

Treatment Options For Tetralogy Of Fallot in Late Presenter

There is no medical treatment for ToF. The structural abnormalities in the heart require surgical correction [15]. Study regarding non-surgical transcatheter palliation in late presentation Congenital Heart Disease (CHD), mainly late Tetralogy of Fallot (ToF) is very rare. Besides becoming a bridging procedure before complete repair, non-surgical palliation catheter was intended to be a salvage procedure for late-presentation that poses high-risk condition for surgery [16]. Palliative shunts are not routinely performed because first remedial treatments are increasingly conducted during infancy.

Palliative options include surgical modified BT shunt, RVOT stenting, ductal stenting, and BPV in high-risk late-presenting ToF patients who are symptomatic with low saturations or frequent cyanotic spell and where institutional policy prevents early intracardiac repair. Each option's advantages and disadvantages would be considered [16], [17]. RVOT stenting is an option for patients with life-threatening symptoms. It helps stabilize cyanosis, improves circulation in the pulmonary artery (PA), improves the function of the left ventricle (LV), and makes elective surgery possible in the future [18].

RVOT Stenting in Late Presenter ToF

In the initial palliation of severely cyanosed and LV dysfunction infants with ToF-type lesions, right ventricular outflow tract (RVOT) stenting has become an alternative to the surgical creation of a modified Blalock–Taussig shunt (mBTS) [18], [19].

In most cases, indications for either mBTS or RVOT stenting were agreed upon in advance during a joint cardiology–cardiothoracic surgery meeting. Decisions were influenced by known risk factors for the performance of mBTS (e.g., low weight, small pulmonary arteries, and significant comorbidities). Initially, the decision to perform RVOT stenting was primarily based on severe associated comorbidities that were

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believed to be adversely affected by mBTS haemodynamics (e.g., prepalliative gut and brain pathologies, prematurity) or where the surgical intervention with potential need for cardiopulmonary bypass was deemed "high risk." [20].

Another reason is that RVOT stenting was recommended for infants with a low weight. Patients with duct-dependent pulmonary blood flow, complex anatomy, and hypoplastic pulmonary arteries (z-score <-1.5) were preferred to be treated with RVOT stenting as experience grew. For surgical correction of CHD, ventricular dysfunction is regarded as a significant risk factor, particularly during cardiopulmonary bypass. It is a crucial factor for excluding this subset from surgical correction [18], [21], [22]. In cyanotic heart disease, hypoxia is believed to be the cause of ventricular dysfunction. Stenting of the right ventricular outflow tract (RVOT) has become a standard initial palliative procedure for Tetralogy of Fallot (TOF) [18], [23], [24]. Until now, there is very limited data that explains the implementation of the RVOT stenting procedure in ToF patients with late presenters.

4. Conclusion

RVOT Stenting is an alternative palliative treatment for ToF patient with sub optimal features of surgical procedures, including LV systolic dysfunction. The purpose of RVOT stent is to diminish cyanosis and chronic hypoxia, improved Left ventricle systolic function

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