Case Report

# Staged Treatment Using Right Ventricular Outflow Tract Stenting for Effective Augmentation of Native Pulmonary Arteries in a Case of Tetralogy of Fallot with Major Aortopulmonary Collateral Arteries

Yuji Doi, MD[1),*], Sung-Hae Kim, MD[1)], Mizuhiko Ishigaki, MD[1)], Keisuke Sato, MD[1)], Jun Yoshimoto, MD[1)], Norie Mitsushita, MD[1)], Masaki Nii, PhD[1)], Kisaburo Sakamoto, MD[2)], and Yasuhiko Tanaka, MD[1)]

[1)] Department of Cardiology, Shizuoka Children's Hospital, Shizuoka, Japan
[2)] Department of Cardiovascular Surgery, Shizuoka Children's Hospital, Shizuoka, Japan
[*]Current affiliation: Kurashiki Central Hospital, 1–1–1 Miwa Kurashiki-city, Okayama710–8602, Japan

Stenting of the right ventricular outflow tract (RVOT) is an emerging alternative for the management of cyanotic tetralogy of Fallot (TOF) in the neonatal or early infancy stage. We describe our treatment of a TOF in the early infancy stage, using RVOT, in a patient for whom the TOF physiology was associated with major aortopulmonary collateral arteries (MAPCAs), all of which had a dual blood supply from the pulmonary arteries. The procedure was successful, with sufficient antegrade blood flow of the native pulmonary arteries achieved for occlusion of all MAPCAs. RVOT stenting provided an effective strategy for the management of cyanotic TOF, without the need for unifocalization, in the presence of MAPCAs with dual blood supply.

Keywords: RVOT stenting, TOF, MAPCAs, unifocalization

## Introduction

Complete repair of a tetralogy of Fallot (TOF) in the neonatal or early infancy period is controversial as it carries a relatively high risk of morbidity and death compared to repair performed later in infancy.[1)] Use of a Blalock-Taussig Shunt (BTS) is the common initial palliation performed for the treatment of cyanotic TOF; however, BTS also carries a relatively high risk of death.[2)] Right ventricular outflow tract (RVOT) stenting has emerged as a safe alternative approach for the treatment of cyanotic TOF, which promotes growth of the pulmonary artery (PA) and has been associated with good clinical outcomes.[3, 4)] In this case report, we describe our successful use of RVOT stenting in the early infancy period in a patient with TOF and major aortopulmonary collateral arteries (MAPCAs), which promoted PA blood flow and eliminated the need for unifocalization (UF) of the MAPCAs at the time of complete repair.

## Case Report

The patient first presented prenatally, at 32 weeks of gestation, to our center for evaluation of a TOF with MAPCAs and a right aortic arch. The baby girl was born at 39 weeks of gestation, by vaginal delivery, with a birthweight of 2,363 g. Her $O_2$ saturation, on room air, was 95% at birth.

The echocardiography performed at birth confirmed the TOF, including a severely hypoplastic pulmonary valve (annular size, 3.7 mm; Z score, −5.8) and MAPCAs. The branch of the PA was also hypoplastic, with a Nakata index of 76 $mm^2/m^2$ and a negative jet

Received: October 31, 2019; Accepted: February 13, 2020
Corresponding author: Yuji Doi, MD, Shizuoka Children's Hospital, 860 Urushiyama, Aoi-ku, Shizuoka-city, Shizuoka420-8660, Japan
E-mail: fools.gold.712@gmail.com
ORCiD: Yuji Doi (https://orcid.org/0000-0002-0686-0197)
doi: 10.24509/jpccs.19-006

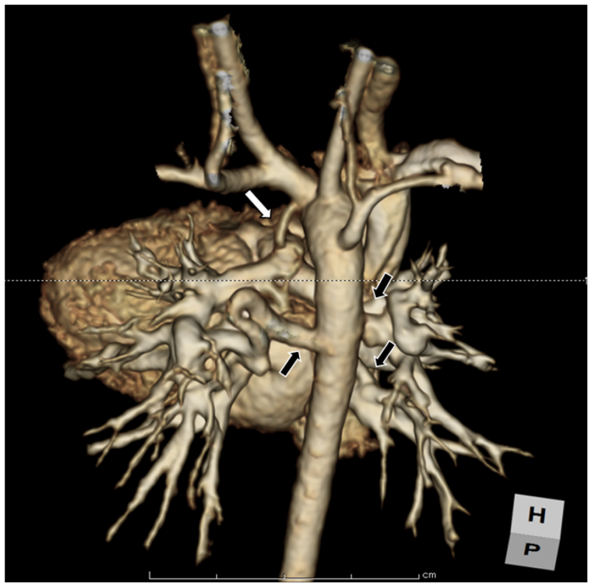


Fig. 1 Contrast enhanced computed tomography image, showing 3 MAPCAs (black arrows) and a small vessel which appear to be a remnant of the patent ductus arteriosus (white arrow).
MAPCA, major aortopulmonary collateral arteries

from the MAPCAs to the native PA observed in the distal portion. The outlet septum was relatively small and did not show prominent RVOT obstruction. Contrast enhanced computed tomography (CT) imaging was performed on post-natal day 29 to assess the MAPCAs (Fig. 1). Three MAPCAs were identified, with an additional small vessel which we considered to be a remnant of the patent ductus arteriosus (PDA).

As the baby gained weight, her $O_2$ saturation decreased below 80%, due to inadequate growth of the pulmonary valve and progression of sub-pulmonary stenosis. We performed a catheterization on post-natal day 61 (weight, 3,900 g) to evaluate the MAPCAs directly and for intervention to increase pulmonary blood flow. The pulmonary annular size measured 3.5 mm (Z score, −7.4) with echocardiography then. We identified two large MAPCAs (MAPCA 1 and 2, Fig. 2) on angiogram, one each for the right and left lungs, with both having a dual blood supply to the native PA, which was hypoplastic (Fig. 2). The small vessel thought to be a remnant of the PDA on CT imaging proved to be a small MAPCA (MAPCA 3, Fig. 2), which also perfused the left lung. The Nakata index for the native PA was 154 $mm^2/m^2$, and a negative jet was again observed. We proceeded with catheterization to promote antegrade PA blood flow and, hopefully, to enable the occlusion of all the MAPCAs. With a pulmonary annular size of 4.0 mm measured on angiography, we proceeded with balloon pulmonary valvuloplasty (BVP), using a Sterling™ 5 × 20 mm balloon catheter (Boston Scientific Corporation, USA). After BVP, her $O_2$ saturation increased to 85% on room air. We, therefore, proceeded with occlusion of the MAPCAs 2 and 3, using an AMPLAZER™ vascular plug-II (AGA Medical Corporation, USA) and an ORBIT GALAXY® device (Codman & Shurtleff, Inc. USA), respectively. We then proceeded with occlusion of MAPCA 1, using a Berman angiography catheter, which caused a drop in her $O_2$ saturation to 72% on room air. An increase in the antegrade PA blood flow was needed to allow occlusion of all the MAPCAs and to promote the growth of the native PA. Thus, we decided to perform RVOT stenting, using an Express™ Vascular SD, 6 × 14 mm, stent (Boston Scientific Corporation, USA). The stent was successfully placed across the pulmonary valve, as the outlet septum was relatively short. $O_2$ saturation increased immediately to 93%, even after occlusion of MAPCA 1, using a Flipper® PDA Closure Detachable Coil (Cook Medical Incorporated, USA). Since stents for RVOT is an off-label use in Japan, we discussed the possible risks of this procedure carefully before the session with the parents, and acquired written consent form. The official approval of institutional review board could not be in time for the procedure, yet it was approved immediately.

Postoperatively, the patient was treated with diuretics to prevent reperfusion injury of the lungs. The patient was able to feed well and gained weight gradually. At 4 months of age, she reached a weight of 5.4 kg, with an $O_2$ saturation of around 90% on room air. Catheter evaluation indicated an increase in the Nakata index to 352 $mm^2/m^2$, with a mean PA pressure of 13 to 17 mmHg. A remaining MAPCA was identified, with some residual flow observed in the previously occluded MAPCAs. We proceeded with occlusion of the new MAPCA, using an AZUR® CX18 and ED COIL (KANEKA MEDICAL PRODUCTS, Japan). At 6 months of age, the patient underwent surgical repair using a transannular patch and the RVOT stent was removed completely, without any difficulty. Her post-operative clinical course was good, and she is being followed on an outpatient basis, remaining in a fairly stable state.

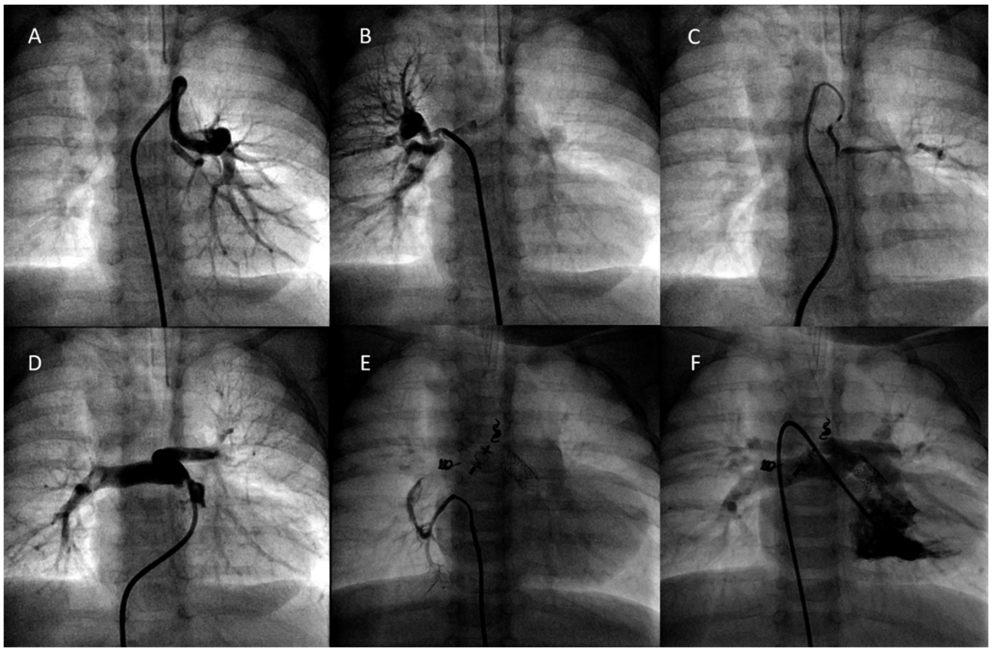


Fig. 2 (A) Selective angiogram of the MAPCA1, which provided perfusion to most segments of the left lung. (B) Angiogram of the MAPCA2, which perfused a large segment of the right pulmonary artery. (C) Angiogram of the MAPCA3, which we first thought to be a remnant of the ductus arteriosus, showing that it perfused a small portion of the left lung. (D) Angiogram of the hypoplastic pulmonary artery, with a Nakata index of 154 $mm^2/m^2$. (E) Angiogram of the MAPCA4, which was occluded at the second catheter. (F) Angiogram of the pulmonary artery after RVOT stenting, with an increase in the Nakata index to 352 $mm^2/m^2$.
MAPCA, major aortopulmonary collateral arteries; RVOT, right ventricular outflow tract

## Discussion

Considering the high risk of complete TOF repair and palliation using the modified BTS in the neonatal and early infancy period,[1–5] RVOT stenting has emerged as a viable alternative palliative option for cyanotic TOF, particularly for infants with severe comorbidities, which might be negatively impacted by a modified BTS procedure and low birth.[3] With accumulating experience, RVOT stenting has also become the preferred option for the treatment of patients with a hypoplastic PA, defined by a Z-score $< -2$.[6] RVOT stenting has also been used for the treatment of pulmonary atresia associated with perforation of the pulmonary valve.[7] RVOT stenting is now widely used as a palliative option for patients with a TOF type physiology.

The management strategy for TOF with MAPCAs is more complex compared to TOF without MAPCAs. Some centers perform neonatal PA rehabilitation, using shunting to promote the PA growth without translocation of the MAPCAs,[8] while other centers opt for single stage UF, which may lead to better long-term outcomes.[9] Regardless of the strategy used, the presence of a native confluent PA is a very important factor to consider. In our case of TOF with MAPCAs, all MAPCAs had a dual blood supply to the confluent native PAs. In such a case, UF may not be necessary, but augmentation of the native PA is often required, prior to intra-cardiac repair, with creation of an aortopulmonary window (APW) being an option for this strategy.[10] In our case, as the angiogram confirmed that all MAPCAs are connected to the confluent native PAs with a dual supply, we performed a palliative intervention to promote antegrade blood flow in the native PA. Although having dual supply MAPCAs is not necessarily the same as an adequate connection that does not require unifocalization, we assumed that our patient had enough pulmonary vasculature for intra-cardiac repair without unifocalization from the angiogram and the CT performed prior to catheter, if we could successfully promote antegrade pulmonary blood flow. Initial BVP was effective, with the $O_2$ saturation increasing from 78% to 85%, but was ineffective

in increasing the antegrade blood flow sufficiently in the native PA to occlude all MAPCAs. Since there was a prominent negative jet from the MAPCAs to the native PA, we thought it necessary to occlude all the MAPCAs to promote antegrade pulmonary blood flow to promote native PA growth. Occluding the MAPCAs later in another session was an alternate option, thought we thought RVOT stenting may facilitate antegrade blood sufficiently to occlude the MAPCAs in one session, reducing the number of catheter and total radiation exposure as well. This is why we performed RVOT stenting, which was successful in increasing the antegrade PA blood flow sufficiently to occlude all MAPCAs, as well as to increase $O_2$ saturation. RVOT stenting is less invasive and provides a higher diastolic pressure compared to BTS or APW, which is preferable with respect to coronary perfusion, and may provide a more stable inter-stage status. As for the selection of the stent profile, we chose the size according to the report from Toronto group,[4] using a stent which is 1 to 2 mm larger compared to the RVOT diameter in diastole. For we have already performed BVP with a 5 mm balloon, we chose a 6 mm stent for stability. As the surgeons had already concluded that there was no way to preserve the native pulmonary valve for it was too small, we decided to place the stent across the pulmonary valve to reduce the risk of stent migration. Since RVOT stenting in a patient with a small outlet septum may carry a risk of impinging the aortic valve and of aortic regurgitation,[11] it is preferable to place the stent at the level of pulmonary valve to avoid lower positioning of the stent as in the present case. Thus, we successfully placed the stent without any complications. The Nakata index increased dramatically after stenting, from 154 $mm^2/m^2$ to 352 $mm^2/m^2$ at 2 months after the intervention, allowing the patient to undergo intra-cardiac repair at 6 months of age. RVOT stenting causes free pulmonary regurgitation hemodynamically, yet it created sufficient antegrade pulmonary blood flow to promote pulmonary artery vasculature growth as described in previous report.[6] Although a previous study indicated that RVOT stents cannot be removed completely in most cases,[12] we were able to remove the stent successfully. Reports with higher removal rate seems to have a relatively shorter palliation period.[13] The successful removal of stent may have been due to relatively short duration of palliation (4 months). Moreover, our surgical management with RVOT stenting was successful in promoting sufficient PA growth for intra-cardiac repair using only one open-heart surgery. Since patients with TOF and MAPCAs will require repeat RVOT repair, it is important to consider a strategy to reduce the number of initial procedures required. And in this context, evading unifocalization is preferable for Mainwaring RD et al. reported that 18% of patients who underwent unifocalization required revision of the distal unifocalized bed and that most of these patients had a single-stage repair.[14]

## Conclusion

RVOT stenting is an effective option in the treatment strategy for TOF with MAPCAs if all MAPCAs have a dual blood supply. RVOT augments pulmonary blood flow and growth of the native PAs, enabling intra-cardiac repair, without any palliative surgery nor UF.

### Conflicts of Interest

The authors have no conflicts of interest to declare.

## References


1) Al Habib HF, Jacobs JP, Mavroudis C, et al: Contemporary patterns of management of tetralogy of Fallot: Data from the STS database. Ann Thorac Surg 2010; **90**: 813–820
2) Petrucci O, O'Brien SM, Jacobs ML, et al: Risk factors for mortality and morbidity after the neonatal Blalock-Taussig shunt procedure. Ann Thorac Surg 2011; **92**: 642–652, discussion, 651–652
3) Stumper O, Ramchandani B, Noonan P, et al: Stenting of the right ventricular outflow tract. Heart 2013; **99**: 1603–1608
4) Dohlen G, Chaturvedi RR, Benson LN, et al: Stenting of the right ventricular outflow tract in the symptomatic infant with tetralogy of Fallot. Heart 2009; **95**: 142–147
5) Castleberry CD, Gudausky TM, Berger S, et al: Stenting of the right ventricular outflow tract in the high-risk infant with cyanotic teratology of Fallot. Pediatr Cardiol 2014; **35**: 423–430
6) Quandt D, Ramchandani B, Stickley J, et al: Stenting of the right ventricular outflow tract promotes better pulmonary arterial growth compared with modified Blalock-Taussig shunt palliation in tetralogy of Fallot-type lesions. JACC Cardiovasc Interv 2017; **10**: 1774–1784
7) Aurigemma D, Moore JW, Vaughn G, et al: Perforation and right ventricular outflow tract stenting: Alternative palliation for infants with pulmonary atresia ventricular septal defect. Congenit Heart Dis 2018; **13**: 226–231
8) Liava'a M, Brizard CP, Konstantinov IE, et al: Pulmonary atresia, ventricular septal defect, and major aortopulmonary collaterals: Neonatal pulmonary artery rehabilitation without unifocalization. Ann Thorac Surg 2012; **93**: 185–191

9) Bauser-Heaton H, Borquez A, Han B, et al: Programmatic approach to management of tetralogy of Fallot with major aortopulmonary collateral arteries: A 15-year experience with 458 patients. Circ Cardiovasc Interv 2017; **10**: 3004952
10) Mainwaring RD, Reddy VM, Perry SB, et al: Late outcomes in patients undergoing aortopulmonary window for pulmonary atresia/stenosis and major aortopulmonary collaterals. Ann Thorac Surg 2012; **94**: 842–848
11) Lee J, Sivakumar S, Alwi M: Stenting of right ventricular outflow tract in Tetralogy of Fallot with subarterial ventricular septal defect: A word of caution. Ann Pediatr Cardiol 2017; **10**: 281–283
12) Sandoval JP, Chaturvedi RR, Benson L, et al: Right ventricular outflow tract stenting in tetralogy of Fallot infants with risk factors for early primary repair. Circ Cardiovasc Interv 2016; **9**: e003979
13) Nokhrin AV, Tarasov RS, Mukhamadiyarov RA, et al: Two stage approach for surgical treatment of tetralogy of Fallot in underweight children: Clinical and morphological outcomes. J Card Surg 2019; **34**: 293–299
14) Mainwaring RD, Patrick WL, Rosenblatt TR, et al: Surgical results of unifocalization revision. J Thorac Cardiovasc Surg 2019; **158**: 534–545