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CASE REPORT

ADVANCED

CLINICAL CASE

Alternative Hybrid Approach to Promote Native Pulmonary Artery Growth in Pulmonary Atresia VSD MAPCAs

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ABSTRACT

We present a case of right ventricle to pulmonary artery hybrid perforation and stenting in a patient with pulmonary atresia with ventricular septal defect major aortopulmonary collaterals and diminutive native pulmonary arteries, then discuss how it compares with established approaches. (**Level of Difficulty: Advanced**.) (J Am Coll Cardiol Case Rep 2022;4:1366–1369) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Pulmonary atresia with ventricular septal defect (PA VSD) can be split into 2 extremes: well-developed branch pulmonary arteries (PAs) that are supplied by a patent ductus arteriosus and pulmonary atresia with main pulmonary blood supply through major aortopulmonary collaterals (MAPCAs).

The first group has traditionally been palliated in the neonatal period with a surgical pulmonary shunt (SPS), which has been replaced in many centers with ductal stenting as a preferred alternative due to greater postprocedural stability and improved patient survival to destination surgical treatment.[1]

Treatment options for PA VSD MAPCAs depend on the development of native PAs. The surgical management has focused on unifocalization of MAPCAs and rehabilitation of native PAs by SPS and right ventricle to pulmonary artery (RV-PA) connection with either a right ventricular outflow tract (RVOT) patch or RV-PA conduit.[2]

With known morbidity and mortality burden of cardiopulmonary bypass in the neonatal period and early infancy,[3] as well as difficulties in pulmonary blood flow regulation using central shunts,[4] it is worth considering whether we should be pursuing transcatheter options for early palliation of PA VSD (MAPCAs), and at what level of acceptable risk. We present a hybrid approach to establish continuity of the right ventricle to the pulmonary artery blood flow

LEARNING OBJECTIVES

- To understand challenges in treatment of PA VSD MAPCAs.
- To explore nonconventional transcatheter options to palliate infants with PA VSD, to promote native PAs growth, and to understand the challenges of a hybrid approach.

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in a patient with PA VSD MAPCAs and diminutive native PAs.

HISTORY OF PRESENTATION

The patient is a term 2.78-kg male infant with antenatal diagnosis of PA VSD MAPCAs and severely hypoplastic PAs, with low percutaneous saturations (60%-70%).

MEDICAL HISTORY

He was initially started on alprostadil, but this was discontinued in view of no patent ductus arteriosus present on computed tomography.

DIFFERENTIAL DIAGNOSIS

High pulmonary vascular resistance or stenosis of MAPCAs was considered as the cause of hypoxemia.

INVESTIGATIONS

Cardiac computed tomography at the age of 2 days showed that the pulmonary blood supply was provided by 3 MAPCAs with a single origin from descending aorta (Video 1); main PAs were diminutive. At cardiac catheterization, pulmonary vein wedge injections demonstrated native 1-mm right pulmonary artery (RPA) and 1.5-mm left pulmonary artery (LPA) (Nakata index 13.4 mm^2/m^2) with miniscule main pulmonary artery (MPA) ending almost 10 mm from RVOT (**Figures 1A and 1B,** Video 2). There was a mild stenosis at the common origin of all 3 MAPCAs from the anterior thoracic aorta. The contrast transit time through the lungs was very prolonged, consistent with elevated pulmonary vascular resistance as the cause of low pulmonary blood flow and saturations.

MANAGEMENT

Due to severe native PA hypoplasia, SPS or RVOT patch were not considered a viable option. A hybrid periventricular RV-PA stent was performed at the age of 6 weeks to establish antegrade pulmonary blood flow. After a midline sternotomy, a micropuncture needle (Cook Medical) was advanced through the RV and into the MPA, and an 0.014 coronary guidewire was advanced. A bolus of heparin 100 U/kg was given. A 4-mm coronary balloon was used to dilate the RVOT-MPA tract because there seemed to be an established RV-MPA connection on inspection. Significant bleeding occurred (confirming true pulmonary atresia) but was controllable, while a 4- x 16-mm coronary covered stent (BeGraft Coronary Stent, Bentley) was positioned directly under fluoroscopy guidance and deployed (**Figure 1C**). The branch PAs filled well antegradely and the bleeding resolved (Video 3). The chest was closed and the patient was moved to pediatric intensive care unit on a therapeutic heparin infusion 20 U/kg/h.

The patient's condition deteriorated after a period of instability (hypotension and acidosis) with profound hypoxemia 6 h after the procedure. He returned urgently to the cardiac catheterization laboratory. The angiogram showed complete thrombosis of the stent. The stent was crossed with a microcatheter from a jugular approach and angioplasty performed with a 4.5- × 15-mm coronary balloon. Two boluses of heparin 100 U/kg were given during the procedure, with final activated clotting time 240 seconds.

Acute management post-stent recanalization focused on maintaining adequate systolic blood pressure, heparin infusion (target activated clotting time 200-240 seconds for the first night), and dual antiplatelet agents administration.

He underwent 2 further percutaneous reinterventions: restenting with coronary uncovered stent (4.5- × 15-mm Resolute Onyx DES, Medtronic) due to proximal stent stenosis at the age of 3 months and stent redilation at the age of 6 months with 6-mm high-pressure balloon (Emerge PTCA Dilatation Catheter, Boston Scientific). Branch PAs at last catheterization (**Figure 1D,** Video 4) showed encouraging development with distal RPA of 3.1 mm and distal LPA of 4.2 mm (Nakata index 61 mm^2/m^2). Both proximal RPA and LPA remain hypoplastic. The common MAPCA from the descending aorta has a proximal stenosis that offers pulmonary blood flow protection (mean PA pressure: 13 mm Hg).

DISCUSSION

Watterson et al[5] described the Melbourne experience using central end-to-side shunt (Mee shunt) in 28 patients with PA VSD MAPCAs and very small pulmonary arteries. Although they managed to achieve a satisfactory pulmonary artery growth in 16 of 24 survivors, the rate of complications was high: central shunt-related complications (congestive heart failure, endocarditis) manifested in 12 patients (50% of survivors), with significant occurrence of proximal branch PA stenosis (75% and 50% for RPA and LPA, respectively).

Zhao et al[2] have published a large study comparing the outcomes of SPS vs surgical RV-PA connection (patch or conduit) to promote the growth of native

ABBREVIATIONS AND ACRONYMS

LPA = left pulmonary artery

MAPCA = major aortopulmonary collateral

MPA = main pulmonary artery

PA VSD = pulmonary atresia with ventricular septal defect

PA = pulmonary artery

RPA = right pulmonary artery

RVOT = right ventricular outflow tract

RV-PA = right ventricle to pulmonary artery

SPS = surgical pulmonary shunt

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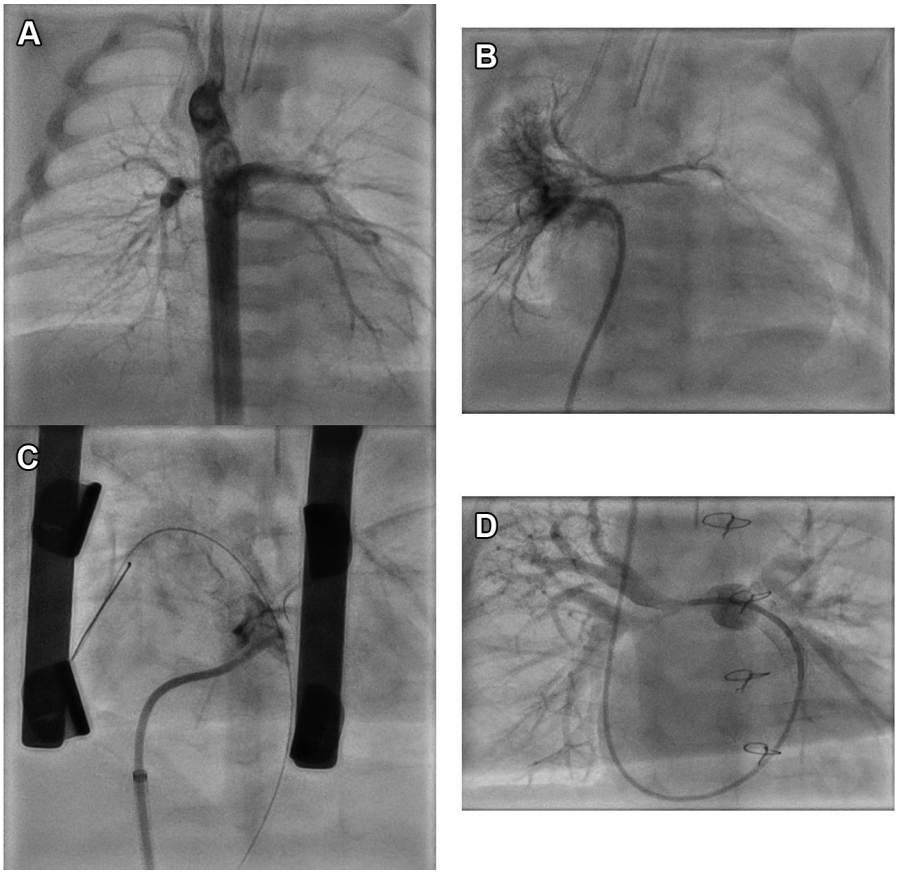
FIGURE 1 Hybrid Periventricular Right Ventricular Outflow Tract Stent in Pulmonary Atresia With Ventricular Septal Defect With Diminutive Branch Pulmonary Arteries



(A) Angiogram in descending aorta showing a major aortopulmonary artery collateral giving off 3 branches that supplied most lung segments. **(B)** Pulmonary venous wedge angiogram demonstrating diminutive central pulmonary arteries. **(C)** Hybrid coronary covered stent placement between the right ventricle outflow tract and pulmonary artery with good forward flow into native pulmonary arteries. **(D)** At last catheterization, distal branch pulmonary arteries have developed well, while severe proximal stenosis persists.

branch PAs in PA VSD MAPCAs. The probability of complete repair was significantly lower in the SPS group (56.0% vs 74.5% after 5 years in the RV-PA group). The increase in the mean Nakata index was significantly lower after SPS. The in-hospital morbidity and mortality after complete repair were similar between the 2 groups. However, suturing either SPS or RV-PA conduit on diminutive branch PAs comes with a high risk of distortion and thrombosis.

Hybrid periventricular RV-PA stenting is an option in these patients and has been reported previously in the literature.[6] It has been described in higher-risk low-birth weight infants,[7,8] who are not candidates for ductal stenting, and have unfavorable anatomy for SPS (hypoplastic branch PAs). Establishing more physiological forward RV-PA flow may promote better growth of the pulmonary tree than a surgical shunt similar to that demonstrated for tetralogy of Fallot patients after RVOT stenting.[9] Choice of the stent used depends on the underlying anatomy; uncovered coronary stent is an option where there is a membranous atresia of the pulmonary valve. In our case, however, use of the covered coronary stent was essential because there was true fibrotic gap between RVOT and MPA), as proven by bleeding after the initial balloon dilatation of the RV-PA segment.

Our case shows that we can achieve a promising result with this approach even in extremely diminutive branch pulmonary arteries, but it also highlights the technical difficulties, delicate periprocedural management, and expected reinterventions to promote optimal PA growth.

FOLLOW-UP

Our patient is now 10 months old and awaiting surgical RV-PA conduit, MAPCAs unifocalization, and fenestrated VSD closure with patch augmentation of the proximal branch pulmonary arteries. We have not decided to occlude MAPCAs because they serve as single pulmonary blood supply to selected lung segments; where dual supply is present, we have no concerns regarding excessive pulmonary blood flow due to MAPCA origin stenosis offering lung protection.

CONCLUSIONS

Transcatheter options to secure pulmonary blood flow in infants with PA VSD are expanding. Further studies are warranted to decide whether these could become the treatment of choice, as has been shown for ductal stenting instead of a surgical shunt. The idea of promoting optimal growth of branch PAs by establishing the continuity between RV and MPA by percutaneous or hybrid stent placement is attractive and should be kept in mind when making an initial management plan.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS periventricular right ventricular outflow tract stent, pulmonary atresia

APPENDIX For supplemental videos, please see the online version of this paper.