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Case Report

Hybrid Right Ventricular Outflow Stent Insertion in a Small Neonate with Muscular Pulmonary Atresia with Intact Ventricular Septum: A Case Report

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Pulmonary atresia with intact ventricular septum (PAIVS) is a rare congenital heart disease that often needs a critical decision on whether to open the right ventricular outflow tract (RVOT). Significant morbidity and considerable mortality might preclude the safe use of percutaneous or surgical right ventricular decompression in patients with muscular PAIVS. We report the case of a 21-day-old neonate weighing less than 3 kg who underwent hybrid RVOT stent insertion as initial palliation for muscular PAIVS and subsequent anatomical correction at 5 months of age, with 6 years of follow-up.

Keywords: Hybrid, Right ventricular outflow stent, Pulmonary atresia with intact ventricular septum, Congenital heart disease, Case report

Case report

A 2.83-kg female was born through vaginal delivery at a gestational age of 39 weeks and 2 days with a diagnosis of pulmonary atresia with intact ventricular septum (PAIVS). Postnatal transthoracic echocardiography demonstrated muscular atresia of the pulmonary outflow, a dysplastic tricuspid valve with severe regurgitation (peak velocity, 3.2 m/sec), and a severely dilated right atrium and right ventricle (RV) with good left ventricular function. The baby was initially treated with intravenous prostaglandin infusion and supplementary oxygen. Daily follow-up transthoracic echocardiography showed gradually increasing peak velocity of the tricuspid regurgitant jet (4.2 m/sec), suggesting that the RV could be an independently functioning ventricle.

As the RV function seemed acceptable, the treatment strategy for this patient aimed to create forward flow through the pulmonary outflow. Given the muscular nature of pulmonary atresia, the only option for creating RV forward flow would be surgical opening with cardiopulmonary bypass (CPB). In addition, since the ductus arteriosus should be eliminated when using CPB, an additional systemic-to-pulmonary artery shunt might be created if oxygen saturation was not acceptable. However, although the surgical approach of an RVOT patch and central shunt on CPB is time-tested, it would involve substantial morbidity for this small neonate. Therefore, we decided to open the RVOT without CPB and not to touch the patent ductus arteriosus as a backup source of pulmonary blood flow immediately after the procedure.

At 21 days of age, the patient was sent to the hybrid operating room and the CPB machine was on standby for emergency situations. Under midline sternotomy, the main pulmonary artery (MPA) was dissected freely. A purse-string suture was placed 2 cm below the junction between the RV and MPA for angiocath cannulation, considering the entry angle of the catheter and length of the stent with help from a preoperative computed tomographic scan (Fig. 1). A 20G angiocath was cannulated and advanced through the junction between the MPA and RV. Through the angiocath, the guide wire was placed under fluoroscopy, and the sheath and stent-deploying device were serially inserted. Finally, a stent (4×15 mm) was placed, crossing the junction between RV and MPA with ballooning. Post-stent angiography showed a well-posi-

tioned stent without violation of the branch pulmonary arteries (Fig. 2).

Since entering the intensive care unit, the patient's oxygen saturation fluctuated for a week, although the ductus arteriosus remained open. Inhaled nitric oxide was required for 10 days, and the patient could be weaned from mechanical ventilation 19 days after hybrid RVOT stenting. The patient could be transferred to the general ward on postoperative day 21, and she was discharged with sildenafil, bosentan, and aspirin. Transthoracic echocardiography at discharge revealed an acceptable amount of forward flow through the RVOT stent, severe tricuspid valve regurgitation (TR), and no remnant flow of the ductus arteriosus. Peripheral oxygen saturation on pulse oximetry was 75%–80% on room air.

During follow-up, the patient showed favorable weight gain (>35th percentile) with good oral intake. A computed tomographic exam and echocardiographic exam at 4 months after hybrid stent placement revealed well-grown branch pulmonary arteries without focal narrowing, a relatively stenotic RVOT stent, severe TR, and acceptable RV function. In cardiac catheterization, the RV end-diastolic pressure was 0 mm Hg and the visual RV contractility was good. Therefore, at 5 months of age, the patient underwent anatomical repair, including tricuspid valve repair, transannular widening of the RVOT using 0.625% glutaraldehyde-treated autologous pericardium after complete extraction of the previous stent, and partial closure of atrial septal defect. The postoperative course was uneventful, and transthoracic echocardiography at discharge showed a mild degree of TR, unobstructed RVOT, severe pulmonary regurgitation, and a small atrial septal defect with a bidirectional shunt and acceptable RV contractility.

At 7 years of age, the patient underwent pulmonary valve replacement with a bioprosthesis and re-repair of the tricuspid valve for significant pulmonary valve regurgitation and TR. The last follow-up transthoracic echocardiography showed a well-functioning prosthesis at the pulmonary position without stenosis, a mild degree of TR without stenosis, and acceptable ventricular function. The patient does not take any cardiac medication except aspirin and is able to play well as of the latest follow-up visit.

This case report was approved by the Institutional Review Board (IRB) of Asan Medical Center (IRB number: S2022-2446-0001; IRB approval date: November 17th, 2022), and the need for informed consent was waived.

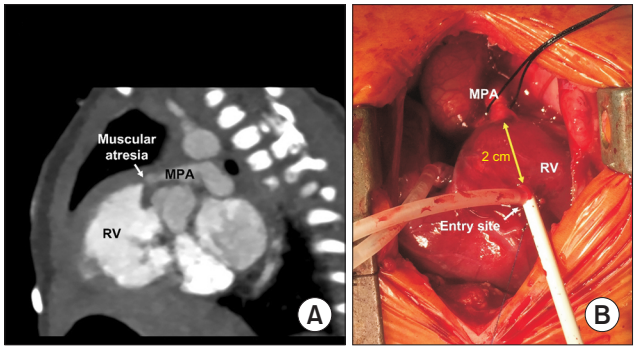

Fig. 1. The morphology of right ventricular outflow tract (RVOT) on preoperative computed tomography and the operative field. (A) Preoperative computed tomography revealed the muscular nature of the atretic segment. (B) An intraoperative photograph showed the entry site on the right ventricle (RV) free wall. MPA, main pulmonary artery.

Discussion

PAIVS is a rare condition, accounting for approximately 1% of cases of congenital heart disease. Despite the improvement of surgical and medical support of PAIVS patients, the surgical mortality rate has remained significantly high, at around 20% to 40% [1,2]. The treatment strategy for PAIVS mainly depends on the morphological heteroge-

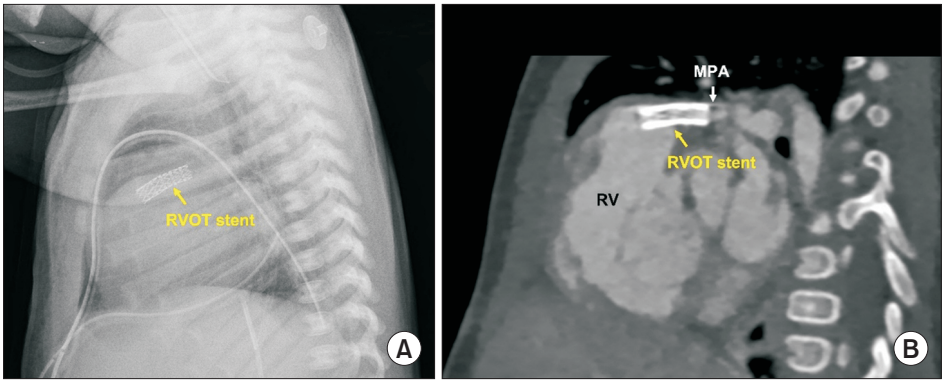

Fig. 2. Postoperative morphology of the right ventricular outflow tract (RVOT) stent. (A) Immediately postoperative chest X-ray. (B) Postoperative computed tomography obtained 4 months after the hybrid procedure. MPA, main pulmonary artery; RV, right ventricle.

neity of the RV, as well as the tricuspid valve. In patients with a small RV or RV-dependent coronary circulation, Fontan-type single ventricular repair could be the sole therapeutic option, but the goal of early palliation in patients with adequate RV is to create forward flow through the RVOT to achieve the growth of the RV for future anatomical repair and supplying blood flow to the pulmonary system [3-5].

Even when the RV is large enough to accommodate systemic venous return, if the RV function is too weak to generate enough forward flow to maintain RV output, the patient should be repaired using a single-ventricle repair strategy. In our case, the RV was large enough for bi-ventricular repair, but the RV function, which was indirectly measured with the peak velocity of the tricuspid valve regurgitant jet (3.2 m/sec), was inadequate for anatomical repair. Fortunately, the peak velocity of the tricuspid regurgitant jet gradually increased and finally reached 4.2 m/sec at 10 days of age; thus, we decided to open the RVOT in order to use the RV as a functioning ventricle.

Percutaneous transcatheter intervention for RV decompression has been a good option for membranous pulmonary atresia in patients with PAIVS; however, for muscular pulmonary atresia, the surgical approach might often be necessary. The hybrid approach for RV decompression in PAIVS was recently introduced [6], and it is performed routinely with comparable outcomes to the surgical approach [4]. In previous reports, the suggested advantages of hybrid RV decompression were higher rates of procedural success, lower rates of procedural complications, lower risk of RVOT perforation, and lower rates of further neonatal surgery than percutaneous catheter-based intervention, as well as the avoidance of neonatal exposure to CPB, which may have long-term detrimental effects, compared to surgical interventions [4]. In our case, we could expect all the advantages of hybrid RV decompression. Unlike the above-mentioned reports, we used a stent for the RV to open the RVOT instead of ballooning due to the muscular nature of the atretic junction between the RV and MPA [7].

In our patient, the key to successful stenting was determining an accurate entry point. In this patient, unlike in patients with membranous PAIVS, since the catheter should pass through the muscular atresia structure, careful consideration regarding the entry angle of the catheter and the length of the stent is needed for determining the proper location of the entry point on the RV free wall. Preoperative computed tomography provided useful information for deciding the entry point to deploy the stent at the atretic point, without violating the branch pulmonary arteries.

In conclusion, the hybrid approach with RVOT stent insertion for RV decompression could be a safe and effective option as initial palliation in small neonates with muscular PAIVS, with the eventual goal of achieving future anatomical correction.

Article information

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Conflict of interest

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