A Devised Option of Neonatal Palliation for Compromised Tetralogy of Fallot with Absent Pulmonary Valve Syndrome

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Neonatal primary repair of tetralogy of Fallot (TOF) with absent pulmonary valve (APV) syndrome is associated with high mortality rates. Our plan involves a staged repair that avoids one-stage intracardiac repair (ICR), with a first palliation that closes the main pulmonary orifice using an expanded polytetrafluoroethylene (ePTFE) patch, pulmonary arterioplication, and an adjustable Blalock-Taussig (BT) shunt. This strategy was used for a neonatal case with TOF/APV syndrome with hypoplastic left ventricle (LV). There was evidence of subsequent progressive increase in the LV size, and bronchial compression was relieved and an ICR was performed successfully at 9 months of age.

Keywords: tetralogy of Fallot with absent pulmonary valve syndrome, neonatal palliation, small left ventricle

Introduction

Absent pulmonary valve syndrome (APV) is a rare disease that is estimated to occur in 3%–6% of patients with tetralogy of Fallot (TOF/APV).1,2) It is characterized by a hypoplastic pulmonary valve, severe pulmonary insufficiency (PI), and largely dilated pulmonary arteries (PAs), which leads to tracheobronchial compression. Neonates with these symptoms require surgery shortly after birth, but primary repair of the TOF/APV is associated with high mortality rates of 9%–33%, that is likely to be higher in neonates.3) Our staged strategy with a first palliation includes a main pulmonary orifice patch closure, pulmonary arterioplication, and an adjustable Blalock-Taussig (BT) shunt, which may reduce the risk of progressive bronchial obstruction by PA wall plication, allow complete regulation of PI, and relieve excess preload on the small left ventricle (LV) compared to the primary repair in a neonate.

Case Report

At 36 weeks gestation, a 2623 g baby boy was born who had a prenatal diagnosis of TOF/APV. After birth, he had progressive respiratory distress and was intubated. Echocardiography confirmed the diagnosis of TOF/APV with a hypoplastic LV and detected that there was limited LV inflow because the dilated PAs were compressing the LA. Left ventricular end-diastolic volume (LVEDV) was too small to be majored using the modified Simpson method and was measured as 38.7% of the normal value using the Teichholz method. Maximum diameter of the right PA was 13.4 mm, left PA was 14.1 mm, and right ventricular end-diastolic volume (RVEDV) was 313% of the normal value, indicating enlargement by severe PI. Enhanced computed tomography showed extremely dilated PAs, with a PA index of 2,550 mm²/m², and compressed and narrowed the left
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bronchi (Fig. 1). Therefore, PI regulation and a reduction plasty of the extremely dilated PA were required.

Because the patient had inappropriate anatomical features for primary intracardiac repair (ICR) in the neonatal period, a staged repair was indicated for the patient who was at 7-day-old and had 2,370g. Beginning with a median sternotomy, a 4-mm expanded polytetrafluoroethylene (ePTFE) tube (GoreTex; W.L. Gore & Associates, FLAGSTAFF, AZ) to brachiocephalic artery anastomosis was performed. A cardiopulmonary pulmonary bypass was established using a bicaval cannulation and an ascending aortic cannulation. After clamping the main and distal bilateral PAs, the anterior wall of the main to left and right first branches of the PA in the hilum of each lung was excised without cardiac arrest. Septation of the main PA just above the pulmonary valve was performed using a 0.6 mm ePTFE patch. The anterior wall of the main to left and right PA was reduced by resection and plication. This extensive reduction PA plasty relieved the compression of the bronchi and LA, which caused an immediate improvement in respiratory distress symptoms and reduced the LV inflow obstruction. The distal side of the 4-mm ePTFE tube was anastomosed to the central pulmonary artery and regulated by clipping the 4-mm ePTFE tube (Fig. 2).

Postoperatively, the patient required ventilator support for 12 days. After extubation, his postoperative hemodynamic and respiratory conditions were stable. Computed tomography at 34 days revealed that compression of the left bronchi by the left PA was relieved, and the PA index had decreased from a preoperative value of 2550 mm²/m² to a postoperative value of 525 mm²/m². Echocardiography at 71 days showed an increase in LV inflow by decompression of the LA and an increase in the LVEDV from 38.7% of normal value to 78.7%, measured using the Teichholz method. At eight months of age, gradual hypoxemia (SpO₂ of approximately 75%) required that the BT shunt flow be adjusted by ballooning and declipping from the ePTFE tube. That resulted in improving the SpO₂ from 72% to 82% and the Qp/Qs from 0.67 to 1.37. Cardiac catheterization at that time confirmed that the small LV had increased and that there was a well-balanced LVEDV and RVEDV (127.2% of normal value.
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and 139.5% of normal value, respectively). The left ventricular ejection fraction was 59%, the PA index was 668.3 mm$^2$/m$^2$ and there was no PI. Therefore, we decided to perform an ICR.

The septation patch that was removed intact and continuous with the main PA. The muscle bar was resected from the right ventricular outflow tract (RVOT), the perimembranous ventricular septal defect (VSD) was closed using a 0.4-mm ePTFE patch, and the RVOT was reconstructed with a handmade monocuspid transannular patch using a 0.1-mm ePTFE membrane for a cusp and a 0.4-mm ePTFE membrane as a patch. The postoperative course was uneventful without respiratory distress or heart failure. Postoperative image was showed (Fig. 3).

Discussion

Surgical TOF/APV strategies for primary repair in patients with this disease remain challenging. Yong and colleagues$^4$ used a multivariate analysis and reported that the risk factors for late reoperation were neonatal primary repair and prematurity, and other authors reported that preoperative ventilation was the only risk factor for overall mortality.$^3$–$^5$ A TOF/APV pathology specimen, reported by Sakamoto and colleagues,$^6$ showed that a patient with TOF/APV who required intubation had irreversible bronchial sclerosis and emphysema-like changes that were caused by compression of dilated PAs. This suggests that surgical intervention should be considered immediately for a respiratory symptomatic baby and a baby with progressively dilated PAs who is in danger of dyspnea. In this situation, either a neonatal one-staged operation or an initial palliation with main PA ligation might be selected.

Our strategy reduces the risk of neonatal TOF/APV compared to one-staged primary repair because we can avoid ICR, cardiac arrest, and right ventriculotomy. Septation of the main PA can also completely regulate PI, which is the main cause of this syndrome, and would prevent progressive dilation of PAs, tracheobronchial compression, respiratory dysfunction, re-operation, and early death. In the part of PA anterior wall resection and plication, we make a target to normalize PA index to under 500 mm$^2$/m$^2$ and relieve the compression of the bronchi which is made sure using bronchoscopy. If the PA volume reduction was insufficient for relieving the compression of the bronchi, the posterior wall resection and plication would be a good option.

For the final ICR procedures, there are many options for the right ventricle outflow tract reconstruction (RVOTR): monocuspid transannular patch, valved conduit, homograft, and valveless transannular patch. Yong and colleagues$^4$ reported that there was no difference in late reoperation rates between valved conduit, monocusp, or valveless techniques. When once the patients get through severe neonatal condition, they have grown up and can be tolerant to be performed our standard procedure for TOF (i.e., conotruncal repair). We think that it is a reliable method and these patients have an excellent long-term prognosis.$^7$ If the patient gets enough growth and body weight to be available for RVOTR with valved conduit, this could be considerable choice because control of pulmonary regurgitation was very important for avoiding the dilatation of PAs.

Conclusion

Staged-repair with first palliation with mPA septation is much safer than primary repair, especially for a TOF/APV patient with a small ventricle. The benefit of a first palliation that allows the continuity of the main PA trunk to be maintained is that it might enable a RVOT standard transannular patch reconstruction without using difficult
techniques. Our strategy would be one good option for a compromised TOF/APV case.

**Disclosure Statement**

The authors have no conflicts of interests.

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