Challenge of Hypoplastic Right Ventricle with Multiple Associated Anomalies: Biventricular Repair or Univentricular Approach?

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Abstract:

Hypoplastic right ventricle is a rare congenital heart disease, as attested to by the small number of cases reported so far. We present a patient with hypoplastic right ventricle and multiple associated anomalies, namely ventricular septal defect, atrial septal defect, and pulmonary stenosis. Our principal concern was whether biventricular repair or Fontan type surgery would be the optimal management in this patient. We succeeded in performing biventricular repair via multi-modality cardiac imaging using intraoperative transesophageal echocardiography.

Key words: Right Ventricle, Hypoplastic

Introduction:

Hypoplastic right ventricle (RV) can be an integral part of such malformations as pulmonary atresia with intact ventricular septum and tricuspid atresia. This anomaly is also occasionally associated with a variety of congenital heart defects like ventricular septal defect (VSD), atrial septal defect (ASD), atrioventricular septal defect, and other complex congenital heart diseases. The existing literature contains only a few cases with isolated hypoplastic RV and ASD; 1-3 nevertheless, what they all unanimously confirm is the fact that the surgical treatment of these patients is certain to throw up many a challenge.

We herein report a case with hypoplastic RV, the crux of which was whether the RV would resume its proper function following ASD and VSD closure without a rise in the caval venous pressure.

Case Report

An 18-year-old man was referred to our Adult Congenital Heart Disease Clinic for Fontan type surgical treatment. The patient had had cyanosis since infancy and exacerbation of dyspnea of a few months' duration. Moreover, he had undergone palliative surgery (modified left Blalock-Taussig [BT] shunt) when he was 2 years of age with the diagnosis of RV hypoplasia, severe pulmonary stenosis, ASD, and VSD with a right-to-left shunt at both ventricular and atrial levels, based on cardiac catheterization conducted at the time. On physical examination, the patient had stable vital signs with severe cyanosis and clubbed fingers and toes. He was well nourished with a body surface area of about 1.6m2. The first and second heart sounds were normal and soft, respectively, and there was a grade 3/6 harsh systolic murmur at the upper left sternal border. Lab data showed secondary erythrocy-

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tosis (hemoglobin=17g/dL). Chest X-ray revealed a normal cardiothoracic ratio with reduced pulmonary vascular marking and hypoperfused lung fields. Transthoracic and transesophageal echocardiographic studies demonstrated a moderate-sized secundum type ASD (15 mm in diameter) with a right-to-left shunt; hypoplastic RV with a small inlet (tricuspid annulus= 28 mm) and a nearly absent outlet portion, which was associated with subvalvular and valvular pulmonary stenosis; and also a moderate-sized mid-muscular VSD (13 mm) with a right-to-left shunt (Figure 1a-b).

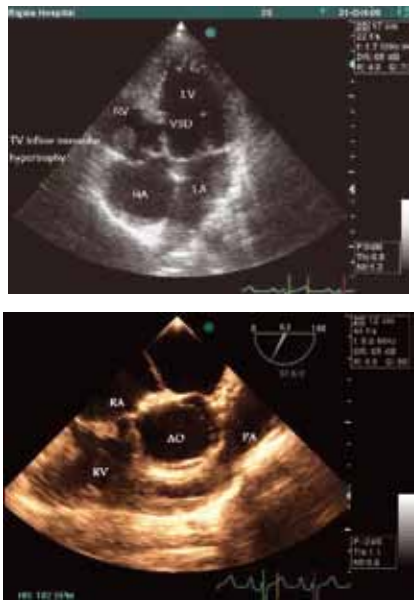

Figure 1. Echocardiographic images. (1a). TTE in four-chamber view showing hypoplastic RV with hypertrophied RV inflow associated with ASD and mid-muscular VSD. (1b) : TEE in RV inflow, outflow view showing hypoplastic infundibulum and main pulmonary artery.

There was a persistent left superior vena cava drain into the coronary sinus and a poor functioning left BT shunt. Cardiac Magnetic Resonance (CMR) study was carried out for a precise assessment of the RV size and function and revealed an RV end-diastolic volume (RVEDV) of 52.25 mL, RVEDV index of 30.68 mL/m2, RV ejection fraction of 35%, pulmonary valve annulus of 12 mm, left ventricular end-diastolic volume (index) of 150.84 mL (88.58 mL/m2), left ventricular ejection fraction of 54%, RV volume of about one third of the LV volume (Figure 2), and the main pulmonary artery before bifurcation 20 mm in diameter pulmonary artery branches (16mm in diameter each). In light of these findings, decision was made to candidate the patient for biventricular repair.

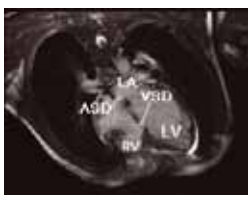

Figure 2. Cardiac MRI showing small RV with ASD and VSD

The patient underwent VSD closure with a Gortex patch in conjunction with partial closure of the ASD, resection of the infundibular muscles and hypertrophied RV via the tricuspid valve, and insertion of an RV to pulmonary artery valve conduit (23-mm homograft). In addition, the previous shunt was sutured.

Post-pump intraoperative transesophageal echocardiography showed a small residual ASD, small residual VSD, significant tricuspid regurgitation, and significant turbulence in the right ventricular outflow tract (RVOT) with a normally functioning pulmonary homograft. Given the unstable hemodynamic allied to significant tricuspid regurgitation and RVOT stenosis, the patient was subjected to a second pump run. The tricuspid valve was repaired, and the RVOT was enlarged with an RVOT patch.

Post-pump intraoperative transesophageal echocardiography demonstrated mild tricuspid regurgitation with no RVOT obstruction. The patient had an uneventful early postoperative follow-up period (Figure 3), and his cyanosis disappeared with 90% oxygen saturation by pulse oxymetry at room air. He was discharged in good physical condition and felt well in a 6 months' follow-up.

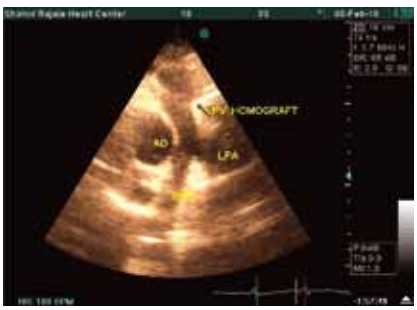

Figure 3.Postoperative TTE showing normally functioning pulmonary homograft

Discussion

Hypoplastic RV is associated with malformed apical, inlet, or outlet portion of the chamber. Our patient had a small and hypertrophied inlet (tricuspid annulus= 28 mm in diameter) and a nearly absent outlet portion, which was as-

sociated with subvalvular and valvular pulmonary stenosis. The challenge in opting for the best surgical modality for the patient was the RV size and function.

CMR is widely deemed the gold standard for the evaluation of the RV size and function. Our literature review showed a variety of figures for the normal range of the RVEDV. In the previously reported cases, there was a minimum RVEDV range of 58-154 mL and RVEDV index of 57-95 mL/m2. Our patient had an RVEDV range of 52.25 mL and RVEDV index of 30.68 ml/m2. He tolerated the surgical operation, whereby his RV was conferred an adequate size via RV inflow muscle resection and RVOT enlargement with patch augmentation. What played a significant role in pre and post-pump assessments was intraoperative transesophageal echocardiography.

With respect to literature review, it is deserving of note that so far only a small number of cases with isolated RV hypoplasia and ASD have been reported, suggesting that suitable patients for the simple closure of ASD be identified through the temporary occlusion of ASD with a balloon catheter1,4. In the present case, our balloon occlusion test was rendered inconclusive by the multiple associated anomalies (ASD, VSD, and pulmonary stenosis) that the patient had. The possibility of the bidirectional Glenn anastomosis method was deemed a proper solution lest the small RV not cope with the altered situation and the superior caval venous pressure exceed 20mmHg with a low cardiac output immediately after weaning the patient off the bypass surgery. Non-invasive cardiac imaging was helpful in the optimal management of our patient, who experienced a successful repair operation and event-free early follow-up period.

Conclusion

Hypoplastic RV with concomitant anomalies could be considered for biventricular repair via multi-modality imaging support. A restrictive, residual ASD or VSD allows a controlled right-to-left shunt, which may prevent excessive venous hypertension and allow adequate left-sided filling following biventricular repair.

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