Case Report
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Heart Transplantation in a Patient with Complex Congenital Heart Disease, Physiologic Single Lung, and Severe Pulmonary Hypertension

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Here we report a case of successful heart transplantation (HT) in a patient with high risk on HT due to her complex congenital heart disease and its consequent complications; physiologic single lung and severe pulmonary arterial hypertension. HT was successfully performed in our patient using meticulous perioperative management techniques, such as control of collateral vessels, appropriate donor selection, pulmonary vasodilator therapy, post-transplant extracorporeal membranous oxygenation support, and atrial septostomy for right ventricular unloading.

Key Words: Heart defects, congenital; heart transplantation; pulmonary arterial hypertension; physiologic single lung; pediatrics

INTRODUCTION

Favorable outcomes of heart transplantation (HT) have been reported in patients with congenital heart disease (CHD) and physiologic single lung.[1-3] However, HT is higher-risk in these patients, who may develop right ventricular (RV) failure related to pulmonary arterial hypertension (PAH) and postoperative bleeding and left ventricular (LV) volume overload by aortopulmonary collateral vessels. In patients with severe PAH, HT is much riskier. Therefore, these patients are excluded from HT and listed for heart-lung transplantation (HLT).[4] Nonetheless, the disadvantages of HLT, such as a longer waiting time, high waiting list mortality, and poor survival due to long-term lung complications, make it a less desirable option. We report a case of successful HT in a patient with complex CHD with physiologic single lung and severe PAH in the remaining lung.

Written informed consent was obtained from the patient.

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•The authors have no potential conflicts of interest to disclose.

CASE REPORT

A female patient diagnosed with pulmonary atresia with ventricular septal defect (VSD), absent intrapericardial native pulmonary artery, and major aortopulmonary collateral arteries (MAPCAs) underwent staged bilateral unifocalization of MAPCAs and a modified Blalock-Taussig shunt at the age of 17 and 20 months. Pulmonary angiogram at 27 months of age revealed hypoplastic pulmonary vasculature with multiple peripheral stenosis. The patient underwent central pulmonary artery reconstruction with the placement of an RV-to-pulmonary artery conduit and mitral valvuloplasty for mitral regurgitation. Her VSD could not be closed due to PAH. During follow-up, the left pulmonary artery was occluded due to massive chronic thrombi (Fig. 1). In the right pulmonary artery (RPA), multiple peripheral branchial stenosis developed, for which multiple attempts of percutaneous arterioplasty were futile. The patient's ventricular function also deteriorated. Magnetic reso-

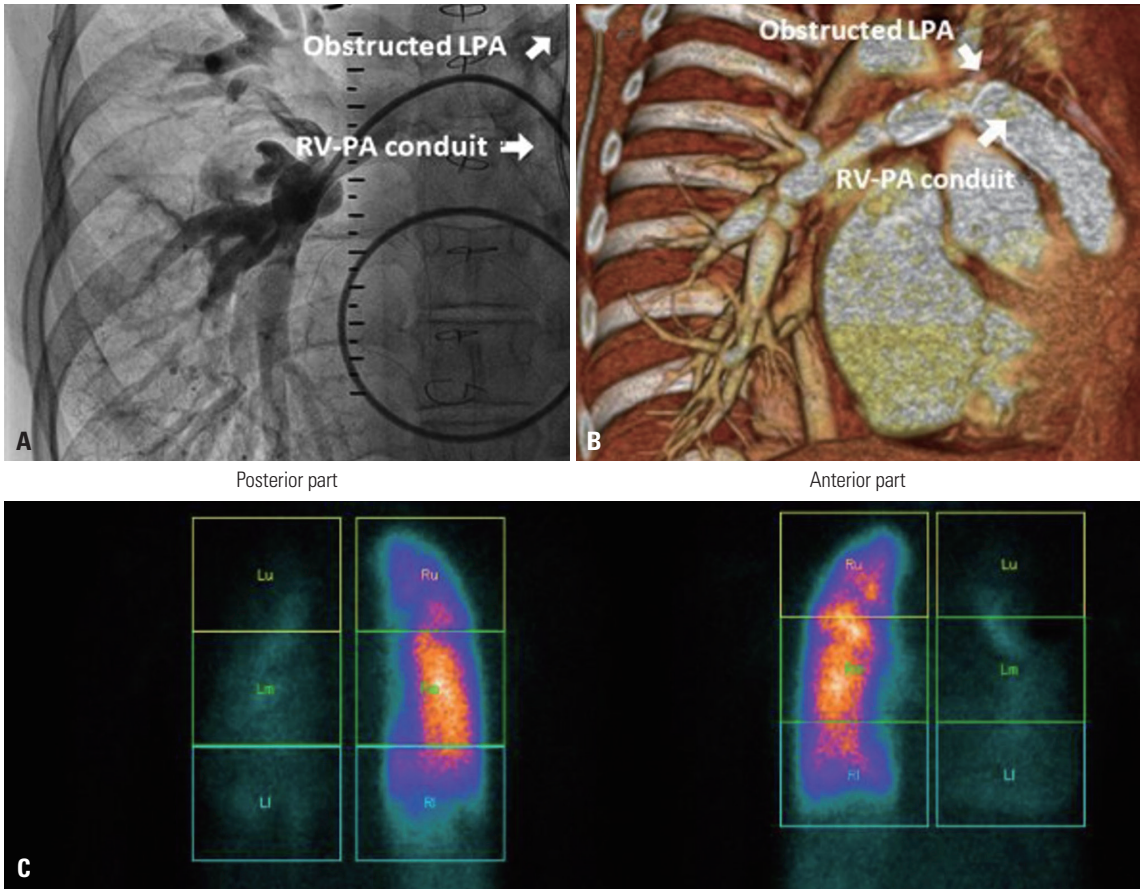

Fig. 1. Pulmonary angiogram (A) and computed tomography scan (B) revealed obstruction of the left pulmonary artery and right pulmonary artery with peripheral stenosis. Lung perfusion scan before transplantation (C) showed the patient's physiologic single lung status. LPA, left pulmonary artery; RV-PA, right ventricle to pulmonary artery.

nance imaging (MRI) revealed right-to-left dominant bidirectional shunt through the VSD, severe mitral regurgitation, and myocardial fibrosis. LV and RV ejection fraction (EF) was 47% and 48%, respectively. RV end-diastolic and systolic volume index was 313 and 162 mL/m^2, respectively. At 17 years of age, the patient had two episodes of ventricular tachycardia associated with syncope. Her SpO_2 was 75%, and she was in NYHA functional class IV. On cardiac catheterization, the main and right upper branchial pulmonary arterial pressure was 90/10 (38) mm Hg when systemic blood pressure was 114/55 (70) mm Hg. Due to the tight segmental arterial stenosis, the middle and lower RPA vascular bed was preserved [pressure was 32/10 (18) mm Hg]. Assumed indexed pulmonary vascular resistance was 11.3 WU*m^2 (Fig. 2). Balloon pulmonary arterioplasty for branchial stenosis was attempted, but it was not effective. An implantable cardioverter defibrillator was placed, and the patient was listed for HLT and HT for intractable ventricular arrhythmia, ventricular dysfunction, and severe cyanosis. Multiple aortopulmonary collateral arteries were embolized to reduce the post-transplantation left heart volume overload. HT was carried out when a heart became available from a larger-physique donor. The peripheral branch pulmonary arterial stenosis could not be addressed since those lesions were too peripheral for surgical approach. As anticipated, after the HT procedure, LV and RV dysfunction was observed in the transplanted heart. Cardiopulmonary bypass was converted to venoarterial extracorporeal membranous oxygenation (ECMO). Inotropes (epinephrine and milrinone) and pulmonary vasodilator therapy using nitric oxide, sildenafil, and bosentan was initiated, and the ECMO flow was gradually decreased for the LV and RV to adapt to the volume and pressure load. The patient was weaned from ECMO on post-transplantation day 5. The patient still had high RV pressure, RV dysfunction, and moderate tricuspid regurgitation. Atrial septostomy was performed, and stent on interatrial septum was inserted to allow RV unloading on post-transplantation day 6. Pulmonary arterial and aortic pressure measurement before atrial septostomy was 58/10 (30) and 108/53 (68) mm Hg, respectively. The signs of right heart failure improved. In 3 days, the patient was weaned from ventilator and free from inotropes on post-transplantation day 19. The patient showed a good clinical course with pulmonary vasodilator and diuretics (iloprost, sildenafil,

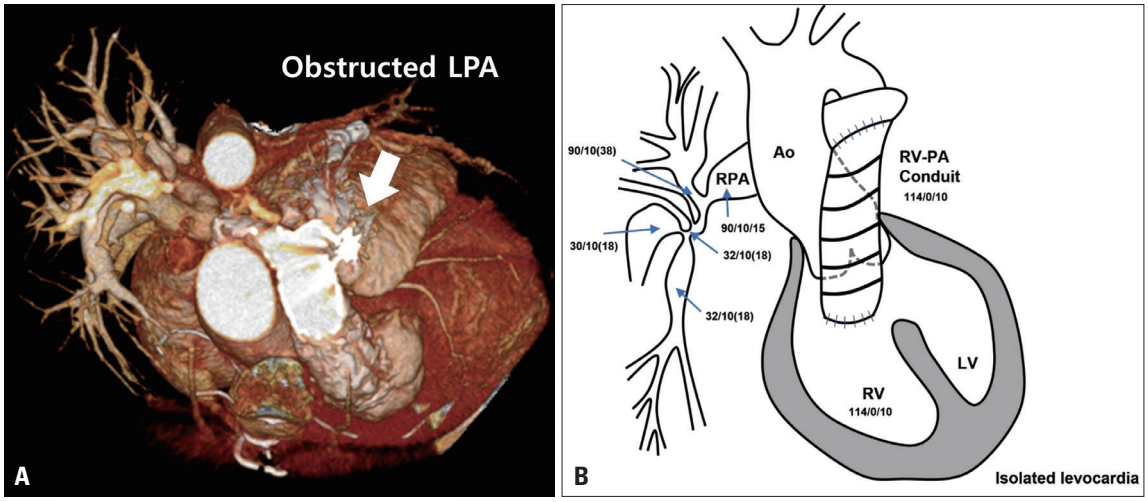

Fig. 2. Computed tomography scan (A) and schematic diagram of the pulmonary angiogram and cardiac catheterization data (B). Arrow indicates obstructed left pulmonary artery. *Numbers represent pressure (mm Hg), shown as systolic/diastolic (mean) pressure or systolic/diastolic/end diastolic pressure. The left ventricular pressure was identical to the RV pressure. PA, pulmonary artery; RPA, right pulmonary artery; RV, right ventricle.

ambrisentan, and furosemide) use at the 2-year follow-up without evidence of rejection on cardiac biopsy. Echocardiography revealed normal LV function and dimensions, an estimated systolic RV pressure of 70 mm Hg, and moderate RV dysfunction. On the latest echocardiogram, RV EF was 46% on MRI, tricuspid annular plane systolic excursion (TAPSE) was 12 mm, and tissue Doppler imaging (TDI) S' was 8 cm/sec.

DISCUSSION

As a consequence of pulmonary arterial arborization anomaly, our patient had physiologic single lung, multiple aortopulmonary collateral arteries, and severe PAH. These patients with severe PAH have been excluded from HT and listed for HLT.[4] Much longer waiting, higher waiting period mortality rate, and poorer long-term outcome of HLT led us to proceed with the HT when a heart organ donor was available. As anticipated, our patient experienced RV failure related to PAH and LV failure related to LV volume overload by aortopulmonary collateral vessels. HT was successfully performed in our patient using meticulous perioperative management such as pre-transplantation control of collateral vessels with embolization, appropriate donor selection, pulmonary vasodilator therapy, post-transplant ECMO support to allow progressive adaptation of the RV to high PA pressure, and atrial septostomy for RV unloading. Although the patient recovered from HT procedure, her high PA pressure is expected to negatively affect the long-term outcomes. It still remains unclear whether HT or HLT would be a better option for this patient. There has been no clear cut-off value to exclude patients with PAH from HT. However, we have learned that HT can be a feasible option than HLT for patients with complex CHD, physiologic single lung, and severe pulmonary hypertension.

AUTHOR CONTRIBUTIONS

Conceptualization: Jo Won Jung, Ah Young Kim, and Han Ki Park. Data curation: Hae Won Lee, Ah Young Kim, and Han Ki Park. Investigation: Hae Won Lee, Ah Young Kim, and Han Ki Park. Methodology: Hae Won Lee, Ah Young Kim, and Han Ki Park. Resources: Ah Young Kim and Han Ki Park. Supervision: Jo Won Jung, Ah Young Kim, and Han Ki Park. Validation: all authors. Visualization: Hae Won Lee and Han Ki Park. Writing—original draft: Hae Won Lee, Ah Young Kim, and Han Ki Park. Writing—review & editing: all authors. Approval of final manuscript: all authors.

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REFERENCES

1. Lamour JM, Hsu DT, Quaegebeur JM, Pinney SP, Mital SR, Mosca RS, et al. Heart transplantation to a physiologic single lung in patients with congenital heart disease. J Heart Lung Transplant 2004;23:948-53.
2. Zuckerman WA, Richmond ME, Lee TM, Bacha EA, Chai PJ, Chen JM, et al. Heart transplantation in the setting of complex congenital heart disease and physiologic single lung. J Thorac Cardiovasc Surg 2015;150:1465-72.
3. Kogon B, Shah N, Villari C, Book W, McConnell M, Parks J, et al. Cardiac transplantation in a patient with effective single left lung physiology. Transplantation 2007;84:128-30.
4. Reinhardt Z. Paediatric heart transplantation: an update. Arch Dis Child 2019;104:1216-22.

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