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Case Report

Orthotopic Heart Transplantation in a Child with Single Ventricle after Pneumonectomy

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We report a 6-year-old with single ventricle physiology secondary to tricuspid atresia who had cardiorespiratory failure who was not a candidate for further single ventricle palliation. The patient underwent planned staged left pneumonectomy for recurrent pneumonias secondary to bronchomalacia followed by orthotopic heart transplantation. This aggressive approach improved the patient candidacy for heart transplantation by removing the source of recurrent infection and respiratory failure (left lung). *ASAIO Journal* 2021; 67;e137–e139

Key Words: single lung, single ventricle, orthotopic heart transplantation, pneumonectomy

Orthotopic heart transplantation (OHT) is the only option for patients who have failed the single ventricle palliative pathway; yet many times, these patients are poor candidates due to multiple organ dysfunction or nutritional status.[1] Some centers consider tracheostomy a relative contraindication to OHT. In addition, persistent recurrent infection is another relative contraindication. Providers are left to weight the benefits of transplant while determining the likelihood of overcoming each of the relative contraindications. Successful transplant in congenital heart disease patients with single lung physiology has been reported previously[2,3] but is a high risk procedure in itself, even without the additional risks of tracheostomy and recurrent infection. We present a case of a 6-year-old female who had chronic respiratory failure and decompensated single ventricle/hemi Fontan physiology who underwent planned pneumonectomy to remove the source of recurrent infection (left lung), which improved the patient's candidacy for OHT and led to successful transplantation.

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Clinical Summary

This is a 6-year-old female with history of Cat Eye Syndrome (supernumerary iso-chromosome 22q11.2) who was admitted with cyanosis, fatigability, and complex medical and surgical history. She was born with functional single ventricle due to tricuspid atresia, large ventricular septal defect, normally related great arteries, supracardic total anomalous pulmonary venous return, left ventricular outflow tract obstruction, and hypoplastic aortic arch with aortic coarctation. She underwent initial palliation with pulmonary artery banding and aortic arch reconstruction in infancy, followed by arch revision and repair of total anomalous pulmonary venous return at 6 months of age. She was then palliated at 13 months of age with Damus-Kaye-Stansel procedure and placement of a modified Blalock-Taussig shunt. The next stage of palliation was a Hemi-Fontan procedure at the age of 3 years 1 month. She underwent postoperative stent placement in her right main branch pulmonary artery and a left hemi diaphragm plication for previous left phrenic paralysis with left-sided decortication for empyema. She has chronically been receiving monthly intravenous immunoglobulin infusions due to hypogammaglobinemia and is gastrostomy tube dependent. She was chronically tracheostomy and ventilator dependent due to left bronchomalacia.

She was deemed not to be a candidate for further surgery due to inadequate hemodynamics for completion of Fontan. Hemodynamics were significant for baseline measurements on 21% FI02 with low cardiac output of 2 L/min/m^2 with Qp:Qs of 0.55:1 and PVRi of 3.62 WUxm^2On 100% FiO_2 plus 40 parts per million of inhaled nitric oxide, her cardiac output decreased further to 1.64 L/min/m^2 with Qp:Qs improving to 0.84:1 and PVRi improving to 2.19 WUxm^2, suggestive of pulmonary vasoreactivity. The left pulmonary artery was diffusely hypoplastic with with elevated saturations suggestive of aortopulmonary collaterals. Lung ventilation and perfusion scans showed that there was only 10% perfusion and ventilation to left lung.

She struggled with recurrent left pneumonia precipitated by chronic left main bronchial obstruction (Figure 1A) with persistent left lung collapse and left lung hypoperfusion (Figure 2). Decision was made to improve/enhance (here or below—2 "optimize" words) the patient's quality of life with pneumonectomy of the compromised left lung to remove the source of recurrent infection in hopes of optimizing outcomewith OHT. Her outcome with an OHT and a native single lung was predicted to be better than a single lung followed by Fontan repair. In addition, heart/lung transplant is exceedingly high risk and was not a favorable option for this patient given multiple thoracotomies and sternotomies, as well as tracheostomy dependence. The patient underwent a

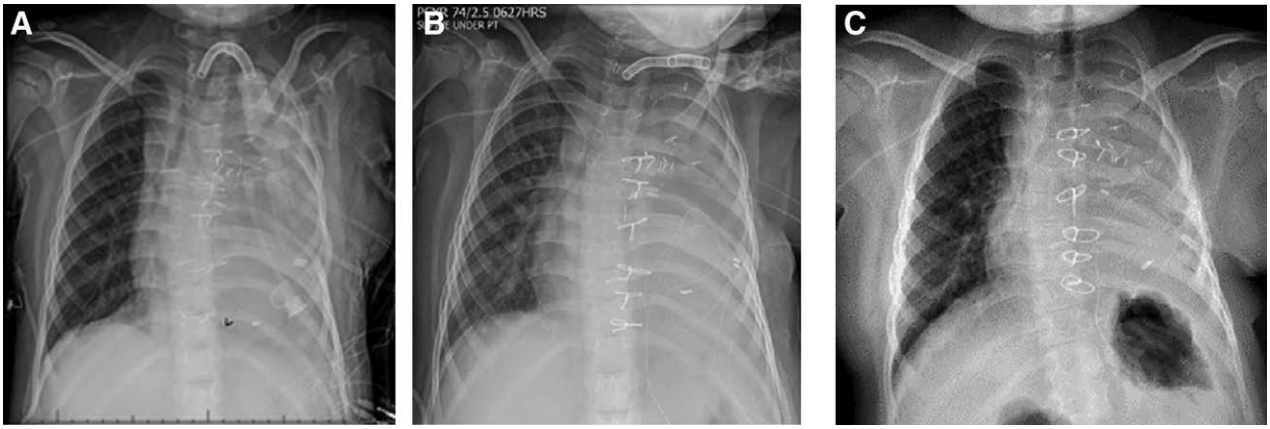

Figure 1. A: Preoperative CXR shows left lung collapse and pneumonia upon admission. **B:** Postoperative CXR postpnemonectomy. **C:** Postoperative CXR post OHT. CXR, chest x-ray; OHT, orthotopic heart transplantation.

left pneumonectomy *via* a redo left posterior thoracotomy at age 6 years (Figure 1B). After her thoracotomy had appropriate time to heal, she was assessed for candidacy for OHT. Cardiac catheterization was not repeated as her PVR was acceptable for transplant before pneumonectomy. Since she was able to wean ventilator to preoperative settings without any recurrence of pneumonia in the right lung, she was listed status 1A for heart transplant. Approximately 2 months following pneumonectomy, she underwent uneventful OHT (Figure 1C). Follow-up echocardiograms demonstrated good biventricular function and normal right ventricular pressures, and she was discharged home 3 weeks posttransplantation on pretransplant ventilator settings. She progressed rapidly from respiratory standpoint, had her tracheostomy removed 7 months post-OHT and at successfully had stoma closure without requiring tracheal reconstruction at 10 months post-OHT. Unfortunately, she developed EBV-associated posttransplant lymphoproliferative disorder diagnosed 13 months post OHT that was successfully treated with rituximab therapy. She is now 18 months post OHT with excellent cardiac function and good quality of life with no respiratory support and complete resolution of cyanosis and clubbing (Figure 3).

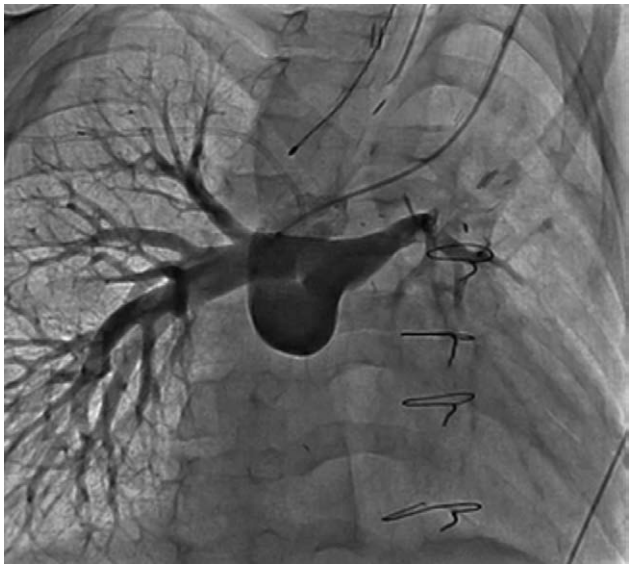

Figure 2. Preoperative pulmonary arteriogram shows left lung minimal perfusion and hypoplastic left pulmonary artery.

Discussion

Although single ventricle palliative strategies carry a relatively high morbidity and mortality risk, we try to delay OHT until patients have demonstrated failure of this pathway. Heart transplantation can provide life-saving treatmentbut is not without risk of morbidity and mortality in itself. The risks of complications are even higher in a congenital heart patient, who is sensitized from prior cardiac surgeries, has recurrent infections, and tracheostomy with ventilator dependence.[4,5] Although there are reports of successful OHT in congenital heart disease patients with single lung physiology,[3] our approach for pneumonectomy to remove source of recurrent infection before transplantation was a novel approach.

The patient described herein was deemed to have no further palliative surgical options given her hemodynamics. There is limited data on success of Fontan physiology to a single lung, especially given the elevated Hemi-Fontan pressures. There is, however, evidence of successful OHT in the adult and congenital heart population with single lung anatomy.[3] In this case, it was thought that the patient would benefit from removing the source of recurrent infection and then proceeding with OHT once stable to provide her the best chance at a successful outcome. Given that there was only 10% of the ventilation and perfusion to the left lung we anticipated she would be able to wean from ventilator once the left lung was removed and would not require the same degree of positive pressure.

Thus far, this strategy has proven very successful, with the patient able to rapidly wean from ventilator, successfully undergo decannulation and stoma closure, and has had normal cardiac function. She has a markedly improved quality of life. For this patient, the tracheostomy allowed for quicker discharge post-OHT with gradual wean in respiratory support. With this experience, we see no contraindication to proceeding with transplant in those that are tracheostomy dependent, if it is anticipated that improved

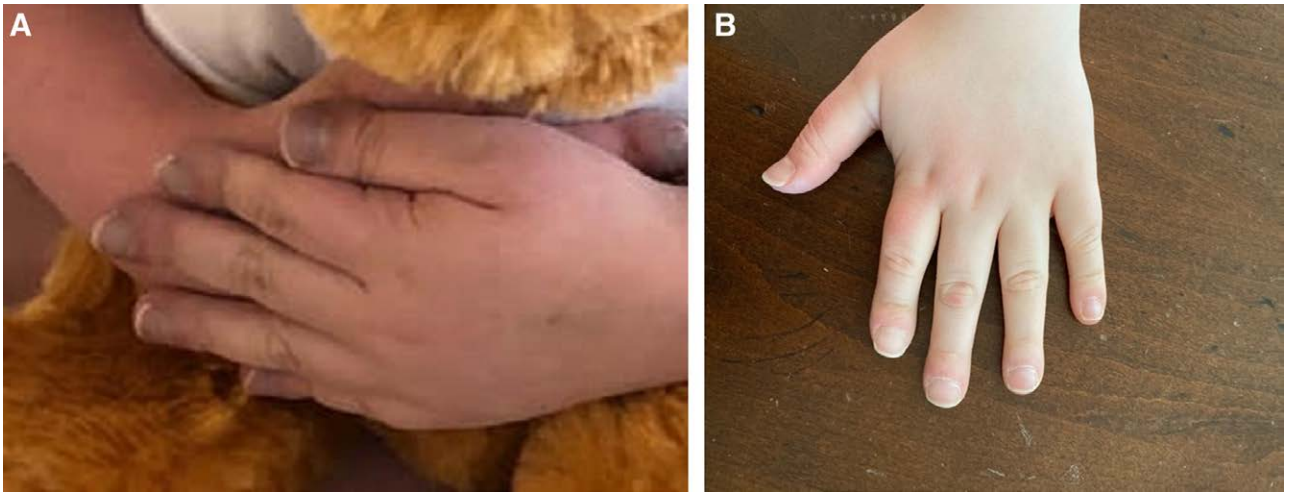

Figure 3. A: Prepnemonectomy photograph of fingernails with cyanosis and clubbing. **B:** Post-OHT photograph with complete resolution of cyanosis and clubbing.

cardiopulmonary function may allow successful weaning of ventilator post transplant.

Conclusion

As a general rule, chronic infection is a contradiction for organ transplantation. Surgical removal of the source of resistant recurrent infection despite maximal medical therapy (in our case left pneumonectomy) was a novel approach to optimize the patient's medical condition and allow candidacy for successful staged OHT with great outcome. In addition, this case report adds to the limited literature on success of OHT with single lung physiology, as well as pediatric heart transplantation with tracheostomy.

References

1. Bernstein D, Naftel D, Chin C, *et al*: Pediatric Heart Transplant Study: Outcome of listing for cardiac transplantation for failed Fontan: A multi-institutional study. *Circulation* 114: 273–280, 2006.
2. Singh TP, Vasquez JC, Delius R, Walters HL III: Neonatal heart transplantation to a physiologic single lung. *J Heart Lung Transplant* 25: 362–364, 2006.
3. Zuckerman WA, Richmond ME, Lee TM, *et al*: Heart transplantation in the setting of complex congenital heart disease and physiologic single lung. *J Thorac Cardiovasc Surg* 150: 1465–1472, 2015.
4. Jacobs JP, Quintessenza JA, Boucek RJ, *et al*: Pediatric cardiac transplantation in children with high panel reactive antibody. *Ann Thorac Surg* 78: 1703–1709, 2004.
5. Pietra BA, Kantor PF, Bartlett HL, *et al*: Early predictors of survival to and after heart transplantation in children with dilated cardiomyopathy. *Circulation* 126: 1079–1086, 2012.