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HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

The Road to Heart Transplant in a Patient With Cardiomyopathy, Shone Complex, and Severe Pulmonary Hypertension

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ABSTRACT

Our case report details the journey of a 16-year-old male patient with Shone complex and advanced heart failure. We highlight the pivotal role of the HeartWare Ventricular Assist Device (Medtronic) implantation in mitigating severe pulmonary hypertension, thereby facilitating his eligibility for a heart transplant. We discuss the subsequent management of post-transplant pulmonary hypertension and right ventricular dysfunction using targeted pulmonary vasodilators and inotropic support, underscoring the intricacies of postoperative care in pediatric heart transplant patients. This case emphasizes our observation of the critical role that left ventricular assist devices play in redefining transplant candidacy and the necessity for complex, ongoing management in pediatric heart transplant scenarios. (J Am Coll Cardiol Case Rep 2024;29:102323) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

CASE PRESENTATION

A 16-year-old male patient with a history of Shone complex and progressive heart failure was referred for advanced heart failure management because of cardiogenic shock and severe pulmonary hypertension (PHT). Born with multiple cardiac anomalies (**Figure 1**), his condition deteriorated by age 16, necessitating intravenous milrinone and dobutamine support. Right heart catheterization revealed severe PHT, with markedly elevated pulmonary vascular resistance and transpulmonary gradient, which improved with nitroprusside.

Given his condition's severity, a HeartWare Ventricular Assist Device (HVAD) (Medtronic) was implanted as destination therapy, significantly improving his PHT within 3 months, leading to his listing for heart transplantation as status 1A. Post-transplant, the patient faced challenges with residual PHT and right ventricular (RV) failure, managed with sildenafil and macitentan, alongside diuresis and intravenous milrinone, allowing successful weaning off epoprostenol. The patient's post-operative course showed marked improvement, with

LEARNING OBJECTIVES

- To discern HVAD's role in pretransplant management of severe PHT and heart failure in pediatric patients.
- To grasp post-transplant care complexities, including PHT and RV support in pediatric patients.
- To appreciate the multidisciplinary approach in pediatric heart transplant evaluations for complex cases like Shone complex.

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ABBREVIATIONS AND ACRONYMS

HVAD = HeartWare Ventricular Assist Device

LVAD = left ventricular assist device

PHT = pulmonary hypertension

RV = right ventricle

eventual weaning off sildenafil and stabilization of cardiac function, demonstrating no signs of rejection or cardiac dysfunction 2 years post-transplant (Table 1).

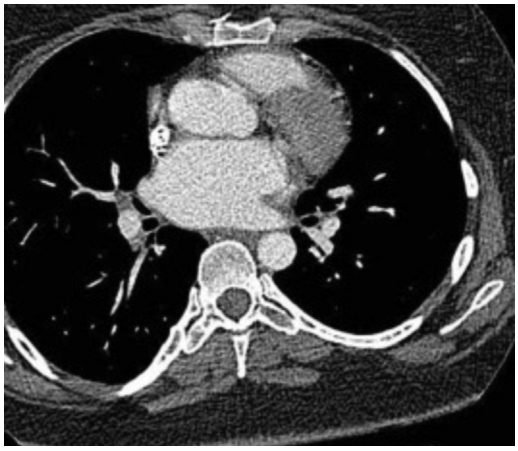
MULTIDISCIPLINARY TEAM'S THOUGHT PROCESS

The decision to proceed with HVAD implantation stemmed from a comprehensive evaluation by our multidisciplinary team, including cardiologists, cardiac surgeons, and imaging specialists. The team considered various alternatives but concluded that HVAD offered the best chance for improving the patient's condition and transplant eligibility due to his severe PHT and complex cardiac anatomy. Advanced imaging techniques, including 3-dimensional reconstructions of the patient's heart pre- and post-VAD placement (Figures 2 and 3), informed our surgical planning and device selection, ensuring a tailored approach to his unique anatomical challenges.

IMPACT OF UNITED NETWORK FOR ORGAN SHARING ALLOCATION CHANGES

Recent changes in United Network for Organ Sharing allocation have significantly influenced our decision-making process, particularly in high-risk patients like ours. These changes prompted our team to consider left ventricular assist device (LVAD) implantation more aggressively as a bridge to transplantation, recognizing the potential for longer wait times and the need for optimal patient stabilization pretransplant. The case of our patient, nearing adulthood, also raised considerations about the transition of care and the impact of allocation changes on pediatric vs adult patients, aligning with observations by Krishnamurthy et al[1] regarding the effects on patients with adult congenital heart disease.

FIGURE 1 Preoperative CT of Chest



Preoperative computed tomography (CT) of chest showing membranous subaortic stenosis.

DISCUSSION

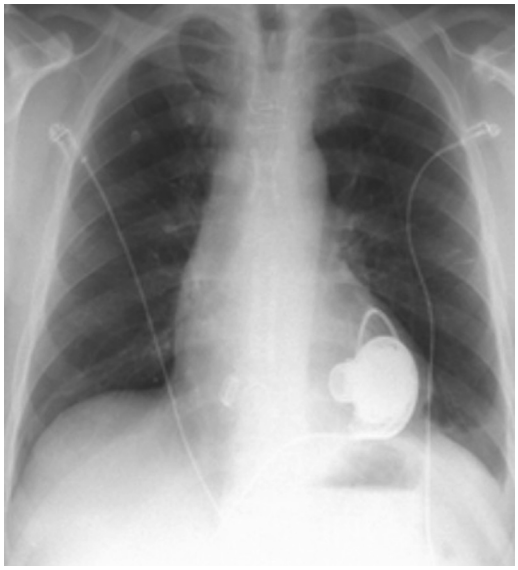
This patient's journey from severe PHT and heart failure to successful heart transplantation and postoperative recovery illustrates the critical role of LVADs in managing severe PHT, particularly in pediatric patients with complex congenital heart diseases. The use of HVAD significantly reduced pulmonary vascular resistance and transpulmonary gradient, aligning with trends noted in prior studies[2-4] and underscoring the efficacy of LVADs in acutely severe cases.[5,6] The case also highlights the utility of 3-dimensional printing for precise device selection and the importance of a tailored approach to post-transplant care, especially in managing residual PHT.[7-9]

TABLE 1 Right Heart Catheterization Data Pre- and Post-HVAD Implantation

	Pre-LVAD	Post-LVAD, 1st	Post-LVAD, 2nd	Post-HT, 1st	Post-HT, 2nd
Date measurement taken	August 8, 2019	August 29, 2019	November 14, 2019	December 4, 2019	February 24, 2020
Hemodynamics					
PA pressure (mean), mm Hg	80/55 (65)	76/25 (44)	57/17 (33)	40/24 (31)	39/18 (27)
PCWP, mm Hg	26	15	17	22	14
CO/CI (Fick), L/min/L/min/m^2	2.9/1.9	2.7/1.8	4.4/2.7	4.6/2.9	5.2/3.2
PVR, WU	13.4	10.7	3.6	1.97	2.5

CI = cardiac index; CO = cardiac output; HT = heart transplant; HVAD = HeartWare Ventricular Assist Device; LVAD = left ventricular assist device; PA = pulmonary arterial; PCWP = pulmonary capillary wedge pressure; PVR = pulmonary vascular resistance.

FIGURE 2 Postoperative CXR Showing HVAD



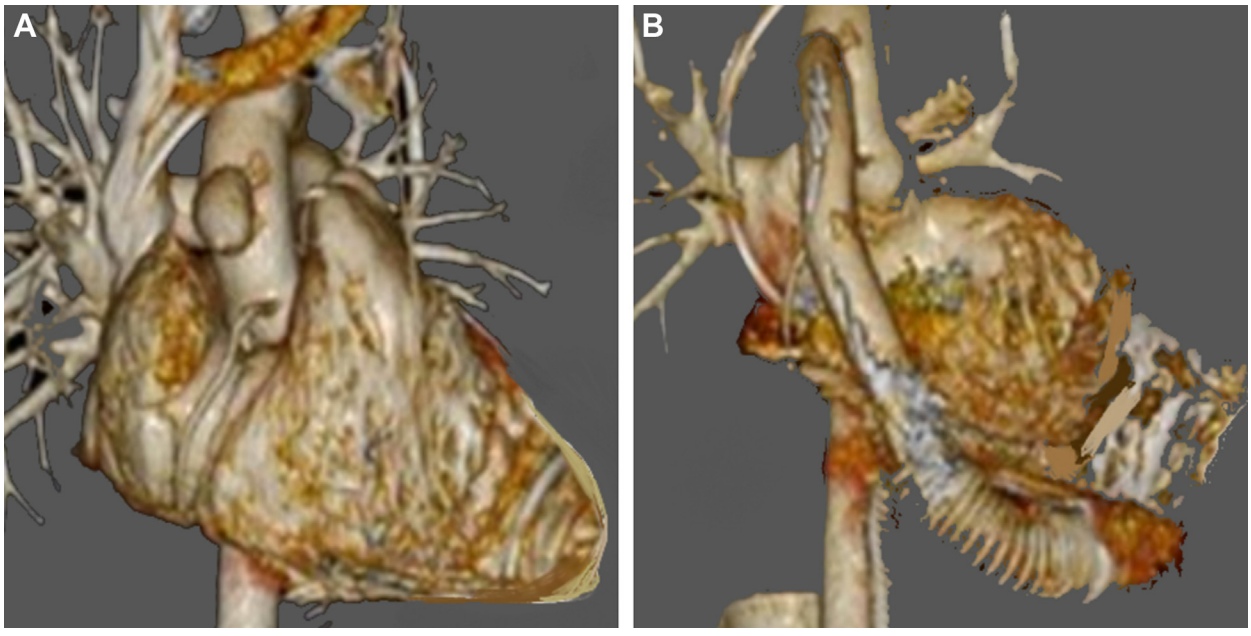
Postoperative chest x-ray (CXR) showing the position of HeartWare Ventricular Assist Device (HVAD).

Two years post-transplant, the patient's positive clinical trajectory, including the successful weaning off pulmonary vasodilators and the absence of rejection or cardiac dysfunction, reflects the sustainable impact of the comprehensive treatment strategy employed. This case contributes to the growing body of evidence supporting the efficacy of mechanical circulatory support in managing PHT in the context of heart transplantation and emphasizes the significance of personalized treatment strategies in complex pediatric heart disease management.

QUESTION 1: HOW DID HVAD IMPLANTATION ADDRESS THE SEVERE PHT HYPERTENSION AND HEART FAILURE IN THE PEDIATRIC PATIENT WITH SHONE COMPLEX?

In this case, the HVAD implantation served as a critical intervention for mitigating severe PHT and heart failure, thereby facilitating the patient's eligibility for heart transplantation. By providing mechanical circulatory support, the HVAD improved cardiac output and reduced the heart's workload, which was essential in managing the complex interplay of conditions presented by the Shone complex and severe PHT.

FIGURE 3 3D Reconstructed CT Pre- and Post-VAD Implant



Three-dimensional (3D) reconstructed CT pre- and post-VAD implant. Abbreviations as in Figures 1 and 2.

QUESTION 2: WHAT WERE THE KEY CONSIDERATIONS IN MANAGING POST-TRANSPLANT PHT AND RV DYSFUNCTION IN THIS PEDIATRIC PATIENT?

Post-transplant management focused on addressing residual PHT and RV dysfunction through targeted pulmonary vasodilators (sildenafil and macitentan) and inotropic support. The strategy aimed at stabilizing the patient's condition, ensuring the transplanted heart's stability, and preventing further complications. The careful selection and dosing of medications, along with close monitoring of the patient's response, were crucial in this phase.

QUESTION 3: WHAT ARE THE CHALLENGES AND SOLUTIONS IN SELECTING AND MANAGING HVAD IN PEDIATRIC PATIENTS WITH COMPLEX CONGENITAL HEART DISEASES LIKE SHONE COMPLEX?

The challenges in this case included selecting an appropriate HVAD size and type to accommodate the patient's small body size and complex heart anatomy. The solutions involved meticulous preoperative planning, including advanced imaging techniques to assess the heart's structure and function, and a tailored surgical approach to ensure the device's optimal placement and function. Postimplantation, the focus was on managing potential complications, such as infection or device malfunction, through vigilant monitoring and patient education.

QUESTION 4: HOW DID THE MULTIDISCIPLINARY TEAM APPROACH CONTRIBUTE TO THE SUCCESSFUL MANAGEMENT OF THIS PEDIATRIC HEART TRANSPLANT PATIENT?

The multidisciplinary team, comprising cardiologists, cardiac surgeons, imaging specialists, and other health care professionals, played a pivotal role in the comprehensive care of this patient. From preoperative assessment, through HVAD management, to post-transplant care, the team's collaborative efforts ensured that all aspects of the patient's complex condition were addressed, leading to a successful outcome.

QUESTION 5: WHAT INDICATORS WERE USED TO GUIDE WEANING OFF OF PULMONARY VASODILATORS POST-HEART TRANSPLANT, AND WHAT WAS THE OUTCOME?

Weaning off of pulmonary vasodilators was guided by indicators such as stable hemodynamics, normal pulmonary artery pressures, and the absence of RV dysfunction. The successful weaning indicated the patient's positive adaptation to the transplanted heart, with no recurrence of PHT symptoms, highlighting the effectiveness of the post-transplant management strategy.

QUESTION 6: REFLECTING ON THE CASE, WHAT ARE THE KEY LESSONS LEARNED ABOUT THE MANAGEMENT OF PEDIATRIC PATIENTS WITH SEVERE PHT AND HEART FAILURE REQUIRING HEART TRANSPLANTATION?

Key lessons include the importance of early and aggressive intervention with mechanical circulatory support in patients with severe PHT and heart failure, the value of a multidisciplinary approach in managing complex pediatric heart conditions, and the critical role of tailored post-transplant care in ensuring the patient's long-term health and quality of life. This case underscores the need for personalized treatment strategies and the potential for successful outcomes even in the most challenging cases.

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KEY WORDS **heart failure, heart transplant, pulmonary hypertension, Shone complex, ventricular assist device**