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Temporary axial-flow mechanical circulatory support and intravenous treprostinil in a patient with D-transposition of the great arteries and atrial switch: A case report

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A 43-year-old woman with D-transposition of the great arteries (D-TGA) with Mustard atrial switch repair, pulmonary venous baffle repair presented with advanced kidney and heart failure (HF). A transthoracic echocardiogram (TTE) showed a severely enlarged systemic right ventricular (RV) size with severely reduced systolic function, normal left ventricular (LV) size with severely reduced systolic function, mild to moderate systemic tricuspid regurgitation (TR) (Video 1) and patent pulmonary venous and systemic venous baffles on cardiac magnetic resonance.

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Cardiac catheterization on hospitalization day (HD) 10 after milrinone-assisted diuresis showed severe pulmonary hypertension [left pulmonary artery (PA) pressure 95/52 mmHg, mean 65 mmHg, pulmonary capillary wedge pressure (PCWP) 30 mmHg], systemic right ventricular end-diastolic pressure (sRVEDP) of 17 mmHg, cardiac index of 2.1 L/min/m^2 on milrinone (0.2 mcg/kg/min) and pulmonary vascular resistance (PVR) of 12.6 Woods units (WU) using sRVEDP. With administration of inhaled nitric oxide (iNO) 40 ppm, PA pressure was 100/50 mmHg with mean of 69 mmHg, PCWP increased to 40 mmHg and cardiac index and PVR improved to 2.3 L/min/m^2 and 9 WU respectively. This was consistent with both intrinsic precapillary pulmonary arterial hypertension (PAH) and post capillary pulmonary hypertension due to restrictive systemic RV function.

Milrinone and diuretic doses were increased and repeat hemodynamics on HD 21 (Table 1) showed decreased PA pressures and PVR by 40 and 50% respectively. At this point, intravenous treprostinil was started and uptitrated with increasing PA and PCWP by HD 31. With iNO administration, cardiac output increased by 40% and PVR decreased by 70% from baseline. However, the sRVEDP increased from a baseline of 17–30 mmHg indicating pulmonary vasoreactivity with restrictive systemic RV physiology. A CardioMEMS™ HF system was placed during that procedure to allow for close hemodynamic monitoring.

Given increasing sRVEDP despite decreasing PVR, we pursued systemic RV offloading with a temporary axial-flow mechanical circulatory support (MCS) device (Impella 5.5®) via right axillary approach on HD 35. After this, PVR decreased to 2 WU, subpulmonic LV function improved (Video 2) and PCWP decreased from 34 to 24 mmHg (Fig. 1). Orthotopic heart transplantation (OHT) and deceased-donor kidney transplantation were performed on HD 43 and 44, respectively.

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Table 1
Hemodynamics according to hospitalization days (HD).

	HD 10	HD 21	HD 31	HD 35
	Milrinone 0.2 mcg/kg/min	Milrinone 0.5 mcg/kg/min, FiO2 30%, iNO 40 ppm	Milrinone 0.5 mcg/kg/min, treprostinil 6 ng/kg/min, iNO 40 ppm	Impella 5.5® at P7, treprostinil 6 ng/kg/min and milrinone 0.5 mcg/kg/min
Systemic venous baffle	14 mmHg	–	–	15 mmHg
Subpulmonary left ventricle	95/17 mmHg	–	–	–
Pulmonary artery	95/52/65 mmHg	60/30/42 mmHg	78/37/52 mmHg	48/31/35 mmHg
Pulmonary capillary wedge	30 mmHg	22 mmHg	34 mmHg	24 mmHg
Systemic right ventricle	103/17 mmHg	88/17 mmHg	96/30 mmHg	–
Cardiac output	3.8 L/min	5.5 L/min	5.5 L/min	7.1 L/min
Cardiac index	2.1 L/min/m^2	3 L/min/m^2	3.1 L/min/m^2	4 L/min/m^2
Pulmonary vascular resistance	12.6 WU	4.5 WU	3.3 WU	2 WU

Intravenous treprostinil was weaned off on POD 4 and vasopressors on POD 5. Following this, the mean PA pressure progressively increased to 42 mmHg associated with RV enlargement with mildly reduced systolic function and severe TR. In consequence, milrinone was increased, and iNO and intravenous treprostinil were restarted on POD 7, achieving regression in RV dilatation and degree of TR by POD 15. She was discharged on POD 30 on subcutaneous treprostinil. On POD 50 she was transitioned to oral treprostinil with stable RV imaging. The patient granted consent for this publication.

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Patients with unpalliated D-TGA had a mortality rate of 90% in the first year of life until the atrial switch operation (via Senning or Mustard techniques) became widespread in developed countries in the 1970s [1]. This restores in-series biventricular circulation via the creation of atrial baffles, and the RV becomes the systemic ventricle. The functional systemic RV decline arises from its fibromuscular architecture, shape and function inherently suited to supply the pulmonary rather than systemic circulation, consequent maladaptive pressure response and volume overload, coronary artery supply mismatch, tricuspid valve failure and progressive RV dilatation [1]. Therefore, this population constitutes a significant proportion of adults with congenital heart disease and HF hospitalizations [2]. In the absence of medical therapies that impact imaging or exercise outcomes [3], many patients will require OHT.

Prior studies have demonstrated up to 33% prevalence of PAH in this population [4]. This is important when considering OHT alone versus combined heart and lung transplantation (CHLT) [5] as demonstrated in two case series. In the first one, of 18 patients with systemic RV, 2 had significantly elevated PA pressures (mean 71 and 47 mmHg) and PVR (8.9 and 4.2 WU). The first patient was already receiving intravenous epoprostenol and required CHLT. The second patient underwent OHT alone, received inhaled epoprostenol intraoperatively but died from pulmonary hypertensive crisis [4]. In the second series of 7 patients with a systemic RV, 2 patients were found to have suprasystemic PA pressures (with PVR 12.3 and 8.9 WU) before undergoing ventricular assist device (VAD) implantation. One of them died on POD 17 due to recurrent VAD thrombosis and the other had significant hemodynamic improvement with decline in mean PA pressure from 111 to 30 mmHg, and PVR from 8.9 to 3.9 WU, leading to a subsequent OHT [6].

As demonstrated here, sequential use of intravenous treprostinil and temporary axial-flow device was successful in achieving a rapid reduction in mean PA pressure, PVR, and PCWP. Additionally, we highlight the rebound pulmonary hypertension seen post-OHT after discontinuation of intravenous treprostinil, which resulted in early allograft RV dysfunction and severe TR, prompting resumption of PAH therapy. This suggest that continuation of intravenous prostaglandins must be considered past the intraoperative period and a very slow wean and transition to subcutaneous pump can be achieved over the next few weeks to months after OHT. Future case series and prospective studies are needed to confirm the efficacy of this approach, which if successful may significantly improve survival not only in the immediate postoperative period but also many years after given the better long-term prognosis offered by OHT when compared to combined heart and lung transplantation. Similarly, in patients whose pulmonary pressures do not fall in the short-term, consideration of subaortic ventricular assist device can be pursued given its demonstrated efficacy achieving lower transpulmonary gradients in nearly 82% of patients 9 months from implantation [7].

Disclosures

None. The patient granted consent for this publication.

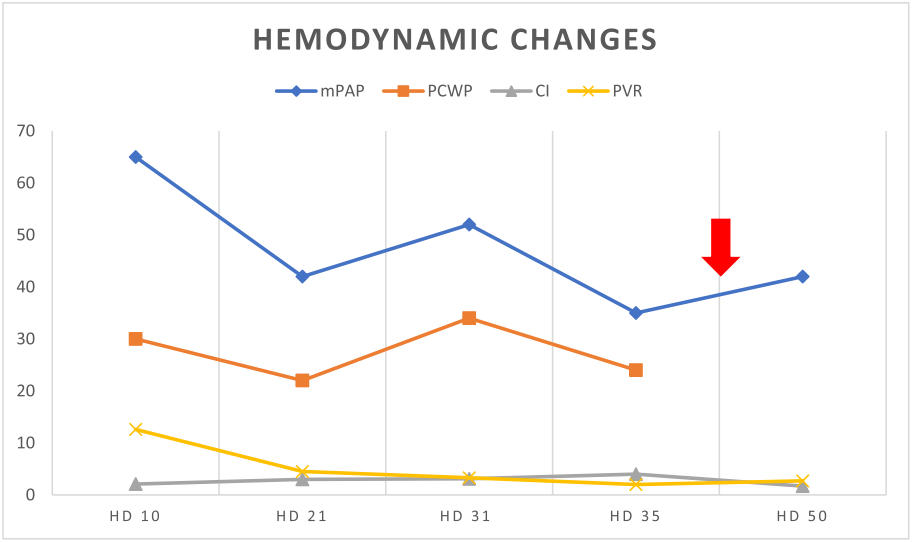

Fig. 1. Hemodynamic changes over time before and after OHT.
*OHT: orthotopic heart transplantation; HD: hospitalization day; mPAP: mean pulmonary artery pressure (mmHg); PCWP: pulmonary capillary wedge pressure (mmHg); CI: cardiac index (L/min/m^2); PVR: pulmonary vascular resistance (WU Woods units). Red arrow indicates day of orthotopic heart transplant (performed on HD 43).

Declaration of competing interest

None declared.

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References

[1] Brida M, Diller GP, Gatzoulis MA. Systemic right ventricle in adults with congenital heart disease: anatomic and phenotypic spectrum and current approach to management. Circulation 2018;137:508–18.
[2] Burchill LJ, Gao L, Kovacs AH, Opotowsky AR, Maxwell BG, Minnier J, Khan AM, Broberg CS. Hospitalization trends and health resource use for adult congenital heart disease-related heart failure. J Am Heart Assoc 2018;7:e008775.
[3] Zaragoza-Macias E, Zaidi AN, Dendukuri N, Marelli A. Medical therapy for systemic right ventricles: a systematic review (part 1) for the 2018 aha/acc guideline for the management of adults with congenital heart disease: a report of the american college of cardiology/american heart association task force on clinical practice guidelines. Circulation 2019;139:e801–13.
[4] Kim YY, Awh K, Acker M, Atluri P, Bermudez C, Crespo M, Diamond JM, Drajpuch D, Forde-Mclean R, Fuller S, Goldberg L, Mazurek J, Mascio C, Menachem JN, Rame E, Ruckdeschel E, Tobin L, Wald J. Pulmonary arterial hypertension in adults with systemic right ventricles referred for cardiac transplantation. Clin Transplant 2019;33:e13496.
[5] Menachem JN, Schlendorf KH, Mazurek JA, Bichell DP, Brinkley DM, Frischhertz BP, Mettler BA, Shah AS, Zalawadiya S, Book W, Lindenfeld J. Advanced heart failure in adults with congenital heart disease. JACC. Heart failure. 2020;8:87–99.
[6] Michel E, Orozco Hernandez E, Enter D, Monge M, Nakano J, Rich J, Anderson A, Backer C, McCarthy P, Pham D. Bridge to transplantation with long-term mechanical assist devices in adults with transposition of the great arteries. Artif Organs 2019;43:90–6.
[7] Roche SL, Crossland DS, Adachi I, Broda C, Jansen K, Hickey E. Mechanical circulatory support for the failing sub-aortic right ventricle in adults. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2021;24:2–9.