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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. Supplemenary data related to this article can be found at https://doi.org/10.1016/j.jjchd.2022.100361.

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/m² 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/m² 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 CardioMEMSTM 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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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.

Supplementary data related to this article can be found at https://doi.org/10.1016/j.ijcchd.2022.100361.

Patients with unpaillated 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].

**Table 1**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>HD 10</th>
<th>HD 21</th>
<th>HD 31</th>
<th>HD 35</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic venous baffle</td>
<td>14 mmHg</td>
<td>–</td>
<td>–</td>
<td>15 mmHg</td>
</tr>
<tr>
<td>Subpulmonary left ventricle</td>
<td>95/17 mmHg</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Pulmonary artery resistance</td>
<td>30 mmHg</td>
<td>22 mmHg</td>
<td>34 mmHg</td>
<td>24 mmHg</td>
</tr>
<tr>
<td>Pulmonary vascular resistance</td>
<td>103/17 mmHg</td>
<td>60/30/42 mmHg</td>
<td>88/37/52 mmHg</td>
<td>14 mmHg</td>
</tr>
<tr>
<td>Cardiac output</td>
<td>3.8 L/min</td>
<td>5.5 L/min</td>
<td>5.5 L/min</td>
<td>4 L/min</td>
</tr>
<tr>
<td>Cardiac index</td>
<td>2.1 L/min/m²</td>
<td>3 L/min/m²</td>
<td>3.1 L/min/m²</td>
<td>4 L/min/m²</td>
</tr>
<tr>
<td>Pulmonary vascular resistance</td>
<td>12.6 WU</td>
<td>4.5 WU</td>
<td>3.3 WU</td>
<td>2 WU</td>
</tr>
</tbody>
</table>

**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/m²); 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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None.

References


