

## CASE REPORT

# Transesophageal Echocardiography in a Case of Dextro-Transposition of Great Arteries with Regressed Left Ventricle

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## ABSTRACT

Patients with dextro-transposition of the great arteries (d-TGA) with an intact ventricular septum (IVS) beyond 21 days of age may develop left ventricular (LV) regression. Perioperative echocardiography-guided assessment of LV for signs of regression is crucial in decision-making for definitive corrective surgery, namely arterial switch operation (ASO), as a regressed LV may not be capable of sustaining the load of systemic circulation. We hereby present transesophageal echocardiography (TEE) findings in a child with d-TGA and regressed LV where decision to perform a primary ASO with integrated extracorporeal membrane oxygenation (ECMO)-cardiopulmonary bypass (CPB) circuit was made. Use of ECMO was planned as a standby technique in the event of LV failure. ECMO was eventually used as the LV was unable to sustain the load of systemic circulation after ASO.

**Keywords:** Dextro-transposition of great arteries, Arterial switch operation, Regressed left ventricle, Transesophageal echocardiography, Extracorporeal membrane oxygenation.

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**Conflict of interest:** None

## INTRODUCTION

Infants with dextro-transposition of the great arteries (d-TGA) with an intact ventricular septum (IVS), left ventricular (LV) regression is a critical issue as it has been accepted that the LV can sustain the load of systemic circulation after corrective surgery only up to 21 days of age.<sup>1</sup> Beyond this period, the LV begins to show signs of regression and is unable to sustain the systemic circulation. Controversy still exists

over the optimal age for a primary arterial switch operation (ASO) in such cases. According to recent reports, age for primary ASO can be extended up to 2 months of age.<sup>2,3</sup> We hereby present transesophageal echocardiography (TEE) findings in a child with d-TGA and regressed LV where decision to perform a primary ASO with integrated extracorporeal membrane oxygenation (ECMO)-cardiopulmonary bypass (CPB) circuit was made.<sup>4</sup> Use of ECMO was planned as a standby technique in the event of LV failure. ECMO was eventually used as the LV was unable to sustain the load of systemic circulation after ASO.

## CASE REPORT

A 4 kg, 2 months old male child (body surface area of 0.24 m<sup>2</sup>) presented with a history of difficulty in feeding and and bluish discoloration for 15 days. Trans-thoracic echocardiography (TTE) showed: situs solitus, atrioventricular concordance and ventriculoarterial discordance, systemic veins draining into the right atrium, pulmonary veins (PV) draining into the left atrium (LA), a small upper muscular ventricular septal defect (VSD) measuring 3.5 mm with restrictive right to left shunt (55 mm Hg gradient), a small patent ductus arteriosus (PDA) and sinus venosus atrial septal defect (SV-ASD) with a left to right shunt. All heart valves were normal. The right atrium (RA) and the right ventricle (RV) were enlarged along with right ventricular hypertrophy. Left ventricle (LV) showed signs of early regression in the form of a crescent-shaped LV and IVS moving synchronous with RV.

## Measurements

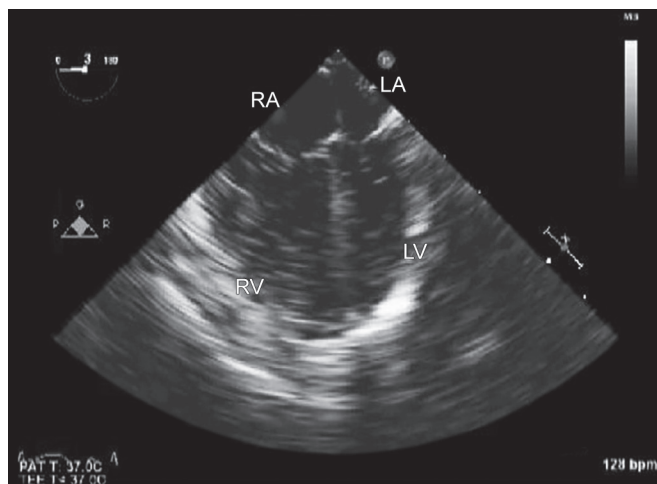
- LV had end-systolic (ES) and end-diastolic (ED) dimensions of 6 and 10 mm with a posterior wall thickness of 3.2 mm ( $\pm$ Z 2.6-4.2 mm) and an interventricular septum (IVS) measuring 3 mm ( $\pm$ Z 2.4-5.2 mm) at the end of diastole.
- Ejection fraction was 76% with normal biventricular function.

The final diagnosis of cyanotic congenital heart disease (CCHD), d-TGA, small restrictive VSD, SV-ASD, PDA and early LV regression was made, and the child

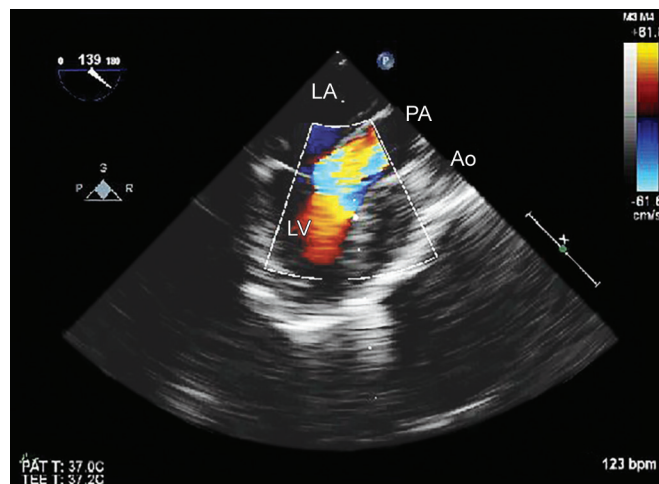
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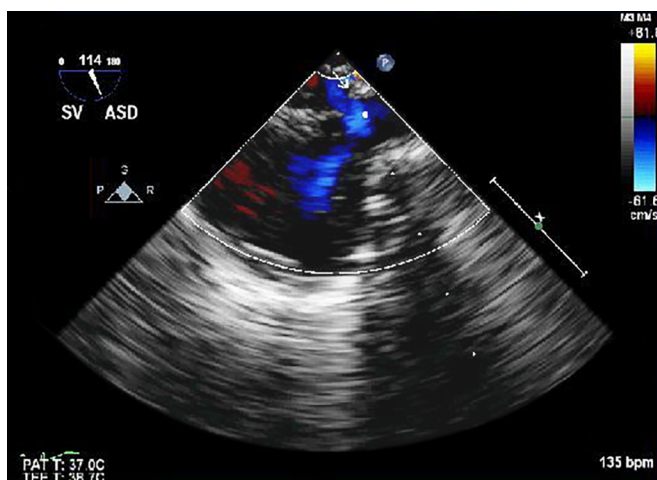
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**Fig. 1:** Midesophageal four-chamber view showing RV with apically displaced tricuspid valve with septal leaflet attachment, coarse trabeculations and moderator band (RA: Right atrium; RV: Right ventricle; LA: Left atrium; LV: Left ventricle)



**Fig. 2:** Midesophageal 120 to 140° long-axis view showing great vessels lying side by side and an adequately sized left ventricular outflow tract (PA: Pulmonary artery; Ao: Aorta; LA: Left atrium; LV: Left ventricle)



**Fig. 3:** The modified bicaval view with color flow Doppler showing the presence of SV ASD with left to right flow (SV-ASD: Sinus venosus atrial septal defect)

was subsequently planned for ASO, ASD and VSD closure with integrated ECMO-CPB support. The patient was taken in OT and after an uneventful induction of anesthesia, a neonatal TEE probe (S8-3t probe attached to Philips iE33x Matrix Echo machine, Bothell, WA, USA) was inserted, and findings of TTE were confirmed.

### Chamber Identification

On TEE, chamber identification and confirmation of the diagnosis was first done. While the central line was inserted from the femoral vein, the guide wire could be visualized entering from the bicaval view into a chamber, confirming it to be RA. The right ventricle was identified by the presence of more apically placed trileaflet atrioventricular valve, presence of moderator band and coarse trabeculations (Fig. 1, Video 1). The identification and spatial relation of the great vessels was then carried out using the midesophageal 120 to 140°

long-axis view which showed great vessels lying side by side in an anteroposterior relationship (Fig. 2, Video 2). The posteriorly placed vessel arising from the LV was then traced vertically upward and seen to bifurcate confirming it to be the pulmonary artery arising from the LV. Additionally, the application of color flow Doppler in the midesophageal aortic valve long-axis view showed an adequately sized left ventricular outflow tract (LVOT). There was no evidence of any LVOT obstruction due to accessory mitral tissue/membrane or bulging of the IVS. There was no insufficiency at the outlet valve of the left ventricle (Fig. 2, Video 2).

### Associated Abnormalities

After the confirmation of anatomical diagnosis, additional findings were confirmed. The probe was slightly rotated from the bicaval view to visualize the SV ASD (Fig. 3, Video 3) which had a left to right flow with a gradient of 2 mm Hg. Midesophageal four chamber view additionally showed a small mid muscular VSD measuring 0.296 cm (Fig. 4, Video 4).

### Assessment of the Ventricles

Assessment of the ventricles for their adequacy was then carried out. Mitral and tricuspid annuli were measured in the midesophageal four chamber view (Fig. 5). Application of color flow Doppler did not show any turbulence across the mitral valve and the LV cavity.

### Position and Movement of IVS

On transgastric view bulging of the IVS was seen into the LV causing it to become crescent/banana shaped (Fig. 6, Video 5). Additionally, the movement of the IVS was seen to be more synchronous with RV.

The ASO was done with an aortic cross clamp time of 101 minutes and CPB time of 240 minutes. TEE was then performed for assessment of LV function and surgical repair during weaning from CPB. But, patient was unable to sustain systemic pressures despite high inotropic support. Patient was subsequently supported on ECMO and shifted to ICU. The ECMO was weaned off after 40 hours and the child's trachea was extubated on post-operative day 7 and shifted to ward on postoperative day 14. Great care was taken during TEE examination and all manipulations were gentle so as to avoid probe-related complications in this infant. A written informed consent was obtained from the father for publishing the case report and the images and loops.

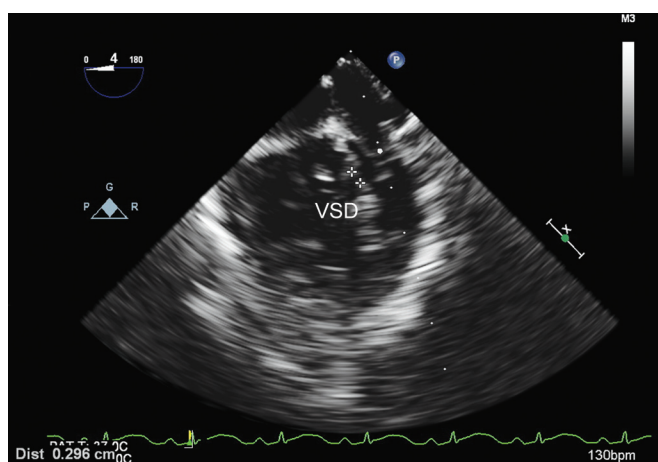
## DISCUSSION

Complete transposition of the great arteries is characterized by ventriculoarterial discordance, i.e. the morphological RV gives rise to aorta, and the morphological LV gives rise to the pulmonary artery. Because of this, systemic oxygenation depends upon

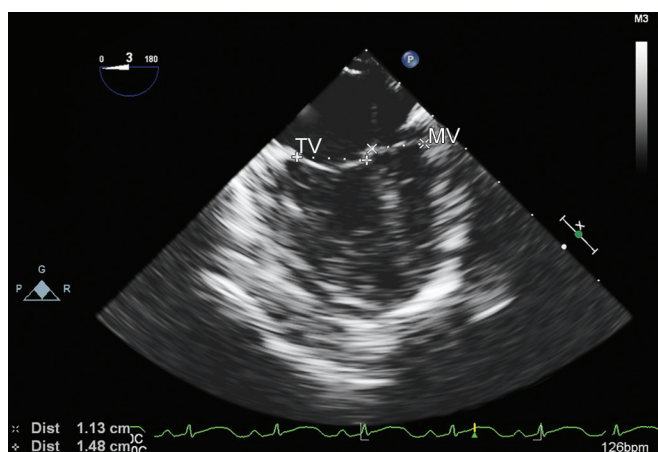
communications between the two circuits, either with an atrial septal defect, a ventricular septal defect (VSD), or at the great arterial level (patent ductus arteriosus).<sup>1</sup> However, in patients with an intact ventricular septum or a small restrictive VSD, fall in pulmonary vascular resistance following birth leads to a decreased afterload to the LV which may then begin to regress. In children with TGA and LV showing signs of regression corrective surgery in the form of ASO is associated with a high risk for LV failure because the regressed LV might be incapable of sustaining the acutely increased work of systemic perfusion.

Differential loading conditions on both the left and the right ventricle alter the geometry of the ventricles. The common wall between the two pumps formed by the interventricular septum undergoes alteration in position (spherical or D-shaped or banana-shaped) and motion (movement with LV or RV) depending upon the extent of decrease in afterload to LV. Left ventricle is considered to be 'prepared' to take up the load of systemic circulation after ASO if the geometry is maintained (spherical or D-shaped) along with motion of the IVS being synchronous with the LV. Left ventricle regression is characterized by an altered LV geometry (crescent/banana-shaped), and the septal motion synchronous with RV.<sup>3</sup> Other important echocardiographic tools for assessment of LV preparedness include LV mass index, LV posterior wall thickness, interventricular septal thickness and LV end diastolic internal diameter.<sup>5</sup>

In our case, after confirmation of the diagnosis, LV was the subsequently examined to assess its preparedness for ASO. The two-dimensional echocardiography showed a crescent/banana-shaped LV with bulging of IVS toward LV and movement of IVS synchronous with that of RV suggesting signs of LV regression. The small restrictive VSD present in our patient (0.29 cm) failed to provide any protection to the LV against the falling pulmonary



**Fig. 4:** Midesophageal four-chamber view showing midmuscular VSD 0.296 cm in size (VSD: Ventricular septal defect)



**Fig. 5:** Midesophageal four-chamber view showing measurements of the MV and TV annuli (MV: Mitral valve; TV: Tricuspid valve)



**Fig. 6:** Transgastric mid papillary short-axis view showing an enlarged and hypertrophied right ventricle with interventricular septum ballooning toward left ventricle



vascular resistance. However, the LV had end-systolic (ES) and end-diastolic (ED) dimensions of 6 and 10 mm respectively, with a posterior wall thickness of 3.2 mm ( $\pm$ Z 2.6-4.2 mm), and an interventricular septum (IVS) measuring 3 mm ( $\pm$ Z 2.4-5.2 mm) at the end of diastole. These values were adequate for the age and body surface area based on the Z values for the age of the child. Also, the mitral valve annulus measured 11.3 mm ( $\pm$ Z 10.9-11.5 mm) and tricuspid valve annulus was 14.8 mm ( $\pm$ Z 13.1-13.7 mm), both being appropriately sized for age and BSA and showed no regurgitation or obstruction to outflow. Thus, with these parameters, LV seemed adequate to support as a systemic ventricle and the decision to undertake an ASO was made with due risk.<sup>6</sup>

Arterial switch operation with the use of ECMO has demonstrated a better outcome in such infants with regressed LV in comparison to the rapid two-stage ASO.<sup>7</sup> With the use of integrated ECMO-CPB circuits, an early and elective initiation of ECMO can be provided without any delay during the episode of low cardiac output at the time of coming off CPB. Studies have shown a better survival (76%) with integrated ECMO-CPB circuit compared to when ECMO is organized either in the operation theater due to inability to wean from CPB or in the postoperative period (25%).<sup>4</sup>

So, echocardiography helps in crucial decision-making for the use of integrated ECMO-CPB circuit, which is costlier than the conventional CPB circuit. Intraoperative TEE also helps in decision-making in situations where there may have been a significant time lapse between the previous TTE and the surgery. Also, intraoperative TEE is useful in de-airing, assessment of surgical repair and LV function after weaning from CPB.

## CONCLUSION

Transesophageal echocardiography assessment in a case of d-TGA with regressed LV is essential for understanding the basic preoperative anatomy, pathophysiology and presence or absence of associated anomalies. It also helps in important decision-making on the use of integrated ECMO-CPB circuit. Moreover, intraoperatively it is helpful in de-airing as well as in post-CPB assessment of surgical repair and LV function.

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