Complete transposition of great arteries associated with total anomalous pulmonary venous connection: An unusual cause for early left ventricular myocardial mass regression

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ABSTRACT

A 24-day-old apparently asymptomatic neonate was found to have complete transposition of great arteries with small patent ductus arteriosus and restrictive patent foramen ovale. The neonate was found to have relatively high saturations (saturations = 88%) despite inadequate mixing communications. Echo findings were suggestive of significant dilatation of right atrium and right ventricle and left ventricular (LV) mass regression. Further echo interrogation revealed coexisting total anomalous pulmonary venous connection (TAPVC) as the cause of relatively high saturations and early LV mass regression. The patient was planned for follow-up and underwent successful Senning repair at the age of 8 months. Hemodynamics and echo findings of this association of TGA with TAPVC have been described in this case report.

Keywords: Left ventricular mass regression, pulse oximetry in newborn, total anomalous pulmonary venous connection, transposition of great arteries

INTRODUCTION

Transposition of great arteries (TGA) with intact ventricular septum (IVS) is a commonly treated congenital heart disease where surgery is performed within first 2–3 weeks of life before the regression of left ventricular (LV) mass. Various factors are known to influence early or late regression of LV mass in TGA. Reported factors promoting early regression include patent ductus arteriosus (PDA) closure and large atrial septal defect (ASD); which may be natural or postballoon atrial septoplasty. Commonly associated cardiac lesions with TGA are ASD, ventricular septal defect (VSD), PDA, pulmonary stenosis, and various coronary anomalies. TGA associated with partial or total anomalous pulmonary venous connections (TAPVCs) is uncommon.

We report a 24-day-old neonate having TGA and TAPVC with early LV mass regression to discuss hemodynamics and echocardiographic clues for diagnosis of such an unusual association.

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CASE REPORT

A 24-day-old male neonate weighing 3 kg presented with mild cyanosis. An echocardiographic diagnosis of TGA with IVS was made before transfer to our facility. On examination, he was active, alert, had mild cyanosis with room air oxygen saturation of 88%, and had no respiratory distress. Cardiovascular examination revealed a single second heart sound and no murmur. Echo revealed complete TGA, 4-mm secundum ASD shunting right to left, dilated right atrium (RA) and right ventricle (RV), small left atrium, and nonapex-forming LV with small mitral annulus (Z score – 4.5) and 1-mm

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PDA [Figures 1 and 2]. LV was regressed with posterior wall measuring 2.7 mm, LV mass of 18 g/m^2, and IVS bulging into LV ("banana shaped LV") [Figure 3]. The conspicuous absence of severe cyanosis in the setting of TGA/IVS and a restrictive PDA along with such early regression of LV prompted us to investigate further. Evaluation revealed that all four pulmonary veins formed a common chamber that drained through the vertical vein into the left innominate vein [Figure 4]. Computed tomography pulmonary angiogram confirmed the above echo findings [Figure 5]. The patient was advised follow-up in view of regressed LV and adequate saturations and underwent successful Senning repair at the age of 8 months.

DISCUSSION

Patients with TGA have good saturations when there is large communication for mixing of blood at atrial, ventricular, or ductal level. Arterial switch operation (ASO) is recommended early in TGA with IVS to avoid LV mass regression usually within first 2–3 weeks. After that period, the progressive drop in pulmonary vascular resistance deconditions the performance of the LV with progressive reduction in its muscle mass that might render the LV incapable of coping with the acutely increased work of systemic perfusion that occurs following ASO.

Exact time when the LV loses its capacity to sustain the systemic function has not been clearly defined, but various factors may influence it. LV mass remains preserved in cases of large VSD, large PDA, obstruction of LV outflow, unregressed PVR, or large aortopulmonary collateral, due to the presence of a higher afterload. In cases of large interatrial communication alone, LV mass is usually not well preserved after fall in neonatal PVR due to the absence of afterload and relatively less inflow to LV. Although the presence of TAPVC leads to better systemic saturations as oxygenated blood from pulmonary veins is being diverted to systemic circulation, it can lead to certain disadvantages too. First, the absence of marked or easily detectable cyanosis in a

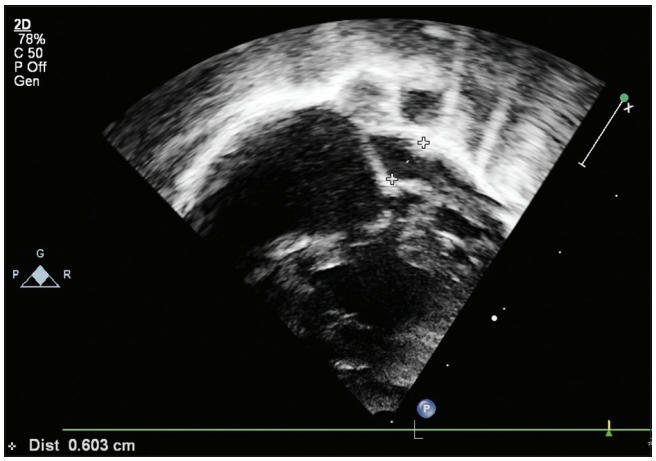

Figure 1: Apical 4-chamber view shows dilated right atrium and right ventricle, small mitral annulus along with small left atrium and left ventricular

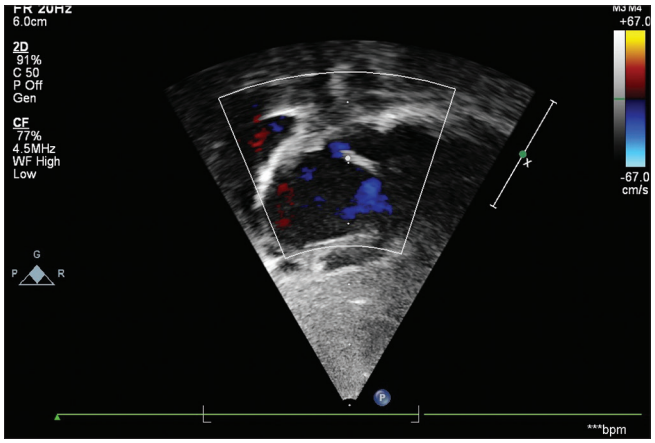

Figure 2: Subcostal view showing small secundum atrial septal defect shunting right to left

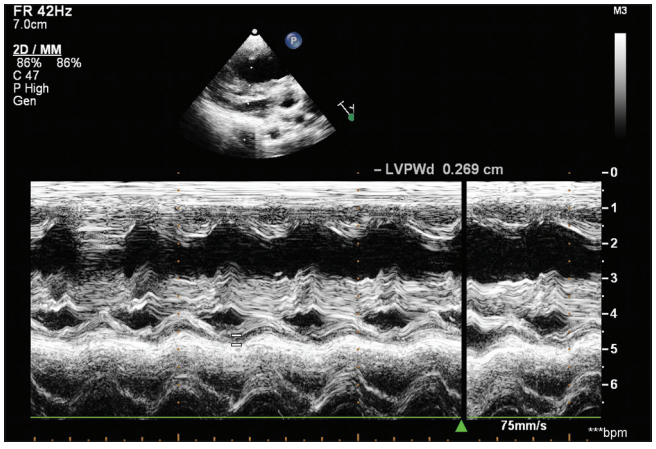

Figure 3: Parasternal long-axis view showing posterior wall thickness in M-mode

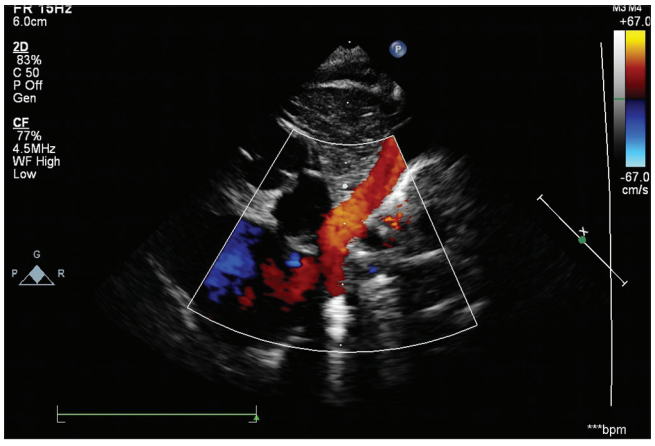

Figure 4: High parasternal short-axis view showing vertical vein draining into the left innominate vein along with d malposed great vessels

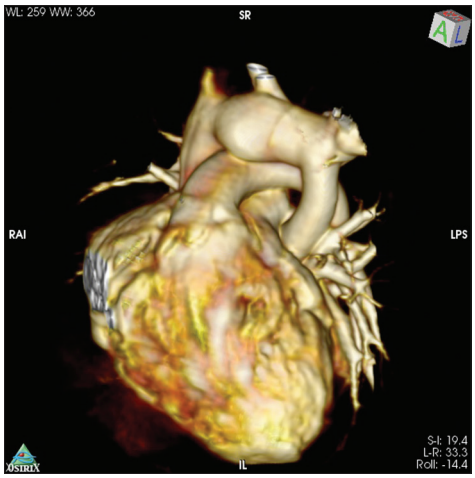

Figure 5: Computed tomography angiogram with three-dimensional reconstruction showing vertical vein drainage to left innominate vein and anterior aorta

reasonably asymptomatic neonate masks detection of a serious underlying congenital cardiac lesion. Second, it leads to very early regression of LV mass due to relatively less amount of LV inflow from times as early as the fetal life. However, it may be noted that obstructed TAPVC will lead to elevated pulmonary arterial pressures and that may be advantageous for LV preparedness.

In the literature, a few authors have reported the successful outcomes with modified techniques of atrial switch based on principles of either the Senning or Mustard.[1-4] One of the studies reports that the ASO, which is the first choice for the TGA, is contraindicated when an association with TAPVC coexists because the left ventricle is unsuitable to tackle systemic workload as it regresses from very early fetal unloaded conditioning.[3] Very few cases have been reported to have successful anatomic repair. Lopes *et al.* reported successful ASO with TAPVC repair in a 22-day-old child having TGA with infracardiac TAPVC.[5] Recently, 2 more case reports of successful anatomic repair were reported in neonates.[6,7] In our case, we preferred to do atrial switch due to significantly small mitral valve annulus though some centers may still do ASO with extracorporeal membrane oxygenation support. In conclusion, complete TGA and TAPVC is a rare combination and should be suspected in cases with early LV regression along with dilated RA and RV and also in neonates presenting with high saturations with small inadequate mixing communications. Management of such an association will be either atrial switch or arterial switch depending on the preference of center.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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