Transposition of the great arteries with total anomalous pulmonary venous connection in a 1½-year-old child: Pulmonary arterial hypertension – An advantage

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ABSTRACT

Transposition of the great arteries with total anomalous pulmonary venous connection is a rare anomaly with varied management approach depending on the time of presentation; the management of cases which present early is usually by anatomical surgical approach while late presentation is by physiologic approach. This is due to early left ventricular mass regression as a result of the absence of volume and pressure load to the left ventricle (LV). We report a late presentation (at 1½ years of age) that had a successful anatomic surgical correction because the LV was "prepared" by both pressure and volume load from pulmonary arterial hypertension and large ostium secundum atrial septal defect.

Keywords: Anatomic correction, pulmonary arterial hypertension, total anomalous pulmonary venous connection, transposition of the great arteries

INTRODUCTION

The earliest write-up on Transposition of the great arteries (d-TGA) with total anomalous pulmonary venous connection (TAPVC) in the literature was an anatomic description made in 1933.[1] It was decades later that the report of a 16-year-old female who had a successful Mustard operation for the anomaly appeared.[2] Both anomalies have high propensity for developing pulmonary arterial hypertension early but through separate mechanisms. In d-TGA, the process starts *in utero* with the preferential channeling of oxygen-rich inferior vena cava blood into the left heart through the foramen ovale, thus causing dilatation of pulmonary vessels and stretch injury of the vascular intima with subsequent development of secondary vasoconstriction. Whereas, in obstructed TAPVC, the increase in pulmonary pressures results from pulmonary venous hypertension and its severity is dependent on degree of obstruction. There is amelioration of the hemodynamic consequences of the individual anomaly in the setting of their coexistence, making early diagnosis to be sometimes missed.

To the best of our knowledge, there is no prior report of a successful anatomic correction of d-TGA beyond the neonatal period in the setting of d-TGA coexisting with TAPVC.

CASE REPORT

We report a case of 1½-year-old female who was diagnosed at 1 month of life with congenital heart

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_65_20

How to cite this article: Abah RO, Prabhu A, Katewa A, Sahu B. Transposition of the great arteries with total anomalous pulmonary venous connection in a 1½-year-old child: Pulmonary arterial hypertension - An advantage. Ann Pediatr Card 2021;14:235-8.

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Submitted: 08-Apr-2020 Revised: 21-May-2020 **Accepted:** 26-Dec-2020 **Published:** 16-Feb-2021

disease following episode of lower respiratory tract infection (LRTI). However, parents were financially constrained and the patient could not get the required surgical intervention till she presented at our facility at 1½ years of age. She had several episodes of LRTI and was on diuretics (furosemide and spironolactone) prior to her presentation at our facility. Her birth weight was 1.9 kg, and she was the first child of a nonconsanguineous marriage. On examination, she was small for age with a weight of 5 kg and length of 64 cm, dusky with SPO_2 of 85%, and tachypneic with a respiratory rate of 30 breaths/min. Her pulse rate was 136 beats/min, apex beat was displaced to 5th left intercostal space midclavicular line, and the heart sounds were SI, SII with a soft systolic murmur. Chest X-ray showed cardiomegaly with a cardiothoracic ratio of 0.7 and plethoric lung fields [Figure 1].

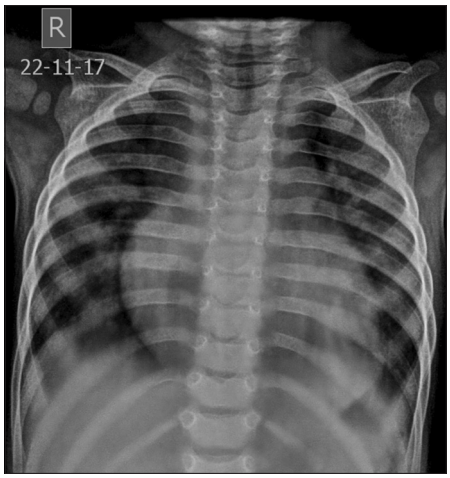

Figure 1: Chest X-ray image

Echocardiographic examination showed situs solitus, levocardia, atrioventricular concordance, ventriculoarterial discordance, dilated right atrium (RA) and right ventricle (RV), large ostium secundum atrial septal defect (OS ASD) with right to left shunt, moderate to severe tricuspid regurgitation (TR), flattened but intact interventricular septum, D-shaped left ventricle (LV), d-TGA, TAPVC with all four pulmonary veins forming a common chamber behind left atrium (LA), and draining via a short right vertical vein to just above the superior vena cava right atrial (SVC-RA) junction. There was flow turbulence at its insertion point with a continuous wave Doppler peak gradient of 20 mmHg and mean of 10 mmHg. The biventricular contractility was good, with a LV mass of 58.6 g/m^2. The diagnosis of d-TGA with obstructed supracardiac TAPVC was thus made [Figure 2].

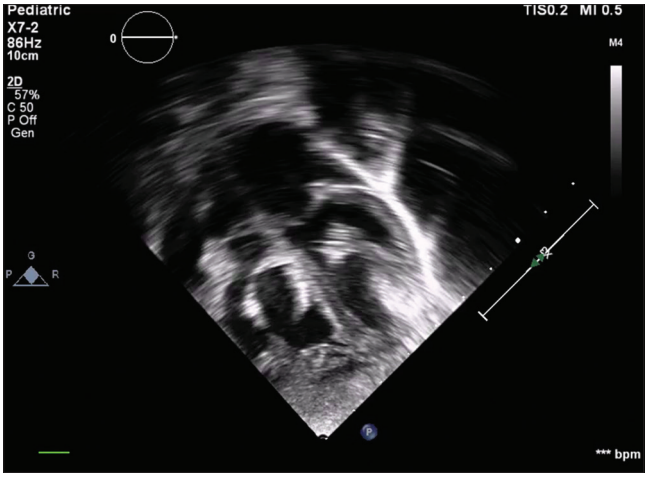

Figure 2: Echocardiographic image (two-dimensional) from the subcostal view showing the ventricles

Intraoperatively, the diagnosis was confirmed with the findings of d-TGA; aorta right and anterior, a main pulmonary artery that was thrice the size of the aorta, supracardiac TAPVC draining via a short right vertical vein to the lower end of the SVC; obstruction was noted to be due to vascular vise between the right pulmonary artery and the right bronchus; large OS ASD and annular dilatation of the tricuspid valve with moderate regurgitation. The systolic pulmonary arterial pressure was 62 mmHg, with a corresponding systemic arterial pressure of 124/85 mmHg [Figure 3].

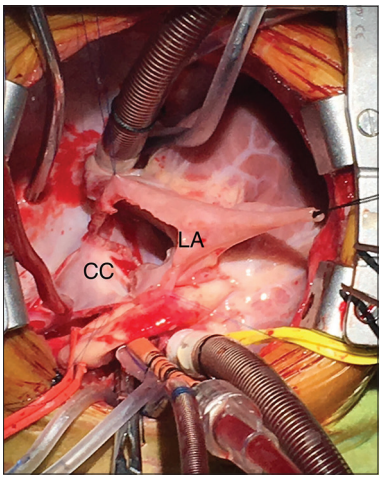

Figure 3: Intraoperative image showing the common chamber and the left atrium

The patient subsequently had an arterial switch operation (ASO) with TAPVC (vertical vein was ligated and anastomosis was created between the common chamber and the LA), ASD, and tricuspid valve repair. Sternum was left open electively throughout postoperative day (POD) 0 but was closed on POD1. The pulmonary hypertension was managed in the immediate post-operative period by keeping the patient sedated and ventilated for 48 hrs along with administration of inhaled Nitroglycerin. Oral Sildenafil and Bosentan were added to the regimen on POD1 and POD2 respectively. She did well postoperatively and was discharged on POD10. Echocardiography evaluation prior to discharge

showed unobstructed pulmonary venous return to LA, well-flowing LeCompte, trivial neo-PR, mild MR, moderate TR with a peak gradient of 30 mmHg, and Grade 1 RV diastolic dysfunction.

One month postoperatively, at her first follow-up visit, she weighed 6.15 kg, with a length of 69 cm and SPO_2 of 98%. Moreover, at 1-year postoperative follow-up visit, she was 8.0 kg in weight, 78 cm in length, and had 100% peripheral oxygen saturation on pulse oximetry.

Echocardiographic finding at 1-year postoperative follow-up visit was trivial TR with no evidence of pulmonary arterial hypertension.

DISCUSSION

The co-existence of d-TGA with TAPVC tends to mask/ameliorate the hemodynamic consequences of each of the individual anomaly and may result in delayed diagnosis.[1,3-5] Cyanosis in isolated d-TGA is usually marked and present at birth, but when d-TGA is in association with TAPVC, the oxygen-rich pulmonary venous blood is channeled into the RA and subsequently through the aorta to systemic circulation; thus, cyanosis is less and may go unnoticed at birth. This was the case in the index patient where diagnosis was made at 1 month of life when she developed LRTI. Furthermore, with isolated obstructed TAPVC, the patient can present from within few hours of birth to early infancy with cyanosis and increased respiratory effort depending on severity of the obstruction. The index patient had a moderate dynamic obstruction to the pulmonary venous flow which coupled with the coexisting d-TGA favored presentation in infancy.

For isolated d-TGA, outcome is bleak without surgical intervention, with an overall mortality of 30% in the 1st week of life, 50% in the 1st month, and 90% in the 1st year.[6] A single-stage anatomic correction of d-TGA, which is the procedure of choice, is best when performed within the first 3 weeks of life and not beyond 8 weeks[7,8] to prevent the regression of the LV mass that tends to be inevitable beyond this period.

Decision on the best treatment option for a patient with TGA and TAPVC is a major determinant of the outcome. For the index patient, the decision to offer her anatomic correction of the TGA with repair of the associated defects was seen as the most appropriate given the preserved LV mass. The preservation of the LV mass was most likely due to the pressure load on it by the presence of the pulmonary arterial hypertension and volume load from the large right to left shunt at the atrial level from the presence of the large OS ASD. Although Gontijo *et al.* had suggested that the occurrence of TAPVC in association with TGA should be seen as a contraindication for ASO,[9] it was evident that this was not the case. Moreover, there had been earlier reports of successful ASO in patients with d-TGA and TAPVC, however, those were in much younger patients of 5-h, 6-day, and 22-day-old babies, respectively.[5,10]

Second, the decision for the chosen surgical approach was also supported by the clinical, radiological, and echocardiographic features which all pointed to the likelihood of the absence of irreversible pulmonary vascular disease despite the presence of pulmonary arterial hypertension. Based on the American Heart Association and American Thoracic Society guidelines for pulmonary hypertension, she was on the lower risk end of the spectrum of pulmonary arterial hypertension severity.[11] The dynamic nature of the pulmonary venous obstruction could be the reason why irreversible pulmonary vascular disease had not yet set in.

The presence of the pulmonary arterial hypertension means that the patient had greater perioperative risk that would require proactive and aggressive measures to ensure good outcome. Hence, postoperatively, ventilation was continued for 48 h in order to reduce the effect of hypercarbia on pulmonary vascular resistance (PVR) as well as sympathetic outflow that could cause an increase in PVR. In addition, administration of inhaled nitroglycerin was commenced to provide targeted effect on pulmonary vasculature without causing any significant reduction in systemic vascular resistance that could jeopardize the delivery of adequate preload to the heart. The patient's growth and cardiac indices noted during the follow-up visits were evidence of improvement and general well-being.

This case has shown that even in the rare case of d-TGA coexisting with TAPVC, ASO is possible beyond the neonatal period, so an individualistic approach is advocated rather than one size fits all in management. This could be considered as one occasion where the presence of pulmonary arterial hypertension was beneficial as it kept the LV prepared yet without causing irreversible pulmonary vascular disease. Moreover, with proactive and aggressive postoperative management, a good outcome was achieved.

Consent

Appropriate permission was sought from and granted by parents.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for their child's images and other clinical information to be reported in the journal. The parents understand that the name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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