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Review Article

Bowled over by posterior Transposition

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ABSTRACT

Background: Transposition of Great arteries (TGA) with posteriorly placed aorta is a rare entity with an incidence of less than 1%. Van Praagh et al. had first described 4 cases with posterior transposition. With 25 cases of this entity reported from all over the world and no case reported from Indian subcontinent, we are reporting a case of 2-month-old male child with TGA, subaortic ventricular septal defect (VSD), posteriorly placed aorta (P) and pulmonary arterial hypertension (PAH).

Case Report: A 2-month-old male child, presented with cyanosis and fast breathing. The echocardiography revealed TGA, moderate subarterial VSD, multiple muscular VSDs, posteriorly placed aorta with aortomitral continuity and PAH. Arterial switch operation was done without the Le Compte procedure and septal defects were closed by trans aortic approach.

Conclusion: We conclude that TGA, with posteriorly placed aorta is an anatomic entity with anecdotal case reports. The diagnosis of this entity in fetal life and preoperative echocardiography is important for diagnosis and surgical approach.

INTRODUCTION

The hallmark anatomic feature of transposition of great arteries is the “ventriculoarterial discordance”. The most common anatomic landmark in complete transposition of great arteries (TGA) with D looped ventricle is aorta arising from the right ventricle (RV) placed anteriorly and the pulmonary artery from the left ventricle (LV) seen posterior to the aorta [1]. Complete Transposition with posterior orientation of aorta is a rare entity with an incidence of less than 1% [2]. Van Praagh et al. have reported 4 cases of transposition with ventricular septal defect (VSD), posterior aorta, anterior pulmonary artery, subpulmonic conus and fibrous continuity between aortic and atrioventricular valves after autopsy [2].

Here, we describe a patient whose echocardiogram revealed TGA with subaortic VSD, multiple lower muscular septal defects, posteriorly placed aorta with aortomitral continuity, anterior pulmonary artery and severe pulmonary arterial hypertension (PAH). These findings were confirmed intraoperatively. To our knowledge, very few cases with posterior aorta have been reported worldwide and echocardiographic information is rare.

CASE PRESENTATION

A 2-month-old male child, weighing 3.12 kg, presented with cyanosis and fast breathing. On examination, there was central cyanosis, saturation 82%, heart rate 120/min, tachypnoea with a rate of 48/min and a systolic blood pressure of 80mmhg in the left upper arm. The cardiovascular exam revealed a single, accentuated second heart sound and a grade 2/6 ejection systolic murmur. Breath sounds were normal. There was no organomegaly on abdominal examination. Chest x-ray showed cardiomegaly with increased pulmonary vascularity. Electrocardiogram showed sinus rhythm, QRS axis of +120, right dominant biventricular hypertrophy. The echocardiography was done on GE VIVID T -8 machine using a 6MHz scan head. The four-chamber view showed both the ventricles to be dilated (Figure 1a). There was no significant atrioventricular valves regurgitation. There was a restricted patent foramen ovale with a left to right flow across it. On cranial sweep across the four-chamber view, there was a VSD in the peri membranous area and the first artery visible from left ventricle was the pulmonary artery (Figure 1b), with its left and right branches followed by the aorta arising from the RV (Figure 1c). There were multiple lower muscular septal defects. The parasternal short axis view showed the great arteries as two circles, with the aorta being posterior and right of the pulmonary artery. Both the arterial valves were visible in diastole and the branching of the pulmonary artery was shown in one intercoastal space upwards. The left parasternal view showed the aorta arising medially from the RV, while the pulmonary artery arose from the LV. A characteristic point, an aorto-mitral continuity was noted but there was no pulmonary mitral continuity. The aorta was overriding the VSD. The subcoastal out flow tract view showed aorta from RV and the pulmonary artery from the LV along with multiple muscular VSD, a sub pulmonic conal tissue was present, however there was no sub aortic conus (5). The coronaries appeared to arise from the anterior and posterior sinus. The LV was preserved due to severe PAH. Left ventricular free wall thickness was 4.2mm with a left ventricular mass/ volume ratio of 1.6. We ordered for a computerised tomography (CT) scan of the chest for coronary artery anatomy and branch pulmonary artery anatomy. The findings coincided with the echocardiography. With these data we planned a complete surgical correction on the following day. The pre surgical investigations were within normal limits. The echo findings were confirmed intraoperatively. The membranous septal defect was closed off by trans aortic route. Multiple lower muscular VSDs were also closed off as much as possible. Le Compte manoeuvre was not required. Unfortunately, we lost the patient intraoperatively.

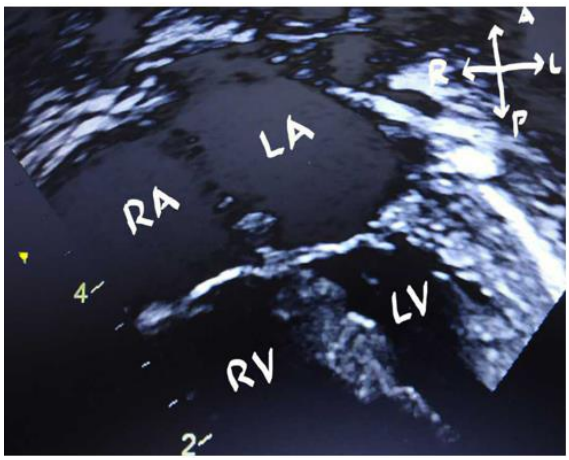

Figure 1a: Showing the four chambers view right atrium (RA), right ventricle (RV), left atrium (LA), left ventricle (LV).

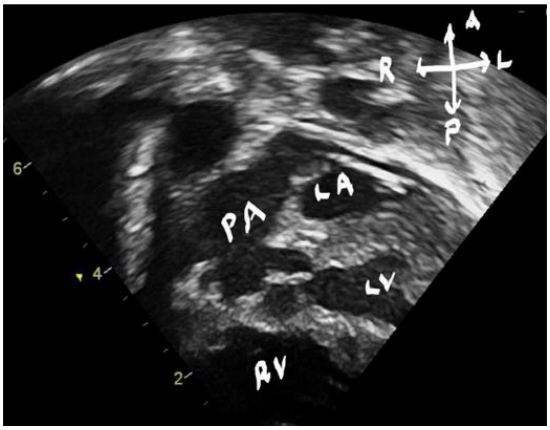

Figure 1b: Showing the five-chamber view with pulmonary artery (PA) arising from the left ventricle (LV)

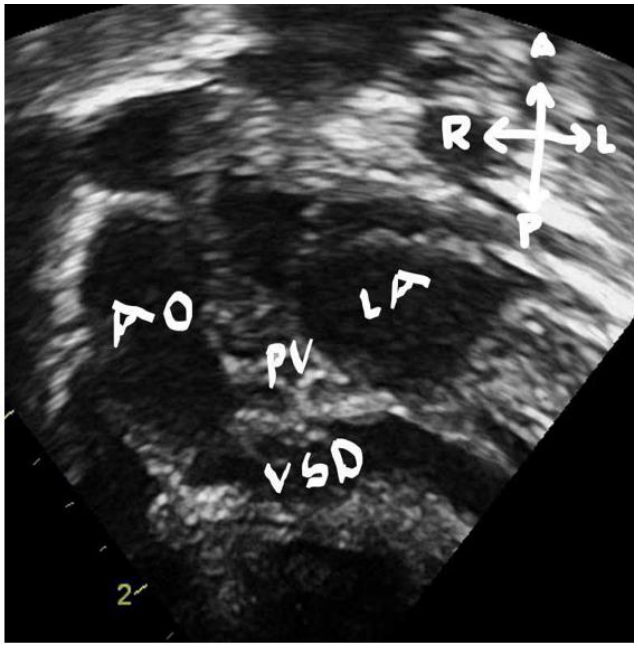

Figure 1c: Showing aorta arising from the right ventricle. The VSD in the peri membranous area is also seen.

DISCUSSION

The echocardiographic diagnosis of a complete transposition of aorta (S, D, D) in a situs solitus heart is made by aorta from right sided RV and pulmonary artery from left sided LV with parallel orientation of the great arteries. Morphologically this accounts for dextro transposition of the aorta in majority of the cases and anterior- posterior or levo- malposition in a few minorities [2]. However the posterior location of the aorta as in our case is quiet rare in literature with a handful of case reports. Van Praagh et al. were the first to report four cases of similar undocumented anatomy diagnosed during post mortem examination. He had termed it as "transposition of the great arteries with posterior aorta, sub pulmonary conus, and fibrous continuity between aortic and atrio-ventricular valves" [2]. Wilkinson et al described similar findings in six of their patients and they were the first to quote the term 'p transposition'. They had also described in detail the anatomic and possible embryological origin of this entity [3]. Marin-Garcia et al demonstrated the similar findings in angiograph [4]. The morphogenetic explanation for this case as described by Van Praag et al. is that the left sided subpulmonic conus developed, but remained much shorter [2]. The pulmonary valve was elevated, but to a subnormal level and was protruded anteriorly but not high enough to rise above the RV. The aortic valve remained depressed inferiorly and posteriorly towards the mitral and left ventricle but to a lesser degree than normal. Development of the subaortic conal musculature is deficient resulting in aortic atrioventricular valve continuity.

We diagnosed this unusual arterial relationship in this case of transposition on echocardiography. The coronary anatomy was confirmed on CT scan of the chest. The unusual arterial relationship was not much discernible on CT. We discussed the case in detail with our surgical team and planned for arterial switch operation. In our case we made a detail description of the anatomy on 2D echo and was confirmed intraoperatively. Some of the salient points noted in the echocardiography are-

1. The absence of pulmonary- mitral continuity (visible in parasternal long axis view and subcoastal coronal view).
2. The aortic and mitral continuity along with a VSD (seen in parasternal long axis view).
3. The posterior location of the aortic valve as against the pulmonary valve in parasternal short axis view.
4. The left sided subpulmonic conus developed, but inadequately causing it to move anteriorly just above LV.

CONCLUSION

TGA, with posterior aorta is a rare entity with anecdotal case reports. We are reporting this case as a reminder that this unique position of great arteries in transposition is of great clinical importance. This anatomic entity is to be noted carefully in this advancing world of fetal echocardiography. The out-flow tract view may reveal the ventriculoarterial discordance, however the short axis view or three vessel view may be misleading showing normal arrangement of the great arteries. The knowledge of morphologic existence of such an entity is essential for accurate perinatal diagnosis. From the surgical point of view, it is also of significance as the Le Compte may

not be required during arterial switch due to the orientation of the naturally corrected orientation of the right pulmonary artery and aorta.

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