Cardiology in the Young

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A unique phenomenon: triple-chambered left ventricle with transposition of great arteries

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Abstract

A rare case of unique and previously unreported variant of transposition of great arteries with bilateral coni, ventricular septal defect, and triple-chambered left ventricle which led to an unexpected and accelerated post-natal cardiac anatomical and physiologic deviation from prenatal and immediate postnatal diagnosis altering the surgical management and patient outcome.

Brief Report

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Case

A premature (35 3/7 weeks; birthweight 2030 g) male infant born with prenatal diagnosis and postnatal confirmation of atrioventricular concordance and ventriculo-arterial discordance, aorta anterior and rightward along with an unusual finding of bilateral coni, and a 4 mm ventricular septal defect that was only partly committed to the aorta. Both the right and left ventricular outflow tracts were widely patent without any obstruction. Pulmonary veins drained normally to left atrium, and patient had two balloon atrial septostomies (day of life 1 and 10) for frequent desaturations below 70% with a plan to allow weight gain and surgical repair at weight around 2.8 kg. Repeat echocardiogram at 2 weeks of age for a louder systolic murmur showed large (6.5 mm) atrial septal defect along with a new finding of prominent left ventricle septal muscle bundle (Fig 1) at mid-cavity level, creating a double chambered left ventricle. The muscle bundle appeared to originate from left ventricle free wall, separate from papillary muscle or attachment to the mitral valve. Though left ventricle apex looked hypertrabeculated, it did not fulfill criteria for definite diagnosis of left ventricle non-compaction. Additionally, the sub-pulmonary conus was noted be severely hypertrophied and elongated, further dividing the distal chamber into two halves resulting in a 3 chambered left ventricle and moderate to severe sub-pulmonary stenosis contributing to worsening cyanosis. These anatomical findings were not present on earlier echocardiograms except for much less prominent septal muscle band noted retrospectively.

A right-sided modified Blalock–Taussig shunt with 3.5 mm heparin-coated Gore-Tex tube graft at 21 days of life improved the oxygen saturation (SaO2). Discharge echocardiogram revealed adequate antegrade flow across the pulmonary valve despite sub-pulmonary stenosis and infant was growing on enteral feeds with SaO2 between 75% and 82% in room air. Over the next month his SaO2 gradually reduced to 60 s as the antegrade pulmonary flow almost abolished because of rapidly progressive massive muscular hypertrophy, requiring balloon dilatation of the Blalock–Taussig shunt and branch pulmonary arteries at 3 months of age.

Few weeks later, he again developed marked cyanosis, echocardiogram showed persistent pulmonary artery stenosis to the left of the Blalock–Taussig shunt. Subsequently, he underwent redo sternotomy for right pulmonary artery (RPA) plasty (Cardiocel patch), ligation of modified Blalock–Taussig shunt, and placement of Laks modification shunt with 4 mm Gore-Tex graft for single ventricle palliation. Because of progressive oxygen desaturation, repeat echocardiogram revealed a new finding of left-sided pulmonary veins stenosis and markedly reduced flow into the left pulmonary artery (LPA). The pulmonary venous stenosis progressed rapidly to atresia of left upper pulmonary vein and near atresia of the left lower pulmonary vein as confirmed on CT angiogram (Fig 1, Video 1). It became increasing difficult to ventilate and oxygenate him despite very high ventilatory support and 100% oxygen supplementation. Doppler interrogation of the Laks shunt suggested severe pulmonary hypertension. Shared decision of the multidisciplinary team and parents concluded that patient was unsuitable for pulmonary vasodilator therapy, mechanical cardiac support, or surgical palliation. He expired at 4 and ½ months of age.

Discussion

To our knowledge, this constellation of congenital cardiac malformations is unique and have not been reported. In usual dextro-transposition of great arteries, the sub-pulmonary conus is either completely resorbed or markedly underdeveloped resulting in fibrous continuity of the mitral

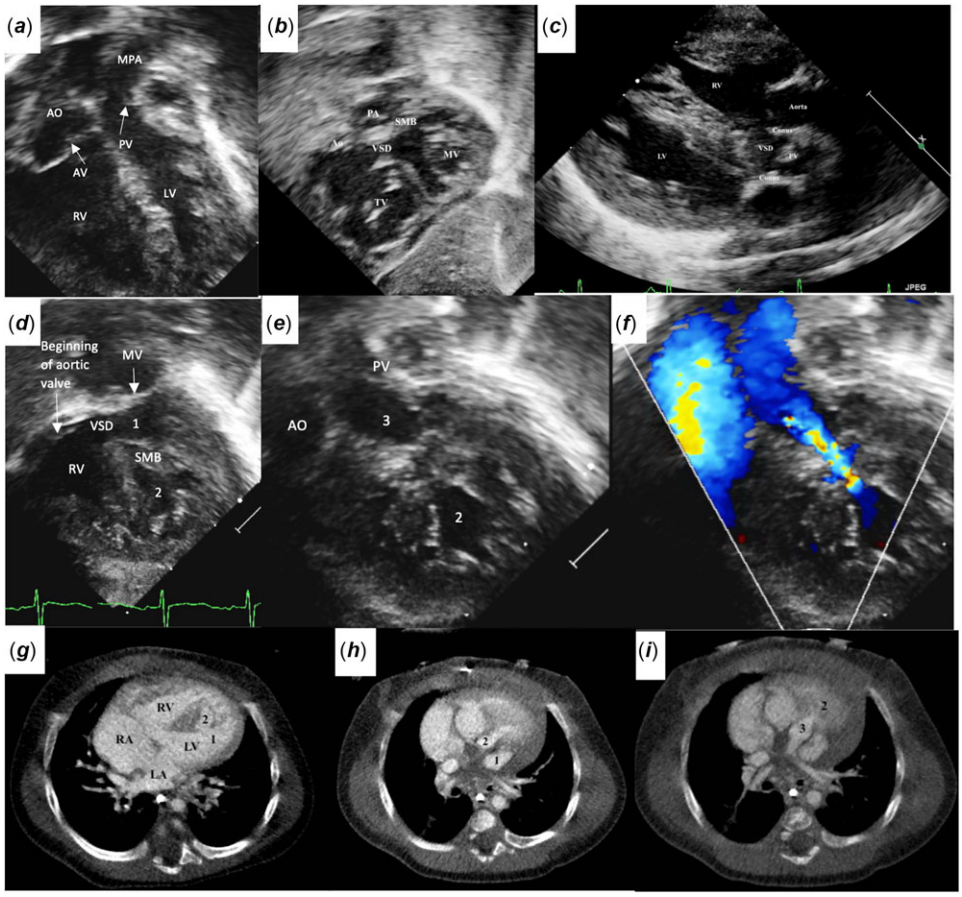

Figure 1. Two-dimensional echocardiogram and color Doppler views (***a***–***f***) showing moderate ventricular septal defect (VSD), D-transposition of great arteries (d-TGA) with bilateral coni, severe sub valvar pulmonary stenosis, and prominent septal muscle bundles (SMB) dividing left ventricle (LV) into three chambers numbered 1 to 3 from inlet towards outlet. CT (***g***–***i***) showing three-chambered left ventricle (LV) with transposition of great arteries; LA left atrium, 1 first chamber, 2 second chamber, and 3 third chamber, RV right ventricle, MV mitral valve, AO aorta, AV aortic valve, PA pulmonary artery, PV pulmonary valve.

and pulmonary valve. Moreover, left ventricular outflow tract as well as main pulmonary artery lie posterior to the aorta as expected in dextro-transposition.[1–3] In other words, in this case despite bilateral coni, patient did not have double outlet right ventricle which was unusual but had no impact on surgical management up to this point. The other anatomical aspect was abnormal left ventricle muscle mass, thick left ventricle septal muscle bundle connecting the left ventricle free wall with the ventricular septum across the left ventricle inlet segment, and a rapidly progressive severe muscular hypertrophy of the elongated sub-pulmonary conus, the later made it impossible to create an unobstructed or minimally obstructed left ventricular outflow tract and establish a normal biventricular circulation which seemed very likely based on the information from prenatal and early post-natal echocardiograms. These abnormal muscle bundles ultimately seemed to divide the left ventricle into three chambers, and the amount of muscle resection required to achieve unobstructed left ventricular outflow tract very likely would have resulted in a non-functioning left ventricle. Furthermore, small and relatively remote ventricular septal defect situated more in the inlet area with extension to the outlet segment was only partly committed to the sub-aortic region. Therefore, a Rastelli-type repair would have involved ventricular septal defect enlargement, right ventriculostomy, and right ventricle to pulmonary artery conduit insertion. Enlarging the ventricular septal defect, tunneling the ventricular septal defect to aorta would have also compromised the right ventricle volume. Finally, left-sided pulmonary venous atresia and stenosis resulting in severe pulmonary hypertension ruled out the option of single ventricle palliation and demise of the patient.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122002736

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