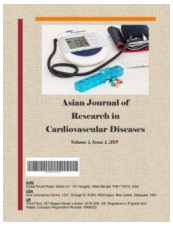

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Congenitally Corrected Transposition of Great Arteries Associated with Interrupted Aortic Arch Type A, Complex and Rare Anatomy Detected by Echocardiography: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case study

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ABSTRACT

We report an extremely rare case of congenitally corrected transposition of great arteries (CCTGA) (Pic. 1) associated with interrupted aortic arch (IAA) type A (Pic. 2) and a ventricular septal defect (VSD), in a preterm infant born at 36 weeks of gestation with a birth weight of 2.5 kg. This baby was diagnosed in utero with a large VSD and severe coarctation of the aorta. A comprehensive echocardiographic examination confirmed the diagnosis and revealed that the congenitally corrected transposition of great arteries was associated with Interrupted aortic arch type A. A review of the literature revealed only one similar case, reported earlier by Cottrell, et al. [1]. In their case, the initial diagnosis was confirmed angiographically as 2D and color Doppler echocardiography were not available in their center. The clarity of echocardiographic images obtained in our institution enabled us to make this diagnosis without needing additional imaging modalities. The infant was managed successfully as he underwent an aortic arch repair using an end-to-end anastomosis and a pulmonary artery (PA) band placement as a palliation towards second stage repair.

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Keywords: Interrupted aortic arch; corrected transposition; ventricular septal defect; Echocardiography.

1. INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) is a rare complex congenital cardiac malformation, which can present with or without associated anomalies [2]. In 1875 Rokistansky V. was the first to describe this defect [3]. In a normal heart, the left ventricle delivers blood to the body and the right ventricle, to the lungs. In congenitally corrected transposition of the great arteries the great arteries are transposed due to ventricular inversion. This occurs when cardiac looping occurs to the left during fetal cardiac development of the heart [4]. This results in both atrioventricular and ventriculoarterial discordance: the right atrium is connected to the left ventricle, supplying blood to the pulmonary artery, whereas the left atrium is connected to the right ventricle, supplying blood to the aorta [5]. It is a rare defect with a prevalence rate of 0.03 per 1 000 live births and makes up for 0.5 to 1% of the congenital cardiac diseases [6,7]. The defect has a slight male predominance [8]. (Figs. 1,2).

In interrupted aortic arch (IAA) there is discontinuity between the ascending and the descending aorta and it is of 3 types: in type A, the interruption occurs distal to the left subclavian artery, in type B the interruption occurs between the innominate artery and left carotid artery [3]. Usually, IAA is associated with other congenital heart diseases like ventricular septal defect (VSD), persistent ductus arteriosus (PDA), and atrial septal defect (ASD) [7]. About 90% of patients with CCTGA are reported to have other cardiac defects like ventricular septal defects, pulmonary artery stenosis and tricuspid or mitral valve abnormalities [8], the first two being the most common [3]. The combination of both CCTGA and IAA has very rarely been encountered. A review of the literature revealed only one similar case that has been reported earlier in 1981 [4].

2. CASE PRESENTATION

We report a preterm baby boy delivered by C-section at 36 weeks of gestation with a birth weight of 2.5 kg and with good Apgar score at one and 5 minutes. He had been diagnosed in utero as a case of large VSD and severe coarctation of the aorta. The baby was shifted to NICU and a 2D and color Doppler echocardiogram performed soon after birth revealed l-looping of the ventricles, with the morphological left ventricle–and the mitral valve on the right side, receiving systemic venous return from the morphologic right atrium and distally—being connected to the pulmonary arteries. There was also a large VSD with bidirectional shunt (Fig. 1). The morphological right ventricle with the tricuspid valve were on the left side, receiving pulmonary venous return from the left atrium and distally being connected to the aorta (Fig. 2). Suprasternal imaging revealed a small transverse aortic arch with interruption of aorta distal to the left subclavian artery. A small PDA was also confirmed with restrictive right to left shunt (Fig. 3).

Prostaglandin infusion was immediately started at a rate of 0.03 micrograms/kg/min. The baby remained stable with oxygen saturation of more than 94%, heart rate of 155 beat per minute, RR of 48 breath per minute, and the non-invasive blood pressure remained at 25th percentile for his age and weight, with no significant difference between the upper and lower limbs .

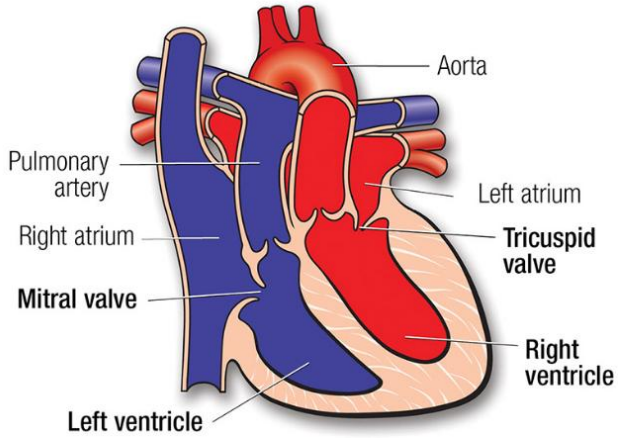
2.1 Systemic Physical Examination

- **CNS (central nervous system):** active, moving four limbs, no abnormal movement. Scaphocephaly Sagittal craniosynostosis (confermed by MRI)
- **Respiratory:** equal air entry bilateral with no added sounds
- **Cardiovascular:** Equally palpable pulse in all four limbs, capillary refill time less than 2 seconds, normal first and second heart sounds + pan systolic murmur 3/6, at left sternal border
- **Abdomen :** soft, no tenderness, liver 3 cm below costal margin
- Normal male genitalia
- **Imaging :**
- **Brain MRI** showed normal brain parenchymal tissue with Sagittal craniosynostosis
- **Renal and abdomen ultrasound :** unremarkable

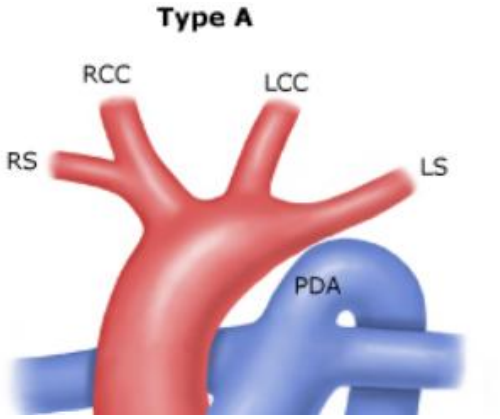
- **Chest X-ray :** cardiomegaly with increase vascular marking , no infiltration
- **Laboratory investigations:**
- **CBC** (cell blood count): white blood cells (WBC) 8.9, Hemoglobin 12.1gram/deciliter , HCT 37% , platelets 385000 per microliter
- **Renal profile :** within normal range for age
- **Liver function test :** within normal range
- **Serology:** negative

3. MANAGEMENT

A multidisciplinary meeting including pediatric cardiologist, pediatric neurologist and cardiac surgeons was held and a decision was made for aortic arch repair and pulmonary artery banding as first stage palliation which is the high risk procedure. Also review of the literature revealed only one similar case, reported earlier at 1981 by Cottrell, et al. [4]. In their case, the initial diagnosis was obtained by cardiac catheterization with angiography, as 2D color echocardiography was not available at their center. The clarity of echocardiographic images obtained in our institution enabled us to make this diagnosis without needing additional imaging modalities. The infant was placed on prostaglandin infusion at a rate of 0.03 micrograms/kg/minutes and remained hemodynamically stable. He underwent a successful aortic arch repair utilizing an end-to-end anastomosis and a PA band placement. The patient evolution was good post-operatively, with uneventful CSICU course .



Pic. 1. Corrected transposition of great arteries



Pic. 2. Interrupted aortic arch type A

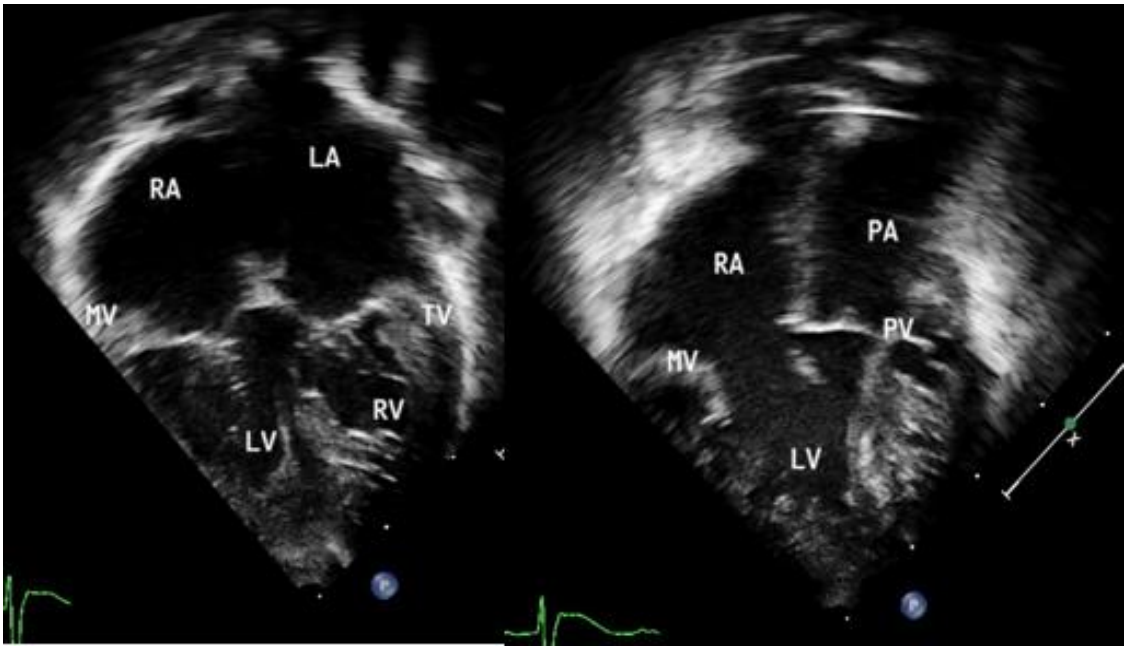


Fig. 1. Apical four chamber views of transthoracic echo showing congenitally corrected transposition of the great arteries, atrioventricular discordance and ventriculoarterial discordance (RA to LV to PA) (LA to RV to AO) (LA: left atrium, RA: right atrium, PA: pulmonary artery, LV: left ventricle , MV: mitral valve, AO: aorta, RV: right ventricle, PV: pulmonary valve)

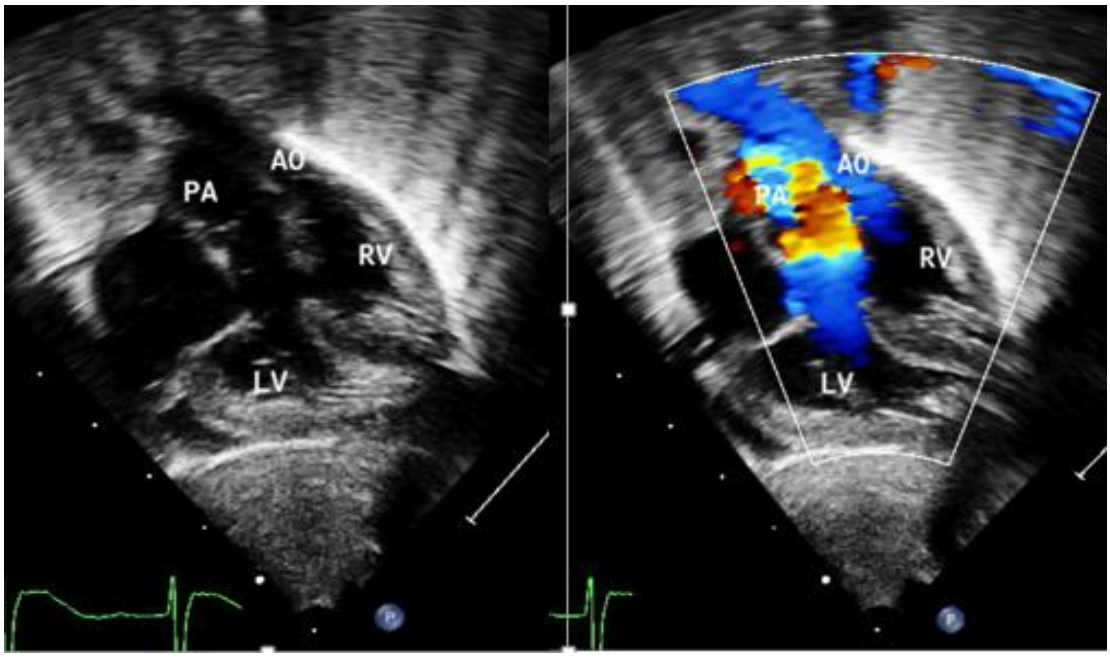


Fig. 2. Apical four chamber views of transthoracic echo showing Ventriculoarterial discordance: pulmonary artery arising from right sided left ventricle and aorta arising from left sided right ventricle and large ventricular septal defect. (LV: left ventricle, RV: right ventricle, AO: Aorta, PA: pulmonary artery)

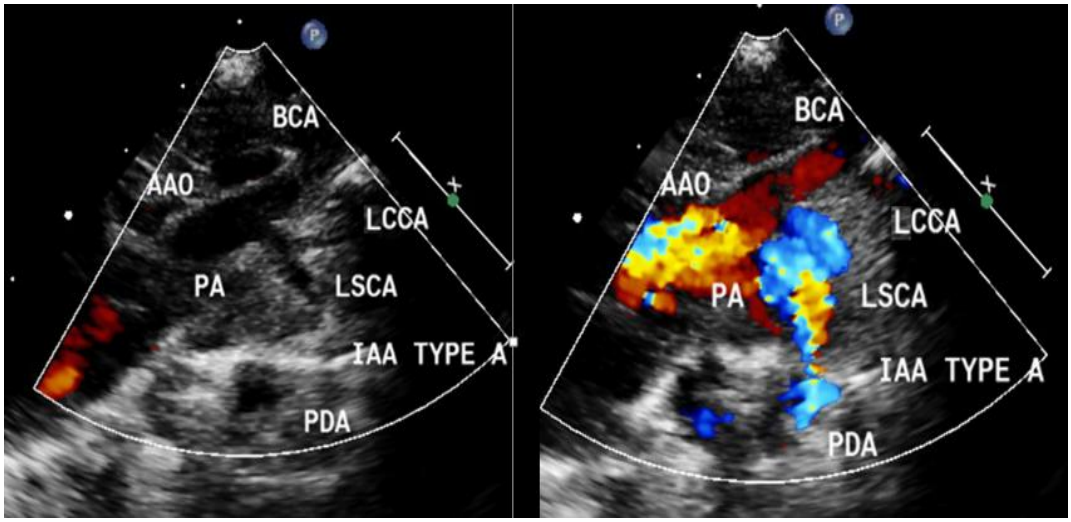


Fig. 3. Supra sternal views of transthoracic echo showing good size ascending aorta and brachiocephalic artery, left common carotid artery, left subclavian artery Smallish transverse arch with Interruption of aorta below the left subclavian artery, Small PDA with restrictive right to left flow.(AAO, Ascending aorta: PA, Pulmonary artery, BCA: brachiocephalic artery, LCCA: left common carotid artery, LSCA: left subclavian artery, PDA patent ductus arteriosus, IAA: interruption of aortic arch)

4. DISCUSSION

The actual cause of CCTGA is unknown,however it is thought to be caused by inappropriate looping of the heart during fetal life and its management is influenced by the associated cardiac abnormality such as Ventricular septal defect (VSD), Pulmonary stenosis (PS), hypoplastic ventricles or abnormal atrioventricular valves.

Also IAA type A is a rare malformation that occurs during fetal life as a result of abnormal fourth aortic arch regression and it is frequently associated with other cardiac defect such as VSD and PDA. The most common type of surgical repair is the total repair performed in the neonatal period.

This case describes an extremely rare association of complex congenital heart defects. This is only the second known case of a congenitally corrected transposition of the great arteries (L transposition) associated with Interrupted aortic arch (IAA) type A (4). The echocardiogram allowed for a thorough and timely diagnosis, with the introduction of appropriate therapy to maintain hemodynamic stability. With the images obtained there was no need for cardiac computed tomography (CT), cardiac magnetic resonance imaging (CMR) or cardiac angiography. Our aim of reporting this case is to share a very rare combination of congenital heart defects and its successful palliative management, and to emphasize the use of echocardiography in identifying such a complex defect without the need for additional routine or advanced cardiac imaging modalities to confirm the diagnosis.

5. CONCLUSION

This case describes an extremely rare combination of complex congenital heart defects. To the best of our knowledge , this is only the second known case of a congenitally corrected transposition of the great arteries (L transposition) associated with Interrupted aortic arch (IAA) type A [1]. A detailed echocardiographic evaluation helped us for timely diagnosis and an appropriate therapy to maintain hemodynamic stability. There was no need for cardiac CT, CMR or cardiac angiography. We believe that echocardiography remains the quintessential tool for the diagnosis of complex congenital cardiac anomalies, especially in centers where other forms of advanced imaging are still not available.

PARENTAL CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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