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Case Report

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Successful Repair of a Complete Atrioventricular Septal Defect Associated with Transposition of the Great Arteries: A Rare Association and a Literature Review

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Abstract

Complete atrioventricular septal defect associated with transposition of the great arteries is a relatively rare congenital heart disease, with only a few cases reported in the literature. Surgical strategy between one-stage or two-stage repair represents a challenging decision-making process. Herein, we report the case of an 11-month-old infant who received a postnatal diagnosis of a complete type A atrioventricular septal defect associated with malposition of the great arteries. The patient underwent a successful one-stage anatomic repair with arterial switch procedure and cAVSD repair with a two-patch technique. His post-operative course was uneventful and the patient was discharged home in stable condition.

This case highlights the suitability of one-stage surgical repair in infants with cAVSD and TGA, resulting in successful outcomes with few complications.

Keywords: Complete atrio-ventricular septal defect; Transposition of great arteries; One-stage repair

Abbreviations: D-TGA = D-loop Transposition of Great Arteries; cAVSD = Complete Atrioventricular Septal Defect; VSD = Ventricular Septal Defects; CPB = Cardiopulmonary Bypass; ASD = Atrial Septal Defect; LVOT = Left Ventricular Outflow Tract; ASO = arterial switch operation; PAB = Pulmonary Artery Banding; LDA/LAD = Left Anterior Descending Artery; CX/LCX = Circumflex Coronary Artery; RCA = Right Coronary Artery; Ao/A = Aorta; PA/P = Pulmonary Artery

Introduction

Complete atrioventricular septal defect (cAVSD) associated with true transposition of the great arteries (TGA) has rarely been reported in the literature and the surgical management of these patients is still debated. We describe an 11-month-old male infant with cAVSD and TGA arrived from abroad and successfully treated by complete anatomic correction. A detailed literature review concerning this interesting and unusual combination is also provided.

Case Report

An 11-month-old boy weighing 6.5 kg was transferred to our center from abroad after a postnatal diagnosis of cAVSD with TGA. On admission, he had mild congestive heart failure symptoms and was on diuretic oral therapy.

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Clinical examination revealed a grade 3/6 systolic murmur in the left sternal border and oxygen saturation of 84% on room air. Chest X-Ray showed an increased cardiothoracic ratio of 0.73 (Figure 1).

He was on sinus rhythm, first-degree atrioventricular block, biatrial enlargement, extreme axial deviation and biventricular hypertrophy.

Transthoracic echocardiography showed complete atrioventricular septal defect (type A Rastelli) with malposition of great arteries (right anterior aorta and left posterior pulmonary artery), moderate regurgitation along the coaptation rhyme of the left part of the common valve, two balanced ventricles, large inlet ventricular septal defect (VSD) without additional associated defects (Figure 2,3, 4A, 4B).

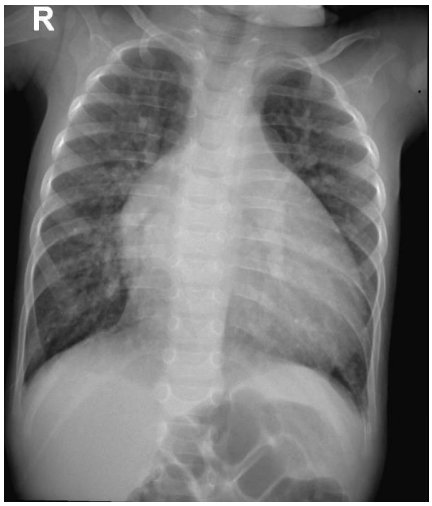


Figure 1: Preoperative Chest X-Ray.

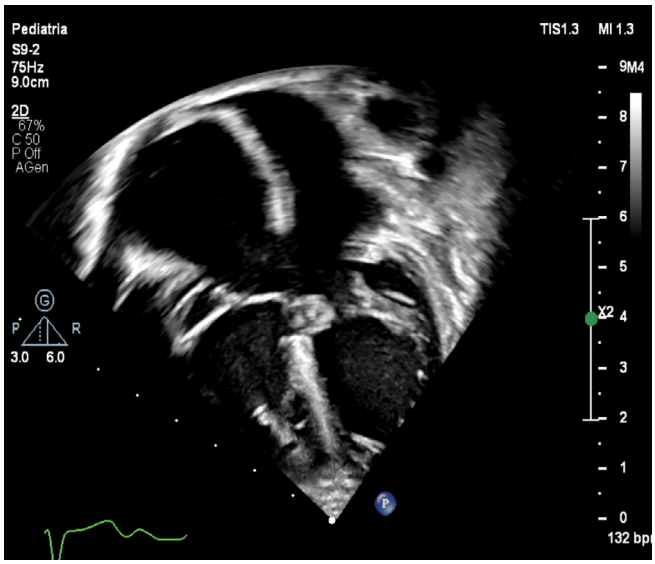


Figure 2: Complete atrio-ventricular septal defect with ostium primum defect and inlet ventricular septal defect.

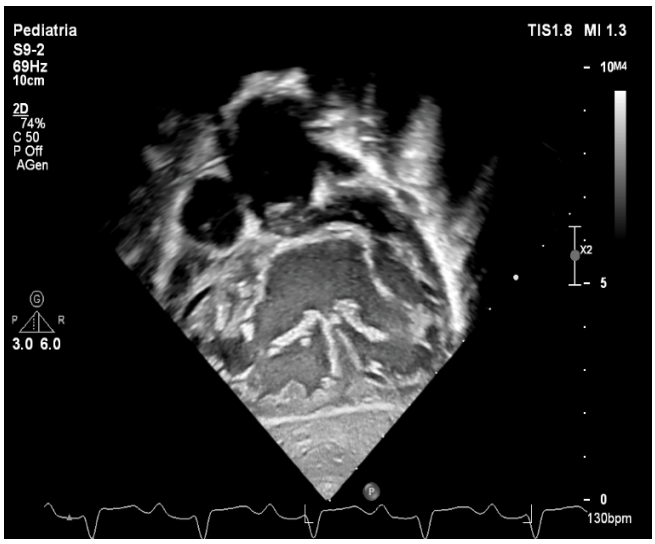
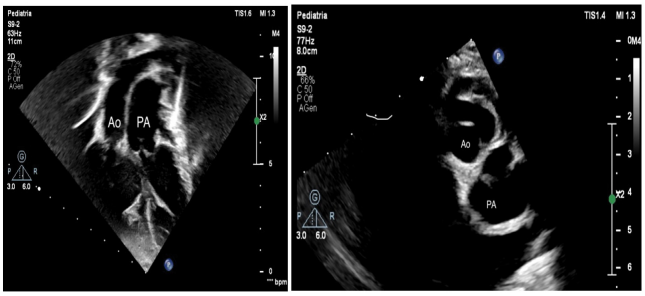


Figure 3: Common atrioventricular valve - Rastelli type A.



(a) (b)

Figure 4: Transposition of the great arteries. 4a: aorta ariese from the morphologically right ventricle and the pulmonary artery from the morphologically left ventricle. (b) Aorta (Ao) is anterior and to the right of the pulmonary artery (PA).

The examination also revealed an unusual coronary pattern. According to the Leiden Classification [1], the anterior descending artery originated directly from the right-facing sinus, while the right coronary artery and the circumflex artery had separate ostia taking off from the left-facing sinus (Figure 5 and 6).

After a median sternotomy and aorto-bicaval cannulation, cardiopulmonary bypass (CPB) was established, and the heart was arrested with Del Nido cardioplegia infusion. A left vent was inserted in the right superior pulmonary vein. After right atrial exposure, the complete atrioventricular septal defect was repaired with a two-patch technique, closing completely the left-valve component cleft with interrupted monofilament sutures. Atrial septal defect (ASD) and VSD were closed with bovine heterologous pericardium patches. Patent ductus arteriosus was doubly ligated and divided.

A single coronary button was harvested from each facing sinus. The arterial switch operation was performed with

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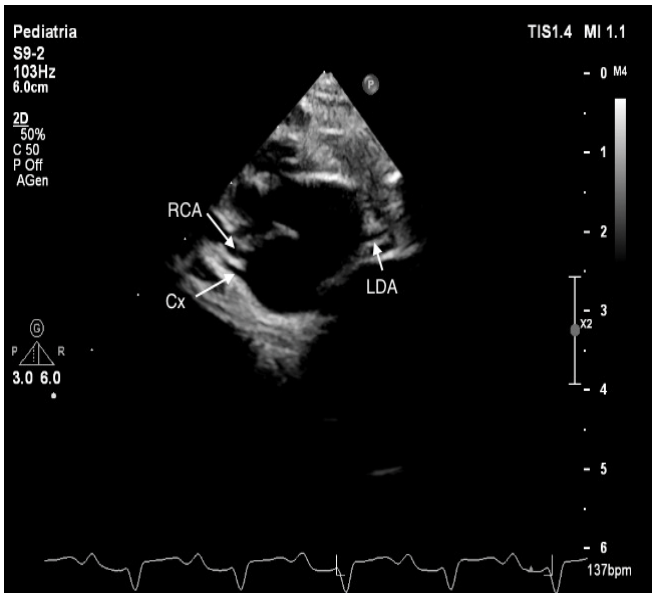

Figure 5: Right coronary artery (RCA) and circumflex artery (Cx) from two separated ostia on the right facing sinus; left anterior descending artery (LDA) from the left facing sinus of Valsalva.

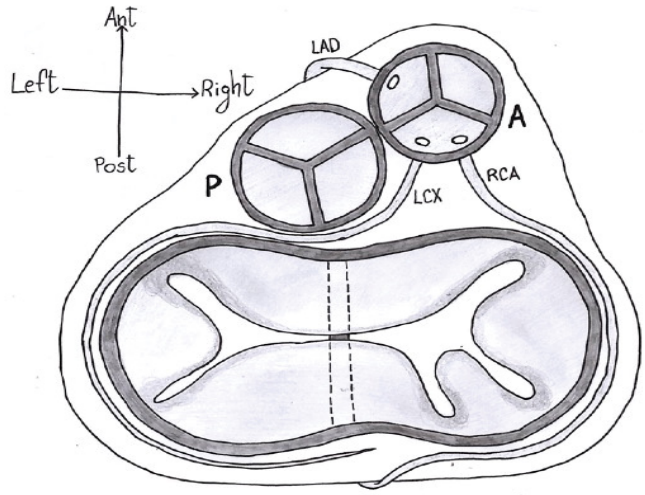

Figure 6: Schematic representation of our patient's anatomy (P = pulmonary valve, A = aortic valve, LAD = left anterior descending coronary artery, LCX = circumflex artery, RCA = right coronary artery).

French manoeuvre. The native pulmonary artery was twice the size of the aorta and a V-shaped reductive plasty of the neo-aortic root was therefore necessary to overcome this mismatch. The cross clamp and CPB times were 125 and 174 minutes, respectively.

The chest was left open and closed on the second postoperative day. The infant was extubated the following day. Inotropic support was discontinued on the third postoperative day. The post-operative course was uneventful except for sporadic and isolated wide QRS extrasystoles successfully treated with beta-blocker. Postoperative transthoracic echocardiography showed good biventricular systolic function with mild diastolic dysfunction, unobstructed left ventricular outflow tract (LVOT), no intracardiac shunts, mild right atrio-ventricular regurgitation, mild left atrio-ventricular regurgitation. The patient was discharged home in stable condition on the 15th postoperative day. At 6 months since the surgical treatment, he is in good health without any issues.

Discussion

In this case report, we have presented the first patient referred to our center with a diagnosis of cAVSD + TGA without other associated cardiac anomalies in whom it was possible to achieve a successful one-stage anatomical repair. The combination of these cardiac defects is extremely rare, and even rarer in the absence of associated cardiac anomalies. Consequently, the management and surgical experience are currently limited, with only a few cases reported in the literature. Till now, only four surgical reports have documented this intriguing combination [2-5].

Patients frequently exhibited one or more clinical symptoms, such as cyanosis, failure to thrive, cardiac murmurs, and signs indicative of congestive heart failure [2,3,5]. Our patient had no specific symptoms at birth, and his growth appeared relatively normal on arrival. Echocardiography served as the primary diagnostic modality in all patients, demonstrating a 100% accuracy rate. Median age was 18 months (range: 14 days – 9 years), and our patient was among the youngest observed.

A two-stage repair strategy was employed in just one patient, initially involving pulmonary artery banding (PAB) to manage pulmonary blood flow and alleviate congestive heart failure symptoms. Subsequently, arterial switch operation (ASO) with a two-patch repair of the cAVSD was performed at 18 months [5]. The predominantly adopted strategy has been instead the one-stage surgical repair [2-4], a successful approach also pursued for our patient, who came to our observation at 11 months of age. Regarding cAVSD repair, in all reported cases, the technique used was the two-patch one. This approach is routinely performed in our center using heterologous bovine pericardium patches.

The survival rate was 100%. This promising result is further reinforced by a post-operative course free of major and minor complications in all patients, except for the one who underwent a two-stage repair. In this case, extracorporeal membrane oxygenation was required due to severe ventricular dysfunction, making weaning from CPB impossible. The mechanical support was discontinued after five days, and at a 14-month follow-up, the patient exhibited good clinical conditions [5]. Our patient also exhibited no complications except occasional sporadic ventricular extrasystoles. This

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also confirms that the one-stage surgical strategy is indeed advantageous, exhibiting a low rate of major and minor complications independently of the age and weight of the patient.

In conclusion, our patient demonstrates the feasibility of one-stage in infants with cAVSD and TGA, achieving successful surgical outcomes with a notably low complication rate.

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Reference

1. Gittenberger-de Groot AC, Koenraadt WMC, Bartelings MM, et al. Coding of coronary arterial origin and branching in congenital heart disease: The modified Leiden Convention. J Thorac Cardiovasc Surg 156 (2018): 2260-2269.
2. Kumar N, Prabhakar G, Wilson N, et al. Total correction of transposition of great arteries with atrioventricular septal defect. Ann Thorac Surg 54 (1992): 989-990.
3. Katogi T, Aeba R, Tsutsumi K, et al. Primary repair of complete transposition of the great arteries with complete atrioventricular septal defect. Jpn J Thorac Cardiovasc Surg 48 (2000): 388-90.
4. Bautista-Hernandez V, Marx GR, del Nido PJ. One-stage neonatal corrective repair for d-transposition of the great arteries and complete atrio-ventricular canal. Eur J Cardiothorac Surg 31 (2007): 135-7.
5. Sabatino ME, Chalela T, Sandoval N. Staged repair of transposition of the great arteries with anomalous coronary anatomy and complete atrioventricular canal. Cardiol Young 30 (2020): 1188-1190.