

# Anatomically Corrected Malposition of the Great Arteries with Atrioventricular Concordance and Left Juxtaposition of the Atrial Appendages

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## Abstract

Anatomically corrected malposition of the great arteries is a rare cardiac malformation. In this condition the great arteries are abnormally related to each other and to the ventricles, but arise from the anatomically correct ventricles. In patients with atrioventricular concordant and the absence of associated anomalies, the circulation is physiologically normal. However, ventricular septal defect and right ventricular outflow tract stenosis are the most common accompanying pathologies that require surgical intervention. Here, we present a 9-day-old female with anatomically corrected malposition of the great arteries with wide atrial septal defect, mild right ventricular outflow tract stenosis, left juxtaposition of the atriums, and wide malalignment subaortic ventricular septal defect that required aortotomy to close the defect.

**Keywords:** Anatomically corrected malposition of the great arteries, echocardiography, juxtaposition, ventricular septal defect.

## Introduction

Anatomically corrected malposition of the great arteries (ACMGA) is a rare form of congenital heart disease in which the arterial trunks arise above the anatomically correct ventricles but are abnormally related to each other and to the ventricles.<sup>1</sup> There may be atrioventricular (AV) concordant or discordant, but there should be ventriculoarterial (VA) concordant. The aorta arises from the morphological left ventricle and the pulmonary trunk arises from the morphological right ventricle. In this situation, the aorta is

located anterior, which may be left or right of the pulmonary trunk. Clinical presentation and management of ACMGA relate to associated anomalies. Ventricular septal defect (VSD), right ventricular outflow tract obstruction (RVOTO), right ventricle (RV) hypoplasia, juxtaposed atrial appendage, and right aortic arch are the most associated anomalies.<sup>2,3</sup> In patients with atrioventricular concordant and the absence of associated anomalies, the circulation is physiologically normal.<sup>2</sup>



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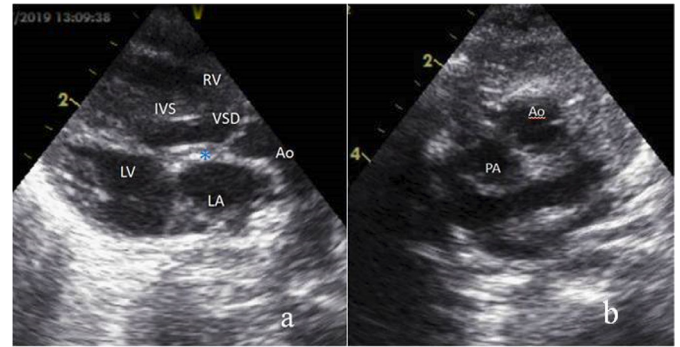
In this article, we present a patient who was diagnosed with ACMGA with large malalignment VSD, large secundum atrial septal defect (ASD), left juxtaposition of the atria, and mild RVOTO, whose VSD margins can only be approached through aortotomy.

## Case Report

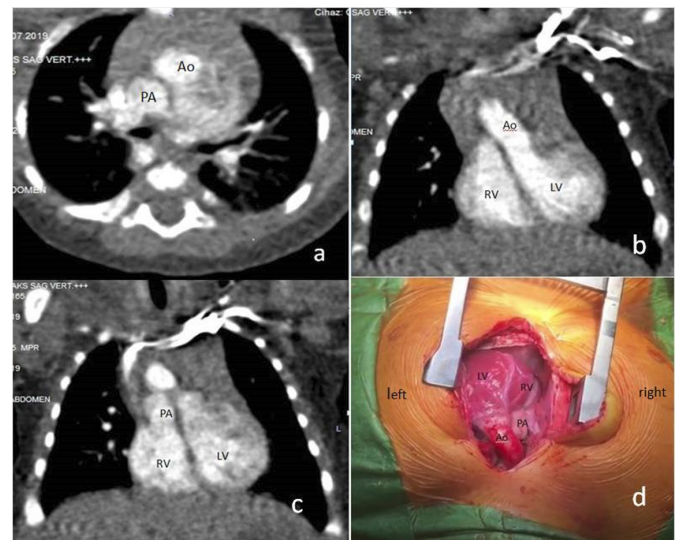
A 9-day-old female patient was consulted for a murmur following for hyperbilirubinemia in the neonatal intensive care unit. Her general condition was good, respiratory rate was 42 / min, heart rate was 148 / b.p.m, oxygen saturation was 98%. A 2/6 systolic murmur was in the pulmonary focus. Other system examinations were normal. Electrocardiography demonstrated normal sinus rhythm, normal axis and incomplete right bundle branch block. Her telecardiogram was normal with cardiac index within referral range. Transthoracic echocardiography showed atrial situs solitus (S); the right atrium was drained to the right sided morphological right ventricle and the left atrium to the left sided morphological left ventricle (D); the aorta arose morphological left ventricle and pulmonary trunk arose morphological right ventricle in a side by side position (AV-VA concordant) with the aorta was anterior leftward (L). The accompanying pathologies were wide subaortic malalignment VSD, wide secundum ASD, thin patent ductus arteriosus (PDA), mild RVOTO with estimate gradient of 30 mmHg, left juxtaposition of the atriums and right aortic arch. The diagnosis of ACMGA (S.D.L.) with subaortic malalignment VSD and mild RVOTO was therefore made (**Figure 1 a,b**). Decongestive treatment was given when she was one month old and followed up until the age of four months. Surgery was decided because the patient did not gain enough weight (4,92 kg, <3%) and had symptoms of heart failure. In addition to the echocardiographic findings, aberrant innominate artery was observed in the preoperative computed tomography angiography. (**Figure 2 a,b,c**). During the operation, median sternotomy incision was made to reach the mediastinal structures. Thymectomy performed. After pericardial incision, morphological findings of ACMGA were confirmed (**Figure 2 d**). Aortic bicaval cannulation performed. Patent ductus arteriosus ligated first then cardiopulmonary bypass instituted; and antegrad cold blood cardioplegia delivered. After diastolic arrest right atriotomy performed. Secundum type ASD was clearly seen but VSD was unable to localized. The main pulmonary artery was then opened to achieving more satisfactory visualization of the VSD margin. However, the VSD could not be seen through the pulmonary artery approach. Aortotomy was performed, then the leaflets of the aortic valve were retracted. The VSD was completely seen under the right coronary cusp of the aortic valve.

A polytetrafluoroethylene (PTFE) patch material was sutured to the margins of the VSD through the aortotomy approach. Aortopexy were performed to aberrant innominate artery pressing trachea anterior. Since the RVOTO was mild and pulmonary outflow tract was normal, its not intervened. Postoperative echocardiography showed an unobstructed pathway

from the LV to the aorta, with no residual shunt and no significant RVOTO. After an uneventfull recovery period, patient discharged in stable condition with sinus rhythm at postoperative day 5. Consent was obtained from the patient's parent for this case study.



**Figure 1.** a) Transthoracic echocardiography showed subaortic malalignment ventricular septal defect and aorta, arising from left ventricle, in five chamber view, b) aorta located anterior leftward in short axis view.



**Figure 2.** a) Computed tomography anjo showed aorta located anterior leftward, b) aorta arising from left ventricle, c) pulmonary artery arising from right ventricle, d) aorta, located anterior leftward, arising from left ventricle and pulmonary artery located posterior, arising from right ventricle in the surgical image.

## Discussion

Anatomically corrected malposition of the great arteries was first reported by Theremin in 1895, later characterized by Van Praagh et al. in 1975.<sup>4</sup> In contrast to transposition of the arterial trunks, which cross the opposite side of the septum and arise from the anatomically inappropriate ventricle, in this condition parallel arterial trunks are normally connected to their appropriate ventricles (VA concordant).<sup>5</sup> It is divided to 4 types based on atrium, ventricle and great arteries segmental analysis. According to Van Praagh's symbolic terminology, there are two types of situs solitus, type 1: (S.D.L.), type 2: (S.L.D.) and two types of situs inversus, type 3: (I.L.D.), type 4: (I.D.L.). Of these pathologies, type 1 and type 3 have normal corrected physiology, while type 2 and type 4 have transposition physiology.<sup>4</sup> According to Ali et al.

review, 78% of the cases are (S.D.L.) and the others are 17.5% (S.L.D.), 4.5% (I.D.L.) types in biventricular ACMGA.<sup>2</sup> In addition to biventricular pathologies, the combination of concordant VA connections with parallel arterial trunks can be found in the setting of isomeric atrial appendages, double inlet left ventricle, and in absence of the right atrioventricular connection.<sup>6</sup> Arterial trunks may also be VA concordant when the morphologically right ventricle was itself left sided, or in the setting of mirror-imaged atrial arrangement but with the morphologically right ventricle in the right-sided position. Thus, the pulmonary trunk can be located on the left as in the normal heart, or on the right as expected for the mirror-imaged atrial arrangement. So, it can be claimed that only the ventricular arrangement is abnormal. Therefore, these unusual situations can be described as isolated ventricular inversion or isolated atrioventricular discordant.<sup>6</sup>

The clinical presentation and management of ACMGA depends on the associated anomalies, such as VSD, ASD, PDA and ventricular outflow tract stenosis (subaortic or subpulmonic). The most common concomitant anomaly is VSD (90%), and the most frequent indication of the surgery is also VSD. Pulmonary stenosis (valvular and/or) often accompanies VSD (59%), and relieve of the pulmonary stenosis may also be required with VSD closure. The other associated anomalies are: 62.5% of the cases have left juxtaposition, 15% of the cases have aortic stenosis.<sup>2</sup> In our patient, there were subaortic malalignment VSD, wide secundum ASD, left juxtaposed appendage, right aortic arch and mild pulmonary stenosis. At the age of four months, VSD and ASD was surgically closed due to growth retardation; pulmonary stenosis was not intervened because it was mild. When S.D.L. is present, the surgical success rate is 92%. However, when AV discordant or hypoplastic RV present, success rate is only 29%.<sup>7</sup> Some syndromes such as VACTERL<sup>8</sup> and Goldenhar Syndrome<sup>9</sup> were reported to be associated with ACMGA. However, there was no accompanying syndrome in our patient.

Awasthy et al.<sup>10</sup> reported that in their patient with ACMGA (S.D.L.) and DORV the presence of subaortic conus (infundibulum) along with the left juxtaposition of the right atrial appendage resulted in the inability to visualize the VSD from either the right atrium or pulmonary artery or even through a ventriculotomy approach, thus had been necessitated a transaortic approach. In our patient, like Awasthy et al.,<sup>10</sup> we could not reach from the right atrium or pulmonary artery; the VSD could be fully visualized in its entirety only by looking down through the aortic valve orifice.

## Conclusion

Because of ACMGA is a rare entity, it can be confused with congenitally corrected transposition of the great arteries and transposition of the great arteries. The segmental approach will help to evaluate the morphology of the heart before surgery and to distinguish it from other similar pathologies. This case also highlights the need to recognize the value of the transaortic approach to VSD, in the segmental anatomy that exists with the left juxtaposition of the atriums in ACMGA (S.D.L.) as reported earlier.

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**Informed Consent:** Written informed consent was obtained from patients who participated in this study.

**Peer-review:** Externally peer-reviewed.

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