

## Anatomically corrected malposition of the great arteries: two case reports

### Büyük arterlerin anatomik düzeltilmiş malpozisyonu: İki olgu sunumu

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**Summary**– Anatomically corrected malposition of the great arteries (ACMGA) is defined as parallel arising of aorta and main pulmonary artery (PA) roots although ventriculoarterial connection is normal. Abnormally related aorta arises from the left ventricle, while abnormally related PA arises from the right ventricle. It can be diagnosed with via echocardiography. In some cases, additional imaging modalities such as computerized tomographic angiography and magnetic resonance are required. In this article, we presented two cases of ACMGA, 5-month-old boy and 1-month-old girl. We wanted to point out the importance of differential diagnosis of other great artery anomalies from this rare pathology.

Anatomically corrected malposition of great arteries (ACMGA) is a very rare congenital heart disease. It is defined as parallel arising of aorta and main pulmonary artery (PA) roots although ventriculoarterial connection is normal. Abnormally connected aorta arises from left ventricle and main PA from right ventricle (RV).<sup>[1]</sup>

Unless well segmental approach is applied, ACMGA can be confused with transposition of great arteries or congenitally corrected transposition of great arteries in which atrial, ventricular and great artery connections are disturbed.

Echocardiography is not always sufficient for accurate description of those pathologies which require different treatment approaches. In some cases, additional imaging modalities such as computerized tomographic angiography (CTA) and magnetic reso-

**Özet**– Büyük arterlerin anatomik düzeltilmiş malpozisyonu (BAADM), ventriküloarteryel bağlantının normal olmasına rağmen aort ve ana pulmoner arter köklerinin birbirine paralel çıkması olarak tanımlanır. Anormal ilişkili aort sol ventrikülden, anormal ilişkili pulmoner arter sağ ventrikülden çıkar. Ekokardiyografi tanıda kullanılmaktadır. Bazı olgularda bilgisayarlı tomografik anjiyografi ve manyetik rezonans incelemesi gibi görüntüleme yöntemlerinden yararlanılabilir. Bu yazıda beş aylık erkek ve bir aylık kız olmak üzere BAADM tanısı konulan iki olgu sunuldu. Böylece nadir görülen bu patolojinin diğer büyük arter anomalileri ile ayırıcı tanısının önemine dikkat çekilmek istenmiştir.

nance imaging (MRI) are required.

In this article, we presented two ACMGA cases, consist of a 5-month-old boy and a 1-month-old girl with 128 slice dual source CTA findings. Hence, we wanted to point out the importance of differential diagnosis of other great artery anomalies from this rare pathology.

#### Abbreviations:

|       |  |
|-------|--|
| ACMGA | Anatomically corrected malposition of great arteries |
| CTA   | Computerized tomographic angiography                 |
| LA    | Left atrium  |
| MRI   | Magnetic resonance imaging                           |
| PA    | Pulmonary artery                                     |
| RV    | Right ventricle                                      |
| VSD   | Ventricular septal defect                            |

#### CASE REPORT

**Case 1**– He was first referred to our hospital with respiratory distress at 3 months of age. His body weight and length were 4000 g and 53.5 cm (<3rd percen-

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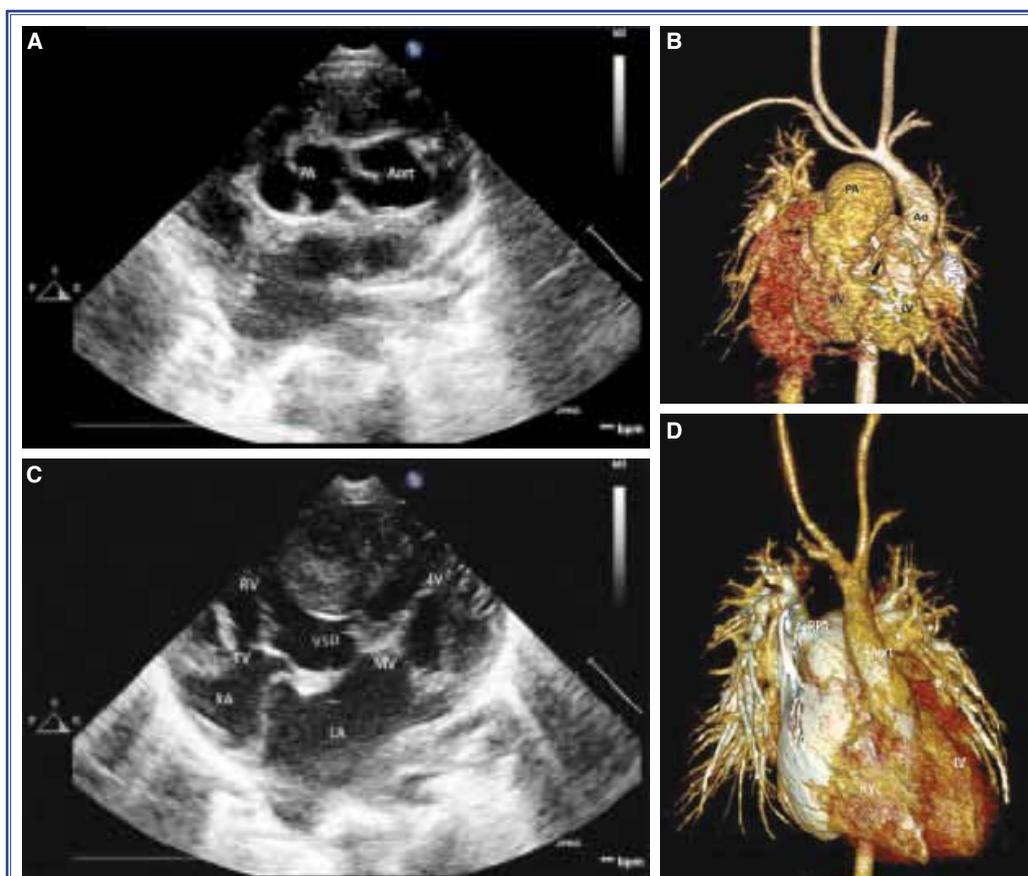


tile), respectively. His heart rate was 160 beats/min and there was a 3/6 systolic murmur over left sternal border. His blood pressure was 72/40 mmHg and respiratory rate was 65/min. The liver was 5 cm palpable and oxygen saturation was 92%. Electrocardiography showed normal sinus rhythm and right axis deviation. The chest X-ray revealed cardiomegaly. On echocardiogram, atrial situs was solitus with levocardia, atrioventricular concordance, and ventriculoarterial concordance. There was a large subpulmonic ventricular septal defect (VSD). However, the aorta was to the left and anterior of the pulmonary trunk, which is different from the usual spatial great artery relation (Figure 1a). Aortic arch was elongated and dextropositioned. Pulmonary arterial pressure was also high that was estimated via tricuspid valve regurgitation which was 55 mmHg.

A 128 slice dual source CTA revealed levocardia with viscerotrial situs solitus and D-looped ventricles (Figure 1b). There was atrioventricular and ventriculoarterial concordance, although there were abnormally related great arteries, with the aorta arising slightly more anterior and to the left of the PA (L-malpositioned). There was conal tissue beneath each great artery, resulting in aortic-mitral and pulmonary-tricuspid discontinuity. These findings are consistent with ACMGA.

Pulmonary banding operation was performed and patient was discharged at post-operative day 7. The patient is being followed since then.

**Case 2**– For the first time, 14 days old female patient was postnatally referred to our clinic following detection of transposition of great arteries in fetal ul-



**Figure 1.** (A) Case 1; echocardiographic parasternal short axis view. PA: Pulmonary artery. (B) Case 1; three-dimensional computerized tomographic view of the heart. PA: Pulmonary artery; Ao: Aorta; RV: Right ventricle; LV: Left ventricle. (C) Case 2; echocardiographic modified apical four chamber view. RA: Right atrium; LA: Left atrium; VSD: Ventricular septal defect; MV: Mitral valve; TV: Tricuspid valve; RV: Right ventricle; LV: Left ventricle. (D) Case 2; three-dimensional computerized tomographic view of the heart. RPA: Right pulmonary artery; Ao: Aorta; RV: Right ventricle; LV: Left ventricle.

trasonography performed for maternal Type 1 diabetes mellitus. Her weight and length were 4700 g and 55.5 cm (>97th percentile), respectively. Her heart rate was 160 beats/min and there was a 3/6 systolic murmur over left sternal border. Her blood pressure was 62/38 mmHg and respiratory rate was 45/min. The liver was 3 cm palpable and oxygen saturation was 95%. In echocardiographic study, situs solitus levocardia, large subpulmonic and muscular VSDs, atrioventricular concordance, and ventriculoarterial concordance were demonstrated. Aorta and PA relation could not be evaluated due to extreme septal hypertrophy. Septal hypertrophy significantly regressed after 3 weeks. Aortic arch was dextropositioned. As no gradient was detected through VSD; PA pressure was estimated systemic (Figure 1c). The CT helped us to identify the relationship between aorta and main PA which could not be assessed by echocardiography. There was atrioventricular and ventriculoarterial concordance, although there were abnormally related great arteries, with the aorta arising slightly more anterior and to the left of the PA (L-malpositioned) (Figure 1d). The patient who had pulmonary banding at 2nd month of her life was discharged at post-operative 8th day. This case is on follow-up without any complication.

## DISCUSSION

This rare form of congenital heart disease was first reported by Theremin in 1895 and later characterized by Van Praagh et al. in 1975.<sup>[1]</sup> It is divided to 4 types based on atrium, ventricle and great arteries segmental analysis.

Type 1 (S, D, L) is situs solitus (right atrium [RA] is on the right of left atrium [LA]), D-loop ventricle (morphologic RV is on the right of interventricular septum), aorta being ahead and on the left of the PA.

Type 2 (S, L, D) is situs solitus, L-loop ventricle (morphologic RV is on the left of interventricular septum) and aorta being ahead and on the right of PA.

Type 3 is (I, L, D) situs inversus (right atrium is on the left of LA), L-loop ventricle and aorta being ahead and on the right of PA.

Type 4 (I, D, L) is situs inversus, D-loop ventricle and aorta being ahead and on the left of PA.

Types 1 and 3 have the features of corrected transposition; Types 2 and 4 physiologic transposition.

Most of described cases in literature (85%) are Type 1. Both of our cases were consistent with Type 1.<sup>[1]</sup>

ACMGA may accompany various cardiac defects such as VSD, RV outlet obstruction, subaortic stenosis, RV hypoplasia, atrial appendix juxtaposition and right aortic arch.<sup>[2,3]</sup> Both cases additionally had VSD and right aortic arch.

Diagnosis of this cardiac pathology which is limited in literature was made with echocardiography and autopsy when first defined.

In some cases, conventional angiography may be useful for clarifying the anatomy and evaluating the hemodynamic data. In recent years, with the development of the imaging techniques, complex cardiac abnormalities can be diagnosed faster and easier with either CTA and/or MRI.<sup>[4,5]</sup> In the both cases, without cardiac angiography, echocardiography and CTA were sufficient to make the correct diagnosis.

The infant of a diabetic mother who has ACMGA is presented in case two. To the best of the our knowledge, this is the first report of such an association.

Surgical results for associated cardiac pathologies are quite well (procedure success, 92%) in the cases of situs solitus and atrioventricular concordance; however, those results are not good in the cases of atrioventricular discordance or RV hypoplasia or both. The overall survival rate was only 29% among these patients.<sup>[6]</sup> Orun et al. recently, reported two cases with ACMGA and atrioventricular and ventriculoarterial concordance. In the first case treatment was achieved by surgically repairing VSD and ASD. In the second case surgically VSD closure and right ventricular outflow reconstruction was performed.<sup>[7]</sup> In both of our cases due to the lack of adequate intact interventricular septum, surgeons could not perform primary VSD closure and performed pulmonary banding. No problem has been observed since the operation. During follow-up, we plan biventricular treatment by closing VSDs for these two patients.

ACMGA is a rare cardiac pathology; which can be diagnosed with echocardiography. CT or MRI can provide better segmental analysis, in some cases. Since the treatment approach, especially in the presence of additional congenital heart disease is different from transposition of great arteries and corrected transposition of great arteries (c-TGA); correct differential diagnosis is crucial.

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**Key words:** Aorta/pathology; great arteries; heart septal defects; infant; pulmonary artery/pathology; transposition of great vessels/diagnosis.

**Anahtar sözcükler:** Aort/patoloji; büyük damarlar; kalp septal defekti; yenidoğan; pulmoner arter/patoloji; düzeltilmiş büyük damar pozisyonu/tanı.