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A Rare Case: Coexistence of Double Aortic Arch and Tetralogy of Fallot in a 1-Year-Old Child

Vascular ring anomalies are approximately 1% of congenital heart diseases. Double aortic arch (DAA) is the most common vascular ring anomaly and forms a complete vascular ring surrounding the trachea and esophagus. Vascular rings are usually isolated, but they may be detected in 1%-1.6% of congenital heart diseases.[1]

Symptoms and progression depend on the degree of tracheal and esophageal compression created by the vascular ring. Double aortic arch patients may present with symptoms such as shortness of breath, stridor, and feeding and swallowing difficulties at the time of infancy; however, it is quite difficult to diagnose asymptomatic DAA.[2] This study aims to present a case of DAA balanced with Tetralogy of Fallot (TOF) in a 1-year-old boy who had no symptoms except cyanosis and murmur.

A 1-year-old male patient was referred to our clinic and diagnosed with TOF. On physical examination, his weight and height were 9 kg and 71 cm, respectively (normal percentile), heart rate was 115 bpm, respiratory rate was 25/min, oxygen saturation was 79%, and heart sounds had ¾ systolic murmur. His family said he had no history of dysphagia, dyspnea, or stridor.

Echocardiography showed a large ventricular septal defect showing malalignment in the perimembranous area (Figure 1A), right ventricle outflow tract (RVOT) narrowed up to 3.5 mm (85 mm Hg pressure gradient) (Figure 1B), and suspected DAA. The patient was diagnosed with the TOF and computed tomography angiography (CTA) and angiography were planned. Computed tomography angiography and angiography confirmed the diagnosis of balanced DAA. In angiography, contrast agent injections were made to the left ventricle at 20˚ left oblique anterior-20˚cranial angulations (Figure 2A) and to the descending aorta in the anterior—posterior position (Figure 2B); it was observed that the aorta was divided into 2 as the right aortic arch and the left aortic arch, and after giving the aortic arch branches, they were united to form the descending aorta. In CTA, the right aortic arch was 8.3 mm and the left aortic arch was 7.3 mm in diameter (Figure 3). The vascular ring surrounded the trachea and esophagus (Figure 4). In view of the

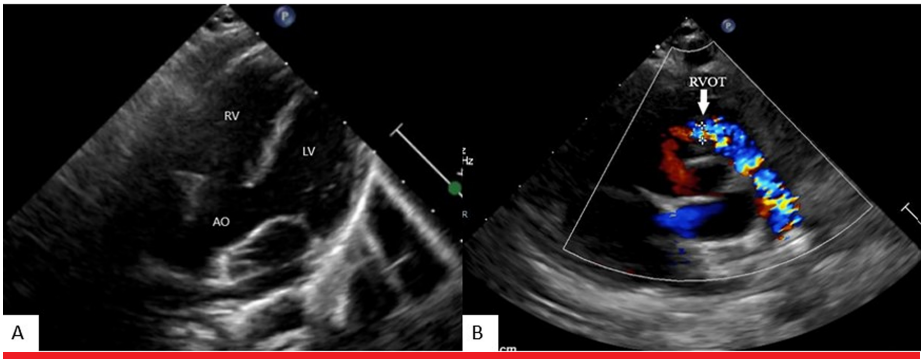

Figure 1. (A, B). Long axis and modified short axis transthoracic echocardiography showing posterior malalignment ventricular septal defect and RV outflow tract. LV, left ventricle; RV, right ventricle; AO, aorta; RVOT, right ventricle outflow tract.

E-PAGE ORIGINAL IMAGE

Doğan Çağrı Tanrıverdi[1]
Murat Şahin[1]
İsmihan Selen Onan[2]

[1]*Department of Pediatric Cardiology, University of Health Sciences, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center, İstanbul, Turkey*
[2]*Department of Pediatric Cardiovascular Surgery, University of Health Sciences, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center, İstanbul, Turkey*

Corresponding author:
Doğan Çağrı Tanrıverdi
✉ dctanriverdi@outlook.com

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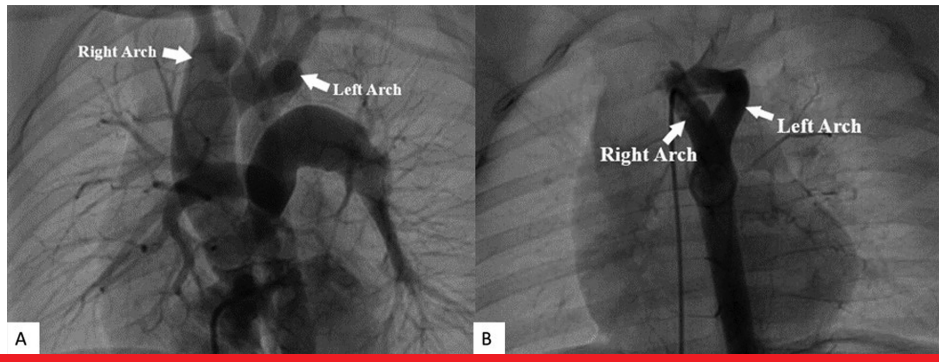

Figure 2. (A, B). Image of double aortic arch seen in aortic root angiograms (A) and descending aorta angiography (B) in anteroposterior position. AO, aorta; DA, descending aorta; PA, pulmonary artery.

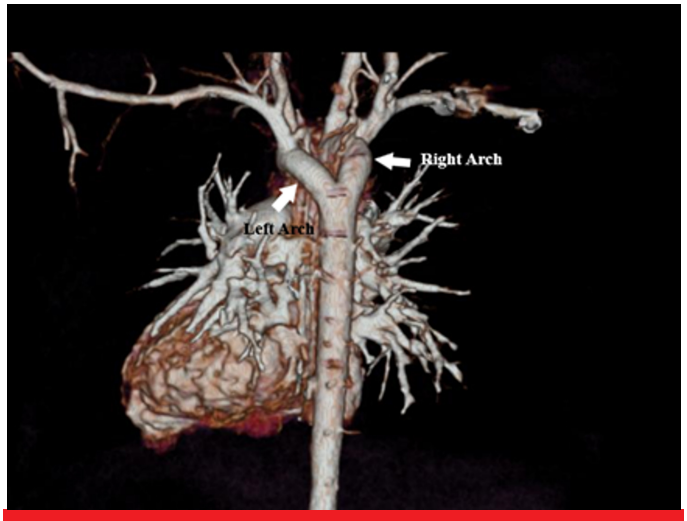

Figure 3. A 3-dimensional volume-rendered image of the double aortic arch seen in contrast-enhanced computed tomography in posteroanterior position.

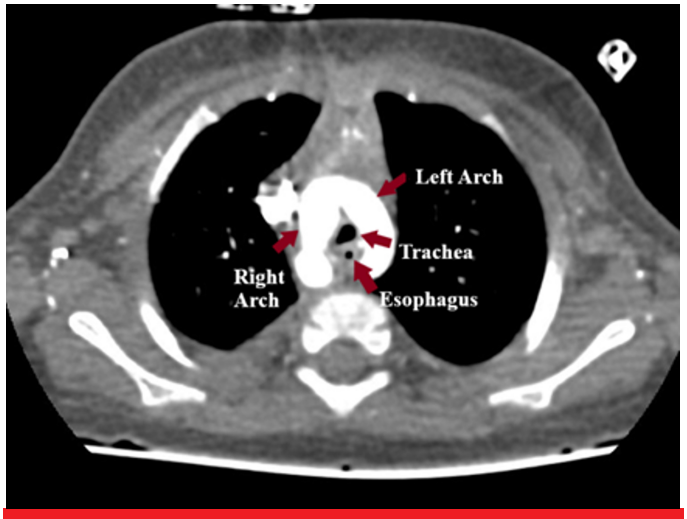

Figure 4. Computed tomography of the thorax showing the vascular ring surrounding the trachea and esophagus.

clinical features and investigations, we decided to perform a 1-stage operation, including TOF repair and left aortic arch dissection.

The message we want to give in this case is: When a pathological finding is found in pediatric cardiology practice, other accompanying congenital heart diseases should not be overlooked. As in our case, knowing the accompanying pathologies pre-operatively is important in terms of surgical planning. It should be kept in mind that the association of TOF and DAA can rarely be seen together.

The authors assert that this work complies with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008.

Informed Consent: This case was approved by the patient's family.

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