

## Double Aortic Arch with Mirror-Image Dextrocardia

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### ABSTRACT

Double aortic arch (DAA) is a very rare congenital vascular anomaly, characterized by the encircling trachea and esophagus, resulting feeding and/or respiratory problems. In this article, a two-month-old girl with situs inversus totalis and DAA was presented. The baby was symptomatic with noisy breathing and coughing since birth. Double aortic arch was detected by echocardiography, barium esophagography, cardiac MRI and conventional angiography. The present report emphasis that vascular ring should have been evaluated in a patient with respiratory and/or feeding problems. Chest radiography, barium esophagography, echocardiography, multislice computed tomography, magnetic resonance imaging angiography and conventional cardiovascular angiography can be used for the diagnosis of DAA.

**Key words:** Double aortic arch, dextrocardia, vascular ring

### Ayna Hayali Dekstrokardi ile Birlikte Çift Arkus Aorta

#### ÖZET

Çift arkus aorta (ÇAA) nadir bir konjenital vasküler anomali olup, beslenme ve/veya solunum problemleri ile sonuçlanan, trakea ve özefagusun çevrelenmesi ile karakterizedir. Bu makalede, situs inversus totalis ile birlikte ÇAA bulunan 2 aylık bir kız hasta sunulmuştur. Bebeğin doğumdan beri sesli soluk alma ve öksürük semptomları bulunmaktaydı. Çift arkus aorta, ekokardiografi, baryumlu özofagografi, kardiyak MR ve konvensiyonel anjiyografi ile saptandı. Bu makalede, solunum ve/veya beslenme problemleri olan bir hastada vasküler halkanın mutlaka araştırılması gerektiği vurgulanmıştır. Göğüs grafisi, baryum özofagografi, ekokardiografi, multislice bilgisayarlı tomografi, magnetic rezonans görüntüleme anjiyografi ve konvensiyonel kardiyovasküler anjiyografi ÇAA tanısında kullanılabilir.

**Anahtar kelimeler:** Çift arkus aorta, dekstrokardi, vasküler halka

### INTRODUCTION

Vascular rings represent approximately 1% of cardiovascular congenital anomalies and it refers to an encirclement of the trachea and the esophagus by an abnormal combination of aortic arch system (1). The usual cause is failure, during embryonic development, of the normal regression of the aortic arches that arise from the truncus arteriosus (2). Vascular rings are formed by double aortic arches with equal right and left components

or with a smaller or atretic right or left component; the descending thoracic aorta may be left or right sided, and there may be a left, right, or bilateral patent ductus arteriosus, or a ligamentum arteriosum (2, 3). Most of the patients are symptomatic due to the compression caused by vascular anomalies. However, some patients are asymptomatic. Double aortic arch (DAA) is the most common type of the vascular rings and also may cause severe and early symptoms. Echocardiography, barium

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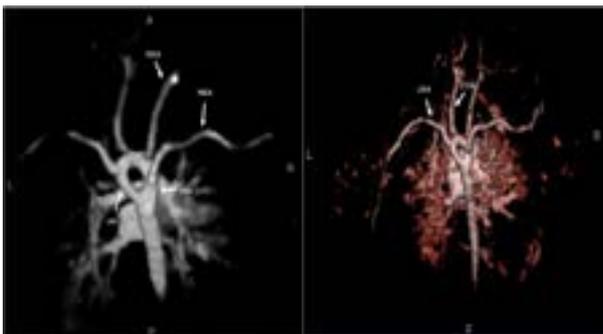
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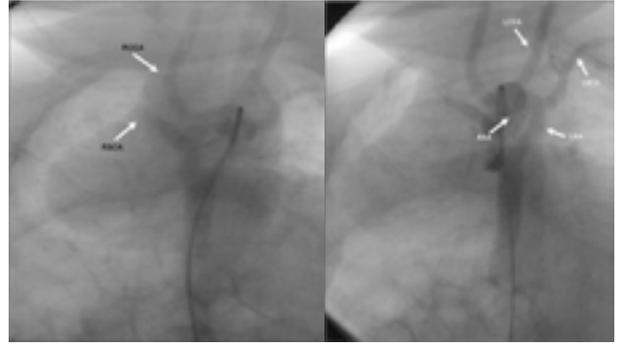


**Figure 1.** Dextrocardia, normal heart size and right-sided gastric air bubble on chest x-ray (a). Posterior and lateral indentation on the upper esophageal shadow on barium swallow study (b).

esophagography, cardiac MRI, conventional angiography and also multislice computed tomography afford a highly accurate preoperative evaluation of the vascular anatomy and its relationship to the trachea and esophagus (4, 5). Surgical treatment can be considered due to the degree of the symptoms. Thoracotomy should be carried out on the side of the smaller or atretic aortic arch and on the side of the persistent ductus arteriosus or ligamentum arteriosum (3, 6). Dextrocardia is defined as a right-sided heart with a base-apex axis directed rightward, resulting from a variation in cardiac develop-



**Figure 3.** The left-sided, smaller aortic arch, left subclavian artery with left common carotid artery both originating from this arch on cardiac catheterization (a) and cardiac MRI (b). (LCAA; left common carotid artery, LSCA; left subclavian artery, LAA; left aortic arch, RAA; right aortic arch)



**Figure 2.** The right-sided; greater and the main aortic arch, right subclavian artery with right common carotid arteries originating from this arch on cardiac catheterization (a) and cardiac MRI (b). (RCAA; right common carotid artery, RSCA; right subclavian artery, LAA; left aortic arch, RAA; right aortic arch)

ment and it occurs in approximately 0.01% of live births (7). The four common types of dextrocardia are classic mirror-image dextrocardia, normal heart displaced to the right side of the chest, L-TGA, and single ventricle (1, 7). Dextrocardia may accompany to asplenia and polysplenia syndromes with situs ambiguus and complicated cardiac defects. Classic mirror-image dextrocardia is composed of situs inversus, atrioventricular concordance, ventriculoarterial concordance and normally related great arteries (1). The present report emphasizes the vascular ring and its relation to the respiratory and/or feeding problems. Also, up to our knowledge DAA with mirror-image dextrocardia has been rarely reported in the literature.

### CASE

The patient, a 2 month-old girl, was referred to our clinic by her pediatrician for persistent noisy breathing and coughing which have been occurring since birth. Her birth history was unremarkable. However, she was born to a nonconsanguineous parent of a Turkish family. She had been evaluated because of respiratory problems in Department of Pediatric Chest Diseases and there was no history of hospitalization, repeated airway infections, and life-threatening respiratory compromise. On physical examination; her weight, height and head circumference were all normal ranges. Blood pressures were

70 mmHg systolic in both arms and legs. Heart sounds were louder over the right side of the chest. The noise was primarily on inspiration and worsened with feeding. There was no hepatomegaly and peripheral pulses were normal. Chest x-ray revealed dextrocardia, normal heart size and right-sided gastric air bubble (Figure 1a). On abdominal ultrasonography left-sided liver and right-sided spleen and stomach were reported. Mirror-image dextrocardia (situs inversus, atrioventricular concordance, ventriculoarterial concordance and normally related great arteries) was showed by echocardiography and no cardiac defect was determined. A barium swallow study revealed a posterior and lateral indentation on the upper esophageal shadow (Figure 1b). On conventional angiography and magnetic resonance imaging angiography, mirror-image dextrocardia and DAA were determined particularly. The right-sided aortic arch; main arch; was greater and right subclavian and right common carotid arteries originate from this arch (Figure 2a, b). However, the left-sided aortic arch was smaller and left subclavian and left common carotid arteries both originate from this part (Figure 3a, b). So, our patient had an asymmetrical DAA. Also, the trachea was narrowed between these two aortic arches. These findings suggest a vascular ring. The patient was mildly symptomatic and there was no growth retardation. Eventually, she has been following up in our clinic without any complication.

## DISCUSSION

Vascular ring is a term given to a combination of vascular and often ligamentous structures that encircle the trachea and esophagus. The usual cause is failure, during embryonic development, of the normal regression of the aortic arches that arise from the truncus arteriosus. The diagnosis can be difficult, because clinical symptoms can be variable and nonspecific. The clinical manifestations vary from asymptomatic to feeding difficulties, repeated airway infections, and life-threatening respiratory compromise. The complete type of vascular ring includes double aortic arch and right aortic arch with left ligamentum arteriosum (1). In this type of vascular ring, abnormal vascular structures form a complete circle around the trachea and esophagus. Incomplete vascular ring refers to vascular anomalies that do not form a complete circle around the trachea and esophagus but do compress the trachea or esophagus.

These include anomalous innominate artery, aberrant right subclavian artery, and anomalous left pulmonary artery (vascular sling) (1, 7).

Double aortic arch is the most common type and clinically recognized form of vascular rings (40%) (8). Double aortic arch usually manifests earlier in infants with symptoms of stridor, dyspnea, cough, and recurrent respiratory infections (1, 8). This anomaly is due to a failure of regression of both the right and left fourth branchial arches, resulting in right and left aortic arches, respectively. These two arches completely encircle and compress the trachea and esophagus, producing respiratory symptoms (1, 9). Also, these arches may be symmetrical or asymmetrical. The patients with DAA are usually symptomatic with respiratory and gastrointestinal symptoms or signs. Respiratory symptoms were stridor, wheezing, cough, recession, bronchiolitis, croup, recurrent chest and upper respiratory tract infections, tachypnea, apnea, pneumonia, asthma, respiratory arrest, shortness of breath, aspiration and increased oral secretion (10). Gastrointestinal signs were choking with feeds, dysphagia, failure to thrive, emesis and cyanosis with feeds (10). Our case had also wheezing frequently but not coughing with feeding.

Chest radiography, barium esophagography, echocardiography, multislice computed tomography, magnetic resonance imaging angiography and conventional cardiovascular angiography can be used for the diagnosis of DAA (11, 12). The development of cross-sectional imaging techniques has facilitated the correct diagnosis and surgical treatment of compression caused by vascular anomalies. In recent years, multislice computed tomography affords a highly accurate preoperative evaluation of the vascular anatomy and its relationship to the trachea and esophagus. In our case, DAA was diagnosed with barium esophagography, especially conventional angiography and magnetic resonance imaging angiography.

The right aortic arch is usually larger than the left arch (70%), but on rare occasions partial obstruction or complete atresia of the left arch may exist (1, 13). Double aortic arch usually requires surgical intervention (14, 15, 16), who have severe respiratory and/or gastrointestinal symptoms. Thoracotomy should be carried out on the side of the smaller or atretic aortic arch and on the side of the persistent ductus arteriosus or ligamentum arteriosum in symptomatic patients. Because of having

the mild clinical symptoms we didn't consider surgical intervention in our patient. Actually, she has been clinically followed up in our clinic without any complication.

Previous studies have shown that associated cardiovascular anomalies are uncommon. When present, the associated cardiac defects are usually cyanotic, including tetralogy of Fallot and transposition of great arteries (10, 13). Other commonly associated cardiac defects include coarctation of the aorta, ventricular septal defect, atrial septal defect, PDA and chromosomal anomalies such as DiGeorge and trisomy 18, 21 and also extracardiac anomalies, esophageal atresia and brachial sinus (10, 13, 17, 18). In our case there was a dextrocardia and, DAA with mirror-image dextrocardia was rarely reported in the literature.

Dextrocardia is defined as a right-sided heart with a base-apex axis directed rightward, resulting from a variation in cardiac development and has four common types; classic mirror-image dextrocardia, normal heart displaced to the right side of the chest, L-TGA, and single ventricle (1, 7). Dextrocardia may be associated with asplenia and polysplenia syndromes with situs ambiguus and complicated cardiac defects. Classic mirror-image dextrocardia is composed of situs inversus, atrioventricular concordance, ventriculoarterial concordance and normally related great arteries (1). In our patient, we have detected classic mirror-image dextrocardia without any associated anomalies.

In conclusion, DAA is an important cause of persistent respiratory symptoms in infants and children, and early investigations should be initiated to prevent mortality and morbidity. When there is clinical evidence of a vascular ring; inspiratory laryngeal stridor, frequent attacks of respiratory tract infection, cough since birth, and dysphagia; physicians should investigate for double aortic arch. Also, findings on chest radiography suspicious of a vascular ring should be correlated with clinical symptoms, and further investigations are needed.

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