

## Sol dominansı olan çift arkus aorta: Olgu sunumu

### Double aortic arch with dominant left arch: case report

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**Özet–** Vasküler halka aort arkusu ve dallarının trakea ve özefagusa bası yapan ve nadir görülen bir anomalisi-dir. Çift arkus aorta en sık görülen formu olup genellikle infant ya da erken çocukluk döneminde bulgu verir. Bu yazıda, tekrarlayan solunum yolu enfeksiyonu ve yutma güçlüğü nedeniyle kliniğimize başvuran yedi yaşındaki bir kız olgu sunuldu. Ekokardiyografide çift arkus aorta düşünüldü. Kardiyak bilgisayarlı tomografide çift arkus aorta tanısı doğrulandı. Kardiyak kateterizasyon ve bilgisayarlı tomografide sol arkusun daha gelişmiş olduğu görüldü. Cerrahi girişim uygulanan hastanın semptomları belirgin olarak geriledi. Tekrarlayan solunum yolu enfeksiyonları ve yutma güçlüğü olan hastalarda vasküler halka düşünülmelidir. Bu hastalarda erken tanı ve tedavi ile tam iyileşme sağlanabilir.

**Summary–** A vascular ring is defined as an anomaly of the great arteries (aortic arch and its branches) that compresses the trachea or esophagus. Double aortic arch is the most common vascular ring. Double aortic arch is very rare and typically becomes symptomatic in infancy or early childhood. We present a 7-year-old girl admitted to our clinic for evaluation of recurrent respiratory infection with dysphagia. Double aortic arch was suspected from echocardiography and diagnosed with cardiac computed tomography Left aortic arcus was larger than the right at computed tomography and cardiac catheterisation. After surgery the symptoms improved strikingly We conclude that vascular ring should be considered in the patients presenting with recurrent pulmonary infections and dysphagia. Early diagnosis and treatment may prevent chronic, irreversible complications.

Vascular rings are seen at an incidence of less than 1 %, among congenital cardiovascular anomalies. Double aortic arch is the most frequently vascular ring anomaly which is firstly reported by Wolman in 1939. The first successful surgical treatment of this anomaly was performed in 1945 by Dr. Robert Gross.<sup>[1,2]</sup> Because of inadequate regression of both right, and left aortic arches, right, and left arches completely surround, and compress trachea, and esophagus leading to respiratory distress, and nutritional problems in the early infancy. In normal individuals right 4. arch regresses, and left 4. arch constructs left aortic arch.<sup>[3]</sup>

In this paper, preoperative echocardiograms, and radiograms, and successful surgical treatment of a case with double aortic arch were presented.

### CASE REPORT

It was learnt that a 7-year female patient had been complaining of respiratory distress beginning from her neonatal period, and hospitalized frequently with the diagnosis of bronchiolitis. At the same time she had been experiencing difficulties while swallowing solid foods. Treatment was initiated with presumed diagnosis of bronchial asthma. Because of persistence of her complaints, she was referred to the departments of pediatric cardiology, and chest diseases with the initial diagnosis of vascular anomaly, and a foreign object within her respiratory tract Her physical examination on admission, rhonchi were heard over both lungs, and prolonged expiratory phase was detected. Physical examination findings of other bodily systems, and biochemical values were within normal limits. On her echocardiograms suprasternal short-axis views disclosed a double aortic arch (Figure 1).

Abbreviation

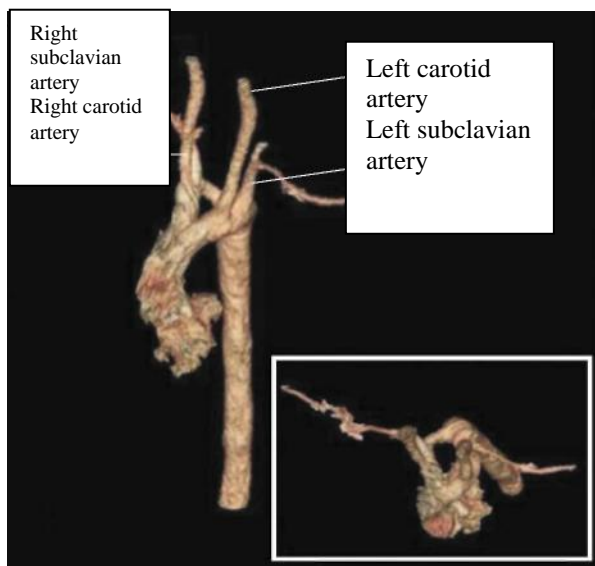
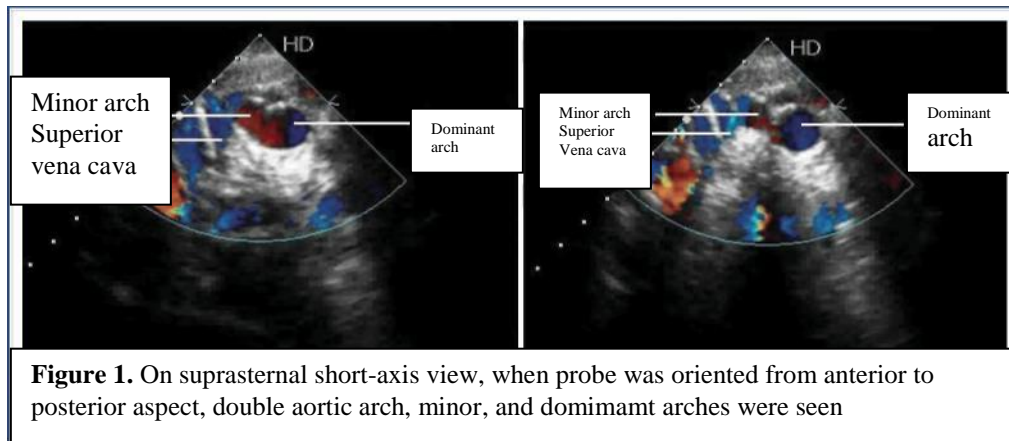
CT computed tomography

Submitted on: 01.03. 2012 Accepted for publication on: : .04. 03. 2012

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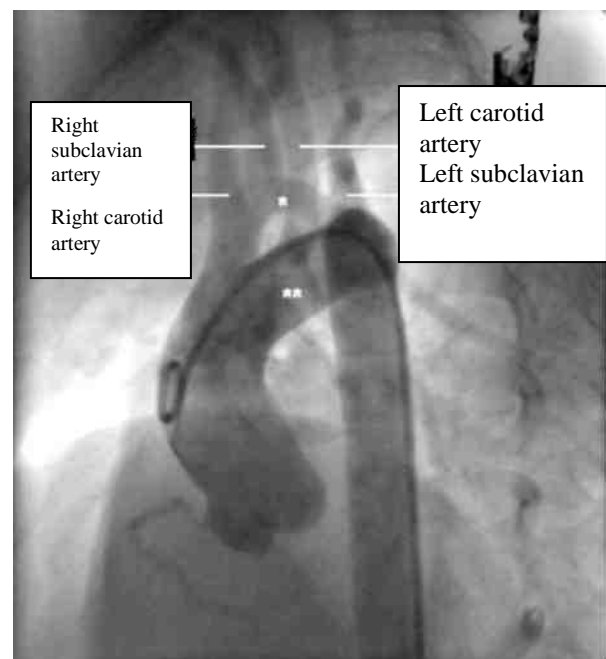
**Figure 2.** Volume rendering image) On CT angiographic examination, through anteroposterior, and superior views, origins of complete vascular ring, and cephalic vessel are seen

Computed tomography was obtained from the patient with a presumed aortic double arch, and diagnosis of double aortic arch was confirmed. From the right aortic arch, right carotid artery, and right subclavian artery originated in that order (Figure 2). Catheter angiograms were obtained to detect additional cardiac anomalies. Angiograms demonstrated origins of double arch, and cephalic vessels. Any additional cardiac anomaly was not detected (Figure 3). Barium esophagographic examination was performed, and esophageal impingement was demonstrated (Figure 4). As a result of these examinations the case was diagnosed as double aortic arch with a dominant left arch, and referred to surgery. Through a right posterolateral thoracotomy thoracic cavity was entered, minor aortic arch, and descending thoracic aorta were demonstrated. For the occlusion test, right aortic arch was clamped.

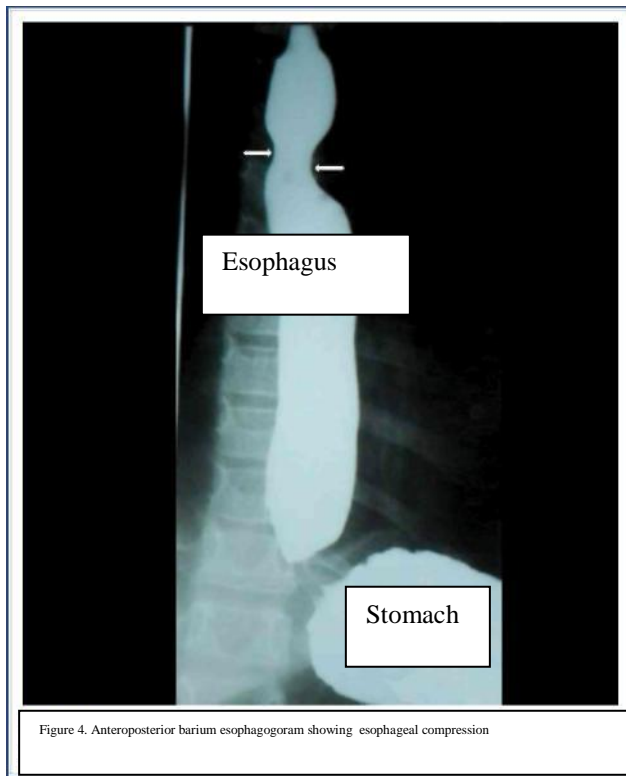
After clamping, pressure gradient was not detected, and any hemodynamic change did not occur. Before removing the clamp, the artery was resected, and both cut ends were sutured (Figure 5a, b) The patient was discharged on the 5. postoperative day with cure. During 18 month-postoperative follow-up, wheezing, and respiratory distress disappeared, and dysphagia was improved. The patient is asymptomatic and still under our supervision

## DISCUSSION

Embryologic development of vascular rings was described by Edwards, and the first successful double aortic arch surgery was realized by Gross.<sup>[2,3]</sup>

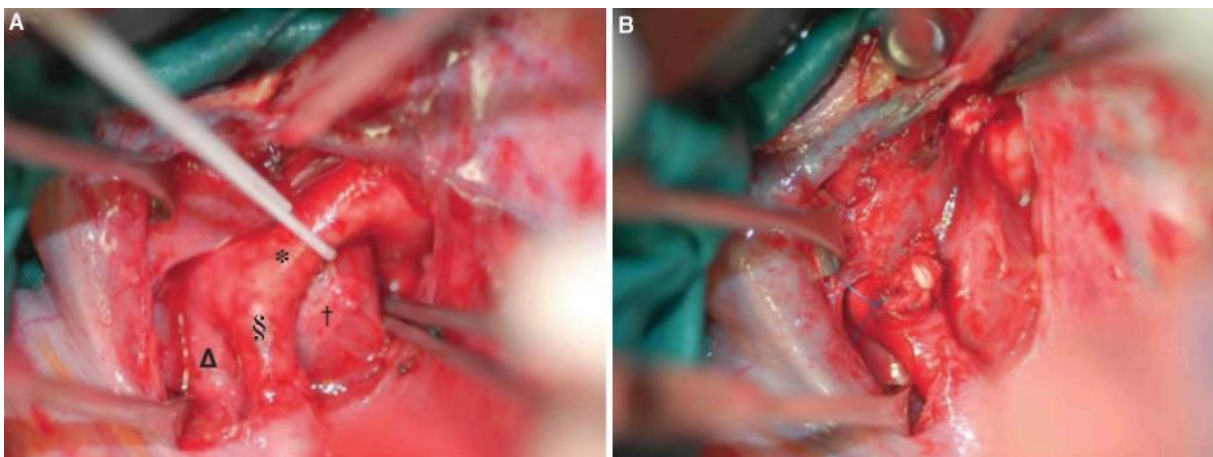


**Figure 3.** On aortic root angiogram obtained with the aid of a catheter passed through dominant aortic arch while the patient was in the left anterior oblique position, origins of vascular ring, and cephalic vessel are seen.. \* Minor arcus; \*\* Dominant arcus



Aortic arch is the most frequently encountered vascular ring which develops because the right 4. aortic arch does not regress during embryologic life. In this anomaly, ascending aorta divides into right, and left aortic arches. Right arch is situated on the posterior, and left arch on the anterior aspect of the aorta, and they compress trachea from the anterior, and esophagus from the posterior. This anomaly forms a tight vascular ring. Both arches are patent, and frequently right arch dominance is seen.<sup>[3]</sup>

However in our case left arch dominance was detected. The most frequently seen symptoms in vascular ring anomaly is inspiratory, and expiratory wheezing, and respiratory distress. Symptoms usually emerge during the neonatal period. Backer et al.<sup>[14]</sup> had operated on 61 patients, and reported onset of symptoms during the first postnatal six months in 92 % of their patients. Our case suffered from respiratory distress since her birth, and she was followed up with diagnoses of bronchial asthma, and foreign substance inside her respiratory tract, and treated accordingly. Double aortic arch is generally an isolated anomaly as is the case with our patient. It may be rarely accompanied with Fallot tetralogy, and transposition of the great vessels.<sup>[3]</sup> Predominantly, chest X-ray, and barium esophagography are used to demonstrate vascular ring, tracheal, and esophageal compression. In our case, barium esophagograms obtained to support the diagnosis demonstrated luminal stenosis because of bilateral impingement with a more prominent left-sided compression. Angiographic examinations were performed to confirm the diagnosis, and detect additional cardiac anomalies. However aortography fails to delineate morphological characteristics of aortic arch anomalies, while CT, and magnetic resonance imaging (MRI) can provide more useful information.<sup>[5]</sup> In our case, both preoperative angiographic, and CT examinations were performed to demonstrate relationships of vascular structures, detect additional anomalies, and plan the surgical procedure. In these cases, diagnostic bronchoscopy has a limited use, but it can be performed for differential diagnosis. In our case, bronchoscopy was performed in another medical center for presumed escape of a foreign substance into respiratory tract, and tracheal compression was observed.



**Figure 5. (A)** After right posterolateral thoracotomy, cranial view of the operation field starting from minor arch, right carotid artery, origin of right subclavian artery, and esophagus (B) After resection of minor arch, esophagus was released from its attachments\* Minor arcus; f Esophagus; § Right subclavian artery; Δ Right carotid artery.

During double aortic arch operations, the patient is placed in the right or left decobitus position based on the laterality of the aortic arch. The purpose in surgical treatment is to divide hypoplastic arch from its junction with descending aorta. Besides, ligamentum arteriosum should be resected, and complete release of trachea, and esophagus from their attachments should be ensured. Occlusion test must be performed to decide which aortic arch is the dominant one.<sup>[3]</sup> In the series by Alsenaidi et al.<sup>[6]</sup> in 81 cases with double aortic arch, right (71 %) and left (20%) aortic arch dominancy were detected. While 9 % of the cases were equally distributed. It was reported that during postoperative period of these patients, respiratory problems persisted frequently, while symptoms related to esophageal compression improved after surgical treatment. In our case during 18 months of follow-up period, complaints related to esophageal compression and respiratory problems were relieved.

In conclusion, vascular anomalies should be suspected in cases with respiratory distress, nutritional problems, and recurrent pulmonary infections starting from early years of life. With early diagnosis, and appropriate treatment complete cure can be achieved.

*Conflict of interest: none declared*

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*Anahtar sözcükler:* Aort, torasik/anormallikler/cerrahi; çift arkus aorta; çocuk, okul öncesi; doğuştan anormalliklerAanı; vasküler halka.

*Keywords:* Aorta, thoracic/abnormalities/surgery; double aortic arch; child, preschool; congenital abnormalities/diagnosis; vascular ring.