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2 **Clinical and Image Findings of Symptomatic Right**  
3 **Aortic Arch and Double Aortic Arch: Case Report**

4  
5 **ABSTRACT**

6 **Aim:** Anomalies of aortic arch are uncommon. They account for 1-3% of all congenital  
7 cardiac diseases. Vascular ring results from abnormal development of aortic arch complex.  
8 Generally, it manifests itself with tracheoesophageal compression symptoms during infantile  
9 period. we aimed to discuss clinical and imaging findings of the two cases with vascular ring  
10 with literature.

11 **Cases:** This article presents two cases of a 4-year-old child diagnosed with right sided aortic  
12 arch and a newborn child diagnosed with double aortic arch in neonatal unit. The first patient  
13 was complaining of coughing, wheezing and shortness of breath while the second patient  
14 was respiratory distress. Both cases diagnosis was made by multi-detector computed  
15 tomography MDCT. Two patients were diagnosed vascular ring with existing findings and  
16 referred for surgery

17 **Discussion:** If airway obstruction is distinct complaints emerge critically in early age. If  
18 symptoms are mild and there is no anatomic compression disease may be diagnosed in later  
19 years of life. The first case is four years old age who suffered cough and shortness of breath.  
20 Another was a neonate who suffered from respiratory distress . Vascular ring diagnosis can  
21 be made by chest radiography ,barium esophagus graphy, transthoracic echocardiography,  
22 MDCT, magnetic resonance imaging and angiography. But pathologies of our patients (right  
23 aortic arch ,double aortic arch, and tracheal compression) are seen effectively by MDCT and  
24 the post-processing images of MDCT

25  
26 **Conclusion:** Vascular ring should be considered in the event of recurrent lung infections,  
27 unexplained cough, wheezing, stridor in childhood period and unexplained respiratory  
28 distress in newborn childs. And MDCT is an excellent modality in patients suspected to have  
29 a vascular ring

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31 **Keywords:** Right aortic arch, double aortic arch, multidetector computed tomography , vascular ring,  
32 Symptoms, Congenital heart disease.

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34 **INTRODUCTION:**

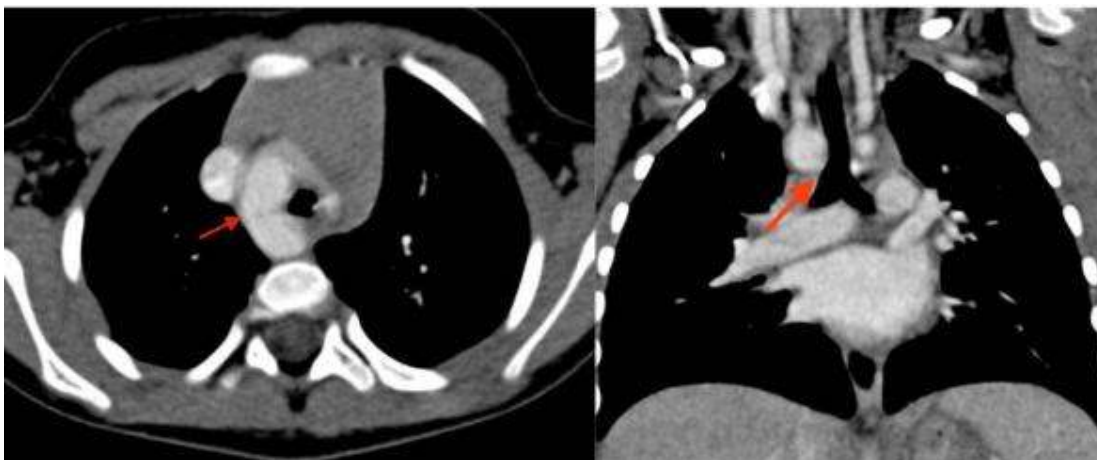
35 That aortic arch and its branches show a complex embryological development may cause many  
36 different types of malformations in this system (1,2). Abnormal relationships can be seen between  
37 tracheobronchial tree and vascular structures as a result of these malformations. In other words,  
38 vascular ring and secondary airway compression on it can be observed. Vascular ring is an important  
39 and unusual reason causing symptoms such as recurrent pulmonary infections, persistent wheezing  
40 and stridor during infancy and respiratory distress in newborns. These symptoms are associated with  
41 the degree of airway obstruction developing secondary to anatomic compression (3). **In this case**  
42 **study, we aimed to discuss the clinical and imaging findings of two cases with vascular ring that**  
43 **symptomatic and rarely appear.**

44

45 **CASES:**

46 A four-year-old female patient admitted to the chest diseases department of our hospital due to  
47 complaints of coughing, wheezing and shortness of breath. The patient's history showed that she had  
48 persistent shortness of breath and wheezing and her complaints increased when playing games and  
49 she had no accompanying systemic complaints. Results of the respiratory system and other systemic  
50 examinations of the patient were normal. According to the patient's medical background, the patient  
51 and her family had no history of atopy. Results of the complete blood count and biochemical tests of  
52 the patient were within normal limits.. Right sided aortic arch and tracheal compression from this right  
53 sided aortic arch (Figure 1, 2) was observed during the examination of the thoracic MDCT taken in our  
54 radiology department ( kV: 100, mAs: 72, reconstruction section thickness 1.0 mm, approximate CT  
55 dose: 2,91 mSv ). Also, it was observed that the left subclavian artery and the left common carotid  
56 artery (the left innominate artery) developed out of the aortic arch as the first branch with a shared  
57 root. Other branches were the right common carotid artery and the right subclavian artery, respectively  
58 (Figure 3). **A surgical operation for vascular ring as identified with the existing findings was planned,**  
59 **but the patient did not accept the surgery.**

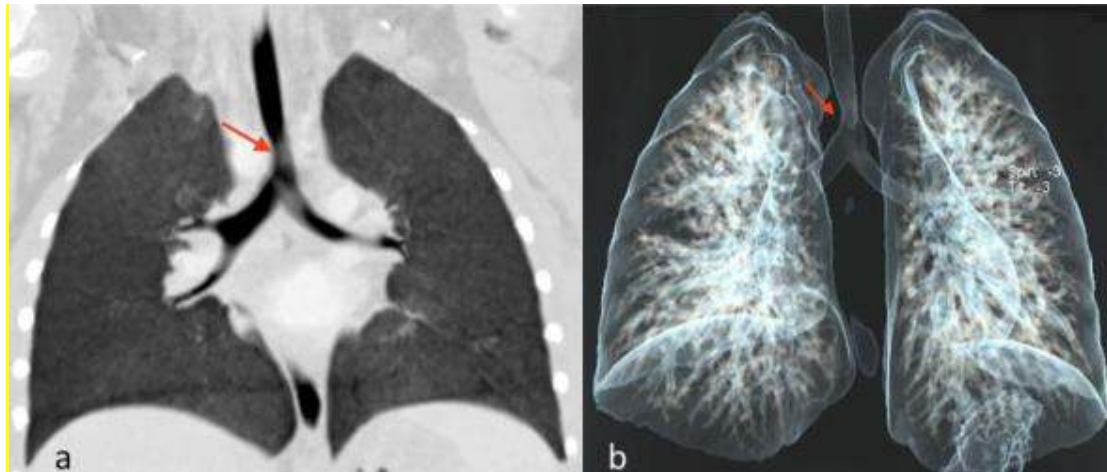
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62 Figure 1: Axial **Computed Tomography** scan shows a right aortic arch **(left image)** and tracheal  
63 compression **(right image)**

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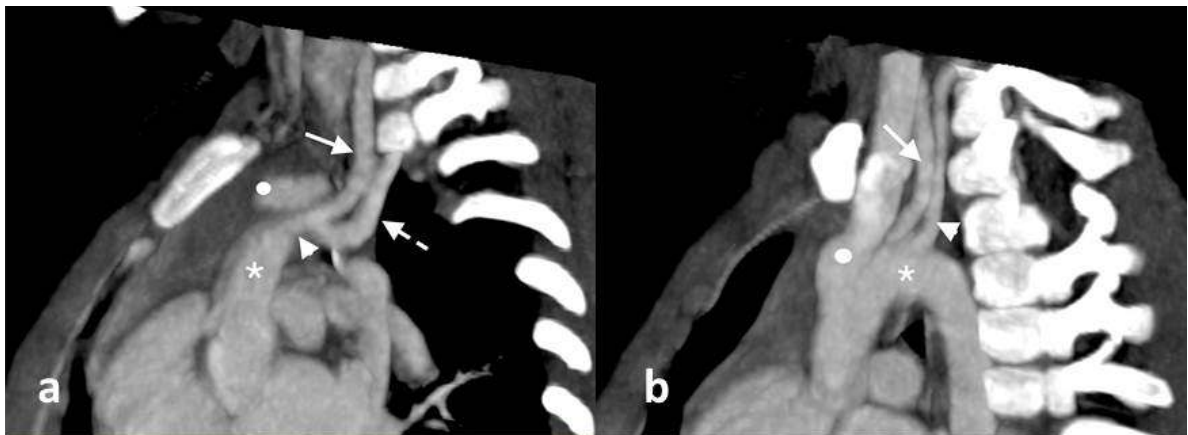
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67 **Figure 2: a) Tracheal compression (arrow) is seen on MinIP image b) Coronal volume-rendered (3D)**  
68 **image of the airway shows narrowing of the airway (arrow) by the right aortic arch**

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71

72 **Figure 3: a) Ascending aorta (asteriks) , left innominate artery (arrowhead) , left main carotid**  
73 **artery (arrow) and left subclavian artery (dashed arrow) are seen MIP sagittal image. b) Aortic**  
74 **arch (asteriks) , right main carotid artery (arrow) and right subclavian artery (arrowhead) are seen**  
75 **other MIP sagittal image. White round is vein in both images.**

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79 The second case was a newborn male infant who had been intubated and monitored from the  
80 birth in the paediatric intensive care unit of our hospital. The case that was the first pregnancy of a 26-  
81 year-old mother was born in the 27th week of the pregnancy. The mother did not have any specific  
82 condition. The case was delivered transvaginally in 1300 gr. The patient who was intubated due to  
83 respiratory distress was initiated an antibiotic therapy with the preliminary diagnosis of pneumonia. It  
84 was attempted to extubate the patient who recovered clinically and radiologically a week later.  
85 However, the patient was re-intubated due to decrease in saturations and respiratory distress **Chest X**  
86 **ray was normal (Figure 4). A MDCT angiography was taken for aorta due to suspected vascular ring.**  
87 **( kV: 80, mAs: 28, reconstruction section thickness 1.0 mm, approximate CT dose: 0,36 mSv ).**  
88 **Findings of a Double aortic arch, four artery sign (figure 5) and tracheal compression were discovered**  
89 **on the cross sections. 3D volume-rendered displayed the ring and sizes of the arches effectively**  
90 **(Figure 6). The patient was referred to surgery. The patient had a surgical division of the right arcus**  
91 **aorta and the compression on the trachea and oesophagus was eliminated.**

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94 **The appropriate ethics committee has approved this study and the patients have provided written**  
95 **informed consent.**

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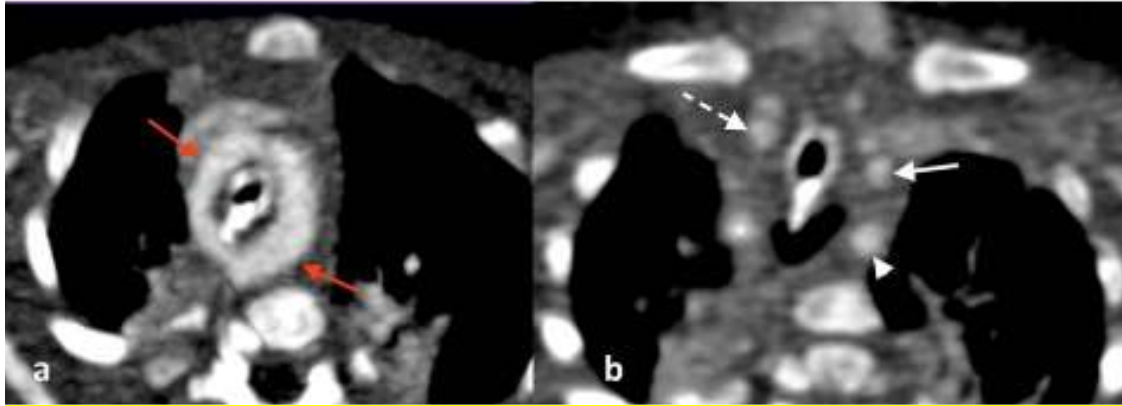
103 **Figure 4: There is no pathologic findings in chest radiography**

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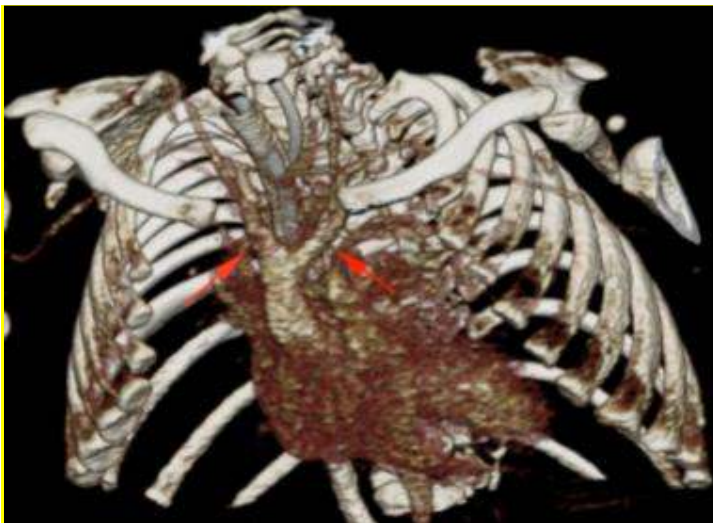
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109 **Figure 5: a) Axial MIP image shows double aortic arch encircling the trachea and oesophagus. b)**  
110 **'four-artery sign' which is characteristics findings of double aortic arch is seen on axial image. Asterix**  
111 **is right subclavian artery, dashed arrow is right main carotid artery, arrow is left main carotid**  
112 **artery and arrowhead is left subclavian artery.**

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115 **Figure 6: 3D volume-rendered image demonstrates the double aortic arch (arrows).**

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## 120 DISCUSSION

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122 Vascular ring, a congenital anomaly, is a defect in which trachea and oesophagus is surrounded by  
123 blood vessels-ligamentous structures developed out of this arch system and remains under pressure

124 as a result of abnormal development of the branchial arch system (4). Vascular ring results from lack  
125 of normal involution of vascular structures forming the aortic in embryonic life arch or from occurrence  
126 of involution of these vascular structures in different sites (5). This defect which accounts for 1-2% of  
127 congenital cardiac diseases is one of the important factors causing airway obstruction neonatal and  
128 childhood period.

129

130 Severity of clinical symptoms varies by the degree of compression (6,7). Dyspnea, stridor, wheezing  
131 and cough are the most common symptoms (6). Vascular ring may both cause severe respiratory  
132 distress developing immediately after birth and be asymptomatic for life (6). The first case was a 3-  
133 year-old patient with complaints of cough and wheezing. The other case was a newborn with  
134 respiratory distress developed immediately after birth and related clinical symptoms.

135

136 Although there are different classifications for vascular ring, more than 95% of these classifications are  
137 grouped in four main categories (Table 1)(8). The most common is double aortic arch anomaly (9).  
138 Typically, both arches are patent and right arch is mostly dominant in this anomaly that forms a tight  
139 ring almost always (2). This anomaly usually shows a clinical picture that is more severe and earlier  
140 than other vascular ring types.. Two arches originate from the ascending aorta, cross on either side of  
141 the trachea-oesophagus and join the descending thoracic aorta. (9). The right (posterior) arch is  
142 retroesophageal. Usually one arch is dominant and the other arch is smaller or may be atretic (9,10).  
143 In our case left side lumen was a little wider than the other one. We did not observe stenosis or  
144 occlusion at any level. This anomaly can be accompanied by cardiovascular anomalies such as  
145 tetralogy of fallot most commonly, coarctation of aorta and patent ductus arteriosus (9,10). No  
146 additional cardiac abnormality was identified in our case.

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**Table 1.** International Nomenclature and Database Conferences for Pediatric Cardiac  
Surgery(1998-1999)

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- I-Double arch aort
  - II-Right arch aorta + left lig.arteriosus
  - III-Innominate artery compression
  - IV-Pulmonary artery sling
- 

148 \*This 4 groups comprises of more than 95% of all vascular rings.

149

150

151 The second most common type of vascular ring anomalies is right sided aortic arch. This anomaly is  
152 classified into three groups (11). The most common branching patterns of the right aortic arch are the  
153 mirror image branching pattern( type II)(9). In this type, the great vessels originate from the arch in the  
154 following order, left innominate artery, right common carotid artery and right subclavian artery(11). And  
155 this type has aberrant left subclavian artery. Type II right aortic arch anomaly was seen In our case. A  
156 right aortic arch can be associated with congenital cardiac malformations such as persistent truncus  
157 arteriosus, tetralogy of Fallot, and pulmonary atresia with ventricular septal defect. Tetralogy of Fallot

158 is seen in 30 % of this patients' group. But additional cardiac abnormality has not been seen in our  
159 case. Intact retroesophageal left ligamentum arteriosum, ductus arteriosus and Kommerell's  
160 diverticulum (an embryological diverticulum of proximal descending aorta) may also accompany right  
161 aortic arch anomaly.

162

163 Diagnosis of vascular ring is established utilizing chest radiography (Posterior-Anterior and Lateral),  
164 barium-contrasted oesophageal graph, transthoracic echocardiography (ECO), contrast enhanced  
165 MDCT, magnetic resonance imaging (MRI) and angiography. Today, ECO, MRI and contrast  
166 enhanced MDCT are the most preferred diagnostic methods (6,9,10). The PA chest radiography  
167 shows that there is a single or double-sided emphysema due to compression and a right or left-sided  
168 tracheal compression, while the lateral chest radiography shows a compression on the anterior of the  
169 trachea. Alsenaidi et al. (12) found in their study conducted with 81 patients that chest radiographies  
170 were normal in 20% of the cases. We also did not observe any significant characteristic in chest  
171 radiographies of the cases in our study. It can be seen in the barium-enhanced oesophagus  
172 radiographies that oesophagus is exposed to anterior, posterior and lateral compression. Indentations  
173 caused by compression can be at different levels and in different sizes (6,10). No barium-enhanced  
174 examination was performed for the both cases. PA chest radiography and barium-enhanced  
175 radiographies can confirm the diagnosis of vascular ring. However, cross-sectional imaging is usually  
176 required to define the exact morphology of the aortic arch malformation and for planning an optimal  
177 surgical approach (10).

178

179 The degree, level and length of the tracheal compression of the aortic arch can be clearly  
180 monitored by MDCT and MRI. The ability to show surrounding structures in addition to vascular  
181 structures and to provide images in three planes are the most important advantages of cross-sectional  
182 imaging (6,9). Mirror image branching of the right sided aortic arch and 'four-artery sign' and marked  
183 tracheal compression, which is the characteristic symptom of double aortic arch, have been clearly  
184 demonstrated in our case.

185

186 MDCT is a fast imaging method. Unlike MRI, it does not require sedation in most cases. Sedation can  
187 also cause serious problems in patients with airway obstruction (6). Patients that need airway  
188 assistance the instruments need to be MRI suitable whereas in CT imaging such concerns are not  
189 needed. MDCT provides more precise information for evaluating airways and lung parenchyma (10).  
190 Multiplanar reformat (MPR) and volume rendering (VR) reconstruction allows for examining images in  
191 each of the three planes and three dimensions (3D). 3D volume-rendered displayed the ring and sizes  
192 of the arches in our case effectively. In addition, minimal compressions on the trachea can be  
193 observed using the minimum intensity projection (MinIP). Also, these compressions can be examined  
194 through 3D methods that reveal only airways and parenchyma. We use these post-processing images  
195 and show tracheal stenosis distinctly. The most significant drawback is the use of x-ray, which use has  
196 a particular importance especially in paediatric period (6,9,10).

197

198 These parameters can be preferred good MDCT images in pediatric patients(9). Tube current : 0–3 kg–  
199 60 mAs/slice, 3–6 kg–80 mAs/slice, 6–10 kg–100 mAs/slice, 10–15 kg–120 mAs/slice . Tube voltage :  
200 80- 100 kV. Reconstructed slice thickness: 0.9 mm. 80 kV value and iterative reconstruction algorithm  
201 can be use to reduce CT doses. We used 80 kV in newborn patient whereas 100 kV in other patient.  
202

203 Conventional angiography is used to specifically determine topographic vascular anatomy. However,  
204 this method has disadvantages such as inability to determine atretic vascular structures, tracheal  
205 and/or oesophageal compression (6,10). It is important to perform ECO in order to exclude  
206 accompanying anomalies in suspected vascular ring (9).

207

208 Surgical intervention for vascular ring is recommended for symptomatic patients with airway  
209 compression identified radiologically (13). The surgeon must make a decision of approach method  
210 according to the vascular ring anatomy and the associated tracheal or cardiac anomalies (11).

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212

## 213 CONCLUSION

214 Vascular ring should be considered in the event of recurrent lung infections, unexplained persistent  
215 wheezing and stridor in childhood and persistent respiratory distress in newborns. And MDBC is an  
216 excellent modality in patients suspected to have a vascular ring. It allows evaluation of the aortic arch  
217 and its branches and provides valuable information for planning surgical management with multiplanar  
218 and three-dimensional (3-D) imaging.

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220

## 221 CONSENT

222

223 All authors declare that 'written informed consent was obtained from the patient (or other approved  
224 parties) for publication of this case report and accompanying images. A copy of the written consent is  
225 available for review by the Editorial office/Chief Editor/Editorial Board members of this journal  
226

## 227 ETHICAL APPROVAL

228

229 All authors hereby declare that all experiments have been examined and approved by the appropriate  
230 ethics committee and have therefore been performed in accordance with the ethical standards laid  
231 down in the 1964 Declaration of Helsinki."

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