

Prenatal Diagnosis of Right Aortic Arch with Mirror Image Branching: Importance of the Three-vessel-trachea View

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ABSTRACT Aortic arch anomalies are rarely diagnosed in the prenatal period. However, due to the higher risk of accompanying chromosomal aberrations, diagnosis of these structural anomalies is of utmost importance. Aortic arch anomalies are detected in the three-vessel-trachea view, which is extremely useful in diagnosing several cardiac disorders and can be visualized during fetal echocardiography. In addition, the presence of a retroesophageal vascular ring should also be checked since it determines postnatal prognosis, and the delivery should be planned in a tertiary center. We report a case of the right aortic arch with mirror image branching and discuss the prenatal diagnosis and management of this anomaly.

Keywords: Echocardiography; aortic arch syndromes; ductus arteriosus

Aortic arch anomalies are a rare occurrence during the development of the embryonic pharyngeal arch system, which results in its altered positioning and branching. The aortic and ductal arches originate from the fourth and sixth pharyngeal arches, respectively. Although the true incidence rate of aortic arch anomalies is unknown, it is estimated to be 1/1000 in the general population.¹ In a current prenatal case series, the incidence of the right aortic arch anomaly was reported to be 0.6% in the general population and 5% in the fetuses with congenital cardiac anomalies. Furthermore, the isolated right aortic arch anomaly accounts for 1.4% of all congenital cardiac anomalies.² It is the least diagnosed cardiac anomaly in the prenatal period, and information regarding its prenatal diagnosis is also sparse. The co-occurrence of right aortic arch and the right ductal arch is even rarer with an only limited number of case reports available.³⁻⁵ We present a case of prenatally diagnosed right aortic arch and right ductal arch with mirror image branching and discuss the prenatal diagnosis and management of the aortic arch anomalies

CASE REPORT

A 29-year-old, gravida 2, para 1 woman was referred to our maternal-fetal medicine unit at 34 weeks of gestation with suspicion of intrauterine growth restriction. Her family history was unremarkable and fetal biometry parameters and amniotic fluid volume were appropriate for the gestational age. Fetal cardiac examination revealed visceral and cardiac situs solitu, and both atri-

oventricular and ventriculoarterial connections were concordant. Three-vessel-trachea view revealed that the aortic arch and the ductal arch were located to the right of the trachea and connected to the descending aorta by forming the right V sign. The diagnosis of the right aortic arch and right ductal arch with mirror image branching was confirmed by the absence of aberrant left subclavian artery in the three-vessel-trachea view (Figure 1, 2). The thymus was visualized at anterosuperior part of the mediastinum in the transverse section of the aortic arch. A detailed sonographic examination revealed no additional anomaly. Prenatal chromosome analysis was offered to the patient in consultation with the medical geneticist and pediatric cardiologist, but the patient did not agree to an invasive procedure. At 38 weeks of gestation, a 3350-gram male baby was delivered spontaneously with APGAR scores of 8 and 9 at 1 and 5 min, respectively. The diagnosis of the right aortic arch and the right ductal arch with mirror image branching was confirmed by postnatal echocardiography performed 2 h after delivery. The post-natal 26th-hour echocardiography showed the closure of the right ductal arch and a retroesophageal ring was absent. The 2-month follow-up did not reveal any complication. A signed written consent was obtained from our patient.

DISCUSSION

Aortic arch anomalies are either obstructive or branching/laterality. The latter type includes left-sided aortic arch with the aberrant right subclavian artery, right-sided aortic arch with mirror image branching of the brachiocephalic arteries, right aortic arch with left ductus arteriosus, and double aortic arch.¹ According to the double aortic arch theory put forward by Edwards in 1948, the right and left aortic arches connect the ascending and descending aorta. The main carotid and subclavian arteries of each side originate from the right and left aortic arches; in normal embryogenesis, the right aortic and ductal arches regress and the left aortic and ductal arches preserve continuity. An aortic arch anomaly arises when the right aortic and ductal arches persist, and the left aortic and ductal arches regress. The regression of the left aortic arch till the



FIGURE 1: The fetal echocardiographic image. The transverse view of the right aortic arch.

L-PA: Left pulmonary artery; RAA: Right aortic arch; SVC: Superior vena cava; T: Trachea.

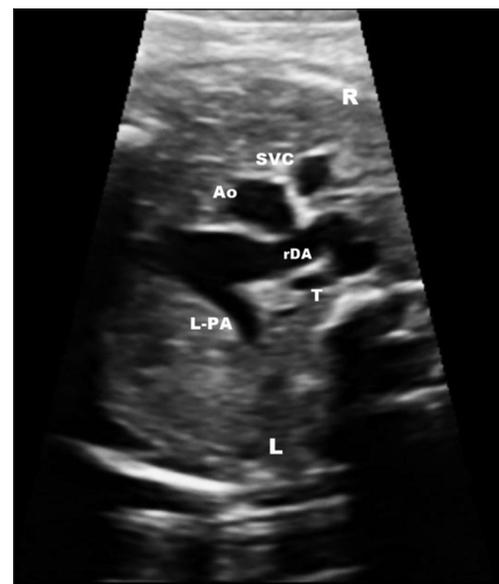


FIGURE 2: The fetal echocardiographic image. The transverse view of the right ductal arch.

L-PA: Left pulmonary artery; rDA: Right ductal arch; Ao: Aorta; SVC: Superior vena cava; T: Trachea.

origin of the left subclavian artery results in the persistence of the right aortic arch and forms the mirror image of the normal left-sided aortic arch. While the left brachiocephalic artery is the first branch of the right aortic arch, the right common carotid artery, and the right subclavian artery also originate from the right aortic arch.⁶

The right aortic arch with mirror image branching is seen in over 90% of complex cardiac anomalies, such as tetralogy of Fallot, pulmonary atresia with a ventricular septal defect, truncus arteriosus, absent pulmonary valve, and tricuspid atresia and double outlet right ventricle.^{3,7}

The aortic arch anomalies are associated with chromosomal aberrations, especially the 22q11 microdeletion and trisomy of 21. The frequency of 22q11 microdeletion is 5–25% in the isolated cases of the right aortic arch and increases up to 46% in the presence of an accompanying cardiac anomaly.^{3,8,9} Therefore, prenatal chromosome analysis should be offered, especially in the presence of an associated cardiac anomaly.

Although the exact cause(s) of the regression of the left aortic arch and the persistence of the right aortic arch are not fully understood, the blood flow pattern of the fetus and certain genetic factors have been implicated in the development and branching of the aortic arch. The 22q11 microdeletion, which is thought to affect the migration and development of the cardiac neural crest in the isolated right aortic arch cases, supports a genetic basis of the aortic arch etiopathogenesis.^{9,10}

The aortic arch is detected in the three-vessel trachea view, which is an axial view obtained in the upper mediastinum, and demonstrates the V-shaped connection of the ductal arch and the aortic arch to the descending aorta. In this view, the great vessels are located from left to right and in descending order of size from the ductal arch, the aortic arch, and the vena cava superior. The aortic and ductal arches are normally located to the left of the trachea and the vertebrae. The aortic arch anomaly is characterized by its presence on the right side of the trachea in the three-vessel trachea view. When the right aortic and right ductal arches are located to the right of the trachea, they appear as V-shaped and are referred to as “right V sign” in the three-vessel trachea view.

The retroesophageal vascular ring is a clinically significant feature of aortic arch anomalies since it may cause tracheal and esophageal compression, respiratory problem, and dysphagia. The

use of color Doppler sonography facilitates the recognition of the retroesophageal vascular ring, especially in the presence of double aortic arch.¹¹ The right aortic arch with mirror image branching does not cause retroesophageal vascular ring formation, and the latter is therefore not expected to affect the postnatal prognosis in the absence of associated structural and chromosomal anomalies.^{3,8,12} However, the tight vascular ring formed around the trachea and esophagus in the presence of double aortic arch, or the right aortic arch and left ductal arch causes compression symptoms. The differential diagnosis of aortic arch anomalies is therefore important in terms of neonatal prognosis. When these anomalies are diagnosed prenatally, delivery at a tertiary center is recommended and the intervention should be planned before the symptoms occur.^{8,12}

The determination of the visceral and cardiac situs facilitates the diagnosis of heterotaxy syndromes and aortic arch anomalies. In addition, our case demonstrated the importance of the evaluation of the three-vessel trachea view, since the four-chamber view is normal or almost normal in many cardiac anomalies. Nearly normal three-vessel-trachea view (right V sign) in the right aortic arch and the right ductal arch with mirror image branching may lead to a false negative diagnosis. Therefore, the order and size of the great vessels and their location relative to the trachea should be evaluated in the three-vessel-trachea view. Since the visualization of the three-vessel trachea plane is easier compared to that of the ventricular outflow plane, the former is gaining importance in the screening of cardiac anomalies. The diagnostic accuracy of the aortic arch anomalies increases as the three-vessel-trachea view becomes a standard part of the fetal cardiac examination.

The increasing awareness of the aortic arch anomalies with advanced visualization methods increases the chances of prenatal diagnosis. After diagnosis, once the possibility of associated cardiac and extracardiac anomalies is excluded, the expecting parents should be counseled regarding the associated chromosomal aberrations and offered prenatal chromosome analysis. The fetuses with

the aortic arch anomalies that cause retroesophageal vascular ring formation should be delivered in tertiary centers, and the newborn should be closely monitored for any respiratory problems. Even though the right aortic and ductal arch with mirror image branching does not lead to the formation of retroesophageal vascular ring, the parents should be informed that the double aortic arch and its complications cannot be excluded in the prenatal period, and the delivery must be planned in tertiary centers. Finally, the prenatal diagnosis should be confirmed postnatally.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Başak Kaya; **Design:** Başak Kaya; **Control/Supervision:** Deniz Kanber Acar; **Data Collection and/or Processing:** Salim Sezer, Helen Bornaun; **Analysis and/or Interpretation:** Başak Kaya, Deniz Kanber Acar; **Literature Review:** Salim Sezer; **Writing the Article:** Başak Kaya; **Critical Review:** Gökhan Yıldırım, Halil Aslan; **References and Fundings:** Salim Sezer; **Materials:** Salim Sezer.

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