



# Prenatal Diagnosis of Double Arcus Aorta Anomalies: Case Report

Tuncay Yüce\*, Erkan Kalafat, Acar Koç

## Abstract

**Introduction:** Double aortic arch is a rare cardiac anomaly that constitutes 1% to 3% of all congenital anomalies. Prenatal diagnoses of such conditions are rare.

**Case Presentation:** We present a case of double aortic arch with compression of the trachea that was diagnosed during the 20th week of gestation. The baby was delivered during the 29th week of gestation via cesarean and the diagnosis was confirmed with computerized tomography (CT) angiography. Later, a surgical repair was undertaken; however, the patient succumbed to pulmonary complications 3 months after delivery.

**Conclusions:** Aortic arch anomalies are a rare and diverse group of congenital heart anomalies. Prenatal diagnosis is possible and beneficial in cases with tracheal compression as it allows for early intervention after birth.

**Keywords:** Congenital anomalies, Cardiac, Prenatal diagnosis, Ultrasonography

## Introduction

Intrauterine development of the aortic arch begins at 4 weeks. A defect at any part of this development period causes aortic arch anomalies. Aortic arch anomalies constitute 1% to 3% of pathologies of all congenital heart diseases. Aortic arch anomalies are 1.4 to 2 times more common in the male population (1).

After Achiron et al (2) proposed routine incorporation of the outflow tracts and the three-vessel and tracheal view (3VT) into the screening for fetal abnormalities, double aortic arch anomalies can be diagnosed prenatally (2). However, the sensitivity for detection of congenital heart diseases varies from 48% to 63% in some series (3). Ultrasound is used for prenatal diagnosis of aortic arch pathologies. Magnetic resonance imaging (MRI) scanning is another important diagnostic method for prenatal cardiac disease (4).

In the case of a delay in prenatal diagnosis of aortic arch pathologies, condition can be diagnosed postnatally after the onset of stridor, wheezing and feeding problems, especially in the first year.

Developmental cardiac pathologies, such as ventricular septal defect, transposing of big arteries, fallot tetralogies and aortic coarctation, cleft lip and palate, subglottic stenosis or DiGeorge syndrome, can be associated with aortic arch pathologies (5).

According to a Medline literature search, our case is important because only a few cases have been diagnosed solely by ultrasound in the early weeks of pregnancy and confirmed by a computerized tomography (CT) angiography after the first postpartum week.

## Case Presentation

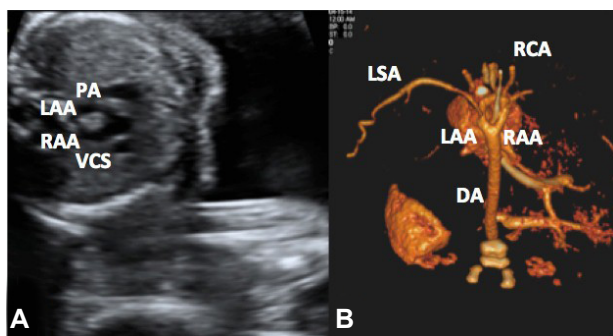
A 29-year-old woman, gravida 2 parity 1, came for her first prenatal visit at 9 weeks of pregnancy. The woman had no history of any genetic disorders. First and second trimester screening tests showed low risk for trisomies. Arcus aorta pathology was diagnosed at a fetal heart screening, which was done at the 20th week during a detailed ultrasound examination. In the three vessels and trachea image, it was determined that a double aortic arch completely surrounded the trachea (Figure 1A). There were no additional cardiac anomalies. Amniocentesis was offered but patient did not consent to the procedure. The patient had vaginal bleeding at 26 weeks and 6 days of pregnancy. She was hospitalized, and after 3 days, she was discharged from the hospital. At 29 weeks and 3 days of pregnancy, the patient was hospitalized again because of premature rupture of membranes (PPROM). After 2 doses of betamethasone and antibiotics, she delivered a baby via C-section, and the baby was intubated for impaired oxygenation due to compression of the trachea. CT angiography was taken after the baby's condition stabilized and the prenatal diagnosis of double aortic arch was confirmed (Figure 1B). Right carotid and subclavian arteries from right arch proximal and left carotid communis and left subclavian arteries from left aortic arch were observed. The baby had an operation on the 13th day. During surgery, the right aortic arch was divided after given branch of right carotid communis and right subclavian arteries. After the procedure, the baby was extubated and its condition stabilized. Because of a pulmonary infection, the baby died 3 months after delivery.

Received 13 February 2015, Accepted 9 October 2015, Available online 1 January 2016

Department of Obstetrics and Gynecology, Ankara University, Ankara, Turkey.

\*Corresponding author: Tuncay Yüce, Faculty of Medicine, Ankara University, Cebeci Campus, Mamak/Ankara, Turkey. Tel: +90 5055943521, Email: [drtuncayyuce@gmail.com](mailto:drtuncayyuce@gmail.com)





**Figure 1.** A) The 3-vessel image of fetal echocardiography. B) Computed tomography (CT) angiogram with 3-dimensional reconstruction. PA; pulmonary artery, LAA; left aortic arc, RAA; right aortic arc, VCS; superior vena cava, DA; descending aorta, RCA; right carotid artery.

### Discussion

Normally, the aorta stems from left ventricle and is minimally angled to the right. The aorta consists of three parts: the ascending aorta, the aortic arch and the descending aorta. The trachea remains to the right side of the aorta. The aortic arch can be seen in an ultrasound examination at 14 weeks. However, as in the case of any anomaly, like a double arcus aortic arch or one-sided aortic arch, a patient must wait until the end of the 20th week of pregnancy for a certain diagnosis (6,7).

Yoo et al published a retrospective analysis of aortic arch pathologies in 2010. In 75 patients with a right-angled aortic arch, only 4 cases had a double arcus aortic arch like our case (8).

Double arcus aortic arch cases are difficult to diagnose prenatally. This condition is rare, but the ultrasound technique for showing aortic arch pathologies is also difficult. Because both arches cannot be visualized simultaneously, discordance in size or sometimes lack of continuity may be misinterpreted (6).

Double aortic arch patients have long-term respiratory problems like stridor and wheezing because of tracheal pressure, and feeding and swallowing problems because of oesophageal pressure; these are the most common causes of morbidity. Because of these problems, early diagnosis of double arcus aorta is becoming more important. In addition, these patients must be screened for associated anomalies, such as 22q11 deletion (DiGeorge syndrome) and facial anomalies (5).

After the fetal echocardiography (three vessels and trachea image), the double aortic arch anomaly was detected and no additional anomalies were found. The patient had undergone a C-section because of PPRM, and the baby was intubated as quickly as possible due to tracheal pressure of the double aortic arch. After the baby's condition stabilized, an operation was planned, and the baby was extubated. All these procedures were quick

because of the prenatal diagnosis.

When performed by a fetal cardiologist, fetal echocardiography will eventually be enough to diagnose a double arcus aortic arch. If fetal heart screening is insufficient, MRI or CT angiography after labour can be used when stridor, difficulty swallowing and feeding problems are encountered with no obvious cause.

### Ethical Issues

The authors have obtained permission before using patient data and images.

### Conflict of interests

Authors of this publication declare no conflict of interest.

### Financial support

None to be declared.

### Acknowledgments

None to be declared.

### References

1. Shah RK, Mora BN, Bacha E, et al. The presentation and management of vascular rings: an otolaryngology perspective. *Int J Pediatr Otorhinolaryngol.* 2007;71(1):57-62. doi:10.1016/j.ijporl.2006.08.025.
2. Achiron R, Glaser J, Gelernter I, Hegesh J, Yagel S. Extended fetal echocardiographic examination in detection of cardiac malformations in low risk women. *BMJ.* 1992;14;304(6828):671-674.
3. Patel CR, Lane JR, Spector ML, Smith PC. Fetal echocardiographic diagnosis of vascular rings. *J Ultrasound Med.* 2006;25(2):251-257.
4. Dong SZ, Zhu M, Li F. Preliminary experience with cardiovascular magnetic resonance in evaluation of fetal cardiovascular anomalies. *J Cardiovasc Magn Reson.* 2013;21:40. doi:10.1186/1532-429X-15-40.
5. McElhinney DB, McDonald-McGinn D, Zackai EH, Goldmuntz E. Cardiovascular anomalies in patients diagnosed with a chromosome 22q11 deletion beyond 6 months of age. *Pediatrics.* 2001;108(6):E104.
6. Galindo A, Nieto O, Nieto MT, et al. Prenatal diagnosis of right aortic arch: associated findings, pregnancy outcome, and clinical significance of vascular rings. *Prenat Diagnosis.* 2009;29(10):975-981. doi:10.1002/pd.2327.
7. Jain S, Kleiner B, Moon-Grady A, Hornberger LK. Prenatal diagnosis of vascular rings. *J Ultrasound Med.* 2010;29(2):287-294.
8. Yoo SJ, Min JY, Lee YH, Roman K, Jaeggi E, Smallhorn J. Fetal sonographic diagnosis of aortic arch anomalies. *Ultrasound Obstet Gynecol.* 2003;22(5):535-46. doi:10.1002/uog.897.