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## Prenatally-Diagnosed Double Aortic Arc with Right Dominance: A Case Report

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### Introduction:

Double Aortic Arc (DAA) is a rare vascular ring form in which the trachea and esophagus are completely surrounded by right and left aortic arcs. The frequency is one in 2000-4000 pregnancies. It is extremely rare to detect with Fetal Echocardiography (ECHO). It is also the most common form of vascular rings, and usually shows symptoms in infant or early childhood period. In symptomatic cases, full recovery can be ensured with early diagnosis and treatment. Here, a case with DAA that was diagnosed in fetal ECHO will be presented, which has a very low prevalence of prenatal diagnosis.

### The Case:

A 21-year-old mother gave birth by cesarean section to a first living baby of first pregnancy born 3210 g at 38<sup>+4</sup> weeks. The baby was hospitalized with a pre-diagnosis of aortic archus. System examinations of the patient was normal: Oxygen saturation: 95%, respiratory count: 55/min, body temperature: 36.5°C. In the fetal echocardiographic examination during the 35<sup>th</sup> gestational week, there was aberrant vascular structure (minor archus) which separated from the aorta-proximal transverse archus line and crossed the trachea from left anterolateral, and it was considered that the patient had double aortic arc in right aortic arcus (major/dominant) on the right of the trachea in normal calibration (Figure-1A, B). In the first 1-hour after the birth, it was confirmed that the patient, who was evaluated by the Pediatric Cardiology Unit, had Double Aortic Arc in the ECHO and in the examination of high parasternal section (Figure-1C, D). Computed Tomography was carried out to the patient who was followed up with the mother in terms of possible unnoticed compression findings. CT examination revealed double arcus aorta on the anterior trachea, and the patient was reported as not having any signs of vascular compression (Figure-2). The patient was discharged with recommendations, and was followed-up at the Pediatric Cardiology Clinic with information on the symptoms that might develop.

### Discussion:

Vascular rings constitute less than 1% of congenital heart anomalies. Double Aortic Arc was reported by Wolman in 1939 for the first time. The first successful surgical repair of vascular rings whose embryological origins were reported by Edwards was performed by Gross for the first time (1-2). In normal embryonic development, while right-side 4<sup>th</sup> archus regresses, the left-side 4<sup>th</sup> archus creates normal archus by proceeding (3). In this anomaly, which occurs as a result of the insufficient regression of aortic archus, the cases become symptomatic as a result of the compression of the ring-forming vascular bodies on the trachea and esophagus causing respiratory distress and nutrition problems in newborn and early infant period. In our case, the archus was divided into right and left arches, and after crossing the trachea from the front and the esophagus from behind, it merged to form the descending aorta. In our case, the front minor archus formed the left archus, and the dominating archus was located on the right, which is reported in the literature mostly as right-dominant archus (3, 4).

The most commonly seen symptom in vascular ring anomalies is inspiratory and expiratory wheezing and respiratory distress, which may appear at early stages like neonatal period. Full

correction operation is carried out in patients who are symptomatic due to compression by eliminating the vascular compression by the dissolution of the minor archus in patients without perfusion loss after occlusion test (5). Since vascular ring anomalies are mostly isolated anomalies, if the doctor sees that only intracardiac structures are normal in the evaluation of obstetric ultrasound in prenatal period does not rule out the diagnosis. It is important that the doctor is careful about the aberrant vascular structures. Vascular ring and other accompanying cardiovascular anomalies may be detected with a detailed fetal echocardiographic examination. Our patient was diagnosed with a double aortic arc in the fetal echo examination in 35<sup>th</sup> gestational week and was followed-up, and the diagnosis was confirmed with early postpartum echo examination. In the computed tomographic examination of thoracic aorta to clarify vascular compression and anatomy, it was seen that the minor archus in the left anterior made a ring; however, in its current form, it did not cause compression. Morbidity and mortality can be reduced by preventing delayed diagnosis and treatment of postpartum patients by detecting these patients in the prenatal period.

### **Result:**

Double AA causes compression in the trachea and esophagus, and results in various symptoms like respiratory obstruction, difficult swallowing, chronic wheezing, vomiting and aspiration. Late diagnosis is frequent in these cases, and may increase permanent structural damage to the trachea because of permanent compression. In cases that do not have compression symptoms, as it was the case in our patient, prenatal diagnosis of experienced perinatologists and pediatric cardiologists is important in preventing delayed diagnosis. Complete recovery can often be achieved with early diagnosis and treatment in these patients.

### **References:**

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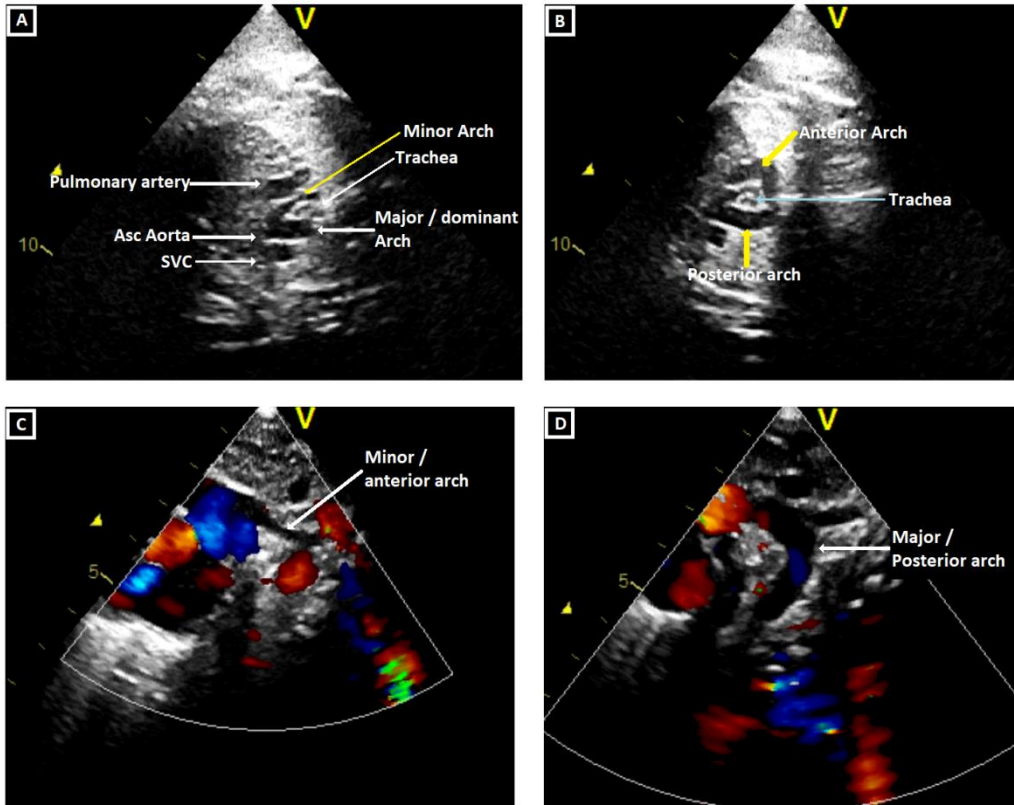


Figure-1. Prenatal Fetal echocardiographic examination shows the dominant arch extending from the aorta to the right trachea, and separated from the ascending aorta and aberrantly following the tracheal left anterior (A). In the posterior tilt of the probe, it is seen that both arches combine in the posterior to form the arch of the pattern and surround the trachea in a ring-shaped manner (B). In the postnatal echocardiographic examination, a minor arch (3,4mm) (C) and a well-developed dominant arch is seen in the suprasternal section (D).

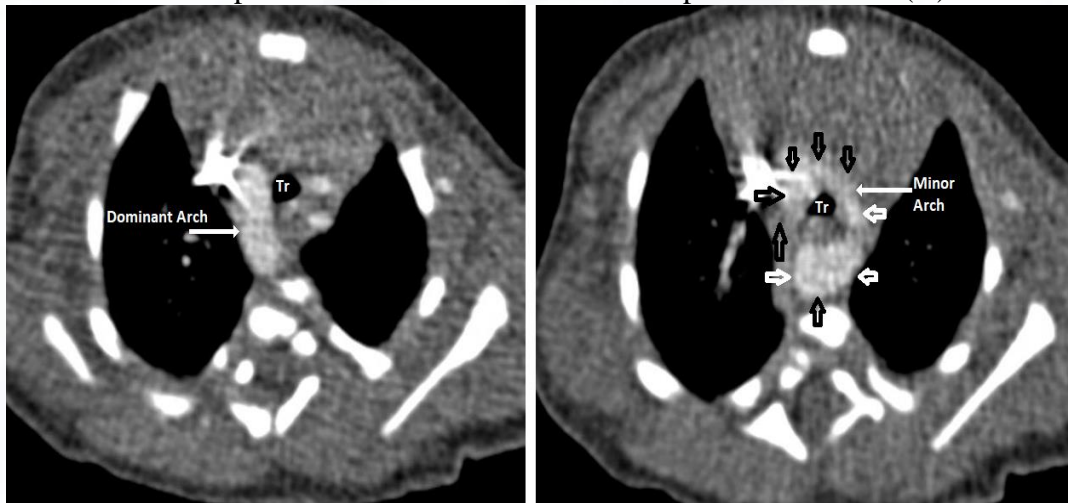


Figure-2. Postnatal thoracic CT angiographic examination revealed that the dominant arch located in the right posterior enveloped the trachea (Tr) via the left and anterior arches, and the two arches joined together to form the descending aorta.