NASCER E CRESCER
BIRTH AND GROWTH MEDICAL JOURNAL
year 2025, vol 34, n.º 2

Nascer e Crescer - Birth and Growth Medical Journal
2025;34(2):90-92. doi:10.25753/BirthGrowthMJ.v34.i2.35493

IMAGING CASES

Right aortic arch: the value of the upper mediastinum view in fetal echocardiography

Arco aórtico direito: o valor do plano do mediastino superior em ecocardiografia fetal

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The authors present the case of a 28-year-old Caucasian pregnant woman, gravida 1 para 0, at 21 weeks of gestation, referred to the hospital for routine fetal ultrasonographic evaluation.

This case involved late pregnancy diagnosis at 20 weeks of gestation; therefore, first trimester ultrasound and screening were not performed.

Evaluation of the fetal heart showed a normal four-chamber-view but an abnormal pattern in the positioning of the major vessels in the three-vessels and trachea view (**Figure 1 and Figure 2**). A sagittal view of the aortic branching is demonstrated in **Figure 3**.

Further morphological assessment confirmed a female fetus with no extracardiac malformations. Non-invasive prenatal testing for aneuploidy screening performed at the time of this ultrasound, indicated a low risk for trisomies 13, 18 and 21.

A follow-up ultrasound was conducted at 23 weeks to reassess fetal morphology, followed by another at 27 weeks due to the late pregnancy diagnosis. This evaluation revealed early severe fetal growth restriction, prompting hospital admission for maternal and fetal surveillance. However, the patient refused hospitalization after seven days and missed the subsequent ultrasound evaluations. At 30 weeks of gestation, she was admitted to the emergency department, where fetal demise was diagnosed. Post-mortem fetal examination confirmed the diagnosis of asymmetric fetal growth restriction and a right aortic arch with atypical *ductus arteriosus* drainage into the descending aorta and no additional findings. Placental analysis revealed diffuse high-grade chronic villitis, leading to fetal vascular dysfunction, which likely contributed to fetal demise.

What is your diagnosis?

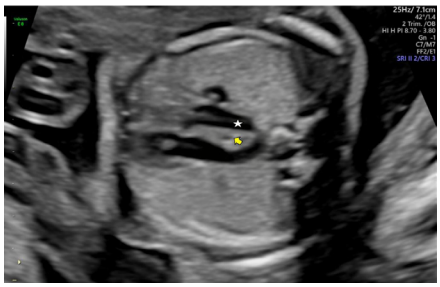

Figure 1 - Ultrasonographic image in B-mode of the three vessels and trachea view (yellow *arrow* points to the trachea, limited downward by the pulmonary artery and upward by the transverse aortic arch marked by the white *star).*

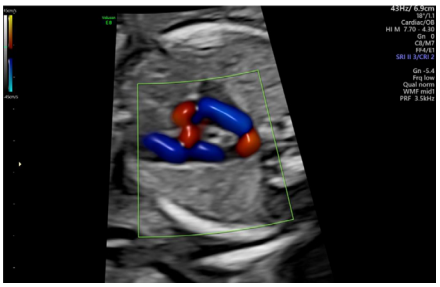

Figure 2 - Ultrasonographic image of the three vessels and trachea view with color Doppler.

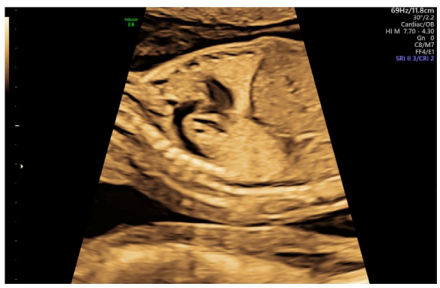

Figure 3 - Sagital view of the fetal thorax demonstrating the branching of the aortic arch.

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DIAGNOSIS

Right aortic arch with mirror-image branching and a patent left *ductus arteriosus* communicating with the descending aorta.

PATIENT MANAGEMENT

The patient was referred for fetal echocardiography at a tertiary specialized center in Pediatrics' Cardiology at 23 weeks of gestation. Invasive prenatal testing was offered but declined by the patient. Due to the late pregnancy diagnosis, serial ultrasound evaluations were conducted every two to four weeks to monitor fetal growth, development and any additional changes.

DISCUSSION

A right aortic arch is characterized by an abnormal position of the aorta and brachiocephalic vessels in relation to the trachea. It is a relatively rare finding in routine ultrasound examination.[1] Estimated prevalence among low-risk fetuses and adults is about 0,1%.[2]

Its trajectory courses to the right of the trachea, crossing the primary right bronchus and descending along the right side of the vertebral column, in contrast to the normal left aortic arch. Rarely, the course may cross the midline posterior to the esophagus, above the level of the carina, and descend along the left side of the vertebral column. This variation is known as a circumflex retroesophageal right aortic arch.

During embryonic development, interruption of the fourth left aortic arch results in the formation of a right aortic arch, characterized by multiples configurations of brachiocephalic branching and its relationship to the pulmonary artery through the *ductus arteriosus*.[1,3,4,6]

The prenatal diagnosis of a right aortic arch is very important because it may be associated with other cardiac or extracardiac anomalies. Sometimes it is also associated with chromosomal anomalies, namely the 22q11.2 deletion.[3,7] Therefore, prenatal invasive testing should be offered with quantitative fluorescence polymerase chain reaction (QF-PCR) and eventually array comparative genomic hybridization (aCGH).[8]

The anomalous arrangement of this vascular territory may predispose to the formation of vascular rings encircling the tracheoesophageal complex, potentially leading to compression symptoms which can be potentially fatal.[3,7] Ultrasonographic evaluation of the fetal heart and upper mediastinum allows the prenatal diagnosis of this anomaly, particularly through the three-vessel and the three vessels and trachea views. The introduction of these views in the systematic evaluation of the fetal heart significantly increased the detection rates of aortic arch anomalies, allowing the identification of two major patterns. The identification of a vascular echocardiographic pattern with a "U-shaped" appearance in which the transverse aortic arch and the *ductus arteriosus* are on opposite sides of the trachea, thus encircling it, is the typical appearance in the presence of a left *ductus arteriosus* communicating directly with the descending aorta or with an aberrant left subclavian artery or innominate artery.[1,3-5] The other pattern is the mirror-image branching which is a mirror-image of the normal left aortic arch.[5] The greater challenge is differentiating between a right aortic arch and a double aortic arch, where the trachea and the esophagus are encircled by the two branches of the aortic arch, causing a vascular loop which is more likely to cause severe compressive symptoms.

This case illustrates the typical ultrasonographic appearance of an aortic arch anomaly and underscores the importance of correctly evaluating the three vessels and trachea view. Specifically, it demonstrates a right aortic arch with a "U-shaped" appearance of the vessels around the trachea in the upper mediastinum view, along with mirror-image brachiocephalic branching demonstrated in the sagittal view of the aortic arch which appears in a more rightward position relatively to the normal left aortic arch.

ABSTRACT

The prenatal diagnosis of a right aortic arch consists of the visualization of a malposition of the aorta crossing the upper mediastinum to the right side of the trachea. The detection rate of this anatomic abnormality has improved with the introduction of the upper mediastinum views in routine fetal ultrasonographic evaluation. The following case-report demonstrates a fetus affected by an isolated right aortic arch emphasizing the importance of the five Yagel views of the fetal heart.

Keywords: aortic arch; fetal echocardiography; fetal heart; prenatal ultrasonography

RESUMO

O diagnóstico pré-natal de arco aórtico direito consiste na visualização de uma anomalia da posição da aorta a cruzar o mediastino superior à direita da traqueia. A taxa de deteção desta anomalia anatómica aumentou com a introdução de planos de visualização do mediastino superior nas avaliações ecográficas fetais de rotina. O seguinte caso demonstra um feto afetado por um arco aórtico direito enfatizando assim a importância dos cinco planos de Yagel para a avaliação do coração fetal.

Palavras-chave: arco aórtico; coração fetal; ecocardiograma fetal; ecografia pré-natal

AUTHORSHIP

Beatriz Sousa Ferreira - Conceptualization; Investigation; Project administration; Visualization; Writing – original draft; Writing – review & editing

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Ana Isabel Correia - Validation; Project administration; Supervision

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Received for publication: 16.04.2024
Accepted in revised form: 04.04.2025