

Prenatal Diagnosis of Vascular Ring with Right Aortic Arch

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ABSTRACT

The diagnosis of vascular ring with right aortic arch allows early treatment after birth preventing morbidity in infants and older children.

We describe the sonographic findings of two patients with right-sided aortic arch associated with aberrant left subclavian artery and left ductus arteriosus.

Two women at 24 weeks pregnancy aged 18 and 24 years old underwent a thorough echocardiographic evaluation of the fetal heart by targeted visualization of the three-vessel view at the Cardiocentro Pediátrico "William Soler", using an ATL HDI 5000 ultrasound machine made in USA with a 3 MHz transducer.

The presence of a right-sided aortic arch, left ductus arteriosus and aberrant left subclavian artery was detected in both cases. Color Doppler ultrasound demonstrated the presence of Kommerell's diverticulum. One of the fetus also presented tetralogy of Fallot and the woman opted for abortion. Both prenatal diagnoses were confirmed, one by postnatal echocardiography and the other by histopathological study.

The presence of right-sided aortic arch, left ductus arteriosus and aberrant left subclavian artery is an uncommon anomaly that can be detected by prenatal ultrasound.

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Key words >

Congenital Heart Defects - Ductus Arteriosus - Prenatal Diagnosis - Echocardiography

Abbreviations >

EAA	Aortic arch	BCT	Brachiocephalic trunk
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BACKGROUND

Aortic arch abnormalities (AA) are present in 1-2% human fetuses and can be related with complex cardiac defects (aortic coarctation, aortic arch interruption or tubular hypoplasia, double aortic arch) or be classified as normal variants that are rarely associated to clinical signs after birth. (1)

There are two typical forms of right AA; the most frequent is called "with mirror image" and the second is referred to as right AA with retroesophageal Kommerell's diverticulum. (2) The infrequency of this type of vascular ring, together with the few reported cases with prenatal diagnosis induced the publication of these cases.

CLINICAL CASES

Two cases are presented, with prior informed consent of the concerned couples, to describe the clinical characteristics and to report prenatal and postnatal

testing results. Patients were evaluated at the Cardiocentro Pediátrico "William Soler" (National Reference Center for Prenatal Diagnosis of Congenital Cardiopathies).

The first case corresponds to an 18 year-old pregnant woman at 24 weeks' gestation, referred to our center for suspected congenital cardiopathy. Upon diagnosis of tetralogy of Fallot, the couple decided to terminate the pregnancy (in our country, abortion for relevant reasons related to the fetus' health is accepted up to 25.6 weeks, with previous genetic counseling and informed consent of the couple).

The second case corresponds to a 24 year-old pregnant woman at 24 weeks' gestation, classified as low obstetric risk, referred to our center for interconsultation due to lack of ultrasound visualization of cardiac structures, leading to several echocardiographic studies throughout pregnancy.

Both patients were examined with an ATL (HDI

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5000) ultrasound machine, made in USA, with a 3 MHz transducer. In each case, a sequential study of the heart was performed, using all the fetal echocardiographic views. (3)

In these two fetuses, the AA was positioned to the right and the echocardiographic three-vessel view (4) allowed the diagnosis.

The following echocardiographic signs were observed in both cases:

Three vessel view and ductus arteriosus (Figure 1)

- Aortic artery slightly more separated than usual from the pulmonary artery
- Descending aorta more anterior and to the right of its normal position.

Transverse aortic arch view (Figure 2)

- Trachea positioned to the left of the AA

Simultaneous view of the arch and ductus arteriosus (Figure 3)

- Trachea positioned between the ductus arteriosus and the AA.

In one of the cases, the right AA was associated with tetralogy of Fallot, (Figure 4) with persistent left superior vena cava, so in addition to the above mentioned signals there were alterations in size (aorta larger than the pulmonary artery), in position (anterior displacement of the aorta) and in the number of vessels (fourth vessel positioned to the left of the pulmonary artery). Following this couple's decision to terminate the pregnancy, the diagnosis was confirmed by histopathological study.

In the other case, the pregnancy reached full term. Childbirth was produced by institutional eutocic delivery, with normal weight and Agpar 9/9. At its last cardiological evaluation performed four months later, the child was asymptomatic. The echocardiographic study showed the presence of the right AA without persistent ductus arteriosus.

The thymus was present in both fetuses, with normal size for the gestational age. A digital back-up was obtained in both cases, for later study and analysis.

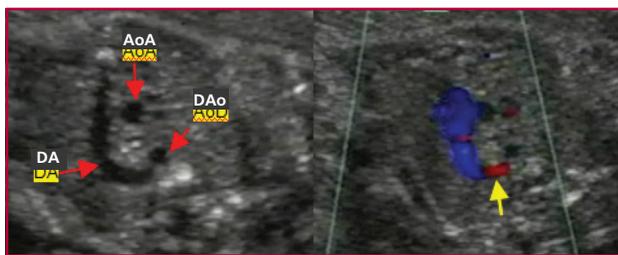


Fig. 1. Three-vessel and ductus view. Aortic artery (AoA) slightly more separated than usual from the pulmonary artery. Descending aorta (DAo) more anterior and to the right of its normal position. Ductus arteriosus (DA) positioned to the left. Color Doppler echocardiography revealed antegrade flow in the ductus and retrograde filling from the ductus toward the descending aorta (arrow).

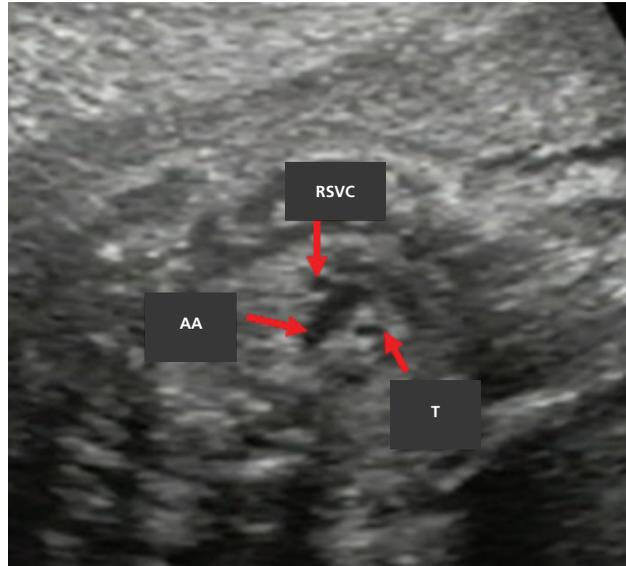


Fig. 2. Transverse view of the aortic arch. An aortic arch (AA) is seen to the right of the trachea (T). RSVC: Right superior vena cava.

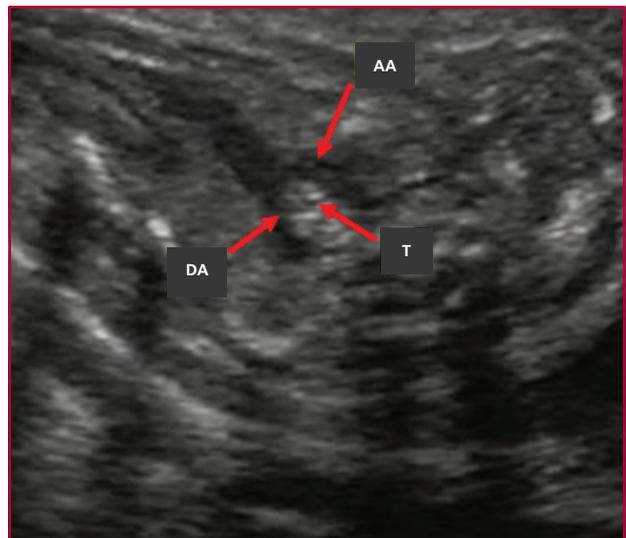


Fig. 3. Simultaneous view of the arch and ductus, where the trachea (T) is positioned between the ductus arteriosus (DA) and the aortic arch (AA).

DISCUSSION

In anatomical terms, a right AA is defined as the aortic arch that courses to the right of the trachea and above the main right bronchus, while the descending aorta can run to the left, middle or right of the medial line outlined by the vertebral column.

In normal conditions, the fourth left AA will give origin to the arch of the aorta which will connect with the ascending aorta, and the fourth right AA will give rise to the arterial brachiocephalic trunk (BCT). In pathological conditions, reabsorption of the fourth left

AA with the persistence of the fourth right AA, will result in a right-sided AA with the BCT located to the left (instead of to the right) and the first aortic vessel will present as described in the introduction. (2)

The case of right AA diagnosis associated with tetralogy of Fallot, with persistent left superior vena cava revealed previously described characteristic echocardiographic signs of both entities. (5, 6)

An important aspect to be considered when faced with a right AA is the position of the ductus arteriosus. In the examined fetuses, the ductus arteriosus was in a contralateral position to the arch, presenting the classical U sign (ascending aorta, right AA to the right of the trachea, aberrant left subclavian artery behind, left ductus arteriosus, the main pulmonary artery to the left and the heart at the front) referred to in the literature (7) (Figure 4).

Color Doppler ultrasound showed antegrade blood flow in the ductus arteriosus and retrograde filling from the ductus toward the descending aorta. This zone described by Burckhard Kommerell in 1936, (7) is known as Kommerell's diverticulum, which in the fetus corresponds to the retroesophageal portion of the left subclavian artery. (Figure 5)

Normally, the ductus arteriosus that originates from the distal portion of the sixth AA is located on the same side of the arch of the aorta, i.e. if the arch of the aorta is right-sided, it is normal for the distal portion of the sixth right AA to persist as ductus and not that of the left. However, if the fourth right AA were arch of the aorta, the distal portion of the sixth left AA would persist as ductus, i.e. if the ductus were contralateral to the arch of the aorta, a vascular ring would form surrounding the developing trachea and esophagus, provided it is associated to an aberrant left subclavian artery. (8)

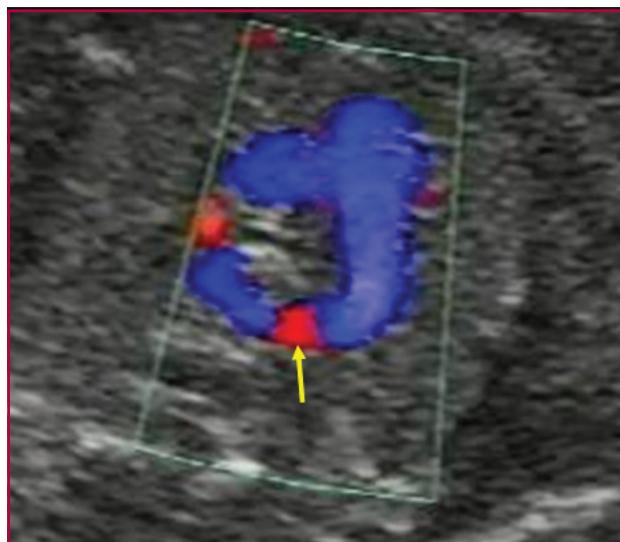


Fig. 4. Use of color Doppler revealed antegrade flow in the ductus and retrograde filling from the ductus toward the descending aorta (classical U sign).

Recent studies (9) emphasize the importance of transverse arch examination, both in the normal fetus as in those with cardiac anomalies. In the latter, due to their high association with chromosomal abnormalities, a cariotype study is recommended, including a detailed analysis of chromosome 22q11 (10) On the other hand, in a case of right AA, derived to fetal echocardiography because of suspect routine study, which does not present additional findings in the fetus, there is no clear indication of a chromosomal study since right AA can occur, more frequently than is known, in the normal population. Moreover, a postnatal assessment to confirm the diagnosis is recommended in all prenatally detected right-sided AA. In asymptomatic infants, magnetic resonance imaging is not justified to examine neck vessels, and neither corrective surgery.

RESUMEN

Diagnóstico prenatal de anillo vascular con arco aórtico derecho

El diagnóstico de anillo vascular con arco aórtico derecho en el feto favorece un tratamiento precoz después del nacimiento, que permite evitar la morbilidad tanto en la infancia como en edades mayores.

En esta presentación se describen los hallazgos sonográficos en dos casos de arco aórtico derecho asociado con arteria subclavia izquierda aberrante y ductus arterioso izquierdo. Se realizó un estudio ecocardiográfico detallado del corazón fetal, con particular énfasis en la vista extendida de los tres vasos, en dos gestantes con 24 semanas de embarazo y edades de 18 y 24 años, atendidas en el Cardiocentro Pediátrico "William Soler"; se utilizó un equipo ATL (HDI 5000), de fabricación estadounidense, con transductor de 3 MHz.

En ambos fetos se detectó la presencia de arco aórtico derecho, ductus arterioso izquierdo y arteria subclavia izquierda aberrante. Al utilizar el Doppler color se apreció una zona dilatada, conocida como divertículo de Kommerell. Uno de los fetos presentaba además tetralogía de Fallot asociada; en este caso, la gestante optó por la interrupción del embarazo. Ambos diagnósticos prenatales fueron confirmados, uno mediante ecocardiografía posnatal y el otro a través del estudio anatomopatológico.

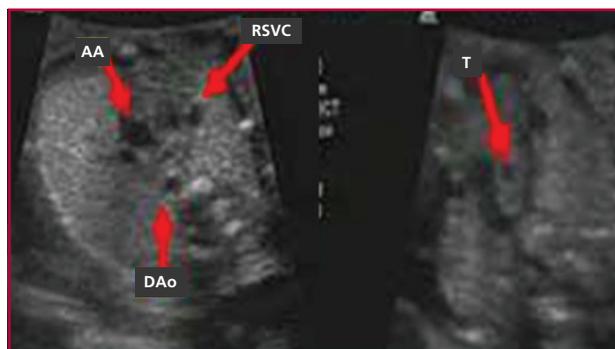


Fig. 5. Case with right aortic arch (AA) associated with tetralogy of Fallot, with persistent left superior vena cava [positioned to the left of the aortic artery (arrow)] of greater size than the pulmonary artery and with anterior displacement. RSVC: Right superior vena cava. DAo: Descending aorta. T: Trachea.

La presencia de un arco aórtico derecho, ductus arterioso izquierdo y arteria subclavia izquierda aberrante constituye una variante poco frecuente, con posibilidades de diagnóstico prenatal.

Palabras clave > Cardiopatías congénitas - Conducto arterial - Diagnóstico prenatal - Ecocardiografía

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