Case Report

Role of Transthoracic Echocardiography in the Diagnosis of Double Aortic Arch

Papel da Ecocardiografia Transtorácica no Diagnóstico de Duplo Arco Aórtico

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Introduction

Double aortic arch is the main cause of vascular ring in cardiovascular surgical case series records. This rare anomaly, caused by the non-involution of one of the embryonic aortic arches, is a challenge for the pediatric echocardiographer, often requiring other imaging methods such as computed tomography or cardiac magnetic resonance. This case report aimed to systematize echocardiographic findings to help diagnose double aortic arch.

Case report

A full-term newborn was born vaginally at 38 weeks' gestation with a birth weight of 3,390 g, and an APGAR 9/10. In the first hours of life, the patient presented respiratory discomfort associated with stridor, wheezing, and increased expiratory time requiring orotracheal intubation and high mechanical ventilation parameters. Chest X-ray showed signs of pulmonary hyperinflation.

The mother reported home contact with a confirmed case of severe acute respiratory syndrome coronavirus 2 approximately 15 days before delivery. A cardiac evaluation was requested due to the possibility of pediatric multisystem inflammatory syndrome, but this was not confirmed later. A cardiovascular system examination detected no changes.

Echocardiography showed coronary artery diameters within the upper limit of normal and an echogenic focus on the left coronary artery wall, preserved cardiac function, and no signs of significant pulmonary hypertension. An aortic arch to the left was visualized in longitudinal suprasternal section with the probe index pointed at 1 o'clock with the emergence of two vessels. Directing the probe index to 11 o'clock revealed another aortic arch of equal diameter with the emergence of two vessels. A double aortic arch forming a complete vascular ring was suggested.

Chest computed tomography angiography confirmed the presence of a tracheoesophageal vascular ring resulting from a balanced double aortic arch associated with tracheal stenosis.

Keywords

Vascular ring, Echocardiography, Heart Defects, Congenital.

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The patient underwent surgical resection of the right aortic arch and release of tracheal compression that postoperatively progressed with extubation failure. Bronchoscopy showed 80% obstruction of the tracheal lumen during inspiration due to tracheomalacia for which a tracheostomy was indicated.

Discussion

Vascular rings represent less than 1% of all congenital heart diseases.[1-4] Double aortic arch, the most frequent type of vascular ring,[4-7] is caused by an embryogenic anomaly with persistent right and left fourth embryonic aortic arches.[4,8,9]

These two patent arches resulted in a bifurcated ascending aorta, whose branches encircle the trachea and esophagus and posteriorly join to form a single descending aorta, forming a ring around these structures that can cause compression.[1,2,4,8] The right arch is often dominant (75%), with only 5% of cases presenting both arches of equal size as in the case reported here.[2,4]

In most cases, double aortic arch occurs as an isolated lesion, although it may be associated with other malformations such as tetralogy of Fallot, truncus arteriosus, and transposition of the great arteries.[4,6] In cases of significant tracheal and esophageal compression, symptoms such as stridor, dyspnea, respiratory failure, and feeding difficulties may be observed soon after birth.[1,3,4,6,8,9]

On the other hand, if compression is minimal, the vascular ring will be accidentally diagnosed in asymptomatic adults.[3,6,8,9]

Treatment is reserved for symptomatic cases and consists of surgical resection of one of the aortic arches, preferably the smaller one.[2-4,6,9] However, after tracheal compression release, respiratory symptoms may persist in a significant number of children (54%) due to tracheomalacia, as in the case described in this report.[2,9]

Although computed tomography and cardiac magnetic resonance imaging are essential for confirming the diagnosis and surgical planning, an initial assessment with transthoracic echocardiography allows the identification of the double arch as in the case reported. Furthermore, it has the advantages of low cost and not requiring the use of contrast, sedation, or radiation.[1,3,4,6-9]

This report presents a systematic approach to diagnosing double aortic arch through transthoracic echocardiography, focusing on its presentation pattern in the different echocardiographic windows. A careful assessment of the aortic arch with determination of its laterality and the branching pattern of brachiocephalic vessels is essential for identifying a double aortic arch by echocardiographic examination.[3,4]

Longitudinal parasternal window

Standard section - probe index pointing at 1 o'clock

Visualization of the aortic arch to the left without the normal branching pattern and origination of two vessels - the left subclavian and common carotid arteries; however, it is impossible to demonstrate the origin of the brachiocephalic trunk[1,3,4,6,7] (Figure 1A); and

Counterclockwise rotation of the transducer - probe index pointing at 11 o'clock

Visualization of a similar image with an aortic arch to the right and origination of the two vessels - the right subclavian and common carotid arteries[1,4,6,7] (Figure 1B).

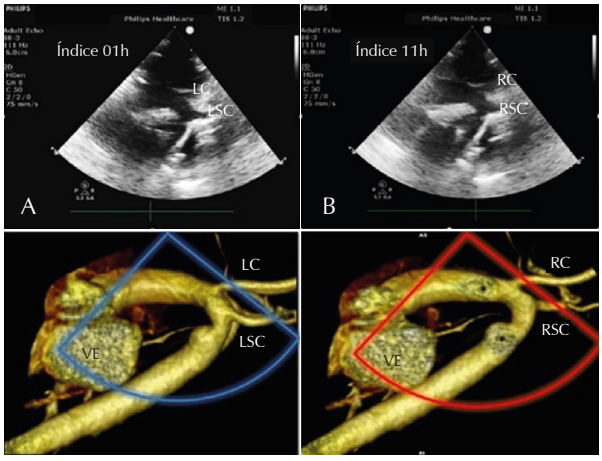
Cross-sectional parasternal window

Probe index pointing at 3 o'clock and with cephalic tilt

Identification of the ascending aorta bifurcating into the left and right aortic arches, with blood flow visualization by color Doppler in both arches[1,3,4,6-10] (Figure 2A); and

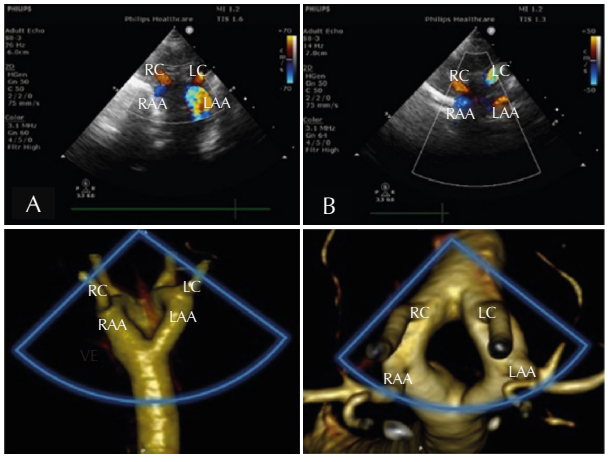
Most superior ultrasound beam tilt

Possible simultaneous identification of the four brachiocephalic vessels, two in each arch. This image is characteristic of the double aortic arch - the "four-vessel sign"[2,4,9,10] (Figure 2B).



LC, left carotid; LSC, left subclavian; RC, right carotid; RSC, right subclavian.

Figure 1 – *Echocardiographic image in longitudinal suprasternal section with the probe index pointing at (A) 1 o'clock showing a left arch originating the left subclavian and common carotid arteries and at (B) 11 o'clock showing an aortic arch of the same caliber originating the right subclavian and common carotid arteries. Comparative image with CT angiography.*



LAA, left aortic arch; LC, left carotid; LSC, left subclavian; RAA, right aortic arch; RC, right carotid; RSC, right subclavian.

Figure 2 – *Echocardiographic image in cross-sectional suprasternal section showing (A) the ascending aorta bifurcated into the right and left arches and (B) simultaneous identification of the four brachiocephalic vessels - the "four-vessel sign" characteristic of a double aortic arch. Comparative image on computed tomography angiography.*

Subcostal or five-chamber apical window

Visualization of a bifurcated ascending aorta similar to the pulmonary artery bifurcation

The aorta bifurcation into the double arch is seen at a significant distance from the aortic valve plane (about 4–4.5 cm) as opposed to the early bifurcation seen in the transposition of the great arteries, type I truncus, hemitruncus, and aortopulmonary window[5-7] (Figure 3).

Conclusion

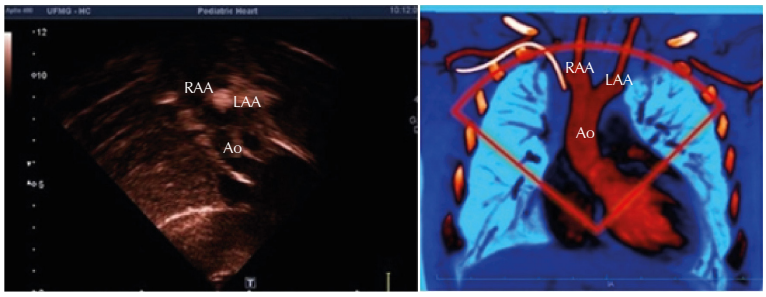
Echocardiography is the ideal method of screening for double aortic arch due to being non-invasive, easily performed, and of low cost. The presence of an aortic arch on the right or left with a normal innominate artery branching pattern into the carotid and subclavian arteries or originating separately from the four cephalic vessels of the same arch excludes the presence of a double aortic arch. Cardiac magnetic resonance imaging and computed tomography should be reserved for diagnostic confirmation and preoperative planning.

Author's contributions

Research conception and design: Souza GG; Data collection: Wang R; Data analysis and interpretation: Guimarães AFM, Castilho SRT; Manuscript writing: Guimarães AFM, Souza GG; Critical review of the manuscript for intellectual content: Tonelli HAF, Araújo FDR.

Conflict of interest

The authors have declared that they have no conflict of interest.



Ao, ascending aorta; LAA, left aortic arch; RAA, right aortic arch.

Figure 3 – *Subcostal echocardiographic image of the left ventricular outflow tract showing bifurcation of the ascending aorta into the right and left arches at a significant distance from the aortic valve plane. Comparative image on computed tomography angiography.*

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