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Review Article

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Double-Lumen Aortic Arch: An Extremely Rare Congenital Heart Anomaly or an Underdiagnosed Disease? – Case Report and Literature Review

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Introduction

Aortic arch anomalies and anatomical variants are relatively common, encompassing a broad spectrum of malformations in embryonic development, occurring in 0.5 to 3% of the population [1,2]. Type I double-lumen aortic arch is one of these anomalies, characterized by the presence of a second systemic-to-systemic channel connecting the ascending and descending aorta components. It's an extremely rare finding, first described in 1969 by Van Praagh, whose incidence remains unknown, as only a few case reports have been published in the literature since then [3].

Here it's described a case of type I double-lumen aortic arch, evidenced during echocardiography evaluation in a patient admitted to a referral hospital for pediatric cardiac surgery in Northeastern Brazil.

Clinical Summary

A female infant, daughter of a hypertensive and diabetic pregnant woman, was born prematurely (31 weeks of gestacional age), with very low birth weight, intrauterine growth restriction and dysmorphic features, without investigation of congenital heart disease during the prenatal follow up. After birth, a heart murmur was heard by neonatologist, thus a transthoracic echocardiogram was performed, revealing a wide perimembranous ventricular septal defect (Figures 1 and 2) and patent ductus arteriosus (Figure 3).

The patient developed signs and symptoms of heart failure within the first few weeks of life, including cardiomegaly, tachycardia, tachypnea and diaphoresis. In this manner, anti-congestive therapy with diuretic drugs was initiated (furosemide and spironolactone), along with fluid restriction. Due to the persistence of decompensated heart failure despite optimization of medical management, transfer to a tertiary center specialized in pediatric cardiac surgery was requested for invasive cardiac intervention.

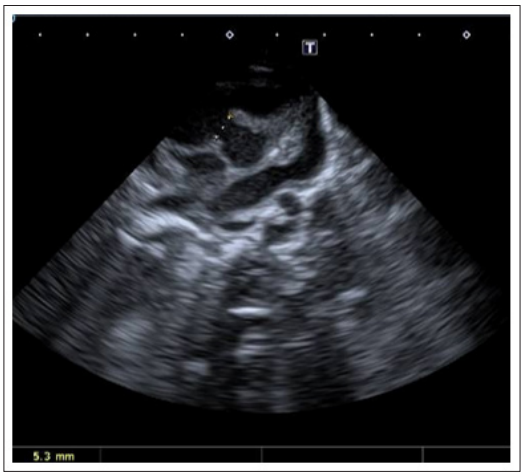

Figure 1: Wide perimembranous ventricular septal defect (VSD). Echocardiogram image in the parasternal short axis view, showing the perimembranous ventricular septal defect.

The patient was admitted to intensive care unit of a cardiology reference center at two months of chronological age, following treatment for infectious complications. On physical examination, she was in fair general condition, acyanotic, normotensive, tachydyspneic, tachycardic, maintaining adequate oxygen saturation levels and pulse amplitude in the four limbs, without asymmetries between lower and upper limbs. Cardiac clinical

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evaluation revealed a hyperdynamic precordium, left ventricular systolic impulses, a prominently audible second heart sound and a grade 3+/6+ systolic murmur best heard at the mid-left sternal border. Chest X-ray showed an enlarged cardiac silhouette and a pattern of pulmonary congestion (Figure 4).

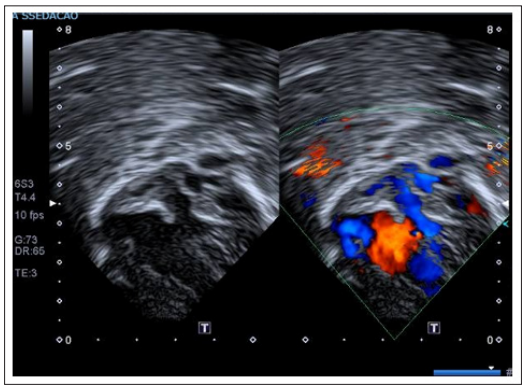

Figure 2: Wide perimembranous ventricular septal defect (VSD). Echocardiogram image in the apical 5-chambers view, showing the perimembranous ventricular septal defect with left to right shunt.

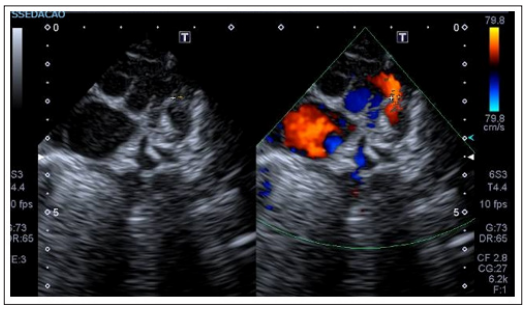

Figure 3: Persistent arterial duct. Echocardiogram image in the parasternal short axis view, showing the persistent arterial duct with aorta to pulmonar artery shunt

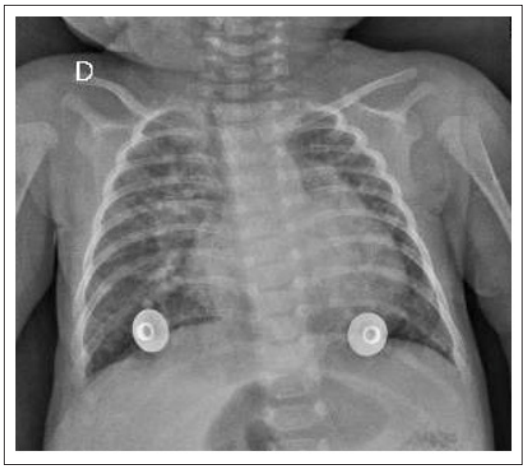

Figure 4: Pre-operatory chest X-ray showing cardiomegaly and a pattern of pulmonary congestion

A transthoracic echocardiography was repeated, confirming the previously described findings, but additionally revealing a hyper-refringent image along the transverse aortic arch, apparently septating the arch, with no evidence of increased flow velocity on continuous-wave Doppler assessment (Figures 5, 6 and 7).

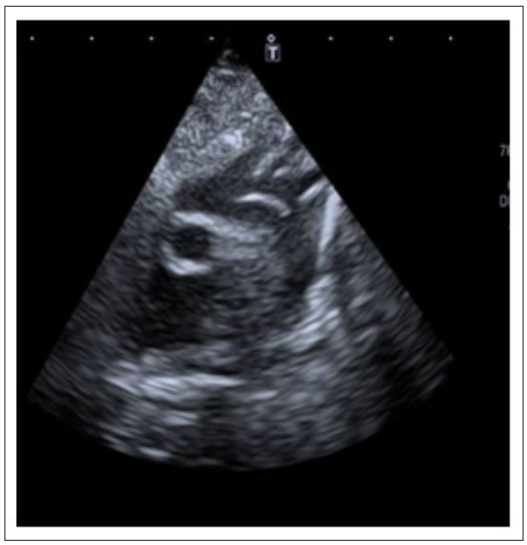

Figure 5: Double lumen aortic arch. Echocardiogram image in the suprasternal axis view, showing type I double lumen aortic arch.

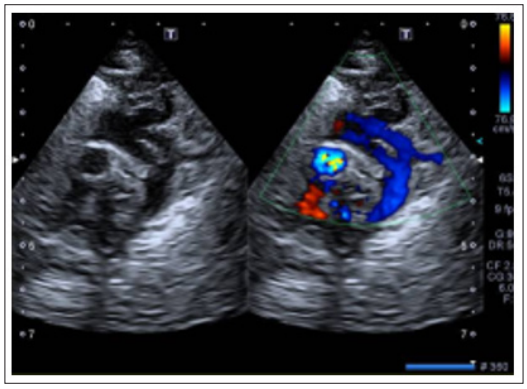

Figure 6: Double lumen aortic arch. Echocardiogram image in the suprasternal axis view, showing type I double lumen aortic arch (left image) and laminar flow on color Doppler assessment (right image)

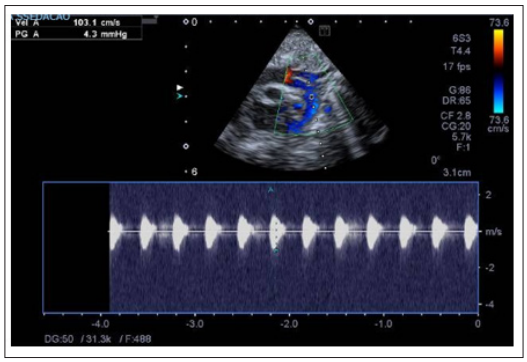

Figure 7: Aortic arch flow. Echocardiogram image in the suprasternal axis view, showing normal flow velocity on continuous-wave Doppler assessment.

For further evaluation, a pre-operatory computed tomography angiography was performed, confirming the presence of a type I double-lumen aortic arch, but no additional aortic arch anomalies were identified (Figures 8 and 9).

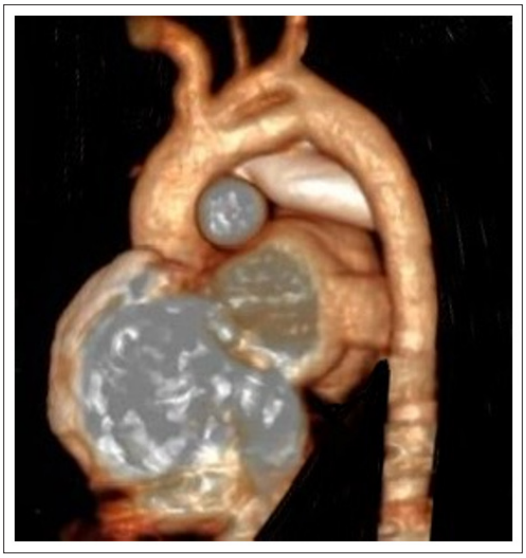

Figure 8: Type I double lumen aortic arch on three-dimensional reconstructed computed tomography angiography.

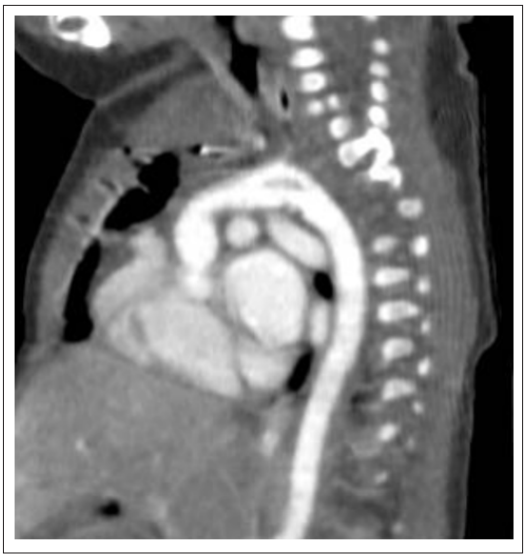

Figure 9: Type I double lumen aortic arch on sagittal section of computed tomography angiography.

Subsequently, the patient underwent palliative pulmonary artery banding and ductus arteriosus ligation for heart failure management. Conservative approach was adopted in relation to the aortic arch anomaly, given that the condition caused no hemodynamical repercussions.

In the postoperative period, the infant had a favorable clinical course and was discharged in good condition for outpatient follow-up. At six months of age, she was electively readmitted for corrective surgical reintervention, during which she underwent ventricular septoplasty and takedown of the pulmonary artery banding. She was discharged for continued outpatient follow-up and is currently clinically stable from a cardiological standpoint, without cardiovascular medications, and under regular clinical monitoring.

Literature Review

The double-lumen aortic arch is a congenital cardiovascular anomaly that was initially attributed to persistence of the fifth aortic arch (PFPA) (Figure 10) [3]. However, its embryological origin remains incompletely elucidated and may alternatively be related to the presence of dorsal collateral arteries connecting the fourth and sixth aortic arches [4-9].

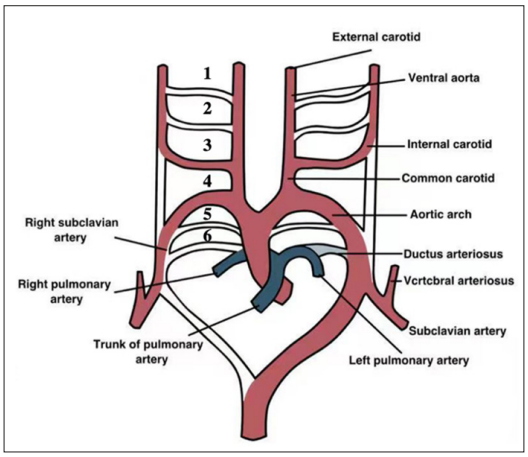

Figure 10: Development of each pair of arterial arches in the embryo. The classical concept derived from the so-called Rathke diagram, in which six sets of arteries (1 to 6) are believed to extend through the pharyngeal arches during the stages of development of the human heart. Source: Adapted from Shan et al. (2023).

That aortic anomaly can be classified into three types: Type I - Double lumen with both lumens patent; Type II -Atresia or interruption of one (upper) lumen and patency of the other; and type III - Connection between the lower lumen and the pulmonary artery. Type I appears to be the most frequently diagnosed [3,10,11].

Although Gerlis et al. reported an incidence of PFPA of approximately 1 in 330 in autopsy series, no precise data are available regarding the true incidence of type I double-lumen aortic arch. By 1989, only 15 cases had been reported, and since then, only a few additional cases have been documented. [5-7, 12-16].

This anomaly is commonly associated with various intracardiac defects [17]. The most frequently reported concomitant congenital abnormalities are aortic coarctation and patent arterial duct [18]. The absence of associated congenital heart defects likely contributes to its underdiagnosis, as the condition often remains clinically silent. Consequently, it is frequently detected incidentally during routine imaging or identified as an additional finding in the assessment of other cardiac malformations.5 Additional factors that may lead to underdiagnosis include the inherent technical limitations of echocardiography in evaluating the aortic arch, the examiner's level of expertise, a limited suprasternal acoustic window in the patient, and the potential for misinterpretation as other cardiovascular anomalies.

Despite the aforementioned diagnostic limitations, the defect can be identified by echocardiography, which is typically the initial imaging modality. Complementary evaluation with alternative imaging techniques, such as chest computed tomography angiography or cardiac magnetic resonance angiography, is

warranted for diagnostic confirmation [19,20]. These modalities allow for a more detailed anatomical assessment of the location, providing highly valuable information, given that hemodynamic procedures via cardiac catheterization may be required later in the patient's life for unrelated clinical indications [21].

It is important to emphasize that, unlike the "double aortic arch"—a condition resulting from persistence of both primitive arches, each located on one side of the trachea—, the double-lumen aortic arch does not form a vascular ring, as there are no structures located between the lumina and both arches are same-sided, running in parallel on the same side of the trachea [20]. Therefore, despite the similarity in nomenclature and anatomy, these entities should not be confused, as they require entirely different management approaches.

Conclusion

Double-lumen aortic arch is a rare and potentially underdiagnosed anomaly, particularly in the absence of associated cardiac defects, as it lacks clinical manifestations. Despite significant advances in imaging modalities, its diagnosis by echocardiography remains challenging, warranting complementary evaluation with chest computed tomography angiography or cardiac magnetic resonance angiography for improved anatomical assessment.

Insufficient awareness of the anomaly may hinder—or even preclude—the performance of hemodynamic procedures via cardiac catheterization, which could be required later in the patient's life for unrelated clinical indications.

Although first described more than five decades ago, this entity remains incompletely understood. A thorough understanding of the anomaly, along with familiarity with the embryological mechanisms involved, may enhance diagnostic accuracy and facilitate earlier recognition in clinical practice.

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