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Case Report

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery in Children: Diagnostic Value of Transthoracic Echocardiography

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Abstract

Introduction: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but life-threatening congenital heart disease. Conventionally, ALCAPA is diagnosed by angiography, which is now replaced by cardiac CT and cardiovascular magnetic resonance. Nevertheless, performing these exams in severely ill infants is being questioned. In this context, noninvasive cardiovascular imaging, mainly transthoracic echocardiography plays an important role. The purpose of this article is to present 2 children with ALCAPA and to focus on the value of transthoracic echocardiography in diagnosis of this cardiac anomaly.

Case Reports: 1. A 5-year-old girl was referred to our pediatric cardiology unit for further evaluation of heart failure. She has been managed in the pediatric department since the age of 7 months because of dilated cardiomyopathy. The diagnosis of ALCAPA was suspected thanks to the electrocardiogram and confirmed by Doppler echocardiography data. The child was successfully operated on.

2. An 18-month-old female infant presented to the pediatric department with respiratory distress and cardiogenic shock. Echocardiography was perfo rmed and demonstrated severely decreased LV function and due to a step by step echocardiography the diagnosis of ALCAPA was made. The child underwent surgical repair: implantation of the left coronary artery directly into the aorta with a good outcome.

Conclusion: The diagnosis of ALCAPA should be suspected in infants who have dilated cardiomyopathy with electrocardiographic changes that suggest ischemia, and in children who have isolated mitral insufficiency. Based on a step by step approach, two-dimensional echocardiogram combined with color-flow Doppler can diagnose anomalous origin of left main coronary artery accurately and give sufficient information for operation.

Keywords: *ALCAPA; Echocardiography; Congenital Heart Disease; Heart Failure*

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but life-threatening congenital heart disease. It occurs once per 300,000 live births and accounts for 0.5% of all congenital cardiac malformations [1]. It is typically an isolated anomaly, but can be seen in association with tetralogy of Fallot, ventricular septal defect, or patent arterial duct [2].

Also termed Bland-White-Garland syndrome, ALCAPA is characterized by a left coronary artery arising from the pulmonary artery instead of the aortic root. This condition causes decreased oxygenated blood to reach the left ventricle, which leads to infarction of the anterolateral left ventricular free wall, mitral valve papillary muscle dysfunction and variable degrees of mitral insufficiency [3].

If untreated, the mortality rate in the first year of life is more than 90% [4].

About 15% have adequate collateralization and can remain asymptomatic until adolescence or adulthood in the setting however of a high risk of sudden death [5,6].

Because of the unfavorable prognosis, it's crucial to make diagnosis and surgical correction early.

Conventionally, ALCAPA is diagnosed by angiography, which is now replaced by cardiac CT and cardiovascular magnetic resonance [7]. Nevertheless, performing these exams in severely ill infants is being questioned. In this context, noninvasive cardiovascular imaging, mainly transthoracic echocardiography (TTE) plays an important role: With improvement in the acquisition of images, echocardiographic technology has markedly improved over the last decade.

Purpose of the Study

The purpose of this article is to present 2 children with ALCAPA and to focus on the value of TTE in diagnosis of this cardiac anomaly.

Case Reports

Case 1

A 5-year-old girl was referred to our pediatric cardiology unit for further evaluation of heart failure. She has been managed in the pediatric department since the age of 7 months because of dilated cardiomyopathy. Investigations didn't reveal any reason for her left ventricular failure. Physical examination revealed blood pressure of 110/75 mmHg, with a regular heart rate of 100 beats/min and a grade 4/6 holosystolic murmur along the apex. Electrocardiogram (ECG) showed left ventricular hypertrophy with left axis deviation and a deep Q wave in Avl and DI and inverted T waves in the lateral leads (Figure 1). A chest X-ray showed cardiomegaly.

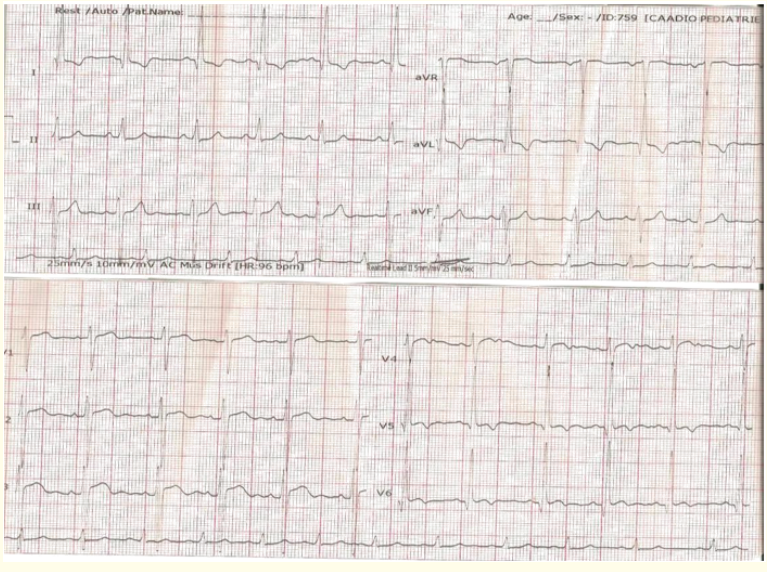


Figure 1: *Electrocardiogram revealed left ventricular hypertrophy with left axis deviation and a deep Q wave in aVL and DI and inverted T waves in the lateral leads.*

Two-dimensional and Doppler echocardiography was done on a VIVID 5 system with a5-MHz transducer. It showed a dilated left ventricle (LV) with a very low function (FEVG = 40%), enlargement of the left atrium and severe mitral regurgitation. Hyper echogenicity of the papillary muscles was also objected (Figure 2). By color mapping, multiple turbulent color flow regions (intercoronary collaterals) in the ventricular septum were found.

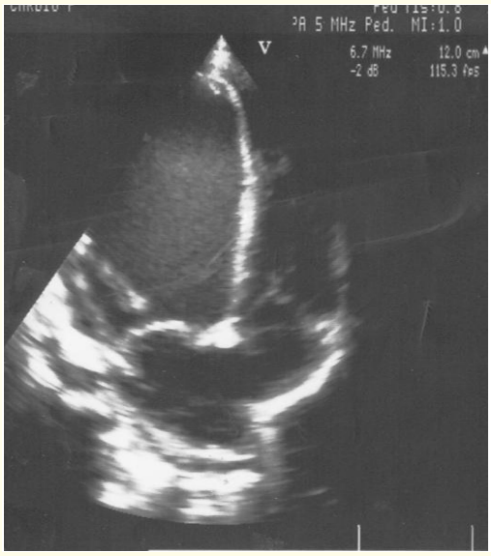


Figure 2: *Apical four-chamber view demonstrating marked left ventricular dilation with hyperechogenic endocardium and papillary muscle.*

From the parasternal long-axis view, a dilated right coronary artery (RCA) arising from the right sinus of Valsalva was visualized (Figures 3).

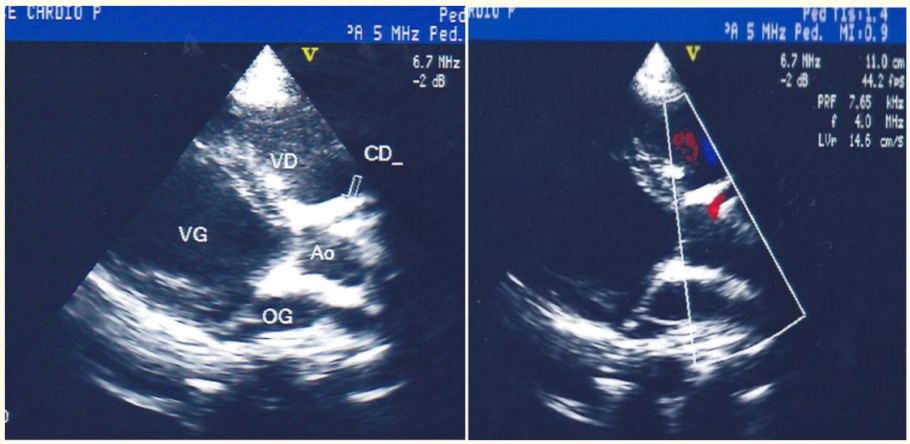
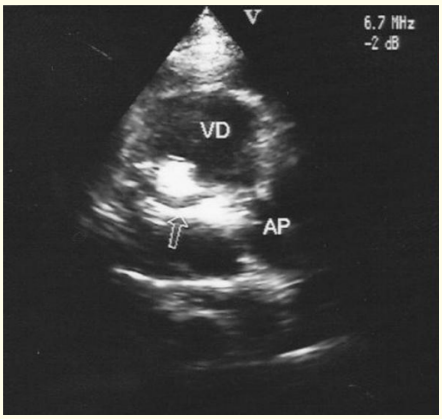


Figure 3: *Echocardiography with parasternal long-axis view shows a prominent large vessel (arrow) arising from the right aortic sinus. Ao indicates aorta; OG: Left Atrium, VG: Left Ventricle; VD: Right Ventricle.*

Modified parasternal short-axis view at the level of great arteries was used to visualize the coronary anatomy: The origin of the LMCA was absent from the left sinus of Valsalva but an abnormal vessel inserting into the pulmonary trunk was seen with abnormal retrograde coronary blood flow (Figures 4 and 5). The diagnosis of LCAPA was confirmed by CT scan (Figure 6).



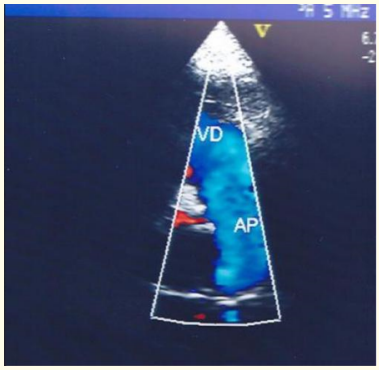


Figure 4 and 5: *Modified parasternal short-axis view identified left main coronary artery from the main pulmonary artery and the shunt from it to the pulmonary artery indicated as a red flow on colour Doppler.*

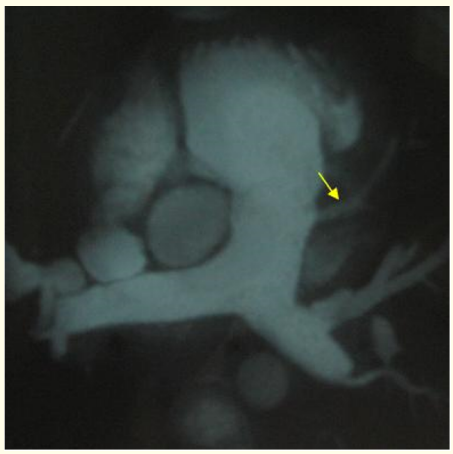


Figure 6: *CT scan showed left coronary artery arising from the pulmonary trunk (arrow).*

The child underwent surgical repair: Reimplantation of the left coronary artery directly into the aorta with a good outcome and alleviation of mitral regurgitation after 12 months of follow up.

Case 2

An 18-month-old female infant presented to the Pediatric Department with respiratory distress and cardiogenic shock. On a first echocardiogram, the ventilated patient showed severely decreased LV function (ejection fraction 26%), fractional shortening 14%. She had no past medical history or significant family history of illness. After clinical improvement within 15 days the patient was referred to our department for further investigation. Electrocardiogram showed deep Q wave in aVL and DI. Echocardiography was performed using a Vivid 9 system with 5 MHZ transducers. Increased echogenicity of the LV myocardium and the mitral papillary muscle, with moderate mitral regurgitation. There was no evidence of intraseptal collateral circulation.

The short axis view at the aortic level showed dilation of the right coronary artery from the right sinus of Valsalva and the ratio of its diameter to that of the aorta ring was 0.3 (Figure 7). The origin of the LMCA was absent from the left sinus of Valsalva There was a retrograde blood flow to the pulmonary artery. The LCA was seen to emerge from the pulmonary artery. The diagnosis was subsequently confirmed at CT Scan. Follow up after surgery revealed no complications with improvement in clinical status.

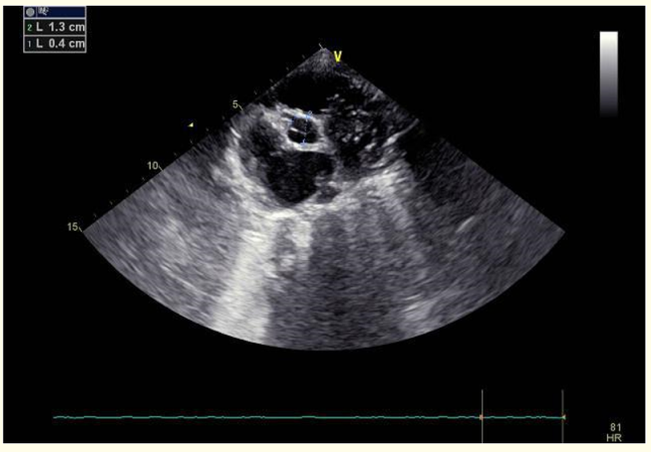


Figure 7: *Parasternal short-axis view of the dilated right coronary artery.*
Movie clip S1: *Multiple regions of color turbulence in the ventricular septum due to flow within coronary collaterals.*

Discussion

The majority of authors [8-9] identify 2 distinct ALCAPA groups: infant and older. Infant type ALCAPA (patients under the age of 2 years) is characterized by a normal sized or mild dilated right coronary artery (RCA), the intercoronary collateral vessels are insufficient or absent and the left ventricle is extremely enlarged, and older type (children aged more than 2 years, adolescents and even adults), is the type where the patient has a markedly dilated RCA and extensive intercoronary collateral system (Figure 8) [10].

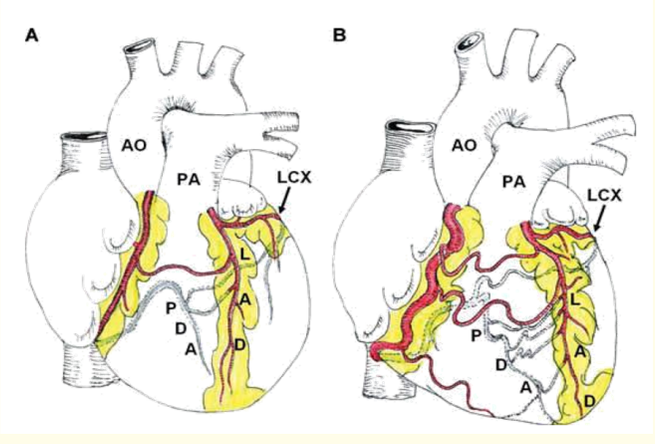


Figure 8: *The anatomic diagram of ALCAPA: A infant type B/ older type [10].*

Echographic findings are derived from these physiopathologic features.

In the past, coronary arterial anomalies have been difficult to diagnose by non-invasive methods. Nowadays with the improvement of images quality, identification of coronary arterial origins is now a routine part of the standard pediatric echocardiogram. Transthoracic echocardiography diagnosis is made using both bidimensional and Doppler modes.

In case of ALCAPA, many echocardiographic features are not directly related to the visualisation of the coronary arterial origin: Many signs should alert the echocardiographist and lead to a step by step echocardiographic approach to identify the left main coronary artery from the pulmonary artery.

The first and crucial step is to remember that any infant presenting with a dilated cardiomyopathy should be considered to have an anomalous origin of the left coronary artery from the pulmonary trunk until proven otherwise.

Also, a child with significant mitral regurgitation who has no history of rheumatic heart disease and no congenital valve anomaly should prompt the search of ALCAPA [11].

The M mode is useful to calculate LV dimensions and ejection fraction: In the infant group we find severe LV dilation and dysfunction. However, in the 2nd group, children have dilation but generally preserved function of the LV or moderate dysfunction.

The myocardium often shows evidence of ischemia, with increased echogenicity of the papillary muscles and the endocardium. This can be seen in all views but particularly in the apical four-chamber and parasternal views. The endocardium may be widely thickened and enhanced confusing with the diagnosis of idiopathic endocardial fibroelastosis (EFE). Thus Keith., *et al.* suggest that in all infant with suspicion of EFE, anomalous origin of coronary artery must be ruled out [2].

Mitral insufficiency can be moderate to severe, typically excentric.

Frequently, abundant intraseptal flow signals attracted the observer's attention. The detection of coronary septal collateralization probably is the key point for the diagnosis of late presentation of ALCAPA [12,13].

This collateral flow can be misdiagnosed as multiple muscular ventricular septal defects. Distinction from ventricular septal defects can be made by pulsed-wave interrogation, which reveals continuous flow in collateral arteries as opposed to systolic flow across ventricular septal defects. In addition, no color flow is seen entering the right ventricle in the presence of collateral arteries [12,14].

The identification of these septal collaterals is the initial clue to the diagnosis in patients aging more than 1 year. Thus, a dilated ostium of the RCA must urge the physician to meticulously look for intercoronary "steal" collaterals [14].

After these striking echocardiographic features, the examiner should concentrate on the direct signs of ALCAPA by studying the origin and size of coronary arteries.

It is best to visualize the coronary arteries in a high parasternal short-axis window [15,16].

The transducer should then be angled down toward the base of the heart. Color flow mapping is helpful and lowering the Nyquist limit permits greater sensitivity. In fact, when the color scale is lowered to 18 - 50 centimeters per second, it is possible to study the direction of coronary arterial flow [17].

Cohen emphasizes on the fact that we should avoid making diagnostic decisions based upon still images because this may lead to a false assurance of a normal origin of the coronary artery when, in fact, it arises anomalously from another location [18].

Normal coronary arteries exhibit antegrade flow and measure less than 3 mm in diameter [19].

If the ostium of the LCA cannot be visualized from the pulmonary trunk, we should pay attention to the proximal right pulmonary artery to exclude anomalous origin of LCA from right pulmonary artery [20,21].

An abnormal vessel inserting into the pulmonary trunk with retrograde flow toward PA confirms the diagnosis of ALCAPA.

Even if the insertion of LCA can't be determined exactly, it's important to reveal the continuous shunt in PA with a low-velocity, prominently diastolic flow pattern which suggests ALCAPA strongly [21].

In our two cases meticulous echocardiography suspected highly the diagnosis that was confirmed also in the operation time.

Echocardiography is also useful after surgical repair: it's needed to assess the recovery of left ventricular performance, as well as the resolution of mitral regurgitation.

Conclusion

ALCAPA is a rare congenital cardiac defect that presents a clinical diagnostic challenge. Awareness of this condition is essential for prompt recognition and early surgical intervention. The diagnosis should be suspected in infants who have dilated cardiomyopathy with electrocardiographic changes that suggest ischemia, and in children who have isolated mitral insufficiency.

Based on a step by step approach, two-dimensional echocardiogram combined with color-flow Doppler can diagnose anomalous origin of left main coronary artery accurately and give sufficient information for operation.

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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