

## Anomalous Origin of the Left Coronary Artery from the Pulmonary Trunk in a Seven-months Baby: Role of Myocardial Strain after Eight Years Follow-up

Thaiene Martins Miranda<sup>1</sup>, Denise Bibiana Masselli<sup>2</sup>, Roberta Longo Machado<sup>2</sup>, Yara Prosdossini Soares de Novaes<sup>2</sup>, Maria Lúcia Bastos Passarelli<sup>3</sup>, Luciana Braz Peixoto<sup>1</sup>, Claudia Gianini Monaco<sup>2</sup>, Manuel Adán Gil<sup>2</sup>, Carlos Eduardo Suaide Silva<sup>1,2</sup>

OMNI-CCNI Medicina Diagnóstica de São Paulo, São Paulo, SP - Brazil<sup>1</sup>, Diagnósticos da América AS (DASA) de São Paulo, São Paulo, SP - Brazil<sup>2</sup>, Santa Casa de Misericórdia de São Paulo, São Paulo, SP - Brazil<sup>3</sup>

### Abstract

Among some pathologies described in the literature, the anomalous origin of the left coronary artery is a cause of heart failure and myocardial ischemia in the early months of life. Here we report a case of a child who at seven months, when in open heart failure underwent echocardiography showed that the left main coronary artery originating from the pulmonary artery and reverse flow in this artery flow mapping in color. Underwent corrective surgery has successfully been followed for eight years, evolving with papillary muscle fibrosis and moderate mitral regurgitation.

**Keywords:** Heart Defects, Congenital; Heart Failure; Myocardial Ischemia; Coronary Vessel Anomalies; Pulmonary Artery/abnormalities; Echocardiography/diagnosis.

### Introduction

The anomaly of the left coronary artery is of rare congenital origin, usually diagnosed in the first month of life, as the child evolves with symptoms of heart failure and electrocardiographic and echocardiography alterations of myocardial ischemia<sup>1</sup>. In the absence of treatment and an adequate collateral circulation, most children, approximately 90%, die during their first year of life. On the other hand, in the presence of an extensive collateral network, patients can reach adulthood. There are reported cases of few symptomatic adults<sup>2</sup>.

Once detected, the CABG surgery should be recommended because of the high incidence of sudden death, cardiomyopathy, ischemia and arrhythmias associated with this anomaly.

We shall present a case in which the diagnosis was delayed at seven months with the child in clear heart failure, surgically corrected by Takeuchi procedure and subsequent progress follow up of eight years.

### Case Report

LGZ, female, seven months old, with clinical CHF and no previous diagnosis of any disease.

The echocardiogram observed significant dilatation and systolic dysfunction of the left ventricle and slight dilatation of the left atrium with mild mitral incompetence.

It was observed the right coronary artery originating normally on the right coronary sinus of Valsalva and the left main coronary trunk originating on the pulmonary artery (Figure 1-A). Color flow mapping showed reverse flow in the left coronary artery (Figure 1-B).

After diagnosis, the child underwent surgical correction by Takeuchi technique, which consists of the creation of an aortopulmonary window connecting the ostium of the left coronary artery with the aorta<sup>3</sup>. The child evolved with a progressive improvement of the symptoms and echocardiography alterations. About six months after surgery the ventricular function was practically normal and the mitral valve showed only a slight escape. It was possible to observe discrete gradient inside the pulmonary trunk of 29.5 mmHg on Doppler (Figure 2B). Throughout its evolution, the ventricular function was preserved, but the mitral regurgitation became more pronounced.

After eight years of development, the child lies in NYHA functional class I and with normal systolic function and mild to moderate mitral incompetence (Figure 2-D).

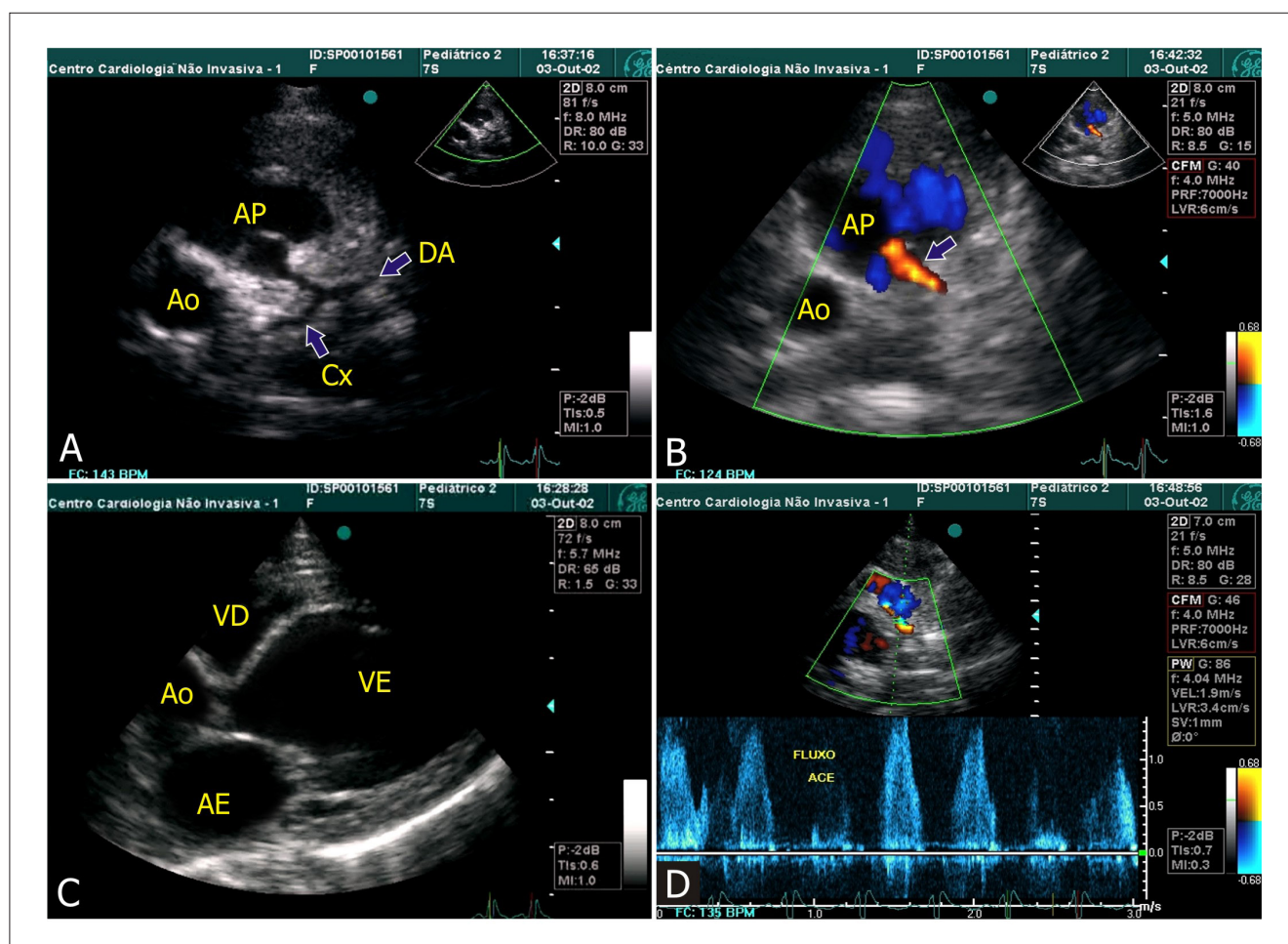
In the last echocardiography study the posteromedial papillary muscle showed increased refringence suggesting fibrosis (Figure 2-C) and the study of systolic function with speckle tracking technique for the evaluation of myocardial

**Mailing Address:** Carlos Eduardo Suaide Silva •

Rua Cubatão, 726, Postal Code 04013-002, São Paulo, SP - Brasil

E-mail: csuaide@cardiol.br

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**Figure 1** - (A) Left coronary artery (and its branches) originating in the pulmonary artery (Movie 1). (B) Flow color mapping showing reverse flow in the left coronary artery. (C) Parasternal longitudinal section showing important dilatation of the left ventricle (and important systolic dysfunction in Movie 2). (D) Pulsatile Doppler of the left coronary artery reserve flow. LA: left atrium, Ao: aorta; Cx: circumflex artery DA: anterior descending artery PA: pulmonary artery, RV: right ventricle, LV: left ventricle.

deformity (two-dimensional strain) demonstrated decreased deformity in the middle region of the inferior wall (Figure 3-B).

## Discussion

The anomalous origin of the left coronary artery was first described in 1886 by Brooks<sup>4</sup>. In 1933, it was then called Bland-White-Garland Syndrome by Bland et al.<sup>5</sup>. In this clinical syndrome, the left coronary usually emerges from the lateral wall or posterior to the trunk of the pulmonary artery. After child birth there is an increase in pulmonary vascular resistance and pulmonary artery pressure, which causes a desaturated flow of the pulmonary artery to the left coronary artery. In some cases there appears a collateral network from the right coronary to the left coronary artery, thus reversing the flow gradually. If this system of collaterals is developed satisfactorily one can visualize on the echocardiogram

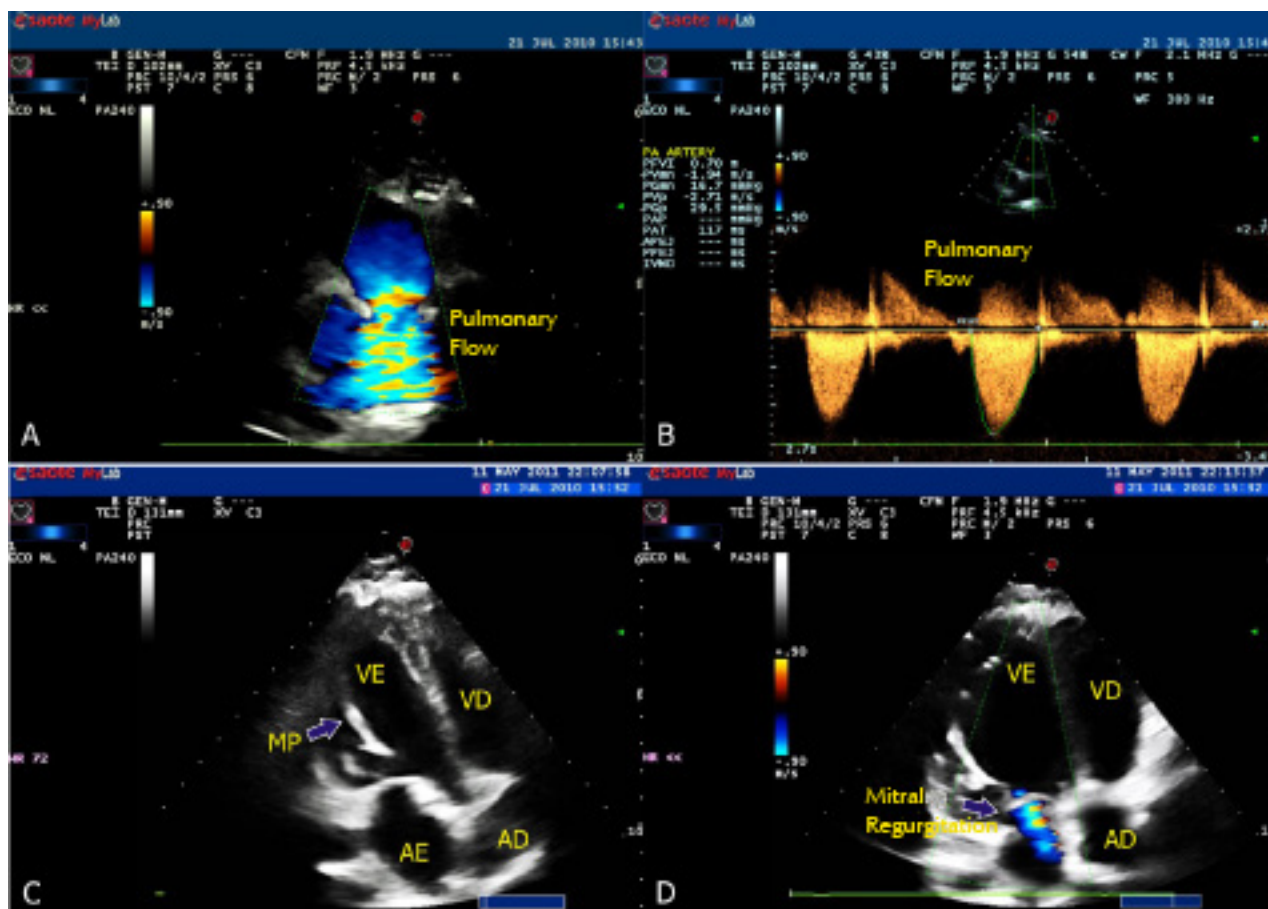
the left-right shunt in the pulmonary artery, resulting in a phenomenon of stolen flow of coronary arteries and late myocardial ischemia.

There are several indirect signs that can be observed on echocardiography with the progression of the disease. Among them, dilated cardiomyopathy and dilatation in the origin of the right coronary artery. Color Doppler may show a turbulent diastolic flow at the site of connection between the two arteries. Still on color flow mapping it is possible to visualize a retrograde flow in the left coronary artery (Figure 1-B).

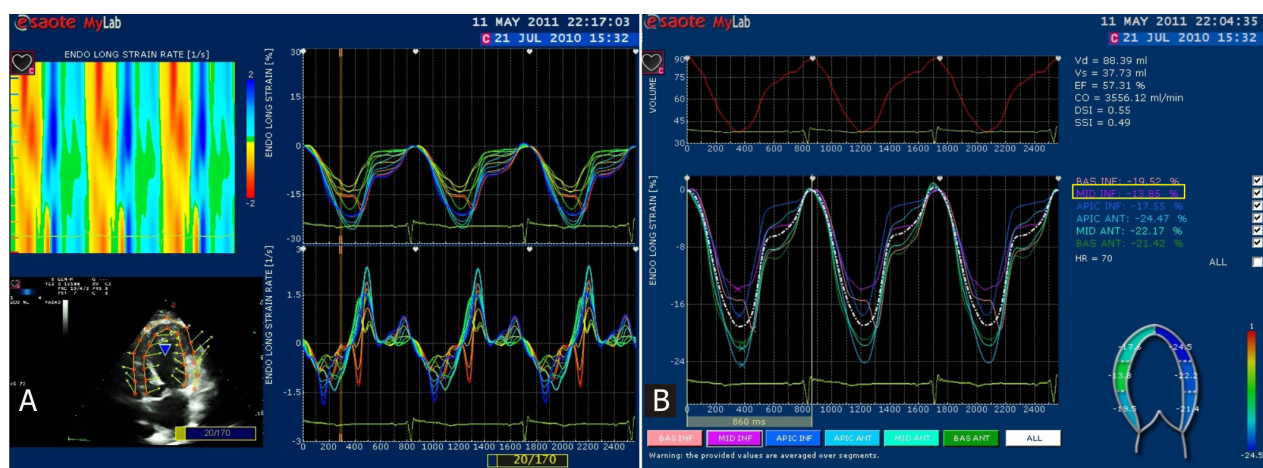
After the diagnosis, the treatment should be done as early as possible and the options are: direct reimplantation of the left coronary artery, use of anastomoses anastomoses and the creation of intrapulmonary tunnels (Takeuchi technique).

Currently Takeuchi surgery and direct reimplantation of ACE have become routine procedures; with low mortality rate and excellent long-term outcome, as in the case of

## Case Report



**Figure 2** - Late postoperative. (A) Turbulent flow in the pulmonary artery by Takeuchi surgery. (B) Continuous Doppler showing pulmonary peak systolic gradient of 29.5 mmHg. (C) Apical section of four-chambers showing posteromedial papillary muscle quite refractile. (D) The same section showing mitral regurgitation in color flow mapping.



**Figure 3** - (A) Map of the strain rate in the M mode (sup-left), two-dimensional mode (inf-left) and curves of strain and strain rate (right). (B) Curves of strain showing, in highlight, decreased deformity (13.85%) in the middle segment of the inferior wall (purple curve).

this report that despite the late diagnosis, the child had a satisfactory clinical evolution without interurrences, only a slight gradient inside the pulmonary trunk frequently observed in these cases<sup>6-8</sup>.

The patient showed a favorable evolution of global ventricular function; however, with the suspicion of fibrosis of papillary muscle at the dimensional echocardiography. a study was done with speckle tracking for assessment of regional deformity which confirmed the decreased value of

the myocardial strain in the middle region of the inferior wall (13.85%). We interpret this finding as a sequel resulting from the several months of ischemia to which this myocardium was subjected until the initial diagnosis.

The use of new echocardiography techniques (two-dimensional strain) can be useful in more detailed assessment of the regional myocardial function and confirm small changes in myocardial contractility suspected in the two-dimensional study even in the absence of global systolic dysfunction<sup>9</sup>.

## References

1. Braunwald E, Libby P, Bonow R O, Mann D L, Zipes D P, - Tratado de doenças cardiovasculares. 8ª ed. Boston:Elsevier; 2008.p.483-7.
2. Jacob NB, Salis FV. Anomalous origin of the left coronary artery from the pulmonary trunk in a 45-year-old woman. *Arq Bras Cardiol.*2003;81(1):199-201.
3. Lenzi AW, Solarewicz L, Ferreira WS, Sallum F, Miyague NI. Analysis of the Takeuchi Procedure for the treatment of anomalous origin of the coronary artery from the pulmonary artery . *Arq Bras Cardiol.*2008;90(3):167-71.
4. Brooks HS. Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: with some remarks upon the effect of this anomaly in producing cirroid dilatation of the vessels. *J Anat Physiol.*1885;20(Pt 1):26-9.
5. Bland E F, White P D, Garland J . Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. *Am Heart J .* 1933; 8: 787-801.
6. Sabinston D C, Neill C A, Taussig H B . The direction of blood flow in anomalous left coronary artery arising from the pulmonary artery. *Circulation.* 1960; 22: 591-7.
7. Cooley D A, Hallman G L, Bloodwell R D - Definitive qualified treatment of anomalous origin of the left coronary artery from pulmonary artery. *J Thorac Cardiovasc Surg.* 1966; 59: 798-808.
8. Lilje C, Lê TP, Ntalakoura K, Weil VJ, Lacour-Gayet F. Noninvasive follow-up of complications after the Takeuchi Operation. *J Am Soc Echocardiogr.*2007;20(12):1415.e3-4.
9. Lopes LM. Avaliação das artérias coronárias na criança. In: Silva CES. *Ecocardiografia: princípios e aplicações clínicas.* Rio de Janeiro: Revinter;2011.