An Insignificant Flow That Intrigues Echocardiographers. Echocardiographic Pattern in 15 Cases of Coronary-Pulmonary Fistula and Literature Review

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Introduction

Coronary fistulas are rare congenital anomalies found in up to 0.2% of coronary angiography scans, which may originate from any of the major coronary arteries and drain into the cardiac chambers or large vessels. They usually drain into low pressure chambers (right ventricle – RV) in 41% of the cases, into the right atrium (RA) in 26% and into the Pulmonary Trunk (PT) in 15% to 17%.[1]

They are defined as channels that communicate the coronary tree with a cardiac chamber or vessel, not going through the capillary bed.

Most patients have no symptoms, although dyspnea, angina, myocardial infarction, pulmonary hypertension, arrhythmia, endocarditis and sudden death have been reported in association with fistulas. Symptoms are usually related to the size of the fistula. There are often incidental findings in imaging scans.[1]

Our objective was to present the most frequent echocardiographic characteristics and patterns in 15 cases of flows suggestive of microfistulas.

Demographic data of patients and report of findings

From 2001 to 2016, we detected, through color Doppler echocardiography, in 15 cases, an intriguing diastolic flow into the PT. The patients were aged 3 months to 58 years (mean of 13.2 years; median of 11 years; standard deviation of 14.09; nine were female), all without any severe heart disease associated, referred for various reasons, such as heart murmur, routine cardiac evaluation and preoperative evaluation; one case was referred to echo under pharmacological stress for investigation of coronary artery disease.

The images were obtained in the cross-sectional parasternal projections at the aortic valve level, with identification of the flows by color flow mapping.

Keywords

Heart Defects, Congenital; Artero-Arterial Fistula; Diagnostic Imaging; Echocardiography, Doppler.

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The PT flow had similar characteristics: small diastolic flow, draining into different places and with an estimated maximum velocity of 3.16 m/s (Figure 1). Although this maximum velocity provides a gradient considered high for a low-pressure bed, we did not find any coronary dilation on the echocardiogram, according to the Z score.

We observed multiple drainage sites related to the proximity to the pulmonary valve and to the appearance of flow. After analyzing the collected data, we created a scheme illustrating the different patterns presented, which we describe below (Figure 2):

Type A: in eight cases, in the third proximal to the pulmonary valve on the PT side opposite to the aortic valve image (cases 2, 3, 6, 7, 8, 9, 11 and 13).

Type B: in two cases in the middle third of the PT, on the side adjacent to the aortic valve (cases 5 and 14).

Type C: in two cases in the distal third of the PT, on the side adjacent to the aortic valve and near the right branch emergence (cases 4 and 12).

Type D: in two cases in the distal third of the PT, opposite to the aortic valve and next to the left bundle branch emergence (cases 1 and 15).

Type E: in one case in the middle third of the PT, opposite to the aortic valve (case 10) (Figures 2 and 3).

In only one of the cases, the fistulous path was confirmed by cardiac catheterization, showing the origin of the fistula in the anterior interventricular artery and drainage into the third proximal to the pulmonary valve (Figure 4). The other cases were not submitted to cardiac catheterization because they had no clinical indication for it.

Discussion

The incidence of coronary fistulas among congenital heart diseases is very low, being reported in 0.2% to 0.4% of the cases.[1,2] Associated anomalies may occur and are described in the literature as persistence of patent ductus arteriosus, tetralogy of Fallot, ventricular septal defect and, also, acquired heart diseases.[2]

Most of the coronary fistulas originate from the right coronary artery (60%), followed by the anterior interventricular branch (35%), draining into low pressure chambers or vessels, such as RV and RA, coronary sinus, superior vena cava and PT.[3] However, different cases show distinct levels of prevalence. In an extensive review in adults, Said[4] reported the characteristics of 304 cases in the literature. Most of the

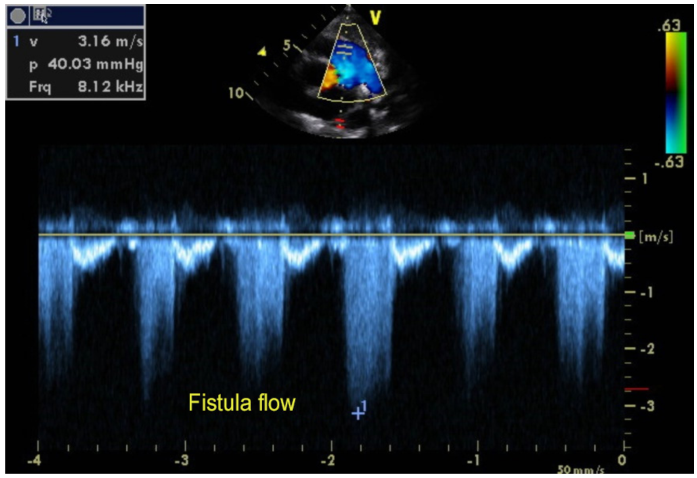

Figure 1 – *Coronary-pulmonary microfistula flow on pulsatile Doppler. Maximum diastolic flow velocity estimated at 3.16 m/s.*

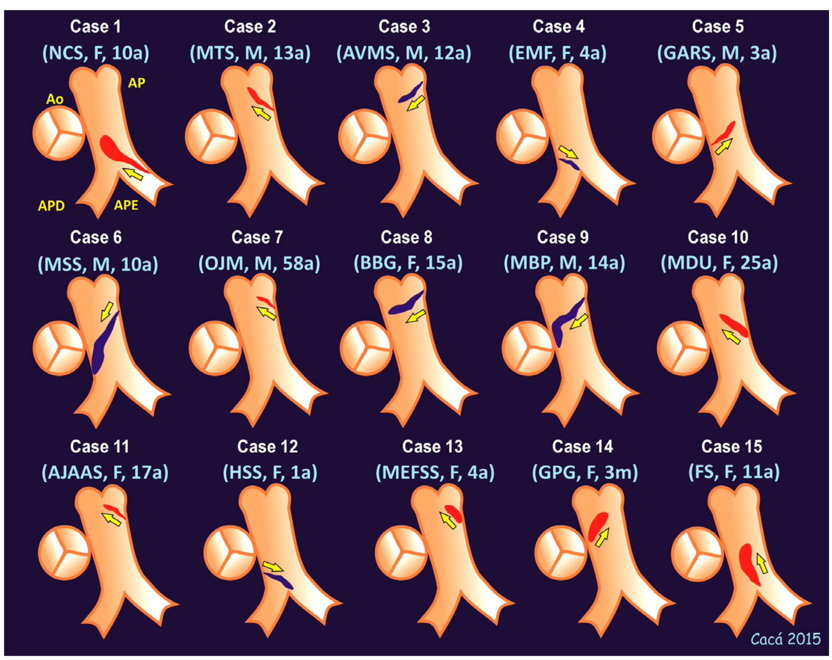

Figure 2 – *Design proposed to schematize the origin and direction of the fistula flow in the 15 cases. M: male; F: female.*

fistulas originating in the left coronary artery trunk and in the anterior interventricular artery flew into the PT (47% and 75%, respectively). Those originating in the circumflex artery had their final orifice in the coronary sinus or in the RA in 52% of the cases, while those originating in the right coronary artery had the same direction in 37% of the cases.[4] In the child population, literature is scarcer.

The cases presented here consisted mostly of children and adolescents who presented with microfistulas draining into the PT (17% of the cases, according to the literature)[1,5] – a rare combination that totaled only 15 cases in 16 years (Table 1). Our only case confirmed by catheterization was that of an adult who came to our service for an echocardiography under pharmacological stress to investigate coronary disease and presented the result of a cinecoronariography performed previously with the presence of microfistula originating in the anterior interventricular artery and flowing into the PT near the pulmonary valve (Figure 4). The echocardiogram revealed the small diastolic flow inside the PT with the same characteristics as the other 14 cases, only differing in site (case 7).

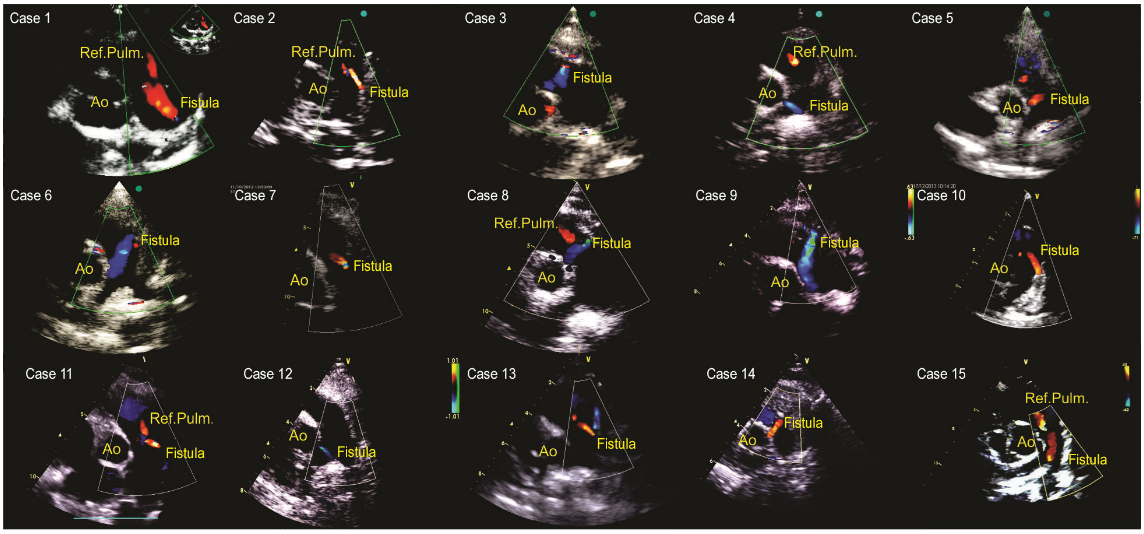

Figure 3 – *Echocardiographic images with color flow mapping of the 15 cases of coronary-pulmonary microfistula. Ao: aorta.*

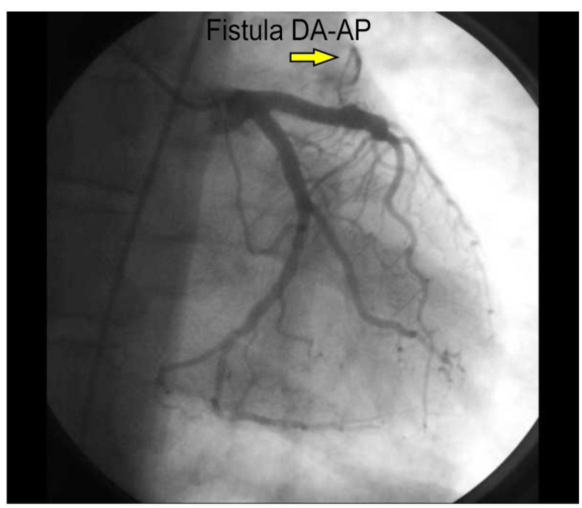

Figure 4 – *Image of the hemodynamic study of case 7 showing the microfistula (arrow).*

Doelder and Hillers published a very similar case of anatomy in a 35-year-old patient with atypical pain in the left submammary region, in addition to diffuse repolarization abnormalities on the electrocardiogram, with diagnosis confirmed by catheterization and multidetector tomography.[6] Because of the size of the fistula and its hemodynamic insignificance, the choice of conduct was clinical follow-up.

The importance of this report lies in the fact that the differential diagnosis of coronary artery fistulas should be made with persistent ductus arteriosus, aortopulmonary window, aortic insufficiency, ruptured sinus of Valsalva aneurysm and pulmonary or thoracic wall fistula.[2] These microfistulas, depending on their location, can often be mixed up with a small arterial canal (as in our cases 1 and 15) and, in these situations, the differential diagnosis between a microfistula and a closing canal is virtually impossible.

In general, differentiation is done according to the Doppler aspect, which, in the microfistula, usually presents only as a diastolic flow (Figure 1), whereas in the ductus arteriosus, it presents a continuous flow (the pressure gradient between the coronary artery and the pulmonary artery is done in the diastole, while from the aorta to the pulmonary artery occurs throughout the cardiac cycle). Moreover, the gradient between the aorta and the pulmonary artery is usually high – close to 100 mmHg (as long as in the absence of pulmonary hypertension) – whereas, in the microfistulas, it usually does not exceed 40 mmHg.

Table 1 – Description of the cases

Case	Name	Gender	Age	Indication
1	NCS	Female	10 years	Heart murmur evaluation
2	MTS	Male	13 years	Routine test
3	AVMS	Male	12 years	Routine test
4	EMF	Female	4 years	Heart murmur evaluation
5	GARS	Male	3 years	Heart murmur evaluation
6	MSS	Male	10 years	Preoperative evaluation
7	OJM	Male	58 years	Evaluation for cardiac ischemia (echocardiography under stress)
8	BBG	Female	15 years	Heart murmur evaluation
9	MBP	Male	14 years	Heart murmur evaluation
10	MDU	Female	25 years	Routine test
11	AJAAS	Female	17 years	Preoperative evaluation
12	HSS	Female	1 year	Heart murmur evaluation
13	MEFSS	Female	4 years	Heart murmur evaluation
14	GPG	Female	3 months	Heart murmur evaluation
15	FS	Female	11 years	Heart murmur evaluation

Today, with the experience acquired over the last 16 years, we have described, in our reports, the presence of a small diastolic flow in the PT, without hemodynamic repercussion, suggestive of a coronary-pulmonary microfistula.

Today, congenital coronary artery fistulas are often diagnosed non-invasively, semi-invasively or invasively.[5,7] These microfistulas do not usually cause symptoms, but on a routine medical examination, depending on their size, they can cause continuous murmur near their location. Only about 45% of the patients may present symptoms.[8] Treatment depends on the location size and the associated pathologies, but the occlusion of these in asymptomatic patients is controversial.[9]

Color Doppler echocardiography remains one of the most valuable diagnostic methods for the detection, in the intraoperative follow-up (when necessary), and in the evolutionary follow-up of patients with coronary fistulas.[5,10] We believe that the proposed classification may help in the follow-up and prognosis of patients.

Conclusion

The recognition of coronary-pulmonary microfistulas and their echocardiographic characteristics allows for correct diagnosis and reassures both the patient and the requesting physician as for its evolution.

Authors' contributions

Research creation and design: Silva CES; Data acquisition: Silva CES, Linhares RR, Peixoto LB, Pimentel MDT, Brecht L, Storti F, Gil MA, Monaco CG; Data analysis and interpretation: Silva CES, Aiello VD; Statistical analysis: Silva CES, Aiello VD; Manuscript writing: Silva CES, Aiello VD, Linhares RR, Peixoto LB; Critical revision of the manuscript as for important intellectual content: Silva CES, Aiello VD, Linhares RR.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate program.

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