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 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 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 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

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

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, Said4 reported the characteristics of 304 cases in the literature. Most of the
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. 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) — 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).
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. 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. 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.
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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