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Occlusion of a large fistula between the right coronary artery and the right ventricular cavity using Amplatzer® Vascular Plug II. Case report and technical description of the procedure

Oclusão de volumosa fístula da artéria coronária direita para a cavidade ventricular direita utilizando Amplatzer® Vascular Plug II. Relato de caso e descrição técnica do procedimento

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ABSTRACT – Coronary arteriovenous fistulas are rare anomalies that connect one or more coronary arteries to a cardiac chamber or other vascular structure. These communications can be occluded using percutaneous treatment, and there are currently several approaches and techniques. We report the case of an adolescent aged 12 years and 10 months, with a large fistula of the right coronary artery draining into the right ventricular cavity, in which the Amplatzer® Vascular Plug II device was used. The technical steps used are described in detail, with excellent immediate results.

Keywords: Congenital abnormalities; Arteriovenous fistula; Septal occluder device

RESUMO – Fístulas coronárias arteriovenosas são raras anomalias que conectam uma ou mais artérias coronárias a uma câmara cardíaca ou a outra estrutura vascular. É possível ocluir essas comunicações por meio de tratamento percutâneo, existindo atualmente várias abordagens e técnicas. Relatamos o caso de uma adolescente de 12 anos e 10 meses, com volumosa fístula da artéria coronária direita drenando na cavidade ventricular direita, no qual foi utilizado o dispositivo Amplatzer® Vascular Plug II. Descrevemos detalhadamente os passos técnicos utilizados, obtendo ótimo resultado imediato.

Descritores: Anormalidades congênitas; Fístula arteriovenosa; Dispositivo para oclusão septal

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INTRODUCTION

Coronary arteriovenous fistulas are rare anomalies that connect one or more coronary arteries to a cardiac chamber or other vascular structure, preferably in the pulmonary vasculature, and may also drain into other venous connections.[1] Some coronary fistulas tend to enlarge over time, whereas others remain trivial.[2] Hemodynamically significant fistulas may lead to ischemia of the myocardial segment perfused by the affected coronary artery, which might cause symptoms of heart failure or, possibly, volumetric overload in the right heart chambers, if the distal drainage site is the right ventricle, the pulmonary arteries or the coronary sinus. More rarely, they may also be complicated with endocarditis or rupture, cause hemopericardium and, consequently, cardiac tamponade.[3]

We describe a rare presentation of fistula between the right coronary artery and the free cavity of the right ventricle, causing overload in the right chambers, and the

occlusion of the fistula using an Amplatzer® Vascular Plug II device (Abbott Medical, Plymouth, MN, USA).

This study was evaluated and approved by a Research Ethics Committee, under CAAE 32012620.4.0000.5608.

CASE REPORT

An otherwise healthy patient, 12 years and 10 month-old, with good weight and height development. At 12 years of age, on a routine visit to a cardiac pediatrician, a heart murmur was noted, which was characterized as a continuous systolic-diastolic murmur, grade 4+/6+. The echocardiogram showed the presence of dilation of the right coronary artery ostium, with possible drainage into the free cavity of the right ventricle. A coronary computed tomography angiography confirmed the diagnosis, with the three-dimensional reconstruction showing a tortuous and large vessel (Figure 1), but the drainage site was difficult to define. The electrocardiogram revealed overload of the right ventricle, with an end conduction delay pattern (Figure 2).

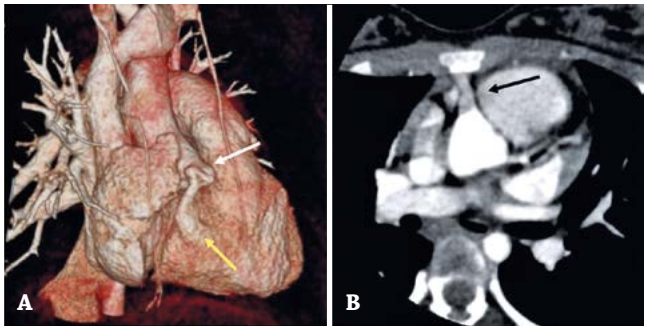


Figure 1. Coronary computed tomography angiography with three-dimensional reconstruction and in the axial plane. (A) Origin of the anomalous vessel (white arrow) with probable drainage into the right ventricle (yellow arrow). (B) Dilatation of the right coronary artery, with a tortuous proximal course (black arrow).

Figure 2. 12-lead electrocardiogram. Sinus rhythm, heart rate of approximately 80bpm, a 41° electrical axis, and end conduction delay are observed, characterizing overload of right chambers.

The procedure was previously planned, and detachable microcoils and vascular plugs of different sizes and diameters were requested, in case of stenoses in the anomalous vessel path that could hinder the deployment of larger-gauge devices.

Under general anesthesia, we punctured the right femoral artery and vein with 6F hemostatic valves. We performed right and left catheterization, with manometry analysis and sample collection to perform the oximetry run, which revealed an oximetric jump of 10% between the atrium and the right ventricle (73% of mean oxygen saturation to 81%, respectively). The calculated systemic-pulmonary flow ratio (Qp/Qs) was 1.75, showing the presence of moderate hyperflow in the right chambers.

The manometry showed normal pressures in the right and left chambers. Aortography revealed the presence of dilation of the origin of the right coronary artery, with proximal tortuosity and drainage of the arterial content directly into the free cavity of the right ventricle. The other chambers and arteries had a normal appearance. A selective injection of contrast in the left coronary artery revealed that the artery had normal origin and caliber, and that the lower left ventricular wall was irrigated by a continuation of the left anterior descending artery, a highly developed branch (Figure 3).

Then, we performed a systemic heparinization (100IU/kg) and administered cefazolin (30mg/kg/dose) for surgical prophylaxis. We selectively catheterized the anomalous right coronary artery with a 3.5 6F Judkins Launcher® guide ca-

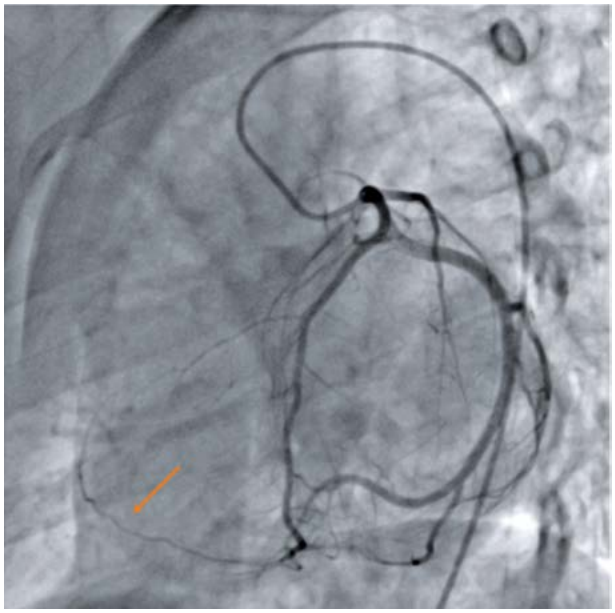
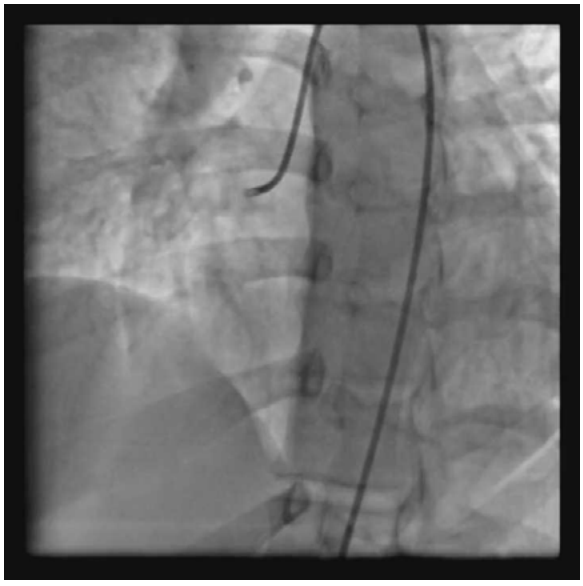


Figure 3. Angiography of the left coronary artery in the left anterior oblique/cranial projection. Normal caliber and course of the left coronary artery are noted. A continuation of the left anterior descending artery irrigates the lower left ventricular wall (orange arrow).

theter (Medtronic, Minneapolis, MN, USA) (Video 1). We used a 0.014"x300cm ChoICE™ Floppy guide wire (Boston Scientific, Marlborough, MA, USA) and navigated it through the fistula until we reached the pulmonary trunk. The next step was to use a 10mm diameter 4F ONE Snare® catheter (Merit Medical Systems, South Jordan, Utah, USA) for foreign body capture, which was inserted into the pulmonary trunk using another right coronary 3.5 6F Judkins guide catheter and, immediately after, we captured the 0.014"x300cm guidewire and externalized it in the intravenous 6F introducer, thus creating an arteriovenous rail.

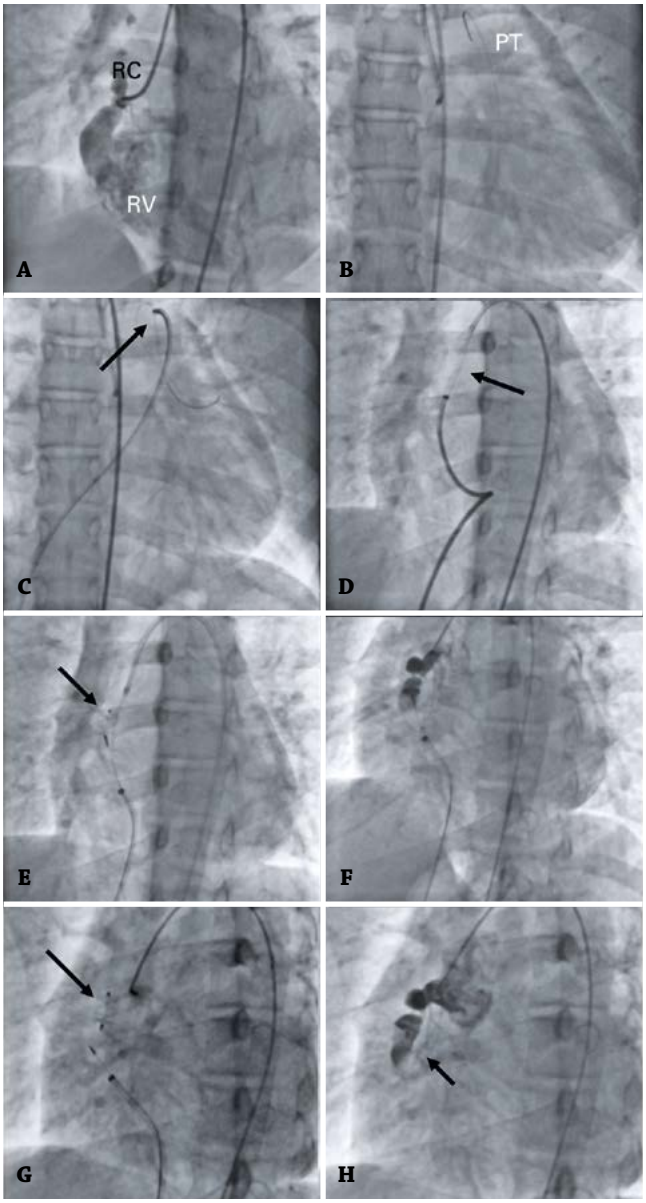


Video 1. Angiography of the right coronary artery in the left anterior oblique projection. The presence of an anomalous, tortuous vessel is noted, which is characterized as a fistula in the right coronary artery draining into the free cavity of the right ventricle.

Through the 0.014"x300cm wire, on the venous side, we inserted a 70cm 6F Flexor® sheath (Cook Medical, Bloomington, Indiana, USA) and carefully placed it in the most proximal portion of the fistula, with angiographic control by contrast media injections using the JR 3.5 6F guide catheter positioned at the arterial origin of the fistula.

As we visualized a stenotic segment in the proximal third of the fistula (3.7mm), we decided to use an 8mm Amplatzer® Vascular Plug II device, i.e., a device with a diameter that was approximately 100% larger than the smallest diameter of the vessel to be embolized, and positioned it proximal to the stenosis. It is important to keep in place the 0.014"x300cm guidewire during the positioning of the prosthesis, to maintain control of the fistula, and, in case of dissatisfaction with the chosen prosthesis size, or even with its correct positioning, to avoid the need of repeating all the steps for creating the arteriovenous rail.

When we were satisfied with the correct positioning of the device, we deployed it with a counterclockwise rotation of the release cable, under fluoroscopic monitoring (Figure 4). Control injections showed a slight residual shunt through the metallic meshes of the device, with adequate positioning thereof (Video 2).



RC: right coronary; RV: right ventricle; PT: pulmonary trunk.

Figure 4. Description of the procedure. (A) Selective catheterization of the right coronary artery and identification of the drainage site of the fistula into the right ventricle. (B) Anterograde placement of a 0.014"x300cm guidewire up to the pulmonary trunk. (C) Capture of the guidewire with a snare through the venous access to create an arteriovenous rail (arrow). (D) Advancement of a 6F 70cm sheath over the arteriovenous rail up to the proximal portion of the fistula (arrow). (E) Positioning of the 8mm Amplatzer® Vascular Plug II, keeping in place the arteriovenous rail, in case the device needs to be repositioned (arrow). (F) Anterograde angiography to control the correct positioning of the device. (G) Removal of the 0.014" wire and deployment of the prosthesis (arrow). (H) Control angiography showing slight residual flow (arrow).

Video 2. Control angiography of the right coronary artery in anterior left oblique projection reveals anomalous vessel occlusion using an 8mm Amplatzer® Vascular Plug II device, with a slight residual shunt at the end of the procedure.

Hemostasis of arterial and venous punctures was performed by usual manual compression, and the patient was referred to the pediatric intensive care unit for recovery, and was discharged from hospital 24 hours after the completion of the procedure.

The postoperative control echocardiogram showed no abnormalities in the pericardium, nor residual shunts draining into the right ventricle. A control chest X-ray revealed normal-sized cardiac area, prominent third arch, and presence of a metallic image compatible with the occlusive device in the right coronary artery topography (Figure 5). The patient was discharged from the hospital with a recommendation for indefinite maintenance of acetylsalicylic acid at a dose of 100mg/day.

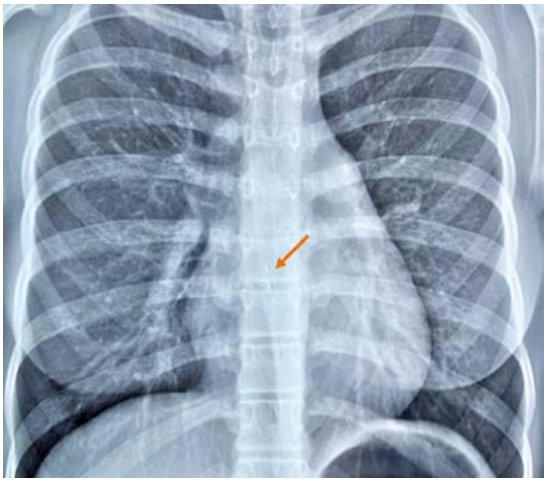


Figure 5. Chest radiography in anteroposterior view. After the procedure, the chest radiography shows prominence of the third arch, denoting pulmonary artery dilation, a prominent pulmonary vasculature, and an image compatible with a metallic device positioned on the topography of the right coronary artery (arrow).

DISCUSSION

Coronary fistulas are rare conditions, accounting for about 0.2% to 0.4% of all congenital heart diseases, and the first description of surgical treatment was made by Bjork and Craaford, in 1947.[4] Most fistulas are small and do not cause symptoms. Large fistulas, on the other hand, may cause several symptoms and complications, such as angina, dyspnea on exertion, weight/height deficits, recurrent respiratory infections, palpitations, and sudden death (by aneurysmal rupture).

With the advent of new, more flexible and lower-profile devices, percutaneous closure of these anomalous vessels has become easier and has more predictable results.[5] Although vascular plugs are devices ideally designed to occlude extracardiac vascular communications, their use for closing intracardiac defects, including arteriovenous fistulas and postoperative paravalvular leaks, possibly using diagnostic catheters (such as Amplatzer® Vascular Plug IV, Abbott Medical, Plymouth, Minnesota, USA), even if off-label, has produced encouraging results, with a reduction in periprocedural morbidity and rapid occlusion of embolized structures, due to its nitinol alloy multi-layered architecture and to the fast clinical recovery of patients.[6,7]

The same can be said of the use of controlled release detachable microcoils, ideally used for the occlusion of cerebral aneurysms, and which yield excellent results in very tortuous coronary fistulas with multiple obstructions in their course, since the adjuvant use of microguides and microcatheters allows navigation of tortuous vessels and deployment of embolizing elements (microcoils) in a safe and effective manner.[8,9] In this case report, since the anomalous vessel had no significant tortuosity or stenosed segments, microcoils were not opted for.

In conclusion, the occlusion of arteriovenous fistulas in the coronary territory with the use of new generation devices is a technically demanding, safe and reliable procedure, and these data are corroborated in the medical literature.[10-12] The knowledge and use of the several currently existing devices are important for selecting the ideal approach for each type of fistula.

SOURCE OF FINANCING

None.

DECLARATION OF CONFLICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: PTT and JFT; data collection: PTT, JFT, WG, VS and GR; data interpretation: PTT, JFT, LAG, VS, WG and GR; text writing: PTT, JFT and LAG; approval of the final version to be published: PTT, JFT, VS, LAG, WG and GR.

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