Percutaneous Closure of Iatrogenic Aortopulmonary Fistula Using the Amplatzer Septal Occluder®

Cierre percutáneo de fistula aortopulmonar iatrogénica con dispositivo Amplatzer Septal Occluder®

To the Editor,

Percutaneous treatment of stenotic lesions in the pulmonary artery system has been shown to be a valid and effective intervention, in both adults and children. 1-2 The development of an iatrogenic aortopulmonary fistula after pulmonary angioplasty is a rare complication described by several authors who have chosen different devices for percutaneous closure of the lesion. 1-6 The present letter discusses this uncommon lesion and considers the percutaneous treatment options.

A 13-day-old boy diagnosed with transposition of the great arteries underwent an arterial switch operation and a Lecompte maneuver. In the operating room, he was diagnosed with a coronary anatomy consisting of an intramural left coronary artery, from which the left anterior descending artery and the right coronary artery originated. There was an independent filiform circumflex artery. To correct the defect, a 3.5-mm PTFE tube was placed between the ascending aorta and the left coronary artery. The postoperative outcome was satisfactory, and the patient was discharged 14 days after the procedure.

In subsequent follow-up, the patient was found to have supravalvular pulmonary stenosis with an echocardiographic gradient of 62 mmHg. A cardiac catheterization procedure was performed when the patient was 1 year old. A stenosis gradient of 52 mmHg was detected, along with right ventricular pressures that were 66% of systemic pressures and stenosis at the origin of the right pulmonary artery (gradient, 18 mmHg). Percutaneous angioplasty of the pulmonary artery and origin of the right pulmonary artery was performed with a balloon catheter measuring 15 × 30 mm and 10 × 20 mm, respectively, using an 8-Fr introducer sheath. Disappearance of the gradient in the branch was confirmed and the gradient in the main artery decreased to 16 mmHg. There was also a mild tear in the intimal layer at the origin of the right pulmonary artery. However, magnetic resonance imaging showed the integrity of the wall of the main artery and branch.

The patient was asymptomatic for the next 4 years until signs of congestive heart failure developed. Echocardiography revealed a progressive increase in the size of the left heart chambers, along with continuous flow at the origin of the right pulmonary branch and retrograde flow in the descending aorta. Because an aortopulmonary window secondary to angioplasty was suspected when the patient was 5.5 years old and weighed 22 kg, cardiac catheterization and transesophageal echocardiography were performed. An 8-mm fistula was observed between the ascending aorta and the origin of the right pulmonary artery. The mean aortic and pulmonary pressures were 65 mmHg and 28 mmHg, respectively, with a QP/QS ratio of 2.3 and a suprapulmonary gradient of 15 mmHg and 8 mmHg at the origin of the right pulmonary branch. The atrial fistula was closed in a percutaneous procedure using a 9-mm Amplatzer® Septal Occluder device, chosen in view of the size of the lesion. The defect was probed from an aortic approach with a 4-Fr distal needle catheter and 0.014” hydrophilic guidewire. Once in the pulmonary lumen, an arteriovenous loop was created by capturing the guidewire with a 10-mm loop catheter, allowing the guidewire to leave via the femoral vein. Once the guidewire was in place, the distal needle catheter was advanced to place a more supportive guidewire (0.035”), over which the sheaf was placed (Amplatzer® 7 Fr Delivery System) and the device was deployed using the pulmonary approach. The procedure was a success; there were no incidents or need for repositioning (Figure 1 and Figure 2). After closure, a residual shunt through the device was observed (Qp/Qs = 1.5), without increasing either the pulmonary gradient or affecting the ascending aorta. The shunt could be detected in echocardiographic studies for 1 month. In addition, there was mild hemolysis that disappeared 6 days after the intervention without requiring any treatment. The patient was asymptomatic on the 5th day after the procedure. After 28 months of follow-up, there have been no new complications.

Percutaneous treatment of stenotic lesions of the pulmonary artery branch has been accepted as an effective and valid option. 1-2 There are isolated reports of patients with an iatrogenic shunt between the aorta and the pulmonary artery, especially in patients who have undergone an arterial switch, as was the case in our patient. 3-6 The etiopathogenesis has been attributed to the widely reported adherence between the aorta and pulmonary artery on performing the Lecompte maneuver in arterial switching. 5

The progressive development of symptoms of heart failure due to the iatrogenic window is due to the progressive increase in the size of the lesion, as explained by Vida et al 4 and as occurred in our patient.

When percutaneous closure is performed, care should be taken when choosing the device so as not to affect normal pulmonary valve function, to ensure sufficient coronary perfusion, and to keep obstruction of the lumen of both arteries to a minimum. Drug-eluting stents have been used during the closure procedure. 3,4 These are indicated in lesions close to pulmonary branching, as another device would lead to protrusion into the lumen of the pulmonary arteries. Amplatzer® Duct Occluder II devices and Amplatzer® Septal Occluder devices have also been used, 6 above all for small lesions. The reason for our choice was the better profile...
that adapts to the vascular lumen without occluding the lumen of the 2 vessels and the outflow of the conduct that irrigates the coronary arteries.

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Complex Diagnosis of Catecholaminergic Polymorphic Ventricular Tachycardia

Taquicardia ventricular catecolaminérgica polimórfica: una entidad de diagnóstico difícil

To the Editor,

Catecholaminergic polymorphic ventricular tachycardia (VT) is a cardiac conduction disorder whereby changes in intracellular calcium regulation increase susceptibility to ventricular arrhythmias with a consequent risk of sudden death despite a structurally normal heart. Affected individuals usually experience exercise-induced syncope and the arrhythmia is characteristically a bidirectional VT.1

The surface electrocardiogram usually shows no abnormalities and diagnosis is complex and based on 24-hour electrocardiographic monitoring and exercise testing. Epinephrine or isoproterenol tests are also useful. Even so, some cases remain undiagnosed despite a clinical manifestation in the form of serious ventricular fibrillation (VF), initially classed as idiopathic.2,3 Recently, genetic testing has become available. Mutations have been identified in up to 5 genes: the ryanodine receptor (RyR2) gene, which is the most common genetic abnormality, cardiac calsequestrin (CASQ2) gene,1 genes coding tight junction proteins, calmodulin gene, and KCNJ2.

The aim of the present study was to investigate the clinical characteristics and the usefulness of different diagnostic tests in a series of 9 patients with catecholaminergic polymorphic VT.

The reason for studying these 9 patients (mean age, 16 [standard deviation, 11.2] years; 55.5% women) was syncope in 7, resuscitation after VF in 1, and pathologic electrocardiogram with multiple ventricular extrasystoles in 1. VF was reported as part of the clinical course in 3 patients (33.3%), all before starting treatment with beta-blockers and after syncope. After therapy was started, no further arrhythmic events were reported except in patient 2, who had an appropriate shock on the only day he did not take beta-blockers (Figure).

No pathologic findings were reported in the electrocardiogram in 55.5% of the patients (Table). The mean QTc interval was 385 (SD, 26) ms (range, 347–425 ms) and the mean U-wave voltage was 0.14 (SD, 0.12) mV.

The complementary test that completed diagnosis was exercise testing in 44.4%, 24-hour Holter monitoring in 22%, epinephrine test in 11.1%, and genetic testing (including RyR2 and CASQ2) in 40%. Although symptoms were triggered by substantial physical or psychological stress in all patients, some patients did not have pathologic values in exercise testing or in the epinephrine test (Table). Interestingly, exercise testing was inconclusive for diagnosis in 3 of 7 patients (42.8%). These patients required the epinephrine or genetic tests (Table). In patients 6 and 7, exercise testing was not performed because bidirectional VT had been

Figure 2. Cardiac catheterization. Lateral aortography. A: Visualization of the window (arrows) between the posterior wall of the main pulmonary artery and right pulmonary artery and anterior wall of the aorta. B: Check for correct deployment of the device (asterisk) without affecting coronary perfusion, with residual shunt through the device. AO, aorta; RPA, right pulmonary artery.

REFERENCES


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