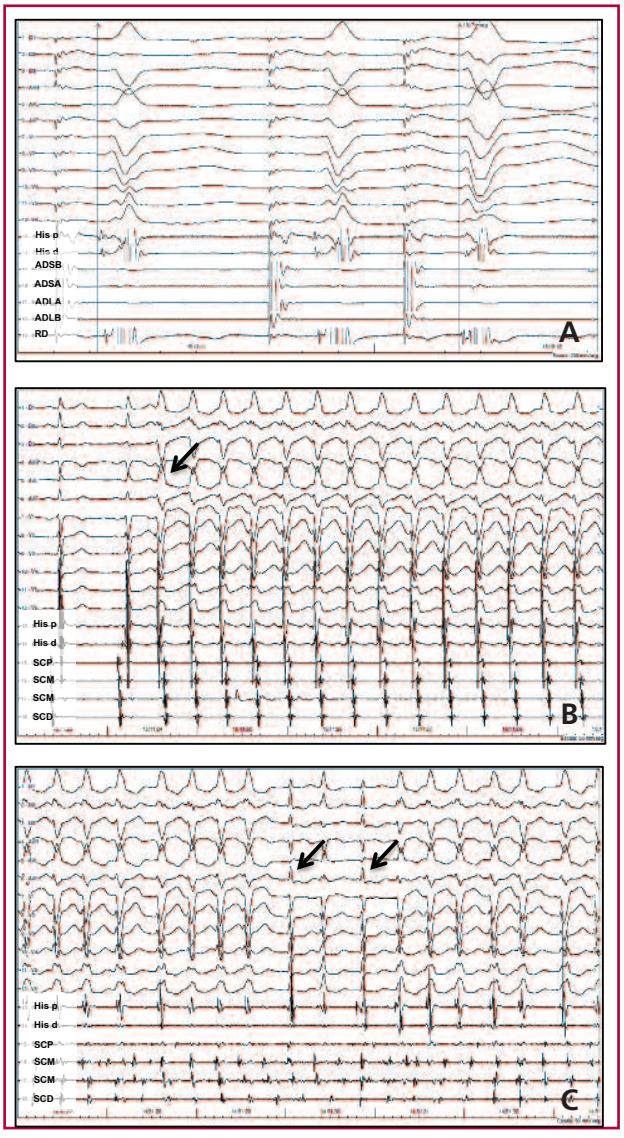

Fig. 2. A. Basal atrial stimulation with minimum pre-excitation and preferential conduction through the atrioventricular node (early His EGM activation with regard to the right branch). With the introduction of atrial extrastimuli (third beat), greater pre-excitation is evidenced, with inversion of ventricular septum activation sequence (EGM activation of the early right branch with respect to His). **B.** Antidromic tachycardia initiated with ectopic beats of the accessory pathway (arrow). **C.** Atrial fibrillation with different degrees of pre-excitation, and without pre-excitation in two beats (arrows). SCP: Proximal coronary sinus. SCM: Mid coronary sinus. SCD: Distal coronary sinus.

posing a difficult differential diagnosis with ventricular tachycardia.

Automaticity is an interesting aspect of this pathway, causing many symptoms in patients suffering from it. This aspect was first described by Kanter et al. in a patient with multiple episodes of nonsustained tachycardia during Holter monitoring. (1) Furthermore, Sternick et al. reported a series of 40 cases of Mahaim fibers, 12.5% of which had accessory pathway automaticity. (2) This property of the fibers resides in that part of the tissue presents functional and histological characteristics similar to those of the atrioventricular node. Proof of this is that pacemaker cells have been found in pathology studies. (3) It is also worth mentioning that ectopic beats with identical morphology to automaticity and/or pre-excitation were observed during radiofrequency, which involves close contact of the catheter with the accessory pathway, predicting a successful ablation, as was the case in our patient. (4)

Patients with accessory pathways are known to have a higher incidence of atrial fibrillation, initiating with episodes of supraventricular tachycardia. Mahaim pathways are related with this association, as seen in the present case.

In summary, we report the case of a female patient with different types of arrhythmias and pre-excitation syndrome due to Mahaim accessory pathway. A thorough diagnostic methodology allowed us to determine a causal relationship between all the arrhythmias and the accessory pathway, all in one, enabling the correct therapeutic approach.

Leonardo Celano[1], Claudio Hadid[MTSAC, 1], Darío Di Toro[MTSAC, 1], Edgar Antezana[1], Néstor Gorini[2], Carlos Labadet[MTSAC, 1]

[1] Department of Electrophysiology, Hospital General de Agudos "Dr. Cosme Argerich". Buenos Aires, Argentina
[2] Department of Cardiology - Hospital "Evita Pueblo". Berazategui, Pcia. de Buenos Aires, Argentina
e-mail: leonardocelano@gmail.com

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EDITOR'S NOTE: The following two scientific letters correspond to the evolution of the same clinical case, consecutively treated in two different institutions.

Multi-Imaging in Adult-Type ALCAPA anomaly

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare entity representing between 0.24% and 0.46% of congenital heart defects, with a prevalence of 1 in 300,000 live births. (1) Left to its natural evolution, its mortality rate exceeds 90% in the first year of life. Those who reach adulthood (10-15%) develop a large-caliber right

coronary artery with extensive collateral circulation, which provides reverse flow in the left coronary circulation draining in the main pulmonary artery. (2)

In general, evolution is asymptomatic, with a 2:1 female predominance. Dyspnea, trepidation, angina and fatigue are the most common forms of presentation in 66% of adult cases. Twenty-seven percent of the patients usually present with ventricular arrhythmia and sudden death in the subgroup of adult patients with mean age of 33±14 years.

All these findings correlate with the ischemic process, even in the absence of symptoms, and explain surgical repair as soon as the diagnosis is made, regardless of age. (2, 3)

Anomalous implantation of the left coronary artery was first reported in 1865 by Krause (4) and in 1885 by Brooks. (5) The first report in adults was discovered at autopsy of a 60-year-old woman following accidental death (5) in 1908. In 1933, a publication reported the case of a 3-month-old infant with autopsy-confirmed ALCAPA by Bland, White, and Garland. (6)

We report the case of a 39-year-old female patient, with history of syncopes in her adolescence during physical activity, and two uneventful natural childbirths at the ages of 24 and 28 years.

The patient reported dyspnea on exertion over the previous year. A gamma camera study detected fixed basal anterior defect, which was informed as fibrosis. Coronary angiography showed large-caliber right coronary artery, significant development of collateral circulation, and fistulae communicating with the anterior descending artery and the pulmonary artery tree, filling with contrast agent when injected from the right coronary artery.

On the first visit to our institution, the patient referred occasional functional class II dyspnea and trepidation over the past 12 months.

Physical examination revealed a protomesosystolic murmur in the pulmonary area, electrocardiogram in sinus rhythm, and right axis deviation, without ST-segment disorders.

Based on her symptoms, the patient received beta blockers, improving her clinical condition. Echocardiography, computed tomography scan and cardiac magnetic resonance imaging (MRI) were performed.

Although echocardiography is not the ideal study to make this diagnosis, it showed a large right main coronary artery (9 mm) (Figure 1 A); color Doppler demonstrated a continuous diastolic flow consistent with collateral branches in several myocardial segments (Figures 1 B and C). The ostium of the left coronary artery (LCA) emerging from the left coronary sinus of Valsalva was not visualized, but a 10-mm round image in the main pulmonary artery was detected in the lateral-posterior region of the main pulmonary artery (Figure 1 D) with predominantly diastolic flow, consistent with the origin of the LCA. Such image was close (6 mm) to the natural origin of the artery, an important point to consider when choosing the surgical procedure.

For better evaluation of the anatomy, a 256-slice CT scan and 3D image reconstruction was performed, showing a large-caliber (12.9 x 9.8 mm diameter) LCA originating from the lateral wall of the pulmonary artery (Figure 2 A and B), 11 mm from the valve plane, and 7.7 mm from the left sinus of Valsalva. The right coronary artery originated in the right Valsalva sinus with 9.6 x 13.5 mm diameter in its origin (Figure 2 C and D). A delayed enhancement cardiac MRI with gadolinium was performed, ruling out fibrosis. Biventricular systolic function was preserved.

Given this situation, with the confirmed diagnosis and the multiple images of the anomaly, and based on the publications of this rare entity, surgical correction of the defect was the treatment of choice due to the high risk of sudden death in these patients, regardless of the presence of ischemia or fibrosis.

There are three surgical techniques for adult patients with this rare condition. In the first place, physiologic restoration of the coronary flow is attempted with reimplantation of the LCA into the aorta. If that is

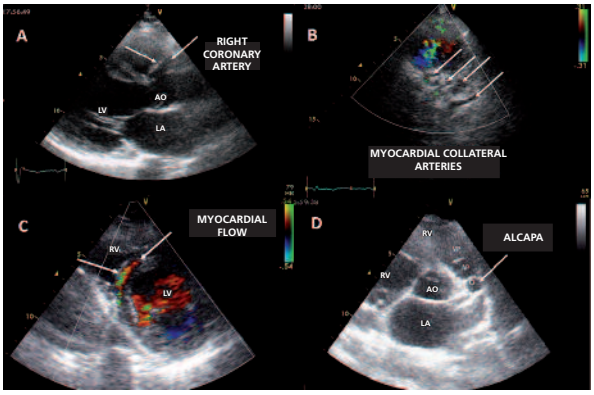

Fig. 1. See text for description.

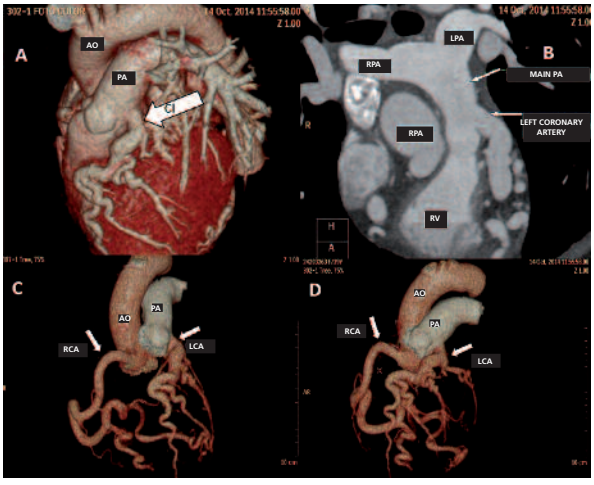

Fig. 2. See text for description.

not possible, mainly due to the LCA friable and fragile wall in adults, a venous or arterial bypass and closure of the LCA ostium in the pulmonary artery with a pericardial patch is performed. The least desirable surgical technique for these patients would be LCA ligation.

The patient was operated and LCA reimplantation into the aorta was performed. A Dacron tube graft was used for the anastomosis of the pulmonary arteriotomy; extracorporeal circulation and aortic cross-clamping times were 85 and 70 minutes, respectively. During the postoperative period, the patient presented with low cardiac output syndrome requiring high doses of inotropic agents and levosimedan infusion, probably due to myocardial stunning. The patient was finally referred to a center for circulatory support and possible heart transplant.

Gustavo Bernabeo[1], Eduardo Fernández Rostello[2], Carlos González[1], María José Bosaleh[3], Luciano Saglietti[1], Roberto Coronel[1]
[1] Sanatorio Sagrado Corazón. OSECAC
[2] Department of Echocardiography "Jonas Salk". OSECAC
[3] Department of Non-Invasive Imaging
Diagnóstico Maipú. CABA, Argentina
e-mail: gbernabeo@hotmail.com

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Rev Argent Cardiol 2015;83:447-449. http://dx.doi.org/10.7775/rac.v83.i5.6408

Cardiogenic Shock Post-Correction of Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

We report the case of a 40-year-old female patient with ALCAPA syndrome who underwent corrective surgery with complete dissection of the pulmonary artery, dissection of left main coronary artery (LMCA), reimplantation into the aorta, and extension of the pulmonary artery with 26-mm prosthesis (extracorporeal circulation time 85 min, cross-clamping time 70 min). The patient progressed with refractory cardiogenic shock during the first three postoperative days, so she was referred to our center for further evaluation, ventricular assistance and possible heart transplant.

On admission, the patient was hemodynamically unstable, requiring high doses of vasopressors and inotropics, and presented with multiorgan failure. Transesophageal echocardiography revealed severe left ventricular systolic function impairment, anterior and anteroseptal hypokinesis, without valvular or right ventricular involvement. Coronary angiography was performed to rule out coronary flow obstruction,

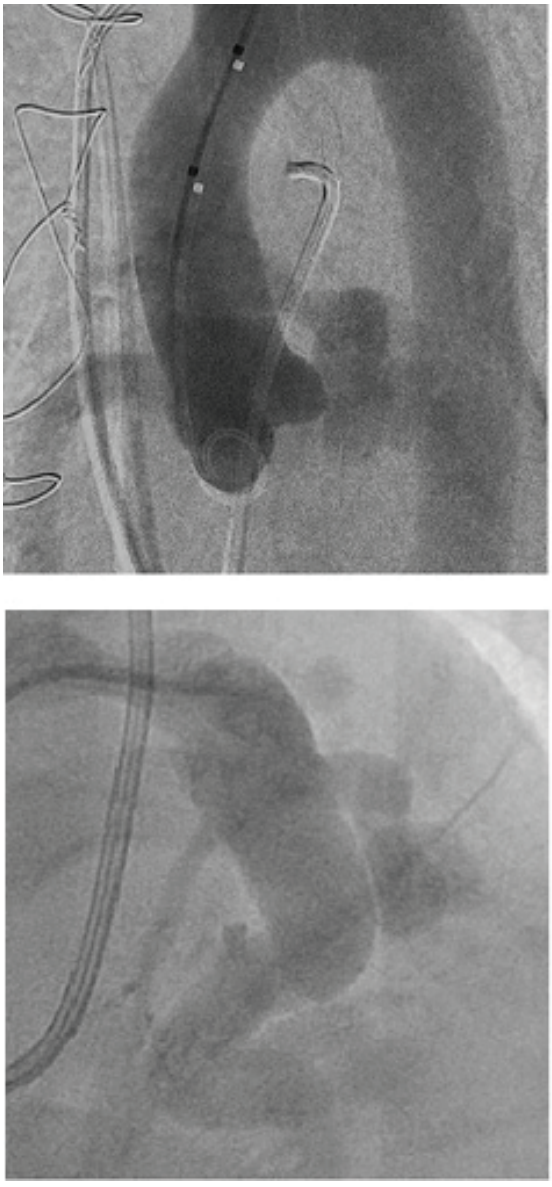

Fig. 1. A. Aortography. The image shows reimplanted left main coronary artery and right coronary artery (both tortuous and with large caliber), originating from the aorta. **B.** Right anterior oblique incidence showing the origin of the left main coronary artery.