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Celano, Leonardo; Hadid, Claudio; Di Toro, Darío; Antezana, Edgar; Gorini, Néstor; Labadet, Carlos

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Various Tachyarrhythmias via a Mahaim Accessory Pathway. All in One

We report the case of a 53-year-old female patient with a 10-year history of hypertension and trepidation, recurrent admissions due to wide QRS tachycardia requiring pharmacological cardioversion with amiodarone on several occasions, who was diagnosed with ventricular tachycardia. Holter monitoring showed multiple episodes of symptomatic tachyarrhythmia suggestive of nonsustained ventricular tachycardia (Figure 1 A) and the ECG revealed atrial fibrillation with wide QRS and left bundle branch block (LBBB) morphology, even under treatment with atenolol, amiodarone or flecainide. Baseline ECG exhibited sinus rhythm without preexcitation. Echocardiography and myocardial perfusion were normal.

The electrophysiological study exposed irregular episodes of wide QRS tachycardia and LBBB morphology, with nodal retrograde conduction decreasing at the bundle of His level (Figure 1 B). With incremental atrial overstimulation, progressive ventricular preexcitation with LBBB morphology was evidenced, associated with smaller increase in the A-delta interval than in the AH interval. Atrial extrastimuli at a fixed pacing train resulted in higher level of pre-excitation with inversion of the right bundle of His branch activation sequence, consistent with a Mahaim accessory pathway (Figure 2 A). Spontaneous antidromic tachycardia was observed, induced by ectopic beats mimicking tachyarrhythmia (Figure 2 B) or by programmed stimulation, which could not be entrained due to the interruption of the arrhythmia. In some cases, antidromic tachycardia degenerated into atrial fibrillation (Figure 2 C). There was 12/12 lead electrocardiographic correlation between the ectopic beats and antidromic tachycardia. Only nodal conduction was evidenced with right ventricular stimulation.

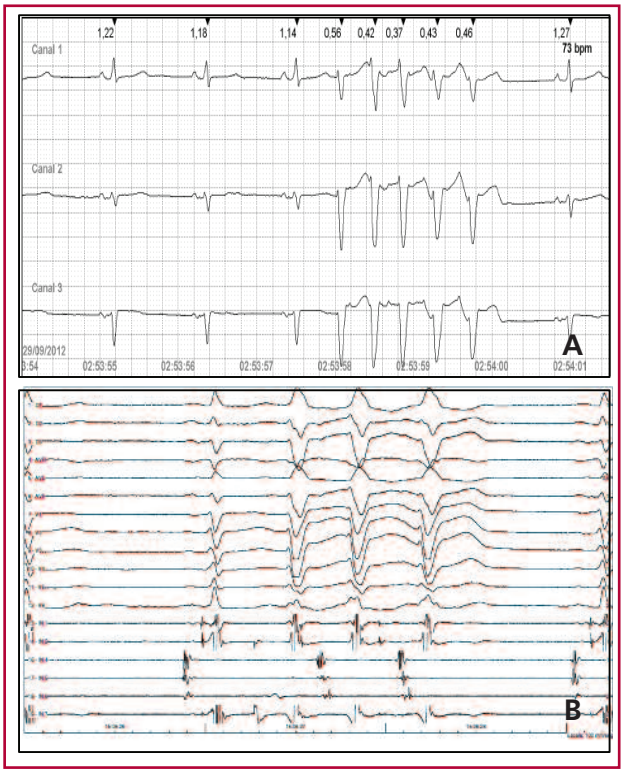

Fig. 1. A. Holter monitoring recording with wide QRS ectopic beats. **B.** Spontaneous ectopic beat tracing of the accessory pathway with decremental retrograde conduction due to prolongation of the HV interval (markers). RA: Right atrium. HL: High lateral. ML: Mid lateral. LL: Low lateral. RB: Right branch.

Following diagnosis of Mahaim accessory pathway, a mapping of the accessory pathway potential was performed during atrial stimulation with a 4-mm ablation catheter in the tricuspid annulus. The catheter was placed at hour 7 of the annulus; 50 W and 60 °C radiofrequency was performed with pre-excitation disappearance. The pathway was ectopic during radiofrequency application. Stimulation maneuvers were then carried out with no connection through the accessory pathway.

The patient made good progress with no recurrence during one-year follow-up.

This case shows all the arrhythmic episodes caused by a Mahaim accessory pathway in the same patient, including repetitive, isolated extrasystoles originated in the anomalous pathway, antidromic supraventricular tachycardia, atrial fibrillation and abnormal automatism caused by radiofrequency. All these arrhythmias disappeared after successful ablation of the accessory pathway.

Mahaim fibers are unusual atrioventricular connections that exhibit decremental antegrade conduction properties, located at the tricuspid annulus and distally inserting into the right ventricle at the fascicular level in the right branch or in the myocardium near it. These pathways cause antidromic tachycardia with wide QRS and image of left bundle branch block,

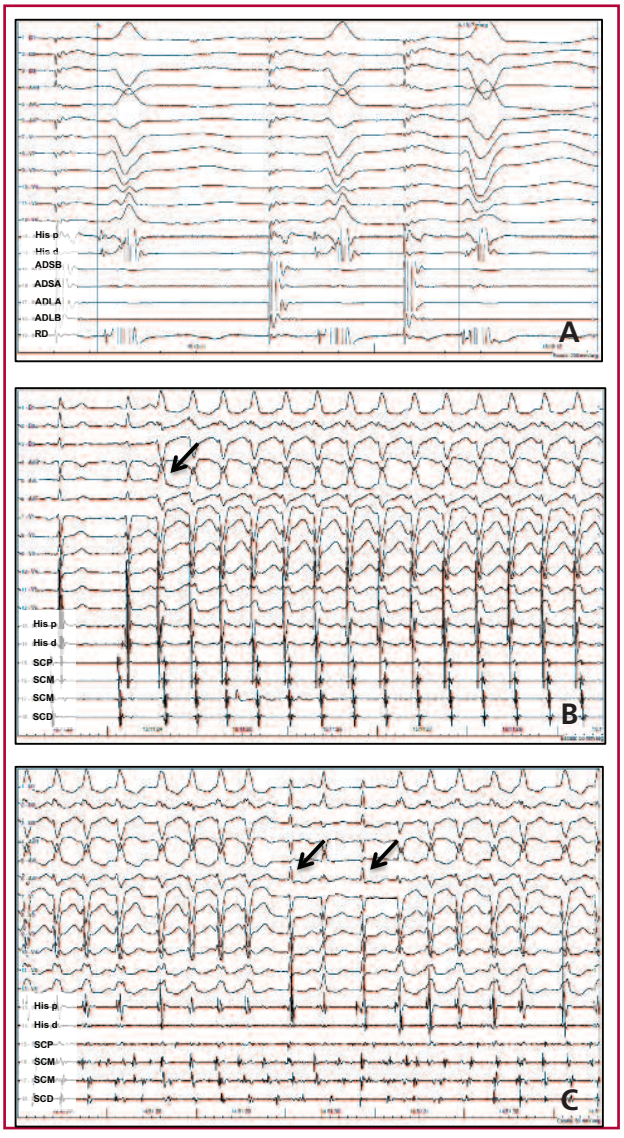

Fig. 2. A. Basal atrial stimulation with minimum pre-excitation and preferential conduction through the atrioventricular node (early His EGM activation with regard to the right branch). With the introduction of atrial extrastimuli (third beat), greater pre-excitation is evidenced, with inversion of ventricular 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