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Case Reports



## Idiopathic ventricular tachycardia of the aortic cusps: Review of the topic apropos of a case

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VT: ventricular tachycardia PVC: premature ventricular complexes

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#### ABSTRACT

Ventricular tachycardia of the aortic cusps is an idiopathic and infrequent tachycardia. Its arrhythmogenic mechanism is the activity triggered by late post-depolarizations. It predominates in young people and its course is generally benign. The radiofrequency ablation is an effective and safe procedure, reserved for patients with failure to antiarrhythmic drugs. We present a 24-year-old male patient, without structural heart disease and very frequent episodes of non-sustained ventricular tachycardia. A successful radiofrequency ablation of the arrhythmic focus located on the right coronary cusp was performed.

Key words: Idiopathic ventricular tachycardia, Aortic cusp ventricular tachycardia, Radiofrequency ablation

## Taquicardias ventriculares idiopáticas de las cúspides aórticas: Revisión del tema a propósito de un caso

#### RESUMEN

La taquicardia ventricular de las cúspides aórticas es una taquicardia idiopática e infrecuente. Su mecanismo arritmogénico es la actividad desencadenada por posdespolarizaciones tardías. Predomina en jóvenes y su curso es generalmente, benigno. La ablación con radiofrecuencia es un procedimiento eficaz y seguro, se reserva para los pacientes con fracaso a los fármacos antiarrítmicos. Se presenta un paciente masculino, de 24 años de edad, sin cardiopatía estructural y con episodios muy frecuentes de taquicardia ventricular no sostenida. Se realizó ablación exitosa con radiofrecuencia del foco arrítmico ubicado en la cúspide coronariana derecha.

*Palabras clave:* Taquicardia ventricular idiopática, Taquicardia ventricular de las cúspides aórticas, Ablación con radiofrecuencia

## INTRODUCTION

The idiopathic ventricular tachycardia (VT) takes place in the absence of

structural heart disease and represents 10% of VTs. About 80% originates in the ventricular outflow tracts and they are generated by the activity triggered by late post-depolarizations. They are more frequent in young people and generally have a benign course  $^{1-3}$ .

The most common form of presentation is the repetitive monomorphic (60-90 %), and it is characterized by frequent premature ventricular complexes (PVC), grouped in pairs, triplets and runs of nonsustained VT, which alternate with brief periods of sinus rhythm. Less common are the sustained VT episodes induced by exercise or emotional stress<sup>1,2</sup>.

The catheter ablation is an effective and safe procedure that should be considered in the event of failure of one or more sodium channel blockers, or when the patient does not wish to undergo prolonged treatment with antiarrhythmic drugs. The procedure should be performed in experienced centers, given the increased risk by mapping the aortic  $root^3$ .

Here is presented a patient without structural heart disease and very frequent episodes of nonsustained VT, who was performed a radiofrequency ablation of the arrhythmic focus located in the right coronary cusp. A bibliographic review of the subject is carried out.

## **CASE REPORT**

A 24-year-old male patient, with a health history, who refers frequent palpitations at rest and after physical activity. The physical examination did not show positive elements and the resting echocardiogram was normal. The electrocardiograms show frequent PVC, sometimes grouped into pairs, triplets and episodes of non-sustained VT (**Figure 1**).



The QRS complex in tachycardia is positive in  $D_{II}$ ,  $D_{III}$  and aVF, congruent with a location in the outflow tract. The precordial transition occurs in  $V_3$ , which is not a specific element to discriminate a right source from the left. However, the transition into tachycardia precedes the transition in sinus rhythm and there is a OS in the lead D<sub>I</sub>, compatible with a location in the left ventricular outflow tract. The algorithm in  $V_2$ , based on the proportion of the R wave in relation to the QRS in tachvcardia and in sinus rhythm [R/R+S (in tachycardia)]/ [R/R+S (in sinus rhythm)],was 0.75, also in favor of a left origin. During the ergometric test, the PVC disappeared in the maximum load and reappeared in the late recovery. An electrophysiological study and a radiofrequency ablation were the choice in this case.



**Figure 2.** Lead  $D_{II}$  and of  $V_3$ - $V_6$  of the surface electrocardiogram. Bipolar and monopolar registers distal and proximal filterers of the ablation catheter. Distal registration of right ventricular. Advancement of 14 ms of the ventricular electrogram of ablation relative to the earlier QRS complex, which coincides with a monophasic QS of rapid drop in the unipolar record. The bipolar record of the sinus beat shows a large ventricular deflection with a small atrial.

# Description of the electrophysiological study and the ablation

After local anesthesia in the right inguinal area, a double femoral venous puncture was performed with the Seldigner's technique. Through introducers 6 and 7F were placed, respectively, a quadripolar catheter of register in the apex of the right ventricular and a non-irrigated ablation catheter, of 4mm length of the distal pole, in the outflow tract of the right ventricular. The PVC were mapped, initially in the right ventricular, in the septal region and free wall of the outflow tract, as well as in the sub and supra-valvular pulmonary areas. Registers with precocities lower than 10ms were obtained, but the radiofrequency for more than 20 seconds in these sites was not effective. Given the close relationship between the right outflow tract and the aortic cusps, it was decided to map the outflow tract of the left ventricular by retrograde arterial tract.

In the aortic subvalvular area, at the level of the Valsalva sinus, the bipolar ventricular electrocardiogram of the probe catheter was of rapid rise and preceded 14 ms to the earlier ORS complex of the surface electrocardiogram. The distal unipolar unfiltered register showed a monophasic QS complex with a rapid negative deflection, coincident with the bipolar. The bipolar register of the sinus pulse showed a ventricular electrogram of high voltage, as a reflection of a far field coming from the activation of the right infundibulum and a small atrial electrogram, in correspondence with a far field of atrial activation of the anteroseptal portion of the tricuspid ring and right atrial appendage (Figure 2). In the right anterior oblique view, the mapping catheter was projected anterior and in the left anterior oblique, to the right (Figure 3). The bipolar registers of the PVC, and of the sinus rhythm as well as the fluoroscopic image, locate the ablation catheter at the level of the right coronary sinus.

There was performed a radiofrequency applica-



**Figure 3.** The ablation catheter (arrow) projects anteriorly into the RAO view, and rightwards in LAO; in relation to a location in the right coronary sinus. LAO (OAI, in Spanish), left anterior oblique; RAO (OAD, in Spanish), right anterior oblique.

tion with fluoroscopy, continuously for 90 seconds, with a maximum power of 30W and a cut temperature of 55°C. At 10 seconds of starting the radiofrequency, a progressive decrease in the frequency of the PVC was observed, until its total disappearance (**Figure 4**). After 30 minutes of the ablation completion and with isoproterenol infusion, there was no spontaneous induction of the tachycardia, not even with stimulation in the right ventricular (increasing frequency and programmed stimulation). After nine months of the procedure, the patient remains asymptomatic and with negative ergometric test.

## COMMENTARY AND REVISION OF THE TOPIC

More than half of idiopathic VTs originate in the outflow tracts and of these, approximately 80% are located in the right outflow tract<sup>4</sup>. Less common is the origin in the left, which includes the VT of the aortic cusps, of the ventricular myocardium located below the aortic valve, of the mitro-aortic continuity, of the upper basal interventricular septum (ventricular summit) and of the epicardial surface of the left ventricular outflow tract<sup>5,6</sup>.

The aortic valve has a central location and it is in contact or continuity with four cardiac chambers

and the remaining valvular apparatus (right ventricular outflow tract, pulmonary and tricuspid valve, interatrial septum, left and right atrial, mitro-aortic continuity and conduction system). It consists of three symmetric and semilunar valves (left coronary, right coronary and non-coronary). The recess of each of these cusps is known as Valsalva sinus. The trunk of the left coronary artery emerges from the left coronary sinus and the right coronary artery from the coronary sinus on that side, which is located posterior to the right and below the pulmonary valve. The right coronary sinus and the union with the left coronary sinus are placed immediately adjacent to the rear portion of the right ventricular outflow tract below the pulmonary valve. The noncoronary sinus is located posteriorly and in relation to the interatrial septum, where it is more frequent a site of origin of atrial tachycardia than of idiopathic  $VT^{7,8}$ .

The activity triggered by cyclic monophosphate adenosine, what produces late postpotentials, is the arrhythmogenic mechanism of these tachycardia; hence, it is also known as "adenosine-sensitive idiopathic VTs". The stimulation of the beta-adrenergic receptors increases the intracellular calcium concentration, which in turn, produces oscillations of the action potential during the fourth phase. The



increases of the heart rate, atrial or ventricular stimulation to high frequency and catecholamine infusion facilitate its induction. The tachycardia's termination is dependent of the direct blocking of the dihydropidinic receptors, the decreased levels of cyclic monophosphate adenosine, inhibition of betaadrenergic receptors and activation of A1 adenosine receptors<sup>1,2,9</sup>.

The adenosine-sensitive VT exhibits two phenotypic forms of presentation. The repetitive monomorphic is the most common (60-90%) and it is characterized by frequent PVC, grouped in pairs, triplets and runs of non-sustained VT, alternating with short periods of sinus rhythm. Usually, it occurs at rest and in periods following the exercise, and decreases during physical activity and can behave as constant. The paroxysmal form is characterized by sustained episodes of VT, precipitated by exercise or emotional stress and separated by long intervals in sinus rhythm, with infrequent  $PVC^{2,10}$ .

It usually occurs in women between 30 and 50 years, with a range between 6 to 80 years. The clinical course is benign and the prognosis favorable. The palpitations are the most common symptom (50%) and syncope is infrequent (10%). The sudden cardiac death is rare<sup>4,10,11</sup>. Tachycardias are triggered by caffeine, emotional stress or physical exertion, during recovery and typically exhibit a circadian variation with peak of occurrence in the morning and late afternoon<sup>12</sup>. In women, the hormonal influx (premenstrual or perimenstrual) can be a particular stimulus<sup>12</sup>.

Some patients are asymptomatic. The PVC or very frequent non-sustained VTs can develop, in 5-7% of patients, a reversible form of systolic ventricular dysfunction, similar to dilated cardiomyopathy induced by tachycardia. The interpolation of the PVC is reported as a risk factor for the development of cardiomyopathy<sup>13,14</sup>.

The diagnosis of idiopathic VT is by exclusion. The structural heart disease, dilated cardiomyopathy and coronary artery disease should be excluded. The differential diagnosis is important with other potentially malignant forms of VT, with the same origin in the outflow tract, such as arrhythmogenic dysplasia of the right ventricular and Brugada syndrome.

The close anatomical relationship between the different components of the output tracts complicates the precise location of the tachycardia site. The only common finding is the lower axis in the frontal plane (positive QRS complex in leads  $D_{II}$ ,  $D_{III}$  and aVF). In the VT of the right ventricular outflow tract of the transition zone in precordials, it is located in  $V_4$ ,  $V_5$  or  $V_6$ , while in those of the left tract it is located in  $V_1$  or  $V_2$ , with a specificity of 93%<sup>2,3</sup>.

The continuity between the rear portion of the right outflow tract and the anterior left ventricular outflow tract makes the transition in the lead  $V_3$  non-specific. Approximately 50% of VTs with transition in  $V_3$  ablate from the right outflow tract, while the remaining percentage is approached from the left, the Valsalva sinuses, the pulmonary artery and the epicardial area of the left ventricular outflow tract, through the percutaneous pericardial puncture<sup>15</sup>.

The VT of the aortic cusps may show a preferential conduction to the right outflow tract through isolated myocardial fibers. In this context, the algorithms for discriminating their origin lose trustworthiness. In a 20% of the coronary sinuses' VTs, the transition can take place beyond the  $V_3^{16}$ .

The relationship between the amplitude of the R wave and the total QRS amplitude in  $V_2$  in tachycardia and in sinus rhythm is useful to differentiate a right or left origin in patients with transition in the  $V_3$  lead, [R/R + S (in tachycardia)]/[R/R + S (in sinus rhythm)]. A ratio greater than or equal to 0.6 predicts a location in the left output tract with a sensitivity of 95% and a specificity of  $100\%^{17}$ . The earlier transition during tachycardia than during sinus rhythm and the morphology of QS in D<sub>1</sub>, also suggest a subaortic origin<sup>18</sup>.

The location of the aortic valve to the right and posterior in relation to the right outflow tract justifies a longer duration and amplitude of the R wave in V<sub>1</sub>, compared to V<sub>2</sub> in the VTs of the aortic cusps  $(R/QRS \text{ duration}>50\% \text{ and } R/S \text{ amplitude}>30\%)^3$ .

The VT of the left aortic cusp shows a triphasic morphology of the QRS complex in  $V_1$  (Mo W), a relationship of the R wave in  $D_{III}/D_{II}$ >0.9 and a QS or

rS in  $D_1$ . In the VT of the right cusp, there is an S wave deeper in aVR than in aVL and a high R wave in  $D_1$ , although in young patients with verticalized hearts, the QRS complex can be negative in this derivation lead. The morphology of the right bundle branch block excludes an origin in the non-right coronary sinus<sup>19,20</sup>.

In patients with symptoms or with ventricular dysfunction, the treatment with sodium channel blockers of the IC group (class I, level of evidence B) is recommended. The catheter ablation should be considered in the case of failure of one or more sodium channel blockers, or when the patient does not wish to undergo prolonged treatment with antiarrhythmic drugs (class I, level of evidence C)<sup>3</sup>. This procedure should be performed in experienced centers, given the increased risk in mapping the aortic root (damage in the coronary arteries and aortic valve, arterial embolism, cardiac tamponade and atrial-ventricular block).

The aim of the ablation is the location of the earliest endocardial activation in tachycardia or of the PVC. Mapping with stimulation is also a useful technique and with a high correlation with endocardial mapping. It is recommended the use of fluoroscopic mapping combined with electrodynamic threedimensional systems. The intracardiac echocardiography, contrast aortography and coronary angiography are useful in mapping the VTs of the aortic cusps; particularly, for identifying the location of the ablation catheter and its relationship with the aortic cusps and with the ostium of the coronary arteries<sup>21,22</sup>.

In some of the aortic cusps' VTs exist preferential conduction in the right ventricular outflow tract and under these conditions, the stimulation mapping is less reliable. When the local ventricular activation does not precede the beginning of the QRS complex in tachycardia, or the ablation is not effective in the right outflow tract, in spite of an adequate correlation in the stimulation map, the mapping of the aortic cusps should be considered<sup>7,21</sup>.

The location of a catheter in the area of the bundle of His in the right ventricular is useful in the ablation of the VTs of the coronary sinuses. The time from the local ventricular activation relative to the beginning of the QRS complex to the ventricular register of the His's catheter is significantly higher in the VT source on the left cusp and at the joint of the right cusp, compared with that originated in the right cusp and in the non-coronary cusp. It is also useful to limit the union between the right coronary cusp and the non-coronary, and to minimize the risk of damage to the conduction system  $^{2,23}$ .

When the registers on the left cusp are early, but the ablation is ineffective, the posterior pulmonary supravalvular and right anterior interventricular areas should be mapped. Rarely, the pulmonary supravalvular region can be approached from the left appendage. Similarly, the superior vena cava is located close to the supravalvular portion of the ascending aorta, above the right cusp<sup>24</sup>.

The electrogram registered in the right coronary cusp shows a ventricular electrogram of near field high voltage, which reflects the activation of the rear portion of the right ventricular outflow tract. In the specific VT of the right coronary sinus, there is -in tachycardia- a small and fractionated electrogram which anticipates the ventricular electrogram of high voltage, resulting from the activation of a small portion of the myocardium that extends into the cusp. An atrial far-field electrogram is registered, closer to the right and non-coronary sinus, representing the atrial activation of the anteroseptal zone of the tricuspid ring and the right appendage. In fluoroscopy, the catheter is oriented rightwards, in the left anterior oblique view, and anterior, in the right anterior oblique<sup>7,22,25</sup>.

The electrogram obtained at the left cusp is the most variable, but typically, the ventricular is larger than the atrial. As it is closer to the union with the right cusp, the ventricular electrogram grows, and to the left and posterior, decreases, and an atrial of far-field and high-voltage is registered, corresponding to the area of the left atrium near the anteroseptal mitral ring<sup>2,3,22</sup>.

A large atrial electrogram that reflects the activation of the interatrial septum is registered on the non-coronary cusp and one small ventricular, relative to the depolarization of the rear portion of the left output tract, the anteroseptal tricuspid annulus and supravalvular area. In the right anterior oblique view, the catheter is projected posterior. In the left oblique, it is difficult to distinguish the location in the non-coronary sinus of the left coronary sinus, since both are oriented to the left. If the torsion against clockwise registered a large atrial electrogram, then, the catheter is located in the noncoronary sinus<sup>2,3,22</sup>.

The main complication of ablation in the aortic cusps is the acute occlusion of the left main coronary artery. Therefore, the use of conventional non-irrigated catheters is recommended, of points with 4 or 5 mm, maximum power between 15-30 W, tem-

perature cut 50°C, application time between 30-60 seconds, in continuous fluoroscopy and with a margin greater than 6 mm between the ostium of the coronary arteries and the ablation site. The radiofrequency should be stopped in case of minimal dislocation of the exploratory catheter, or, if after 10 seconds of application, the tachycardia is not interrupted<sup>21,23,26</sup>.

The radiofrequency at the ablation site produces rapid PVC with morphology of the QRS similar to tachycardia, which gradually decreases until their complete disappearance; a finding with high specificity and low sensitivity. There are generally needed from 2 to 11 applications, with an average of 5. Sometimes, after several applications, it is noticed that the morphology of the QRS in tachycardia is not exactly the same, which suggests modifications in the exit site or a new focus of tachycardia. In this situation, close applications between 1 and 2 cm from the first site eliminate the VT with the second morphology of the QRS<sup>2,23,26</sup>. The immediate success of ablation is greater than 90% and recurrence occurs between 7-10%, usually within 24 to 48 hours<sup>2,3</sup>.

In patients with VT of the coronary sinuses, the ablation with radiofrequency is not the first choice of treatment because of the potential complications of the procedure, but it is effective and safe when performed in experienced centers.

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