

Voltage-directed cavo-tricuspid isthmus ablation using a novel ablation catheter mapping technology in a myotonic dystrophy type I patient

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A successful case of maximum voltage-directed cavo-tricuspid isthmus (CTI) ablation using a novel ablation catheter mapping technology in a myotonic dystrophy type I (DM1) patient is reported. The patient complained recurrent episodes of atrial flutter, revealed by the atrio-ventricular electrograms analysis during the routine pacemaker controls.

Key words: myotonic dystrophy, atrial flutter, atrial fibrillation, cavo-tricuspid isthmus ablation, microelectrodes

Introduction

Myotonic dystrophy type 1 (DM1) is an autosomal dominant disorder resulting from an amplified CTG trinucleotide repeat (> 50) in the 3-prime untranslated region of the dystrophin myotonia protein kinase gene (DMPK gene) on chromosome 19q13.3. DM1 is the most common muscular dystrophy of the adult life with an incidence of 1 in 8000 births and a worldwide prevalence ranging from 2.1 to 14.3/100.000 inhabitants (1). Cardiac involvement is noticed in about 80% of cases, often preceding the skeletal muscle one (2). Paroxysmal supraventricular tachy-arrhythmias (atrial fibrillation, atrial flutter, atrial tachycardia) represent a common finding on 12 lead electrocardiograms (ECG) or 24 hour ECG-Holter monitoring, with a prevalence up to 25% (3). The patients are often asymptomatic. Atrial tachycardias are observed in up to 7.3% of patients, both as un-sustained and sustained forms (4). Atrial fibrillation/flutter (AF/AFI) frequently

occurs in DM1 patients up to 17% (3, 4), some times representing the first manifestation of the muscular dystrophy in young patients (5). AF/AFI seems to increase mortality in this population (6). We report a successful case of maximum voltage-directed cavo-tricuspid isthmus (CTI) ablation using a novel ablation catheter mapping technology in a DM1 patient with recurrent episodes of typical atrial flutter.

Description

A 71-year-old woman with DM1 and arterial hypertension, with previously dual chambers pacemaker implantation for advanced atrio-ventricular block, was referred to our Division for device check and cardiologic therapy optimization. She never reported palpitations or arrhythmias-related symptoms, she didn't take anti-arrhythmic medications. ECG showed a dual-chamber VDD paced rhythm at 60 bpm. Transthoracic echocardiogram showed normal left ventricular systolic function associated to grade I diastolic dysfunction. The device interrogation showed several episodes of sustained supraventricular tachycardia, with a cycle length of 289 ms (Fig. 1). AFI diagnosis was made by 24 h ECG Holter monitoring that showed the common features of typical CTI dependent atrial flutter. The patient underwent electrophysiological study (EPS) for catheter ablation of the arrhythmogenic substrate. Two decapolar diagnostic catheters were placed in the coronary sinus and in the lateral right atrium wall. An IntellaTip MiFi XP® (IntellaTip

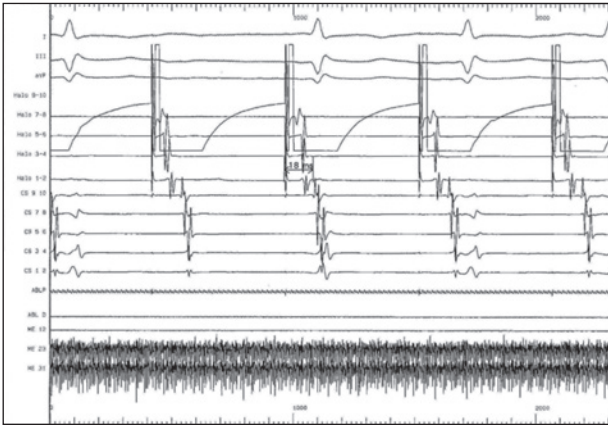


Figure 4. Baseline pacing trans-isthmus interval of 118 ms as measured from the pacing stimuli on electrode 1/2 of the HALO catheter to the atrial electrogram detected by electrode 9/10 on the coronary sinus catheter. In basal conditions the electrical impulse from the posterolateral tricuspid spreads around the tricuspid with two wave fronts, one in a clockwise direction (along the side wall) and another counterclockwise (along the rear wall and medial).

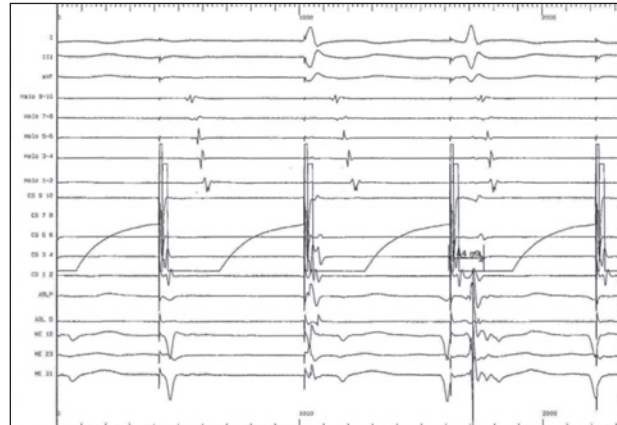


Figure 5. Post-procedure increased trans-isthmus interval as measured from the pacing stimuli on coronary sinus electrode 9/10 (144 ms) to the atrial electrogram detected by electrode 1/2 on the Halo catheter. The electrical impulse is forced to activate the atrial peritricuspid myocardium only counterclockwise.

for 40-60 s (60-70 W, 60-70 uC); this process was then repeated and continued until all sharp atrial electrograms detected on the mini-electrodes along the CTI were eliminated and only low-voltage double potentials were visible. A total of 3 radiofrequency ablation lesions were applied.

We performed a post-procedure evaluation of pacing intervals from both electrode 9/10 of coronary sinus catheter and electrode 1/2 of the HALO catheter. We showed an increased trans-isthmus interval measured by both pacing from coronary sinus electrode 9/10 to the atrial electrogram detected by electrode 1/2 on the Halo catheter and from HALO electrode 1/2 to the atrial electrogram detected by electrode 9/10 on the coronary sinus catheter, of 144 and 140 ms, respectively confirming the bidirectional block (Figs. 5, 6).

The procedure was completed without complications. Six months later, the ambulatory interrogation of the device did not reveal any recurrent atrial arrhythmias; so no long-term anticoagulation therapy was necessary.

Discussion

Cardiac involvement in DM1 patients occurs as a degenerative process with progressive fibrosis and fatty replacement of the myocardium, not limited to the specialized conduction system, but involving also areas – initially unaffected – of the atrial myocardium (8). This anatomopathological substrate, causing the discontinuous and

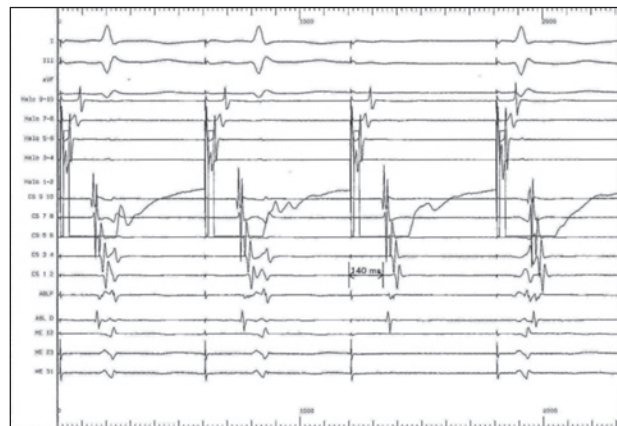


Figure 6. Post-procedure increased trans-isthmus interval as measured from HALO electrode (140 ms). The electrical impulse provided by the posterolateral wall side is forced to activate the peritricuspid myocardium only clockwise.

inhomogeneous propagation of sinus impulses and the prolongation of atrial conduction time, may facilitate the onset and the perpetuation of atrial arrhythmias in DM1 patients (9-12), as usually it happens in other clinical conditions (13-18). The modern pacemakers, including detailed diagnostic functions and therapeutic algorithms, may facilitate the diagnosis and management of frequent paroxysmal atrial tachy-arrhythmias that may remain undetected during conventional clinical follow-up (19-29). Radiofrequency ablation of typical atrial flutter is a very successful procedure with reported acute success rates of 90-95%. The conventional ablation technique requires cre-

ation of a complete line of conduction block across CTI, from the tricuspid valve to the inferior vena cava. Such an approach has a high overall success rate but results in variable, sometimes lengthy procedure times, mainly due to anatomic variation (30-35). The “maximum voltage-guided” (MVG) ablation technique targets high-voltage isthmus electrograms to ablate the functionally important anatomic muscle bundles alone, without drawing a complete anatomic line. Previous reports suggest that this method reduces the mean ablation time and shortens the procedure duration (36, 37). The ablation tip of IntellaTip MiFi catheter may enhance the available data for such a signal dependent technique. In this catheter, bipolar signals can be recorded between the three 0.8 mm-wide electrodes that are arranged radially 1.3 mm from the end of the catheter, alongside the standard distal and proximal bipolar recordings. The use of the Intellatip MiFi radiofrequency ablation catheter has been suggested to improve mapping resolution with a more precise localization of the points with the highest amplitude, potentially allowing less but more effective RF application in less time, in a technique that relies on signal amplitude (38). These features make the maximum voltage guided cavo-tricuspid isthmus ablation using a novel catheter with three mini electrodes within the ablation tip (IntellaTip MiFi, Boston Scientific, Boston, MA) particularly suitable for efficacy and safety during the ablation procedure in DM1 patients with an higher potential risk of complications for the diffuse atrial fibrosis. This novel ablative approach is particularly useful in this category of patients for their inability to maintain for long time the supine position.

Conclusions

We showed a successful case of maximum voltage-guided cavo-tricuspid isthmus ablation using a novel catheter with mini electrodes within the ablation tip in a myotonic dystrophy type 1 patient with typical atrial flutter episodes revealed by atrio-ventricular electrograms analysis during the routine pacemaker controls.

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