

CASE REPORT

Usefulness of the external loop recorder in a patient with Myotonic Dystrophy type 1 and recurrent episodes of palpitations: evaluation of the follow-up from diagnosis to 6 month-post-cardiac interventional treatment

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A patient with Myotonic Dystrophy type 1 and recurrent episodes of palpitations is reported, in which the application of an external loop recorder (ELR) was useful for the diagnosis and post radiofrequency ablation follow-up of typical atrial flutter.

Key words: Myotonic Dystrophy, atrial flutter, atrial fibrillation, cavo-tricuspid isthmus radiofrequency ablation, mapping technology, external loop recorder

A 17-year-old male patient affected by Myotonic Dystrophy type 1 (DM1) and recurrent episodes of palpitations was referred to our Division for an arrhythmologic evaluation. He didn't take antiarrhythmic medications. ECG showed sinus rhythm at 65 beats/minute (bpm), atrio-ventricular (PR interval: 120 ms) and intraventricular (QRS duration: 90 ms) conduction within the normal limits, and no ST-T abnormalities (QTc interval: 360 ms). The transthoracic echocardiogram showed a normal ventricular systolic function (Simpson's biplane ejection fraction: 60%) and normal left atrial size. Three conventional 24-hour Holter monitoring, performed during the previous three months, didn't show significant arrhythmias.

Considering the persistence of the symptomatology, a thirty-day high capacity memory external-loop-recorder (ELR, SpiderFlash-T R, Sorin CRM, France) was applied. The recorder was connected to the thorax by lead wires and disposable adhesive electrodes. The patient was trained to

change them daily after taking a bath, as far as to press a button on the recorder to store ECG tracings in case of symptoms. Otherwise, no other manipulation was required. The patient was asked to also annotate on a special diary, the onset of palpitations. After twenty days, the ELR interrogation showed a sustained episode of supraventricular tachycardia at 155 bpm with the common features of typical atrial flutter, in concomitance with the onset of palpitations (Fig. 1).

The patient underwent electrophysiological study (EPS) which confirmed the diagnosis of cavo-tricuspid isthmus (CTI)-dependent atrial flutter (AFI) (Fig. 2). A voltage-directed cavo-tricuspid isthmus ablation was performed using a novel ablation catheter mapping technology (IntellaTip MiFi, Boston Scientific, Natick, MA) in order to only ablate the conducting bundles of the CTI (Fig. 3). In fact, according to previous experiences (1) we avoid to ablate the intervening non-conducting fibrous tissue. The procedure was completed without complications.

At discharge from the hospital, an ELR was applied to the patient for post-ablation ECG monitoring. At six months follow-up, no experienced episodes of palpitations were reported by the patient nor supraventricular arrhythmias were documented at the ELR monitoring.

Discussion

DM1 is the most common muscular dystrophy of the adult life. The cardiac involvement is noticed in about 38-

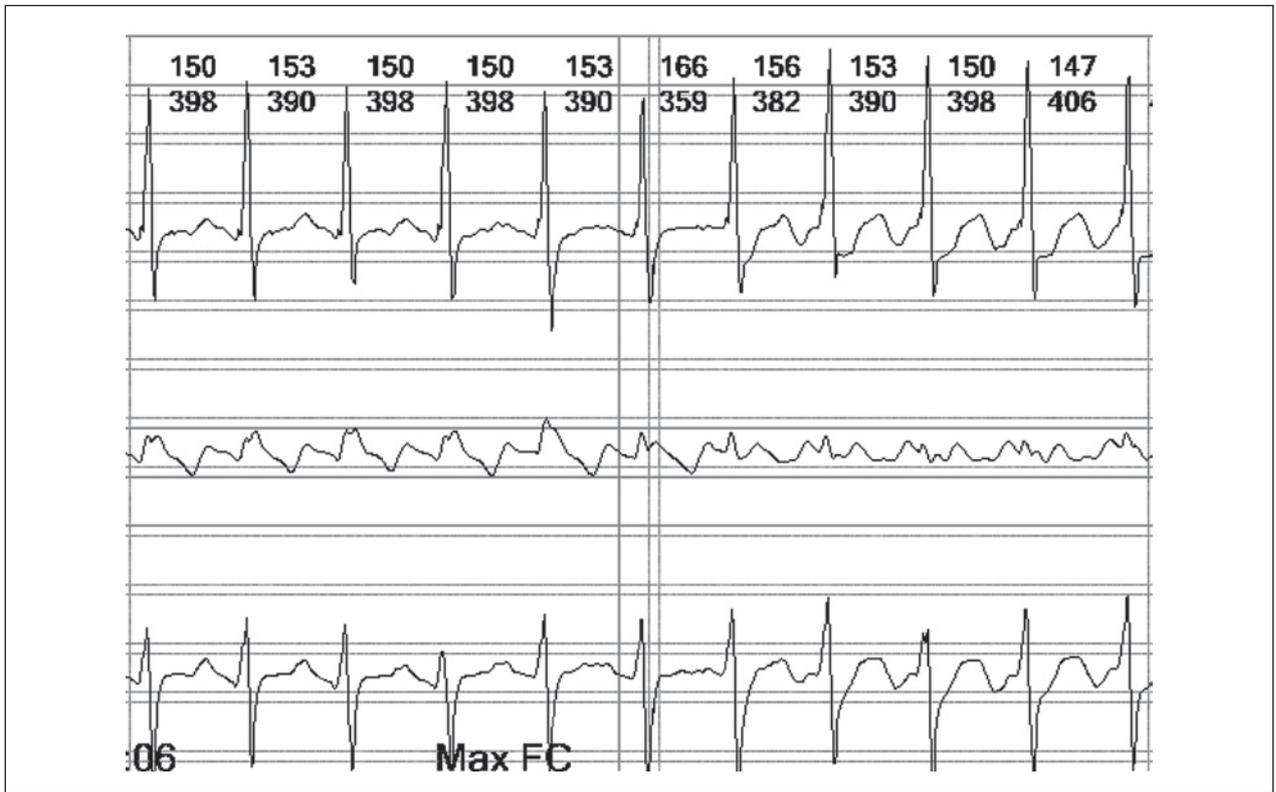


Figure 1. Sustained episode of supraventricular tachycardia compatible with typical atrial flutter diagnosis.

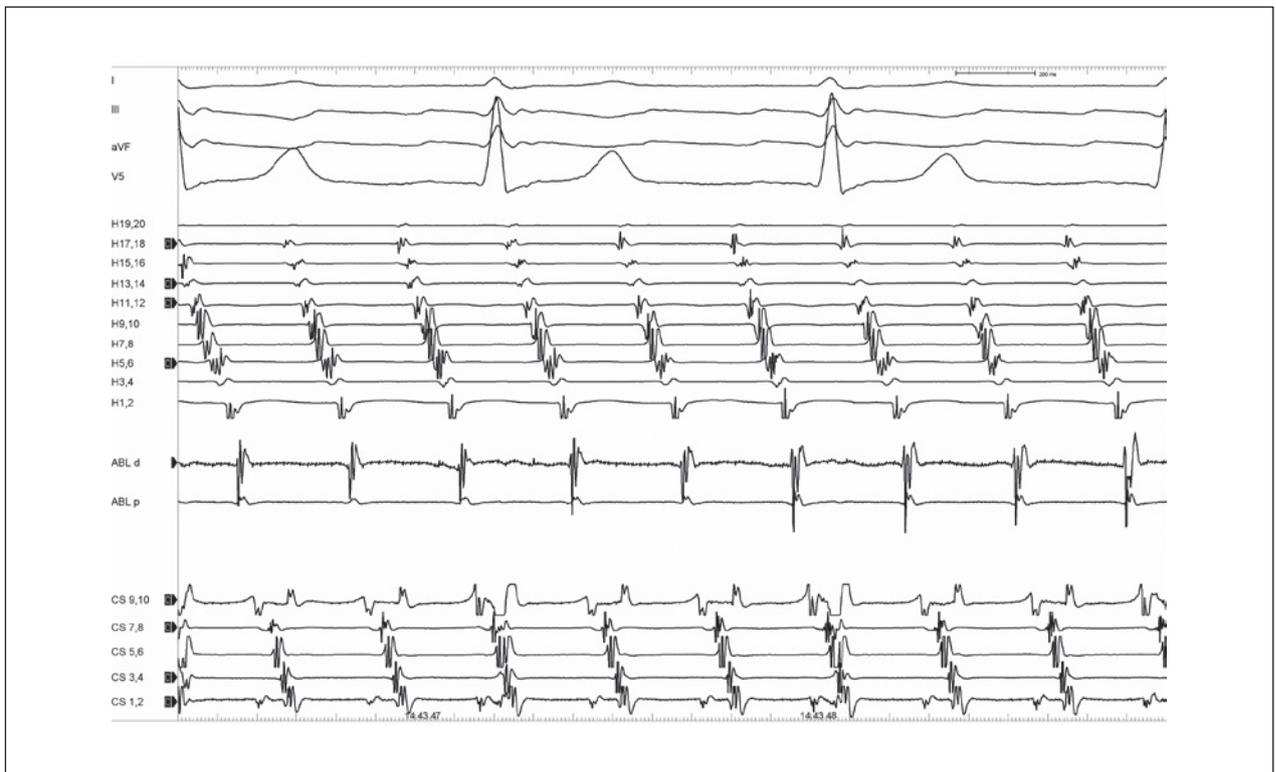


Figure 2. Typical counterclockwise atrial flutter electrograms at electrophysiological study.

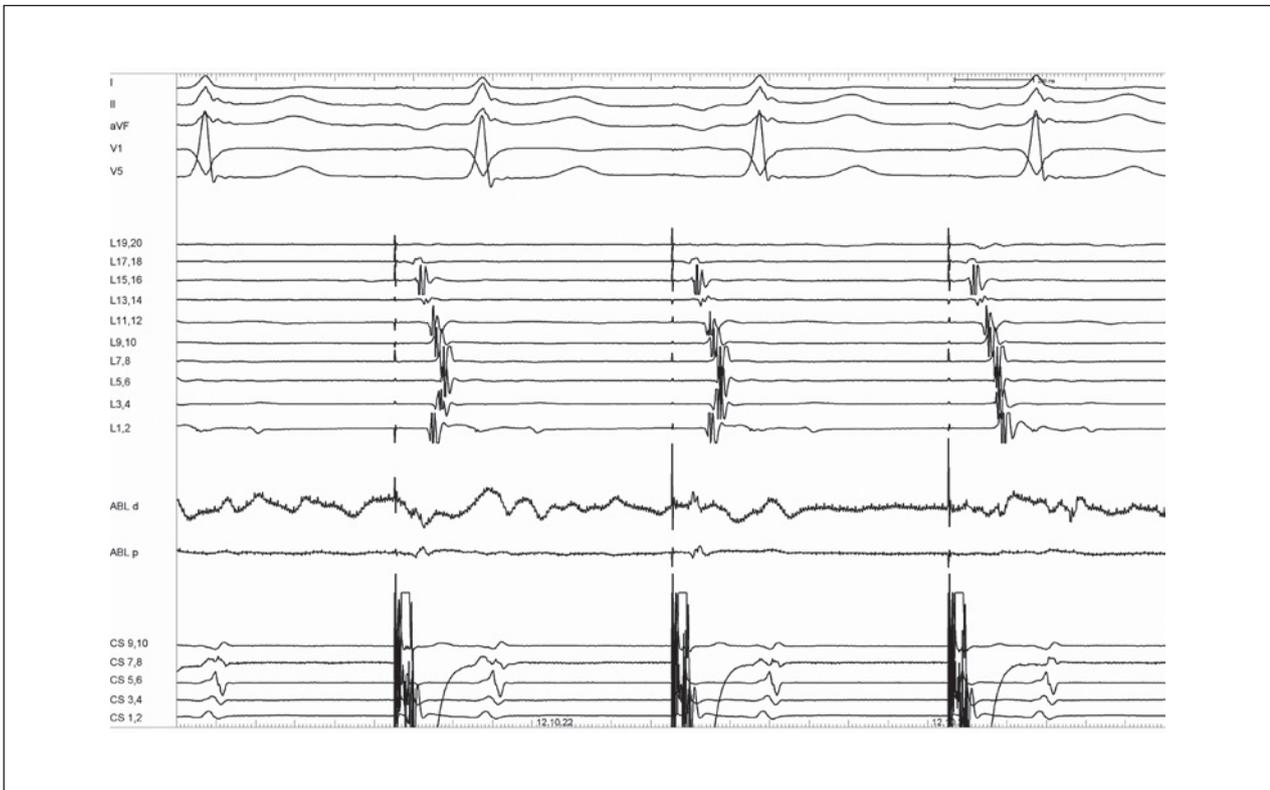


Figure 3. Abrupt change with complete reversal of depolarization sequence on the multipolar mapping catheter during RF energy application.

80% of cases, and it often precedes the skeletal muscle one. It occurs as a degenerative process with progressive fibrosis and fatty replacement not only limited to the specialized conduction system, but also to initially unaffected areas of the atrial myocardium (2). This anatomopathological substrate, causing the discontinuous and inhomogeneous propagation of sinus impulses (3, 4) and the prolongation of atrial conduction time (5, 6), may facilitate the onset and the perpetuation of atrial arrhythmias in these patients (7-15), as in other clinical conditions (16-24). Paroxysmal atrial arrhythmias such as atrial fibrillation, atrial flutter and atrial tachycardia show a prevalence up to 25% in DM1 patients and seem to increase mortality in this population (25).

Considering the arrhythmic risk and its consequences, an early identification of patients with high arrhythmic risk is of pivotal importance for the optimization of the clinical follow-up and medical therapy. For its auto-trigger capability that automatically detects and records predefined and programmable rhythm disturbances such as pauses, bradycardia, or supraventricular/ventricular tachy-arrhythmias, ELR represents a useful tool for early diagnosis of arrhythmic disorders in patients with recurrent palpitations, not diagnosed by conventional 24-h ECG Holter monitoring (26).

The early detecting of supraventricular arrhythmias in DM1 patients consents in turn to earlier evaluate the opportunity of interventional or pharmacologic approach. Furthermore, considering the high prevalence of supraventricular arrhythmias in these patients and the low diagnostic yield of conventional 24-hour Holter monitoring, the early use of ELR in recurrent palpitations might be a feasible and useful strategy, to detect patients with high risk of arrhythmias.

In the case report here described, the use of ELR allowed us to early detect and treat a symptomatic atrial flutter in a young patient with DM1 and to confirm during the follow-up the successful of the radiofrequency ablation procedure.

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