

## Early onset “electrical” heart failure in myotonic dystrophy type 1 patient: the role of ICD biventricular pacing

*Miyotonik distrofi tip 1 hastada erken başlayan “elektriksel” kalp yetersizliği: ICD biventriküler pacing’in rolü*

### Introduction

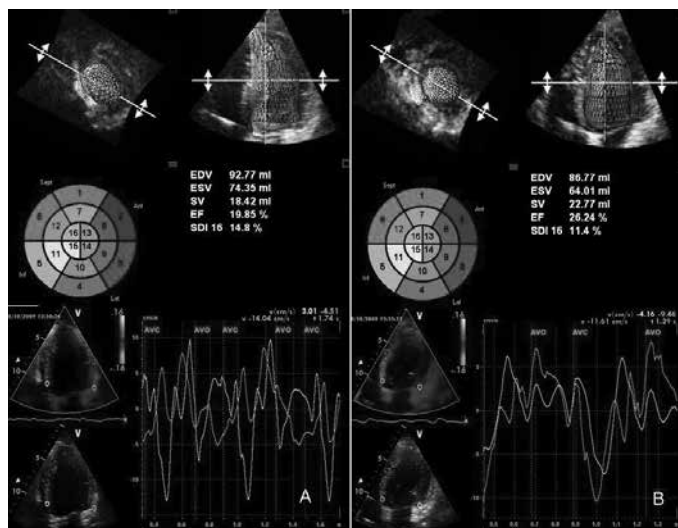
Myotonic dystrophy type 1 (MD1) has been identified as an autosomal dominant disorder associated with the presence of an abnormal expression of a cytosine-thymine-guanine trinucleotide repeat on chromosome 19q13.3. The classical symptoms are myotonia, muscle atrophy, cataract and a characteristic facial appearance (1, 2). The cardiac involvement, noticed in about 80% of cases, predominantly affects the conduction system, while myocardial contractile function is less commonly impaired in MD1 patients. Heart failure (HF) often occurs late in the course of the disease as consequence of cardiac myopathy due to progressive scar replacement (3). Cardiac resynchronization therapy (CRT) is an innovative new therapy that can relieve HF symptoms by improving the coordination of the heart’s contractions.

### Case Report

A 24-year-old man with myotonic dystrophy type 1 was referred to our division for dyspnea and palpitations. On physical examination crackles at the basal fields of lungs were detected. Electrocardiogram (ECG) revealed sinus rhythm, normal axis, prolonged PR interval and complete left bundle-branch block (LBBB) with QRS duration of 160 ms. Transthoracic echocardiography showed dilated cardiomyopathy with evident left systolic dysfunction and overt intraventricular and interventricular dyssynchrony. In particular, left ventricular (LV) ejection fraction (EF) was 20%, as calculated by both the Simpson’s biplane method and by 3D echocardiography, and significant intraventricular mechanical dyssynchrony was documented by both tissue Doppler and by 3D echocardiography (Fig. 1). At admission, the patient was taking angiotensin converting enzyme inhibitor and coenzyme Q10. The electrophysiological study showed a baseline prolonged AH (147 ms) and HV interval (75 ms); the programmed ventricular stimulation, using up to triple extrastimuli, revealed a sustained right bundle ventricular tachycardia (VT) with right inferior axis and cycle length of 290 ms, treated by external DC shock. According to the characteristic findings of overt ventricular dyssynchrony, complete LBBB, inducible VT, a biventricular implantable cardioverter-defibrillator (ICD) was implanted. At one month follow up, we performed the echocardiographic optimization of the atrioventricular and interventricular intervals during cardiac resynchronization. At six months follow-up the patient experienced symptom relief; ECG revealed paced ventricular rhythm with narrow QRS complexes; Echocardiogram showed increased ejection fraction and LV stroke volume, while LV mechanical dyssynchrony was significantly reduced (Fig. 2, 3). Six months later ambulatory interrogation of device revealed one proper and effective ICD shock, occurring during an episode of ventricular tachycardia (Fig. 4).

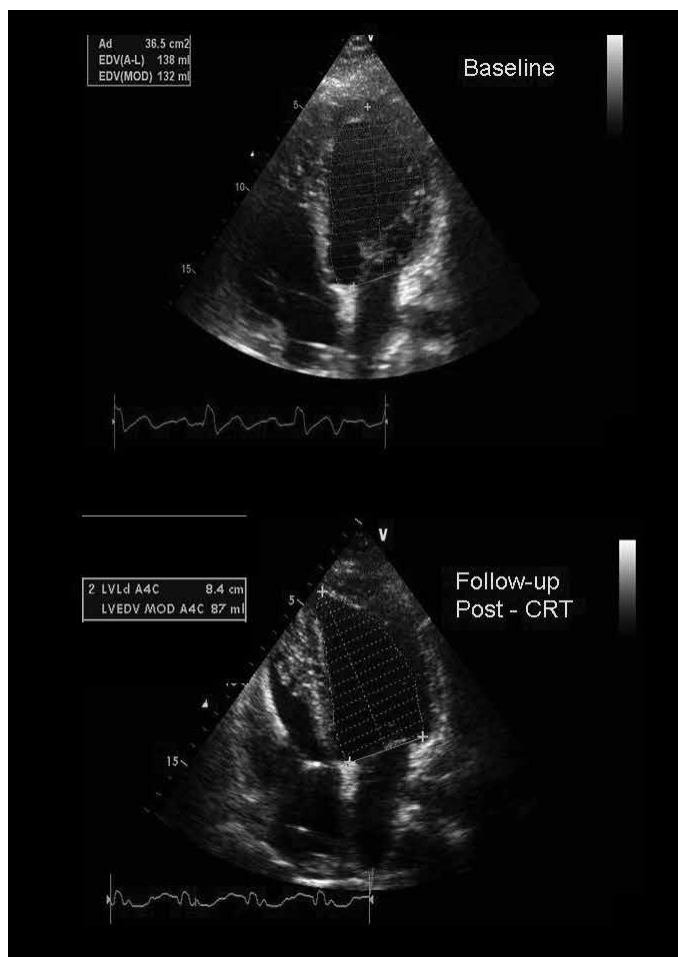
### Discussion

Heart failure is rare in myotonic dystrophy type 1 and often occurs late in the course of the disease. The clinical recognition of heart failure



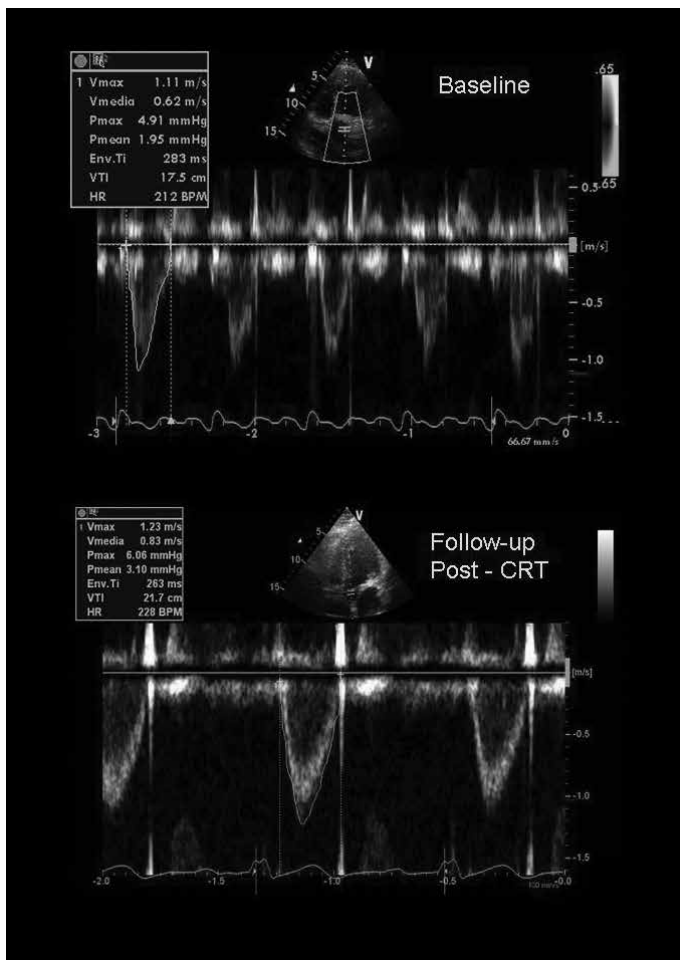
**Figure 1. Three-dimensional evaluation of LV ejection fraction and tissue Doppler evaluation of LV dyssynchrony without (Panel A) and with (Panel B) biventricular stimulation. Note the immediate increase in LV ejection fraction and the immediate reduction of systolic dyssynchrony index during active biventricular pacing**

LV - left ventricle



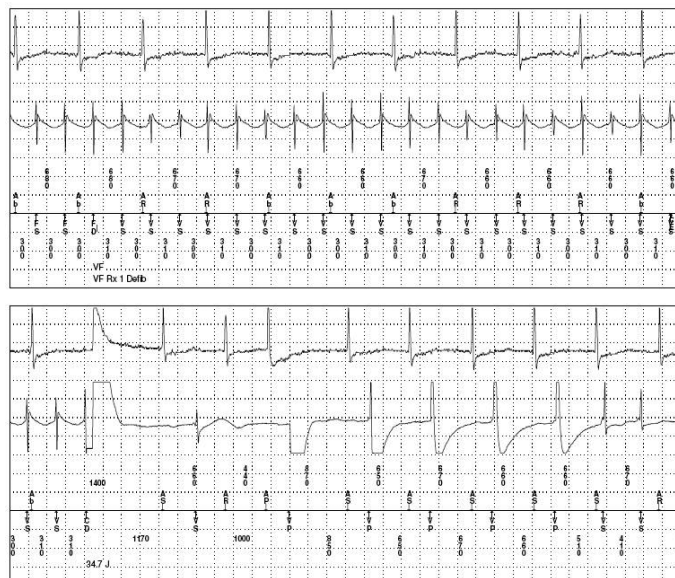
**Figure 2. LV end-diastolic volume at baseline (upper panel) and during 6-months follow-up after CRT. A significant reduction in LV end-diastolic volume was observed**

CRT - cardiac resynchronization therapy, LV - left ventricle



**Figure 3.** Time-integral of LV outflow tract systolic flow measured by standard Doppler at baseline (upper panel) and during 6-months follow up after CRT. Note the significant increase of this measurement during follow-up, as expression of improved LV stroke volume

CRT - cardiac resynchronization therapy, LV - left ventricle



**Figure 4.** Stored electrogram of ventricular tachycardia with a cycle length of 310 ms and appropriate intervention with shock of 34.7 J

in muscular disease is more difficult than in patients with a normal muscular function as fatigue is inherent to the muscular weakness and exercise tolerance is already impaired by the muscular disease itself. According to ESC 2007 Guidelines for Cardiac Pacing (4), there is neither a clear consensus about biventricular pacing nor the usage of implantable cardiac defibrillator for patients with overt myotonic heart disease. Basing on progressive deterioration of the left ventricular function, progression of atrioventricular conduction disturbances and on the occurrence of ventricular tachyarrhythmia, Said et al. (5) hypothesized a role for biventricular ICD in MD1 patients who necessitated permanent pacemaker implantation. Kılıç et al. (6) described the first case of efficacy cardiac resynchronization therapy in MD1 patient with heart failure and complete LBBB with ventricular asynchrony; the intra-cardiac defibrillator implantation was not performed because of no induced serious life threatening ventricular arrhythmia in the EPS.

In our patient, the early onset heart failure may be related to the electromechanical delay caused by both intra- and interventricular asynchrony. ICD-CRT can be a useful therapy in MD1 presenting with heart failure, cardiac dilatation with low EF, complete LBBB and inducible ventricular tachyarrhythmias. The spontaneous ventricular tachycardia, occurred in our patient at twelve months follow up, suggests that the improvement in ejection fraction may not reduce the arrhythmic risk in MD1 patients.

## Conclusion

In MD1 patients with early onset heart failure and complete LBBB, ICD-CRT implantation may improve "electrical" left ventricular dysfunction; induce reverse remodelling and relief in symptoms of heart failure. ICD-CRT implantation may be a life-saving treatment modality especially in high-risk MD1 patient with inducible malignant ventricular arrhythmias. The improvement in ejection fraction does not seem reduce the arrhythmic risk in MD1 patients. The early deterioration of the left ventricular function related to left bundle-branch dyssynchrony and the occurrence of ventricular tachyarrhythmia poses the question whether a biventricular ICD should be the first choice management in MD1 with early onset heart failure and complete LBBB.

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## Successful catheter ablation of symptomatic premature ventricular contractions originating from mitral annulus

*Mitral anülüsten kaynaklanan semptomatik ventriküler erken atımların başarılı kateter ablasyonu*

### Introduction

Premature ventricular contractions (PVC)/ventricular tachycardia's (VT) rarely originate from mitral annulus (1). In these cases, radiofrequency catheter ablation (RFCA) is an important treatment option.

Herein, we present a patient with PVC refractory to medical therapy, who was successfully treated with RFCA.

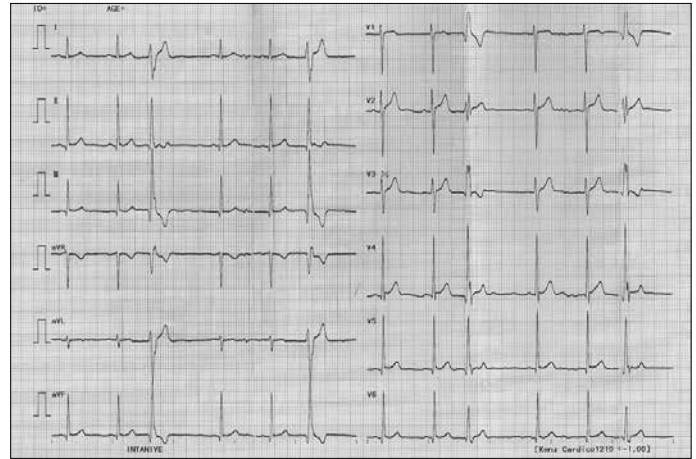
### Case Report

A 20-year-old male patient was admitted to our department with the complaint of palpitation. He had been having palpitations for 4 years. Medical treatment with calcium channel blocker and beta-blocker was unsuccessful. Physical examination of cardiovascular and other systems was normal. Resting electrocardiogram (ECG) showed PVCs with a right bundle branch block morphology and inferior axis (Fig.1). QRS notching was observed in the inferior leads of the PVCs. Exercise ECG and transthoracic echocardiography were within the normal range. The monomorphic PVCs (8000 beats/day) were detected in Holter ECG. Electrophysiological study was performed. Programmed ventricular stimulation did not induce ventricular tachycardia. Electrophysiological mapping was performed during PVCs. During PVC, the earliest ventricular activation was seen in the distal electrode of the coronary sinus. After placing the steerable 4-mm-tip ablation catheter (Mariner; Medtronic, Minneapolis, MN, USA) to the left ventricle with retrograde aortic approach, mapping of the aortic cusps and left ventricle outflow tract was performed. Early activation site was not detected at the aortic cusp and left ventricle outflow tract. With left ventricular mapping, earliest ventricular activity during PVCs was recorded in the anterolateral of the mitral annulus. In this site during the PVC, local ventricular

activation preceded the QRS onset by 28 ms (Fig. 2). Radiofrequency ablation applied to this site and PVCs disappeared (Fig. 3). PVCs were not observed during follow-up examinations at second month.

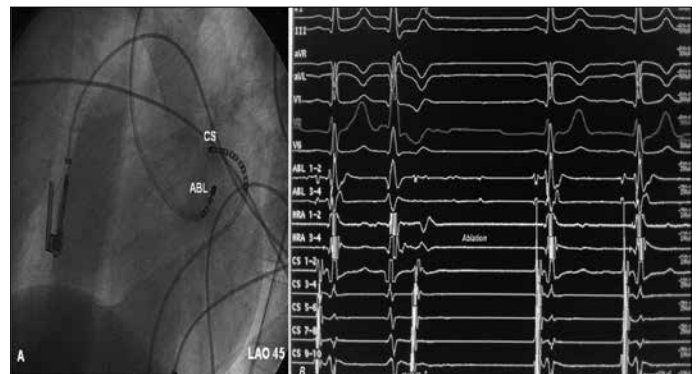
### Discussion

Idiopathic PVCs mostly originate from ventricular outflow tracts. RFCA is successfully performed to these sites. Premature ventricular



**Figure 1. Surface ECG of the patient with premature ventricular contractions**

ECG - electrocardiogram



**Figure 2. A) Left anterior oblique position, current catheter positions are shown while premature ventricular contractions disappeared. B) Surface and intracardiac electrograms recording at the site of successful ablation of the premature ventricular contractions**

ABL-ablation, CS- coronary sinus



**Figure 3. Premature ventricular contractions are not observed on the ECG after the procedure**

ECG - electrocardiogram