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Letter to Editor

Left Bundle Branch Optimized Implantable Cardioverter Defibrillator (LOT-ICD) Implantation in a Patient with Myotonic Dystrophy

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Abstract
Left bundle branch (LBB) optimized implantable cardioverter defibrillator (LOT-ICD) is a recently emerged alternative to the standard biventricular pacing - ICD (BiVp-ICD). The IS-1 connector pin of the ICD lead was closed with a protective cap and the LBB pacing (LBBP) lead was inserted into the ICD generator at the ventricular P/S hole. Herein, we presented a myotonic dystrophy patient who underwent LOT-ICD device implantation because of a high degree of atrioventricular block and heart failure with mid-range ejection fraction. A DDD-ICD (DF-1) was implanted in our patient which has been attached to the atrial lead (RA hole), the LBBP lead (ventricular IS-1 hole), and the ICD lead (ventricular DF-1 hole). The IS-1 pin of the ICD lead was closed accordingly. The LOT-ICD should be considered an option to standard BiVp-ICD associated with lower cost, lower procedure time, and no phrenic nerve capture.

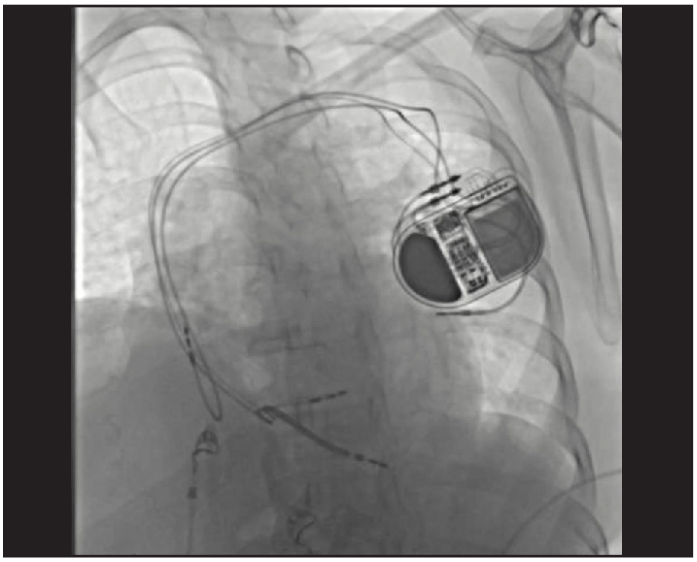
Keywords: Implantable cardioverter defibrillator, left bundle branch pacing

Letter to Editor,

Patients with myotonic dystrophy type 1 (MD1) have an increased risk of sudden cardiac death because of cardiac involvement and conduction abnormalities [1, 2]. Permanent pacemaker implantation is recommended in MD1 patients with advanced atrioventricular block (AVB) independent of the symptoms [3]. Cardiac resynchronization therapy (CRT) may be considered in DM1 patients who have permanent pacemaker dependency and left ventricular ejection fraction (LVEF) of <50% [4]. Although overt myocardial involvement in DM1 is infrequent, CRT implantation in DM1 patients was also reported previously [5-7].

Left bundle branch pacing (LBBP) is emerged as an alternative pacing method to biventricular CRT [8, 9]. The LBBP is achieved by inserting an electrode into the interventricular septum [10]. Several recent studies implicated that, LBBP is a more effective method and could be an alternative to biventricular CRT [11, 12]. Furthermore, in a recent research, another novel method was emerged and implemented as LBB optimized implantable cardioverter defibrillator (LOT-ICD) due to its high cost of biventricular CRT-D (IS-1 pin of the ventricular lead was closed with a protective cap and LBBP electrode was inserted into ventricular-P/S hole of ICD generator). This method had a lower cost and showed stable lead parameters. Herein, we presented a myotonic dystrophy patient with heart failure with mid-range ejection fraction (HFmrEF) and advanced atrioventricular block (AVB) in whom the LOT-ICD method was used.

A written and verbal informed consent was taken from the patient for submission of this paper. We did not use artificial intelligence (AI)– assisted technologies in the production of submitted work. A 51-year-old female patient with the diagnosis of MD1 and HFmrEF has been hospitalized for a rehabilitation program and consulted us for symptomatic bradycardia (dizziness and pre-syncope). She was under treatment of metoprolol 50 mg/day, Ramipril 2,5 mg/day, and spironolactone 25 mg/day for HFmrEF. She had no known history of coronary artery disease and her coronary angiography which was performed 7 years ago was normal. Cardiac magnetic resonance imaging (MRI) at the time of HFmrEF revealed no septal scar and LVEF of 41%. Her electrocardiography (ECG) showed sinus rhythm (40 bpm) and a trifascicular block (right bundle branch block, alternating left anterior/posterior hemi block, and first-degree AV block with PR interval of 400 ms) (Figure 1A). Furthermore, another ECG showed a second-degree Mobitz type 2 AVB and ventricular rate of 35 bpm (Figure 1B). Laboratory tests were unremarkable. Transthoracic echocardiography revealed an LVEF of 43% and moderate mitral and tricuspid regurgitation. Metoprolol treatment was stopped. As the patient was symptomatic, a temporary transvenous pacemaker was implanted to bridge the waiting period for metoprolol clearance (Figure 2). However, no improvement in the patient's rhythm was observed, and recurrent non-sustained ventricular tachycardia (NSVT) episodes were documented on telemetry recordings. As the patient would be pacemaker dependent and the LVEF was between 36-50%, we planned to implant a dual-chamber LOT-ICD device functioning as a CRT-D. Under local anesthesia and sedation, a left axillary vein puncture was performed. DF-1 ICD lead was placed into the right ventricle apex. Then, Solia S60 ventricular lead was inserted deep into the mid-interventricular septum with the support of Selectra 3D 55-39 (Biotronik, Berlin, Germany) delivery sheath and unipolar pacing parameters were compatible with the non-selective LBBP (V6 R wave peak time of 72 ms and V6-V1 inter-peak delay 40 ms, and a QRS transition during threshold test) (Supplementary Video 1). Pacing parameters of unipolar and bipolar pacing threshold/impedances were 0.6 and 0.7 V @ 0.4 ms / 615 and 773 ohms, R wave sense amplitude: 8.6 mV. Thereafter, atrial lead was placed into the right atrial appendage. After slitting the delivery sheath and fixation of the leads to the muscular tissue with the sleeves, the atrial lead pin was inserted into the RA hole, the defibrillation lead pin was inserted into the ventricular DF-1 hole, and the LBBP lead was inserted into the ventricular IS-1 port of the DDD-ICD (DF-1) pulse generator. IS-1 pin of the ventricular lead was closed with a protective cap and had left freely (Figure 3A & B). Post-procedural ECG showed a paced QRS duration of 120 ms with bipolar pacing of the device (Figure 4). No periprocedural complication was observed and the remaining hospitalization was uneventful. Beta-blocker treatment was re-initiated accordingly. The patient was asymptomatic and the pacemaker parameters were normal in the 1st and 3rd follow-up visits.



Supplementary Video 1. Contrast injection via delivery sheath showed the depth of the left bundle branch pacing lead in the septum. (Available at https://eurjther.com/index.php/home/article/view/2038/1546)

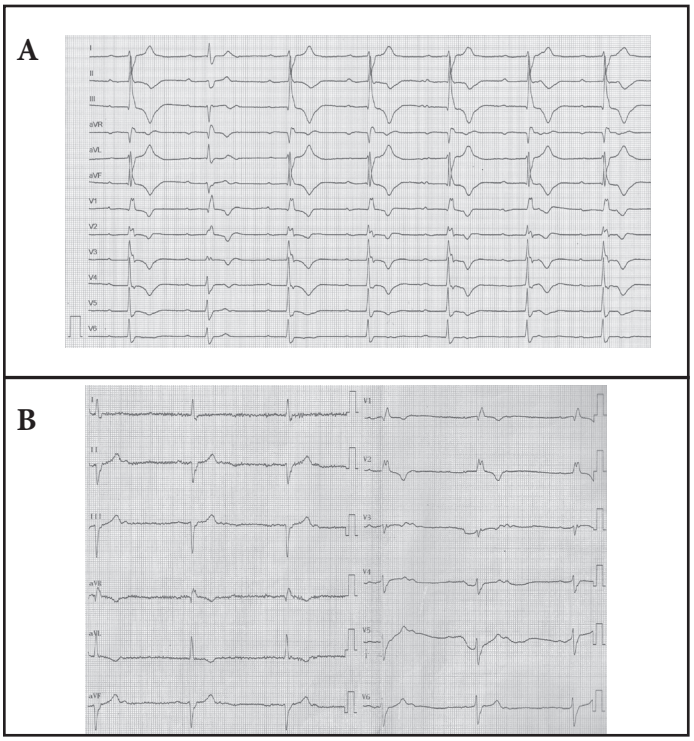


Figure 1. A. Electrocardiography showed sinus rhythm (40 bpm) and a trifascicular block (right bundle branch block, alternating left anterior/posterior hemi-block, and first-degree AV block with PR interval of 400 ms). **B.** Follow-up electrocardiography showed a second-degree Mobitz type 2 AV block and ventricular rate of 35 bpm.

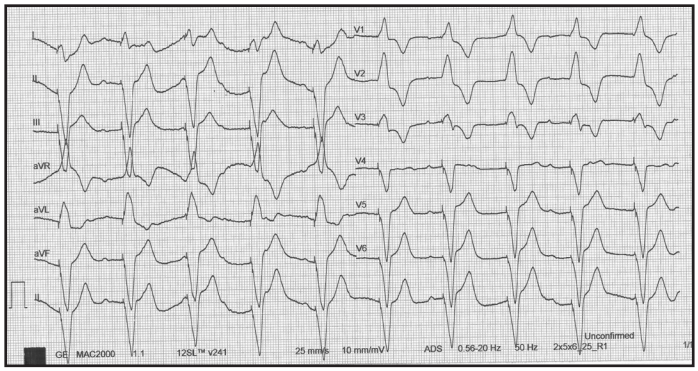


Figure 2. Electrocardiography showed a pacemaker rhythm with a paced QRS duration of 200 ms

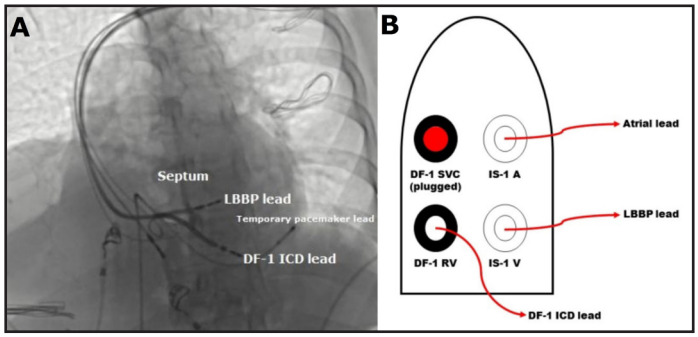


Figure 3. A. The fluoroscopy at the left anterior oblique view shows the localizations of the LOT-ICD device leads. **B.** The connections of the LOT-ICD device lead to the pulse generator.

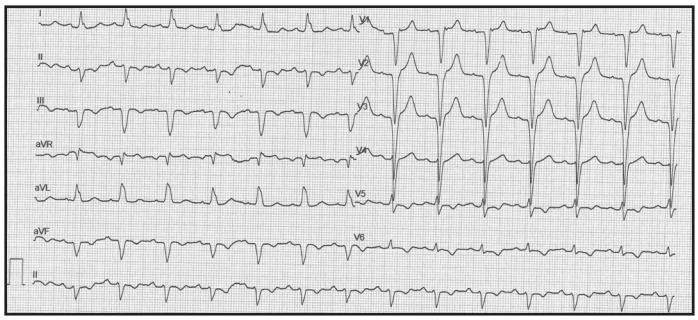


Figure 4. Post-procedural electrocardiography showed a paced QRS duration of 120 ms with bipolar pacing of the LOT-ICD device and terminal small r wave in V1 derivation.

In the current paper, we presented a female patient with MD1 and HFmrEF who consulted us for symptomatic bradycardia in whom a dual-chamber LOT-ICD device was implanted. MD1 is an autosomal dominant neuromuscular disorder characterized by multi-systemic involvement including the heart. It has been reported that sudden cardiac death can occur in MD1 patients because of myocardial and conduction system involvement [1, 2]. Overt myocardial involvement was rarely reported [5-7]. Although there were various data about conduction system abnormalities and permanent pacemaker implantation [1, 3], the data about biventricular CRT implantation in MD1 patients was scarce [5-7]. Furthermore, there is no clear consensus about biventricular CRT or ICD implantation in MD1. Groh et al. [1] reported the presence of atrial tachyarrhythmia, a PR interval > 240 milliseconds, aberrant QRS conduction, and advanced AV block as significant predictors of sudden death. In a recent study, Benhayon et al. [13] showed that the presence of AV conduction abnormalities in MD1 patients was also associated with an increased risk for ventricular tachyarrhythmia. Their study also revealed that one-third of the MD patients who received a primary prevention ICD experienced ventricular arrhythmia during a 22-month follow-up. Thus, it has been suggested that there is a role for CRT-ICD in DM1 patients who require permanent pacemaker implantation based on the progressive nature of the cardiac involvement [14]. Following the previous literature, our patient experienced multiple NSVT episodes with the risk factors for ventricular tachyarrhythmia and sudden cardiac death such as an advanced AV block, aberrant QRS conduction, and mid-range myocardial involvement. Thus, we implanted a pacemaker with a defibrillator function.

LBBP has emerged and is adopted very fast in clinical practice [10]. It has better LV synchronization and acute hemodynamic response compared to biventricular CRT [15]. Recent studies showed that CRT via LBBP was more effective than biventricular CRT [11, 12]. Interventricular septal scar and severe His-Purkinje conduction abnormality were prognostic factors for ineffective LBBP alone, and cMRI-guided LBBP might be a better option to be successful [16, 17]. LOT-CRT or HOT-CRT are better options in case of extensive septal scar or severe His-Purkinje conduction abnormality. LOT-ICD is a recently emerged cost-effective resynchronization technique. DDD-ICD generator ventricular P/S hole is occupied by LBBP lead, and the IS-1 pin of ICD lead is closed with a protective cap. It is shown that the LOT-ICD is associated with stable R wave sensing in arrhythmia detection, low cost, and low procedure time that cause lower radiation dose.

Furthermore, the LOT-ICD resulted in significant improvement of LVEF, LV end-diastolic diameter, and QRS durations of the patients [18]. It also overcomes the limitations of biventricular CRT implantation such as coronary sinus lead dislodgement, absence of appropriate coronary sinus branch, non-response to biventricular CRT, and phrenic nerve capture [19]. The main reason for using a LOT-ICD device rather than a biventricular CRT device is its lower cost of the pulse generator. Ponnusamy et al. [18] reported a 30% reduction in the cost of the therapy without any obstacle in clinical response with the LOT-ICD device. This novel cost-effective resynchronization method can be preferred in developing or under-developed countries with limited health sources and no reimbursement for device therapies [20]. Our MD1 patient had a symptomatic advanced AV block, documented NSVT episodes, and HFmrEF (LVEF 43%). It is recommended to perform a cardiac physiological pacing (LBBP or His bundle pacing) or biventricular CRT in such a patient who requires substantial ventricular pacing, NSVT episodes, and LVEF of 36-50%. The presence of myocardial involvement and an advanced AV block were thought as significant risk factors for sudden death in our patient. Thus, we prefer to perform LOT-ICD functioning as a CRT-D.

In conclusion, LOT-ICD is a promising alternative to biventricular CRT-D that has lower cost and mostly better pacing and procedure-related properties. Future large-scale randomized studies comparing LOT-ICD with biventricular BiVp-ICD are needed.

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