A patient with Duchenne muscular dystrophy and complete AV block undergoing a successful left bundle area pacing implantation

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SOUHRN

Stimulace septa jako nová technika kardiostimulace se doporučuje zvláště u pacientů se srdečním selháním a s potřebou stimulace ≥ 40 % doby za 24 hodin. Proveditelnost a účinnost stimulace oblasti levého Tawarova raménka u pacientů s Duchenneovou muskulární dystrofií nejsou známy. Naše kazuistika je prvním popisem pacienta s Duchenneovou muskulární dystrofií, který ukazuje použitelnost stimulace převodního systému.

ABSTRACT

The use of septal pacing, a novel pacemaker technique, is particularly recommended for patients with heart failure and a pacing requirement of ≥40% within a 24-hour period. The feasibility and effectiveness of left bundle area pacing remain unknown in Duchenne muscular dystrophy patients. Our case represents the first instance in Duchenne muscular dystrophy patients demonstrating the applicability of conduction system pacing.

Introduction

Duchenne muscular dystrophy (DMD) is a progressive skeletal and cardiac muscle disease. The major cause of death in individuals with DMD is of cardiac origin, primarily due to cardiomyopathy and arrhythmia. Cardiac arrhythmias encompass sinus tachycardia, ventricular tachycardia, sudden death, bundle block, and complete AV block.[1]

A pacemaker is a proposed therapy for patients with complete AV block. Additionally, an intracardiac defibrillator (ICD) should be employed in patients with an ejection fraction ≤35% to prevent sudden cardiac death. However, right apical ventricular pacing might induce or exacerbate heart failure in individuals with complete AV block. The use of septal pacing, a novel pacemaker technique, is particularly recommended for patients with heart failure and a pacing requirement of ≥40% within a 24-hour period.[1,2]

However, septal pacing has not yet been conducted in patients with DMD. In our study, we successfully implemented left bundle area pacing and an ICD in a patient with DMD, marking a pioneering achievement.

Case presentation

A 29-year-old male patient with DMD underwent a routine cardiac assessment. Electrocardiography revealed complete AV block with a heart rate of 57 bpm and a QRS duration of 90 mms **(Fig. 1A)**. Left ventricular ejection fraction was measured at 30% using the Simpson method, while right ventricular ejection fraction was within the normal range, and the interventricular septum thickness was 10 mm.[3] His life expectancy was projected to exceed 1 year.

We opted to conduct cardiac resynchronization therapy using the left bundle area pacing method. Initially, after puncturing three brachial veins, a 6935-shock lead (Medtronic Inc., USA) was positioned in the right ventricu-

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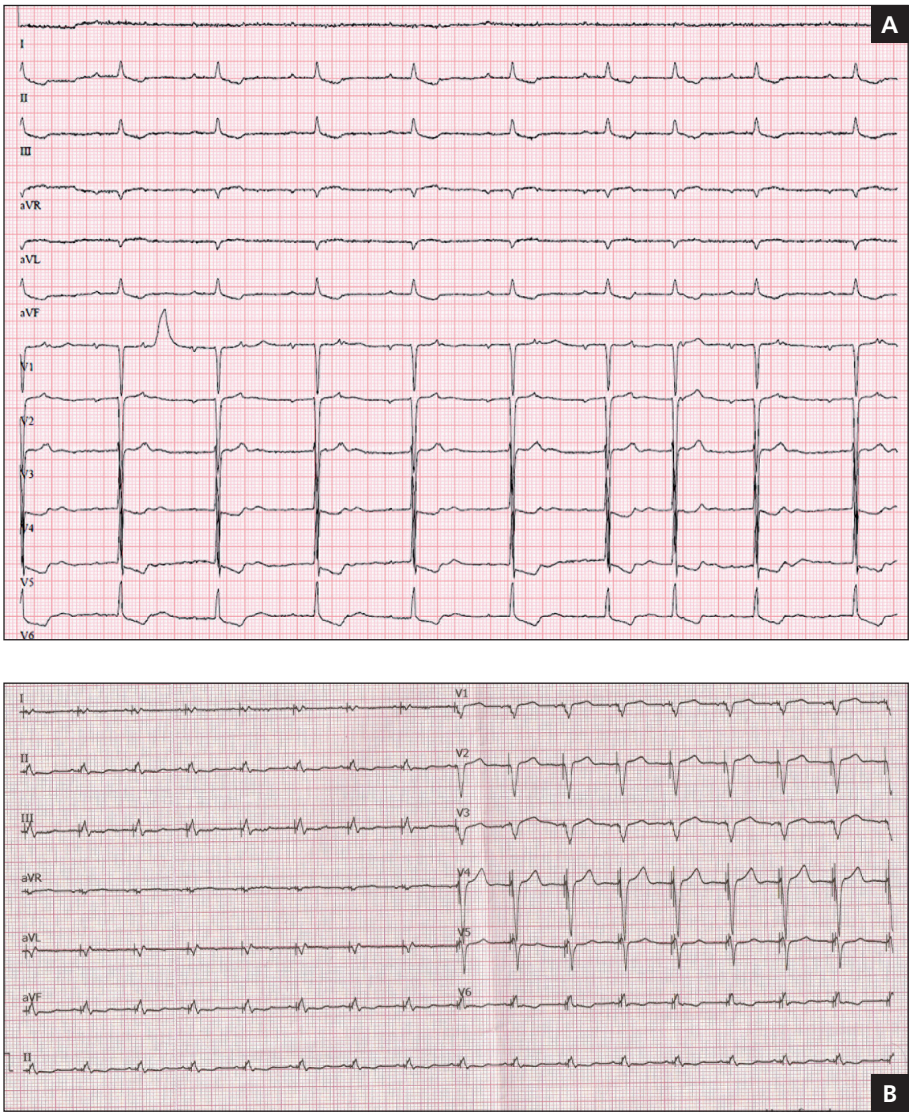

Fig. 1 – (A) Bazal electrocardiogram, (B) pacemaker electrocardiogram.

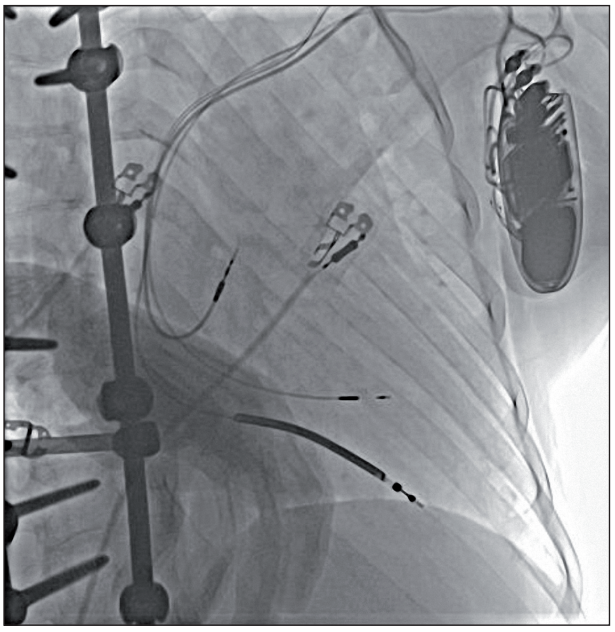

Fig. 2 – Fluoroscopy image of left bundle area pacing.

lar apex. Following that, a Medtronic 3830 lead (Medtronic Inc., USA) and a C315 his bundle sheath (Medtronic Inc., USA) were advanced toward the right ventricle. The 3830 lead (Medtronic Inc., USA), supported by the C315 sheet, was inserted into the upper mid-septum area of the left ventricle region, and visualized through fluoroscopic RAO 30-degree view. Contrast dye was injected through the sheet in the LAO 30-degree fluoroscopic view to assess the depth of the lead. Both the sensing and capture thresholds exhibited satisfactory levels. The atrial lead was inserted into the right atrial appendix. Following this, the sheath was skilfully extracted through a cutting procedure. Subsequently, the three leads were securely connected to a biventricular pacemaker generator. The left area pacing lead (3830) was linked to the biventricular pacemaker generator (DTMC2D4, Medtronic Inc., USA) in lieu of the coronary sinus lead. Postoperative electrocardiography indicated a pacemaker rhythm with a QRS duration of 120 ms (**Fig. 1B**). The positioning of lead 3830 in the upper mid-septum was confirmed through echocardiography and fluoroscopy (**Fig. 2**). The procedure duration was 58 minutes, with a fluoroscopy time of 8 minutes.

Discussion

Right ventricular pacing can potentially lead to heart failure, especially when there is a high pacing burden in patients with a normal ejection fraction. Additionally, patients who already have heart failure may experience worsened symptoms due to right ventricular pacing. In the context of the Multicenter Automatic Defibrillator Implantation Trial (MADIT) II study, a right ventricular pacing burden exceeding 50% was associated with an almost two-fold increase in the likelihood of developing or exacerbating heart failure symptoms.[4]

For patients with a reduced ejection fraction (ejection fraction <40%), regardless of their New York Heart Association (NYHA) functional classification, who require ventricular pacing due to high-degree AV block, it is advised to opt for cardiac resynchronization therapy over right ventricular pacing. This choice aims to minimize morbidity associated with the condition.[5] Given our patient's ejection fraction of 30%, we initially attempted to implant biventricular pacing through the coronary sinus. However, we were unable to identify a reliable vein. Echocardiography indicated that the septum's thickness was suitable for left bundle area pacing. Therefore we opted for conduction system pacing.

Nevertheless, the conduction system might be affected in patients with DMD. Conduction system degeneration often occurs at a slower rate compared to muscle degeneration. Pathological studies have demonstrated fat infiltration, fibrosis, and Purkinje fibre degeneration in the AV node and sinoatrial node.[6] The feasibility and effectiveness of left bundle area pacing remain unknown in DMD patients. Our case represents the first instance in DMD patients demonstrating the applicability of conduction system pacing.

Conflict of interest
The authors declare that they have no competing interests.

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Ethical statement
The study complied with the Declaration of Helsinki and informed consent has been obtained from the participant.

Informed consent
The patient consent statement was taken.

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