Case Report

Cardiac Resynchronization Therapy in Becker Muscular Dystrophy

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A 44-year-old male patient with known Becker muscular dystrophy and concomitant non-ischemic dilated cardiomyopathy presented to our department because of worsening heart failure and presyncope. Upon admission, the patient was in New York Heart Association functional class III despite optimal pharmacological treatment; his ECG showed sinus rhythm with left bundle branch block and a wide QRS complex. Non-sustained ventricular tachycardia was recorded during 24-hour Holter monitoring. A complete three-dimensional echocardiographic study was performed and documented the dilatation and concomitant hypertrabeculation of the left ventricle (LV), with severely depressed systolic LV performance (ejection fraction 20%), as well as mechanical dyssynchrony—mainly in terms of intraventricular delay. A biventricular cardioverter–defibrillator (CRT-D) was implanted in this patient, with the LV lead in a lateral vein and the right ventricular defibrillating lead in the apical part of the interventricular septum. Echocardiography-guided device programming was performed in order to achieve the optimal atrio-, inter-, and intraventricular resynchronization. The patient's clinical condition was substantially improved within one month after the implantation.

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Becker muscular dystrophy (BMD) is a dystrophinopathy, due to mutations in the X-linked gene of the sarcolemmal protein dystrophin.[1,2] Cardiac involvement is common in patients older than 40 years,[3] while the role of cardiac resynchronization therapy in patients with advanced heart failure is largely unknown.[3]

We present the case of a 44-year-old male patient with known BMD and non-ischemic cardiomyopathy (NICM), who presented with advanced heart failure (New York Heart Association, NYHA functional class III), syncope and non-sustained ventricular tachycardia. Significant electrical and mechanical dyssynchrony were documented on the electrocardiogram and echocardiogram, respectively. Soon after biventricular cardioverter–defibrillator (CRT-D) implantation, there was a significant improvement in his clinical status and related echocardiographic parameters.

Case presentation

The 44-year-old male patient was first hospitalized nine years ago, because of heart failure symptoms. The diagnosis of NICM was based on the findings of the echocardiogram and coronary angiogram, indicating left ventricular (LV) dilation and moderate systolic dysfunction. Four years later, BMD was diagnosed by skeletal muscle biopsy. The patient was on optimal, maximal tolerated medical therapy, including b-blockers, angiotensin-converting enzyme antagonist, aldosterone antagonist, and loop diuretics, and had a stable functional status (NYHA class I-II). During the last eight months, he presented a

gradual worsening in his heart failure symptoms (NYHA class III), with fatigue and dyspnea on minimal exercise and at rest, despite up-titration of diuretics. In addition, he suffered a syncopal attack and was referred to our department for further evaluation and treatment.

Upon admission, his electrocardiogram (ECG) showed sinus rhythm with left bundle branch block and a wide QRS complex (130 ms; Figure 1A). Episodes of non-sustained ventricular tachycardia were recorded during 24-hour Holter monitoring.

A complete three-dimensional echocardiographic study was then performed using a Vivid 7 ultrasound system (GE Medical Systems, Horten, Norway). Three cardiac cycles were stored in cineloop format for off-line analysis using dedicated software (Echopac, GE Medical Systems, Horten, Norway). The findings included significant LV dilatation with concomitant hypertrabeculation – resembling non-compaction (Figure 2) – and severely depressed LV systolic function (LV ejection fraction, LVEF approximately 20%), as well as moderately severe mitral and tricuspid valve regurgitation. Pulsed tissue Doppler was used to determine the interventricular mechanical delay (IVMD) by measuring the time from QRS onset to the peak myocardial systolic velocities of the right ventricular (RV) free wall (tricuspid annulus) versus the same time for the LV lateral mitral annulus (apical 4-chamber view). An IVMD of 90 ms was observed. Off-line color tissue Doppler-derived tissue velocity imaging (TVI) was used to assess intraventricular dyssynchrony in the longitudinal plane. At least 3 cardiac cycles were recorded during held respiration. Before performing measurements, aortic valve opening and closure were marked by means of a previously recorded pulsed-wave Doppler recording of the LV outflow tract, in order to avoid confusion between systolic (normal) and post-systolic (abnormal) contraction. Intraventricular dyssynchrony was detected between the LV lateral wall and the interventricular septum (90 ms), and the anterior–posterior wall (80 ms).

The patient underwent a CRT-D implantation (Medtronic Protecta, MN, USA). The active fixation right atrial and ventricular leads were positioned in the right appendage and the apical region of the interventricular septum, respectively. The LV lead was inserted, after coronary venography, into a lateral-posterolateral vein, with optimal values in terms of sensitivity, impedance and stimulating threshold. The CRT-D implantation was completed uneventfully.

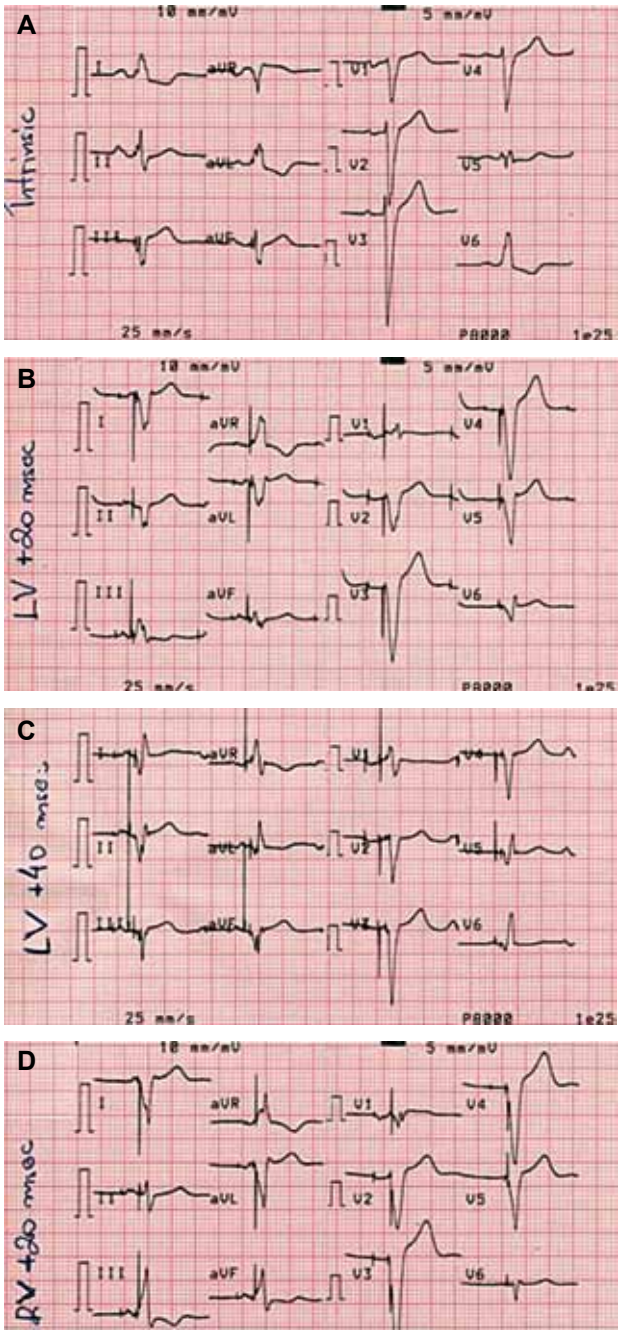

Figure 1. Twelve-lead ECG during intrinsic rhythm (A), biventricular pacing with the left ventricular lead preceding the right ventricular lead by 20 ms (B), by 40 ms (C), and with the right ventricular lead preceding by 20 ms (D).

The day after implantation, an echo-guided programming of the device was performed in order to achieve the best atrio-, intra- and inter- ventricular resynchronization. Optimization of the atrioventricular (AV) delay was guided by a Doppler recording of transmitral inflow (mitral inflow method), which requires recording the mitral inflow pattern at a sweep

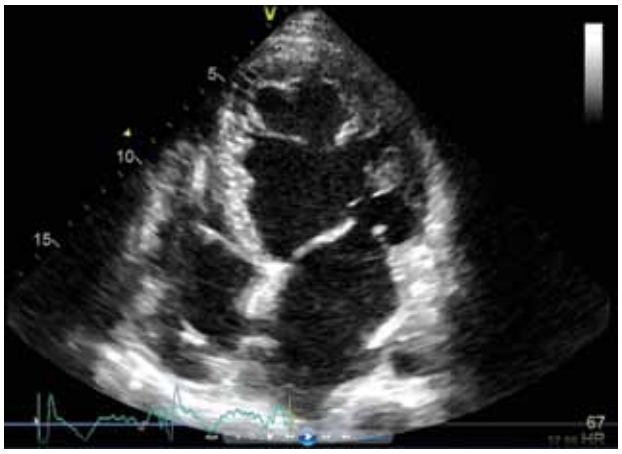

Figure 2. Significant left ventricular dilatation with concomitant hypertrabeculation resembling noncompaction

speed of 100 mm/s, with the sample volume placed at the tips of the mitral leaflets; mitral valve closure must be clearly defined at the time of the ECG R wave. The final pacing programming was as follows: DDDR mode, biventricular pacing, with LV offset (LV→RV) 40 ms, 60-130 bpm, sensed AV delay 100 ms, paced AV delay: 130 ms. The ECG in various pacing modes is presented in Figure 1 and shows a markedly changed depolarization pattern compared to the patient's intrinsic rhythm.

One month after implantation, the patient reported significant improvement in his clinical status, and remained asymptomatic, with a concomitant decrease in diuretic dose. Amelioration of the echocardiographic parameters was also documented, with an increase in LVEF (approximately 30%) as well as a decrease in atrioventricular valve regurgitation. No arrhythmic episodes have been recorded by the device. After 5 months of follow up the patient remains in good condition and his clinical course is uneventful.

Discussion

Becker and Duchenne muscular dystrophies, together with X-linked dilative cardiomyopathy, constitute the family of dystrophinopathies.[1,2] Mutations in the dystrophin gene on chromosome Xp21.1 cause abnormality or absence of this sarcolemmal protein, which participates in cytoskeleton architecture.[1,2] Pathologic findings consist of normal myocardium replacement by fibrofatty tissue. Cardiac involvement is rather frequent in BMD, especially in patients older than 40 years (approximately 75% of these patients).[3] The cardiac disease phenotype may vary from myocardial hypertrophy with preserved systolic function to extensive LV dilatation with wall motion abnormalities and significantly diminished cardiac function, depending mainly on the stage of the disease diagnosis.[4] There are also a few reports of left ventricular hypertrabeculation, as in our case, similar to noncompaction cardiomyopathy, based on echocardiographic and cardiac magnetic resonance imaging.[5,6]

We presented a patient with BMD and advanced heart failure, due to NICM with noncompaction morphology of the LV, and associated electrical, as well as mechanical dyssynchrony. The patient became nearly asymptomatic (NYHA class I) shortly after implantation, while the improvement in LV function was also documented by 3D-echocardiography. To our knowledge, there have been very few published cases of CRT therapy in BMD,[7] and for this reason specific guidelines on device therapy are not available for this type of disease. Despite the paucity of the existing evidence, it would be plausible to suggest the inclusion of Becker muscular dystrophy and associated cardiomyopathies in the broad spectrum of diverse etiologies of NICM that may respond favorably to resynchronization therapy. Our case strongly supports the expansion of the CRT indication in BMD patients.

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