Successful cardiac resynchronisation therapy in Duchenne muscular dystrophy: A 5-year follow-up

Resynchronisation myocardique dans la myopathie de Duchenne : un suivi de 5 ans

Case report

A 34-year-old man with genetic proven Duchenne muscular dystrophy (DMD), on wheelchair, was admitted to hospital because of asthenia, leg oedema and ascites. His past medical history was pertinent for scoliosis, tracheotomy for home ventilation because of restrictive respiratory deficiency, chronic heart failure (known for 12 years) and implantation of dual chamber pacemaker because of complete atrio-ventricular block (1 year before). He took daily angiotensin-converting enzyme inhibitor (perindopril 2 mg), beta-blocker (bisoprolol 1.25 mg) and loop diuretic (furosemid 80 mg). On admission, the patient was apyretic with blood pressure 95/55 mmHg and heart rate at 105 beats/min. Oxygen saturation was 98% with home mechanical ventilation. The electrocardiogram showed sinus rhythm with prolonged PR, complete left bundle branch block with unpaced prolonged QRS (130 ms) (figure 1). Laboratory findings were without any particularities. Chest X rays showed cardiomegaly with discrete pleural effusions. Doppler-echocardiography found moderate left ventricle (LV) dilation (LV end diastolic diameter at 59 mm), a decrease of the LV ejection fraction (LVEF at 30%) and a significant arterial pulmonary pressure (57 mmHg). The assessment of cardiac asynchrony with Doppler-echocardiography disclosed intra-ventricular and inter-ventricular asynchrony (table I). Upgrade from a dual-chamber to a biventricular pacemaker was performed and 1 month after, we noted stabilization of LV systolic function (LVEF at 30%), regression of inter-ventricular asynchrony and intra-ventricular asynchrony with a decrease of systolic pulmonary artery pressure (40 mmHg). After 5 years of follow-up, LV systolic function increased with LVEF at 45% and we noted a decrease of LV end diastolic diameter (41 mm).

Discussion

We report a rare case of successful cardiac resynchronisation therapy (CRT) in a patient with DMD who suffered from severe

Figure 1

EKG at admission (paper speed 25 mm/s): note the sinus rhythm with prolonged PR and wide QRS
heart failure. Hor et al. [1] reported a significant prevalence of mechanical ventricular dyssynchrony (17%) in DMD boys with LVEF < 55% but claimed that CRT is unlikely to improve heart failure in DMD. DMD is an inherited myogenic disorder due to mutations in the dystrophin gene on chromosome Xp21.1. It occurs in 1/3500 live-born males [2]. DMD has the onset of its manifestations at about the age of 5 years and is characterized by progressive loss of strength of muscles of pelvic and shoulder girdles. By the age of 12, most patients are confined to wheelchair. Cardiomyopathy is present in about 90% of patients [2]. Prognosis is affected by heart and respiratory failure. Cardiac involvement is segmentary initially (postero-basal segment of the left ventricular wall) and progressively affects all the heart. With heart failure, cardiac medical management relies on angiotensin-converting enzyme (ACE) inhibitor, beta-blocker and aldosterone antagonist. Conduction system disease (complete atrio-ventricular block) has been reported in Duchenne muscular dystrophy [3] and needs pacemaker implantation. However, right ventricular apical pacing in patients with cardiomyopathy can deteriorate progressively LV function because of dysynchrony [4]. CRT is an adjuvant treatment for patients with symptomatic (NYHA III or IV) heart failure (LVEF 35%) and QRS > 120 ms. It has been shown acute and sustained hemodynamic improvement, reversal of LV-remodelling and reduction of symptoms of heart failure in patients with heart failure after CRT. Cardiac asynchrony can be assessed using electrocardiogram (QRS interval) and echocardiography Doppler (pulsed Doppler and tissue Doppler imaging). In DMD patients, assessing cardiac symptoms (NYHA stratification) is challenging because of skeletal muscle failure and wheelchair state. Moreover, an increase of the posology of cardiac drugs is not well tolerated in patients with DMD because of arterial hypotension that limits a pharmacological approach. Normally, CRT is indicated in patients with heart failure who remain symptomatic despite optimal medical therapy. Also, CRT is usually considered not relevant in patients with low contractile reserve. Ionotropic contractile reserve is found to be a strong predictor of response to CRT [5]. This case report illustrates the possibility and the efficiency of CRT implantation in this group of patients that suffer from severe heart failure and ventricular dyssynchrony.

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References

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Table I

<table>
<thead>
<tr>
<th>Echocardiography finding parameters before and after CRT</th>
<th>Before CRT</th>
<th>1 month after CRT</th>
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</thead>
<tbody>
<tr>
<td>At admission LVEDD</td>
<td>58 mm</td>
<td>56 mm</td>
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<tr>
<td>LVEF</td>
<td>30%</td>
<td>30%</td>
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<tr>
<td>Inter-ventricular asynchrony delay</td>
<td>56 ms</td>
<td>40 ms</td>
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<tr>
<td>Intra-ventricular asynchrony delay using TDI (septum-lateral)</td>
<td>88 ms</td>
<td>8 ms</td>
</tr>
<tr>
<td>Intra-ventricular asynchrony delay using TDI (anterior-inferior)</td>
<td>92 ms</td>
<td>4 ms</td>
</tr>
</tbody>
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CRT: cardiac resynchronisation therapy; LVEDD: left ventricle end diastolic diameter; LVEF: left ventricle ejection fraction; TDI: Tissue Doppler Imaging.