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ORIGINAL RESEARCH

Cardiac asynchrony in Duchenne muscular dystrophy

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Abstract Duchenne muscular dystrophy (DMD) is an inherited myogenic disorder due to mutations in the dystrophin gene on chromosome Xp21.1. Heart failure is a classical complication in this disease. Little data are available about systolic dyssynchrony in DMD. We sought to assess the prevalence of left ventricular dysfunction and systolic asynchrony in DMD patients using echocardiographic parameters. We performed electrocardiography and echocardiography for adult's patients with DMD. For systolic dyssynchrony assessment, echocardiography-Doppler was performed and completed by tissular Doppler imaging. 48 DMD were included in our study. Age ranged from 20 to 37 years. QRS duration >120 ms was present in 10 patients/48 and 1 patient disclosed a QRS duration >150 ms. Left ventricular (LV) ejection fraction (EF) ranged from 10 to 62 % with a median of 43 %. Inter-ventricular asynchrony was found in 11.9 % of patients with EF < 35 % and in 2.6 % of patients with EF > 35 %. Intra-ventricular asynchrony was present in 6 % of patients with EF < 35 %. We found a high prevalence of LV dysfunction in DMD. Systolic ventricular asynchrony seems frequent particularly in patients with EF < 35 %.

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1 Introduction

Duchenne muscular dystrophy (DMD) is an inherited myogenic disorder due to mutations in the dystrophin gene on chromosome Xp21.1. Dystrophin is a component of the dystrophin–glycoprotein complex and links the actin to the extracellular matrix. DMD occurs in 1/3,500 live-born males [1]. DMD is characterized by progressive loss of strength of muscles of pelvic and shoulder girdles. By the age of 12 years, most patients are confined to wheelchair. Cardiomyopathy is present in 90 % of adult's patients. Systolic dyssynchrony is potentially a marker for disease severity and may act as a prognosticator. Little data are available about systolic dyssynchrony in DMD. We sought to assess the prevalence of left ventricular dysfunction and systolic asynchrony in DMD patients using echocardiographic-Doppler parameters.

2 Materials and methods

We performed electrocardiography (EKG) and echocardiography for adult's patients with DMD. Standard 12-lead EKG was performed at a paper speed of 25 mm/s and a scale of 10 mm/mV. QRS duration was measured manually from one of leads V1–V4.

For systolic dyssynchrony analysis, Doppler-echocardiography was performed and completed by tissular Doppler imaging (TDI). The following parameters were analyzed: LV end-diastolic diameter (LVDD), LV ejection fraction

(EF) assessed by Simpson's equation, ventricular electro-mechanical delays.

The intra-ventricular delay was examined by spectral pulsed TDI from the apical 4 chamber view. The time to the onset of ejection phase contraction in the systolic wave was measured from the basal segment of septal and lateral walls.

The inter-ventricular electromechanical delay was calculated as the difference between the aortic and pulmonary pre-ejection time intervals where aortic and pulmonary ejection flows were recorded from the five-chamber apical and para-sternal views, respectively.

The intra-ventricular asynchrony was calculated as the difference between the systolic electro-mechanical delay of the lateral free wall of the left ventricular and the systolic electro-mechanical delay of the septal annulus.

A systolic delay >50 ms was considered significant for intra-ventricular asynchrony.

An inter-ventricular electro-mechanical delay >40 ms was considered significant for inter-ventricular asynchrony.

3 Results

48 DMD were included in our study. Age value ranged from 20 to 37 years with a median of 27 years. All patients were disabled and were on wheelchair. 75 % of patients were ventilated with tracheotomy.

3.1 EKG and echocardiographic results

Mean QRS duration was 117 ± 29 ms in patients with EF < 35 % whereas mean QRS duration was 98 ± 17 ms in patients with EF > 35 %.

QRS duration >120 ms was present in 10 patients/48 and one patient disclosed a QRS duration >150 ms. 5 patients disclosed left bundle branch block (LBBB).

Mean indexed LVED diameter was 30 mm/m^2 with a median of 29. EF value ranged from 10 to 62 % with a median of 43. 11 patients/48 (23 %) had EF < 35 % whereas 37 patients/48 (77 %) had LVEF >35 %.

3.2 Echocardiographic asynchrony results

Mean pre-aortic ejection time interval was 138 ± 22 ms in patients with EF < 35 % and mean pre-pulmonary ejection time interval was 110 ± 23 ms in patients with EF < 35 %.

In patients with EF > 35 %, mean pre-aortic time was 98 ± 16 ms and mean pre-pulmonary ejection time was 91 ± 12 ms. Inter-ventricular asynchrony was found in 11.9 % of patients with EF < 35 % and in 2.6 % of patients with EF > 35 %. Left intra-ventricular asynchrony was present in 6 % of patients with EF < 35 % and in 2 % of patients with EF > 35 %.

Table 1 illustrates the repartition of patients with QRS duration >120 ms regarding left ventricular function (EF). Table 2 illustrates the repartition of patients with QRS duration >150 ms regarding EF.

The Tables 3 and 4 summarize data about inter-ventricular and intra-ventricular asynchrony in patients with DMD.

4 Discussion

DMD is an inherited muscular dystrophy due to mutations in the dystrophin gene on chromosome Xp21.1. Cardiomyopathy is present in about 90 % of the patients [1]. Despite medical management, prognosis is poor because of severe heart failure. Cardiac resynchronisation therapy (CRT) is an adjuvant treatment for patients with symptomatic, drug refractory heart failure and cardiac asynchrony. Cardiac asynchrony can be assessed using electrocardiogram (QRS interval) and echocardiography Doppler (TDI and 2D strain imaging). According the European Society of Cardiology guidelines [2], CRT is a class I and level A indication in patients with reduced ejection fraction (35 %) and ventricular dyssynchrony (QRS ≥ 120 ms) and who remain symptomatic (NYHA III–IV) despite optimal medical therapy. Moreover, CRT is a class I and level A indication in NYHA II patients with EF < 35 % and a QRS duration >150 ms despite optimal medical therapy.

Table 1 Number of patients with QRS duration >120 ms regarding LV ejection fraction (EF)

QRS duration	QRS duration <120 ms	QRS duration >120 ms	
EF ≤ 35 % (n)	6	5	11
EF > 35 % (n)	32	5	37
Total	38	10	48

n number of patients

QRS duration >120 ms

Table 2 Number of patients with QRS duration >150 ms regarding LV ejection fraction (EF)

QRS duration	QRS duration <150 ms	QRS duration >150 ms	
EF ≤ 35 % (n)	10	1	11
EF > 35 % (n)	37	0	37
Total			48

n number of patients

QRS duration >150 ms

Table 3 Doppler-echocardiographic asynchrony: data about inter-ventricular asynchrony using TDI

n (%)	Absent	Present	
≤35 %	4 (9.5)	5 (11.9)	9
>35 %	32 (76.2)	1 (2.6)	33
Total	36 (86)	6 (14)	42

n number of patients

Table 4 Doppler-echocardiographic asynchrony: data about intra-ventricular asynchrony using TDI

n (%)	Absent	Present	
≤35 %	8 (17)	3 (6)	11
>35 %	36 (75)	1 (2)	37
Total	44 (92)	4 (8)	48

n number of patients

In our study, QRS duration >120 ms was present in 10 patients/48 and 1 patient disclosed a QRS duration >150 ms. 5 patients disclosed LBBB. This group of patients are eligible for cardiac resynchronisation therapy.

With echocardiography Doppler, inter-ventricular asynchrony was found in 11.9 % of patients with EF < 35 % and in 2.6 % of patients with EF > 35 %. We found a left intra-ventricular asynchrony in 6 % of patients with EF < 35 % and in 2 % of patients with EF > 35 %. However, we assessed mainly longitudinal asynchrony in our study. Hor et al. [3] analysed mechanical dyssynchrony in boys with DMD using MRI and found circumferential dyssynchrony in 31.2 % of patients with abnormal EF.

5 Limitations

We use classical TDI for the analysis of cardiac asynchrony. This technology is limited because of angle dependence. 2D strain is more accurate for the assessment of LV dyssynchrony.

6 Conclusion

We found a high prevalence of LV dysfunction in DMD. Systolic ventricular asynchrony seems frequent particularly in patients with EF < 35 %. Cardiac resynchronization therapy has to be discussed in selected patients with muscular dystrophy.

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Conflict of interest None.

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