

POSTER PRESENTATION

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Left ventricular function by echocardiography correlates poorly with cardiac MRI measures in Duchenne muscular dystrophy

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Background

Duchenne muscular dystrophy (DMD) causes skeletal muscle weakness and cardiomyopathy (CM). Current recommendations are for annual left ventricular (LV) function assessment after age 10 years. Although echocardiographic image quality in DMD patients can be affected by scoliosis and adipose tissue, recent reviews recommend echocardiography as the standard imaging modality. We hypothesized that objective and subjective LV functional assessment by echocardiography in DMD is suboptimal compared to cardiac MRI (CMR).

Methods

Twelve DMD patients prospectively enrolled; echocardiography and CMR performed median of 0 days apart (max 22 days). Echocardiography was performed by sonographers with DMD imaging expertise. Cardiologist blinded to CMR results measured the following echocardiographic parameters: 1) M-mode fractional shortening (MMFS); 2) 2-dimensional FS (2DFS), 3) biplane LV ejection fraction (LVEF); 4) single plane LVEF; 5) 3-dimensional LVEF; 6) peak circumferential strain (ϵ_{cc}); 7) subjective LVEF. CMR measures included: 1) LVEF; 2) HARP ϵ_{cc} analysis of tagged images; 3) Subjective segmental function. Segmental assessments by echocardiography and CMR were performed using 17-segment model. Agreement between echocardiography and CMR assessed with intraclass correlation coefficient (ICC) and Spearman correlation; subjective LVEF evaluated with weighted kappa.

Results

Mean age was 15.8 years (range 10-27). Mean LVEF by CMR was $47.4 \pm 8.9\%$; 8 patients had CM defined as LVEF $< 55\%$ (Table 1). Subjective echocardiographic image quality rated good in 4/12 (33.3%), average 2/12 (16.7%), poor 3/12 (25%) and inadequate 3/12 (25%); none rated excellent. For echocardiography, only MMFS was measurable in all patients. Only moderate correlations were seen between MMFS and CMR LVEF ($r = 0.59$, $p = 0.042$) and echocardiographic ϵ_{cc} and CMR ϵ_{cc} (ICC = 0.52, $p = 0.045$). A strong correlation was seen between 2DFS and CMR LVEF ($r = 0.79$, $p = 0.033$) but 2DFS was only obtainable in 58% of patients. No significant correlations were found between other measures,

Table 1 Results of Objective Measures of LV Function

Measures of LV Function	Mean \pm SD	N
Echocardiographic Measures		
M-mode FS	24.5 ± 6.1	12
2-Dimensional FS	27.6 ± 3.9	7
Biplane LVEF	52.1 ± 8.1	4
4 chamber LVEF	44 ± 12.2	7
3-Dimensional LVEF	36.2 ± 13.3	5
ϵ_{cc}	-16.5 ± 4.4	8
CMR Measures		
LVEF	47.4 ± 8.9	12
ϵ_{cc}	-12.9 ± 3.5	12

LV (left ventricular) SD (standard deviation) FS (fractional shortening) LVEF (left ventricular ejection fraction) ϵ_{cc} (peak circumferential strain)

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Table 2 Comparison of Echocardiography and CMR measures of LV function

Echocardiographic Measures	Adequate Echocardiographic Image Quality (N = 12)	Correlation	P-value
CMR LVEF			
M-mode FS	12/12 (100%)	0.59 ¹	0.042
2-Dimensional FS	7/12 (58.3%)	0.79 ¹	0.033
Biplane LVEF	4/12 (33.3%)	-0.07 ²	0.531
4 chamber LVEF	7/12 (58.3%)	0.5 ²	0.09
3-Dimensional LVEF	5/12 (41.7%)	0.28 ²	0.168
Subjective global function	12/12 (100%)	0.08 ³	0.35
CMR ϵ_{cc}			
ϵ_{cc}	8/12 (66.6%)	0.52 ¹	0.045

LVEF (left ventricular ejection fraction) FS (fractional shortening) ϵ_{cc} (peak circumferential strain) ¹Spearman Correlation ²Intraclass Correlation Coefficient
³Weighted Kappa

including subjective LVEF (Table 2). Subjective segmental assessment was possible in 202 of 204 segments by CMR and only 137 of 204 segments by echocardiography. Of 69 segments not visualized by echocardiography, 39 had abnormal wall motion by CMR. Inferior and inferolateral walls at mid-ventricular level were most common sites of wall motion abnormalities.

Conclusions

Objective and subjective echocardiographic measures of LV function were not possible in many DMD patients and had limited correlation with CMR. Only 3 studies were rated inadequate, suggesting that, even in the face of “adequate” imaging, functional analysis by echocardiography had suboptimal correlation and unrecognized wall motion abnormalities. These discrepancies could adversely impact patient care. We recommend early consideration for CMR for annual, accurate assessment of DMD function.

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