



ORIGINAL ARTICLE

Importance of three-dimensional speckle tracking in the assessment of left atrial and ventricular dysfunction in patients with myotonic dystrophy type 1

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Received 25 June 2017; accepted 28 October 2017

KEYWORDS

Myotonic dystrophy;
Three-dimensional
echocardiography;
Three-dimensional
speckle tracking;
Strain

Abstract

Introduction and Aim: Myotonic dystrophy type 1 (DM1) is a multisystem disease in which cardiac involvement is common. The aim of this study was to identify early changes in left atrial (LA) mechanics and left ventricular (LV) systolic function in patients with myotonic dystrophy type 1 using three-dimensional (3D) speckle tracking echocardiography (3D-STE).

Methods: This observational study included 25 patients with DM1 and 25 healthy volunteers. We assessed LA and LV global strain parameters using 3D-STE.

Results: Patients with DM1 showed significantly lower longitudinal LA strain ($22.85\% \pm 5.06$ vs. $26.82\% \pm 5.15$; $p=0.008$ in univariate analysis and $p=0.026$ in multivariate analysis) and global LV longitudinal strain ($-13.55\% \pm 1.82$ vs. $-16.11\% \pm 1.33$; $p<0.001$ in univariate analysis and $p<0.001$ in multivariate analysis), which was not observed with LA area tracking ($p=0.412$) or LV global circumferential strain ($p=0.879$), global radial strain ($p=0.058$), area tracking ($p=0.092$) or twist ($p=0.992$).

Conclusion: LA and LV global longitudinal strain is significantly decreased in patients with DM1, which may be an early marker of subclinical dysfunction in these patients.

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<https://doi.org/10.1016/j.repc.2017.10.011>

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PALAVRAS-CHAVE

Distrofia miotônica;
Ecocardiografia 3D;
3D speckle tracking;
Strain

Importância da tecnologia 3D *speckle tracking* na avaliação da disfunção auricular e ventricular esquerda em doentes com distrofia miotônica do tipo 1

Resumo

Introdução e objetivos: A distrofia miotônica do tipo 1 é uma doença multissistémica com o envolvimento cardíaco a ser comum. O objetivo deste estudo foi identificar alterações precoce no *strain* da aurícula esquerda e função sistólica do ventrículo esquerdo em doentes com distrofia miotônica tipo 1, com o uso da tecnologia 3D *speckle tracking*.

Métodos: Estudo observacional, que incluiu 25 doentes com distrofia miotônica do tipo 1 e 25 voluntários saudáveis. Foram avaliados os parâmetros de deformação global da aurícula esquerda e do ventrículo com o uso de 3D *speckle tracking*.

Resultados: Os doentes com distrofia miotônica do tipo 1 apresentaram *strain* longitudinal da aurícula esquerda ($22,85\% \pm 5,06$ versus $26,82\% \pm 5,15$; $p=0,008$ na análise univariada e $p=0,026$ na análise multivariada) e *strain* global longitudinal do ventrículo esquerdo ($-13,55\% \pm 1,82$ versus $-16,11\% \pm 1,33$, $p<0,001$ na análise univariada e $p<0,001$ na análise multivariada) significativamente menores, o que não se verificou com a *area tracking* da aurícula esquerda ($p=0,412$) ou com o *strain* global circumferencial ($p=0,879$), *strain* global radial ($p=0,058$), *area tracking* ($p=0,092$) e *twist* ($p=0,992$) do ventrículo esquerdo.

Conclusão: O *strain* longitudinal global do ventrículo esquerdo e da aurícula esquerda encontrase significativamente diminuído em doentes com distrofia miotônica do tipo 1, pode ser um marcador precoce de disfunção subclínica nesses doentes.

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List of abbreviations

DM1	myotonic dystrophy type 1
LA	left atrial
LV	left ventricular
3D	three-dimensional
STE	speckle tracking echocardiography
2D	two-dimensional
LS	longitudinal strain
AT	area tracking
GLS	global longitudinal strain
GCS	global circumferential strain
GRS	global radial strain
ICC	intraclass correlation coefficient

ventricle in early diastole, and active contraction in late diastole.^{4,5}

In a comparison of volumetric real-time three-dimensional (3D) echocardiography and 3D speckle tracking echocardiography (3D-STE), the two methods were found to provide comparable and reproducible measurements, and to be interchangeable for quantification of LA dimensions and functional properties.⁶

Echocardiographic myocardial tracking systems using 3D-STE have provided new insights into ventricular function and atrial mechanics, with many potential clinical applications.^{7,8} 3D formats more closely represent reality than two-dimensional (2D) formats, not only in the assessment of LV function but also in the diagnosis of valvular or myocardial disease. Determination of LV strain by 2D speckle tracking echocardiography (2D-STE) enables identification of changes in LV systolic function in patients with DM1.^{9,10} However, the applicability of 3D-STE in these patients is not fully established and the available data are scarce.

The aim of this study was to identify early changes in LA mechanics and LV systolic function in patients with DM1 using 3D-STE.

Methods

Study population

This was a single-center observational study. After written informed consent was obtained, all 25 patients with DM1 followed in our center and 25 healthy volunteers (control group) were consecutively selected. All DM1 patients had genetic confirmation of the diagnosis and had no known

Introduction

Myotonic dystrophy type 1 (DM1) is a multisystem neuromuscular disease with autosomal dominant transmission that affects smooth and skeletal muscle.¹ About 30% of mortality in adults with DM1 is due to cardiac involvement.² Arrhythmias, conduction system disturbances and myocardial fibrosis are the most frequent changes.³ Detection of early markers may facilitate the identification of patients at risk of developing cardiac complications.

Left atrial (LA) function is a complex process that includes receiving pulmonary venous return during left ventricular (LV) systole, transfer of blood passively into the left

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