

How Echocardiographic Deformation Indices Can Distinguish Different Types of Left Ventricular Hypertrophy

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We present cases of athlete's heart, idiopathic HCM, and glycogen storage cardiomyopathy (PRKAG2).

The two non-athlete patients (Pts) underwent genetic studies and myocardial biopsies.

Echocardiography showed moderate to severe LVH in all cases.

Case 1, Figure 1-A, A-3 – Athlete, male, 26, intense exercise practice. Automated function imaging: 2D LV strain bullseye map showing normal Longitudinal Regional Myocardial Deformation (LRMD), despite LVH. GLS (global longitudinal strain) -20.4%.

Case 2, Figure 1-B, B-1, B-2, B-3 – Male, 26, tachycardia and palpitations with myosin essential light chain 3 mutation. LRMD is typically reduced where hypertrophy is more accentuated. GLS -14.0%.

Figure B-1 – Section of RV ventricular myocardium in HCM, demonstrating marked myocyte hypertrophy and disorganization (HE-stained).

Figure B-2 – Gomoririchrome stain (GS) showing intense fibrosis in extracellular matrix (blue) and cardiomyocyte architecture disarray.

Case 3, Figure 1-C, C-1, C-2, C-3 - Male, 22, palpitations and tachycardia. Genetic analysis found missense mutation, a heterozygous pathogenic variant for PRKAG2 c.905g > A p.(Arg302Gln). LRMD shows deformation levels in a striped pattern. GLS -10.5%.

Keywords

Speckle Tracking; PRKAG2 Mutation; Deformation Indices; Left Ventricular Hypertrophy, Cardiomegaly; Echocardiography/diagnosis.

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Figure C1 - HE-stained RV showing myofiber vacuolization with gross granular glycogen inclusions within vacuoles, without cardiomyocyte architecture disarray.

Figure C2-GS showing intense myofiber vacuolization (white) and extracellular matrix collagen fibers without fibrosis (blue).

STE (speckle tracking echocardiography) differentiates LVH and infiltrative disorders. We tried to instantaneously identify disease-related patterns.

To our knowledge, we present the first pattern in a PRKAG2 mutation Pt bullseye map, differentiated from other causes of LVH.¹ We recommend GLS polar map analysis to improve accuracy in echocardiographic examinations involving moderate LVH. STE can suggest the etiology, critically important to improve therapeutic strategies.

Author contributions

Conception and design of the research, acquisition of data and analysis and interpretation of the data: Pena JLB, Santos WC, Araújo SA, Dias GM, Sternick EB; writing of the manuscript: Pena JLB, Santos WC; critical revision of the manuscript for intellectual content: Araújo SA, Dias GM, Sternick EB.

Potential Conflict of Interest

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Study Association

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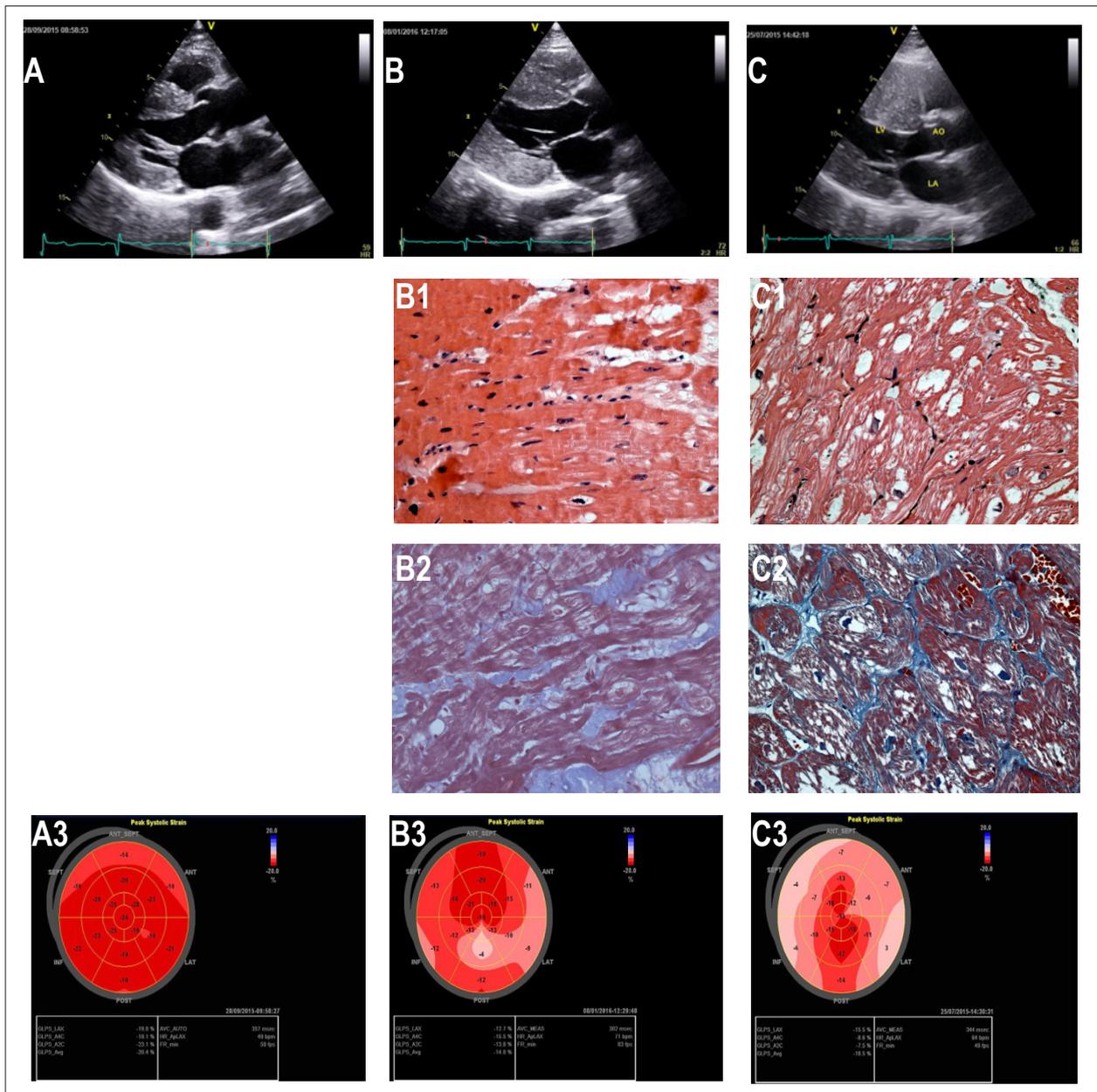


Figure 1 – Two-dimensional echocardiography (A, B, C), endomyocardial biopsies (B1, B2, C1, C2) and bullseye maps (A3, B3, C3).

Reference

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