

## CASE REPORT

# Left ventricular noncompaction in a Para athlete

## Miocárdio não compactado em paratleta

Ednei Costa Maia<sup>1</sup>, Filippo Aragão Savioli<sup>1</sup>, Sanna Roque Pinheiro<sup>1</sup>, Leandro Santini Echenique<sup>1,2</sup>, Japy Angelini Oliveira Filho<sup>1</sup>

<sup>1</sup> Universidade Federal de São Paulo, São Paulo, SP, Brazil.

<sup>2</sup> Hospital Israelita Albert Einstein, São Paulo, SP, Brazil.

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### ABSTRACT

The left ventricular noncompaction is a congenital cardiomyopathy characterized by the presence of abnormal trabeculations in the left ventricle. The present study describes the case of a 14-year-old female Para athlete, who plays goalball. She was asymptomatic, with history of congenital nystagmus and mild visual impairment, who presented nonspecific electrocardiographic abnormalities during pre-competition screening. Cardiac magnetic resonance imaging showed left ventricular non-compaction (non-compacted to compacted layer ratio equal to 2.5) and mild biventricular systolic dysfunction. Initially, the patient was excluded from sports participation and clinical follow-up was performed every three months. Patient remained asymptomatic during the one-year follow-up, with no history of unexplained syncope, marked impairment of systolic function or significant ventricular arrhythmias at the exercise stress test. Finally, she was released for competitive goalball participation and clinical follow-up was continued every 6 months. There is no consensus regarding the eligibility criteria for sports participation in cases of left ventricular non-compaction. Thus, it is prudent to individualize the decision regarding practice of sports, as well as to consider participation in competitive sports for asymptomatic individuals and with no disease repercussions.

**Keywords:** Isolated noncompaction of the ventricular myocardium; Cardiomyopathies; Congenital abnormalities; Exercise

### RESUMO

O miocárdio não compactado é uma cardiomiopatia congênita caracterizada pela presença de trabeculações anormais no ventrículo esquerdo. O presente estudo descreve o caso de uma paratleta de *goalball*, 14 anos, sexo feminino, assintomática, com história pessoal de nistagmo congênito e leve deficiência visual, que apresentou alterações eletrocardiográficas inespecíficas durante avaliação pré-participação. A ressonância magnética cardíaca evidenciou presença de não compactação miocárdica (relação entre camada não compactada/camada compactada igual a 2,5) e disfunção sistólica biventricular leve. Inicialmente, a paciente foi afastada da prática de esportes, e o seguimento clínico foi realizado a cada 3 meses. A paciente permaneceu assintomática durante o período de 1 ano de seguimento, sem história de síncope inexplicada, comprometimento significativo da função sistólica ou taquiarritmias ventriculares importantes ao teste de esforço. Por fim, ela foi liberada para prática competitiva de *goalball*, e o seguimento clínico foi mantido a cada 6 meses. Não há consenso quanto aos critérios de elegibilidade para a prática esportiva nos casos de miocárdio não-compactado. Assim, é prudente individualizar a decisão quanto a prática esportiva, bem como considerar a participação em esportes competitivos para indivíduos assintomáticos e sem repercussões da doença.

**Descritores:** Miocárdio ventricular não compactado isolado; Cardiomiopatias; Anormalidades congênitas; Exercício

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#### Corresponding author:

Ednei Costa Maia  
Avenida Jerônimo de Albuquerque, S/N, Calhau  
Zip code: 65074-220 – São Luís, MA, Brazil  
Phone: (55 98) 3268-2710  
E-mail: ednei.maia@hotmail.com

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## INTRODUCTION

Left ventricular noncompaction (LVNC) was first described by Grant, in 1926.<sup>(1)</sup> Since then, the disease has received different denominations in the literature: spongy heart, left ventricle (LV) hypertrabeculation, or isolated abnormal LV trabeculation.<sup>(2)</sup> During the normal embryologic development, the trabecular compaction process takes place between the 12<sup>th</sup> and 18<sup>th</sup> weeks of gestation. This process initially occurs on the base of the heart, and progressively evolves towards the apex. In LVNC cases, the myocardial compaction does not occur as expected due to still unknown reasons.<sup>(3)</sup>

This congenital cardiomyopathy is characterized by the presence of abnormal trabeculations in the LV, mostly in the apex. It may be associated to LV dilation or hypertrophy, systolic and/or diastolic dysfunction, or to congenital heart diseases.<sup>(3,4)</sup> The prevalence of LVNC in the general population has not been definitely established. However, the prevalence in adult patients referred to echocardiography laboratory was 0.014% (34 cases in 15-year follow-up).<sup>(5)</sup> And the prevalence in members of affected families ranges from 18% to 50%.<sup>(6)</sup>

Nonetheless, these data are probably underestimated considering more knowledge about the disease and improved imaging diagnosis techniques, which have led to increased cases among young and asymptomatic individuals, including athletes.<sup>(7)</sup>

The objective of the present study was to describe a case of goalball athlete with disability presenting with LVNC, but asymptomatic.

## CASE REPORT

A 14-year-old female goalball athlete with disability, went to the Sport Cardiology Department with a history of electrocardiographic abnormalities during a pre-competition assessment. Patient was asymptomatic and was training, on average, 6 hours per week. In her past medical history, she was diagnosed as congenital nistagmus and associated visual impairment, what enabled her to take part in official competitions of goalball (visual class B3 – athletes who are able to define images). She had no other comorbidities. Family history was negative for cardiovascular diseases or sudden death.

Upon cardiovascular clinical examination, heart rate of 62bpm, blood pressure 110/70mmHg, normal heart sounds, no murmurs or other significant signs of cardiovascular disease. In the neurological examination, visual impairment and horizontal nystagmus were

observed, with no other motor or sensory deficits. The physical examination presented no other significant findings.

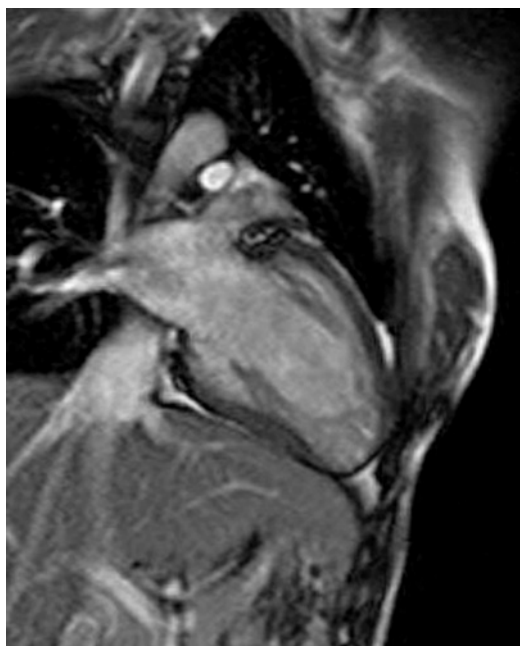
To complement investigation, a resting electrocardiogram (Figure 1), was requested, and showed sinus rhythm with ventricular extrasystoles, QRS electrical axis  $\approx -30^\circ$ , heart rate 56bpm, left anterosuperior fascicular block, and left ventricular overload by Cornell index (voltage), besides presence of Q waves in D1 and AVL. Laboratory tests, treadmill stress test, echocardiogram and 24-hour Holter electrocardiography were normal.

The patient was oriented to discontinue sport activities until defining diagnosis. Later, a cardiac magnetic resonance imaging (CMR) was performed with intravenous administration of gadolinium, and showed left ventricular noncompaction (non-compacted to compacted layer ratio equal to 2.5) and mild biventricular systolic dysfunction (Figures 2 and 3). The combined findings were considered sufficient to make diagnosis of LVNC, and the patient was oriented to not practice sports activities. A prophylactic treatment with acetylsalicylic acid 100mg/day was prescribed. The clinical follow-up was initially performed every three months.

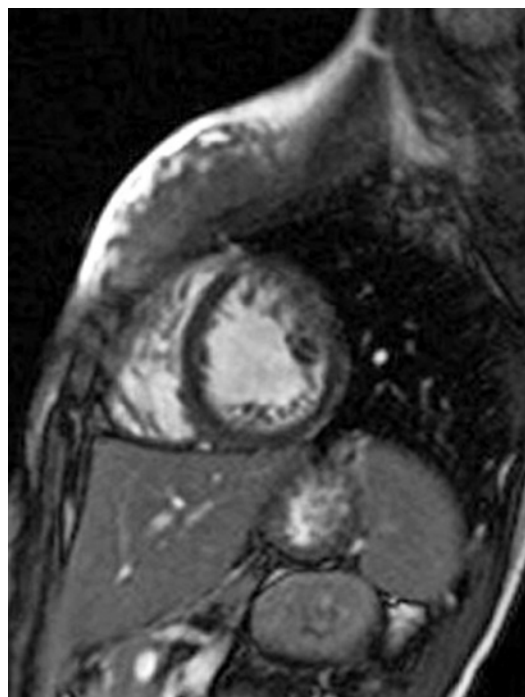
Since the patient remained asymptomatic during the follow-up period of approximately one year, with no history of syncope, significant impairment of ventricular function or marked tachyarrhythmia in treadmill stress test, she was allowed to return to goalball competitive participation.<sup>(8)</sup> Clinical follow-up was performed every 6 months.



**Figure 1.** Resting electrocardiogram shows signs of left ventricular overload (Cornell index – voltage), Q wave in inferolateral wall and isolated ventricular extrasystole



**Figure 2.** Cardiac magnetic resonance imaging in coronal plane shows cardiac chambers with preserved dimensions and increased myocardial trabeculation



**Figure 3.** Cardiac magnetic resonance imaging in sagittal plane shows left ventricular noncompaction: a non-compacted layer of 10mm and a compacted layer of 4mm

## DISCUSSION

Left ventricular noncompaction is considered a rare congenital cardiomyopathy, with a distinct

morphological presentation of the myocardium (trabeculation), which is classified in the group of primary genetic cardiomyopathy.<sup>(9)</sup> Most cases in adults present autosomal dominant disorder, but family inheritance and X-linked inheritance patterns have been described. The presence of three morphological elements characterizes the disease: prominent LV trabecular layer, compacted layer and intertrabecular recesses communicating with the left cavity. By definition, noncompaction occurs primarily in the LV, but it may involve the right ventricle and present as biventricular condition, or even an isolate right ventricle variant.<sup>(10,11)</sup>

The clinical spectrum of LVNC varies extensively and comprises from total absence of symptoms to clinical manifestations of heart failure, arrhythmias or thromboembolism.<sup>(12)</sup> The mean time for onset of symptoms after diagnosis is approximately 3.5 years.<sup>(13)</sup> Upon diagnosis, the most frequent clinical manifestations include dyspnea (79%), heart failure functional class III and IV (35%), chest pain (26%) and chronic atrial fibrillation (26%).<sup>(5)</sup> The associated arrhythmias most often described are atrial fibrillation and ventricular tachycardia. The incidence of sudden death varies between 0% and 18% in observational studies.<sup>(12)</sup> The differential diagnosis of LVNC should include dilated cardiomyopathy, hypertensive heart disease, apical hypertrophic cardiomyopathy, athlete's heart, infiltrative cardiomyopathy and endomyocardiofibrosis.<sup>(12)</sup>

The electrocardiographic findings most frequently found in LVNC cases are T wave inversion, prolongation of corrected QT interval, ST depression, left ventricular hypertrophy, intraventricular conduction disorder and prolongation of PR interval.<sup>(11)</sup> However, there is no electrocardiographic findings pathognomonic of LVNC. Traditionally, echocardiography is the initial method employed for diagnosis of LVNC, which is made when the non-compacted to compacted layer ratio is greater than two. Nonetheless, it is difficult to visualize the apical region of heart in echocardiography. Therefore, other imaging modalities, such as CMR, computed tomography and left ventriculography gained relevance to confirm or rule out diagnosis.<sup>(9)</sup> The imaging method most often chosen is CMR, for providing a more detailed heart morphology. The most accepted CMR criterion defines the ratio between non-compacted to compacted layers, measured at the end of diastole, greater than 2.3 to establish diagnosis of the disease.<sup>(3,12)</sup>

Treatment is usually directed to three most frequent complications of LVNC: heart failure, arrhythmias and thromboembolic complications. Some authors suggested electrophysiological study

and 24-hour Holter electrocardiography as initial assessment, and yearly follow-up of patients, given the inherent risk of sudden cardiac death.<sup>(14)</sup> Antiplatelet agents or systemic anticoagulants should be considered, especially when the atria or LV are dilated. In children, antiplatelet agents could be an option in cases of left ventricular systolic dysfunction, evidence of spontaneous echocardiographic contrast, severe LV dilation or atrial dilation.<sup>(4)</sup>

A recent official statement of the American Heart Association (AHA) recommends considering participation in competitive sports in cases of asymptomatic patients diagnosed as LVNC, provided the athletes present normal systolic function, no evidence of significant ventricular tachyarrhythmia in outpatient clinic control or treadmill stress test, and have no past history of unexplained syncope (Class IIb; evidence level C).<sup>(8)</sup> However, it has been suggested that orientation for sports participation be individualized, taking into account some factors, such as presence or absence of symptoms (especially during exercises), importance of sports for the athlete, myocardial dysfunction grade and extension of trabeculations. There is still a discussion about the definition of more strict diagnostic criteria for the athletes' population, aiming to minimize the risk of overdiagnosis in this group.<sup>(11)</sup>

## CONCLUSION

There is no consensus on eligibility criteria for sports participation in cases of left ventricular noncompaction. Therefore, it is prudent to individualize the decision about sports practice, and consider participation of asymptomatic individuals in competitive sports, with no repercussion on the disease.

## AUTHORS' INFORMATION

Maia EC: <http://orcid.org/0000-0001-9255-3478>  
 Savioli FA: <http://orcid.org/0000-0002-8871-4512>  
 Pinheiro SR: <http://orcid.org/0000-0003-4874-313X>  
 Echenique LS: <http://orcid.org/0000-0001-5182-2192>  
 Oliveira Filho JA: <http://orcid.org/0000-0002-5591-0285>

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