

# Isolated left ventricular noncompaction in an asymptomatic athlete (RCD code: III-5A.1.0)

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## Abstract

Left ventricular noncompaction (LVNC) is a rare form of cardiomyopathy due to abnormal morphogenesis of the endocardium and myocardium that occurs in early stages of foetal life. Given the increased risk of sudden cardiac death associated with LVNC, athletes with this diagnosis should be excluded from most competitive sports with the possible exception of those of low intensity (class IA) in selected cases. We report de novo diagnosed LVNC in an asymptomatic young footballer. JRCV 2014; 1 (7): 21–23

**Key words:** left ventricular noncompaction, athlete, cardiomyopathy, sudden cardiac death

## Background

Left ventricular noncompaction (LVNC) is a rare form of cardiomyopathy due to abnormal morphogenesis of the endocardium and myocardium that occurs in early stages of foetal life [1]. Given the increased risk of sudden cardiac death associated with LVNC, athletes with this diagnosis should be excluded from most competitive sports with the possible exception of those of low intensity (class IA) in selected cases [2]. We report de novo diagnosed LVNC in an asymptomatic young footballer.

## Case report

A 21-year-old professional football player had undergone cardiovascular assessment as a part of routine screening. He had no family history of cardiac diseases or sudden deaths. No abnormalities in physical examination were found. Resting ECG showed sinus bradycardia 50/min and early repolarization (recording within normal limits). Wenckebach-type AV block and nocturnal sinus pauses (the longest 2.3 sek) were recorded in 24-h ECG monitoring. Moreover episodes of atrial ectopic rhythm were found. Cardiopulmonary exercise test was performed in a protocol designed for professional athletes. Exercise lasted for 17 min. The patient achieved 16,2 METs. No ST-segment changes nor arrhythmias

during the test were recorded. Peak oxygen uptake was 42 ml/kg/min (the value within the normal range).

Transthoracic echocardiography showed enlargement of four cardiac chambers and mildly impaired left ventricular function (EF 42%) (Figure 1). Periapical thickening of left ventricular myocardium was noted. The ventricular wall in this region was inhomogeneous in appearance. Compacted, epicardial layer and a thick, noncompacted endocardial zone could be distinguished. The ratio of the thickness of noncompacted to compacted myocardium layers was 2.3. No additional pathologies were found. The presence of two-layer structure of the left ventricular wall with diastolic non-compacted to compacted ratio 3,8 confirmed the diagnosis of LVNC (Figure 2). The patient's case history was discussed during multidisciplinary consultations. According to expert's consensus aspirin as a prevention of thromboembolic complications and ACEI should be considered in asymptomatic patients with diagnosed LVNC. Echocardiographic screening in asymptomatic family members has been advocated. The athlete was excluded from competitive sport.

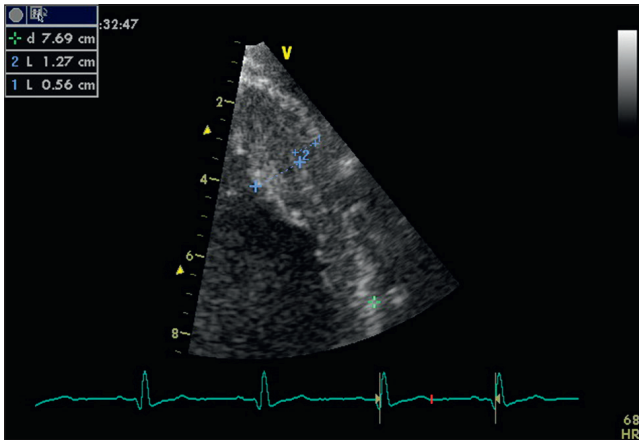
## Discussion

According to the definition, the professional athlete is a person who participates in an organized team or individual sport that requires regular competition against others as a central compo-

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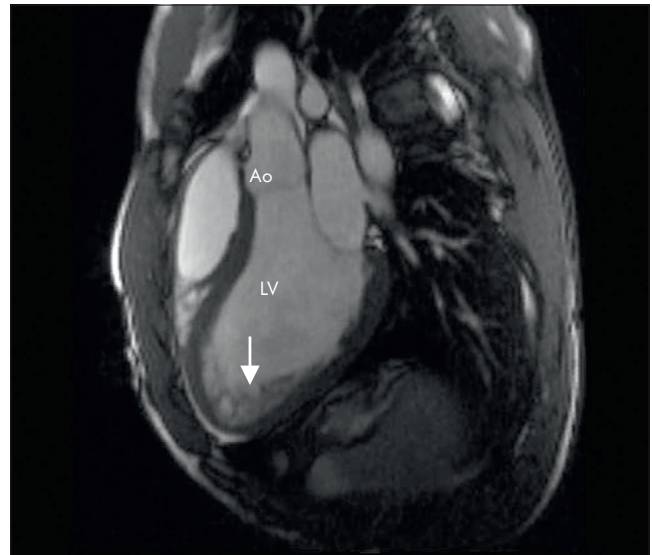


**Figure 1.** Transthoracic echocardiography. Apical region of the left ventricle. The measurement of the ratio of thickness of noncompacted to compacted myocardium layers

ment, places a high premium on excellence and achievement, and requires some form of systematic (and usually intense) training [2]. Competitive sports activity is associated with a significant increase of the risk of sudden death (2.5-fold in age group between 12–35 years old) and its incidence is estimated on 1–2 per 100 000 athletes per annum [3,4]. Sport is not “per se” cause of the increased mortality but it acts as a trigger of cardiac arrest in those athletes with previously undetected cardiovascular conditions. The causes of SCD among athletes are strongly correlated with age. In adolescents and young athletes (<35 years), the leading causes are congenital cardiac diseases, particularly hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, congenital coronary artery anomalies, and other forms of cardiomyopathies [5]. By contrast, in older athletes (>35 years), the most common cause of fatal events is coronary artery disease.

Left ventricular noncompaction is a rare form of cardiomyopathy due to abnormal morphogenesis of the endocardium and myocardium that occurs between 5-th and 8-th week of fetal life [1]. It is characterized by prominent myocardial trabeculations accompanied by deep inter-trabecular recesses which lie in continuity with the left ventricular cavity. There are discrepancies in LVNC classification, as the American Heart Association has classified LVNC as a primary genetic cardiomyopathy whereas the European Society of Cardiology places this pathology in the category of unclassified cardiomyopathies [6,7]. Defects of genes encoding various proteins including taffazin, alpha-dystrobrevin, beta-myosin heavy chain, lamin and calsequestrin have been identified in near 50% of patients with LVNC [8].

Phenotype of LVNC frequently resembles typical characteristic of dilated cardiomyopathy and its symptoms are unspecific and include dyspnoe, impaired exercise tolerance, syncope, thromboembolic complications and sudden cardiac death [9]. LVNC is diagnosed within a wide range of ages. The course and symptoms of the disease differ from case to case. The more advanced symptoms of cardiac insufficiency at the time of diagnosis, the worse prognosis can be expected. Vast majority (87%) of patients with LVNC have abnormal ECG [10]. Echocardiography is a primary diagnostic tool of LVNC, but due to various diagnostic criteria and suboptimal



**Figure 2.** Cardiac magnetic resonance. An arrow showing trabeculations in the left ventricle (arrow). LV – left ventricle, PA – pulmonary artery, Ao – aorta

echo quality, in most of cases magnetic resonance is recommended. Final diagnosis of LVNC should reflect clinical picture and multimodality imaging [11].

LVNC is a rare cardiovascular disease and no specific therapy is available. We previously reported a case of the **left ventricular non-compaction in patient with congenital diaphragmatic hernia causing cardiac dextroposition** [12]. **In our opinion** aspirin as a prevention of thromboembolic complications and ACEI should be considered in asymptomatic patients with diagnosed left ventricular noncompaction and mildly impaired left ventricular function. Currently, taking into account asymptomatic course of disease and some conduction problems (second-degree AV block type I), the decision to indicate beta-blockers was postponed. According to position statement of the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases screening is indicated in first-degree relatives of patients with LVNC [13]. Electrocardiogram and echo should be started in newborns and then systematically repeated until 50–60 years of age.

## Management strategy

Currently, only medical history, physical examination and resting ECG are obligatory in pre-participation screening of competitive athletes [14]. Until now, sparse cases of athletes with diagnosed LVNC were described and all of them were symptomatic [15]. The most common symptom was brief syncope during exercise; thromboembolic events including stroke, transient ischaemic stroke and pulmonary embolism were also described. The athlete described above was asymptomatic, had no family history of cardiac diseases or sudden cardiac death and no abnormalities in resting ECG. The suspicion of LVNC was based on transthoracic echocardiography, which is obligatory part of screening of competitive athletes in our department. In our opinion, TTE per-

formed by cardiologists experienced in sports cardiology should become an indispensable element in pre- participation screening of competitive athletes.

## Conclusion

Data on optimal management of LVNC are limited and there is no specific therapy. The athlete with diagnosed LVNC should be excluded from competitive sport. Aspirin as a prevention of thromboembolic complications and ACEI are an important part of treatment of asymptomatic patients with LVNC.

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