

# ICD implantation in left ventricular noncompaction: A case report and review of the literature

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## Abstract

*Left ventricular noncompaction (LVNC) is an uncommon cardiomyopathy characterized by the persistence of fetal myocardium with a pattern of prominent trabecular meshwork and deep intertrabecular recesses, systolic dysfunction and left ventricular dilatation. It is thought to be caused by the arrest of normal endomyocardial morphogenesis. There is no consensus on the definition, diagnostic criteria, pathogenesis or treatment of LVNC. We report the case of a 43 year-old patient with LVNC, nonsustained ventricular tachycardia and family history of sudden cardiac death (SCD). An implantable cardioverter-defibrillator (ICD) was prophylactically implanted because of the individual's high SCD risk. Although ICD is an effective option for preventing SCD, data on the long-term follow-up of patients with LVNC is limited. (Cardiol J 2011; 18, 6: 691–694)*

**Key words:** left ventricular noncompaction, implantable cardioverter-defibrillator, ventricular tachycardia, heart failure, arrhythmia

## Introduction

Left ventricular noncompaction (LVNC), first described in 1990, is an uncommon cardiomyopathy characterized by the persistence of fetal myocardium with excessive prominence of trabecular meshwork and deep intertrabecular recesses, systolic dysfunction and left ventricular (LV) dilatation [1]. With a prevalence of 0.014%, men are more affected than women [2]. Although LVNC has been classified as a primary cardiomyopathy of genetic origin, its definition, diagnostic criteria and treatment modalities are still being debated. It has been suggested that it is caused by an arrest of the normal process of intrauterine endomyocardial and myocardial morphogenesis [1]. Clinical manifestations are highly variable, ranging from no symptoms to a progressive deterioration in cardiac function that results in congestive heart failure, systemic thromboemboli, arrhythmias, and sudden cardiac

death (SCD) [3, 4]. Outcomes and appropriate therapies remain poorly defined. Pharmacological treatment and implantable cardioverter-defibrillators (ICD) may be considered for the management of ventricular arrhythmias (VA) in patients with this cardiomyopathy. The prognosis is generally poor, but may be improved by early diagnosis with risk stratification and proper management.

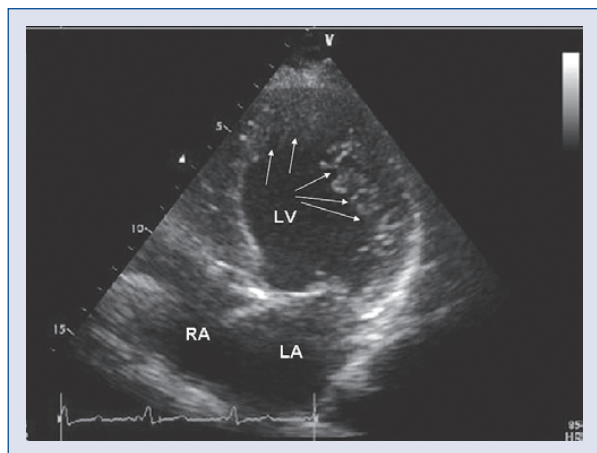
## Case report

A 43 year-old female patient was admitted to our unit with shortness of breath. She had dyspnea, decreased exercise tolerance and palpitations of two years' duration. On admission, the patient was in a good hemodynamic condition and in New York Heart Association (NYHA) class II. Physical examination and laboratory tests revealed no abnormality. The electrocardiogram (ECG) showed normal sinus rhythm with findings of LV hypertrophy.

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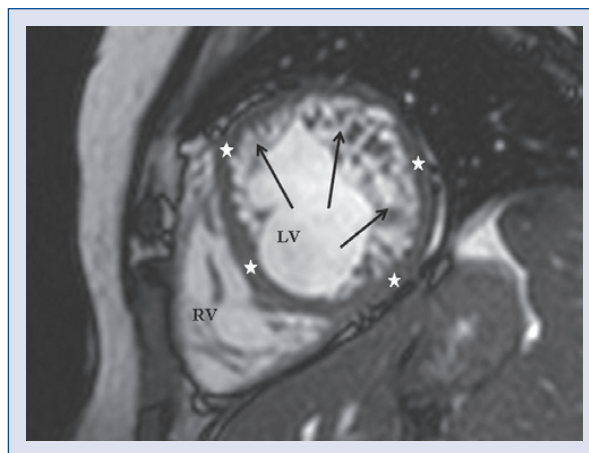
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**Figure 1.** Heavy trabeculation of the left ventricle's apical and lateral walls; LA — left atrium; RA — right atrium; LV — left ventricle.

Echocardiography revealed LV dilatation (end-diastolic volume 173 mL), normal wall thickening (both interventricular septum and posterior wall were 8.5 mm), widespread LV hypokinesia with an ejection fraction (LVEF) of 33%, restrictive type diastolic filling pattern with mitral E/E' ratio of 17, moderate mitral regurgitation and pulmonary hypertension (estimated systolic pulmonary artery pressure: 45 mm Hg). Heavy trabeculations were noted at the LV apical and lateral walls (Fig. 1). Coronary angiography revealed normal coronary arteries. Basal and contrast-enhanced cardiac magnetic resonance imaging (MRI) confirmed the presence of trabeculation and intertrabecular recesses, with a two-layered structure of the endocardium with an increased noncompacted to compacted ratio (> 2.0) in the LV lateral wall, as well as global hypokinesia and an increased LV volume, which were all compatible with LVNC (Fig. 2).

The patient received aspirin 100 mg/day, carvedilol 12.5 mg/day, and spironolactone 25 mg/day. She was considered for a possible ICD implantation for primary prevention of SCD. The 24-hour ECG Holter monitoring showed sinus bradycardia, frequent episodes of ventricular bigeminy, and non-sustained ventricular tachycardia (VT). During hospitalization, several episodes of asymptomatic VT was observed on ECG monitoring. Considering LVNC with decreased LVEF, nonsustained VT episodes and a family history of SCD, the patient was a class I (evidence level B) according to the recent guidelines for the management of patients with VA and the prevention of SCD [5]. A single-chamber ICD device (Maximo VR, Medtronic, Inc., Minne-



**Figure 2.** Cardiac magnetic resonance imaging end-diastolic short axis bright blood B-TFE sequence cine images showing a markedly dilated left ventricle with compacted myocardium (\*) and areas of substantial trabeculations (ie noncompacted myocardium) in the anterior, septum and lateral wall (arrows); LV — left ventricle; RV — right ventricle.

apolis, MN, USA) was implanted. The patient was discharged without complications. Follow-up at six months revealed no episodes of VT or syncope.

## Discussion

LVNC is characterized by electrical abnormalities including low voltage and scar areas, mainly related to the presence and extent of myocardial fibrosis rather than noncompacted myocardium [6]. In previous studies, ventricular tachyarrhythmias have been reported in up to 47% of symptomatic patients referred to a tertiary referral center, and SCD reported in 13–18% of (mostly adult) patients with LVNC [2, 7]. Such findings support the hypothesis that noncompacted ventricular myocardium may be a highly arrhythmogenic substrate with a high recurrence rate in patients with a history of syncope and/or VT. Although all the three main mechanisms responsible for arrhythmogenesis (re-entry, triggered activity, and abnormal automaticity) have been implicated in the genesis of VA in patients with LVNC, myocardial macrore-entry is probably the responsible mechanism. The histological examination of patients with LVNC shows evidence of continuity between the LV endocardium and the deep intertrabecular recesses that may be suitable for substrate formation for propagation of various reentrant circuits subjacent to scar [7].

Although a high incidence of VT and SCD is expected, little consensus exists on the treatment of ventricular and supraventricular arrhythmias in this cohort of patients. What is suggested at the moment is that patients with LVNC should be closely monitored by conventional cardiologic diagnostic tools, with appropriate measures taken only if indicated [8]. Although implantation of ICD is a treatment option, its superiority over medical therapy is still being debated and there is limited data on long-term follow-up [4]. While some authors favor ICD implantation to prevent SCD, others prefer choosing the patient at highest risk. Kobza et al. [9] suggested that ICD therapy might be effective for primary and secondary prevention in these patients. They conducted a retrospective study on 12 patients with LVNC, who underwent ICD implantation for secondary and primary prevention. During a median follow-up of 36 months, five patients received appropriate ICD therapy. Similarly, Duru et al. [10] reported a patient with LVNC and ICD who had developed numerous different VT episodes that were appropriately treated by the device. Celiker et al. [11] reported a six year-old child with LVNC and ICD who had three ventricular fibrillation episodes which were treated with appropriate shocks during a follow-up of 16 months. On the other hand, Stanton et al. [12] analyzed the follow-up data of 30 patients with LVNC and reported no appropriate therapies during a mean follow-up of 2.5 years among the 11 LVNC patients who were implanted with ICD. They also did not find any difference in mortality between patients with LVNC and patients with dilated cardiomyopathy during follow-up. They concluded that deaths in the LVNC group (three patients out of 30) were observed only in patients with decreased LVEF, suggesting that ICD therapy might be reserved for this subgroup.

While the necessity of ICD implantation is being argued, nearly all reports have pointed out that supraventricular tachyarrhythmias, especially new-onset atrial fibrillation, might lead to inappropriate ICD discharges [12]. Patients with LVNC may develop supraventricular tachyarrhythmias; atrial fibrillation had been reported in up to 29% of patients [2, 7]. Kobza et al. [9] reported that intermittent supraventricular tachyarrhythmias were observed in 66% of patients, and that patients with supraventricular tachyarrhythmias were more prone to inappropriate ICD shocks. Therefore, they suggested devices with reliable detection enhancements be considered in these patients.

It is extremely hard to establish a correct stratification of the arrhythmogenic risk in these patients.

The LV systolic dysfunction assessed either by echocardiography or cardiac MRI and detection of sustained VT during 24-hour Holter monitoring might be useful in determining the risk [13]. Prolonged monitoring by means of a loop recorder might also provide better information [14]. The role of electrophysiological study (EPS) to determine the risk for VA and SCD is not well established in LVNC. Kobza et al. [9] reported that the three patients in whom a sustained VT had been induced during EPS received appropriate ICD shocks. However, they stated that the number of patients in their study was too small to allow them to devise parameters that would predict risk for SCD. Some authors prefer to implant an ICD even if no sustained VT or fibrillation is observed during EPS [14]. A recent study pointed out that symptoms of heart failure, a history of sustained VT or an enlarged left atrium were associated with an unstable course and more severe prognosis [15]. Oechslin et al. [7] reported that certain clinical characteristics were more frequently observed in non-survivors compared with survivors with LVNC, including higher LV end-diastolic diameter on presentation, NYHA class III–IV, permanent or persistent atrial fibrillation, and bundle branch block. Patients with these high risk features might be candidates for early, aggressive intervention, including consideration of ICD implantation and evaluation for transplantation.

In our case, we decided to perform ICD implantation on the basis of the young age of our patient, the family history of SCD, the LV systolic dysfunction and the documentation of nonsustained VT on Holter monitoring.

## Conclusions

Life-threatening VA may occur in patients with LVNC. Its low prevalence and the limited data available in the literature do not allow us to make a firm conclusion on the prognosis of these patients. The best therapeutic decision should be based on the patient's own clinical features and the physician's judgement. We would currently recommend that a patient with LVNC be implanted with an ICD in cases of aborted cardiac arrest, sustained VT, syncope related to VA, family history of SCD, or severely impaired LVEF.

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