## CASE REPORTS

# ICD Implantation in Noncompaction of the Left Ventricular Myocardium: A Case Report

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*Isolated noncompaction of the ventricular myocardium (INVM) is an uncommon cardiomyopathy characterized by the persistence of fetal myocardium with prominent trabecular meshwork and deep intertrabecular recesses, often associated with systolic dysfunction and ventricular dilatation. A 23-year-old man from Burkina Faso was referred to our operative unit with a diagnosis of INVM, made with echocardiogram and magnetic resonance imaging and nonsustained ventricular tachycardia. The literature reports the incidence of malignant ventricular arrhythmias in as many as 47% of the patients and sudden cardiac death in almost 50% of them and this supported our decision to perform implantable cardioverter-defibrillators implantation. (PACE 2009; 32:1092–1095)*

***sudden cardiac death, ICD, left ventricle noncompaction***

### Introduction

Isolated noncompaction of the ventricular myocardium (INVM), also known as "spongy myocardium," is an uncommon cardiomyopathy characterized by the persistence of fetal myocardium with excessive prominence of trabecular meshwork and deep intertrabecular recesses and is caused by the interruption of the normal process of endomyocardial and myocardial morphogenesis. These features are also associated with systolic dysfunction and ventricular dilatation.[1–3] Echocardiography is the gold standard procedure for diagnosis. In the largest series of patients with INVM, the prevalence of those referred to echocardiography laboratory and describing similar findings was 0.014%; men were more affected than women.[4] Clinical manifestations ranging from no symptoms to congestive heart failure, systemic thromboemboli, and also arrhythmias and sudden cardiac death (SCD).[1–3]

Implantation of implantable cardioverter-defibrillators (ICD) in these patients is a treatment option, but data on long-term follow-up are limited. The aim of this case report was to analyze the management of a patient with INVM undergoing ICD implantation.

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### Case Report

A 23-year-old man from Burkina Faso was referred to our unit with a diagnosis of INVM based upon echocardiography and cardiac magnetic resonance imaging (MRI) performed after a history of decreased exercise tolerance and palpitations. The first surface electrocardiogram (ECG) showed sinus rhythm, normal QRS duration, left ventricular hypertrophy, early repolarization, T-wave "lability," and negative U wave in $V_5$–$V_6$ leads; the 24-hour ECG Holter monitoring showed sinus bradycardia, frequent episodes of ventricular bigeminism, and nonsustained ventricular tachycardia (NSVT). Echocardiographic evaluation showed left ventricle dilatation (end-diastolic volume 180 mL), normal wall thickening (interventricular septum and posterior wall 10 mm), widespread left ventricular hypokinesis with a left ventricle ejection fraction (LVEF) of 35%, heavy trabeculation of the left ventricle's apical and lateral wall, while the left atrium, aortic root, right chambers, and inferior cava vein were normal; color Doppler displayed normal diastolic filling pattern, mild-to-moderate grade mitral regurgitation, and mild grade tricuspidal regurgitation in the absence of pulmonary hypertension. Neither interventricular nor intraventricular dyssynchrony was described. Basal and contrast-enhanced MRI confirmed the presence of trabeculation and intertrabecular

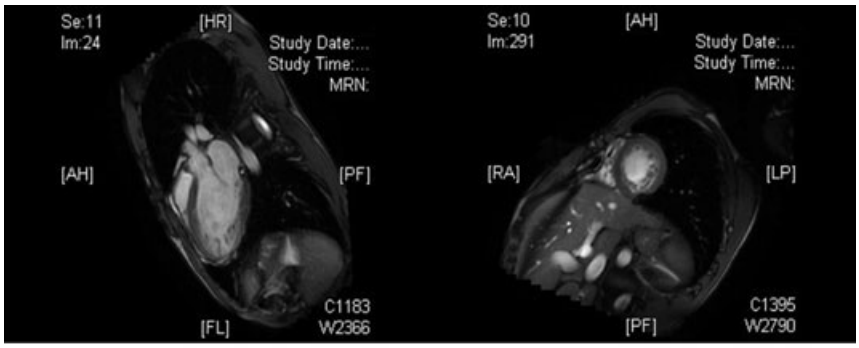


**Figure 1.** *Long-axis (A) and short-axis (B) cardiac MRI showing apical and anterolateral trabecular meshwork and deep intertrabecular recesses.*

recesses as well as global hypokinesis (EF 27%) and an increased left ventricle volume (Fig. 1). Several episodes of asymptomatic NSVT occurred while performing the MRI Virus isolation, previously performed, was negative; the coronaroventriculography showed normal vessels and noncompaction of the left ventricle. According to all these findings and the clinical manifestations of the disease, the patient received aspirin, carvedilol, and enalapril drug therapy and was referred to our unit to evaluate a possible ICD implantation for primary prevention of SCD as a bridge to heart transplant.

Upon admission, 3 months after the diagnosis of INMV, the patient was in good hemodynamic conditions and in New York Heart Association (NYHA) class II. We performed a surface ECG (Fig. 2) and an echocardiogram that confirmed the previous results, except for the LVEF that we had estimated as 30%; this could, however, be caused by disease progression or operator-dependent variability (Fig. 3). All routine blood tests were normal, and the ECG monitoring performed during the patient's stay in our intensive care unit before ICD implantation showed persistent sinus bradycardia and short runs of atrial tachycardia.

In summary, the patient was in NYHA class II and was affected by INVM. The ICD implantation guidelines showed a class II (evidence level B) indication according to the recent Italian Association of Arrhythmology and Cardiostimulation guidelines, although considering LVEF, the patient was a class I (evidence level B) according to the American College of Cardiology/American Heart Association/European Society of Cardiology (ACC/AHA/ESC) guidelines for the management of ventricular arrhythmia (VA) patients and SCD prevention.[5,6] We performed an electrophysiological (EP) study in which the VA induction protocol included programmed stimulation at three basic cycle lengths (500, 400, and 330 ms) and up to three extrastimuli with a minimum coupling interval of 200 ms from two sites in the right ventricle (apex and outflow tract); a third extrastimulus was introduced during a basic drive cycle length of minimal 500 ms after completion of programmed ventricular stimulation with one and two extrastimuli during paced cycle lengths of 500, 400, and 330 ms. We observe no sustained ventricular tachycardia or fibrillation during the test, but our decision to perform ICD implantation was supported by literature data reporting a presence of supraventricular arrhythmias and an incidence of malignant VA in as many as 47% of the patients and SCD in almost 50% of the patients for an adult population of INMV patients.[1,3,7] Because of the high prevalence of supraventricular tachyarrhythmias in these patients and based on our patient's persistent sinus bradycardia and need for $\beta$-blockers, we opted for a dual-chamber device. The patient was discharged with indications for a clinical follow-up and monitoring for potential need of a heart transplant. Upon the first ICD check, 1 month after implantation, no sustained VAs were recorded; the patient was in a good hemodynamic state and was still in NYHA class II.

## Discussion

According to literature data of last 20 years, clinical manifestations of INVM vary from asymptomatic to heart failure, systemic thromboemboli, arrhythmias, and SCD. While medical treatment of heart failure is established by international guidelines, less consensus exists on the treatment of ventricular and supraventricular arrhythmias in this cohort of patients. According to the published data, the incidence of malignant VA is reported in as many as 47% and SCD in 50% of adults affected by INVM[1,3,7]; a better prognoses is reported in the pediatric population.[3,8]

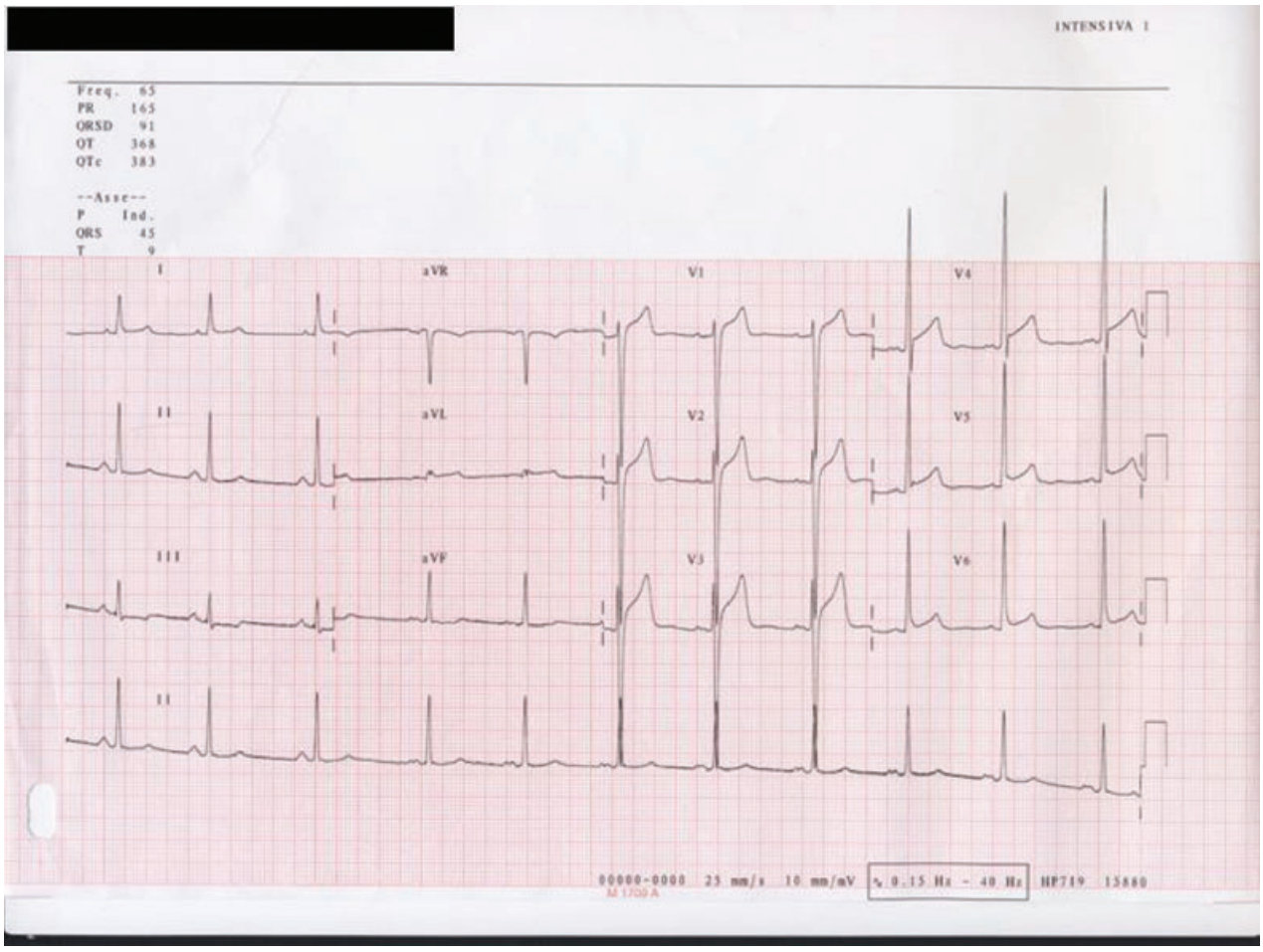


**Figure 2.** *Patient's surface ECG.*

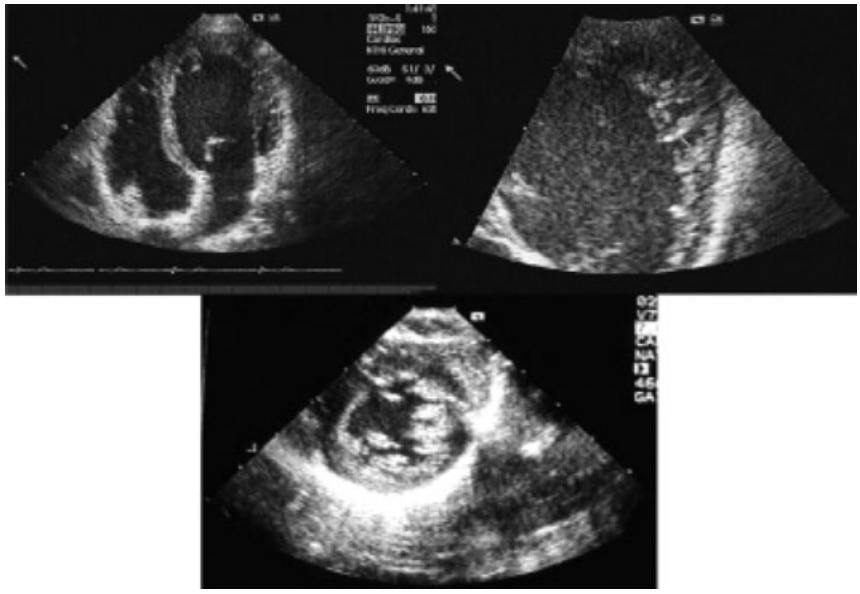

**Figure 3.** *INVM echocardiographic finding.*

A recent study conducted on an adult population seems to reduce the arrhythmogenic role of this cardiomyopathy even in the presence of left ventricle systolic dysfunction.[9] On the contrary, other recent evidences demonstrates a potential arrhythmogenic role in patients with INVM who underwent ICD implantation for secondary and primary prevention. In particular, Kobza et al. and Duru et al. showed how potentially life-threatening VA may occur in these patients and how ICD therapy may be effective also for those who did not show sustained ventricular tachycardia or fibrillation during EP study.[10,11] Similarly, Sato et al. illustrated how fatal VA might occur in INVM in the absence of left ventricular dysfunction and/or clinical evidence of myocardial ischemia.[12] It should be underlined, however, that VA have been associated with mortality in idiopathic dilated cardiomyopathy, and in some studies, increased ventricular volumes and impaired EF have put patients at a higher risk for SCD.[13]

All the three main mechanisms of arrhythmogenesis, reentry, trigger activity, and automatism, have been implicated in the genesis of VA in patients with IDC. Moreover, these arrhythmogenic substrates are often potentiated by electrolyte imbalance secondary to diuretic treatment or concomitant renal failure, by antiarrhythmic drugs, or by bradycardia.[14] In INVM patients, myocardial macroreentry is probably the mechanism responsible for VA, probably because trabecular meshwork and deep intertrabecular recesses represent a good anatomic substrate for its onset. In our opinion, it is extremely hard to establish a correct stratification of the arrhythmogenic risk in these patients, not only because INVM is an uncommon disease but also because patients are often affected by asymptomatic systolic dysfunction, representing an independent risk factor for SCD. Also, in our opinion, the 24-hour Holter monitoring is not the most appropriate diagnostic tool for arrhythmias in this small cohort of patients. A prolonged monitoring by means of a loop recorder would most likely provide us with better information.[9]

## Conclusion

VA and SCD are described in patients affected by INVM. The disease's low prevalence and the limited data available in literature do not allow to reasonably establish the prognosis of these patients; also, the therapeutic treatment is left to the individual physician and to his/her medical knowledge. On the basis of the young age of the patient, the history of NSVT, and the systolic dysfunction and also supported by the current guidelines,[5,6] we decided to perform ICD implantation. We consider our patient to be at a high risk for SCD and ICD was not only a life-saving treatment for his condition but also a bridge to a potential heart transplant.

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