

CASE REPORT

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## Isolated Left Ventricular Noncompaction Cardiomyopathy diagnosed by Transesophageal Echocardiography

Tariq Bhat<sup>1</sup>, James Lafferty<sup>2</sup>, Sumaya Teli<sup>3</sup>, Georges Abou Rjaili<sup>2</sup>, Yefim Olkovsky<sup>2</sup> and Thomas Costantino<sup>2</sup>

<sup>1</sup>Department of Medicine, Staten Island University Hospital, New York 10305, USA. <sup>2</sup>Division of Cardiology, Staten Island University Hospital, New York 10305, USA. <sup>3</sup>University of Sheffield School of Medicine, UK.

Corresponding author email: [mohiuddin\\_bhat@yahoo.com](mailto:mohiuddin_bhat@yahoo.com)

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**Abstract:** Isolated noncompaction of the ventricular myocardium has often been misdiagnosed as other cardiomyopathies because it is a relatively recently described cardiomyopathy with literature limited to case reports and case series and little awareness among physicians. We are reporting a case of isolated left ventricular noncompaction cardiomyopathy that was misdiagnosed for over two decades.

**Keywords:** ILVNC, cardiomyopathy, noncompaction, TEE, diagnosis of ILVNC

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## Introduction

Isolated noncompaction of the left ventricle is a congenital abnormality in the structure of ventricular myocardium due to a defect in morphogenesis during embryogenesis, characteristically leading to hypertrabeculations and deep recesses in the ventricular wall. Majority of patients clinically present with heart failure, arrhythmias, embolic events, conduction disorders, neurological abnormalities and recently reported mitral regurgitation. Echocardiography is considered the reference standard for diagnosis; even so the diagnosis may be overlooked. In a recent case series, diagnosis was initially missed in almost 90% of the cases.<sup>1</sup> We are reporting a case of isolated left ventricular noncompaction cardiomyopathy in our patient—a 78-year-old female, which was initially diagnosed in 1986 as hypertrophic cardiomyopathy. She carried this diagnosis for more than 20 years before she was actually found to have noncompaction cardiomyopathy with the help of transesophageal echocardiography and ventriculography.

## Case

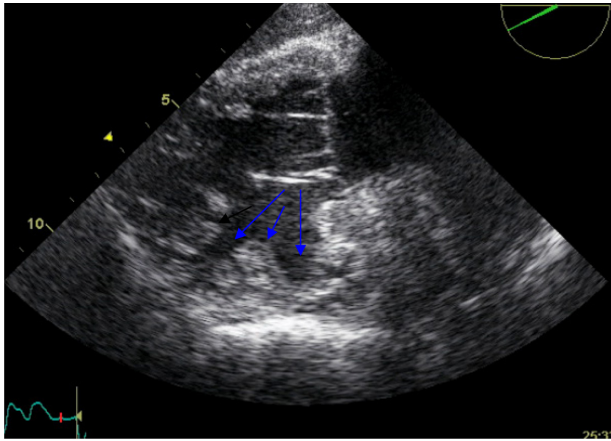
A 78-year-old female, who presented to our hospital with a chief complaint of shortness of breath at rest; she had reported progressive worsening of dyspnea on exertion, fatigue and worsening lower extremity edema for a few days. She was recently hospitalized for similar complaints in Norway where she had been treated for worsening congestive heart failure. Patient denied any chest pain and palpitations. She has a 24-year history of atrial fibrillation, which was noticed for the first time when she was admitted for an embolic stroke in 1986 and was diagnosed as having hypertrophic cardiomyopathy on transthoracic echocardiogram, which we believe was misdiagnosed ILVNC (isolated left ventricular noncompaction) due to a limited knowledge of this disorder at that time. Patient was started on coumadin for anticoagulation and is still on it. She was followed with no history of any cardiovascular events. During these years she slowly progressed to NYHA class II and was stable on small dose of furosemide and quinapril till this episode. Previous workup for ischemic cardiomyopathy had been negative including coronary angiogram. Now patient had progressed to NYHA class III. Her medications on presentation were Furosemide, Coumadin, Metoprolol Aspirin, Quinapril, Calcium and

Fosamex. No other past medical and surgical history. She never smoked or drank alcohol and had no allergies.

On examination the patient was found to be afebrile, in atrial fibrillation with a heart rate of 55 bpm, blood pressure of 116/67 mmHg and respiratory rate of 16 pm. Her height was measured to be 63 inches and weight to be 121 pounds. Cardiac auscultation revealed an irregularly irregular rhythm, and a grade 3/6 systolic murmur at the cardiac apex radiating to axilla. Bibasilar crepitation was noticed on lung auscultation. There was no hepatomegaly or splenomegaly, and there was 2+ lower extremity pitting edema. The electrocardiography (ECG) showed atrial fibrillation with nonspecific intraventricular conduction block. Chest X ray showed features of congestive heart failure and routine labs were within normal limits except high BNP. Patient was treated with intravenous diuretics for her worsening heart failure.

Two-dimensional and Doppler echocardiography (TEE) revealed an enlarged left atrium and hypertrophic left ventricle, mildly decreased LV systolic function (ejection fraction (EF) = 40%, moderate to severe mitral valve regurgitation). LVIDd (left ventricular inner diameter during diastole) was measured 4.2 cm and LVIDs (left ventricular inner diameter during systole) was measured to be 2.8 cm. Right ventricular size and morphology, as well as the function of the aortic and pulmonary valves, were normal. Right ventricular systolic pressure was measured at least 85 mmHg, and inferior vena cava was found dilated. All previous admissions and available cardiac imaging modalities were reviewed. There was suspicion of ILVNC based on previous left ventriculogram. A Trans-Esophageal Echocardiography (TEE) was done and showed apical and posterior trabeculations, which met the criteria for ILVNC (Figs. 1 and 2). Mitral valve showed moderate to severe regurgitation with dilated annulus, no vegetation or stenosis was found.

We performed left and right cardiac catheterization, left ventriculography to exclude any other cardiac abnormalities and confirm left ventricular noncompaction. The angiogram showed normal coronary arteries (Figs. 3 and 4). The left ventriculogram showed global depression in left ventricular function (Estimated EF = 30%) with hypertrophy and extensive trabeculations consistent with isolated noncompaction of the left ventricular myocardium and

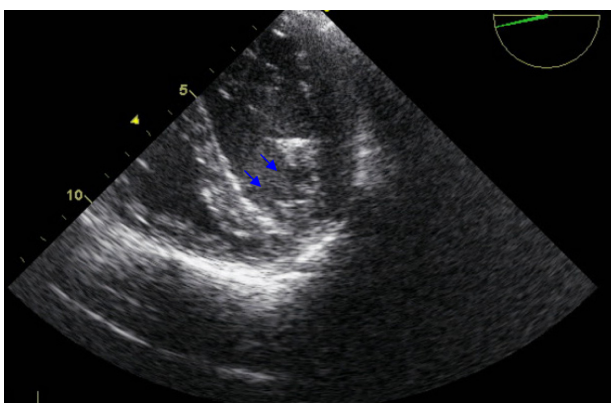


**Figure 1.** Trabeculations of left ventricular posterior wall shown by short arrows and recesses shown by long arrows on transesophageal echocardiography.

severe mitral regurgitation (Fig. 5). Although cardiac MRI would have been ideal for characterization of the myocardium, the presence of AICD was deemed as a contraindication in our patient. Cardiac CT was not clinically justified, because the risk of radiation and contrast exposure outweighed any benefit to the patient and her management.

## Discussion

Isolated noncompaction of left ventricular myocardium is a rare cardiomyopathy due to abnormal endomyocardial morphogenesis.<sup>2</sup> Awareness of ILVNC in the recent past has increased tremendously especially in the elderly. It is characterized by numerous excessively prominent ventricular trabeculations and deep intertrabecular recesses. Noncompaction was initially reported in the pediatric population and in association with complex congenital heart disease,<sup>3</sup> but is now

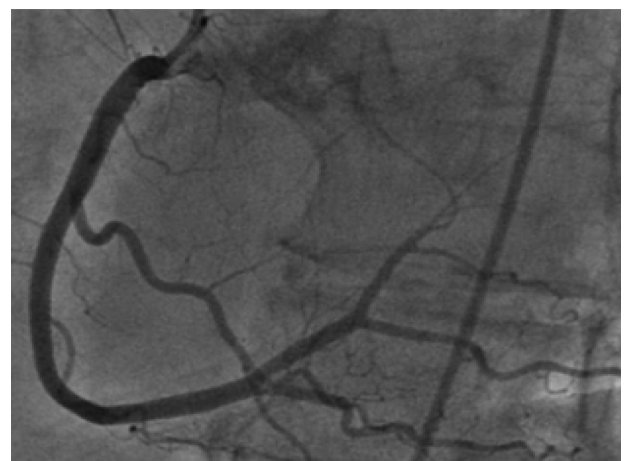


**Figure 2.** Left ventricular apical trabeculations shown by arrows during transesophageal echocardiography.



**Figure 3.** Normal left coronary circulation.

increasingly reported as IVNC in the elderly population.<sup>4</sup> ILVNC is usually familial, both autosomal and X-linked inheritance have been described, but sporadic cases have also been reported.<sup>5</sup> Even though this disorder has been believed to be rare but recent reports indicate it may be more common than previously thought. Due to lack of large studies and prolonged follow up, clinical presentation and long-term event rate is not well defined leading to either misdiagnosis or lag between clinical presentation to diagnosis. The majority of patients with ILVNC clinically present with heart failure, arrhythmias, embolic events, conduction disorders, and neurological abnormalities.<sup>2,4</sup> Severe mitral regurgitation associated with ILVNC has been also documented recently.<sup>6</sup> Common



**Figure 4.** Normal right coronary circulation.



**Figure 5.** Left ventriculogram showing contrast filling recesses between trabeculations.

patterns of arrhythmias and conduction disorders include atrial fibrillation, bradycardia, WPW syndrome and incomplete left bundle branch block can be observed.<sup>7</sup> Our patient presented for the first time when she was in her fifties with embolic stroke and atrial fibrillation, which was probably secondary to presence of underlying left ventricular noncompaction at that time. She received a diagnosis of hypertrophic cardiomyopathy at that time, which we believe was misdiagnosed ILVNC. Her systolic function got progressively worse. Her clinical course was complicated by episodes of ventricular arrhythmias and severe mitral regurgitation requiring AICD and mitral valve replacement. Although the outcome of patients with ILVNC is not clear, a recent small study has shown a trend towards worse prognosis than in the general population, but was found similar to dilated cardiomyopathy patients.<sup>8</sup> Early diagnosis of ILVNC is important due to many reasons, primarily because of its high mortality in symptomatic patients, secondly for screening first degree relatives as familial occurrence is known and thirdly because early institution of carvedilol has been described to improve ventricular mass and function in such patients.<sup>9</sup> Diagnosis and management of ILVNC is not only difficult but also controversial. Echocardiography is usually utilized for diagnosis of ILVNC, which is considered the reference standard for diagnosis. Jenni et al<sup>10</sup> established four echocardiographic criteria for ILVNC diagnosis and all four are required for diagnosis. Where as Chin

et al<sup>2</sup> proposed  $X/Y < \text{or} = 0.5$  for diagnosis, where X is the distance from epicardial surface to the trough of the trabecular recess and Y is the distance from epicardial surface to peak of trabeculation. Other imaging modalities that can be diagnostic as well as determine the severity and prognosis are CMR, CCT and left ventriculography.

Management of patients with ILVNC is similar to that of patients with other cardiomyopathies and should therefore include appropriate treatment for heart failure, management of arrhythmias, and oral anticoagulation to prevent systemic emboli in patients with impaired left ventricular function.<sup>5</sup> Implantation of an internal cardioverter defibrillator system and early listing of symptomatic patients for heart transplantation must be seriously considered.<sup>5</sup>

## Conclusion

This case highlights that noncompaction cardiomyopathy may be missed or misdiagnosed with two dimensional and Doppler echocardiography, which is considered standard and reliable imaging modality for this cardiomyopathy. The universal use of standard echocardiography as the sole diagnostic modality may lead to under diagnosis of noncompaction cardiomyopathy. New imaging modalities can be diagnostic as well as determine the severity and prognosis. These including MRI or MCE, CCT and ventriculography and should be strongly recommended in patients with heart failure and poor echocardiographic image quality and under high suspicion. Even though Trans-esophageal echocardiography has not been used often for diagnosis of left ventricular noncompaction, it proved helpful in our case and ventriculography confirmed the diagnosis.

## Author's Contributions

JL, YO and TC analyzed and interpreted the patient data. TB, GA and ST were involved in doing the literature review and manuscript preparation and TB was also instrumental in obtaining informed consent.

## Disclosures

This manuscript has been read and approved by all authors. This paper is unique and not under consideration by any other publication and has not been published elsewhere. The authors report no conflicts of





interest. The authors confirm that they have permission to reproduce any copyrighted material. Written and informed consent was obtained from the patient for publication of this case report along with images.

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