

## Left Ventricular Noncompaction in a Patient of Afro-Caribbean Descent: A Call for Risk Assessment in Immigrants from Tropical Regions

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### Introduction

Noncompaction of the left ventricular myocardium (LVNC) is a rare cardiomyopathy believed to be caused by intrauterine arrest of normal embryogenesis of the endocardium and myocardium. Noncompaction is characterized by presence of prominent trabeculae, deep intertrabecular recesses and thickening of the myocardium in two distinct layers: compacted and noncompacted. LVNC may or may not be associated with other congenital cardiac defects. Studies in heart failure patients of Afro-Caribbean (black) origin reveal a high prevalence (up to 30%) of myocardial trabeculations and the potential diagnosis of left ventricular noncompaction. It is unclear whether the myocardial morphology is representative of LVNC or whether it represents an ethnicity related epiphenomenon to increased cardiac preload. With increasing phenomenon of immigration from tropical countries to the United States, it is important to identify potential cardiovascular risks that can be attributed to ethnic background. Clinical presentation of LVNC may vary from an incidental finding on an echocardiogram with no symptoms to symptoms attributable to congestive heart failure, cardiac arrhythmias, and systemic thromboembolic phenomenon. Echocardiography is the initial modality of choice for diagnosis [1-5].

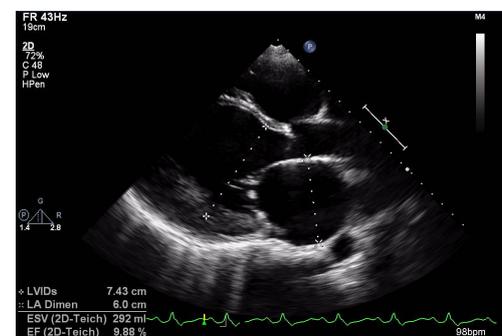
### Case Presentation

A 44 years old male patient of Afro-Caribbean descent with a past medical history significant for depression and anxiety presented at our heart failure clinic with complaints of worsening shortness of breath. Patient had recently moved to North Carolina from Barbados. Patient indicated a previous history of systolic heart failure diagnosed a year ago, which was believed to be secondary to his previous heavy alcohol intake. Patient quit alcohol use 2 years ago. Patient also had three previous hospitalizations in a community hospital for heart failure exacerbations and had received an implantable cardioverter defibrillator (~3 months ago) for primary prevention of sudden cardiac death. His primary care physician referred him to our clinic for further work up. His main symptoms were profound shortness of breath, abdominal bloating, fatigue and swelling of his feet. On exam his vital signs showed a blood pressure of 88/70 mm of mercury, pulse 102 per minute, respiratory rate of 22 per minute, with oxygen saturation of 96% on room air. Neck exam revealed elevated jugular venous pulsation at 10 cm above sterna angle. Chest auscultation revealed bibasilar crepitations and normal S1 and S2 sounds were heard with a summation gallop. Patient also had a 2/6 soft holosystolic murmur at the apex on auscultation. Abdominal examination was benign, with no obvious ascites. Patient did have bilateral ankle edema.

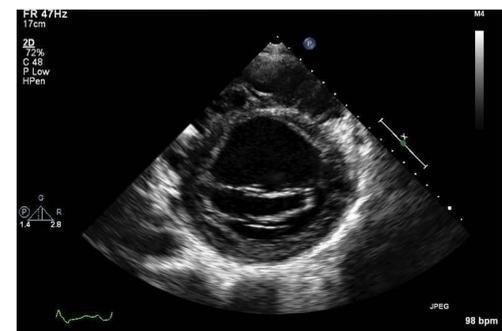
Laboratory investigations showed complete blood counts within normal limits. Blood chemistry was notable for elevated serum creatinine at 1.49 mg/dl and pro- Brain Natriuretic Peptide at 8720 pg/ml. Rest of the blood chemistry was within normal limits.

Transthoracic echocardiogram was ordered which showed left ventricular systolic dysfunction with myocardial contour suggestive

of non-compaction (Figures 1-5), decreased left ventricular ejection fraction at 10-15%, dilated left ventricle (Figure 1), diastolic left ventricular dysfunction with severe mitral regurgitation (Figure 6) and dilated left atrium. There was also evidence of moderate pulmonary hypertension with dilated pulmonary artery, moderate contractile right ventricular dysfunction and moderate tricuspid regurgitation. Patient was admitted and started on intravenous diuretic therapy.



**Figure 1:** Left Parasternal Long-Axis view showing dilated LV and LA with spongiform appearance of LV free wall.



**Figure 2:** Parasternal Short-axis view at the level of mitral valve showing spongiform appearance of the noncompacted myocardial layer and adjacent compact myocardium.

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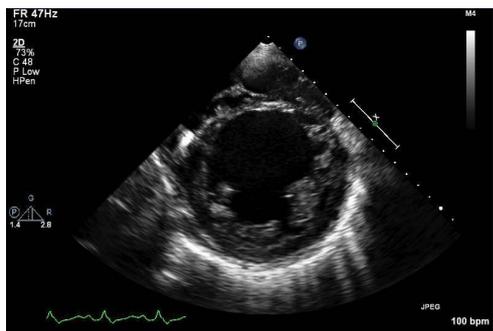


Figure 3: Parasternal Short-axis view at the level of papillary muscles showing trabecular spaces in noncompacted myocardium.

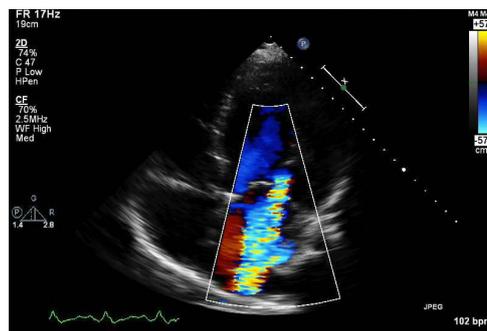


Figure 6: Color Doppler showing moderate mitral regurgitation in apical 4-chamber view.



Figure 4: Parasternal short-axis view at the LV apex showing spongiform appearance of the LV Anterior, anterolateral, lateral and inferior walls.

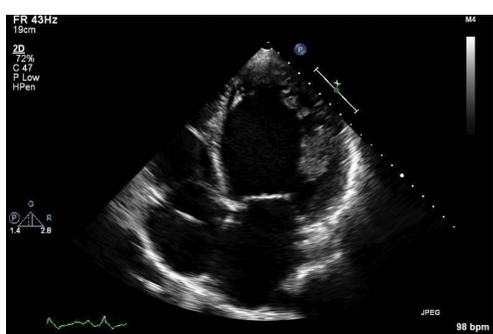


Figure 5: Apical four chamber view showing noncompacted myocardium in the LV free wall and apex. Also seen is a dilated LA and LV.

In view of his low cardiac output symptoms and inadequate diuretic response initially, he was subsequently started on inotropic therapy with milrinone infusion 0.5 mcg/kg/min along with guideline based heart failure pharmacotherapy. It was apparent after reviewing his echocardiogram that his left ventricular systolic dysfunction was likely due to noncompaction along with likely contribution from his heavy alcohol abuse. Cardiac magnetic resonance imaging could not be done as patient had an implantable cardioverter defibrillator in situ. Due to inotrope dependence, patient was referred to a tertiary care center for consideration for advance heart failure therapies including heart transplant or ventricular assist devices.

## Discussion

LVNC is a rare disorder and cause of noncompaction is not clearly understood. Some cardiac defects found associated with LVNC are

Ebstein's anomaly, bicuspid aortic valve, ventricular septal defect and transposition of great vessels. LVNC may also occur in some genetic and metabolic disorders like the Barth syndrome, Charcot-Marie-Tooth disease and the Melnick-Needles syndrome [6-9].

Oechslin et al. [10] reported a case series of 34 patients with male predominance (74%). Familial occurrence of left ventricular noncompaction was reported in 6 patients out of 34 (18%). End-systolic thickness of the noncompacted and compacted myocardium was measured at the site of maximal thickness and the ratio of noncompacted to compacted layer had to be more than 2:1 for diagnosis. The mean age at diagnosis was  $42 \pm 17$  years. The most common presenting symptom was shortness of breath (79%) and only 26% reported chest pain. Symptoms attributable to heart failure were present in over 50% of the patients [10].

Non-specific electrocardiographic abnormalities are seen in most patients, with 56% patients having bundle branch block in the study by Oechslin et al. [10] Atrial fibrillation was reported in 29% patients by Ritter et al. [11] Wolff-Parkinson-White syndrome has been described in up to 15 patients of pediatric patients with LVNC by Ichida et al. [12]

Diagnosis of NCLV can be made by 2-dimensional and color Doppler echocardiography. Echocardiogram shows multiple prominent trabeculations with deep intra trabecular recesses. Color Doppler can visualize blood flow through these deep recesses in continuity with the ventricular cavity [13]. Different echocardiographic criteria have been proposed for diagnosis of LVNC. However, the criteria proposed by Jenni et al. and Frischknecht et al. are more accurate and hence more commonly used [14,15]. The ratio of non-compacted myocardium to compact myocardium should be more than 2:1 at the end of systole per criteria proposed by Jenni et al. Cardiac magnetic resonance imaging provides more detailed description of the cardiac morphology in any imaging plane and is the modality of choice to confirm or rule out the diagnosis [14,16].

The main complications attributable to LVNC are thromboembolism, risk of cardiac arrhythmias and progressive heart failure. Standard guideline-based therapy is recommended for systolic and diastolic dysfunction, with implantable cardioverter defibrillator placement for those with Left Ventricular Ejection Fraction (LVEF) less than 35% despite optimal heart failure therapy. Some authors have advocated use of anticoagulation for patients with LVNC with low LVEF (<40%), but this suggestion is not guideline based yet. [17]. Prognosis is variable and depends upon degree and progression of heart failure, presence of thromboembolic complications and cardiac arrhythmias.

## Conclusion

LVNC is a rare condition, which is being increasingly diagnosed among immigrants from tropical regions, likely due to improved cardiac imaging techniques. It may be associated with other cardiac or extracardiac abnormalities. A need exists for future research to ascertain a definite etiology for LVNC and studying the effect of demographic factors, ethnicity and region of origin on the incidence of LVNC.

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