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# A rare case of non-compaction cardiomyopathy presented with acute coronary syndrome

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## Abstract

Non-Compaction syndrome is a rare disease. It should be considered as it has complications as Heart Failure, cardiomyopathy or arrhythmia. Echocardiography is the method of diagnosis and cardiac MRI to confirm. We are here presenting a case of a non-compaction syndrome associated with Acute Coronary Syndrome.

## Keywords

Non-compaction; cardiomyopathy; papillary muscle hypertrophy; echocardiography.

## Case Report

A 67-year-old man with PMH of HTN, HLD who presented to the ED with chest pain. He described his pain as substernal, non-radiating pain, 10/10 in severity, aggravated with inspiration that started 10 days before presentation. Initially, the pain was intermittent with exertion and progressively worsened. It was associated with dyspnea, diaphoresis, nausea and one episode of non-biliary non-bloody emesis. Dyspnea started 2 months ago on walking even 1 block, with gradual onset and progressive course. He is an ex-smoker, 20 PPD, non-alcoholic. Vitals were stable on admission, CBC and BMP were WNL, Troponin was >50, EKG showed STE in the anterolateral leads (Figure 1) Patient was evaluated by the cardiology team and Cath lab was activated. Cardiac cath was done and showed 80% mid LAD, distal LCX 60%, stenting the mid LAD with DUS. The patient then transferred to the CCU for observation and started on aspirin, ticagrelor, lisinopril, metoprolol, spironolactone.

The patient was symptoms free, Echocardiography was done the next day and showed EF of 30%, biventricular hypertrophy, filling defect likely hypertrophied papillary muscle, and filling defect within the wall. The patient was diagnosed with non-compaction syndrome (Figure 1, 2 & 3). The patient was discharged on the previous medications and the life vest as temporary pacing to protect from any arrhythmia.

The patient followed up in 3 months but has been non-compliant with medications and Echo repeated to show the same EF and the same findings, Entresto was added. In 6 months follow up, the patient reported improvement in physical activity without shortness of breath.

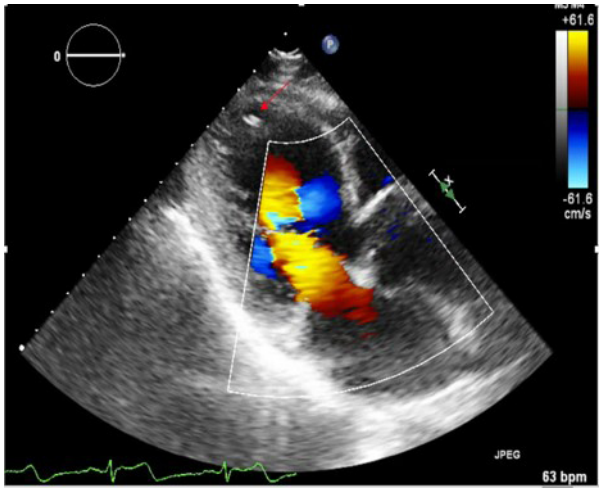


**Figure 1:** (red arrows): shows deep endocardial recess.

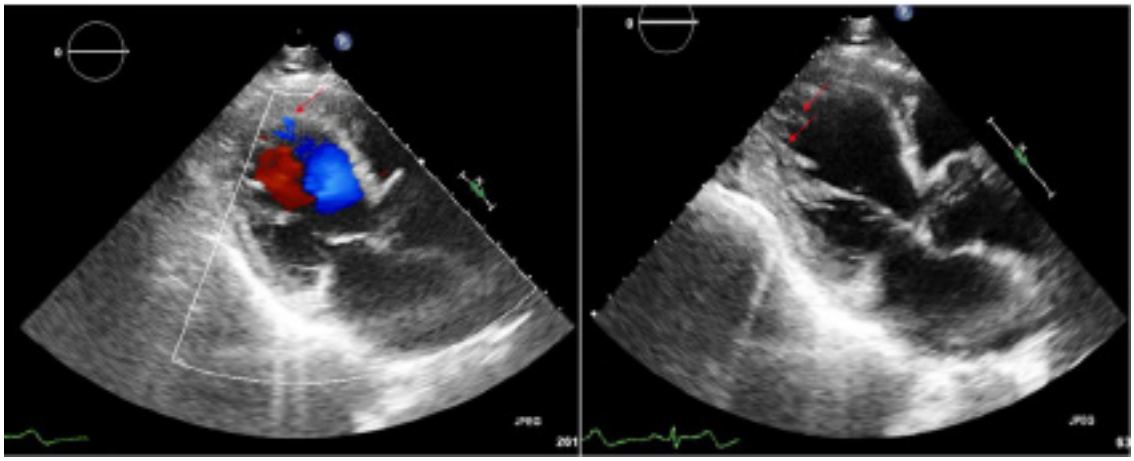

**Figure 2 & 3:** Red arrows

## Discussion

Non-Compaction Cardiomyopathy (NCCM) is an embryological defect of the heart, affecting the endocardial and myocardial layers of the left ventricle [1]. It is characterized by very compact thin epicardial and noncompaction of the endocardium which results in deep recess and prominent trabeculations that connect with the left ventricle cavity but not with the coronary circulation [2].

Some patients with this condition can be asymptomatic and the symptoms in others can vary depending on the position and location of non-compaction. Symptoms may include dyspnea, fatigue, dizziness/light-headed, syncope, palpitations and edema of the legs, ankles or feet [1].

In many cases, NCCM can be misdiagnosed as either hypertrophic or dilated cardiomyopathy due to non-specific symptoms. Isolated non-compaction cardiomyopathy can often present with ventricular tachycardia and thromboembolic events [3].

In our case, the patient presented with chest pain that was found to be acute coronary syndrome and treated accordingly. Non-compaction was found as accidental findings on echocardiography.

Though it is more commonly congenital, acquired forms of this defect were noted. Screening of first-degree relatives is recommended as it is correlated with a mutation in MYH7 or MYBPC3 gene with an autosomal dominant inheritance pattern [4].

There are really no universally agreed diagnostic criteria for this condition as it is extremely rare but two-dimensional echocardiography can be used to diagnose this condition [5].

Some criteria should be taken care of on diagnosis of the non-compaction syndrome: should be evaluated in the apical view, Differential diagnosis with thrombi, false tendons, apical hypertrophic cardiomyopathy, fibroma, obliterative processes, intramyocardial hematoma, cardiac metastases, and intramyocardial abscesses must be considered [6].

A cardiac MRI is an alternative and at times preferred diagnostic tool as it can definitively confirm or rule out left ventricular noncompaction because it allows for proper visualization of the apex, a harder task when using echocardiography [7]. Findings on cardiac MRI is described as a double-layered appearance on the 4-chamber view and marked trabeculations and intertrabecular recesses in the inferno-lateral segments on the short axis view [8].

The treatment regimen for NCCM can vary based on symptoms. Patients with a reduced ejection fraction should be placed on Beta-blockers, ACE inhibitors, Diuretics to manage the systolic and diastolic dysfunction [9].

Patients with end-stage or refractory heart failure may require a transplant. Some with NCCM are at risk for SCD and arrhythmias ergo annual Holter monitoring, EP studies, and anti-arrhythmic therapies may be warranted. ACID placement and bi-ventricular pacemakers can also help reduce the risk of SCD due to NCCM [10].

## Conclusion

Non-compaction cardiomyopathy is a diagnosis of interest. Although there is no specific treatment for it, the associated condition should be treated. Non-compaction cardiomyopathy might be associated with acute coronary syndrome, heart failure or arrhythmia. Treatment should be directed to the associated condition. Some authors recommended treatment as a treatment for cardiomyopathy.

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