# Triple Valvular Crisis and Fulminant Heart Failure in Noncompaction Biventricular Cardiomyopathy

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## Introduction

A rare case of biventricular noncompaction and triple valvular insufficiency in a 59-year-old male with decompensated biventricular heart failure requiring triple valve surgery and aggressive diuresis supported with milrinone.

## Case

- 59-year-old Costa Rican Male

- **Past medical history:**
  - Heart failure with preserved ejection fraction, NYHA II
  - COPD
  - Current tobacco use
  - Prior cocaine abuse
  - Hypertension
  - Hyperlipidemia
  - Poor social support

- **Chief Complaint:**
  - Progressively dyspnea with presentation consistent with fulminant decompensated heart failure

- **Physical Exam:**
  - 3+ Pitting pedal edema to the knees
  - Audible holosystolic murmur  at apex
  - Tachycardiac
  - Bilateral rales

- **Labs:**
  - H&H 14.4/43.8, WBC 9.8
  - BUN 42 Cr 1.01
  - Bilirubin 2.3 d(1.0) , AST 85 ALT 108
  - Pro-BNP 13,264
  - High sensitivity troponins 22
  - Negative blood cultures and infectious work up

## What is Noncompaction?

Ventricular noncompaction is an anatomical variant descripting the intertrabeculation of the thin inner layer of the left ventricles but may also involve the right ventricle as either biventricular or isolated noncompaction. Its presence does not indicate cardiomyopathy as it can be observed in otherwise healthy individuals. In rare forms when associated with LV dilation and dysfunction it is classified as a form of genetic cardiomyopathy. Due to limited understanding of the etiology, most cases display late presentation of advanced heart failure and arrythmias. There tend to be a strong family history with an autosomal dominant pattern of inheritance, in up to half the cases. Additionally, the gene mutations affected are also notably implicated in hypertrophic and dilated cardiomyopathy. Several genetic mutations have been noted including LDB3, 1q21.1 deletion syndrome, TTN, RBM20 and LMNA. These defects may result in an arrest of endomyocardial morphogenesis with the presence of ‘sinusoids’ within the ventricles. Based on reports, the left ventricle is most commonly involved. The noted prevalence of noncompaction amongst heart failure patients is estimated at 3-4%. Clinical presentation varies tremendously and can range from asymptomatic to fulminant congestive heart failure, LV dysfunction, ventricular tachycardia, sudden cardiac death, and thromboembolic complications. These complications carry a higher prevalence in females and are associated with more catastrophic outcomes including cardiac arrest and ischemic strokes.

## Hospital Course and Beyond

- **Medication and Therapy**:
  - Aggressive diuresis
  - IV inotrope to maintain diuresis and cardiac output

- **Imaging**:

  - **Transthoracic echocardiogram:** Presence of moderately reduced biventricular systolic function, LVEF 35% and prominent LV trabeculation raising suspicion for noncompaction cardiomyopathy. Several valvular abnormalities were also noted

  - **Transesophageal echocardiogram:** Congenital bicuspid aortic valve with severe aortic insufficiency, posterior leaflet flail with anteriorly directed severe mitral regurgitation, and moderate tricuspid regurgitation

  - **Left heart catheterization:** normal coronary arteries by angiography.

  - **Right heart catheterization:** RA 16 mmHg, RV 29/0 mmHg, PA 70/34 (46) mmHg, PCWP 40 mmHg, likely falsely elevated due to giant V waves, PA sat 46% and Ao Sat 94% on room air and a thermodilution CO of 3.63 L/min with a CI of 2.19 L/min/m2

  - **Cardiac MRI:** Did not illustrate any disease specific enhancement on late gadolinium imaging. It further confirmed the TEE findings and noted a ratio of non-compacted to compacted myocardium ranges from 2.3 - 3.0

- **Procedures:**
  - St.Jude Epic 9 mm valve in the mitral position
  - St.Jude Epic 23 mm valve in the aortic position
  - 30 mm Triad ring in the tricuspid position

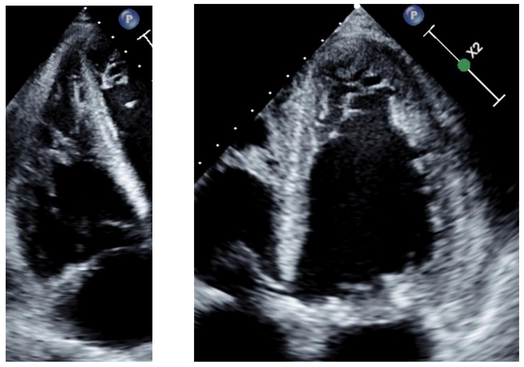

**Image 1.** TTE illustrating  LV (right) and RV (left) with trabeculation in the Apical 4 and RV focus imaging

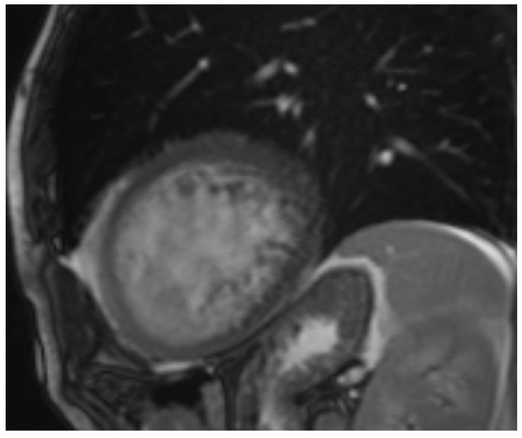

**Image 2.** Cardiac MRI, further illustrating the intertrabeculation of the ventricular

## Conclusion

It is important to maintain a high index of suspicion for early diagnosis and aggressive treatment of patient with noncompaction cardiomyopathy to minimize the associated complications, as delay in therapy can be fatal. As illustrated, delayed diagnosis can result in late presentation and complex interventions with potential severe complications.

## References

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