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Case Study

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LEFT VENTRICULAR NONCOMPACTION CARDIOMYOPATHY IN AN INFANT - RARE CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Noncompaction cardiomyopathy (NCCM), also termed left ventricular noncompaction (LVNC), is an uncommon form of cardiomyopathy that arises from an arrest in normal endomyocardial embryonic development. We describe a 2-month-old infant presenting with heart failure who demonstrated classic echocardiographic findings of LV NCCM, including a markedly dilated left ventricle with excessively prominent trabeculations and deep intertrabecular recesses.

KEYWORDS: Left ventricular noncompaction, Cardiomyopathy, Heart failure, Intercostal retractions, Noncompaction cardiomyopathy.

INTRODUCTION

In healthy children and adults, the left ventricle (LV) typically contains no more than three prominent trabeculations and is therefore less trabeculated than the right ventricle [1, 2]. On rare occasions, however, the presence of more than three prominent trabeculations—referred to as LV noncompaction of the ventricular myocardium—may be identified at autopsy (Figures 1–3) or detected through various imaging modalities, including echocardiography and MRI.

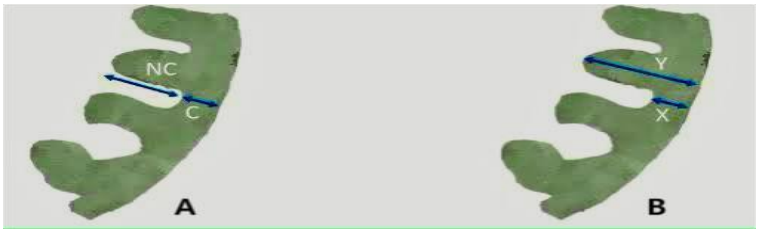


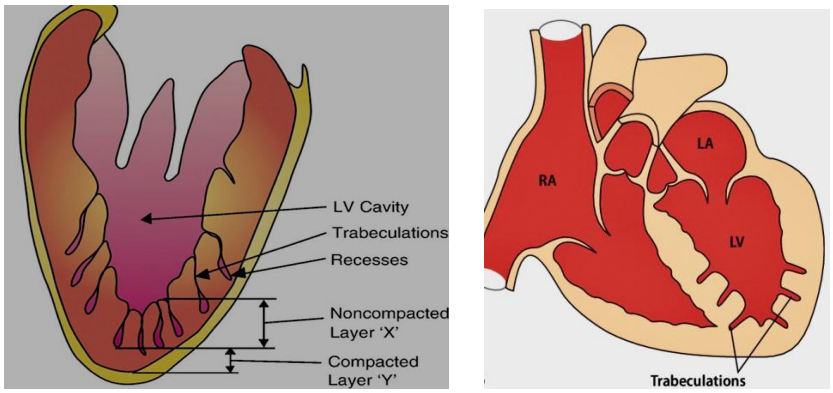
Figure 1: Schematic depiction of LV noncompaction.

(A) Jenni et al. criteria[3]

1. **Presence of a bilayered myocardial structure at end-systole, consisting of a thin compacted (C) layer and a markedly thickened non-compacted (NC) layer.**
2. **An NC/C ratio exceeding 2.**
3. **Color Doppler demonstration of blood flow entering the intertrabecular recesses from the ventricular cavity.**
4. **Absence of associated structural cardiac abnormalities.**

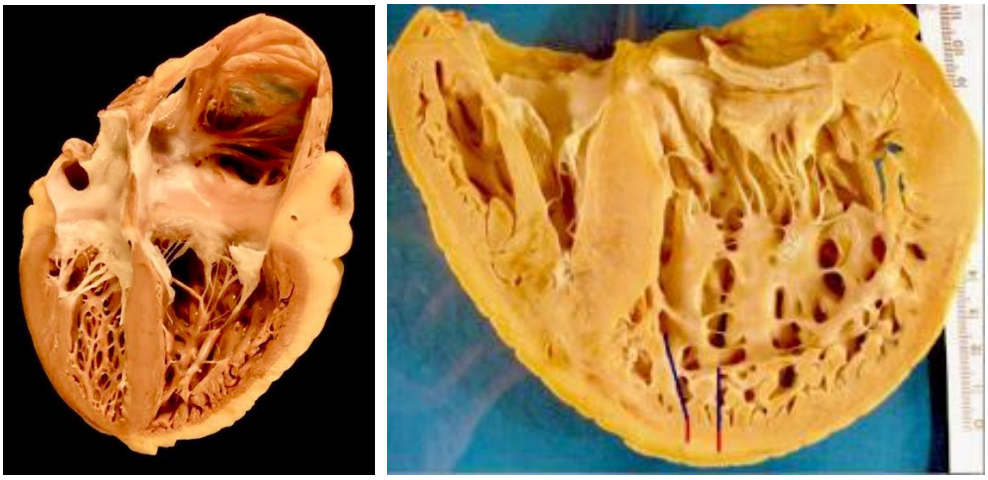
(B) Chin et al. criteria[4]

1. **Measurement of the distance from the epicardial surface to the trough of the trabecular recess (X) and from the epicardial surface to the peak of the trabeculation (Y) at end-diastole.**
2. **An X/Y ratio of ≤ 0.5.**
3. **Emphasis on trabeculations localized at the LV apex.**



(A) **(B)**

Figure 2: Graphic representation of the morphopathological features of left ventricular noncompaction (LVNC). (A) Artistic illustration of LVNC depicting the characteristic two-layered myocardial structure and markedly increased trabeculations near the left ventricular apex. A non-compacted to compacted myocardial thickness ratio (X/Y) > 2 is considered indicative of LVNC. (B) Additional schematic illustration highlighting the typical appearance of left ventricular noncompaction.



(A) **(B)**

Figure 3: Pathological specimens demonstrating (A) a normal heart and (B) left ventricular non-compaction cardiomyopathy. In the LVNC specimen, prominent trabeculations (blue line) and deep intertrabecular recesses (red line) extend throughout the entire left ventricular apex.

NCCM has recently been classified as a Genetic Cardiomyopathy in the 2006 cardiomyopathy classification system.[5] It arises from an abnormality in endomyocardial morphogenesis, leading to incomplete compaction of the trabeculated embryonic myocardium during development.[6] In adults, one or more left ventricular segments—most commonly the apical, mid-lateral, and mid-inferior regions—and occasionally both ventricles, exhibit numerous sinusoids or trabeculations that are excessively numerous, unusually prominent, and associated with deep intratrabecular recesses. These recesses are lined by endothelium that maintains continuity with the ventricular endocardium.

A variety of imaging modalities have been employed to describe, characterize, and diagnose LVNC, including magnetic resonance imaging, two-dimensional echocardiography (2DE), contrast-enhanced 2DE, and angiography.[7, 8] Among these, 2DE remains the most widely used diagnostic tool. Based on echocardiographic data, the estimated prevalence of LVNC in the general population is approximately 0.05%.[9]

Diagnosis

Diagnostic criteria

Three echocardiographic criteria sets are currently used for the diagnosis of LVNC:

(i) the Jenni criteria, which emphasize the identification of a two-layered myocardial structure;

(ii) the Chin criteria, which evaluate the ratio of trabecular recess depth to trabecular height[3, 4]; and

(iii) the Stollberger criteria, which highlight the number of trabeculations and the ratio between non-compacted and compacted myocardial layers.[10]

A comparative summary of these three criteria sets is provided in Table 1.

Table 1: Echocardiographic-based Left Ventricular Non-Compaction diagnostic criteria.

Chin Criteria (1990)[4]	Jenni Criteria (2001)[3]	Stollberger (2002)[10]
Absence of any other coexisting cardiac structural abnormality	Absence of any other coexisting cardiac structural abnormality	Absence of any other coexisting cardiac structural abnormality
Numerous. excessively prominent trabeculations and deep intertrabecular recesses	Numerous. excessively prominent trabeculations and deep intertrabecular recesses	> 3trabeculation protruding from LV wall apically to papillary muscle in 1 imaging plane
Views: parasternal long axis, subxyphoid, and apical	Views: parasternal short axis, and apical	Views: non standard view
Focus on depth of recesses	Focus on a 2-layer structure	Focus on a 2-layer structure
Measured in end-diastole	Measured in end-systole	Measured in end-systole
Ratio of distance from the epicardial surface to the trough of the trabecular recesses and distance from the epicardial surface to peak of trabeculation ≤ 0.5	Ratio of thick noncompacted layer to thin compacted $\geq$ 2 Perfused intertrabecular recesses supplied by intraventricular blood on color Doppler analysis	Ratio of thick noncompacted layer to thin compacted ≥ 2

Modified Rotterdam Criteria of Noncompaction Cardiomyopathy

Despite their widespread use, most existing diagnostic criteria show limited validation against true pathoanatomic correlates.[3, 4, 11] Notably, nearly one-quarter of patients with heart failure meet one or more of these echocardiographic criteria, indicating that current definitions may lack sufficient specificity for distinguishing non-compaction cardiomyopathy (NCCM) from other conditions.

To address these limitations, Soliman and colleagues proposed the Rotterdam criteria, which integrate traditional trabeculation-based parameters (i.e., the Jenni criteria) with septal thickness assessment (Figure 4).[12] This framework improves differentiation between definite NCCM and physiological hypertrabeculation patterns commonly observed in athletes, individuals of African descent, and patients with chronic hypertension.[12]

In addition, recognizing asymptomatic individuals who exhibit normal ECG findings, preserved cardiac function, and unremarkable genetic profiles is essential, as these cases do not constitute disease in the strict sense and misclassification may pose substantial

psychological, social, legal, and insurance implications. Importantly, the presence of concomitant congenital heart disease (CHD) or neuromuscular disease (NMD) should not be considered exclusionary when applying these criteria.

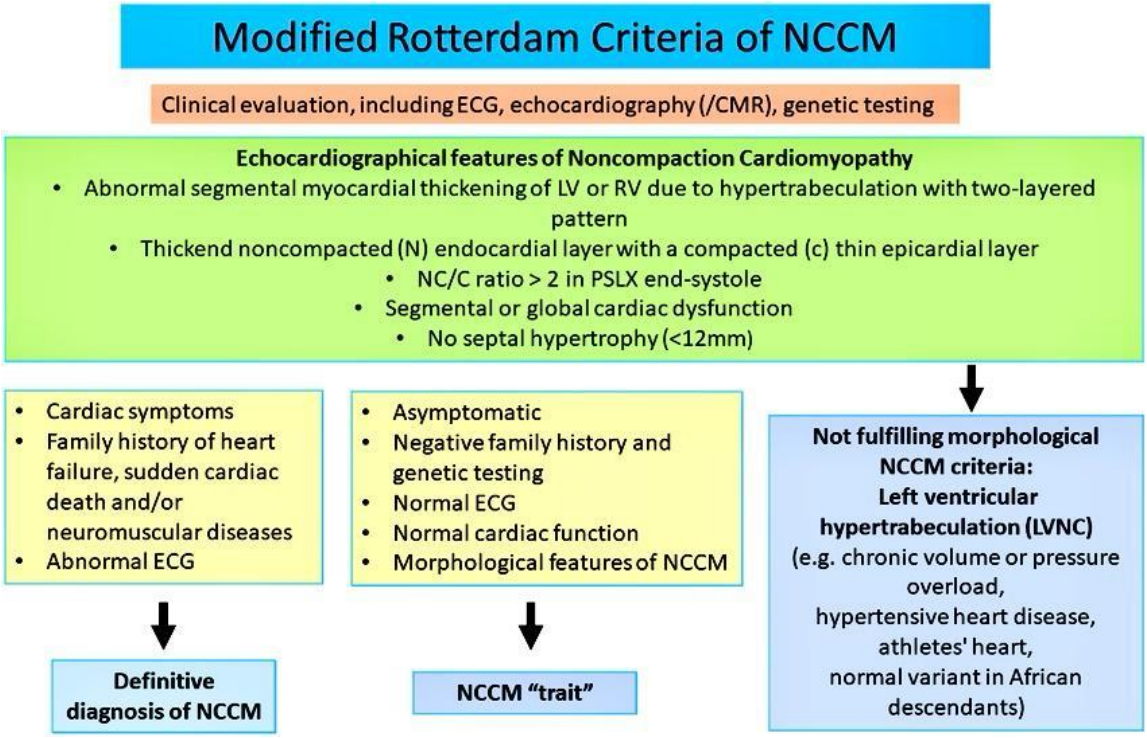


Figure 4: Diagnosing NCCM according to the Rotterdam criteria.[12] LVNC left ventricular noncompaction, NCCM noncompaction cardiomyopathy, NMD neuromuscular disease, PSLX parasternal long axis view.

CASE REPORT

A two-month-old male infant was referred to our center for a comprehensive evaluation for suspected congenital heart disease. According to the parents, he had experienced severe breathlessness since birth.

On presentation, the infant had an average build but showed facial puffiness, marked respiratory distress, and profuse sweating over the forehead (Figure 5). Prominent intercostal retractions were observed along with significant sternal indrawing. His anthropometric and vital parameters were as follows: weight 3.9 kg, height 56 cm, blood pressure 78/40 mmHg, heart rate 147/min, respiratory rate 88/min, and SpO_2 99% on room air. Cardiovascular examination revealed a grade 2/6 systolic murmur, best appreciated in the left infraclavicular region. The second heart sound was notably loud with a closely split component. The remainder of the systemic examination was unremarkable.

Figure 5: Facial appearance of our index patient. Marked breathlessness, sweating over forehead and puffiness of face was recognized.

The chest X-ray in the posteroanterior (PA) view (Figure 6) demonstrated marked cardiomegaly accompanied by pulmonary edema.

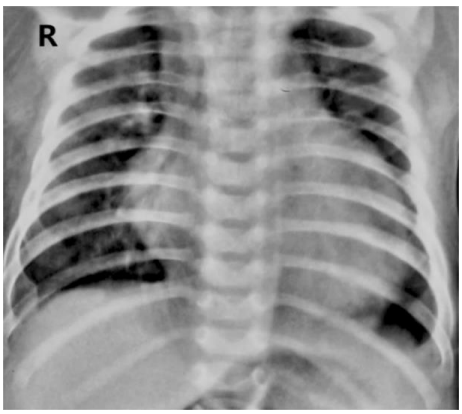


Figure 6: X-ray chest (PA view). Gross cardiomegaly with pulmonary edema was discerned.

The resting ECG (Figure 7) demonstrated markedly increased precordial QRS voltages.

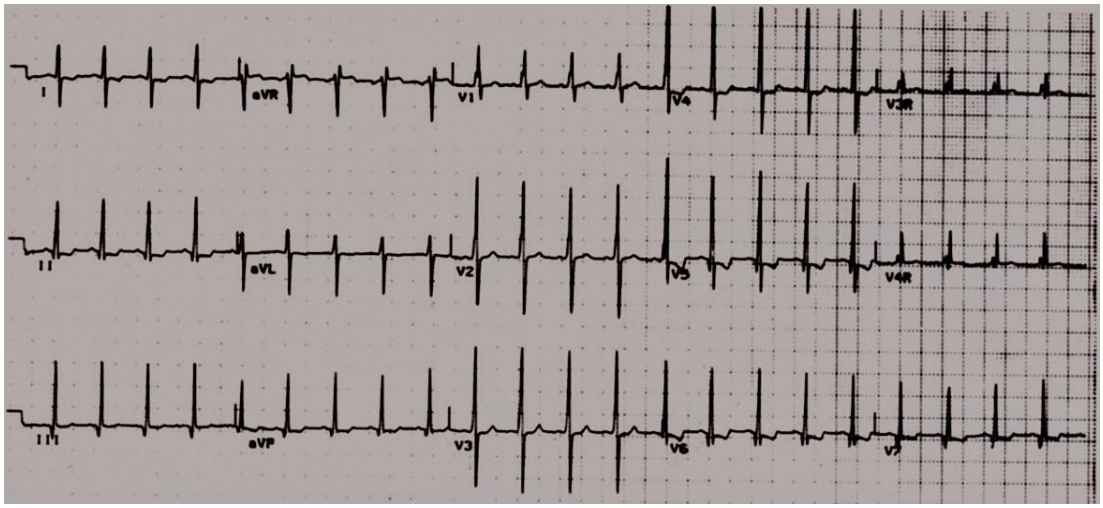


Figure 7: Resting ECG. Note the prominent precordial voltage (1/4 standard).

Transthoracic Echocardiography

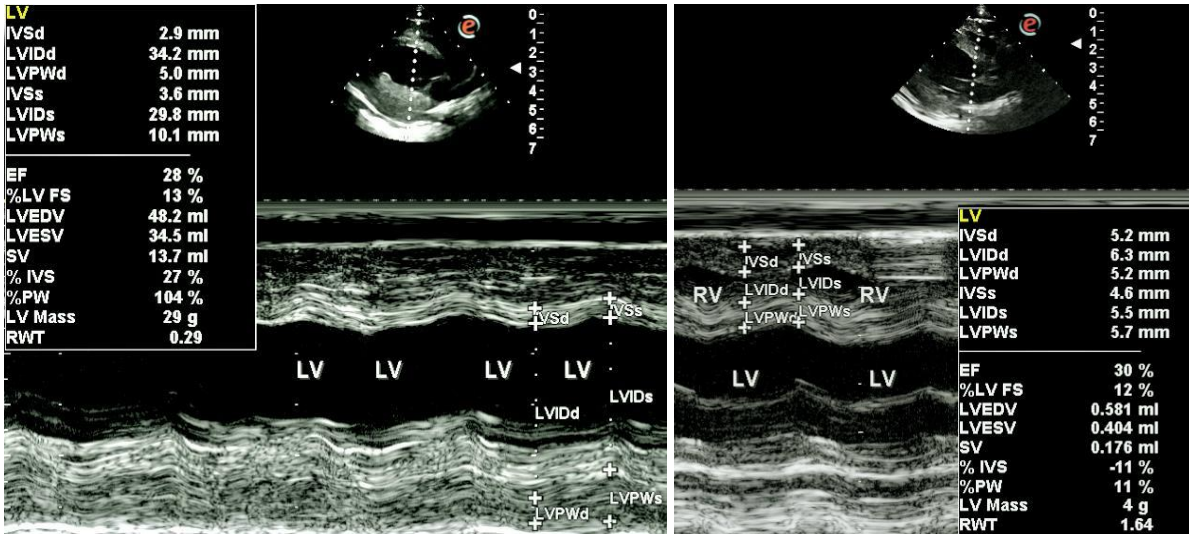
Transthoracic echocardiographic assessment was performed using the MyLab X7 4D XStrain system (Esaote, Italy) equipped with a pediatric probe. A sequential segmental evaluation was conducted utilizing standard imaging windows, including the subcostal, parasternal long-axis (LX), parasternal short-axis (SX), apical four-chamber (4CH), apical five-chamber (5CH), and suprasternal views.

M-mode Echocardiography

M-mode echocardiography of both the left and right ventricles was obtained, and the corresponding measurements are summarized in Table 2 and Figure 8.

Table 2: Calculations of M-mode echocardiography.

Measurements	LV	RV
IVS d	2.9 mm	5.2 mm
ID d	34.2 mm	6.3 mm
PW d	5.0 mm	5.2 mm
IVS s	3.6 mm	4.6 mm
ID s	29.8 mm	5.5 mm
PW s	10.1 mm	5.7 mm
EF	28 %	30 %
% FS	13 %	12 %
EDV	48.2 ml	0.581 ml
ESV	34.5 ml	0.404 ml
SV	13.7 ml	0.176 ml
Mass	29 g	4 g
IVS, interventricular septum, ID, internal dimension; PW, posterior wall, d, diastole; s, systole; FS, fractional shortening; EDV, end-diastolic volume; ESV, end systolic volume; SV, stroke volume; EF, ejection fraction.		



(A) **(B)**

Figure 8: M- mode estimation of LV and RV indices - demarcated marked dilatation of LV, slit like RV with severely reduced bivenricular systolic function. LVEF & RVEF was 28 % & 30 %, respectively and LV mass & RV mass were 29 g & 4 g, respectively.

Summary of M-mode echocardiography

The M-mode assessment demonstrated a dilated left ventricle with a slit-like right ventricle and markedly impaired biventricular systolic function. The LVEF and RVEF were 28% and 30%, respectively, while the LV and RV masses measured 29 g and 4 g, respectively.

2-Dimensional-Transthoracic Echocardiography

Transthoracic echocardiography (TTE) was systemically performed by the sequential segmental approach (SSA) and the echocardiographic characteristics which were documented are enumerated below (Figure 9-15):

- Levocardia
- Situs solitus
- Atrioventricular concordance
- Ventriculo-arterial concordance
- Concordant d-bulboventricular loop
- Normally related great arteries
- Left aortic arch
- Confluent pulmonary arteries
- Normal systemic and pulmonary venous drainage.
- Dilated LV with global hypokinesia.

- Conspicuous dyssynchronised motion of LV.
- The LV posterior and lateral wall is distinctly divided into compacted and non compacted layers. The compacted portion is also significantly thickened.
- Striking, multiple trabeculations and deep intertrabecular recesses were recognized on the posterior and lateral wall of LV.
- Mitral regurgitation (trace)
- Dilated LV, slit like RV.
- Biventricular concentric hypertrophy with severely reduced systolic functions.
  - LVEF and RVEF were 28 % and 30 %, respectively.
- There was no evidence of ASD, VSD, PDA, COA, , AS, PS.

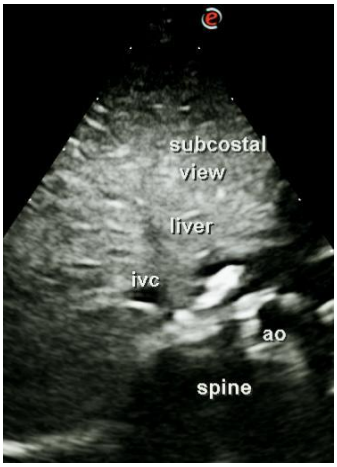


Figure 9: Situs solitus. In the subcostal view liver and inferior vena cava were right sided and aorta and spleen were left sided. ivc, inferior vena cava; ao, aorta.

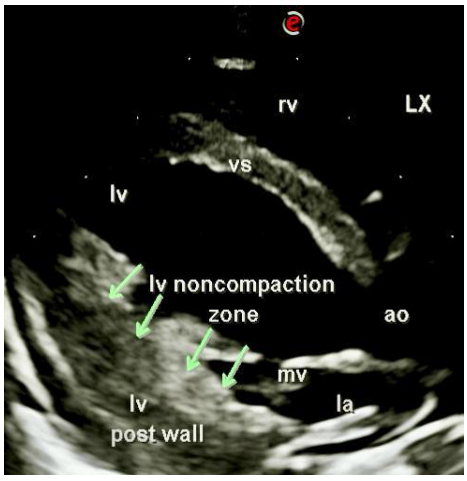


Figure 10: Transthoracic Echocardiography (TTE). In the LX view a salient LV non compacted zone of post wall is demonstrated. lv, left ventricle; la, left atrium; mv, mitral valve; ao, aorta; vs, ventricular septum; rv, right ventricle.

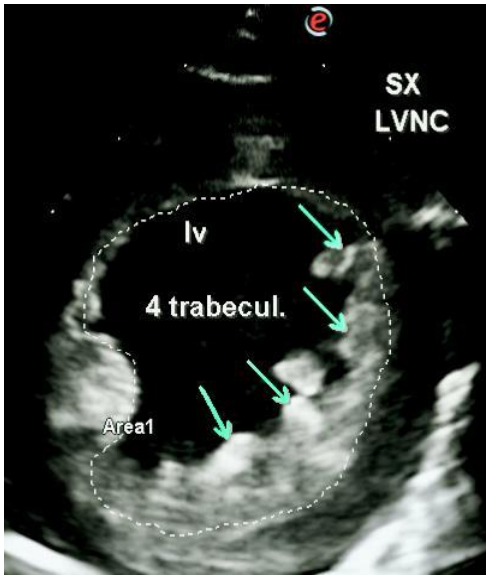
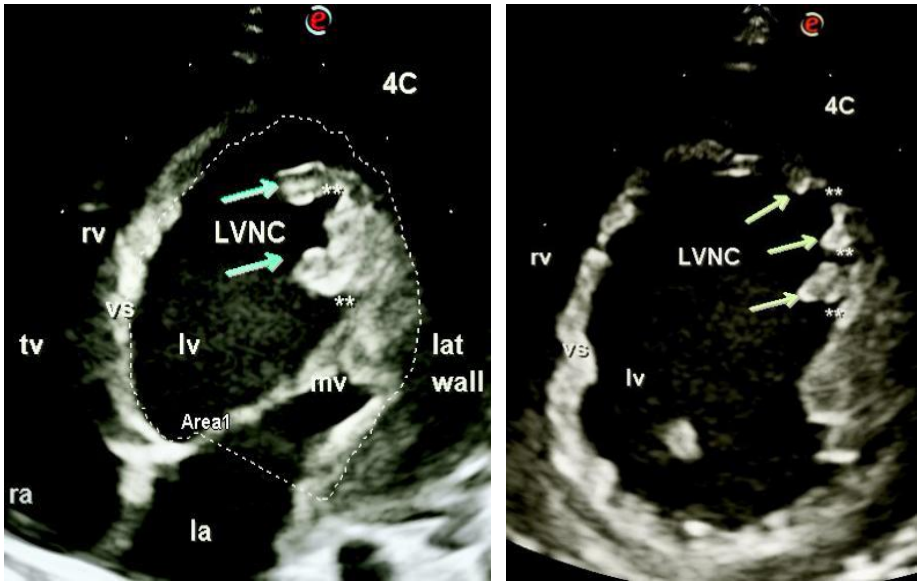


Figure 11: TTE in SX view depicted a large area of non compaction in the lateral wall of LV. At least 4 prominent trabeculations were delineated. LVNC, left ventricular non compaction; lv, left ventricle; trabecul, trabeculations.



(A) (B)

Figure 12: TTE in 4C view. Two images of apical 4C view are shown to highlight the presence of lv non compaction. Distinctive trabeculations and recesses were illustrated. LVNC, lv non compaction; lv, left ventricle; vs, ventricular septum; ** depict recesses; mv, mitral valve; la, left atrium; ra, right atrium.

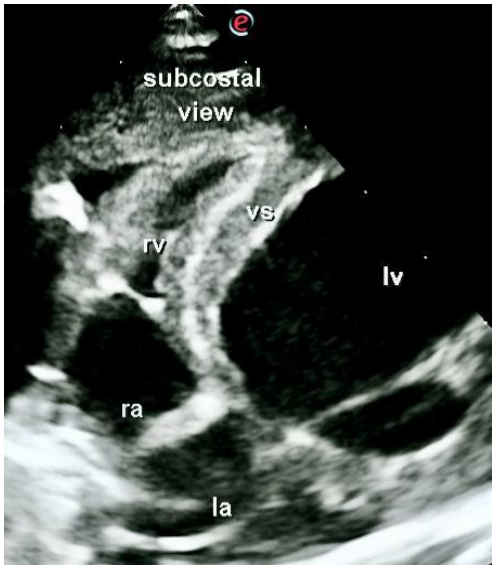
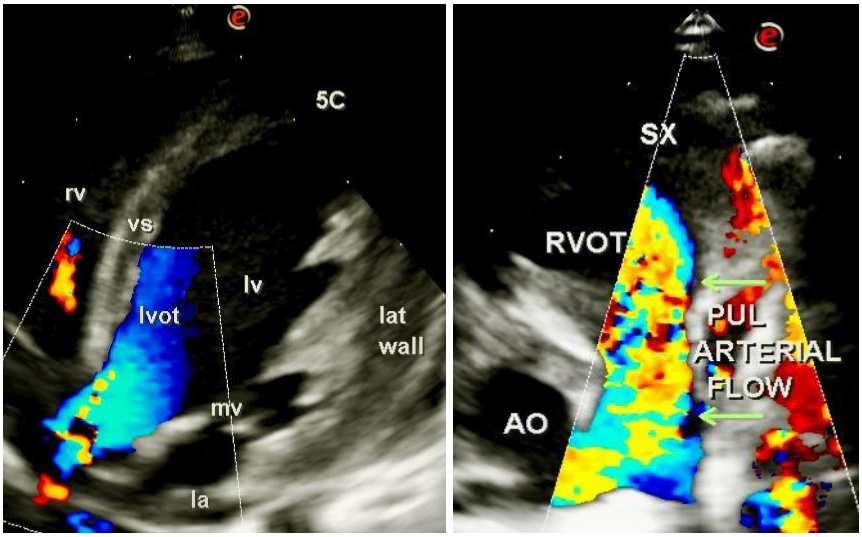
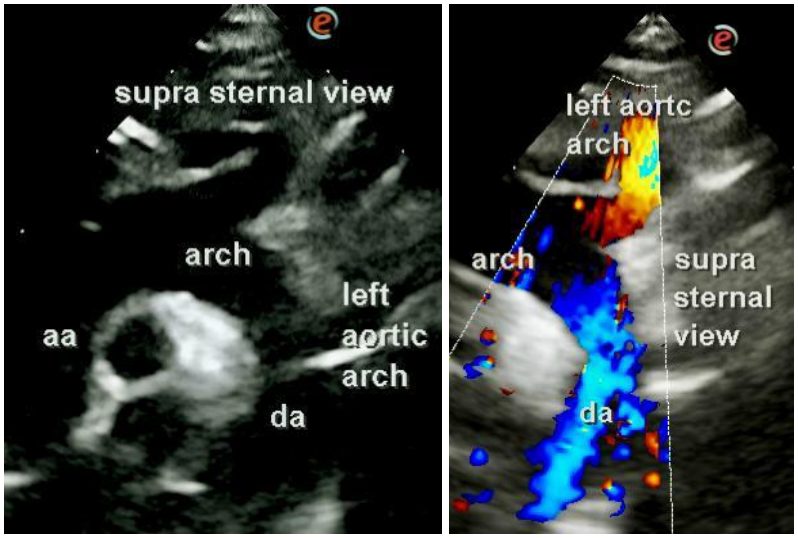


Figure 13: In the subcostal view a considerably dilated LV with a slit like RV was portrayed. rv, right ventricle; vs, ventricular septum; lv, left ventricle; la, left atrium; ra, right atrium.



(A) (B)

Figure 14: Color flow mapping (CFM) across left and right outflow tracts. The CFM was normal across both outflow tracts without any obstruction. lvot, left ventricular outflow tract; vs, ventricular septum; lv, left ventricle; mv, mitral valve; la, left atrium; RVOT, right ventricular outflow tract; AO, aorta.



(A) (B)

Figure 15: Suprasternal view depicting left aortic arch. (A) without and (B) with color flow mapping. aa, ascending aorta; da, descending aorta.

Summary of Color flow doppler echocardiography

Color flow Doppler imaging revealed pronounced regions of noncompaction involving the posterior and lateral walls of the left ventricle, accompanied by marked LV dilation and significant dyssynchronous motion, resulting in a severely reduced LVEF of 28%. Biventricular concentric hypertrophy was also evident. In addition, although the right ventricle appeared slit-like, the RVEF was markedly diminished at 30%.

DISSCUSION

LVNC is a heterogeneous form of cardiomyopathy characterized by multiple prominent trabeculations and deep intertrabecular recesses. It is a relatively new and uncommon clinical entity, formally recognized as a distinct cardiomyopathy only since 2006.[5] Since its initial description by Engberding et al. in 1984[13], the condition has appeared in the literature under several names, including spongy myocardium, NCCM, and left ventricular hypertrabeculation (LVHT).[3, 14]

Although considered rare, LVNC represents the third most common type of cardiomyopathy (CMP) in children, following dilated cardiomyopathy (DCM) and hypertrophic cardiomyopathy (HCM).[15] LVNC may lead to heart failure and is associated with thromboembolic complications and potentially fatal arrhythmias in adults.[16] It may occur as either an isolated or non-isolated phenotype, with the latter frequently accompanied by

congenital heart diseases (CHDs), features overlapping other cardiomyopathies, and/or neuromuscular disorders (NMDs).[17, 18]

The pathophysiology of LVNC remains incompletely understood, and several hypotheses have been proposed to explain the development of excessive trabeculations. One commonly cited theory suggests that LVNC results from impaired embryologic compaction of the myocardium, producing a hypertrabeculated, honeycomb-like ventricular structure.[19, 20] However, this theory does not fully account for cases first diagnosed in adulthood. Numerous genes have been implicated in LVNC, yet none of the suggested pathogenic variants or chromosomal abnormalities have been definitively linked to a disrupted fetal compaction process. Moreover, many of these (likely) pathogenic variants are known to produce diverse phenotypes.[21]

Epidemiology

The precise incidence of LVNC in children remains uncertain, largely due to its relatively recent classification as a separate cardiomyopathy. In addition, the absence of universally accepted diagnostic criteria contributes to diagnostic delays and the possibility of misclassifying LVNC as other cardiomyopathies, such as DCM.[12] Distinguishing LVNC from DCM, or from LVNC with DCM-like features, is essential because management strategies, prognosis, and familial recurrence rates may differ significantly.

Current estimates suggest an incidence of approximately 0.12 per 100,000 in children up to ten years of age and ≤ 0.81 per 100,000 in infants under one year.[12] Thus, LVNC is considered the third most common cardiomyopathy in pediatric populations.[22] Similar rates have been reported in smaller pediatric cohorts. An Australian study identified LVNC in 9.2% of all children under ten diagnosed with a primary cardiomyopathy between 1987 and 1996.[15] Additionally, data from the Pediatric Cardiomyopathy Registry—which includes 98 centers across the United States and Canada—indicated that 4.8% of children with any form of cardiomyopathy had isolated LVNC over an 18-year period.[23] Despite these findings, standardized diagnostic criteria and international multicenter registries remain essential for determining the true incidence and prevalence of LVNC.

Pathogenesis

During normal embryologic development, the myocardium initially consists of a spongy network of muscle fibers and trabeculations separated by deep recesses (Figure 16).

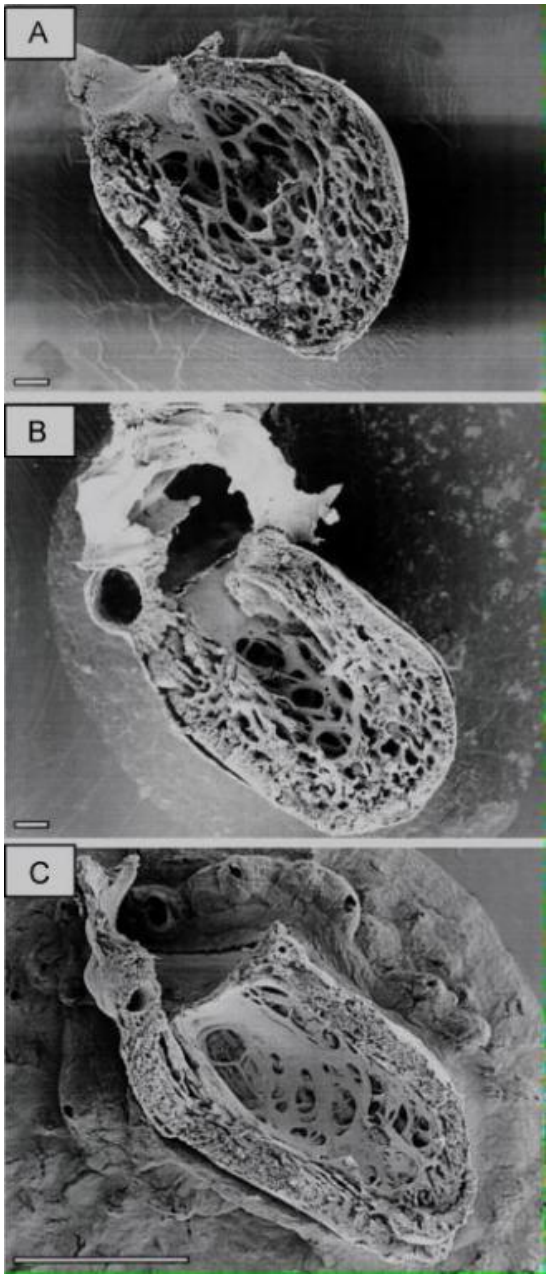


Figure 16: Parietal views of sagittally dissected human embryonic left ventricles illustrating the normal process of trabecular compaction. (A) At 6 weeks, the ventricular wall exhibits abundant fine trabeculations. (B) By 12 weeks, these trabeculations begin to solidify at their basal regions, contributing to progressive thickening of the compact myocardial layer. (C) In the early fetal period, completion of this compaction process results in the compact layer constituting the majority of the myocardial mass.

Before the coronary vasculature develops, the intertrabecular recesses, or sinusoids, communicate directly with the ventricular cavity and serve as the primary source of myocardial perfusion. Once the coronary circulation forms, the ventricular myocardium

gradually undergoes compaction, during which the larger intertrabecular recesses are transformed into capillaries. Trabecular compaction typically occurs between 12 and 18 weeks of gestation, beginning at the base of the heart and progressing toward the apex.[24, 25] For reasons that remain unclear, this transition fails to occur in patients with LVNC, resulting in a markedly thickened, non-compacted endomyocardial layer composed of prominent trabeculations that remain in continuity with the LV cavity, exhibit deep recesses, and lack communication with the epicardial coronary circulation. A thin compacted epicardial layer is also characteristic of this condition.[26, 27]

Histologically, LVNC does not exhibit pathognomonic features, nor are there consistent differences between isolated forms (without associated congenital anomalies) and non-isolated forms. However, interstitial fibrosis and endocardial fibroelastosis have been documented in endomyocardial biopsies.[28] Additionally, autopsy studies reveal that the absence of well-formed papillary muscles is strongly associated with LVNC.[29]

LVNC may also occur in familial forms. In one study, 6 of 34 patients (18%) had a positive family history of LVNC.[11] Although no single genetic mutation consistently produces the LVNC phenotype, variants in several genes—including Z-band alternatively spliced PDZ-motif protein (ZASP), α-dystrobrevin (DTNA), tafazzin (TAZ-G4.5), and sarcomeric protein–encoding genes—have been implicated.[30] More recently, mutations in hyperpolarization-activated cyclic nucleotide–gated channel 4 (HCN4) have been reported in families with sinus node dysfunction and LVNC.[31, 32] Consequently, some asymptomatic individuals have been diagnosed through cascade screening of relatives.

Natural History and Prognosis

Outcomes in pediatric LVNC vary considerably. An Australian cohort reported that within 10 years of diagnosis of dilated-phenotype LVNC, half of the children died or underwent heart transplantation (HTx), and only 20% were alive with normal cardiac function at 15-year follow-up.[15] A Toronto study reported a 72% three-year transplant-free survival rate among pediatric patients with LVNC, 77.3% of whom exhibited ventricular dilatation.[33]

Differences in prognosis likely reflect variations in cohort composition. Pediatric patients with mixed cardiomyopathy phenotypes carry a significantly increased risk of death or HTx (hazard ratio 6.35) compared with those without a mixed phenotype.[23] Left ventricular dilatation has also been identified as an independent predictor of adverse outcome

(HTx/death).[34] Similarly, five-year transplant-free survival is lower in children with LVNC and concomitant cardiomyopathy compared with those with isolated LVNC.[28] Notably, among children with dilated LVNC, time to death or HTx does not differ significantly from those with isolated DCM (p = 0.22). Five-year composite endpoints occurred in 37% of patients with dilated LVNC compared with 47% of those with DCM alone.[23]

Interestingly, children with LVNC and Barth syndrome demonstrated a trend toward better 15-year survival compared with those with LVNC alone (71% vs 36%, p = 0.08), likely owing to routine family screening and earlier diagnosis.[15]

Overall, presentation within the first year of life is associated with markedly increased mortality (HR 2.1, p = 0.02) [23, 35], with the majority of deaths (90%) occurring in the first year after diagnosis. These findings underscore the need for internationally standardized diagnostic criteria to facilitate early identification of LVNC.

Management

No disease-specific medical or surgical therapy has yet been established for LVNC. Standard heart failure therapy—including beta-blockers, angiotensin-converting enzyme inhibitors, and/or angiotensin II receptor blockers—may promote beneficial LV remodeling.[36] Data regarding the use of newer therapies such as sacubitril/valsartan in children with LVNC are currently lacking. Patients should be closely monitored for clinical deterioration and arrhythmias, with rhythm disturbances managed according to established protocols. The role and efficacy of implantable cardioverter-defibrillators (ICDs) in pediatric LVNC remain to be clarified. Further investigation is needed to determine whether preventive anticoagulation or antiplatelet therapy is warranted.

For children with end-stage heart failure secondary to LVNC who do not respond to maximal medical therapy, HTx remains the only definitive option. Approximately 4% of pediatric patients listed for HTx in North America have isolated LVNC. Experience with mechanical circulatory support in this population is limited and largely restricted to case reports.[37–39]

CONCLUSION

Childhood LVNC is an uncommon but increasingly recognized disorder characterized by heterogeneous phenotypes, diverse clinical presentations, and potentially life-threatening complications. Clinical manifestations range from asymptomatic status to severe heart failure

and sudden cardiac arrest. Improved awareness and operator training in echocardiography and CMR can reduce rates of missed diagnosis. This case highlights the importance of early recognition of LVNC in infants—a rare but serious condition requiring timely evaluation. Our patient exhibited a left ventricular ejection fraction of 28%, underscoring the need for prompt echocardiographic assessment. Because LVNC may be asymptomatic, careful evaluation of infants—particularly those with cardiomegaly or a positive family history—may allow earlier diagnosis and intervention. Although current therapeutic options are limited, individualized management can still improve outcomes. Further research is essential to identify safe and effective treatment strategies for LVNC in infancy.

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