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Case Report

# Biventricular noncompaction induced heart failure in premature newborn ☆

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ABSTRACT

Deep intertrabecular recesses and overly pronounced trabeculations in one ventricle are the hallmarks of noncompaction cardiomyopathy (NCCM), a rare congenital cardiomyopathy but very rarely right ventricle (RV), or both ventricles may be involved. We reported a 5-day-old preterm newborn with signs of congestive heart failure that the transthoracic echocardiography (TTE) revealed deep intertrabecular recesses perfused from the left ventricle (LV) and RV cavity, as well as significantly increased wall thickness of the right ventricles and hypertrabeculations in the apical and midventricular segments.



## Introduction

The interruption of myocardial fiber compaction during embryogenesis is the cause of noncompaction of the left ventricular myocardium. Although it can also affect the right ventricle, it is typically seen in the left ventricle [1]. The condition is identified using 2-dimensional echocardiography or magnetic resonance imaging, which reveals deep intertrabecular recesses with apparent flow as well as large, prominent trabeculations. Additionally, the diagnosis depends on the 2-layered wall structure, composed of the thick, noncompacted endocardium and the thin, compacted epicardium [2]. It has been associated with a single point mutation in the beta-myosin heavy chain gene, while the exact etiology is yet unknown which may be related to mutations in several genes, including tafazzin, dystrobrevin, and ZASP [3,4]. The estimated incidence of NCCM in children aged 0 to 10 years old is 0.12 per 100,000, while in infants aged 0 to 12 months, it can reach up to 0.81 per 100,000 [5]. Despite being a rare heart condition, NCCM stands third among pediatric cardiomyopathies, behind dilated cardiomyopathy (DCM) and hypertrophic cardiomyopathy (HCM) [6]. Adults with NCCM are at risk for thromboembolic events, arrhythmias, and heart failure [7]. NCCM can either manifest as a nonisolated or isolated phenotype. Congenital cardiac disease may coexist with nonisolated NCCM. We report a case of biventricular noncompaction cardiomyopathy in a preterm 5-day-old girl (34 weeks gestational age) presenting with signs of congestive heart failure.

*Abbreviations:* NCCM, noncompaction cardiomyopathy; RV, right ventricle; TTE, transthoracic echocardiography; LV, left ventricle; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy.

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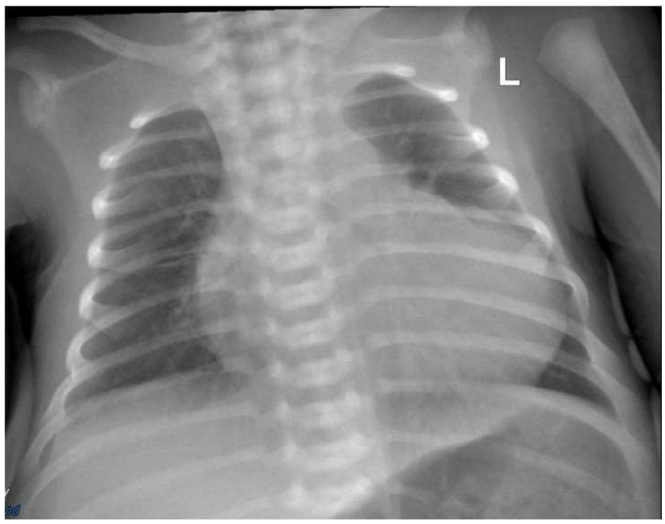


**Fig. 1 – Chest Xray showed Chest X-ray showed mild cardiomegaly and pulmonary congestion.**

## Presentation of case

A preterm 5-day-old girl, 34 weeks gestational age, weighing 2.6 kg, had poor feeding and persistent respiratory distress. The clinical evaluation showed a hyperdynamic precordium, tachypnea (67 bpm), tachycardia (150 bpm), and 98% SpO2. A chest X-ray revealed moderate cardiomegaly (Fig. 1).

Echocardiography revealed a mildly dilated left ventricle, right ventricle hypertrophy, and about 56% left ventricular ejection fraction (Fig. 2D). The left ventricle's anterior wall, lateral wall, apex, and mid-and apical septum were all found to have prominent trabeculations and intertrabecular spaces, which gave the structure its characteristic spongiform look (Figs. 2A and B). Furthermore, the right ventricle was shown to have a prominent trabecular pattern with intertrabecular recesses (Fig. 3A). In both ventricles, there was low-velocity Doppler and color Doppler across the intertrabecular recesses (Fig. 2C and 3B). NT-proBNP level was 605.5 pg/mL, Troponin T hs level was 2553 pg/mL.

Initially treated with furosemide and captopril, she needed oxygen therapy via nasal cannula for 18 days. She had no signs of heart failure or respiratory distress when she received her discharge from the hospital on day 20 of her life.

## Discussion

In 1984, Engberding and Bender published the first description of NCCM. The American Heart Association classifies NCCM as a significant inherited cardiomyopathy, characterized by a reconstructed myocardial wall with deep intertrabecular recesses and prominent trabeculae [2].

In the literature, the following diagnostic standards for isolated left ventricular noncompaction on echocardiography have been applied a bilayered myocardium, a noncompacted to compacted ratio greater than 2:1, Doppler-demonstrated communication with the intertrabecular space, the lack of concomitant cardiac anomalies, and the appearance of many prominent trabeculations at end-systole are among these requirements [8]. Typically, the myocardium compresses between weeks 5 and 8 of fetal life, moving from the septal to the lateral walls and from the basal to the apical segments. It is thought that in congenital NCCM, mutations in genes or epigenetic regulation of particular cardiac pathways inhibit the normal process of myocardial compaction, leaving the myocardium with 2 layers: the endocardium, which is a honeycomb-like structure with deep intertrabecular recesses and extensive ventricular trabeculation, and the epicardium, which is a compact layer [9].

Systolic function changes between temporary increases and decreases, and this corresponds to an undulating phenotype associated with NCCM which is frequently a part of a mixed phenotype, which includes HCM-DCM (28%), HCM (27%), and DCM (19%) [10].

In children with NCCM, arrhythmia is a recognized risk factor for some morbidities and mortality. It is therefore necessary to conduct additional studies on the precise incidence and severity of the various types of arrhythmias as well as the management of problems for pediatric NCCM patients [10]. The incidence of stroke and other thromboembolic events is another factor that is commonly associated with NCCM. It is thought that blood clots in the myocardium's honeycomb-like structure are likely to form, resulting in blood clots in some areas of NCCM patients [11].

For the treatment of NCCM, no particular medicinal or surgical approach has been effectively used to date. However, medication utilizing an angiotensin II receptor blocker, angiotensin-converting enzyme inhibitor, or a beta-blocker may result in beneficial remodeling of the left ventricle. In addition, children with NCCM should have careful surveillance for any problems or deteriorations, and any arrhythmia should be managed according to medical recommendations. It is necessary to assess these patients' ICD therapy's efficacy. Further investigation is necessary to ascertain whether antiplatelet medication or preventive anticoagulation is necessary [12].

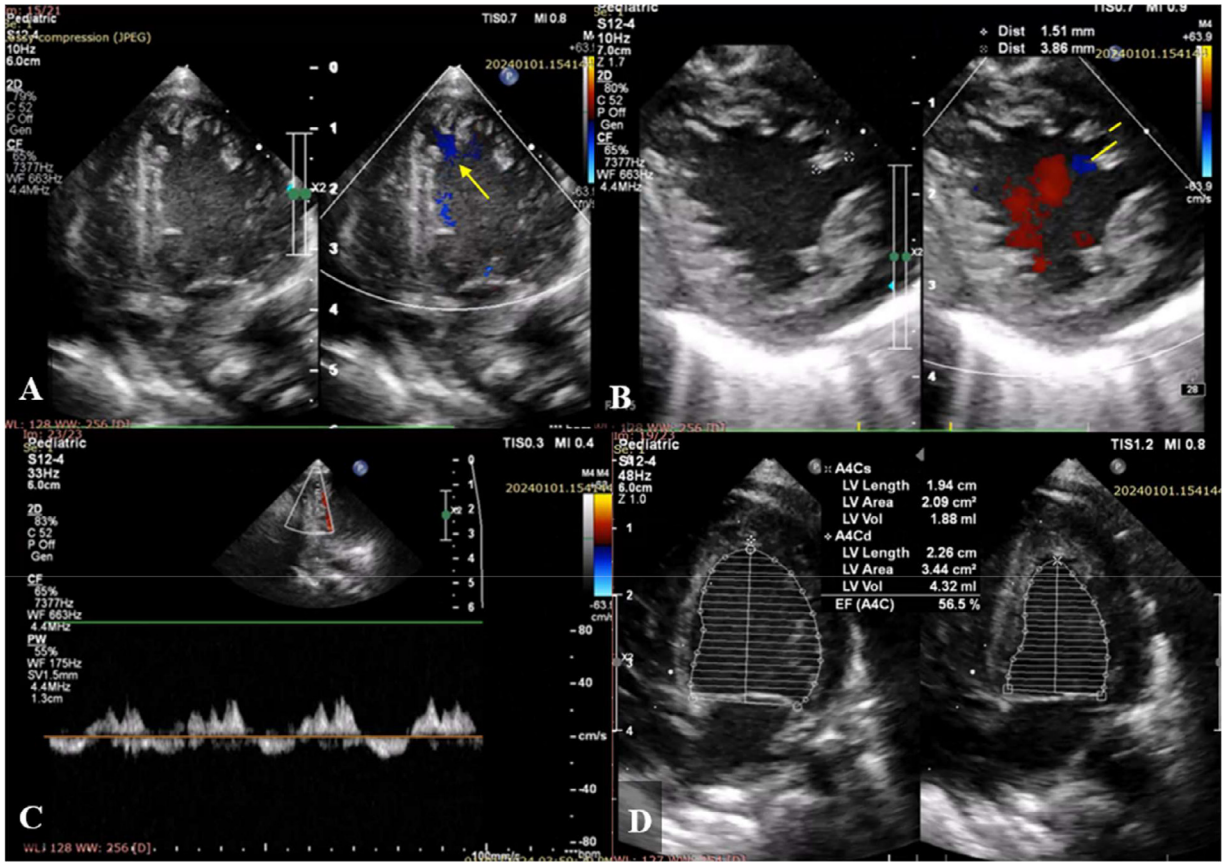


**Fig. 2 – Echocardiography showed: (Picture A) increased trabeculation in the apex and color Doppler flow (Yellow arrow). (Picture B) noncompacted/compacted ratio >2.0 in end-systole (Yellow line); (Picture C) intertrabecular spaces perfused from the low-velocity Doppler; Picture D: Left ventricular ejection fraction was 56%.**

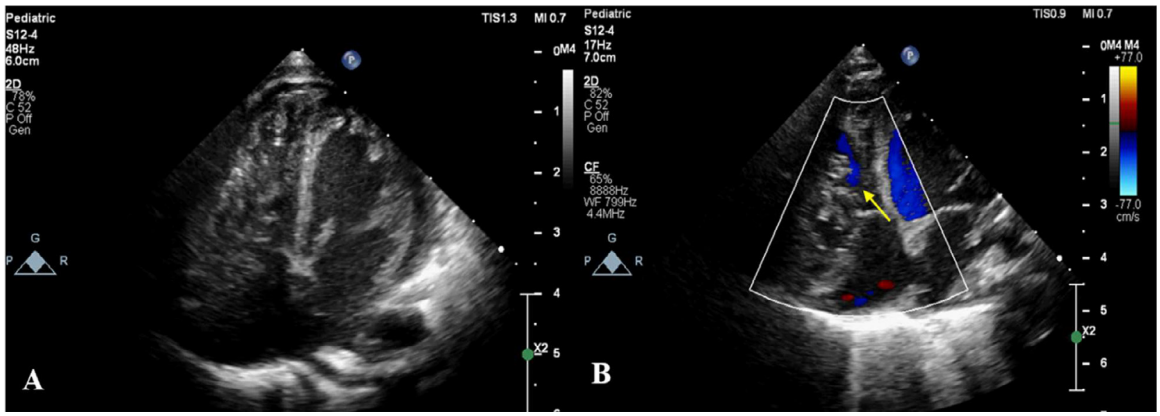


**Fig. 3 – Echocardiography showed: (Picture A) The right ventricle was shown to have a prominent trabecular pattern with intertrabecular recesses; (Picture B) Intertrabecular spaces perfused from the low-velocity Doppler (Yellow arrow).**

Compared to NCCM patients without a mixed cardiomyopathy phenotype, pediatric NCCM patients with mixed cardiomyopathy phenotypes have a higher risk of transplant or death [13]. According to an Australian study, just 20% of young patients with dilated NCCM were still alive and had normal cardiac function after a 15-year follow-up, and half of those kids either died or had heart transplants within 10 years after being diagnosed [6].

## Conclusion

Although controversial, biventricular involvement is even more uncommon than NCCM. Usually, this disorder is accompanied by potentially fatal consequences. The spectrum of clinical presentation is broad, including asymptomatic, congestive heart failure, and sudden cardiac arrest. Doppler

echocardiography is still the most common initial test that analysis helps suggest the diagnosis of NCCM and may lead to further evaluation.

## Availability of data and materials

Data and materials used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

## Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

## Patient consent

Written informed consent for patient information to be published in this article was obtained.

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