Pediatric Oncall Journal

DOI: https://doi.org/10.7199/ped.oncall.2025.60

PEDIATRIC ONCALL
CHILD HEALTH CARE

TEACHING FILES (GRAND ROUNDS)

BRONCHIOLITIS UNVEILING A HIDDEN HEART: A CASE OF LEFT VENTRICULAR NON-COMPACTION

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ARTICLE HISTORY
Received 18 January 2025
Accepted 01 February 2025

KEYWORDS
Bronchiolitis, left ventricular non compaction, pediatric cardiology.

Clinical Problem

An 8-month-old male infant presented with a three-day history of cough, rhinorrhoea, and low-grade fever and one day history of fast breathing and feeding difficulties. He was born full-term with no significant past medical history. Physical examination revealed an irritable child with tachypnoea, tachycardia, subcostal, intercostal retractions, and bilateral wheezes. Oxygen saturation was 92% on room air. Other systemic examinations were normal. Complete blood count was unremarkable. C-reactive protein was mildly elevated. Chest X-ray showed hyperinflation and patchy atelectasis consistent with bronchiolitis. The infant was managed conservatively with supportive care, including supplemental oxygen, nebulized bronchodilators, and adequate hydration. However, despite initial improvement, the child continued to exhibit tachypnoea and tachycardia. Due to the persistent symptoms, a transthoracic echocardiogram was performed which revealed a left ventricle with prominent trabeculations and deep intertrabecular recesses, raising suspicion for Left ventricular non compaction (LVNC). Left ventricular systolic function was reduced with an estimated ejection fraction of 45%. A detailed cardiac evaluation, including a repeat echocardiogram and electrocardiogram (ECG), was performed. The repeat echocardiogram confirmed the presence of prominent trabeculations and deep intertrabecular recesses in the left ventricle, consistent with LVNC. ECG was unremarkable. Cardiac magnetic resonance imaging (CMR) was performed to further characterize the LVNC and assess for any associated abnormalities. CMR confirmed the diagnosis of LVNC with moderate severity. On consultation with the paediatric cardiologist, the infant was started on diuretics and an angiotensin-converting enzyme inhibitor to manage heart failure symptoms associated with LVNC to which the child responded well and was discharged on day 7 of admission. The infant is currently asymptomatic and receiving regular cardiology follow-up. This case highlights the importance of considering underlying cardiac conditions in infants with persistent respiratory symptoms following an episode of bronchiolitis. A comprehensive evaluation, including careful clinical assessment, thorough investigations, and close follow-up, in infants with suspected cardiac involvement can aid in early diagnosis and prompt treatment.

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What is left ventricular non compaction?

Discussion

Left Ventricular Noncompaction (LVNC) is a rare primary cardiomyopathy with a likely genetic origin. It primarily affects the left ventricle, characterized by a distinct two-layered myocardial structure. An abnormally trabeculated, thick, spongy inner layer communicates with the ventricular cavity, while a thin compacted outer layer forms the exterior. LVNC may occur in isolation or be associated with other cardiomyopathies, congenital heart diseases, neuromuscular disorders, or reversible conditions like pregnancy, athletic activity, and hemoglobinopathies.[1] The prevalence of LVNC ranges from 0.014% to 0.26%, with a higher incidence in males.[2] Clinical presentations can vary widely, ranging from asymptomatic cases to those with cardiac failure symptoms (dyspnea, exercise intolerance, peripheral edema), arrhythmias, thromboembolic events, and sudden cardiac death.[3]

Diagnosis

Two-dimensional echocardiography (2D-ECHO) is the initial diagnostic tool. A ratio of the trabeculated spongy layer to the compact layer exceeding 2 in end-systole on short-axis views is a key diagnostic criterion. 2D-ECHO also helps rule out associated congenital heart defects. Cardiac magnetic resonance imaging (CMR) can confirm the diagnosis and provide additional information, such as ventricular thrombi and myocardial fibrosis.[4]

Management

Management is primarily supportive. Close monitoring is crucial for patients with normal ventricular function to detect complications. Aspirin or oral anticoagulants is recommended for patients with reduced ventricular function due to the increased risk of clot formation associated with hyper-trabaculation. Anticoagulants is also advised for LVNC patients with atrial fibrillation, as these individuals are at higher risk of systolic dysfunction over time. Other arrhythmias should be managed according to standard protocols including implantable cardiac defibrillator in patients with severe arrhythmias to prevent sudden death. Patients

with congestive heart failure may benefit from diuretics, angiotensin-converting enzyme inhibitors (ACE inhibitors), or angiotensin receptor blockers (ARBs). Beta-blockers may be necessary in cases with concomitant hypertrophic cardiomyopathy. Approximately 12% of LVNC patients may progress to end-stage disease requiring cardiac transplantation.[5]

Prognosis

The prognosis of children with LVNC is variable. However, a significant proportion are likely to develop systolic dysfunction over time. Mortality rates are influenced by factors such as ejection fraction and the presence of complications like arrhythmias and thromboembolic events.[6]

Compliance with ethical standards

Funding: None

Conflict of Interest: None

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