Left ventricular non-compaction in children
A Case Report

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Isolated left ventricular non-compaction (LVNC) is a genetic abnormality characterized by prominent ventricular trabeculations and deep inter-trabecular or sinusoidal cavities communicating with the left ventricular cavity [1]. LVNC can be isolated or associated with cardiomyopathies, congenital heart disease and complex syndromes involving the heart. The American Heart Association classifies LVNC as genetic cardiomyopathy (2).

We report the case of a 2-year-old girl, with a history of asthma, admitted to pediatric emergencies for respiratory distress. The physical examination found a heart rate of 260 beats per minute, a tachypnea, grunts, signs of right venous congestion with hepatomegaly, low cardiac output: low energy, pallor, cold extremities.

A chest x-ray showed no parenchymal abnormalities, the cardiac chest index was 71% and the electrocardiogram found supraventricular tachycardia with fine QRS.

Echocardiography demonstrated dilated cardiomyopathy with (FE = 41%, FR = 19%). Color Doppler reveals an enlarged spongiform left ventricle with deep trabecular cavities. The diagnosis of dilated cardiomyopathy most likely related to isolated left ventricular non-compaction. The treatment was based on digital diuretics, a vasodilator and an amiodarone-type antiarrhythmic agent.

Left Ventricular Compaction Cardiomyopathy (LVNC), was described by Grant in 1926, Various types of electrocardiographic abnormalities, arrhythmias, and conduction disturbances have been reported in children with LVNC. In two series of children with LVNC, ECG abnormalities were present in 81% of cases [3]. The most common abnormality was ventricular arrhythmia, isolated single ventricular extra systoles, sustained ventricular tachycardia and ventricular fibrillation, which can be fatal. Another frequent association is the presence of WPW syndrome [4]. Our clinical case illustrates this, or there was a complicated supraventricular tachycardia of FA. Echocardiography was the diagnostic procedure of choice for LVNC. Although computed tomography and magnetic resonance imaging are alternative means of diagnosis, their superiority over echocardiography has not yet been conclusively proven [5].

Echocardiography was the diagnostic procedure of choice for LVNC. Although computed tomography and magnetic resonance imaging are alternative means of diagnosis, their superiority over echocardiography has not yet been conclusively proven [6]. The management of the disease depends on the associated comorbidities, including heart failure, thromboembolic events and arrhythmias.

Echocardiatic Diagnostic Criteria for left Ventricular non compaction

Author and Status	Criteria
Validated by Jenni et al.	Presence of prominent ventricular trabeculations , predominantly in the apical and midventricular areas of both the inferior and lateral walls Two-layered appearance of the myocardium with a thin , compacted outer(epicardial) band and thicker, non compacted inner(endocardial) layer;end-systolic ratio between non compacted and compacted myocardium is greater than 2 Multiple deep intertrabecular recesse communicating with the ventricular cavity, as visualized color Doppler imaging
New diagnostic criterion introduced by Stollberger et al.	End-diastolic ratio between non compacted myocardium greater than 2 in apical four-chamber view
Additional criterion introduced by Gebhard et al.	Systolic thickness of compacted Myocardium less than 8mm may facilitate differentiation of left ventricular non compaction from normal hearts and potentially from those with myocardial hypertrophy

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In children with signs of dilated cardiomyopathy, left ventricular non-compaction LVNC should be discussed. The frequency of the disease in patients with dilated cardiomyopathy is higher than suggested, and in these patients, attention should be paid to the formation of thromboembolic events and arrhythmias.

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