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CLINICAL ASPECTS OF THE COURSE OF LEFT VENTRICULAR NONCOMPACTION IN CHILDREN

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RELEVANCE

The relevance of noncompaction cardiomyopathy (NCCM) in children is обусловед by a number of clinical, diagnostic, and prognostic factors that have increasingly attracted the attention of pediatricians and pediatric cardiologists in recent years [3,5]. Noncompaction cardiomyopathy is a rare but potentially life-threatening form of cardiomyopathy characterized by impaired normal embryonic myocardial compaction, resulting in prominent trabeculation and deep intertrabecular recesses. Despite its relative rarity, this condition is being diagnosed more frequently due to the widespread use of modern imaging techniques such as echocardiography and cardiac magnetic resonance imaging [1,6,9].

NCCM is of particular significance in the pediatric population, as its clinical manifestations are highly variable, ranging from an asymptomatic course to severe heart failure, life-threatening arrhythmias, and thromboembolic complications. In children, the disease often presents at an early age and may lead to disability or death [2,5,7].

Recent studies, including those by the authors of this article (2022; 2026), emphasize that the clinical course of noncompaction myocardium in children is characterized by marked polymorphism of symptoms and varying degrees of hemodynamic impairment, which complicates early diagnosis and necessitates in-depth clinical and instrumental evaluation. Their findings indicate that left ventricular noncompaction (LVNC) is often detected incidentally or at the stage of already established heart failure, underscoring the need for more detailed investigation of the clinical aspects of this condition [1,4,7].

The absence of specific clinical signs in the early stages makes timely diagnosis difficult, thereby increasing the risk of an unfavorable prognosis [2,8,10].

Diagnostic challenges in NCCM are also associated with the lack of universally accepted criteria, especially in pediatric practice. Different echocardiographic and MRI criteria may yield inconsistent results, leading to both overdiagnosis and underestimation of the true prevalence of

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the disease. This highlights the need to improve diagnostic approaches and develop clear clinical guidelines [1,3,6].

Thus, noncompaction cardiomyopathy in children represents a relevant medical problem requiring further investigation. The need for early diagnosis, clarification of pathogenetic mechanisms, development of unified diagnostic criteria, and optimization of treatment strategies determines the importance of conducting scientific research in this field and implementing its results into clinical practice.

Objective. To evaluate and analyze the clinical manifestations of left ventricular noncompaction in children.

MATERIALS AND METHODS

The study was conducted at the cardiology department and the functional diagnostics department of the Andijan Regional Children's Multidisciplinary Medical Center. The study included 20 children aged 8 to 16 years with a verified diagnosis of noncompaction cardiomyopathy (NCCM).

The diagnosis of NCCM was established based on the presence of clinical manifestations of heart failure, signs of cardiomegaly, and reduced myocardial contractility.

Verification of the diagnosis was based on a comprehensive approach, including analysis of anamnestic data obtained through parental interviews and review of medical records, as well as the results of clinical examination, electrocardiography (ECG), echocardiography (EchoCG), and chest radiography.

Electrocardiographic examination was performed in 12 standard leads using a six-channel electrocardiograph. Assessment of the morphofunctional state of the heart was carried out based on echocardiographic data obtained in M-mode and B-mode (one-dimensional and two-dimensional imaging), with the use of Doppler mode to analyze intracardiac hemodynamics.

RESULTS

The study revealed that girls with left ventricular noncompaction outnumbered boys by four times (80% vs. 20%).

At the time of hospitalization, approximately two-thirds of the children were assessed as being in severe condition, accompanied by clinical signs of class IIB heart failure.

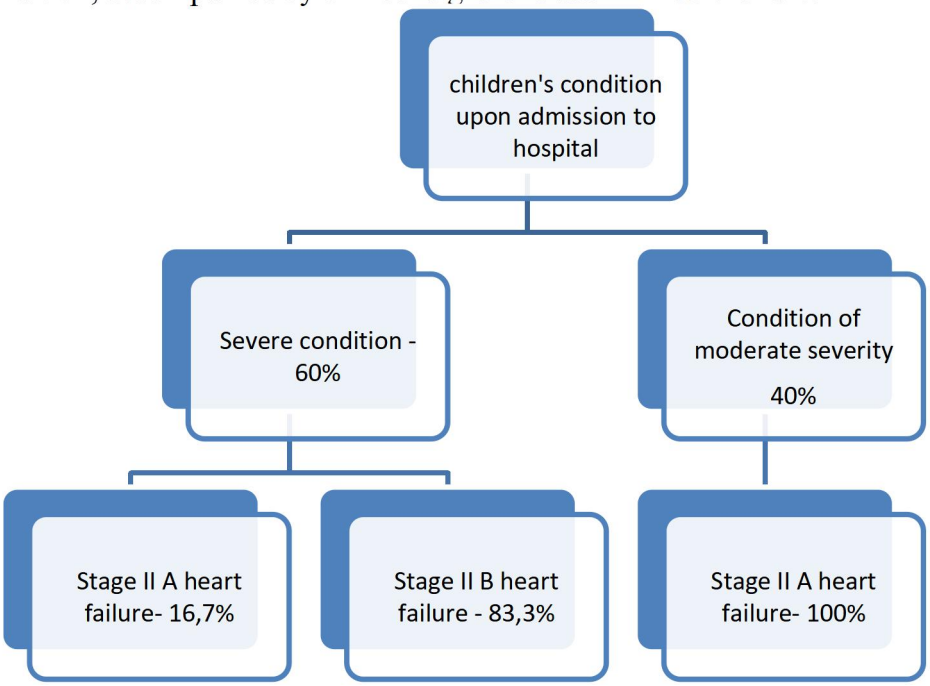

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Figure 1. Condition of children upon admission

Analysis of the distribution of patients by the severity of heart failure showed a predominance of girls among those with more severe clinical manifestations (class IIB heart failure), whereas class IIA heart failure was observed with equal frequency regardless of sex.

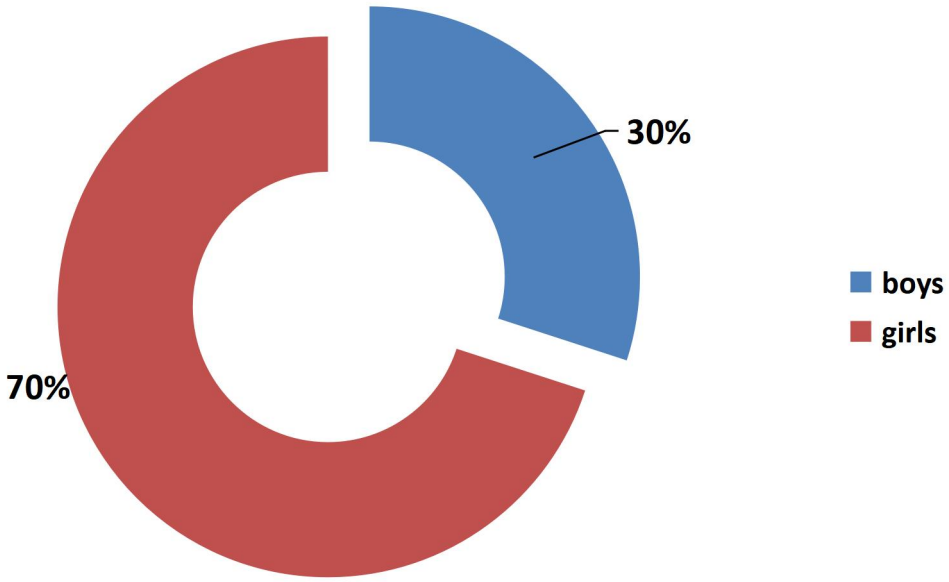
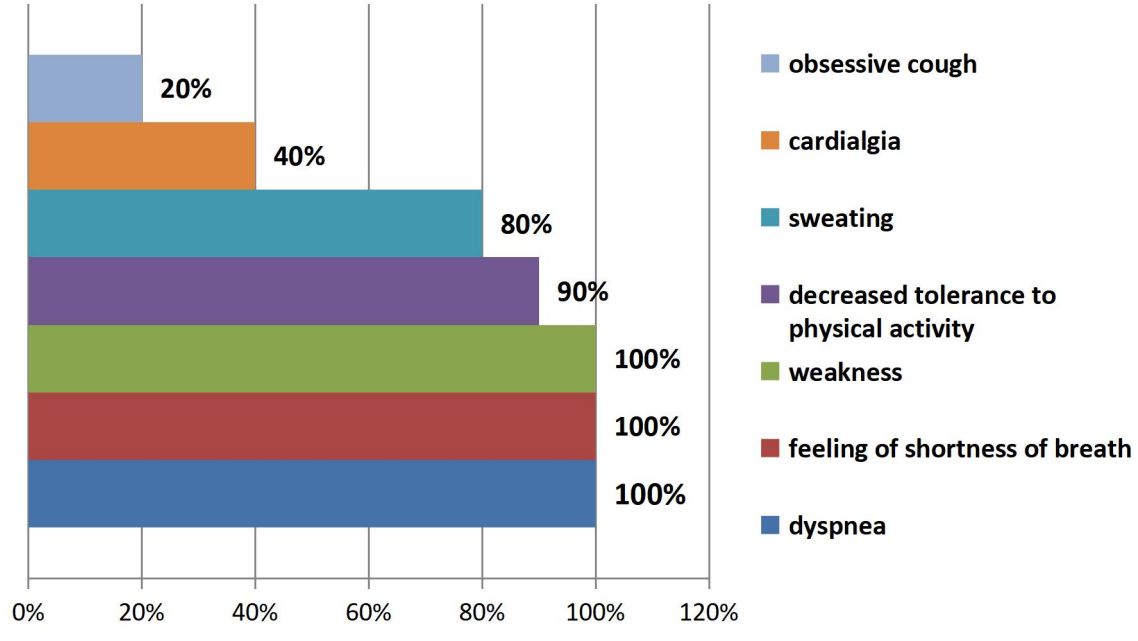


Figure 2. Frequency of stage II B heart failure by gender (n = 12).

Subjective clinical manifestations upon admission to the hospital are presented in Figure



3.

Figure 3. Subjective symptoms of the disease (n = 20).

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The results of the objective examination revealed that all children had a systolic murmur of varying intensity, localized at the cardiac apex.

Table 1. Objective signs of the disease (n = 20).

Clinical examination findings	abs	%
Pale skin	20	100%
Mottled skin tone	3	15%
Cyanosis	16	80%
Shortness of breath	20	100%
Muffled heart sounds on auscultation	20	100%
Systolic murmur	20	100%
Tachycardia	18	90%
Bradycardia	2	10%
Hepatomegaly	16	80%

CONCLUSIONS

In the majority of children with noncompaction cardiomyopathy (60%), the condition at hospital admission was assessed as severe, with pronounced signs of class IIB heart failure, indicating late referral and a progressive course of the disease.

The obtained data confirm that noncompaction cardiomyopathy in children is characterized by a severe clinical course with early development of symptoms of chronic heart failure and signs of systemic hypoperfusion.

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