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Research Article

CARDIOMYOPATHIES IN CHILDHOOD: CLINICAL COURSE, DIAGNOSIS AND CARE TACTICS

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ABSTRACT

The article examines aspects of the clinical course, diagnostics and treatment of various forms of cardiomyopathy (dilated, hypertrophic, restrictive) in children. Due to the relevance of the problem, the obtained data on the features of the clinical course, diagnostics and tactics of patient management are of interest to cardiologists, general practitioners and pediatricians of family clinics.

A total of 37 children aged 0-17 years were examined. Among those examined, the frequency of various forms of cardiomyopathy was: dilated - 62.1%, hypertrophic - 32.4%, restrictive - 5.4%. Anamnestic data, severity of the course, features of ECG, echocardiography, radiography for each form of cardiomyopathy were studied.

Latent, mild, but more often moderate and severe course of the disease was noted. Patients with DCM and HCM often have various rhythm and conduction disorders. Outpatient observation and timely (continuous) therapy of CHF are indicated for children with cardiomyopathies. In severe cases of HCM with obstructive forms of HCM, the issue of surgical treatment is considered.

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KEYWORDS

Dilated, hypertrophic, restrictive cardiomyopathy, children, chronic heart failure.

INTRODUCTION

Cardiovascular pathology in childhood is considered a serious problem due to its prevalence, difficulties in recognition, features of the course, treatment and outcomes [1-9]. Cardiomyopathy in pediatric cardiology remains the least studied and, unfortunately, untreatable heart pathology. High disability and mortality, non-specificity of clinical and laboratory markers, lack of effective treatment methods, difficulties in heart transplantation in childhood determine the relevance of the problem. In recent years, an increase in the incidence of various forms of cardiomyopathy in children has been noted, and this is primarily due to improved diagnostics. A true increase in the incidence of cardiomyopathy is also possible [7, 10-18].

Dilated cardiomyopathy (DCM) is the most common form of cardiomyopathy in both adults and children. DCM in children is a primary myocardial disease of unknown etiology, characterized by dilation predominantly of the left ventricle with a decrease in the contractile ability of the myocardium. Most likely, DCM has a polyetiological heterogeneous nature, including viral persistence, genetic determination, and autoimmune factors [10-16,18,20]. The incidence in childhood is 0.5%. Genetic forms of DCM account for approximately 30%. The incidence does not depend on gender or age. The average age of children at the time of diagnosis is 6-13 years [11,16,19].

Hypertrophic cardiomyopathy (HCM) is a genetically determined disease of the heart muscle, characterized by massive hypertrophy of the myocardium, most often the left ventricle, with the obligatory involvement of the interventricular septum. The population frequency among children is 2.5 per 100,000. According to numerous studies in different geographical locations around the globe, the frequency of HCM is 0.2-1.1%. Symptoms of obstruction are detected in only 20% of individuals with HCM [10-12,16].

Restrictive cardiomyopathy (RCM) is the least rare of all cardiomyopathies. The disease is based on widespread interstitial fibrosis, combined with a sharp thickening of the endocardium. With this type of cardiomyopathy, diastolic function of the myocardium suffers with little change in systolic function. The etiology of the disease is unknown. It can occur in children at any age, with a predominance in girls. The incidence is about 5% among all forms of cardiomyopathy. The mortality rate in children from

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the time of diagnosis is 63-75% from 3-6 years, respectively, from the onset of the disease [10,11,16].

All forms of cardiomyopathy, given the increase in the frequency of recognition, severity of the disease, lack of effective methods of therapy, and high mortality, require further study.

The goalto study the course, diagnostic criteria and treatment tactics to improve the quality of life and optimize the prognosis.

METHODS

The study included 37 patients who were inpatients in the cardiology department and subsequently followed up as outpatients at the ODMMC from 2019 to 2024. The observed children were aged 0-17 years, including 19 boys (51.4%) and 18 girls (48.6%). All children underwent echocardiography, ECG, chest X-ray, ultrasound of the abdominal organs, and general clinical tests. The main criteria for their determination according to echocardiography data (in 4 patients, additionally, MSCT of the heart and vessels with contrast) are: the presence of dilation of the cavity or hypertrophy of the myocardium of the left (in some cases, the right) ventricle, the predominance of atrial lesions (in patients with RCM) or ventricles, the degree of change in the functional state of the heart (decreased ejection fraction).

RESULTS AND DISCUSSION

When studying the anamnesis, a family history of cardiomyopathy was found in 8 (21.6%) children, all of whom had HCM, 2 (5.4%) had intrauterine TORCH infection, and 12 (33.3%) children were frequently ill. In our observation, we did not aim to conduct a genetic examination; according to the anamnesis of our patients with DCM, no familial cases were identified. However, according to the literature, about 30-40% of cases of familial isolated DCM have an established genetic origin; more than 50 associated genes have been identified, and this number is constantly increasing as new genes are discovered [4,11,13,16]. In all examined patients, the diagnosis of cardiomyopathy was made at least several months after the onset of the first symptoms, after ineffective therapy for other diseases (acute and recurrent upper respiratory tract infections, pneumonia, carditis). According to the severity of the disease, severe course was observed in 24 patients (64.9%), moderate course - 8 (21.6%), mild and asymptomatic - 5 (13.9%). Severe course was observed mainly in patients with DCM, mild and asymptomatic - in 5 children with HCM. Cardiomyopathies in children were accompanied by circulatory failure of HC I degree (FC1) - 7 (18.9%), HC II degree (FC2-3) - 20 (54.1%), HC III degree (FC4) - 5 (13.5%). After clinical and instrumental examination, the following forms of cardiomyopathy were revealed: DCM - 23 (62.2%), RCM - 2 (5.4%) and HCM - 12 (32.4%), among the latter, the obstructive form - in 5 children (13.9%). These data are consistent with the literature on

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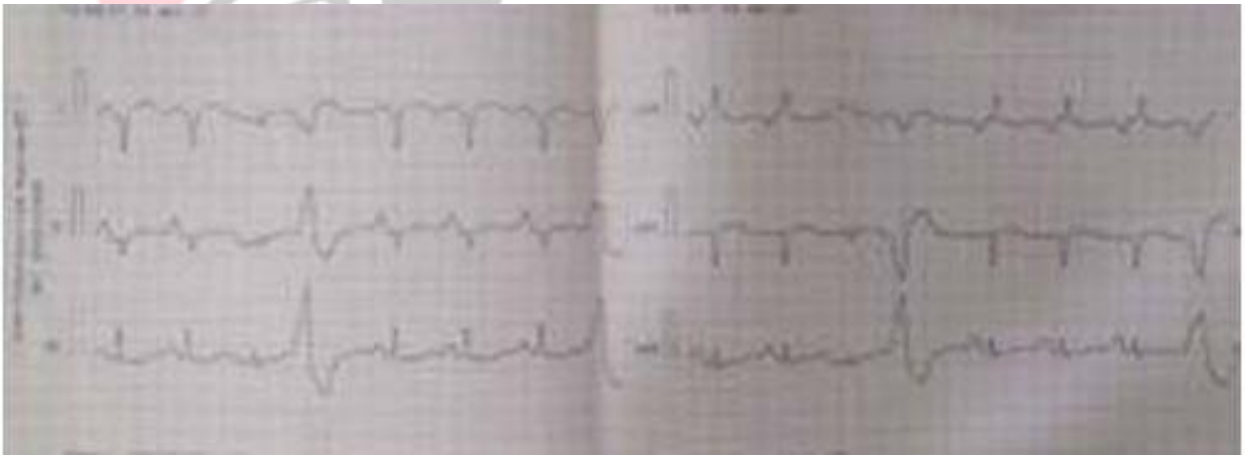
the incidence of various forms of cardiomyopathy [10,11,15,16]. The distribution by gender was as follows: among patients with DCM, there were 10 boys (43.5%), 13 girls (56.5%), 8 boys (66.7%) and 4 girls (33.3%) with HCM, 1 boy (50%) and 1 girl (50%) with RCM.

ECG of patients with DCM reveals signs of left ventricular hypertrophy (enlargement) (100%), atrial overload (69.6%), and repolarization disorders, often of ischemic origin (56.5%). Rhythm disorders include sinus tachycardia (78.3%), supraventricular tachycardia (17.4%), pacemaker migration (4.8%), ventricular extrasystole (39.1%), and polytopic extrasystole in one patient (4.3%), and atrial flutter (4.3%) (Fig. 1). In ECG in HCM we found signs of left ventricular hypertrophy with its overload (100%), repolarization disorders in the form of ST depression and deep negative T waves (Fig. 2) (58.3%) - a characteristic sign of HCM, bundle branch block in the form of left anterior block (25%), left posterior block (8.3%), we found right bundle branch block in (25%), extrasystoles - in (16.7%). In RCM, signs of severe right atrium hypertrophy are the most characteristic sign (100%), in our observations, patients had rhythm disturbances only in the form of sinus tachycardia (100%). The frequency of arrhythmias in children with cardiomyopathy is also shown in the works of foreign authors [11,13-17]. Arrhythmias are the most common cause of sudden death in cardiomyopathy [11,16,19,20].



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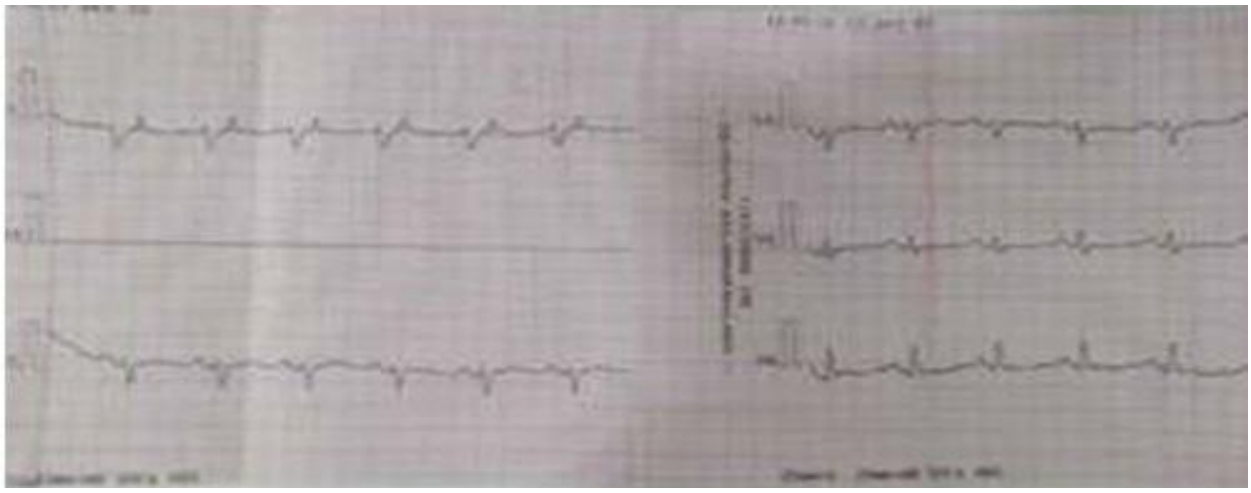

Fig. 1 ECG of patient M. with DCM. Sharp deviation of the EAH to the right, ventricular extrasystoles, signs of overload of both parts of the heart, changes in the ventricular myocardium, decreased voltage

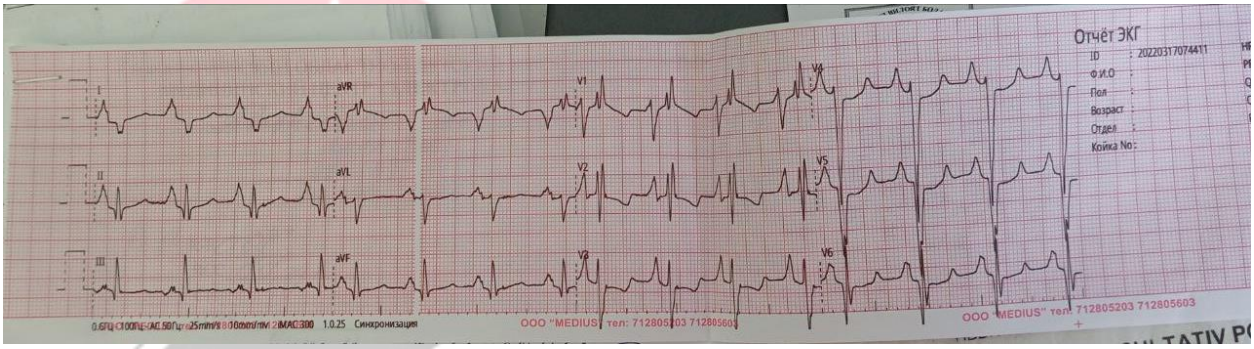

Fig. 2. ECG of patient A. with RCM. Deviation of the EAH to the right. Signs of severe RA overload. Imcomplete blockade of the right bundle branch.

According to X-ray data, in DCM the heart is significantly enlarged in cross-section due to the left or both ventricles (Fig. 3). CTI was 0.60-0.71. The heart is of a characteristic spherical or trapezoidal shape. In (52%) patients, venous congestion was observed. In HCM the heart shadow is enlarged in cross-section mainly due to the left ventricle. CTI up to 0.66. In RCM the pulmonary pattern is not enriched on the X-ray. The heart shadow is slightly enlarged in cross-section. CTI - up to 0.60. The arches are smoothed.

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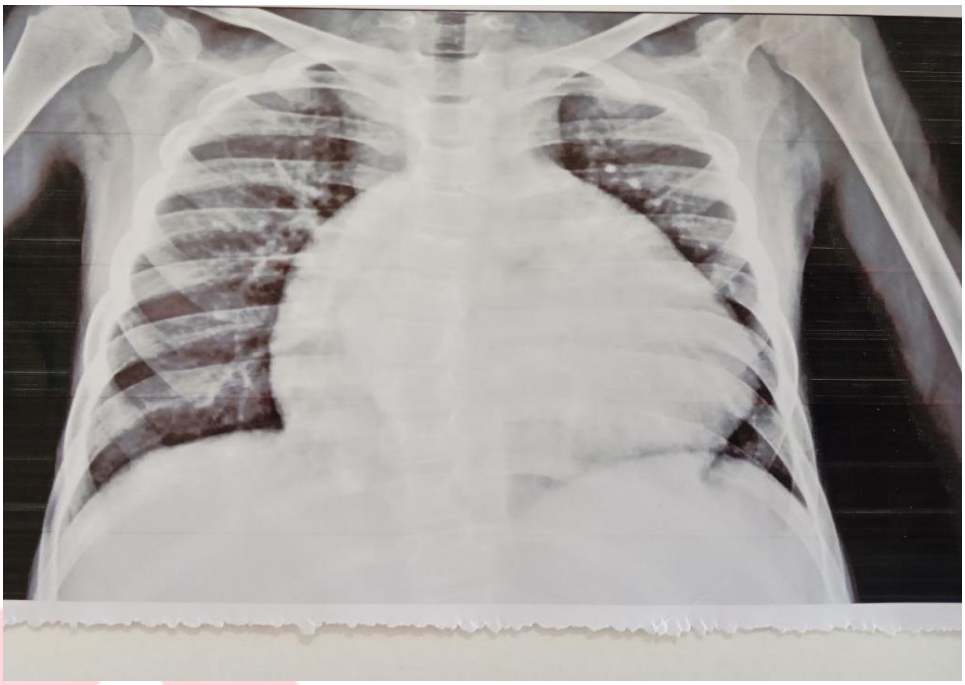

Fig. 3. X-ray of patient M. with RCM.

In patients with all three forms, regardless of the severity of the patient's condition, chronic heart failure of varying degrees was noted. In 3 (25%) patients with HCM, chronic heart failure was absent during the survey and examination, in 2 (16.7%) it corresponded to FC1. All patients with DCM had chronic heart failure. Upon admission to the hospital FC3-4, upon discharge - FC2. Patients with RCM were admitted to the hospital with CHF FC3, and later treated outpatients with CHF FC2.

In the hospital, children with DCM received cardiotonic therapy with cardiac glycosides, dopamine if indicated, ACE inhibitors, beta-blockers (carvedilol), diuretic, cardiotrophic and symptomatic therapy. Positive dynamics were noted against the background of the therapy in the hospital. However, after discharge, despite outpatient treatment, CHF gradually progressed again. 2 (9.1%) girls with DCM died. Their disease duration was more than 5 years. The remaining children in the follow-up are under dispensary registration at the place of residence and receive symptomatic therapy for CHF. Patients with HCM received treatment with beta-blockers (egilok) on an outpatient basis. 1 child was operated on in the cardiac surgery department of the ODMMC, the condition after the operation improved significantly, continues to receive egilok. 2 children with RCM received therapy for CHF, but without the use of inotropic drugs; they were treated jointly with a gastroenterologist, since

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congestion in the liver and ascites led to gastrointestinal dysfunction. All patients with DCM and RCM received anticoagulant therapy.

Thus, the frequency of various forms of cardiomyopathy in children in the overall structure of cardiovascular diseases is currently increasing. Diagnostics has improved, and, consequently, the detection of all forms. Protocols for the treatment of cardiomyopathy have been developed. Most patients with severe cardiomyopathy, with proper monitoring and treatment, have a disease history of more than 5 years. However, a large number of complications requires that this pathology be classified as a disease with an unfavorable prognosis.

CONCLUSIONS

1. Among those examined, the frequency of various forms of cardiomyopathy was: DCM - 61.1%, HCM - 33.3%, RCM - 5.6%.

2. Almost all forms of cardiomyopathy (with the exception of 3 patients with HCM) were accompanied by moderate and severe persistent CHF.

3. Often, late detection of the disease in the majority of patients dictates the need for at least an annual examination of all children using the echocardiography method.

4. Most patients with DCM and HCM exhibit various rhythm and conduction disturbances.

5. Dispensary observation and timely (continuous) therapy of CHF in children with cardiomyopathy are indicated.

6. For patients with obstructive forms of HCM, the issue of surgical treatment is considered.

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