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RISK FACTORS AND PROGNOSIS IN CHILDREN WITH CARDIOMYOPATHY

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Abstract:

Cardiomyopathies remain one of the least studied and most difficult problems in pediatric cardiology. The manifestation of cardiomyopathies can develop at any age and range from asymptomatic forms to pronounced clinical manifestations with a poor prognosis. One of the most common in children is dilated cardiomyopathy, and the most insidious is hypertrophic, which often ends in sudden cardiac arrest. Restrictive cardiomyopathy, which is detected during targeted examination, is quite difficult in terms of diagnosis. Non-compact cardiomyopathy is a frequent, but poorly understood, cardiomyopathy, in most cases complicated by arrhythmic syndrome.

Keywords: dilated, hypertrophic, restrictive, non-compact cardiomyopathy, heart failure, diastolic dysfunction, cardiac arrhythmias, subepicardial ischemia.

RELEVANCE. In recent years, the structure of diseases of the cardiovascular system in children has changed significantly, with a decrease in the number of patients with bacterial heart disease and an increase in myocardial diseases of unclear ethiology. It is in this group that cardiomyopathies are classified as one of the most difficult problems of pediatric cardiology . Cardiomyopathies remain one of the least studied cardiac diseases in children, due to their nonspecific clinical manifestations. Despite the development of high-tech diagnostic methods, morbidity and mortality from cardiomyopathy remains high. The lack of generally accepted algorithms and treatment regimens for children with cardiomyopathies leads to the progression of the disease and often death.

It is far from always possible to define a clear boundary between different forms of cardiomyopathies, since there are similar clinical symptoms and one type of cardiomyopathy may develop into another over time.

According to the data of pathological studies during life, only about a quarter of cases are diagnosed among those who died from sudden death syndrome at the age from birth to 18 years. Non-coronary changes in the myocardium are found with a frequency of 20-40% .The manifestation of cardiomyopathies can develop at any age and clinical manifestations can be expressed to varying degrees. Some children feel quite well without making any complaints or alarming their parents. It is these children who are susceptible to sudden death.

Dilated cardiomyopathy (DCMP) is one of the most common clinical forms of cardiomyopathy in children. The disease is characterized by pronounced morpho-structural changes in the myocardium, the result of which is the expansion of the cavities of the heart, especially the left ventricle, a decrease in myocardial contractility and the development of progressive congestive heart failure, refractory to on going drug therapy.The prognosis of the disease is serious, because the course of DCMP can be different and remains unpredictable. The data on the prevalence of DCMP in children are very different, which is due to the difference in methodological approaches, used research methods, the lack of specific diagnostic criteria.

The clinical symptoms of DCMP are identical to those in patients with heart failure of a different ethiology. Symptoms of heart failure may appear earlier than instrumental examination reveals DCMP, and symptoms of stagnation in the systemic circulation very quickly join the pulmonary stagnation. ECG changes in DCMP are nonspecific: a decrease in the voltage of the waves, a change in the ST interval and the T wave, often sinus tachycardia. The occurrence of atrial fibrillation is a prognostically unfavorable sign indicating the progression of the disease, and an extended QT interval is considered a predictor of mortality .

The most insidious form of cardiomyopathy in children is hypertrophic cardiomyopathy (HCMP). The clinical course of HCMP is very polymorphic: in some cases,

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the disease is asymptomatic, which is the reason for its late diagnosis; in others - with vivid clinical manifestations, due to hemodynamic disorders. Difficulties in the timely diagnosis of HCMP are due to the fact that in some cases the disease is detected by chance when examining children for systolic murmur in the heart. A feature of HCMP is sudden cardiac arrest, which may be the first and only manifestation of the disease. Heart rhythm disturbances are a characteristic attribute of HCMP. The spectrum of arrhythmias in children with HCMP is very diverse and it is the arrhythmic syndrome that determines the clinical and prognostic significance of the disease. Restrictive cardiomyopathy (RCMP) is a rare disease with a progressive course in childhood onset. The prognosis of RCMP is extremely unfavorable, since as a result of restrictive hemodynamics, severe diastolic dysfunction with heart failure, resistant to drug therapy, develops. Diagnosis of RCMP is a complex clinical task. Clinical manifestations, as a rule, are heterogeneous and ambiguous for an objective assessment. It is almost impossible to recognize RCMP only by complaints, since cardiac symptoms may be absent, and signs of muscle abnormality are detected only with targeted examination. Since the onset of symptoms of RCMP, the prognosis of the disease is poor. Non-compact cardiomyopathy (NCMP) is a new concept in cardiology and is currently isolated as a separate variant of cardiomyopathies. NCMP is a fairly common but poorly studied disease with a variety of clinical manifestations from asymptomatic to progressive systolic dysfunction with symptoms of heart failure and arrhythmic complications. Considering the achievements of modern instrumental research methods, there are no generally accepted criteria for the diagnosis of NCM in children, and there is no evidence to consider non-compact myocardium as a pathological condition, a normal variant of myocardial trabecularity, or a stage of progress of other cardiomyopathies.
Despite many years of research on the study of cardiomyopathies, this disease remains a serious problem in pediatric cardiology.

PURPOSE OF THE STUDY: to present the results of clinical features and the nature of cardiac arrhythmias in children with various forms of cardiomyopathy.

THE SCOPE AND METHODS OF RESEARCH: The work was carried out in the department of cardiology of the regional children's multidisciplinary medical center in the city of Andijan. 50 children in the age range from 3 to 16 years old were examined. In our studies by gender, the ratio of boys and girls was approximately the same - 26 (52%) and 24 (48%). The collection of information was carried out during a conversation with parents, and data from case histories were also studied. In each case, we adhered to a specific plan for collecting an anamnesis. The stages of heart failure were determined taking into account clinical criteria in accordance with the classification of N.A. Belokon (1987). We received confirmation of the diagnosis by performing an EchoCG and a standard ECG.

RESULTS. In our studies, we included children with various forms of cardiomyopathies: 30 children (60%) with DCM; 10 children (20%) - with HCM; 5 children (10%) - with RCMP and 5 children (10%) with non-compact cardiomyopathy. Of the total number of examined children, children with primary hospitalization accounted for 30%, the rest of the children had a history of two or more hospitalizations. In addition to the underlying disease, the cause of repeated hospitalizations was respiratory diseases, regardless of the form of cardiomyopathy.

Since there is no evidence base on the degree of influence of the pathological course of the anti- and perinatal periods on the formation of cardiomyopathy and heart rhythm, we analyzed this aspect of the issue. Regardless of the clinical forms of cardiomyopathy in all mothers the examined children had 2-3 or more risk factors during pregnancy and during delivery. The most significant were: anemia (100%), fetoplacental insufficiency (24%), threatened abortion (56%), intrauterine infection (28%), somatic diseases during pregnancy (76%). All children are born naturally, well-fed. Small newborns accounted for 24%. In all examined children, the general condition was assessed as severe.

According to the analysis of anamnestic data for the main share of children (63.6%), the development of DCMP was associated with a previous viral infection, and 68% of them are children from 3 to 8 years old. The severity of the condition upon admission to the hospital was associated with heart failure: stage II A - 40% of children and stage II B - 60%; at the same time, in 76.6% of children it is predominantly of the left ventricular type. The leading complaints of children with DCMP were: shortness of breath (at rest - 33%, with physical activity - 67%), attacks of obsessive cough (70%), feeling short of breath (60%), hepatomegaly (80%). We classified as clinical manifestations of DCMP: diffuse apical (73.7%) and heart atack (26.3%); expansion of the borders of the heart to the left (76.7%) and in both directions (23.3%); systolic murmur of varying

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intensity at the apex (100%). ECG signs: left ventricular hypertrophy (83.3%) with left atrial overload (16.7%); sinus tachycardia (100%); incomplete right bundle branch block (16.6%);AV blockade of I degree (13.3%) and II degree (10%); WPW syndrome (in two children); subendocardial ischemia of the left ventricular myocardium (43.3%).

Echocardiography revealed left ventricular dilatation with dysfunction in 50% of children. The range in terms of the expulsion fraction ranged from 35% to 53%. It was not possible to find out the reason for the formed HCMP during a conversation with parents. All children were admitted to the hospital in serious condition with clinical manifestations of heart failure (II B - 8 children; II A - 2 children). Dyspnea and cyanosis occurred in all examined children. 4 children (40%) of the surveyed had complaints of syncope, which, according to the mother, were provoked by stressful situations and emotional stress. Syncope states were manifested by a sharp pallor in younger children (up to 5 years old - 50%) in combination with vomiting and weakness; in children over 5 years old - dizziness and darkening in the eyes. Strengthening of the apical impulse was noted in 7 children; systolic murmur with the epicenter in the VI intercostal space to the left of the sternum was present in all children.

Among the features of the ECG we have noted: sinus tachycardia (8 children); sinus bradycardia (2 children); incomplete right bundle branch block (4 children); intra atrial block (2 children); WPW syndrome (in 2 children); subendocardial ischemia (in 2 children). Signs of left ventricular hypertrophy were diagnosed in all children; 6 children had asymmetric HCMP with obstruction of the left ventricular outflow tract. The asymmetry index in these children exceeded 1.4. The expulsion fraction ranged from 60% to 88%.

The leading complaints in all children with RCMP (5 children) were dyspnea, acrocyanosis, aggravated by physical exertion. All children were admitted to the hospital in serious condition. Symptoms of stage II A heart failure were diagnosed in 2 children, stage II B symptoms in 3 children, with severe hepatomegaly, and in one child, cervical vein swelling was visualized. From the physical data - a systolic murmur at the apex.

ECG - sinus tachycardia (100%); complete blockade of the right bundle branch block (in 3 children); AV blockade of the I degree (in 2 children); subendocardial ischemia (in 2 children); signs of left ventricular hypertrophy (1 child), right atrial hypertrophy (2 children). Echocardiographic signs of dilatation of the atria, with a predominance of the left atrium and not disturbed diastolic size of the left ventricle. Secondary mean systolic reversal of blood flow through the mitral and tricuspid valves (3 children); Echo signs of pulmonary hypertension in all children.

Among children with NCMP (5 children), 2 children were admitted to the hospital in serious condition. All children complained of shortness of breath associated with physical activity, weakness, palpitations. On the ECG, in addition to sinus tachycardia, one child was diagnosed with supraventricular paroxysmal tachycardia; atrial overload (in 2 children) and left ventricular hypertrophy (100%). EchoCG of all children revealed increased trabecularity of the left ventricular myocardium with areas of intertrabecular spaces and foci of compact and non-compact layers of the left ventricle. The ejection fraction ranged from 55% to 59%. EchoCG was diagnosed in all children with NCMP with a congenital heart defect - a defect of the interventricular septum.

CONCLUSIONS:

1. Our results showed that in the majority of children (52%) the cause

development of cardiomyopathies could not be established and this does not contradict the data of most researchers.

2. For all types of cardiomyopathies, there are many things in common.

clinical symptoms.

3. Regardless of the type of cardiomyopathy, the majority of children (76%)

were admitted in serious condition with clinical manifestations of stage II B heart failure (58%).

4. The leading complaints of children with cardiomyopathy were: shortness of breath

(100%), tachycardia (96%), cyanosis (100%), exercise intolerance.

5. Syncope conditions occurred in 4 children with a characteristic asymmetric form of hypertrophic cardiomyopathy.

6. Arrhythmic syndrome was present in all variants

cardiomyopathy. The most threatened cardiac arrhythmias: WPW syndrome (8%); supraventricular paroxysmal tachycardia (1 child); AV blockade of I degree (12%) and II degree (8%). Myocardial damage in 34% of cases was accompanied by subendocardial ischemia.

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