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MITRAL VALVE PROLAPSE AND ITS ROLE IN THE DEVELOPMENT OF CARDIAC AND EXTRACARDIAL DISORDERS FOR CHILDREN

Efimenko O.V.

Associate Professor of the Department of Hospital Pediatrics

at the Andijan State Medical Institute

Khaidarova L.R.

Senior Lecturer at the Department of Hospital Pediatrics at the

Andijan State Medical Institute

Summary: An analysis of modern data on the prevalence and causes of the formation of mitral valve prolapse in children is carried out. Cardiac and extracardial manifestations of mitral valve prolapse are described. The results of electrocardiographic and echocardiographic examination of children are presented. The most frequent phenotypic manifestations of connective tissue dysplasia in children with mitral valve prolapse were identified.

Key words: mitral valve prolapse, electrocardiographic and echocardiographic examination, connective tissue dysplasia, cardiac and extracardial manifestations, mitral regurgitation, myxomatic degeneration.

Relevance. Mitral valve prolapse is an urgent medical and social problem, being considered perhaps the most common outpatient pathology of the heart. Questions about the prevalence of mitral valve prolapse are still not fully clarified, including among children and adolescents. The prevalence of this pathology according to modern research, according to the Framingham Heart Study, is 2.4% and does not depend on gender and age. [2,5,9]

Unfortunately, there are still no clear generally accepted criteria for the diagnosis of mitral valve prolapse. However, in everyday practice, mitral valve prolapse is considered to be the deflection of one or both valves into the left atrium cavity during systole, regardless of the etiology of the disease. [1,3,10]

There are many theories in the literature explaining the origin of mitral valve prolapse, but there is no consensus among researchers. Some researchers emphasize the role of physiological stress in the development of mitral valve prolapse. Others consider mitral valve prolapse to be a pathology that develops due to imbalances between the size of the mitral valve and the left ventricle. A wide variety of vegetative vascular disorders in mitral valve prolapse, changes in the degree of flap prolapse, and even its disappearance over time point to the theory of functional disorders of the valve. There is also evidence that mitral valve prolapse is caused by weakness of connective tissue structures and mycomatous changes in the valvular apparatus of the heart, due to genetic disorders, namely, as a result of

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mutations in the filamin A (FLNA) gene and the DCHS1 gene responsible for the development of mitral valve prolapse. Many authors point to a violation of the metabolism of trace elements as the main reason for the development of mitral valve prolapse, and first of all, this is due to magnesium deficiency, the lack of which reduces the ability of fibroblasts to produce collagen. [6,7,10]

Thus, mitral valve prolapse is the result of a combination of genetic and acquired disorders leading to connective tissue dysplasia, leading to an elongation of the valves, their thickening and degeneration. [4,8]

Mitral valve prolapse is a pathology that not all doctors treat unambiguously, as there are still controversial issues regarding the assessment of its clinical significance. Isolated mitral valve prolapse is not a severe pathology of the heart, however, in cases of myxomatous degeneration of the mitral valve flaps, severe cardiac disorders often develop: heart failure, rhythm and conduction disturbances, and even sudden death. [3,6,9]

Cardiac symptoms associated with connective tissue dysplasia of the mitral valve are very often combined with extracardial manifestations, namely, episodes of arterial hypotension, fainting, prolonged weakness due to a violation of the autonomic nervous system. Since the connective tissue defect is generalized, in children with mitral valve prolapse, symptoms of connective tissue dysplasia from the musculoskeletal system, skin, eyes, etc. are also determined, manifested as a set of phenotypic signs of the body. [3,4,8,10]

The literature data indicate that mitral valve prolapse in children is a systemic anomaly that requires further study to clarify the nature of the disease, predict possible complications, and develop management tactics for children in a polyclinic. [5,7,9]

In this regard, our **goal** was to study the nature of cardiac and extracardial manifestations of mitral valve prolapse in children.

Materials and methods. The work was performed on the basis of the Regional Children's Multidisciplinary Medical Center in Andijan. 40 children with a verified diagnosis of mitral valve prolapse were under observation. The diagnosis of mitral valve prolapse was established after a comprehensive clinical, instrumental and laboratory examination. The complex of mandatory instrumental studies included: ECG (in 12 conventional leads to detect various rhythm and conduction disorders, the presence of hypertrophy of the heart chambers) and EchoCG (the examination was performed in 2D mode using Dopplerography). Mitral valve prolapse was diagnosed with a maximum systolic displacement of the mitral valve flaps beyond the line of the mitral valve ring by more than 2 mm. The thickness of the valves was measured in a diastole in their middle part, a thickening of more than 5 mm indicated their myxamatous degeneration. The length of the valves was also determined in diastole from the point of attachment to the mitral valve ring to the free edge. The degree of mitral regurgitation was assessed visually by the area of the blood stream.

The diagnosis of mitral valve prolapse was established with a combination of two main signs: auscultation and EchoCG. The presence of signs of dysplastic development of

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connective tissue structures was assessed by anamnesis, constitutional features, skeletal structure, skull, thorax, skin condition and joint mobility.

The results of the study. 40 children in the age range of 3-17 years were examined. The largest group consisted of school-age children (7-17 years old), and girls predominated by gender. (Table 1).

Table 1. Distribution of children by age depending on gender (n=40)

Paul	**Age**							
	3 – 6 years old		**7-9 years old**		**10-13 years old**		**14-17 years old**	
	Abs.	**%**	**Abs.**	**%**	**Abs.**	**%**	**Abs.**	**%**
Girls	**6**	15%	**9**	22,5 %	**8**	20 %	**4**	10 %
Boys	**2**	5 %	**3**	7,5 %	**8**	20 %	**-**	-
Total	**8**	20%	**12**	30%	**16**	40 %	**4**	10 %

The majority of the examined children (80%) were diagnosed with idiopathic mitral valve prolapse for the first time.

Based on the features of the perinatal history, we found that 85% of children were born from a pathological pregnancy and childbirth. Perinatal adverse factors were of great importance in the perinatal history of the examined children, among which pregnancy toxicosis (70%), the threat of miscarriage (30%), and maternal anemia (100%) should be highlighted.

Fetal and newborn asphyxia (30%) and impaired duration of labor (15%) were the most common of the intranatal risk factors.

According to the analysis of the somatic status, 25% of children are classified in the group of acute respiratory diseases.

Considering the most characteristic and common symptoms in the examined children with mitral valve prolapse, we identified both cardiac symptoms and extracardial manifestations of connective tissue dysplasia.

Cardiac symptoms were found in all children with mitral valve prolapse. **Table 2. Frequency of cardiac complaints in children with mitral valve prolapse.**

Complaints	N = 40

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	Abs	%
Cardialgia	10	25 %
Interruptions	3	7,5 %
Heartbeat	6	15 %
Feeling of lack of air	27	67,5 %
Shortness of breath during physical activity	4	10 %

The leading detection rates were cardialgia, palpitations, shortness of breath, and a feeling of lack of air. Complaints of pain in the heart area were made by school-age children aged 12-17 (35%). The pain was short-lived and stabbing. When analyzing the factors provoking pain in the heart area, such causes as stressful situations (31.1%) and overwork at school (38.9%) were identified. In 30% of children, cardialgia occurred spontaneously. The appearance of shortness of breath (55%) and palpitations (70%) in children with mitral valve prolapse was associated with physical exertion, and complaints such as the need to periodically take deep breaths, a feeling of lack of air, and dissatisfaction with breathing were found in 67,5% of all examined children with mitral valve prolapse. All children have a characteristic auscultation phenomenon.

Table 3. Frequency and causes of hyperventilation syndrome. (n=27)

Characteristics of hyperventilation syndrome	Abs	%
The need to take deep breaths periodically	8	29,7 %
Having a feeling of lack of air	14	51,8 %
Dissatisfaction with breathing	1	3,7 %
Shortness of breath during physical activity	4	14,8 %

All children underwent an ECG examination in 12 conventional leads. Analyzing the ECG results, various rhythm and conduction disorders were found in all children. Sinus tachycardia was most often diagnosed (35%), and sinus bradycardia was less common (18%). The analysis of the distal part of the ventricular complex in children with mitral valve prolapse revealed violations of repolarization (17,5%). Cardiac impulse conduction disorders in our studies are represented by incomplete blockage of the right leg of the Gis bundle (27,5%) and grade 1 atrioventricular block (15%).

Mitral valve prolapse in all examined children was determined on the basis of typical echocardiographic signs – the presence of sagging mitral valve flaps into the left atrium cavity by more than 2 mm. As the flaps deflected, the maximum group consisted of children with the first degree of prolapse, and the 3rd degree of prolapse was much less common.

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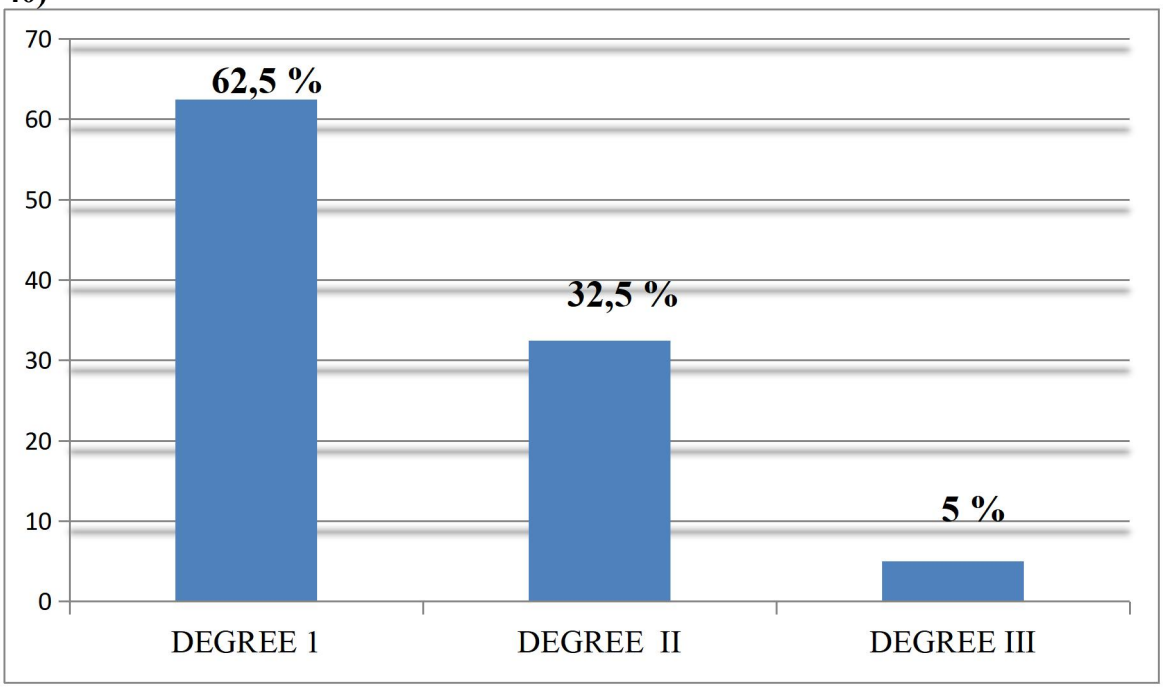
Figure 1. Distribution of children with varying degrees of mitral valve prolapse (n = 40)



By the time of occurrence, 35% of children had early prolapse of the anterior mitral valve leaflet; the remaining children had early prolapse, but of both mitral valve leaves. In two children aged 7 and 13, a thickening of the anterior mitral valve flap to 5 mm was found, which we regarded as their myxomatous degeneration.

Along with studying the structural features of the mitral valve in children with mitral valve prolapse, we evaluated mitral regurgitation. Mitral regurgitation was detected in all children with mitral valve prolapse, regardless of the degree of valve prolapse.

Of the 25 children with grade I prolapse, 20% had grade I regurgitation, and 80% had grade II regurgitation. In 13 children with grade II prolapse, 69,2% of children with grade II mitral regurgitation and 30,8% of children with grade III mitral regurgitation. Among children with grade III prolapse of the valves, grade II and III mitral regurgitation occurred with the same frequency.

Symptoms of vegetative dystonia were very characteristic in the clinical picture of children with mitral valve prolapse, represented by headache (77,5%) of varying intensity and localized mainly in the temples; a tendency to tachycardia (35%); excessive sweating (87,5%) and palm hyperhidrosis (47,5%).

The results of a physical examination of children with mitral valve prolapse showed a high frequency of various phenotypic signs of connective tissue dysplasia. The leading phenotypic manifestation of connective tissue dysplasia in our studies was joint hypermobility syndrome. The incidence of joint hypermobility syndrome among children with grade I (n=25) mitral valve prolapse was 76%; grade II (n=13) – 69.2% and grade III (n=2) – 50%. The joint hypermobility index ranged from 4 to 6 points. 1/3 of the children complained of joint pain, while there were no clinical signs of arthritis.

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The second most common phenotypic manifestation of connective tissue dysplasia was the asthenic type of constitution, established in 44% of cases among children with grade I mitral valve prolapse; in 46,1% - with grade II and in 50% of children with grade III mitral valve prolapse.

Among the rare phenotypic manifestations of connective tissue dysplasia among all children with mitral valve prolapse, we identified: funnel-shaped chest deformity (15%), cranial deformity (5%) and scoliosis in one child.

Conclusions. The use of modern echocardiographic criteria will make the diagnosis of mitral valve prolapse more specific. The examination algorithm for children with mitral valve prolapse must necessarily include an analysis of cardiac, extracardial manifestations and phenotypic signs of connective tissue dysplasia. Children with thickened valves should be classified as a high-risk group due to the possible development of life-threatening complications. These children should be examined at least once a year to assess changes in the cardiovascular system**.**

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