International Journal of Scientific Pediatrics
published: 28 February 2023
doi.org/10.56121/2181-2926-2023-2-2-47-50
Article / Original Paper

FREQUENCY AND CLINICAL MANIFESTATIONS OF MITRAL VALVE PROLAPSE IN CHILDREN ASSOCIATED WITH PHENOTYPIC MANIFESTATIONS OF CONNECTIVE TISSUE DYSPLASIA

O.V. Efimenko[1] **L.R. Khaidarova**[1]

1. Andijan State Medical Institute, Associate Professor, Andijan, Uzbekistan.

OPEN ACCESS
IJSP

Correspondence
O.V. Efimenko, Andijan State Medical Institute, Andijan, Uzbekistan.

e-mail: efimenkooksana1955@gmail.com

Received: 06 February 2023
Revised: 15 February 2023
Accepted: 22 February 2023
Published: 28 February 2023

Funding source for publication: Andijan state medical institute and I-EDU GROUP LLC.

Publisher's Note: IJSP stays neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Abstract. As an expression of a generalized connective tissue defect, mitral valve prolapse (MVP) is often combined with other dysplastic changes that manifest themselves as a set of phenotypic signs of the body. The aim of this study was to identify the frequency and clinical manifestations of mitral valve prolapse in children associated with phenotypic manifestations of connective tissue dysplasia. In the course of the work, not only clinical, but also instrumental methods were used to study 60 children with a verified diagnosis of mitral valve prolapse who are being treated in the department of cardiorheumatology of the Andijan regional Children's Multidisciplinary Medical Center. As a result of the work carried out, it was found that the high frequency of phenotypic manifestations of connective tissue dysplasia in children with MVP indicates that the diverse clinical symptoms are not limited only to the defeat of the cardiovascular system.

Key words. cardiovascular system, mitral valve prolapse, mitral regurgitation, connective tissue dysplasia, phenotypic manifestations, asthenic constitution.

Relevance. Mitral valve prolapse (MVP) is one of the most common small heart abnormalities and the most studied manifestations of connective tissue dysplasia (CTD). (1,4,7)

Connective tissue dysplasia is defined as an independent nosological form, polygenic-multifactorial genesis, characterized by genetic heterogeneity and relatively benign course, combined into syndromes and phenotypes based on the commonality of external and/or visceral signs. (4,5,8)

However, external phenotypic manifestations of connective tissue dysplasia are not always combined with dysfunction of internal organs, and vice versa, dysplastic changes in internal organs are not always associated with phenotypic signs of connective tissue dysplasia. (2,3,8)

Connective tissue makes up 50% of body weight and CTD is more common, less often local with a predominant lesion of any organ or system. (1,3,4,7)

CTD in pediatrics is characterized by high frequency, pronounced clinical polymorphism and complexity of diagnosis and treatment. Along with pronounced syndromic variants, there are many erased, transitional forms of CTD that significantly change the clinic and prognosis of CTD -associated diseases of internal organs. (4,5)

Among the variety of clinical manifestations of CTD in children, the most important place is given to changes in the cardiovascular system, where the leading place is occupied by MVP. (1,3,6,10)

In accordance with modern concepts, the basis of MVP is a genetically determined inferiority of collagen, which leads to a change in the mechanical properties of the mitral valve flaps and their ability to withstand normal pressure in the left ventricular cavity. Being an expression of a generalized connective tissue defect, MVP is often combined with other dysplastic changes that manifest themselves as a set of phenotypic signs of the body. (4,5,9)

The literature data indicate that MVP is one of the manifestations of connective tissue dysplasia that requires further study, due to the polymorphism of clinical manifestations and the complexity of diagnosis and treatment. (1,3,5,10)

Objective: to identify the frequency and clinical manifestations of mitral valve prolapse in children associated with phenotypic manifestations of connective tissue dysplasia.

Materials and methods. The work was carried out on the basis of the Regional Children's Multidisciplinary Medical Center of Andijan, Republic of Uzbekistan. There

were 60 children under observation with a verified diagnosis of mitral valve prolapse, of which 40 patients with MVP had phenotypic manifestations of CTD. The diagnosis of MVP was established after a comprehensive clinical, instrumental and laboratory examination. The complex of mandatory instrumental studies included: ECG (in 12 generally accepted leads for detecting various rhythm and conduction disorders, the presence of hypertrophy of the heart chambers) and EchoCG (the examination was carried out in 2D mode using Dopplerography). MVP 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 flaps 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 ring of the mitral valve to the free edge. The degree of mitral regurgitation was assessed visually by the area of the blood stream.

The diagnosis of MVP was established with a combination of two main signs: auscultation and echocardiography. The presence of signs of dysplastic development of connective tissue structures was assessed by anamnesis, features of the constitution, the structure of the skeleton, skull, chest, skin condition and joint mobility.

Results. In our studies, the age range of children with MVP was 3-17 years, with the largest proportion in the age group of 10-13 years (40%). By gender, there were twice as many girls as boys – 67.5% and 32.5%.

In connection with the multifactorial origin of MVP, we have studied possible risk factors that are likely to influence the formation of mitral valve flap anomalies.

From the features of the perinatal anamnesis, we found that children were born from pathologically occurring pregnancy and childbirth in mothers over the age of 30 years (82%). Perinatal adverse factors were of great importance in the perinatal history of the examined children, among which pregnancy toxicosis (95%), the threat of miscarriage (57.5%), anemia in the mother (100%) should be highlighted. Of the intranatal risk factors, fetal and newborn asphyxia (30%) and violation of the labor act in duration (15%) were the most frequent. According to the analysis of the somatic status, 25% of children were assigned to the group of frequently ill children for acute respiratory diseases.

During an external examination of the examined group of children with MVP, we noted the predominance of children with asthenic constitution as a phenotypic sign of connective tissue dysplasia, and in boys asthenic physique was observed more often than in girls.

The asthenic constitution can be explained by the heterochrony of the development of individual organs and systems, which is embedded in the genetic program of human ontogenesis, however, other genetic or external factors can cause significant individual deviations from a given program both in the intrauterine and postnatal periods, when some parts of the body or organs grow faster than others, or vice versa, growth selectively slows down.

However, regardless of the constitution of children, the leading phenotypic sign of connective tissue dysplasia in terms of the frequency of detection was joint hypermobility syndrome, which in 83.8% of cases was accompanied by a pronounced pain symptom. Arthralgia occurred most often in the knee joints (15%), in small joints of the arms and legs (17.5%) and in 3 children arthralgia was generalized. Joint pains were of a nagging nature, with a tendency to increase in the evening and with varying duration. Instrumental examination of this category of children with MVP revealed no signs of arthritis.

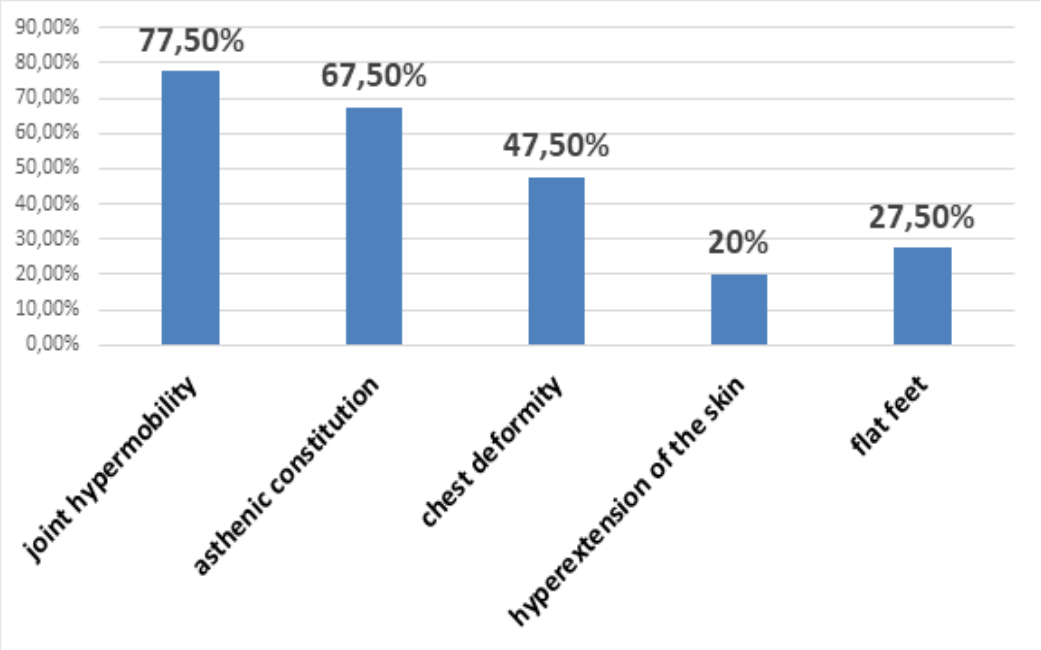
Of the other phenotypic manifestations of CTD in children with MVP, there were: chest deformity, hyperextension of the skin and flat feet. **(figure 1)**

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

Cardiac symptoms were found in all children with MVP. Leading in the frequency of detection were cardialgia, palpitations, shortness of breath, a feeling of lack of air. Complaints of pain in the heart area were made by school-age children 12-17 years old (25%). The pain was short-term, had a stabbing character. When analyzing the factors provoking pain syndrome in the heart area, such causes as stressful situations (30%) and overwork at school were identified (35%). 40% of children had cardialgia spontaneously. The appearance of shortness of breath (10%) and palpitations (15%) was associated with physical activity by children with MVP, and complaints such as the need to periodically take deep breaths, a feeling of lack of air, dissatisfaction with inhalation were found in 67.5% of all examined children with MVP. Cardialgia and palpitations were found with the

same frequency among both boys and girls, while the feeling of lack of air was 3 times more common in girls of the older age group.

Figure-1



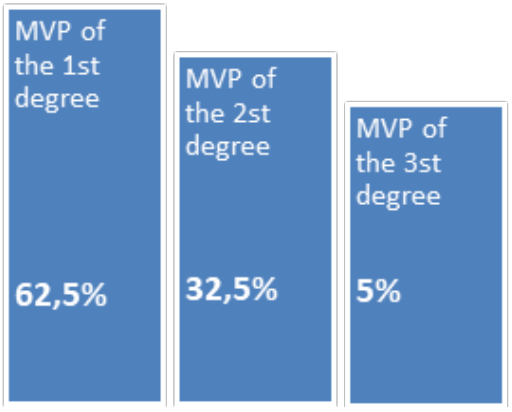
Frequency of phenotypic manifestations in children with varying degrees of MVP.

All children had characteristic auscultative manifestations of MVP in the form of isolated systolic noise (52.5%), isolated click (27.5%) and a combination of clicks with systolic noise (20%).

All children underwent an ECG study in 12 conventional leads. Analyzing the ECG results, 90% of children have various rhythm and conduction disorders. Sinus tachycardia is most often diagnosed (35%), less often sinus bradycardia (18%). Analysis of the distal part of the ventricular complex revealed repolarization disorders in children with MVP (17.5%). Supraventricular extrasystoles were found in 3 children (7,5%). Cardiac pulse conduction disorders in our studies are represented by incomplete blockade of the right leg of the Gis bundle (27.5%) and atrioventricular dissociation (15%).

Mitral valve prolapse in all examined children was determined on the basis of typical echocardiographic signs – the presence of sagging of the mitral valve flaps into the left atrium cavity by more than 2 mm. With the severity of the deflection of the flaps, the maximum group consisted of children with the first degree of prolapse; the second and third degrees of prolapse were much less common (Figure 2). According to the time of occurrence, 35% of children with early prolapse of the anterior flap of the mitral valve; 47.5% of children had late prolapse in the second half of the systole. In two children (5%) aged 7 and 13, a thickening of the anterior flap of the mitral valve to 5 mm was found, which we regarded as their myxomatous degeneration.

Figure-2



The frequency of the degree of prolapse of the mitral valve flaps

Along with the study of the structural features of the mitral valve in children with MVP, we evaluated mitral regurgitation. Mitral regurgitation was detected in all children with MVP examined by us, regardless of the degree of prolapse of the valves.

Of the children with MVP, 70% - with the I degree of regurgitation; 30% - with the II degree of regurgitation. Mitral regurgitation of the III degree in the process of echocardiography with Dopplerography was not detected among the subjects.

Comparing the frequency of phenotypic manifestations of connective tissue dysplasia depending on the degree of MVP, we found that asthenic type of constitution and hypermobility of joints with severe pain syndrome were more often detected among children with I degree of MVP. In children with grade II and III MVP, the leading phenotypic manifestations of CTD were chest deformity and flat feet.

Table-1

Phenotypic manifestations	MVP 1 egree	MVP 2 degree	MVP 3 degree
Constitution type: Asthenic Normosthenic	 76% 24%	 53,8% 46,2%	 50% 50%
Joint hypermobility syndrome: With arthralgia Without arthralgia	 80% 16%	 38,5% 7,7%	 50%
Chest deformity	24%	84,6%	100%
Hyperextension of the skin	28%	7,7%	-
Flat feet	16%	53,8%	50%

Frequency of phenotypic signs of connective tissue dysplasia in children with varying degrees of MVP

Conclusions. Thus, the high frequency of phenotypic manifestations of connective tissue dysplasia in children with MVP indicates that the diverse clinical symptoms are not limited only to the defeat of the cardiovascular system. The presence of certain connective tissue dysplasias in children should serve as a basis for conducting EchoCG to diagnose asymptomatic MVP.

LIST OF REFERENCES

[1] AFA Ibatova ShM, Abdulkadyrova NB. Prevalence of small heart abnormalities in children according to Echocardiography. International Journal of Scientific Pediatrics 2022:11–5.

[2] Zemtsovsky E.V. Connective tissue dysplasia of the heart. St Petersburg: Politex 2019:95.

[3] Muratkhodzhaeva A.V. Small anomalies of heart development in children and adolescents. Eurasian Bulletin of Pediatrics 2019;1:48–56.

[4] Kleimenov A.V. Mitral valve prolapse: clinical variants, modern concepts. Attending physician 2019:65–9.

[5] Mukhsinova M.Kh., Khuzhaeva F.S., Abduvakhidov Zh.Z. Small anomalies of the heart in children. Re-Health 2021:173–81.

[6] Grigoriev K.I., Solovyova A.L. Mitral valve prolapse in children: when opportunities create difficulties. Medical nurse 2021;23:19–28.

[7] Efimenko O.V., Khaidarova L.R., Khodzhamberdiev Sh.Kh. The nature of cardiac and extracardial manifestations of mitral valve prolapse in children. A new day in medicine 2020:669–72.

[8] Medrazhevskaya Ya.A., Kuleshov A.V., Malyk S.L. Cardiac arrhythmias in children with mitral valve prolapse and ways of correction. Medicine and Pharmacology 2019;6.

[9] Eidem B.W., Cetta F., O'Leary P.W. Echocardiography in Pediatric and Adult Congenital Heart Disease. Philadelphia 2019:500.