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MITRAL VALVE DISEASE IN CHILDREN AND ADULTS

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Abstract

Mitral valve / MV / is a functional complex that is based on the normal morphology, geometrical relations and function of all constituents. Its role is triple: 1. regulate blood flow to the LV during diastole with low pressure gradient, preventing the recurrence of systolic blood flow in LA, 2. contribute to the formation of LV outflow tract in systole and 3. its integrity is essential for maintenance of a normal size and geometry function of LV. The disease of heart valves, mitral and aortic caused by degenerative changes, is quite common in adults. In a young person, the more common causes are: inflammatory diseases, cardiomyopathy and ischemic heart disease. Using color Doppler echocardiography, the morphological valve changes and damage to its function can be diagnosed. The goal of treatment is: to reduce or eliminate symptoms of disease, to avoid damage to LV function or right ventricle, improving the quality of patient's life, prevention of complications and mortality reduction. Treatment options for valvular heart disease are: medical therapy, balloon valvuloplasty and surgical therapy: valve replacement or valve reconstruction. In children there are predominantly acquired valvular heart diseases of rheumatic etiology. Mitral stenosis is rare, more common is mitral regurgitation /MR/. Treatment of MR involves: SBE prophylaxis, afterload reducing agents, diuretics and digoxin. Surgical correction prefers reconstruction over MV replacement. Asymptomatic patients with MV prolapse do not require treatment, and symptomatic patients need beta blocker agents. Reconstructive surgery or MV replacement is indicated in rare patients with severe MR.

Key words: mitral, valve, disease

Mitral valve /MV/, located between the left atrium /LA/ and left ventricle /LV/, is a functional complex that is based on the normal morphology, geometrical relations and function of all constituents: the left atrium, the mitral annulus, the mitral leaflets, the subvalvular apparatus (tendinous chords and papillary muscles) and left ventricle. Its role is triple: 1. regulate blood flow towards the LV during diastole at low pressure gradient, while preventing systolic backflow in LA, 2. contribute to the formation of LV outflow tract during systole and 3. its integrity is essential to maintain normal size, geometry and function of LV. Normal mitral valve function depends on perfect function and complex interaction between various structures. The broader concept of “mitral valve complex” – mitral annulus, mitral leaflets, chordae tendineae, papillary muscles, left ventricular wall, left atrium – allows a better characterization of both normal and abnormal valvular function. It is important to recognize that the leaflets of the mitral valve constitute only a portion of the mitral valve apparatus and that diseases resulting in mitral dysfunction are often caused by abnormalities in the overall apparatus rather than in the actual leaflets (1).

During the phase of contraction, the mitral valve closes so that all the blood is ejected into the aorta. When mitral valve is diseased, valve can be stenotic, or not closing properly during blood ejection, which is then insufficient, and is called: mitral regurgitation. Early mitral regurgitation consists of: increased LV end diastolic pressure, increased LV contractility via increased preload, decreased or stable afterload due to filling of LA, preserved / hyperdynamic LV systolic function. Chronic compensate regurgitation has: dilatation of LV with hypertrophy, dilated LA with normal pressure, normal to hyperdynamic LV function. Chronic decompensated regurgitation is characterized by enlarged LV chamber and decline of LV function. Mitral stenosis used to be the main cause of mitral valve disease due to untreated streptococcal infections of the throat. Today, given the widespread use of antibiotics, mitral stenosis is increasingly rare, and is present in economically depressed areas (2). Insufficiency of the mitral valve - mitral regurgitation is now the most common form of mitral disease. It is mostly due to mitral valve prolapse generated on the basis of degenerative process in the mitral valve leading to elongation and cracking of tendinous chords that control valve movement. In mitral regurgitation, the valve does not close during the blood ejection phase, so only the portion of the blood goes into the aorta, and a portion is returned back into the left atrium and towards the lungs. Over time this leads to the heart load, and deterioration of the heart muscle, accompanied by fatigue,

suffocation and rhythm disorders. Mitral regurgitation may also occur with coronary artery disease and myocardial infarction. Infarction of left ventricular inferior wall usually leads to the so-called: ischemic mitral regurgitation.

Heart valve disease is quite common in adults (3-5). The most common diseases are of aortic and mitral valve, rarely tricuspid and pulmonary valve. The valve diseases include narrowing or stenosis and insufficiency or incomplete closure of the valve. In elderly patients, valvular heart disease is usually caused by degenerative changes, but in younger persons inflammatory bowel disease, cardiomyopathy, and ischemic heart disease are more common causes. A multiple simultaneous disease of the heart valves could be diagnosed in some patients. The most common symptoms that patients have are: dyspnoea or shortness of breath, feeling of increased heart rate or palpitations, intolerance to physical effort and faster fatigue, fainting, chest pain, and swelling of legs. A diagnosis of valvular heart disease is based on patient's history, clinical examination and diagnostic procedures including: electrocardiogram, X-ray of lungs and heart, transthoracic and transesophageal echocardiography, cardiac catheterization and coronary angiography, stress test - exercise test, magnetic resonance imaging and computerized tomography. The mitral valve was the first of the four cardiac valves to be evaluated by echocardiography (6-9). This was due to the relatively high prevalence of rheumatic heart disease and the relatively large excursion of the mitral valve leaflets, which made them easier targets for early M mode techniques. Using Doppler echocardiography and Color echocardiography, valvular morphological changes and their function damage can be diagnosed. This diagnostic method is used to estimate the severity of heart valves disease. Real-time-3 dimensional echocardiography of MV allows easy identification of different anatomical segments of MV including both commissures (10). The "en face" view of MV can be constructed and refers to exposure to the MV from the atrial perspective, similar to the surgeons view during MV surgery.

After clinical examination and diagnostic procedures, a decision is made on the choice of therapy (10, 11). The goal of treatment of diseases of the heart valves is to: reduce or eliminate symptoms of the disease, avoid damage to the functions of the left and right ventricle, improve the quality of patient's life, prevent complications and reduce mortality. Treatment options for valvular heart disease are: medical therapy, balloon valvuloplasty and surgical valve replacement therapy or surgical therapy for valve reconstruction. Following drugs are recommended: vasodilators, beta-blockers, ACE-inhibitors and for some groups of patients anticoagulant therapy (12). Balloon

valvuloplasty and percutaneous mitral commissurotomy are the way of treating mitral stenosis. Through peripheral artery, a catheter with a balloon is introduced and positioned to the place of narrowing of the mitral valve, then, by increasing the pressure and expanding the balloon, the stenotic area of the mitral valve is expanded (13). One of the possibilities of surgical treatment of valvular heart disease is valve repair which lately brings more benefits than the insertion of an artificial valve. By reconstructing the valve, the pathological changes in the mitral ring, or tricuspid valve, then the valve leaflets, the chordes tendineas or papillary muscles, can be corrected. Another option is surgical treatment for an artificial valve insertion which may be: mechanical, biological or homograft with the treatment of associated diseases such as coronary heart disease. Following insertion of the mechanical valve, patient should take lifelong anticoagulant therapy with regular control of coagulation parameters to determine the optimal dose of the drug. After the reconstruction of the valve or biological valve installation or bio prosthesis, such therapy is required in the first three months after surgery, while maintaining the target value (INR 2.5).

Acquired valvular diseases in children are mostly of rheumatic etiology. Involvement of MV occurs in $\frac{3}{4}$ and of aorta in $\frac{1}{4}$ of all cases of rheumatic heart disease. Stenosis and regurgitation of the same valve usually occur together. Mitral stenosis /MS/ is rare in children, because MS requires a period of 5-10 years from initial attack to develop. MS is the most common valvular disease in adult rheumatic population. Maintenance of dental hygiene with SBE prophylaxis is essential. In the case of atrial fibrillation development, anticoagulants are necessary with digoxin and quinidine as well. Balloon valvuloplasty was initially tried as an alternative to the closed commissurotomy – the results were comparable. MR is the most common valvular disease in children with rheumatic heart disease (14-16). Treatment includes SBE prophylaxis, afterload reducing agents, diuretics and in the case of atrial fibrillation (rare in children), digoxin. Surgical treatment prefers correction of the mitral valve rather than replacement due to lower mortality and the fact that anticoagulants are not necessary. Its use is necessary in case of the prosthetic valve. Mitral Valve Prolapse (MVP) occurs in 5% of children, in 50% of cases it is idiopathic (17). MVP is observed in primary form of autosomal dominant inheritance, and congestive heart failure occurs in 30% of cases. Thick and redundant mitral valve leaflets due to myxomatous degeneration bulge into mitral annulus. The posterior leaflet is more commonly and more severely affected than the anterior leaflet. Almost all patients with Marfan syndrome have MVP;

it may occur with diseases of connective tissue disorders. Asymptomatic patients do not require treatment or activity restriction. SBE prophylaxis is recommended in patients with MR or thickened mitral valve. Symptomatic patients (palpitations, dizziness, syncope) or those with arrhythmias, are subjected to 24-hour Holter ECG and/or stress test. Propranolol or other beta blocker is the drug of choice in patients with arrhythmias or chest pain. Reconstructive surgery or MV replacement is indicated in few patients with severe MR.

Although valvular heart disease (VHD) is infrequent in industrialized countries in relation to coronary artery disease, heart failure, or hypertension, guidelines are important in this area because the VHD are frequent, and often require interventions (18-20). The decision on intervention is complex because the VHD is often seen in the elderly age, resulting in a higher frequency of comorbidity, which contributes to increased risk of interventions. Another important aspect of contemporary VHD is the growing number of previously treated patients who are presented with further problems. In contrast, rheumatic valve disease remains a major public health problem in developed countries, where it affects mostly the younger population. When compared with other heart diseases, there are few studies in the field of VHD and randomized clinical trials are particularly rare. A large number of guidelines are issued in recent years by the European Society of Cardiology as well as by other societies and organizations. Because of their impact on clinical practice, quality criteria have been established to develop guidelines so that all decisions would be clear to the user. The relationship between clinical research, developing the guidelines and their implementation in clinical practice, can be complete only if surveys and registries confirm that daily practice is keeping in line with what is recommended in the guidelines. Such surveys and registries also make possible the calculation of impact of the implementation of recommendations on patient's outcome. The guidelines, however, do not cover the individual responsibility of health professionals to make the appropriate decision in the case of an individual patient, in consultation with that patient, and – where appropriate and necessary – with the person who takes care of the patient. It is also the responsibility of health professionals to uphold the rules and regulations applicable to drugs and devices at the time of prescribing. The objectives of the evaluation of patients with VHD is to diagnose, quantify and assess the mechanisms of VHD, as well as its consequences. Consistency between the results of diagnostic tests and clinical findings should be checked at every stage of the decision-making process (20-22). It would be ideal if the decisions are made by the “Heart team”, with special

expertise in the field of VHD, which includes: cardiologists, cardiac surgeons, imaging specialists, anesthesiologists and, if necessary, general praxis physicians, geriatric specialists, or intensive care specialist. If decision has to be made in paediatric patient, then the Heart Team consists of: paediatrician, paediatric cardiologists, imaging radiologists, intensivists, cardiac paediatric surgeons, anesthesiologists, psychologists, social workers and nutritionists. This approach, in the “Heart Team”, is particularly advisable in the management of high-risk patients, and it is important also for other subgroups, such as asymptomatic patients, where assessment of the possibility of reconstructing the valve is very important in decision making. Finally, indications for intervention – and what type of intervention should be chosen – is mainly based on the comparative assessment of spontaneous prognosis and results of intervention according to the characteristics of VHD and associated comorbidity.

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BOLESTI MITRALNOG ZALISKA KOD DJECE I ODRASLIH

Sažetak

Mitralni zalistak (MV) je funkcionalni kompleks koji se temelji na normalnoj morfologiji, geometrijskim odnosima i funkcioniranju svih konstituenata. Njegova uloga je trostruka: 1. reguliše protok krvi u LV tijekom dijastole s niskim gradijentom pritiska, sprečavajući ponavljanje sistoličkog protoka krvi u LA, 2. doprinosi formiranju odlivnog trakta LV u sistoli i 3. njegov integritet je od suštinskog značaja za održavanje normalne veličine i geometrijske funkcije LV. Bolesti srčanih zalistaka, mitralne i aortne, uzrokovane degenerativnim promjenama, uobičajene su kod odraslih. Kod mladih osoba, češći uzročnici su: upalne bolesti, kardiomiopatija i ishemijske bolesti srca. Korištenjem kolor dopler ehokardiografije mogu se dijagnosticirati morfološke promjene zaliska i oštećenja njegove funkcije. Cilj liječenja je: smanjiti ili eliminirati simptome bolesti, izbjeći oštećenja funkcije LV ili desne komore, poboljšati kvalitetu života bolesnika, spriječiti komplikacije i smanjiti stopu smrtnosti. Mogućnosti za liječenje bolesti srčanih zalistaka su: medicinska terapija, balonska valvuloplastika i kirurške terapije: zamjena zaliska ili rekonstrukcija zaliska. Kod djece nalazimo pretežno stečena oboljenja srčanih zalistaka reumatske etiologije. Mitralna stenoza je rijetka, dok je češća mitralna regurgitacija (MR). Tretman MR-a uključuje: SBE profilaksu, agense za smanjenje afterloada, diuretike i digoksin. Kod hirurških korekcija preferira se rekonstrukcija u odnosu na zamjenu MV-a. Asimptomatski bolesnici s MV prolapsom ne zahtijevaju liječenje, dok pacijenti s izraženim simptomima trebaju beta blokatore. Rekonstruktivna hirurgija ili zamjena MV-a se koriste kod rijetkih bolesnika s teškom MR.

Ključne riječi: mitralni, zalistak, oboljenje