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Mitral valve prolapse syndrome

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Abstract

Mitral valve prolapse is a common valvular defect. In its overt clinical form, the condition known as mitral valve leaflet prolapse syndrome, or also known as Barlow's syndrome or flaccid valvesyndrome, is a significant clinical problem, worsening the quality of life of patients, and in its severe forms is a risk factor for diseases such as congestive heart failure, endocarditis or cardiac arrhythmias, or even lead to death from sudden cardiac death.

The development of methods, as well as the increasing general availability of echocardiography methods, makes a specific diagnosis early enough to allow the right treatment strategy to be chosen. Thanks to reliable reports in recent years, it has been possible to identify a group of patients whose cardiovascular risk predisposes them to a more severe course of the disease and thus these patients require special observation and attention in the process of diagnosis and treatment.

Recent years have allowed the development of a new method of interventional treatment in patients burdened with multimorbidity for whom conventional surgery would be associated with a significant risk of worsening of the general condition.

Keywords: Sudden cardiac death, Mitral valve prolapse syndrome, Mitral valve prolapse

Mitral valve prolapse syndrome(MVP) this is a condition that occurs during ventricular systole in which one or two mitral valve leaflets are displaced into the left atrial lumen. If this phenomenon is clinically overt in the form of symptoms such as chest pain, shortness of breath, palpitations, dizziness, fainting, then we can talk about mitral valve leaflet prolapse syndrome (MVPS) (Barlow syndrome) these symptoms are dependent on autonomic system dysfunction, disorders in the renin-angiotensin-aldosterone system, as well as neuroendocrine disorders [5,7,8].

In patients affected by this syndrome, the morphology of the valve very often takes the form of thickened and elongated leaflets, i.e. changes characteristic of myxomatous degeneration. In the mechanism of development of this pathology, there is an accumulation of mucopolysaccharides in the connective tissue leading to fragmentation of collagen fibers and elongation of the leaflets and tendon fibers [8,9].

We can distinguish between two types of MVP: primary and secondary. Primary being a consequence of changes in the leaflets and tendon cords due to myxomatous degeneration may be related to familial occurrence, as confirmed by the identification of mutations and a site in chromosomes 16, 11 and 13 responsible for the genetic predisposition. We can identify classic mitral obturator leaflet prolapse with significant regurgitation and it is appropriate to investigate family members [8,9].

Secondary MVP in the course of connective tissue diseases such as Marfan syndrome, where a crucial role is attributed to the transforming growth factor TGF-beta responsible, in cooperation with metalloproteinases, for leaflet dislodgment, ischemic heart disease with subsequent rupture of the papillary muscle or acute endocarditis causing rupture of the stapes cord [5].

A significant proportion of patients also predispose to lesions within other valves. Where it is estimated that with the highest frequency because in about 40% of patients co-morbid lesions of the tricuspid valve leaflet. We will observe coexisting lesions of the aortic and pulmonary trunk valve leaflets much less frequently.

Epidemiology

The incidence of MVP in the general population was determined to be between 5% and 15%, but according to current criteria, this result should be estimated in the range of 2-3% with equal frequency in both genders [5, 18,20].

Clinical disease manifestation

Among the physical manifestations are typical auscultatory signs in the form of intrasystolic and/or late systolic clicks; however, their presence is not sufficient to make the diagnosis, but their absence on examination reduces the likelihood of MVP.

Among the subjective symptoms of MVP that make up the picture of the syndrome, we can distinguish non-specific chest pain, palpitations, dizziness, loss of consciousness, dyspnea unrelated to physical activity, weakness, anxiety attacks predominate [8,20].

Natural history

MVPS is characterized by a wide spectrum and we may expect to observe a mild form - characterized by far the highest frequency in the population - as well as rarer forms, moderate and severe, associated with a high risk of death.

If MVP is clinically silent or manifests itself as a mild form with a favorable echocardiographic picture, such patients require clinical follow-up every 3-5 years with assurance of a good prognosis. In patients with moderate and severe forms ofMVP, volume and pressure overload begins to impact left ventricular function also in the form of changes in adverse morphological remodeling of the myocardium. These changes occur at a rate dictated by the etiology, degree ofregurgitation as well as pre-existing organic heart changes. The early stages carry compensatory changes in the form of left ventricular enlargement, myocardial hypertrophy and the development of auscultatory phenomena in the form of a cardiac murmur [8].

A complication found relatively rarely in the course of MVP is sudden cardiac death having a potential connotation of supraventricular tachycardia, abnormalities in the ECG in the form of ST-T wave, The states particularly predisposing to the development of this dangerous complication are prolapse of both leaflets, excessive leaflet surface, fibrotic process involving the left ventricular muscle or papillary muscles [2,8,11].

Among other possible complications ofMVP, the authors also mention stroke or infective endocarditis, emphasizing, however, their controversial cause-and-effect relationship [6]. It has been estimated that the incidence of sudden cardiac death episodes is relatively low, ranging from 217 patients per 100,000 per year [16].

Patients with imaging findings of mitral annular dysfunction were characterized by an increased long-term incidence of clinical cardiac arrhythmias.

One of the significant problems inpatients with mitral valve leaflet prolapse syndrome is the life-threatening stress associated with the onset of complaints that may indicate a potentially life-threatening emergency condition. These situations result infrequent visits to health care facilities, abuse of medications and reduced physical activity. Therefore, a key role in improving the patient's quality of life is to ensure a good prognosis among patients who make up the vast majority ofMVPS patients and are not at risk of developing severe complications. Among this group of patients, a normal lifestyle and regular exercise are recommended [8].

Diagnosis

Only in some individuals, a basic, commonly available electrocardiogram (ECG) may suggest the need for further diagnostic testing with much more specific tests, while noting that most patients will not present any abnormalities on this test. In some patients, however, we may observe non-specific ST-T changes in the form of T-wave inversion, as well as QTc interval prolongation. These changes can be a valuable tool in the process of initial diagnosis and estimation of the risk of dangerous complications in a patient with MVP [2,3,8].

Diagnosis with ECG does not allow to make a specific diagnosis, but only in some cases to start expanding the diagnosis with tests of higher accuracy [8].

The main test for diagnosing MVP is echocardiography.

MVP in this examination is visualized as functional changes in the form of displacement of the edge of one or more mitral valve leaflets beyond the plane of the mitral annulus > 2 mm on the left atrial side during the phase of myocardial contractions using the most common positioning of the transducer in the parasternal projection in the long axis. Slight leaflet deflection less than or equal to 2 mm from the annular plane does not admit the diagnosis of prolapse; however, if features ofmyxomatous degeneration coexist, these features may indicate a genotype predisposed to mitral valve leaflet prolapse [4,6,8].

An even more accurate method that provides more precise localization of pathology than transthoracic examination is transesophageal echocardiography, which is recommended as a particularly useful method when considering cardiac intervention on the valve.

Transesophageal echocardiography can help identify abnormal positioning of the mitral valve leaflets in relation to surrounding structures and further identify mitral annular dysfunction and allows exclusion of infective endocarditis [6].

Ring dysfunction is associated with mitral valve leaflet prolapse and this pathology in itself is a marker for the risk of ventricular arrhythmias and sudden cardiac death, but the management and risk stratification of these patients needs to be systematized in the form of new guidelines to help make decisions [14].

A method currently increasing in popularity is cardiac imaging using magnetic resonance imaging. It may facilitate diagnosis and may have the added benefit of allowing detection of structural changes accompanying MVP such as fibrosis within the myocardial wall or the papillary muscle itself [7,11,19].

Pharmacological treatment methods

Treatment is not recommended inpatients without significant mitral regurgitation and significant myxomatous changes.

If these lesions do occur, however, there are no methods of treating the underlying cause. The strategy in such patients is based on prevention of complications and improvement of quality of life by reducing the occurrence of acute complaints such as chest pain, shortness of breath, palpitations, anxiety, etc.. To this end, the use of B-blockers is reasonable, the use of which will reduce the effects resulting from their inhibitory effect on the adrenergic system which excitation is one of the components of MVP syndrome.

Patients with orthostatic syncope in the course of MVPS, may benefit from the inclusion of more fluids and salt and mineralocorticosteroid use should be considered in more severe cases [8].

Among patients with an episode of transient cerebral ischemia, acetylsalicylic acid at a dose of 75-325 mg is recommended [8].

On the other hand, post-stroke patients with mitral regurgitation, atrial fibrillation or left atrial thrombus should take chronic anticoagulation with a vitamin K antagonist so that the INR is kept close to the value of 2.5. However, when it is mitral regurgitation with paroxysmal or persistent atrial fibrillation, one can reach for anticoagulants in the form of new oral anticoagulants [8].

There are no specific data on the efficacy of Beta-blockers in preventing sudden cardiac death; however, they are often used as first-line drugs because of their effect on combating premature ventricular excitation [21].

In view of the potential benefit to patients with severe arrhythmia and concomitant mitral valve leaflet prolapse, it is reasonable to undertake studies evaluating the efficacy of therapeutic intervention in the form of implantable cardioverter-defibrillators (ICDs), targeted ablation or surgical repair procedures for patients in this risk group [13,15,19].

The European Society of Cardiology guidelines recommend that when acute mitral regurgitation occurs, pharmacotherapy should use nitrates and diuretics to reduce filling pressures, while sodium nitroprusside in this case can be used to reduce afterload and regurgitation fraction. On the other hand, the use of an inotropic agent and an intra-aortic balloon pump can be used if hemodynamic instability or hypotension occurs.

On the other hand, if the course of the disease develops complications in the form of overt heart failure,it should be treated in accordance with current guidelines for this entity [1].

Invasive treatment methods

Such methods are for patients with severe symptomatic valvular regurgitation in a patient with MVP. As for asymptomatic patients, it is necessary to evaluate the benefit/risk ratio in deciding on a potential procedure [5].

Regarding symptomatic patients, both the American Heart Association and the European Society of Cardiology recommend valve surgery among patients with severe mitral regurgitation caused by MVP [6].

The ambiguous recommendations of the above-mentioned societies in relation to asymptomatic patients makes the decision about surgery should be made individually for each patient, while assessing the benefit-risk ratio, taking into account, among other things, comorbidities such as atrial fibrillation or pulmonary hypertension [6].

Reconstructive mitral valve surgery is associated with lower operative mortality, increased postoperative ejection fraction percentage, as well as better long-term survival and the possibility of reduced anticoagulant therapy compared to valve replacement surgery [6].

Patients who require predictably complex repair should be operated on at experienced repair centers with high repair rates, low operative mortality and durable outcomes. When repair is not possible, mitral valve replacement with preservation of the subvalvular apparatus is preferred [1].

A relatively new treatment method used among patients with high perioperative risk is percutaneous intervention, which involves mitral valve plication with an "edge-to-edge" technique where a cobalt-chromium leaflet clip is used [1,8].

Summary

MVP is a common valve pathology. There is a spectrum of disease, from isolated single leaflet prolapse to myxomatous, multi-lobe Barlow disease. Familial presentation is with several identified genetic loci. Echocardiography is crucial to diagnose and identify the severity of mitral regurgitation and the hemodynamic impact on the left ventricle. Although the great majority of patients have no clinical consequences, complications include progression of mitral regurgitation and infective endocarditis. In addition, the risk of sudden death and cerebrovascular incidents has been identified. In patients with significant mitral regurgitation, careful risk stratification is required to determine the optimal timing and method of intervention.

Disclosures

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