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MANAGEMENT OF MITRAL VALVE PROLAPSE IN PEDIATRIC POPULATION

Review Article

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Abstract

Mitral valve prolapse is a relatively common condition that associates various controversies with regard to therapeutic diagnosis and management. Generally, it is a benign pathology, although it is correlated with nonspecific symptoms such as precordial pain, dyspnoea or even panic attacks. This pathology can be randomly diagnosed following a routine cardiology check in both asymptomatic and symptomatic patients. The mitral valve prolapse is in most cases not progressive, but a minority of patients has the risk of developing mild mitral insufficiency, infectious endocarditis, or even sudden death. The clinician's responsibility is to identify low cases of complications as well as those with unfavourable development to initiate treatment.

INTRODUCTION

The mitral valve prolapse (PVM) consists in the prolapse of the mitral valve leaflets during the systole in the left atrium, with the occurrence of a phenomenon of regurgitation of a variable volume of blood.
The mitral valve prolapse (PVM) consists in the prolapse of the mitral valve leaflets during the systole in the left atrium, with the occurrence of regurgitation of a variable volume of blood.
In most cases, its progression is benign but the mitral valve prolapse may be complicated with mitral failure, infectious endocarditis, cerebral ischemic stroke or sudden death (Devereux, et al, 2011, Flack, et al, 1999, Freed, Levine & Evans, 1999, Pellerin et al, 2002).

CLASSIFICATION

It can be have two forms, namely, a classical and a non-classical form. The classic form is also referred to as mitral valve prolapse syndrome (SPVM), the lesions are not limited to the valvular apparatus but also extend pervalvularly to connective tissue in the tendinous chords and valvular ring.
It also correlates with extracardiac manifestations such as bone, connective tissue anomalies or neuropsychiatric manifestations. As for the non-classical form, the defect is limited to the valvular apparatus and does not involve systemic manifestations (Naggar et al, 1986, Rozmus & Fedorowski, 2002).

EPIDEMIOLOGY

PVM has become the most common valvular disorder in developed countries with a reported prevalence between 2-5% in different populations (Devereux, et al., 2001).

ETIOLOGY

Etiologically, mitral valve prolapse is mostly described as a primary pathology with dominant autosomal transmission (Devereux, et al., 1989).
However, there are situations where mitral valve prolapse appears as a secondary manifestation in connective tissue hereditary diseases such as *Marfan syndrome* (Pini et al., 1989), *Ehlers-Danlos syndrome* (Leier et al, 1980) *osteogenesisimperfecta* (Lebwohl et al., 1982), *pseudohanthomaelasticum or nodousperiarthritis* (Rozmus & Fedorowski, 2002).

PATHOPHYSIOLOGY

The pathophysiological mechanism of mitral valve prolapse is based on myxomatous degeneration of the valvular apparatus. It will cause the change in collagen structure of all elements of valve leaflets and in the structure of the chords (Rabkin et al, 2001, Tamura, et al, 1995). Tendinous chords involved in myxomatous valves contain an increased amount of glycosaminoglycans (Grande-Allen et al., 2001) leading to their weakening and eventual rupture. Chordal rapture is a common consequence of mitral valve prolapse due to mechanical weakness together with the hemodynamic stress caused by valvular tissue surplus (Barber et al., 2001).

CLINICAL PICTURE

The clinical picture described in the mitral valve prolapse is symptomatically poor, patients being diagnosed through auscultation clinical examination and abnormal mitral valve motility detected by echography and angiography. There are cases where complaints such as palpitation, dyspnea or anxiety-related symptoms have been described (Crawford et al, 2010, Ciomaga, et al., 2014). In isolated cases, syncopal or presyncopal episodes of orthostatic hypotension have been described (Devereux et al., 2001).
In clinical practice, PVM is often recognized by auscultation that is performed with the patient in a seated position and left lateral decubitus, the stethoscope being placed in supination.
The auscultation features are represented by a mesosystolic click of mitral valve closure and a telesystolic murmur of mistral regurgitation.
The occurrence of the click results from the sudden tension of the mitral valve apparatus when the mitral valve leaflet is prolapsed to the atrium cavity during the systole. The breath is moderate or high in intensity and is better perceived towards the apex due to turbulence caused by the volume of the regurgitated blood (Rozmus & Fedorowski, 2002).

PARACLINICAL EXAMINATIONS

To accurately determine the diagnosis of mitral valve prolapse, the following paraclinical examinations will be performed:
Echocardiography is currently the gold standard in the diagnosis of mitral valve prolapse in symptomatic patients or in patients with a suggestive family history due to good anatomical and functional visualization of the mitral valve (Rozmus & Fedorowski, 2002, Crawford et al, 2010).

The suggestive view of M-mode echocardiography consists of a posterior displacement of approximately 2 mm of one or both valve leaflets. 2D echocardiography is useful in the diagnosis of severe mitral valve prolapse anomalies (Rozmus & Fedorowski, 2002, Bonow, et al., 1998). These changes are accompanied by severe mitral regurgitation (Bonow, et al, 1998).

Electrocardiogram in most cases presents normal morphology but can also identify the presence of biphasic or negative T-waves in DII, DIII and aVF derivatives, and sometimes supraventricular or ventricular extrasystoles are recorded (Fauci, et al., 2003).

Cardiac MRI is useful in assessing the dimensions of the mitral ring, a matter of particular importance for patients requiring replacement of the valve.

It also provides information on the severity and volume of regurgitation (Christiansen et al, 2011).

Cardiac CT allows the visualization of adjacent anatomical structures, such as coronary sinus or coronary arteries but compared to MRI examination, it does not provide information on tissue characteristics (Durst & Gilon, 2015).

MANAGEMENT AND PROFILAXY

From the perspective of mitral valve prolapse management, the focus has been on keeping blood pressure and body mass index within normal limits, as well as on avoiding coffee, tobacco, alcohol or stimulant medication that may contain epinephrine or ephedrine (Crawford et al, 2010; Ciomaga et al., 2014).

According to the latest European Society of Cardiology Guidebook 2009, among patients receiving prophylaxis against infectious endocarditis, there have been mentioned specific selected cases such as operated patients or patients with valvuloplasty, patients with endocarditis history and with cyanogenic cardiac malformations, unoperated or corrected (Habib et al, 2009).

Revaluation by auscultation and echocardiography is recommended at moderate 5-year intervals to maintain the same risk group (Crawfordet al, 2010).

TREATMENT OF ARRHYTMIAS

The existence of arrhythmias requires the introduction of an antiarrhythmic treatment that in case of ventricular extrasystoles includes beta-blockers, namely, Bisoprolol (1.25-2.5/ mg/day) (Romanciuc & Revenco, 2012).

Supraventricular arrhythmias could be treated with Verapamil (0.1-0.3 mg / kg - no more than 5 mg - iv for 2 min, second dose should be no more than 10 mg, it may be administered after 30 min) (Medscape, 2013).

In case of recurrence of supraventricular tachycardia episodes, these will be stopped by means of ablation (Crawford et al, 2010).

TREATMENT OF CARDIOVASCULAR SYMPTOMATOLOGY

Treatment of cardiovascular symptoms in patients with anxiety, who may develop panic attacks and be susceptible to palpitations, may begin with Digoxin (10-17.5 mcg/kg) or beta-blockers (Romanciuc & Revenco, 2012).

TREATMENT OF AUTONOMIC DYSFUNCTION

The mitral valve prolapse may be associated in specific cases with the presence of an autonomic dysfunction syndrome manifested by episodes of vertigo resulting from syncope episodes or prolonged orthostatism. Their treatment includes the administration of Bisoprolol at the usual single dose of 1.25-2.5 mg/day (Romanciuc & Revenco, 2012).

SURGICAL TREATMENT

Surgical repair of the mitral valve is indicated in cases of severe regurgitation bringing the advantage of lower perioperative morbidity and mortality and better left ventricular function (Enache, 2011, Enriquez-Sarano, et al., 1995, Goldman, et al., 1987, Chesler et al, 1983). Patients with severe mitral insufficiency may also benefit from mitral valve replacement (Enriquez-Sarano, et al, 1995).

PRONOSTISAND COMPLICATIONS

The prognosis in children with PVM is very good, most of them remaining asymptomatic for many years. A limited number of patients may develop a number of complications over time.

Mitral insufficiency is the most common complication of mitral valve prolapse and results from progressive mitral regurgitation. It can be detected by Doppler echocardiography presenting a large regurgitation jet, dilated left ventricle and various anomalies of the valvular apparatus (Rozmus & Fedorowski, 2002).

Infectious endocarditis has a high incidence among patients with systolic murmur (Rozmus & Fedorowski, 2002).

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Cerebral ischemia associated with atrial fibrillation will require antiplatelet and anticoagulant therapy aimed to prevent embolic episodes. Smoking and consumption of oral contraceptives (Corrado, 2003) will also be avoided.
Sudden death has a low incidence and its production mechanism is still not fully understood (Chesler, King & Edwards, 1983).
Cases of sudden death have been reported in young athletes suffering from silent cardiovascular diseases, predominantly cardiomyopathy, coronary artery disease or Marfan syndrome (Corrado, et al, 2003,Markiewicz-Łoskot, et al, 2009).

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