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Floppy Mitral Valve Syndrome

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The floppy mitral valve syndrome (FMVS) could be considered as part of a well-recognized syndrome of heritable connective tissue disorders, such as Marfan syndrome, Elher-Danlos syndrome, adult polycystic kidney disease, Ebstein's anomaly, muscular dystrophy, Grave's disease, scoliosis. Not rarely floppy mitral valve (FMV) patients present some findings of Marfan disease, like thoracic deformities, long limbs, skin abnormalities and aortic dilatation, indicating "a phenotype continuum" [1-4]. Floppy tricuspid and aortic valves have been described [5,6].

From a genetic perspective, two forms of inheritance have been identified at the moment: the former is an autosomal dominant form with a variable degree of penetration (three gene loci have been identified: chromosome 16, 11 and 13), the latter is transmitted through the X-chromosome [7-11]. Prevalence of FMVS in general population is <1%. FMVS patients show a particular mitral valve anatomy, characterized by diffuse or regional thickening and redundant leaflets, with altered 2:1 ratio of the anterior to posterior surface area, elongated and/or ruptured chordae tendineae, annular dilation. As a consequence of these anatomic findings some changes occur over time in the global function of mitral valve apparatus: one, both or a portion of leaflets (usually scallop P2 and segment A2) extend above the atrio-ventricular plane during systole, initially during the mid- and late- phase and, over time, during all systole. Two effects occur: the prolapsing leaflet (or its portion) occupies part of the left atrium, creating a third chamber between mitral annular plane and prolapsing leaflet, in which an amount of blood flows during systole, reducing the effective left ventricular stroke volume; the prolapsing leaflet (or its portion) and annular dilatation can modify the leaflets apposition and coaptation, creating an effective regurgitant orifice and causing mitral regurgitation of various degree; in this way the left ventricular stroke volume decreases and the left atrial pressure increases. A myxomatous degeneration with mucopolysaccaryde infiltration, elastin fragmentation, collagen disruption and dissolution in the pars fibrosa of the leaflets and in the cordae tendineae have been demonstrated by histology in patients who had reconstructive mitral valve surgery for severe regurgitation and prolapse of posterior and/or anterior leaflets [12-14].

It is necessary a long-term follow-up and several clinical and echocardiographic evaluations to understand the natural life history of FMVS. The most common symptoms are palpitations, sensation of "a thumping heart", cardiac arrhythmias, syncope, dispnea, chest pain (not related to coronary artery disease), fatigue, difficulty breathing or shortness of breath, often when lying flat or during physical activity. In many cases these symptoms precede significant mitral valve regurgitation and are more common in women than in men. These symptoms seem related to neuroendocrine or autonomic nervous system functional abnormalities and to anatomic and functional mitral valve alterations [15-17].

Other symptoms can be attributed to complications such as progressive valve regurgitation, an acute and severe valve regurgitation, congestive heart failure, infective endocarditis, cardiac arrhythmias, tromboembolic events. The sudden appearance of severe mitral regurgitation and of pulmonary oedema in patient with known mitral valve disease is usually due to the acute rupture of the cordae tendineae and related hemodynamic consequences. Physical examination revealed in many patients' skeletal abnormalities such as pectus excavatum, scoliosis, a narrow anteroposterior chest diameter, an altered body height to weight ratio compared to normal subjects. On cardiac auscultation, a mid-telesystolic click alone or combined with mid-telesystolic murmur at the apex radiating to left sternal border or to axilla and/or spine, with typical changes with posture, is generally present. Echocardiography (M-mode, 2D, Doppler, Color, 3D) is the most widely used method for diagnosis and follow-up of FMVS, allowing the precise evaluation of mitral apparatus anatomy and function, LV and LA dimension and function, systolic pulmonary artery pressure in basal and stress condition, aortic dimension. In our experience 2D-TTE, performed by an experienced echo-lab, demonstrated a very good diagnostic accuracy in defining the anatomy and function of mitral apparatus, localizing the scallops/segments involved in degenerative mitral regurgitation, particularly for the middle ones (P2-A2), which represent almost the totality of prolapses [18]. More invasive, time consuming and expensive exams (TEE and/or 3D-Echo, Cardiac Magnetic Resonance, cardiac catheterization) should be reserved to selected cases addressed to surgery [19] or other repair techniques such as mitra-clip [20].

International Guidelines [21-23] clearly identified all conditions in which is recommended surgical repair or replacement with prosthetic valve or in which transcatheter mitral valve repair, using mitra-clip device, is preferable because of prohibitive surgical risk.

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