



REVIEW ARTICLE

# Floppy Mitral Valve (FMV) – Mitral Valve Prolapse (MVP) – Mitral Valvular Regurgitation and FMV/MVP Syndrome



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**Abstract** Mitral valve prolapse (MVP) results from the systolic movement of a portion(s) or segment(s) of the mitral valve leaflet(s) into the left atrium during left ventricular (LV) systole. It should be emphasised that MVP alone, as defined by imaging techniques, may comprise a non-specific finding because it also depends on the LV volume, myocardial contractility and other LV hemodynamics. Thus, a floppy mitral valve (FMV) should be the basis for the diagnosis of MVP. Two types of symptoms may be defined in these patients. In one group, symptoms are directly related to progressive mitral regurgitation and its complications. In the other group, symptoms cannot be explained only by the degree of mitral regurgitation alone; neuroendocrine dysfunction has been implicated for the explanation of symptoms in this group of patients that today is referred as the FMV/MVP syndrome. When significant mitral regurgitation is present in a patient with FMV/MVP, surgical intervention is recommended. In patients with a prohibitive risk for surgery, transcatheter mitral valve repair using a mitralclip device may be considered. Furthermore, transcatheter mitral valve replacement may represent an option in the near future as clinical trials are underway. In this brief review, the current concepts related to FMV/MVP and FMV/MVP syndrome will be discussed.

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## I. Introduction

Few diseases have evoked more interest and controversy during the previous century than floppy mitral valve (FMV)/mitral valve prolapse (MVP). These controversies are related, at least in part, to the following four reasons.<sup>1,2</sup>

1. There is a lack of a precise definition for FMV/MVP. MVP occurs when one, both, or a portion of the mitral valve leaflets extend above the plane of the atrioventricular junction during left ventricular (LV) systole.<sup>1–6</sup> However, it should be noted that MVP may be a non-specific finding because it also depends on LV haemodynamics, such as myocardial contractility, LV volume, and heart rate.<sup>3,6</sup> Furthermore, the evolution of technology over the years has resulted in various definitions of MVP. For a long period of time, MVP was the central or only issue, and the diagnosis of MVP was completely dissociated from the FMV morphology.<sup>3,6</sup> However, to date, it is apparent that FMV is the central issue in the MVP - mitral valvular regurgitation story. The term FMV originates from surgical and pathological studies and refers to the expansion of the area of the mitral valve leaflets with elongated chordae tendineae, chordae tendineae rupture, and mitral annular dilatation. Thus, rather than asking the question does this individual have MVP, the question should be does this individual have FMV?<sup>2,3,6?</sup>

2. The natural history of FMV/MVP and the occurrence of mitral regurgitation requires long-term follow-up with several evaluations to understand the life history of the disease. The follow-up of an uncomplicated course during a relative short period of time has resulted in the misconception that FMV/MVP is benign.<sup>2,7,8</sup>

3. A subgroup of patients with FMV/MVP may have symptoms that are not directly related to the severity of mitral regurgitation, but rather neurohumoral activation and other abnormalities (subsequently discussed); these patients today are referred to as having the FMV/MVP syndrome. The lack of an association between the severity of mitral regurgitation and symptoms was also a cause of confusion for a long period of time.<sup>1,3,8</sup>

4. FMV/MVP may be a part of a well-recognised syndrome of heritable connective tissue disorders, such as Marfan syndrome, Ehlers-Danlos syndrome, and adult polycystic kidney disease.<sup>9–12</sup> The fact that clinicians were often not able to separate isolated FMV/MVP from FMV/MVP that was a component of a systemic heritable connective tissue disorder was also a cause of confusion. Furthermore, FMV/MVP often shares several manifestations of Marfan syndrome, including long limbs, thoracic cage deformities, striae atrophicae, and, in some cases, mild dilatation of the aorta or abnormal aortic function. The acronym MASS (mitral, aorta, skeleton, skin) was introduced to emphasise the involvement of the mitral valve, aorta, skeleton and skin.<sup>12</sup> Thus, the clinical phenotype of FMV/MVP represents a heterogeneous group of patients with mitral valve or other abnormalities from mild to severe and constitutes a continuum from Marfan syndrome at one extreme to isolated FMV/MVP at the other extreme. To date, isolated FMV/MVP is considered a cardiovascular abnormality with a connective tissue origin, which, in most instances does not fit the presently recognised heritable connective tissue disorders.<sup>2,9,12</sup> By

virtue of its high frequency in the general population, FMV/MVP comprises a larger group of patients with a connective tissue abnormality of the heart. In this brief review, the current concepts related to FMV/MVP and FMV/MVP syndrome will be discussed.

## 2. Inheritance

From a genetic perspective, FMV/MVP is a heterogeneous group.<sup>2,3,13–17</sup> To date, at least two forms of inheritance exist. FMV/MVP may be transmitted by an autosomal dominant form (most common) with a variable degree of penetration. Another less common form is transmitted through the X-chromosome (chromosome Xq28).<sup>16</sup> For autosomal dominant transmission, three gene loci have been reported: chromosome 16 (16p12.1-p11.2); chromosome 11 (11p15.4); and chromosome 13 (13p31.3-p32.1).<sup>15</sup> The elucidation of the genetic details in the near future will enable a better classification of this common valvular abnormality and will result in novel diagnostic pathways.

## 3. Pathology - Histopathology

1. *Pathology.* Surgically excised mitral valves from patients with FMV/MVP and significant mitral valvular regurgitation have a substantial surface area on both leaflets (18–25, Fig. 1). The typical 2:1 ratio of the anterior to posterior leaflet surface area is altered because of the enlargement of all portions of the posterior leaflet. The mitral annular size is also increased, and the chordae tendineae are frequently thin, elongated or ruptured.<sup>18–20</sup> Pathologic studies have suggested that prolapse is limited to the posterior leaflet in 67% of cases, the anterior leaflet in 10% of cases, and both leaflets in 23% of cases.<sup>20</sup> In a recent study of 98 patients with FMV/MVP who had reconstructive mitral valve surgery for severe mitral regurgitation, prolapse of the posterior leaflet was identified in 53 patients, the anterior leaflet in 4 patients, and both leaflets in 41 patients.<sup>26</sup> Mitral valves with diffused thickening are currently referred to as Barlow's valves, whereas regional thickening of the mitral valve leaflets are referred to as fibroelastic deficiency valves. To date, it is not clear whether these two entities are genetically different or represent a different spectrum of abnormalities of the same disease (or both). For this reason and to avoid further confusion, we believe that it is better to define patients with FMV/MVP as having diffuse or regional thickening of the mitral valve leaflets.<sup>3</sup>

2. *Histology.* The gross pathologic changes occur in the presence of histological evidence of myxomatous degeneration, collagen disruption and dissolution, mucopolysaccharide infiltration, and elastin fragmentation.<sup>18–24</sup> The most specific fundamental and characteristic changes appear to comprise collagen dissolution and disruption in the pars fibrosa of the mitral valve leaflets, which is accompanied by proteoglycan accumulation. Similar histological abnormalities have also been demonstrated in the chordae tendineae.<sup>24</sup> Continuous pressure and stress during LV systole on the mitral valve leaflets and chordae tendineae contribute to the gradual progression of these histological abnormalities. In some cases, similar histological

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