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Mitral valve surgery: Does it really decrease ventricular arrhythmia in patients with mitral valve prolapse?

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Mitral valve (MV) prolapse (MVP) is characterized by fibromyxomatous changes in the mitral leaflet tissue, with superior displacement of one or both leaflets into the left atrium. It is not an uncommon finding on echocardiographic screening, afflicting 2–3% of the general population, though most of them remain asymptomatic [1–3]. MVP can be distinguished into primary (non-syndromic) and secondary (syndromic) MVP. Secondary MVP occurs in the presence of connective tissue disorders such as Marfan syndrome, Loeys-Dietz syndrome, Ehlers-Danlos syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum, and aneurysms-osteoarthritis syndrome. MVP has also been observed in hypertrophic cardiomyopathy (HCM) and may contribute to the pathophysiology of obstruction [1].

MVP is generally regarded as a benign condition [4–6], however, the outcome is widely heterogeneous, and its manifestations such as mitral regurgitation (MR), atrial fibrillation, congestive heart failure, endocarditis, and stroke are well known. Cardiac mortality is best predicted by the presence of mitral regurgitation (MR) and left ventricular dysfunction at the time of diagnosis. Risk factors for cardiac morbidity include age $\geq$50 years, left atrial enlargement, MR, the presence of a flail leaflet, and atrial fibrillation [4,7–11]. MR can occur due to a spectrum ranging from single prolapsing valve segment to diffuse myxomatous degeneration with bileaflet prolapse and annular dilatation. Degenerative mitral valve disease, most commonly related to MVP, is the most repairable form of surgical mitral valve disease, and repair is the most recommended surgical approach and represents 60–70% of surgical mitral regurgitation (MR) in industrialized nations [12–15].

Cardiac arrhythmias are frequently detected in patients with mitral valve prolapse (MVP) [16,17]. Atrial ectopics, couplets, atrial tachycardia, paroxysmal or sustained atrial flutter or fibrillation as well as ventricular premature contractions (VPCs), multiform VPC, VPC couplets, and runs of three or more sequential VPCs (salvos of ventricular tachycardia) have been described to occur in presence of MVP. The proper mechanisms causing atrial and ventricular arrhythmias in patients with MVP have not been fully investigated, though arrhythmias correlate with age, female gender, presence of MR, left atrial diameters, left ventricular end-diastolic diameter, anterior mitral leaflet thickness and bileaflet prolapse in various studies [4,16–20]. Sudden cardiac death (SCD) has also been reported in patients with MVP [21].

From a pathophysiological perspective, the mechanism of ventricular arrhythmias in patients with MVP with trivial or absent mitral regurgitation remains speculative [21,22]. MVP-related factors have been advocated such as the excessive traction on the papillary muscles by the prolapsing leaflets, the mechanical stimulation of the endocardium by the elongated chordae with afterdepolarization-induced triggered activity, the diastolic depolarization of muscle fibers in redundant leaflets with triggered repetitive automaticity, and the endocardial friction lesions with extension into the myocardium [23–25]. The coexistence of extravalvular diseases has been suggested, including autonomic nervous system dysfunction [26], conduction system abnormalities [27], fibromuscular dysplasia of small coronary arteries [28], and occult cardiomyopathies [29,30].

Cardiovascular magnetic resonance imaging (CMRI) can not only identify MVP by similar anatomic and functional criteria, but in addition, CMRI can identify myocardial fibrosis involving the papillary muscle in MVP patients. Delayed contrast enhancement (DCE) of papillary muscles, indicative of fibrosis, which may act as a focus for arrhythmias, is often present in a subgroup of patients with complex ventricular arrhythmias [17,31–33]. Mitral annulus disjunction is a feature of arrhythmic MVP with LV fibrosis. The excessive mobility of the leaflets caused by posterior systolic curling accounts for a mechanical stretch of the infero-basal wall and papillary muscles, eventually leading to myocardial hypertrophy and scarring [34].

Naksuk N et al. [35] have retrospectively described effect of mitral valve surgery (predominantly mitral valve repair) on pre-existing ventricular arrhythmia in patients having MVP. They conclude that mitral valve surgery does not uniformly reduce VPC frequency in patients with bileaflet MVP and patients who have at least a 10% reduction in overall VPC burden tend to be younger than those who do not. They therefore argue that MVP related mechanical stress on the papillary muscles is unlikely a cause of arrhythmia. Intervention at an earlier age decreases the structural changes related to MVP and MR, and therefore may decrease the arrhythmia post operatively in younger age groups.

MVP and mechanical stress/papillary muscle scarring cannot explain the presence of VPCs arising from other than the papillary

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muscle apparatus, which were also seen in this study. Presence of idiopathic outflow tract ectopics may be unrelated to MVP or can be a manifestation of an occult cardiomyopathy. This association remains undetermined presently. Secondary MVP associated with various connective tissue disorders can also result in cardiac connective tissue changes, resulting in possible arrhythmic substrates from different myocardial sites. Therefore, mitral valve surgery will not be able to decrease ventricular arrhythmia in this scenario. However, no increase in incidence of ventricular arrhythmia with secondary MVP has been observed. Though some centres have also described a possible reduction in ventricular arrhythmias after MV surgery [36–38], the results of surgery have not been encouraging in the study by Naksuk et al. in this aspect.

Management of low risk symptomatic/asymptomatic ventricular arrhythmia needs to be conservative with or without medications. Risk assessment and frequent monitoring should be done in such patients. CMRI can help to understand the structural abnormality associated with ventricular arrhythmia. High risk, drug resistant, symptomatic ventricular arrhythmia should undergo electrophysiology study and radiofrequency ablation. Rarely an automated implantable cardiac defibrillator implantation is required for prevention of SCD due to malignant ventricular arrhythmia related to MVP.

Currently the etio-pathogenesis of ventricular arrhythmia in MVP is not fully understood and also the role of mitral valve surgery to decrease ventricular arrhythmia is not proven. Prospective studies need to be done to establish the role of surgery in managing ventricular arrhythmia in patients with MVP. Further insights into the genesis of arrhythmia in future would one day provide targeted therapy in such patients of MVP having ventricular arrhythmia.

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