Brief Review

Mitral valve prolapse

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Mitral valve prolapse (MVP) was first described as a clinical–pathological entity in the mid 1960's[1,2] when the auscultatory findings of a mid-systolic click were correlated with angiographically demonstrated valve prolapse. Prior to that time, these findings were thought to be caused by extra-cardiac pathology. Since the advent of echo-cardiography, this entity has been increasingly recognized and diagnosed.

Incidence

Mitral valve prolapse is the most common valvular cardiac abormality with published prevalence rates ranging from five per cent to 20 per cent.[3–6] The ongoing Framingham study[6] has sought to determine the prevalence and features of mitral valve prolapse in the general population. The incidence of mitral valve prolapse in men was a constant two to four per cent in all age groups; however, in women it varied with age. The incidence in women aged 20–29 was 17 per cent but declined with each subsequent decade to 1.4 per cent in women over 80 years of age[3] (see Figure). The reason for the age-related decline in women remains unclear.

Aetiology

The aetiology of mitral valve prolapse is unclear. There is a definite familial occurrence of MVP and in some kindreds it follows an autosomal dominant patern of inheritance.[9] There is also an association with certain other congenital heart diseases (Table I) although these are rare occurrences.[10] MVP is also associated with certain skeletal and connective tissue abnormalities such as the straight back syndrome. Finally, there may be an embryologic aetiology suggested by the incidence of MVP associated with tricuspid, thoracic skeletal and dermatologlyphic abnormalities all of which develop during the fifth to sixth gestational week.

Pathophysiology

The principal anatomic and histologic abnormalities found in the mitral valve in this syndrome are redundancy and myxomatous degeneration of the leaflets, dilatation of the mitral valve annulus and variable chordal pathology (both thinning and elongation of the chordae, as well as shortening).[9,10] Normally, the mitral valve remains in a subannular position during ventricular systole. However, in this syndrome, the redundant leaflets act as an "unfurled sail" that catch the "wind" of ventricular systole and prolapse into the left atrium. This subjects the valve leaflets, the chordae, the papillary muscles and the underlying ventricular wall to abnormal and destructive stresses. The valvular endothelium may be traumatized providing a nidus for sterile vegetations or for bacterial endocarditis. There is no histological evidence of cardiomyopathy, and coronary arteries tend to be normal, although there may be an association with coronary artery spasm in these patients.

Functionally, the mitral valvular apparatus may be thought of as being "too big" for the ventricle or, inversely, the ventricle is "too small" for the valve. Factors which increase left ventricular volume tend to improve the prolapse. The prolapse occurs later during systole, is less severe and has less mitral

TABLE I Congential abnormalities associated with mitral valve prolapse

Ostium secundum atrial septal defect
Hypertrophic cardiomyopathy
Marfan's syndrome
Ehlers-Danlos syndrome
Ebstein's anomaly of the tricuspid valve
Turner's syndrome
Wolf-Parkinson-White syndrome

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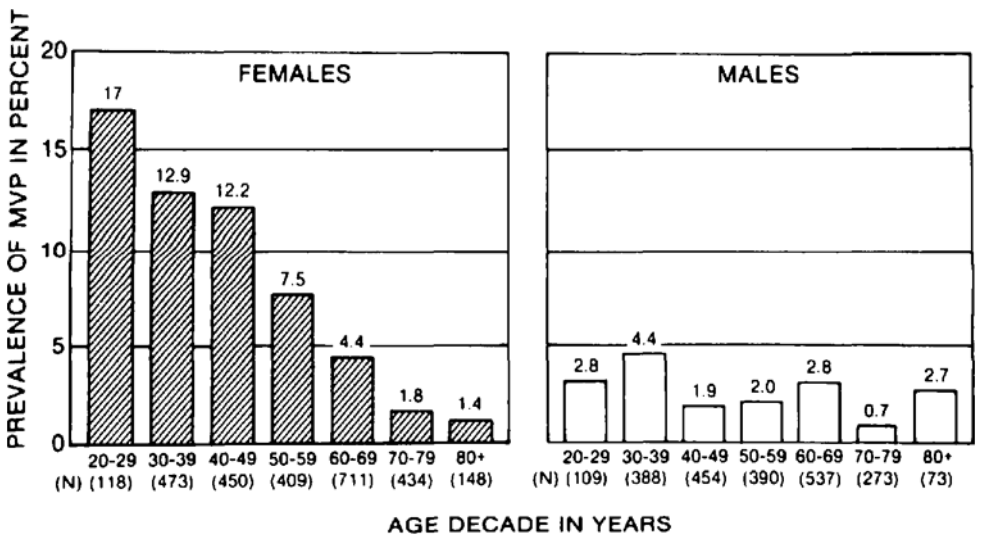

FIGURE Age and sex related prevalence of mitral valve prolapse. From Savage DD, *et al.*[6] Mitral valve prolapse in the general population. 1. Epidemiologic features: the Framingham study. Am Heart J 1983; 106: 571–6. Used with permission.

regurgitation. Thus, anything that increases afterload or decreases contractility such as squatting, leg raising, administration of vasoconstrictors or of beta adrenergic receptor blocking agents, will diminish the prolapse. Conversely, anything that decreases left ventricular volume (e.g. standing, peripheral vasodilatation or afterload reduction) will cause the prolapse to occur earlier and to be more severe.

Clinical features

The vast majority of patients with MVP are asymptomatic.[7,9] Patients may complain of symptoms of palpitations, atypical chest pain, syncope or fatigue but these symptoms are no more common than in patients without MVP.[7] On physical examination individuals with MVP are taller and thinner than average, and may have associated findings such as high arched palate, thoracic skeletal abnormalities (straight back syndrome, pectus excavatum, scoliosis) or hyperextensible joints. Auscultation reveals the classic mid to late systolic click. The pathognomonic feature of the click is that it is predictably mobile. As previously stated, factors that decrease left ventricular size cause the click to occur earlier in systole and factors that increase left ventricular size cause it to occur later. Murmurs can be heard, either a late systolic murmur, beginning with the click or a holosystolic one. Again, the late systolic murmur will occur earlier and be louder when left ventricular afterload is reduced.

The vast majority of patients have normal ECG's; however, a small percentage have ST segment changes and/or T wave flattening or inversion.[7] These are most commonly found in the inferior leads (II, III, aVF) and may be induced either with exercise or inhalation of amyl nitrate. Dysrhythmias may also occur; the most common being supraventricular tachyarrhythmia, and premature atrial or venticular contractions (PAC or PVC). Bradyarrhythmias have also been associated with MVP.[8]

The principal method of diagnosing MVP is by means of 1-D or 2-D echocardiography, showing the characteristic late systolic prolapse of the mitral valve. Interestingly enough, the Framingham study found that in people with echocardiographically proven MVP, only a minority had auscultatory findings. Clicks were found only in 13 per cent of males and eight per cent of females with MVP and

TABLE II Complications

Arrhythmias
Sudden death
Embolic phenomena (cerebral vascular accidents)
Mitral insufficiency
Chordal rupture
Bacterial endocarditis
Dissection of the aorta

murmurs only in three per cent of males and eight per cent of females.[7] Conversely, a click does not indicate mitral valve prolapse. Only 50 per cent of subjects with systolic clicks had evidence of prolapse.[7]

Natural history and complications

The information available about the long-term prognosis of MVP is rather sketchy and influenced by the biases of case selection in each study. There are a number of serious complications that have been reported to be associated with this entity (Table II).

The exact incidence of these complications is not known. The Framingham study found 50 per cent of subjects with MVP had multiform or repetitive PVC's and 25 per cent had runs of PVC's. However, 42 per cent of controls also had multiform or repetitive PVC's, which is not statistically different.[8]

Information from long-term follow-up varies with the type of study. A British study of 62 patients with MVP who had no prior complications except PVC's, followed these patients for from 9–22 years.[12] Five developed infective endocarditis, one developed chordal rupture and one died of mitral regurgitation at age 75. In an American study of 53 patients followed for ten to 22 years, three developed infective endocarditis (one died), five developed significant mitral regurgitation, one patient died suddenly and one was resuscitated after spontaneous ventricular fibrillation.[13] In another series based on postmortem findings of a floppy mitral valve, 102 out of 1,376 cases had evidence of mitral valve prolapse.[10] Of these patients, there were no cases of sudden death, seven cases of endocarditis, one case with a mycotic cerebral embolus. Of interest, is the fact that the average age of death of patients with MVP was greater than that of controls.

Regarding surgery or dental procedures, most authors suggest that patients with MVP are at risk for developing endocarditis and should receive antibiotic prophylaxis.[9,14,15] Clemens *et al.*, in a retrospective study, concluded that patients with MVP have a substantially higher risk of developing endocarditis than normals and that in patients with prolapse *and* a murmur, this risk is even higher.[11]

Mitral valve prolapse and anaesthesia

The anaesthetist will meet patients with MVP in one of three probable situations. One is the patient with known MVP presenting for surgery. Secondly, is the asymptomatic patient in whom a click and/or murmur is discovered on preoperative examination. Finally, is the patient who presents under anaesthesia with unexplained dysrhythmias; either supra-ventricular tachyarrhythmias, ventricular dysrhythmias or occasionally bradyarrhythmias.

Patients with known MVP may be receiving a variety of medications. Beta blockers are the most common medication to treat both atypical chest pain as well as ventricular or supra-ventricular arrhythmias. They may also be receiving any of a large number of antiarrhythmics. These medications should be continued up to and including the day of surgery. If a patient has had a cerebral vascular accident secondary to MVP, he may be receiving aspirin and/or dipyridamole or even coumadin therapy. Patients should be monitored according to physiologic status and the type of procedure being undertaken. Invasive monitoring is not required just because of the presence of MVP. One of the inferior ECG leads should be monitored because of the higher incidence of positive findings.[9] Antibiotic prophylaxis for subacute bacterial endocarditis is recommended.[9,14,15] These patients should be well sedated and premedicated. Drugs causing tachycardia (atropine) or alpha blocking agents (droperidol) should be avoided.[15] In terms of anaesthetic technique, it is more important to understand the underlying pathophysiology and treat the patient accordingly, than to specify general versus regional anaesthesia. The principles of management include maintenance of a large left ventricular end diastolic volume by keeping the patient well volume loaded, and avoidance of sudden afterload reduction. An extreme head-up position should be avoided.

Factors increasing myocardial irritability such as acidosis, hypercarbia, hypoxia, electrolyte dis-

turbances, etc. should be avoided as well as potentially dysrhythmic agents like adrenalin. In the selection of anaesthetic agents, tachycardia causing or sympathomimetic-type agents such as ketamine, atropine, and gallamine are best avoided.[15] If vasopressor therapy is required, an alpha adrenergic agonist like phenylephrine or methoxamine is the agent of choice. Induced hypotension with vasodilators may well aggravate the mitral prolapse. If dysrhythmias do develop under anaesthesia, it is important to exclude the more common causes (blood gas abnormalities, electrolyte disturbances, depth of anaesthesia) before attributing them to mitral valve prolapse.

If MVP is found in the asymptomatic patient pre-operatively, SBE prophylaxis is recommended.

In summary, mitral valve prolapse is a very common condition. These patients, especially those with murmurs, should receive SBE prophylaxis. Asyptomatic patients with no ECG abnormalities should otherwise be treated as normal, healthy people. However, there is a small subgroup who do present with dysrhythmias, embolic phenomena, congestive failure and sudden unexplained death. As yet it cannot be predicted in whom or why these problems will develop.

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