ORIGINAL

Mitral Valve Prolapse

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INTRODUCTION

The presence of mid or late systolic clicks and late systolic apical murmurs were first shown by Barlow[1] to indicate mild mitral valve prolapse. In these circumstances mitral valve function is only mildly abnormal. However marked mitral prolapse may also occur and this is often associated with more severe mitral regurgitation. The wide range in severity of prolapse is parallelled by a wide variety of causes.

MATERIAL

Sixty five patients have presented to the Cardiology Clinic at Green Lane Hospital during a six year period with findings of mitral valve prolapse (Table 1). In 25 cases chordal rupture was responsible and this most often occured in elderly patients where at operation the chordae appeared thin and attenuated and the leaflet tissue atrophic. Hypertension was a significant factor in three of these cases, while in a further three instances reptured chordae had followed bacterial endocarditis. During the review period eight patients were seen in whom prolapse was associated with myxomatous degeneration of mitral leaflets, this being often accompanied by dilatation of the valve ring. Myxomatous change in the mitral valve was confirmed by histological examination in seven of these patients who underwent surgery (Fig. 1).

Of special interest has been the referral of patients with mild mitral prolapse often associated with apical systolic clicks and a late systolic murmur. The frequency of the disorder is of interest and opinions differ. In a screening survey of approximately one thousand female students Rizzon et al. found a prevalence rate of 0.33 percent[2]. Amongst 4379 new patient referrals to the Cardiac Clinic at Green Lane Hospital during the past six years 32 cases have been recognised (Table 2). The incidence in our deprtment has been approximately 0.7 percent. Others have claimed a much higher incidence of prolapse than our figures suggest and in a study by Markiewicz and his associates of one hundred presumed healthy volunteers, 17 were found to have non-ejection clicks and/or late systolic murmurs[3]. The abnormalities were detected either with the subjects resting suprine, in the upright position, or by provocation using an inhalation of amyl nitrate. Twenty subjects had echo cardiographic abnormalities which were thought to indicate prolapse.

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DISCUSSION

Echocardiography has been a useful aid in the study of this disorder (Fig 2) and the specificity of the method has recently been tested by Haikal et al who found M node echocardiography a reliable indicator of the presence of mitral valve prolapse[4]. It is likely however, that undue reliance on abnormal echo findings will result in over diagnosis of this condition. Pansystolic "hammocking" with multiple echoes should not be considered diagnostic of prolapse. Another false positive appearance sometimes seen is late systolic bowing with non-continuous mitral valve echos during systole. This is believed to be due to movement of the heart during systole which allows different parts of the mitral valve apparatus to be recorded[5]. False-negative appearances are also sometimes seen in patients who have clinical and angiographic evidence of mild mitral valve prolapse but normal echocardiograms. The application of 2 dimensional echocardiography has provided additional information and should significantly reduce the incidence of a false-diagnosis.[6]

Most of our 32 patients with the "systolic click syndrome" had both a click and a late systolic murmur. However, four patients were seen who had a click only and six patients had late systolic murmurs without clicks. Prolapse was suggested by the auscultation and echo

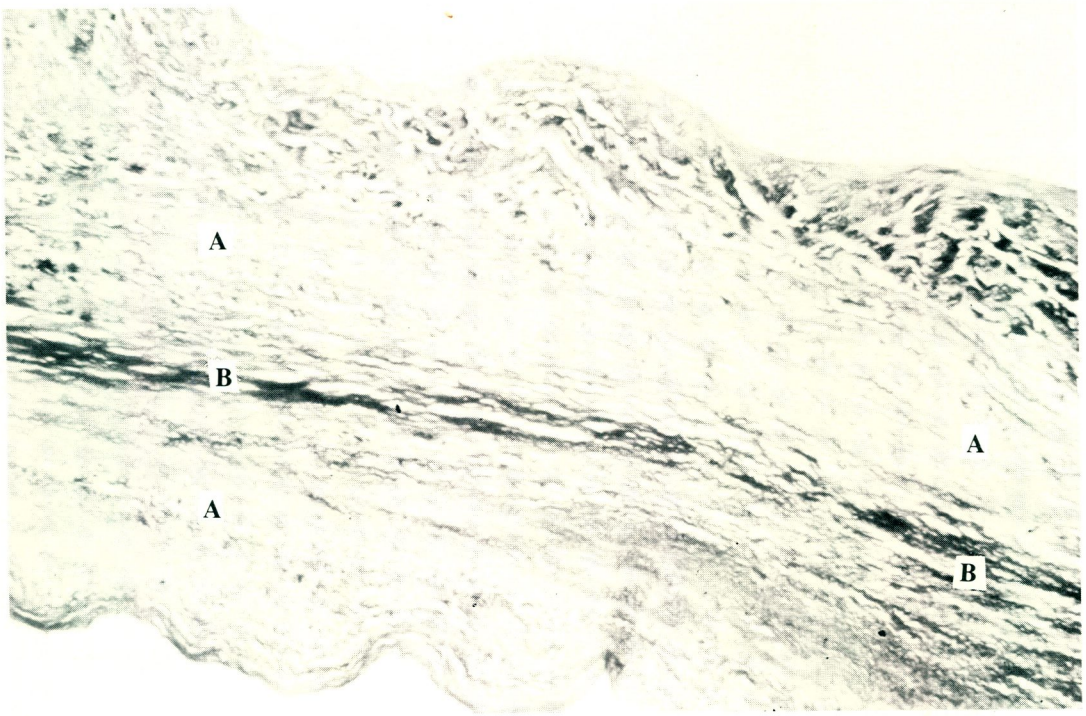

Figure 1 *A Mitral valve leaflet in a patient with the "Floppy Valve" syndrome. Note marked increase in acid mucopolysaccharide and Sparse elastic and collagen tissue.* **A** **B**

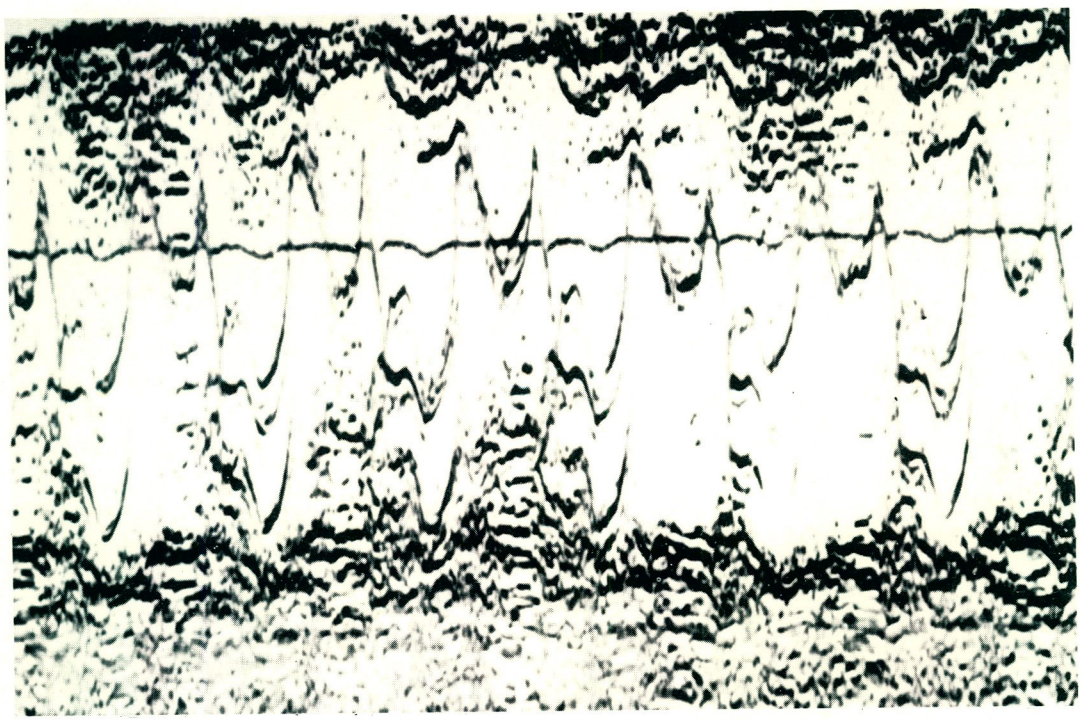

Figure 2 *Echocardiogram demonstrating typical appearances of mitral valve prolapse. Note systolic bowing of mitral valve and the continuous nature of the echoes.*

findings. A higher incidence has been reported amongst women and this was confirmed in our small series where 20 females and 12 males were affected. Eight of the 32 patients were unusually tall and exhibited some of the features of Marfan's disorder. Approximately half the patients had been referred for the assessment of heart murmurs but were asymptomatic. Thirteen complained of palpitations and some of these patients were also troubled by breathlessness or chest pain. Syncope had occured in three. Nine patients were shown to have arrhythmias and in eight these were due to ventricular extra systoles. In only four patients were ectopic beats frequent and these patients were treated with either quinidine or a beta-adrenergic blocking drug.

Electrocardiographic abnormalities are common in the prolapse syndrome and include QT prolongation, myocardial infarction patterns, T wave abnormalities, prominent U waves, conduction disturbances and both atrial and ventricular arrhythmias[7] [11]. Ventricular ectopic beats appear to occur frequently and in complex form in approximately half of affected individuals. Only about 25% of patients with mitral prolapse are free of arrhythmias. Winkle[9] and Sloman[8] have noted the poor correlation between recorded arrhythmias and the occurence of symptomatic palpitations. Many of their subjects who were free of symptoms were found to have arrhythmias when monitored. Methods involving exercise testing and ECG Holter monitoring have been used extensively to investigate the problem, and ambulatory Holter monitoring has proved the most reliable indicator of ectopy.

Among those who have speculated on the association of ventricular premature beats and the prolapse syndrome have been De Maria[10] and Cobb[12] who believe that the billowing leaflets induce abnormal mechanical stresses on the supporting papillary muscles and underlying left ventricular wall, thus triggering ectopic beats. Abnormal contraction of the left ventricle has been noted by Gooch[7] and also reported in papers by Jeresaty[13]. These authors have all noted an abnormal internal contour of the left ventricle during systole so that the postero-interior wall appeared to bulge into the cav-

TABLE 1

Mitral Valve Prolapse, 65 patients.
Green Lane Hospital, New Zealand.
(6 year experience)

Ruptured chordae	25
Floppy valve ± ruptured chordae	8
Systolic click & murmur	22
Late systolic murmur	6
Mid systolic click	4

TABLE 2

Mitral Systolic Click/Late Systolic Murmur Syndrome

32 patients
Green Lane Hospital, New Zealand.
(6 years experience).

Aetiological factors	Rheumatic fever	1
	Abnormal body build	8
	Unknown	23
Symptoms	Nil	14
	Palpitations	13
	Chest pain	6
	Breathlessness	6
	Syncope	3
Documented Arrhythmias		
	Paroxysmal atrial tachycardia	1
	Ventricular premature beats or VT	8

ity causing a convex ridge in some subjects. Others, including oursevles, have usually noted normal left ventricular contractility in this disorder.

Although ventricular ectopic beats are common, sudden death appears to be rare. In a review of the medical literature, Jeresaty[13] reported 12 cases where death had occured unexpectedly. In one of these patients mitral regurgitation was severe, and in another hypokalemia due to diuretic therapy may have been a contributing factor. It is clear that the patients who died, did not form a homogeneous group. The rarity of sudden death in this syndrome was also stressed in a review article by Devereux[14] who noted four cases among 387 patients. The possibility that individuals with the familial form of this syndrome may be at greater risk from sudden death has been reported in family studies.[15, 16] In a further report Campbell[11] placed emphasis upon abnormalities in the resting electrocardiogram. He showed that the occurence of major ventricular arrhythmias in a group of 20 patients with the syndrome was more common in the eight who had infero-lateral ST and T wave abnormalities.

While the mechanism of ventricular extra systoles remains undefined in patients with mitral valve prolapse and the disorder is still an enigma, anti-arrhythmic therapy is often beneficial. This should certainly be recommended in the management of patients with symptoms, or when monitoring demonstrates frequent or complex ventricular ectopic beats. It should also probably be recommended in those with a familial history of the disorder and in whom ventricular ectopic beats are demonstrated. Several authors have shown that propranolol is sometimes effective in suppressing ectopic beats but most have experienced difficulty in this regard. Quinidine, propranolol, or both drugs used in combination have been the favoured methods of treatment, until recently, when renewed interest has developed in amiodarone hydrochloride. This drug is very effective in the treatment of both supraventricular and ventricular arrhythmias. But there have been a number of adverse reactions reported following its use, of which the most serious is probably diffuse interstitial pneumonitis.[17] The use of this drug should probably be reserved for the treatment of patients with intractable and life threatening abnormalities of cardiac rhythm. It remains uncertain whether anti-arrhythmic agents in present use will prevent the rate but lethal arrhythmias that are sometimes seen in association with mitral valve prolapse.

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