Mitral Valve Prolapse Syndrome: Experience In Pakistan

By

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Mitral valve prolapse is a clinical syndrome in which there is a spectrum of auscultatory findings characterised by non-ejection click, a late systolic murmur or mid-systolic click followed by a late systolic murmur. The murmur which is caused by late systolic regurgitation associated with upward displacement of the anterior, posterior, or both leaflets of the mitral valve into the left atrium may be due to a congenital or acquired anomaly of the valve apparatus. The click occurs at the point of initial buckling of the leaflet. The murmur occurs due to the occurrence of mitral regurgitation. While usually there is both a click followed by a murmur, a click alone or an isolated murmur may exist (1).

This condition has been intensively written about in the last decade, and multiple names have been given to it. It was in the early sixties that Reid (2) proposed that the click and the murmur were mitral valvular in origin, and associated with myxomatous degeneration of the mitral valve. Barlow (3) confirmed the presence of mitral regurgitation (M.R.) in patients with mid-systolic click and late-systolic murmur. Ronnan (4) subsequently demonstrated by intracardiac phono that the click and the murmur arose from the mitral complex, and Criley (5) demonstrated the relationship between the mitral valve prolapse and the systolic click.

We present here the experience with mitral valve prolapse in Pakistan since echocardiography became available in 1978.

MATERIAL AND METHODS: Between Dec. '78 and Feb. '80 270 patient seen in the OPD and wards of the N.I.C.V.D. had Echocardiography evaluation 38 patients were found to have isolated mitral valve prolapse (MVP). Also, 18 patient had MVP with some other associated abnormality. Of the patients with MVP, 20 (53%) had a pre-echo suspicion of MVP. A detailed clinical history and a complete physical examination was recorded in all patients. A CP & ESR urinalysis, ASOT, ECG and chest X-Ray P.A View were done routinely in all patients. Echo was done using SKI EKOLINE A system with a 2.25 Mega Hz. transducer focused at 10 CM. Patients were examined from the standard left parasternal window in the supine, left lateral and in the sitting positions. The measurements were made in the standard manner (6). The diagnosis of MVP was made when the following criteria were fulfilled:

1. Mid or late systolic buckling of the leaflets (Fig. 1A).
2. Holosystolic retraction which is seen as a hammock-shaped deformity in which a concave motion throughout systole is recorded from both valve leaflets, with the

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transducer in the standard mid-position along the LSB (Fig. 1B).

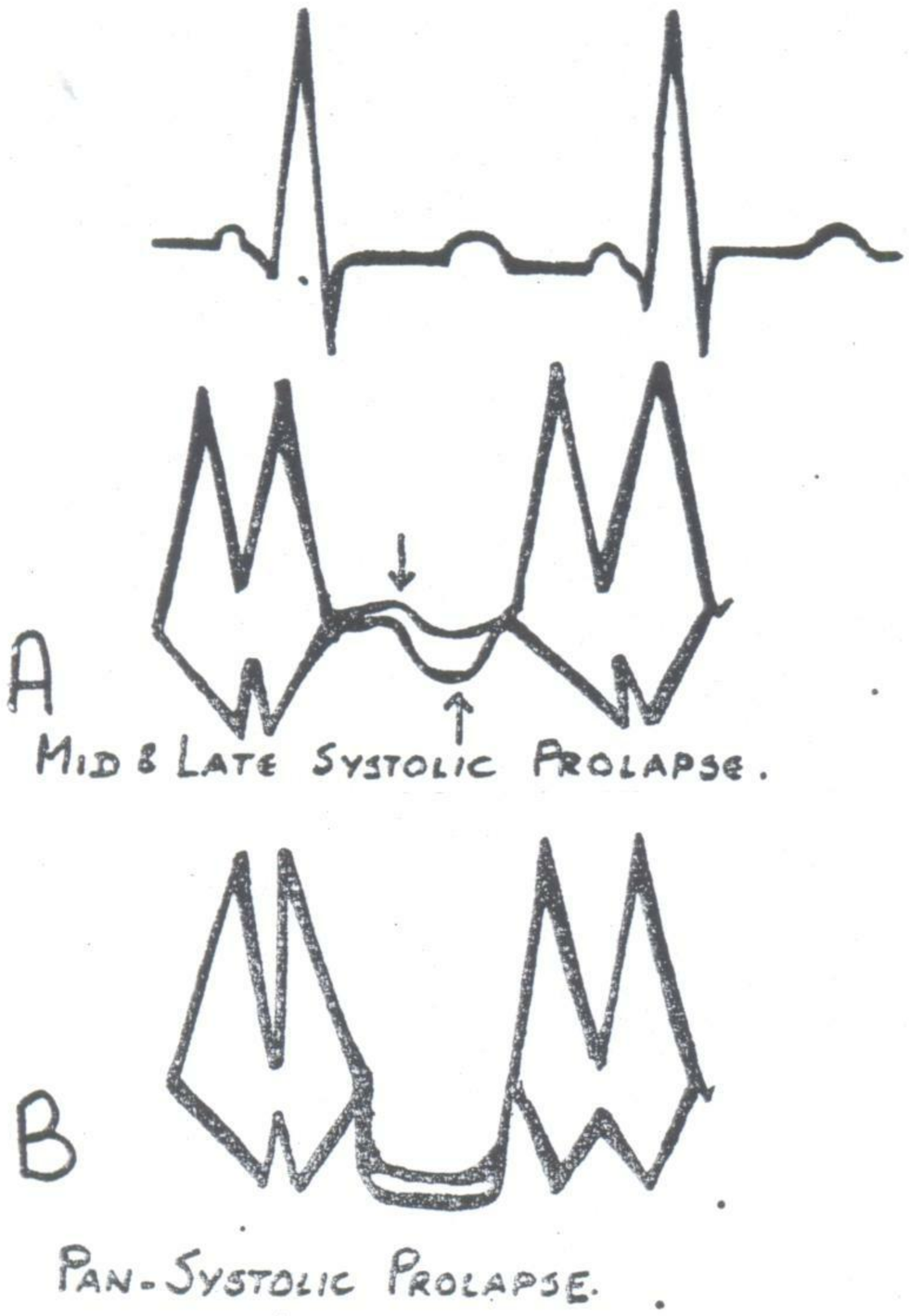

Figure 1.

There is some controversy regarding the exact echo diagnosis of MVP, as a large spectrum of abnormalities has been reported. We made a definitive diagnosis of MVP only when one of the above two patterns were recorded. Care was taken to eliminate the chances of artifactually producing the prolapse pattern by steep angulation upwards from the apex.

Results:

Total No. of patients 38.

Age :—ranged from 3-55 years, mean 24 years. Majority of the patients were between 17-30 years.

Sex :—Males 17, Females 21.

Clinical Presentation.

Table I: Presenting Symptoms

Atypical chest pain	21 pltients	(45%)
Palpitation	10 ,,	(26%)
Shortness of breath	4 ,,	(11%)
Fever	4 ,,	(11%)
Anxiety	1 ,,	(3%)
Failure to thrive	1 ,,	(3%)
Hemiplegia (young female; late post-partum)	1 ,,	(3%)
Asymptomatic	7 ,,	(18%)

(*Note :* Several subjects had multiple complaints *!*)

Table 2: Auscultatory Findings

Systolic murmur	25 patients	(66%)
Systolic murmur plus Systolic click	10 ,,	(26%)
Systolic click only	1 ,,	(3%)
No murmur or click	2 ,,	(5%)

Table 3: ECG

Non-specific ST-T changes	21 patients	(55%)
Increased voltage	4 ,,	(11%)
Normal	11 ,,	(29%)
Tall R VI	2 ,,	(5%)
Arrhythmias (on short 12-lead ECG only)	0 ,,	(0%)

Chest X-Ray:

P.A. view only were taken. Lateral views were not available. therefore we cannot comment on any patient having any skeletal abnormality, e.g. pectus or Straight oack etc. However 1 patient had pectus excavatum, and 2 patients had pectus cavinatum. Thirty-two patients had normal cardio-thoracic ratio (CT<0.5), 3 pati-

ents had borderline cardiomegaly (CT=0.5), 1 patient had cardiomegaly (CT>0.5), 1 patient had a small heart size (CT<0.25), 2 patients had pulmonary vascular congestion.

Table 4: Echocardiographic Findings

Isolated prolapse of mitral valve:		
(*a*) Mid and late systolic	24	Patients
(*b*) Holosystolic	14	,,
Total	38	,,
Coarse diastolic fluttering of anterior leaflet	7	,,
Anterior MV excursion (D-E) greater than 25 m.m. or anterior MV hitting IVS in diastole	10	,,
LV Volume Overload (i.e. EF normal or increased with the LVEDD increased)	5	,,
LA enlargement (> 40 m.m.)	6	,,
MV thickening (> 3 m.m.)	2	,,
Paradoxically moving (flail) posterior leaflet	2	,,
Aorta—No abnormality		
RV—No abnormality		
IV Septum and LV Posterior Wall showed no abnormality		
MVP seen with other conditions:		
(not included in this study)		
pericardial effusion	6	,,
(pseudo-prolapse)		
Volume Overload RV (ASD)	5	,,
PS	2	,,
Bicuspid Aortic Valve	3	,,
Complex Cyanotic heart disease	1	,,
Sub-valvular AS	1	,,

Discussion:

Mitral Valve Prolapse syndrome was anecdotally considered to be rare in Pakistan and a monograph was read at one of the local symposium presenting a few cases confirmed by phonocardiography (7). There is however no previous publication on this subject from Pakistan. The main reason for this may have been the absence of the facility of echocardiographic confirmation of this diagnosis and perhaps a low index of suspicion. In an early communication (8) from this Institute describing the experience of the first year of Echo in Pakistan we had mentioned the relative frequency of mitral prolapse. With the free use and availability of Echo locally, mitral prolapse has become one of the common problems referred for confirmation to us. However, it is still significant that in this selected group of patients we describe here, only 53% had a pre-echo diagnosis of mitral prolapse and in 47% some other diagnosis had been made.

As in the western world (9) the typical Pakistani patient with mitral valve prolapse is a young female or male, usually but not always with a slender build who presents with atypical chest pains and palpitation. Most of them have an isolated systolic murmur and only one fourth of our patients had the classical early systolic click followed by a mid-late systolic murmur. The ECG in the majority showed the usually described nonspecific ST and T wave changes usually in the inferior leads and the right sided chest leads. However, a good 29% had a normal resting electrocardiogram. One surprising finding was the absence of arrythmias in our group. What must be remembered is that only a 12 lead ECG was done and patient did not have exercise ECG or holter recording performed. Most

patients had a normal X-Ray chest. We cannot comment on the straight back and sternal deformities said to be commonly present in this group as routine lateral X-Ray chest views were not done. Clinicially however 2 patients had pectus cavinatum (pigeon chest) and 1 patient had pectus excavatum. The patients with cardiomegaly, pulmonary vascular congestion were those who had developed significant mitral incompetence.

The echo showed the usual type of mid-late systolic prolapse in 64% of patients and holosystolic prolapse in 36%. Patients with holosystolic prolapse did tend to be somewhat more symptomatic and the patients with significant mitral regurgitation had holosystolic prolapse. Coarse diastolic flutter on the anterior mitral leaflet thought to represent flail anterior leaflet (6) was seen a number of times but surprisingly in the absence of significant mitral regurgitation. Only those patients with clinically apparent significant mitral regurgitation had an enlarged left atrium and a pattern of the touching of the septum by the anterior mitral leaflet in diastole. Mitral valve thickening especially towards the tips of the leaflets possibly due to myxomatous modules was seen occasionally.

While mitral valve prolpase pattern was seen in association with a variety of other diseases as noted earlier, these cases were excluded from the overall group of isolated mitral valve prolapse. In our small group there was one case of hemiplegia in a young woman—an association noted increasingly in the West (10).

The importance of this disease process lies in the facts that it is a very common cardiological problem in Pakistan and is responsible for the ECG changes described above which had frequently been mislabelled ischaemic changes in the patients referred to us. Treatment involves mostly reassurance and permission to lead a full life, occasionally use of propranolol for symptoms and bacterial endocarditis prophylaxis in those with a clear cut systolic murmur. A lot of disability and anxiety can be relieved by recognition of this disease which is compatible with long term survival and good prognosis.

Summary

The features of mitral valve prolapse syndrome as seen in Pakistan are described. The importance of the recognition of this problem lies in the fact that a number of these patients are misdiagnosed and given labels and prognosis much worse than warranted.

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