Mitral Valve Prolapse in Children

A Problem Defined by Real-time Cross-sectional Echocardiography

DAVID J. SAHN, M.D., HUGH D. ALLEN, M.D.,
STANLEY J. GOLDBERG, M.D., AND WILLIAM F. FRIEDMAN, M.D.

SUMMARY The cross-sectional echocardiographic features of mitral valve prolapse were defined in 26 children (ages 2–18 years) using a real-time, multiple-crystal ultrasound scanner. In each patient the physical findings of the mitral valve click-murmur syndrome were present and mitral valve prolapse had been diagnosed previously by conventional single crystal echocardiography. Mitral prolapse occurred in a familial setting in eight patients and was associated with the Marfan syndrome in five. Real-time two-dimensional echocardiography uniformly disclosed maximum mitral arching and the superior-posterior prolapse. These visual observations were confirmed by M-mode recordings derived from single elements within the array of 20 crystals. The method allowed a complete M-mode description of the phasic motion of the entire mitral apparatus and observations of the spectrum of prolapse from discrete late systolic prolapse to “hammock-like” holosystolic prolapse. Further, the recording of multiple systolic M-mode lines occurred when the ultrasound beam intersected the arched leaflets more than once. Pseudo-systolic anterior motion was observed often and resulted clearly from a superimposition of echoes from the mitral annulus and from the posterior-superiorly arched prolapsed leaflets. A major finding in 22 patients was the association with prolapse of biconvex enlargement of the aortic sinuses of Valsalva and a significant increase in the diameter of the aortic root. Aortic root dilatation was most marked in, but not confined to, patients with the Marfan syndrome and was a prominent finding in six patients with minor musculoskeletal abnormalities. The presence of aortic root dilatation in children with normal body habitus raises important questions concerning the generalized nature of an abnormality of cardiac connective tissue in patients with mitral prolapse. The cross-sectional approach significantly enhances the noninvasive evaluation of mitral valve prolapse and provides an explanation for many of the single crystal observations reported previously.

CONVENTIONAL ECHOCARDIOGRAPHIC techniques are of proven utility in the detection of mitral valve prolapse.1–5 It would appear that the echocardiographic features of this disorder are similar in children and adults since members of all ages are affected when the lesion occurs in a familial setting.3,4 Despite echocardiographic6–9 and angiographic10–12 reports of varying patterns of mitral valve motion and abnormalities in ventricular shape and contraction patterns,13,14,15 a unified concept does not exist of the primary abnormality in this disorder. In the present study, real-time, multiple-crystal, cross-sectional echocardiography16 was employed to evaluate the anatomy and phasic characteristics of the mitral complex and the proximal aorta in 26 children with the typical physical findings of click-murmur syndrome and standard, single crystal echocardiographic evidence of mitral valve prolapse. The cross-sectional analysis was augmented by selected, single element M-mode recordings from known locations within the multi-crystal array.17 These studies provided an explanation for previously described single crystal findings in mitral valve prolapse and uncovered new information concerning the association of an aortic root abnormality in this disorder.

Methods

Patient Material
Seventeen females and nine males, aged 2 to 18 years, (mean 9.42 ± 0.66 [se] years) underwent ultrasound examination with an Echocardiovisor real-time, multiple-crystal cross-sectional echocardiographic system.17 These children were included in the present study after consecutive presentation (over nine months) as newly referred outpatients with physical findings of click-murmur syndrome and positive standard single crystal echocardiograms. Mid-systolic click was an isolated auscultatory finding in eight patients; a holosystolic murmur was heard in five patients; and a mid-systolic click and late systolic murmur was audible in 13 (table 1). Mitral prolapse unassociated with the stigmata of the Marfan syndrome occurred in a familial setting in eight patients (five females, three males) from three separate families. The morphologic characteristics of the Marfan syndrome were present in one boy and four girls (three had an affected parent, none had affected siblings).18,19 Although the remaining 21 patients had normal height and weight distribution, six were noted to have minor musculoskeletal abnormalities (pectus excavatum) (4), high arched palate (1), arachnodactyly with hyperextensible joints (2). In none of the patients was there a history of rheumatic fever or clinical evidence of other cardiac abnormalities.

The control group of 25 age-matched normal children were selected siblings of patients presenting in our outpatient department for a variety of pediatric problems, who underwent echocardiographic study to evaluate the normal spectrum of mitral valve motion. None of these children had systolic murmurs or clicks at rest or when squatting. None had family histories of congenital or acquired valvular heart disease.

Echo Techniques

Real-time, multiple-crystal cross-sectional echocardiographic studies were performed as described previously.20 In brief, sagittal visualization of the heart was obtained in resting and supine subjects with a 4.5 MHz transducer.

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placed vertically along the left sternal border in the long axis of the heart (fig. 1). Provocative testing by handgrip or amyl nitrite was not employed. Mitral valve motion was visualized in real-time on an oscilloscope and was recorded on Super-8 mm motion picture film and video tape. Still frames were obtained by electrocardiographic gate with a Polaroid system at 1/30 second exposure. The M-mode output of single elements within the multiple-crystal array was recorded on a Honeywell 1856 fiberoptic strip chart. To obtain single element M-mode recordings, mitral valve motion was visualized in cross-section, the index crystal selected by number, and the oscilloscope position of the single crystal identified electronically as a bright horizontal line. Cooperative patients held their breath at end expiration while the A-mode output from the single crystal was displayed for fine gain adjustment. Thereafter, line position with reference to the mitral valve was rechecked in the cross-sectional mode and a Polaroid still was obtained to document and index the line position of the M-mode recording. In this manner, a complete M-mode profile of motion of the

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**Marfan Syndrome**

*16 CM A, P + WHIP-T DIL HSP
*17 HSM A, P + WHIP DIL HSP
18 HSM A, P + T DIL HSP
*19 HSM A, P + WHIP DIL HSP
20 CM A, P + DIL HSP

**Skeletal Abnormalities—Pectus excavatum—21-24; Arachnodactyly—25-26**

21 CM A, P + WHIP DIL HSP
22 HSM A, P + WHIP DIL HSP
*23 CM A, P + NL HSP
24 CM A, P + DIL HSP
25 CM A + NL HSP
26 SC A, P WHIP DIL HSP

*Familial cases.

Abbreviations: CM = click and murmur; SC = systolic click (no murmur); HSM = holosystolic murmur; T = thickened leaflet tissue; DIL = fishbowl-like dilatation; HSP = holosystolic prolapse—both leaflets; LSP = late systolic prolapse—both leaflets; NL = normal contour; A = anterior; P = posterior; * = conventional single crystal echo showed LSP only.

**Figure 1.** Normal motion of the mitral complex is illustrated in still frames from an 8 mm movie. Left panel: The anterior (AML) and posterior (PML) mitral leaflets are separated in diastole. The aortic valve (AO) is closed. With the onset of pre-ejection systole (center panel), the mitral leaflets coapt with a slightly arched configuration while the aortic valve remains closed. In late systole (right panel) the aortic valve is open. S = septum, LV = left ventricle.
anterior leaflet of the mitral valve from its free edge to the aortic root was obtained by serial, ascending individual crystal recordings.

Dimensional studies were obtained from echocardiographic M-mode output recordings. Aortic root outer diameter and left atrial inner diameter were measured at the level of the aortic valve, the latter at end systole. Aortic root diameter and left ventricular end-diastolic dimensions were measured at the onset of the QRS complex of ECG. The ventricular diameter and mitral valve D-E excursion were determined at the level of the posterior mitral leaflet. All dimensional data were normalized for body surface area and compared to normal values described previously.21

Results

Normal Mitral Valve Motion: Cross-sectional Visualization

In diastole, the mitral valve leaflets separate with the anterior leaflet appearing as an inferior extension of the aortic root (fig. 1). The posterior leaflet arises from the posterior heart wall at the atroventricular junction. At the onset of pre-ejection systole, coaptation of both leaflets creates the appearance of a curvilinear funnel. As ejection proceeds, the atroventricular ring moves inferiorly and the line of leaflet coaptation moves forward, lagging behind the aortic root. Thus, the mitral apparatus becomes somewhat more horizontal within the cavity of the left ventricle and slight arching of both leaflets is noted. A spectrum exists in the degree of arching seen in normal children. However, superior motion of the leaflet body was not seen in the normal group.

Mitral Valve Prolapse: Cross-sectional Visualization

Prolapse of the mitral valve appears as a superior and somewhat posterior displacement of both leaflets. The leaflets arch on their chordae with the body of the leaflets billowing toward the left atrium. While a spectrum exists in the degree of prolapse on cross-sectional examination, superior motion of the leaflet body was the common diagnostic feature of real-time examinations. The body of the posterior leaflet is driven superiorly toward the left atrium while portions of the leaflet near the atroventricular junction billow posteriorly. Isolated posterior leaflet prolapse was not observed in any patient. In every patient, a motion abnormality of the anterior leaflet was recorded on single crystal output from the area of maximal arching of the valve. Prolapse of leaflets into the cavity of the left atrium during systole (fig. 2) occurred in ten patients, including all five patients with the Marfan syndrome and all of those in whom a holosystolic murmur was detected by auscultation. Abnormal "whip-like" diastolic mobility of the leaflets was observed in real-time in 11 patients. This was associated with increased mitral valve DE excursion in single crystal output studies (table 2). Of major interest was the detection in 17 patients of dilatation of the aortic root at the sinuses of Valsalva (fig. 2).

Single Crystal Profile of Mitral Valve Prolapse

In most previously reported single-crystal echocardiographic studies,5,6 maximum prolapse has been described when the transducer was angled upwards so that the structure visualized behind the mitral valve leaflet was actually the left atrial wall (line 4, fig. 7). Thus, the echo beam transsected the maximum posterior-superior prolapse arch of the mitral valve. The line output selected in figure 3 passes through the arch of the elongated anterior leaflet. When single crystal outputs are examined both above and below

![Figure 2. Polaroid still frame cross-section (left) and line drawing (right) illustrate arching of the anterior (AML) and posterior (PML) mitral leaflets which are prolapsed into the left atrium (LA). The aortic root (AO) is significantly dilated in this patient with the Marfan syndrome.](image-url)
By selecting the appropriate single-crystal outputs from the multiple-crystal array, holosystolic prolapse was identified in 20 patients and late systolic prolapse in six. No correlation existed between the type of prolapse and the findings in physical examination. Thus, in the 20 patients with holosystolic prolapse, six had a systolic click only on auscultation, while nine had both click and murmur and five had pansystolic murmur. In the six remaining patients, only late systolic prolapse was recorded (table 1). In this regard, while it has been suggested that concordance exists in the phono-cardiographic timing of the click and murmur and the echo timing of posterior leaflet prolapse,4 DeMaria et al.4 reported five patients with isolated mid-systolic click and hammock-like holosystolic prolapse without simultaneous auscultatory mitral motion correlations.

The ability to create a total profile of mitral motion provided an additional observation of great interest. A pseudo idiopathic hypertrophic subaortic stenosis pattern (systolic anterior motion of the mitral valve) has been described in patients with mitral valve prolapse. It was apparent in the present study that this appearance is created by superimposition of the mitral annulus and mitral valve leaflets.23 In 17 patients, the anterior motion of the annulus was superimposed on posteriorly-placed mitral valve tissue during the posterosuperior motion of the valve (fig. 5).

Tricuspid Valve Motion

Tricuspid valve prolapse has been described angiographically20 and echocardiographically26 as a component of the systolic click-murmur syndrome. However, it should be recognized that variations exist in tricuspid systolic motion patterns in normal children21,28 that may cause uncertainty regarding the association of tricuspid valve pathology and mitral prolapse. In the present study, tricuspid valve M-mode echoes were considered suitable for analysis in 18 of 26 patients. Although a minimal suggestion of late prolapse could be visualized (fig. 4). Although it has previously been suggested that these patterns represent different forms of prolapse,5 it was quite clear that simultaneous views of different portions of the same prolapsed valve accounted for each pattern. Our current observations suggest that the echocardiographic pattern recorded from the free edge of the valve has the closest association temporally with the appearance of the systolic click and the murmur of mitral regurgitation.

With the leaflets arched toward the left atrium, limitations of beam width (lateral resolution) resulted in the beam subtending various portions of the valve at the same time. Multiple line sequences were produced by the depth differences of various portions of the valve; examination of these multiple line sequences disclosed that the faintest echoes were most posterior (fig. 4). The portion of the leaflet most severely prolapsed was the most difficult to record because of its aposition to the posterior left atrial wall and its obtuse angle with reference to the echocardiographic beam. The real-time motion sequences demonstrated that as the valve leaflet moved posteriorly, its echo signal became fainter. Thus, in five patients, disappearance of leaflet tissue echoes (echo dropout) was observed in cross-section as leaflet angulation progressed during prolapse.

This beam position, the entire spectrum of M-mode abnormalities, from late systolic prolapse to holosystolic (hammock-like) prolapse could be identified in the same patient. Moreover, both holosystolic and late systolic prolapse were identified at the same time in the same patient from portions of the valve subtended by the echocardiographic beam (fig. 4). Although it has previously been suggested that these patterns represent different forms of prolapse,5 it was quite clear that simultaneous views of different portions of the same prolapsed valve accounted for each pattern. Our current observations suggest that the echocardiographic pattern recorded from the free edge of the valve has the closest association temporally with the appearance of the systolic click and the murmur of mitral regurgitation.

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systolic dipping was observed in five children, unequivocal tricuspid valve prolapse did not exist in any instance.

Cardiac Chamber and Vessel Dimensions

Dimensional data are presented for the prolapse patients in table 2. When compared to normal values corrected for body surface area, six of the ten patients with the most severe mitral prolapse (#9, 16–18, 22 and 25) had mild left atrial and ventricular dilatation. Less commonly, an isolated increase in left atrial (#20) or left ventricular diameter (#19 and #26) was noted. A major finding was the detection of aortic root dilatation in 22 of 26 patients, including all with the Marfan syndrome, in the absence of clinical findings of aortic regurgitation. Aortic root dimensions in each of these 22 patients exceeded the 95th percentile by at least two millimeters (fig. 6). While aortic root dilatation was most marked in the children with the Marfan syndrome, five of six patients with minor musculoskeletal abnormalities had marked increases in aortic root dimensions (table 2).

Only one of the 25 normal children had a dimensional abnormality: an eight-year-old championship swimmer (body surface area 1.1m²) who had a 2.6 cm aortic root diameter. All of the other echo dimensions were within normal limits in the control population.

Discussion

Concern has been expressed recently that echocardiography may be an oversensitive diagnostic indicator of leaflet involvement in mitral valve prolapse when compared to cineangiography.

The results of the present investigation demonstrated that real-time cross-sectional echocardiography in combination with M-mode outputs derived from known locations within a multcrystal array enhances substantially the noninvasive delineation of the anatomy and physiology of mitral valve prolapse. It must be appreciated that the cross-sectional illustrations that accompany this report are but single still frames derived from real-time motion studies that are visualized best on an oscilloscope. Furthermore, they have been subjected to photographic enlargement from a small format film. Anatomic details are substantially clearer and structure identification facilitated greatly by review of the moving images.

Real-time cross-sections readily identified the posterior-superior motion of the mitral valve prolapse and arching of either or both mitral leaflets. Most importantly, cross-sectional visualization allowed identification of the most appropriate leaflet site from which to derive single-crystal recordings of maximal prolapse. Moreover, creating a complete profile of phasic mitral motion via M-mode recordings from all portions of the multiple-crystal array provided a substantially more comprehensive description of the mitral disorder. Thus, in eight of 13 patients with late systolic prolapse by conventional single-crystal examination, holo-systolic prolapse was detected on portions of the total profile. In this same regard, some normal children with normal cross-sectional examinations had trivial degrees of late systolic posterior motion recorded from the body of the leaflet and were observed to have normal motion on recordings from the free edge of the leaflets. We have found that a posterior mitral valve displacement during the CD slope which is 20% of the mitral valve DE excursion occurring at the free edge correlates with click-murmur. Tracings obtained from the body of the leaflet (where the anterior and posterior leaflet traces do not come together) with a superior or inferior transducer angulation may have artifactual minor systolic motion abnormalities.

The integrated single crystal multiscan approach clarified the mechanisms responsible for such poorly-defined past observations in patients with mitral prolapse as multiple line
sequences,⁴,⁵ faintness of echoes from the most severely prolapsed portion of a leaflet, and the pseudo systolic anterior motion artifact.⁶ A proper interpretation of echoes from a prolapsed mitral leaflet requires an appreciation of the superior-posterior nature of the abnormal motion and the problems imposed by the limits of lateral resolution and reception of echoes from an obliquely-oriented valve. Multiple line sequences represent a superimposition of echoes from various portions of the valve that result directly from their obliquity and the influence of beam width errors. Lateral resolution is a function of depth and depends on frequency, acoustic element size, and the magnitude of amplification of returning echoes. Although the appearance in the final image is different, both single-crystal M-mode and multiple-crystal cross-sectional results are subject to the same physical limitation. The multiscan array has smaller individual elements than conventional single-crystal instruments and superimposed echoes were recorded from these smaller elements. However, the total profile provided by all of these elements made it clear that individual echo beams subtended mitral valve tissue several times (line 2, fig. 7). Similar multiple line sequences are often recorded on M-mode echoes from patients with mitral stenosis in whom a dome is formed at the valve orifice in diastole, allowing the echo beam to intercept mitral tissue several times.⁷ Likewise, the systolic anterior motion pattern observed in idiopathic hypertrophic subaortic stenosis may appear as multiple line sequences when the anterior mitral leaflet moves forward, and portions of the valve are obliquely related to the echocardiographic beam.

Echocardiographic imaging of the most severely prolapsed portion of the mitral valve may be difficult since the apex of the arch created by the leaflets is often oriented obliquely to the echo beam. This mechanism accounted for echo dropout or faintly received signals on both cross-sectional visualization and on M-mode outputs. The approach utilized in the present study demonstrated that the superimposition of echoes derived from the anterior mitral annulus upon echoes from the posterosuperiorty prolapsed leaflets provided the source of the pseudo systolic anterior motion artifact observed on M-mode recordings (line 1, fig. 7).

Angiographic correlates of the mitral valve motion abnormalities seen in this study were not obtained as none of the children in the study group underwent cardiac catheterization. The involvement of both anterior and posterior leaflets may represent a unique feature of this pediatric patient group since previous angiographic features have commonly suggested isolated posterior leaflet prolapse in adults.⁸¹⁰ Nevertheless it is possible that the left ventricular angiogram may be a less sensitive indicator of anterior leaflet abnormality when compared to posterior leaflet visualization.

A major finding of the present investigation was the association of aortic root dilatation with mitral prolapse in 85% of our patients, many of whom had subtle abnormalities of body habitus. In this regard, the association of prolapse with flat feet, kyphoscoliosis and a high arched palate prompted Rizzon⁹ to speculate that prolapse might be a component of a forme fruste of the Marfan syndrome. Additional patients have been reported in whom severe prolapse has been associated with straight-back syndrome.⁵ Read and co-workers⁸ have described musculoskeletal abnormalities in patients with myxomatous degeneration of the mitral valve. Likewise, an increased frequency of thoracic skeletal anomalies associated with mitral prolapse has also been emphasized in other recent studies.²⁹, ³⁰ The high incidence of skeletal abnormalities and aortic root dilatation in the present study lends support to the hypothesis that an intermediate group may exist between mitral prolapse patients with normal body habitus and those with the Marfan syndrome. Brown et al.³¹ have suggested that aortic root dilatation and mitral valve prolapse are echocardiographic markers for the Marfan syndrome. Our own data would not support this notion since the two abnormalities were associated often in children with normal body habitus. Whether or not such children have a generalized disorder of cardiac connective tissue remains unknown, but it would appear that the question deserves more intensive study.

References

![Figure 7](http://circ.ahajournals.org/)

**Figure 7.** Diagrammatic representation of mitral valve prolapse shows the posterosuperior orientation of the prolapsed leaflets. The M-mode output from line 1 demonstrates the pseudo-systolic anterior motion artifact caused by superimposition of echoes from the valve annulus and the anterior leaflet. The output from line 2 shows a multiple line sequence from the various portions of arched leaflets subtended by the beam. The echo beam at line 3 shows the pattern of prolapse demonstrated at the free edges of the leaflets. Line 4 represents the direction from which most published single crystal recordings have been obtained. Aortic root dilatation (AO) is also shown. LA = left atrium, LV = left ventricle.
M-mode Echocardiography in the Evaluation of Patients for Aneurysmectomy

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SUMMARY In order to determine whether echocardiography could be useful in predicting surgical mortality of aneurysmectomy, preoperative condensed M-mode echocardiographic scans were taken from both mid (standard position) and low (nearer apex) intercostal spaces and/or from the subxiphoid area in eighteen patients who were sent to surgery for aneurysmectomy. Eleven of the eighteen patients survived aneurysmectomy. All eleven had mid left ventricular dimensions less than 3.3 cm/m² and low dimensions of 3.8 cm/m² or less.

The incidence of left ventricular aneurysm has been reported to be from 3.5% to as high as 38% in patients surviving myocardial infarction. Many reports indicate that an aneurysm may markedly shorten a patient's life span, and surgical intervention is being used increasingly for symptomatic patients. One of the major limitations of surgical therapy is the problem of leaving enough normally contracting myocardium after removing a large aneurysm.

Of the seven patients who died, the mid and low left ventricular dimensions exceeded 3.3 cm/m² and 3.8 cm/m², respectively, with one exception. The combination of abnormal mitral valve closure, a dilated mid dimension and lack of normal motion in opposing wall segments was only seen in six nonsurvivors. Echocardiography can provide information concerning the state of the left ventricle in patients with ventricular aneurysms and these findings may be helpful in predicting surgical mortality for aneurysmectomy.

This study was undertaken to see if M-mode echocardiography could be of value in selecting patients for aneurysmectomy by seeing whether any echocardiographic measurements of left ventricular performance could be useful in predicting surgical mortality.

Methods

A group of 18 patients selected to undergo surgical resection of ventricular aneurysms secondary to coronary artery disease were studied using M-mode echocardiographic techniques. All 18 of these patients had undergone selective coronary cineangiography as well as ventriculography in the right anterior oblique (RAO) position. Each patient was examined by M-mode echocardiography utilizing an Ekoline 20-A echocardiograph coupled to an 1856 Honeywell recorder. A 2.25 MHz 0.5 inch

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