Echocardiographic Observations on the Association between Mitral Valve Prolapse and Asymmetric Septal Hypertrophy

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SUMMARY One hundred and ninety consecutive patients with mitral valve prolapse (MVP) were studied by echocardiography. Asymmetric septal hypertrophy (ASH) was noted in 16 patients. Three patients had syncope which was associated with supraventricular arrhythmias. Three others had episodes of presyncope which were not related to rhythm disturbances. One of the patients with MVP and ASH had a family history of idiopathic hypertrophic subaortic stenosis. The septal thickness ranged from 1.6 to 3.1 cm, mean = 2 cm. The posterior wall thickness ranged from 0.7 to 2.1 cm, mean = 1.0 cm. The ratio of the thickness of the interventricular septum to that of the posterior wall ranged from 1.5 to 2.5, mean = 1.9. The percentage of thickening of the septum in systole was reduced in 13 patients. The excursion of the interventricular septum was reduced in three patients. In nine patients the left ventricular end-systolic dimension was below the lower limit of normal. Percentage fractional shortening of the left ventricle was increased in eight patients. Since MVP predisposes to cardiac arrhythmias which are poorly tolerated in the setting of ventricular hypertrophy and reduced left ventricular compliance, the recognition of this combination of MVP and ASH is of clinical importance.

MITRAL VALVE PROLAPSE is a common entity. Although the majority of patients with mitral valve prolapse have a benign clinical course, some subjects experience serious complications such as sudden death, infective endocarditis and severe mitral regurgitation. Mitral prolapse has been associated with skeletal abnormalities, ventricular and atrial arrhythmias, abnormal patterns of ventricular contraction on the angiogram, and tricuspid valve prolapse.

Numerous reports have pointed out the usefulness of echocardiography in the diagnosis of mitral valve prolapse. Diagnostic ultrasound is a useful modality in the detection of asymmetric septal hypertrophy.

In this report we describe the association between mitral valve prolapse (MVP) and asymmetric septal hypertrophy (ASH).
Materials and Methods

One hundred and ninety consecutive patients with mitral valve prolapse were studied by echocardiography. All patients had either the classical mid systolic, or holosystolic prolapse of the mitral valve. Sixteen patients had asymmetric septal hypertrophy. Detail echocardiographic studies of left ventricular parameters in these sixteen patients form the basis of this report.

The group consisted of eleven females and five males. Their ages ranged from 14 to 76 years. Seven patients were under the age of 40. Five patients complained of atypical chest pain, three others had chest pain suggestive of angina pectoris. One of these patients had cardiac catheterization; she had severe retrosternal pain associated with ST depression during the procedure. The left ventricular end-diastolic pressure (LVEDP) was 27 mm Hg. Sublingual nitroglycerin resulted in prompt relief of pain and return of the LVEDP to normal levels. Her coronary arteriograms were normal. The left ventriculogram showed mitral valve prolapse and excellent left ventricular contractions. A ventriculogram in the LAO view was not performed and hence the presence of septal hypertrophy could not be documented.

Four subjects had recurrent episodes of syncope (two of them had a history consistent with angina and another had atypical pain). In three of them syncope was associated with supraventricular arrhythmias. One of these patients was monitored in the hospital. She had episodes of atrial flutter which resulted in marked hypotension and syncope. Supraventricular arrhythmias were detected in the other two patients by Holter monitoring. Three others had bouts of near syncope which were not associated with arrhythmias (one had dyspnea and another had atypical pain). Two patients complained of dyspnea on exertion (one had presyncopal spells and the other had atypical chest pain). Six patients admitted to a history of palpitations (three had experienced syncopal episodes, and three others had atypical chest pain). Two patients presented with fatigue and two were asymptomatic. Four patients had a history of mild systemic hypertension.

On examination a nonejection systolic click was noted in nine patients, five of whom had systolic murmurs following the click. A fourth heart sound was heard in five patients. Five had systolic ejection murmurs and an isolated late systolic murmur was noted in two patients. The electrocardiogram revealed left ventricular hypertrophy (LVH) in six patients, nonspecific ST-T changes in six, and four had normal ECGs. One of the patients with LVH had multiple ventricular extrasystoles.

Cardiac catheterization was performed in three patients. The presence of mitral valve prolapse was confirmed by left ventricular angiography in all three patients. A septal bulge suggestive of septal hypertrophy was noted in one patient who had a ventriculogram performed in the left anterior oblique (LAO) view. The other two did not have angiograms in the LAO projection and hence no comment could be made regarding septal hypertrophy; however, both of them demonstrated hypercontractility of the left ventricle with obliteration of the apex.

The patients were supine or in the left lateral decubitus position during the echocardiographic examination. Echocardiography was performed using an Ekoline 20 ultrasonoscope, a 2.25 MHz, 7.5 cm focus, 0.5 inch transducer and a Honeywell 1856 or Electronics for Medicine DR8 recorder. The tracing was done from a standard interspace, i.e., the interspace from which the mitral valve could be visualized by perpendicular or near perpendicular placement of the transducer. Care was taken to avoid inferior angulation of the transducer during the mitral valve recording. An M-mode sector scan ultrasound study was performed on each patient. After completion of the baseline recording amyl nitrite was administered and the mitral valve echocardiogram repeated in order to assess whether systolic anterior motion could be seen. The left ventricular end-diastolic dimension was measured at the apex of the R wave. The end-systolic dimension was taken as the shortest distance between the left septal and posterior wall endocardial echoes. The amplitude of the posterior wall endocardial echo and the left septal echo were measured according to previously described methods. The left ventricular dimensions, the amplitude of the left septal echo and the endocardial echo of the posterior wall of the left ventricle, and the thickness of the interventricular septum were measured at a level just caudal to the mitral valve. Percentage thickening of the septum or posterior wall was expressed as systolic thickness minus diastolic thickness divided by diastolic thickness, and this fraction was multiplied by 100. Fractional shortening of the left ventricle (FS) was determined by subtracting the end-systolic left ventricular dimension (LVES) from the end-diastolic dimension (LVED) (measured at the apex of the R wave), dividing this value by the end-diastolic dimension and expressing it as a percentage:

$$\text{FS} = \frac{\text{LVED} - \text{LVES}}{\text{LVED}} \times 100.$$ 

Results

The results of the investigation are summarized in table 1. All sixteen patients had the classical mid systolic variety of mitral valve prolapse. None had systolic anterior motion of the mitral valve after provocation with amyl nitrite. The ratio of thickness of the interventricular septum to that of the posterior wall was 1.5 or greater in all patients. Septal hypertrophy was present in all patients (septal thickness in 30 normal subjects studied in our laboratory ranged from 0.8 to 1.2 cm, mean = 1.0 cm). The septal thickness ranged from 1.6 to 3.1 cm. The thickness of the left ventricular posterior wall ranged from 0.7 to 2.1 cm (the normal range for our laboratory is 0.7 to 1.1 cm, mean = 0.9 cm). Only four patients had hypertrophy of the posterior wall (> 1.1 cm), three of whom had a borderline increase in wall thickness (1.2 cm). The percentage systolic thickening of the interventricular septum was reduced in thirteen patients (the normal range for our laboratory is 28 to 50%, mean = 41%). Systolic thickening of the left ventricular posterior wall was reduced (< 38%) in one patient; and it was increased in three patients (the normal range for our laboratory is 40 to 75%, mean = 57%). The excursion of the interventricular septum was reduced in three patients and increased in one patient (the normal range for our laboratory is 0.4 cm to 1.0 cm, mean = 0.7 cm). Motion of the left ventricular posterior wall endocardial echo was reduced in one patient and increased in two patients (the normal range is 0.9 to 1.4 cm, mean = 1.1 cm).
Reduction of the left ventricular end-systolic dimension below the lower limit of normal for our laboratory was noted in nine patients (the normal range is 2.5 to 3.5 cm, mean = 3.1 cm). Percentage fractional shortening of the left ventricle was increased above the upper limit for our laboratory in eight patients (the normal range is 28-41%, mean = 34%).

Figure 1 shows an example of mitral valve prolapse. Mid-systolic posterior motion of the valve is present. His left ventricular echocardiogram demonstrates asymmetric septal hypertrophy (fig. 2).

Discussion

The role of echocardiography in the diagnosis of mitral valve prolapse has been clearly established.13-14 Asymmetric septal hypertrophy can be reliably diagnosed by echocardiography.14 Although the echocardiographic features of mitral valve prolapse have been studied extensively, the echocardiographic detection of the association between mitral prolapse and asymmetric septal hypertrophy has not been described previously. Jeresaty observed a high incidence of mitral valve prolapse on the angiograms of patients with idiopathic hypertrophic subaortic stenosis (IHSS) and in subjects with "nonobstructive left ventricular hypertrophy with obliteration of the apex."14 He postulated that a small left ventricular volume which is a common feature of IHSS may be responsible for the genesis of mitral valve prolapse in this group of patients. He termed this type of prolapse secondary ballooning as opposed to primary ballooning which is associated mainly with myxomatous degeneration of the valve. Creiley and Kissel stated that mitral valve prolapse was due to "ventriculovalvular disproportion in which the valve is 'too big' for the ventricle or the ventricle is 'too small' for the valve during the course of ventricular systole.15 They felt that in IHSS there was excessive contraction of the inferobasal left ventricle which pushed the papillary muscle inward permitting posterior leaflet prolapse. It is of interest that in nine of our patients, the end-systolic dimension (LVES) was less than 2.5 cm, which was the lower limit observed in a group of normal subjects studied in our laboratory. Thus, it is likely that the end-systolic volume was decreased (less than the lower limit of

![Figure 1. Echocardiogram of patient 15 shows mitral valve prolapse (vertical arrow). AMV = anterior mitral leaflet, PMV = posterior mitral leaflet.](http://circ.ahajournals.org/)

![Figure 2. Left ventricular echocardiogram of patient 15 illustrates asymmetric septal hypertrophy. IVS = interventricular septum, ENDO = endocardium, EPI = epicardium.](http://circ.ahajournals.org/)
normal) in these patients. It is conceivable that in this group of patients the etiology of their mitral valve prolapse was ventriculovalvular disproportion in which the ventricle was too small for the valve, as stated by Criley. However, it is possible that in addition to a small ventricular volume, some degree of valvular redundancy may be required to permit the occurrence of prolapse. There were two subsets in our group of patients with ASH. Group 1 consisted of 13 patients who had diminished thickening of the interventricular septum; three of these patients had reduced excursion of the septum. Group 2 patients had normal septal thickening and excursion. Rosen and associates reported a much more marked reduction in septal thickening and excursion in a series of patients with IHSS than was seen in our group. It is conceivable that ASH seen in some of our patients is pathologically different from that seen in IHSS. However, three of our patients (patients 1, 2, 12) clearly had a reduction both in excursion and in systolic thickening of the septum which was similar to that observed in the patients described by Rosen and his co-workers.

Although our patients had ASH none of them had systolic anterior motion of the mitral valve even after provocation with amyl nitrite. This suggests that there was no left ventricular outflow tract obstruction. Septal hypertrophy, together with a ratio of the septum to posterior wall thickness of 1.5 or more, was defined as asymmetric septal hypertrophy. We used the more stringent criterion of 1.5 instead of 1.3 to define ASH because we felt that a ratio of 1.5 is more specific for this entity. In a recent study of patients with systemic hypertension, we observed three patients with a septum to posterior wall ratio of greater than 1.3, but in each instance the ratio was less than 1.5 (unpublished observations). Criley and his associates have made similar observations in a group of patients with malignant hypertension.

Several reports have indicated that atrial and ventricular arrhythmias are common in patients with mitral valve prolapse (MVP). It is conceivable that serious hemo-dynamic deterioration could result if an arrhythmia were to occur in a patient with ventricular hypertrophy and diminished left ventricular compliance. Three of our patients experienced recurrent supraventricular arrhythmias which were associated with syncope. Because of the possibility of serious sequelae, we recommend a careful search for arrhythmias by Holter monitoring and exercise testing, and rigorous antiarrhythmic therapy if rhythm disturbances are detected in patients who have MVP and ASH. Three others had episodes of near syncope which were not accompanied by arrhythmias. These spells were probably due to decreased filling of the poorly compliant left ventricle during tachycardia induced by exercise. It should be stressed that a systematic attempt to define the incidence of arrhythmia was not made in the rest of the patients. Similar symptoms have been described in patients with ASH with or without obstruction to left ventricular outflow. Winkle and others reported syncope during sinus rhythm on long-term ECG monitoring of patients with MVP. It is possible that some of their patients had associated ASH.

It should be emphasized that none of our patients had electrocardiographic or clinical evidence of right ventricular hypertrophy, which has been shown to produce ASH. Four patients had mild systemic hypertension. In three of them the posterior wall thickness was normal and the fourth had a mild increase in wall thickness. Thus, it is unlikely that hypertension played a significant role in the genesis of severe septal hypertrophy in these patients.

Chest pain is a common presenting complaint of patients with MVP. In the majority of patients, the pain lacks the characteristics of angina pectoris. Three of our patients had typical anginal type retrosternal pain. One of these subjects had attacks of rest pain associated with marked ST depression. Coronary arteriography revealed normal coronary arteries. Relative ischemia to the hypertrophied muscle is a possible explanation for this patient's chest pain.

The familial occurrence of ASH has been observed by several workers. Clark and his associates demonstrated that ASH is transmitted as an autosomal dominant trait. One of our patients (#5) gave a strong family history of ASH: her brother (only sibling) and her father had IHSS diagnosed by cardiac catheterization.

In summary, the association between mitral valve prolapse and asymmetric septal hypertrophy was noted on the echocardiograms of sixteen patients. Since MVP predisposes to cardiac arrhythmias which are poorly tolerated in the setting of ventricular hypertrophy and reduced left ventricular compliance, the recognition of this combination of abnormalities is of clinical importance.

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Phonoangiography by Autocorrelation

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SUMMARY Phonoangiography, as a noninvasive quantitative analysis of arterial bruits, was conducted just prior to standard invasive radiographic angiography in 135 patients. Sound records from 162 carotid arteries were analyzed with a new processing technique employing a high speed analog acoustic analyzer, the autocorrelator. In 18 arteries with carotid stenosis, a correlation coefficient of 0.87 resulted between phonoangiographic diameter predictions and radiographic diameter estimates. Bruit analysis identified two patients with patent lumen diameters, but tortuous carotid arteries. One hundred thirty-three carotid arteries had no bruits that could be analyzed, but angiograms showed no extracranial stenosis. Four arteries from which bruits could not be analyzed were found to be totally occluded. Carotid phonoangiography appears applicable in approximately one of seven patients now requiring angiography of head and neck vessels. When applicable, phonoangiography is significantly correlated with radiographic findings.

IT IS DESIRABLE to supplement arteriographic diagnosis of vascular disorders with reliable noninvasive techniques. Phonoangiography, described by Lees and co-workers, is a procedure for quantitative analysis of arterial bruits. Analysis of 50 carotid bruits in 48 patients by these investigators provided estimates of residual carotid lumen diameters which differed from the radiographic values by less than 1 mm in 83% of cases studied. The patients reported by Lees were selected because of identified cervical bruits. We report here a second evaluation of the method in a patient group selected because of a need for cerebral or cervical angiography. We have employed a different procedure for bruit analysis. Our findings confirm the validity of the original hypothesis underlying phonoangiography and extend previous observations regarding a problem with the method when stenosis severely reduces flow. In addition, we have found that two bruits associated with tortuous carotid arteries, but patent lumen, could be differentiated from major stenosis by this method.

Methods

Patients

During a period of four months, patients over 40 years of age undergoing carotid and/or aortic arch arteriography at Los Angeles County — USC Medical Center were sound-recorded and evaluated for turbulent bruits. In addition, 51 patients under 40 years whose clinical histories suggested vascular disease and 36 patients with trauma involving the arteries of the neck were recorded.

Procedures

A miniature Sony electroet condenser microphone model ECM-50 was adapted for skin application with a parabol-shaped polycarbonate bell. Acoustic signals were selectively amplified to enhance turbulent spectral characteristics. A variable-bandwidth Princeton Applied Research model 113 preamplifier was set for low-frequency roll-off at 1000 Hz, a high-frequency roll-off at 3000 Hz and a gain setting of 500. Signals were stored with a Tandberg Cross-Field Series 3300X tape recorder.

Transmitted heart sounds and background noise were defined initially by inching the microphone from the base of the neck to the angle of the mandible and accepting for analysis only those individuals whose bruits were loudest near the carotid bifurcation. In subsequent batch processing of recorded data, bruits were rescreened for predominantly turbulent flow sounds on an oscilloscope display. Figure 1 illustrates an acceptable bruit sequence.

Criteria for acceptability were low background noise and the abrupt onset and gradual decay of sound, each sound burst occurring periodically with each pulse wave. Recordings of bruits were discarded when the bruit signal was low in relation to baseline noise or if the only significant signal was the transmitted pulse waves or heart sounds (fig. 2).
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