Detection of Dysrhythmia in Pediatric Patients with Mitral Valve Prolapse

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SUMMARY  The association of dysrhythmias with mitral valve prolapse in pediatric patients was investigated by graded treadmill exercise testing and 24-hour ambulatory ECG monitoring. Twenty-six unselected patients with a clinical diagnosis of mitral valve prolapse confirmed by echocardiography or angiography or both were studied. On 24-hour ambulatory ECG monitoring, six patients (23%) had potentially serious ventricular dysrhythmias. Five of these patients also manifested ventricular dysrhythmias on treadmill exercise testing. Neither standard ECG abnormalities nor clinical symptoms correlated with detected dysrhythmias. This study shows that potentially serious ventricular arrhythmias are common in pediatric patients with mitral valve prolapse and that ambulatory ECG monitoring and treadmill exercise testing are useful for detecting dysrhythmias.

MITRAL VALVE PROLAPSE is now recognized as a common form of mitral valve dysfunction. Although previous reports suggest that the prognosis for isolated mitral valve prolapse in childhood is excellent,1,2 syncope and sudden death have been reported and are felt to be related to cardiac arrhythmias.2 The association of mitral valve prolapse with arrhythmias manifested on ambulatory monitoring or stress testing has been well substantiated in adults.4,5 To date, there have been few reports concerning this association in pediatric patients.6 In this paper we report the results of treadmill exercise testing and 24-hour ambulatory ECG monitoring in a group of unselected pediatric patients with mitral valve prolapse.

Methods

Patient Population

The study group consisted of 26 consecutive patients seen during a 1-year period in the Pediatric Cardiology Clinic with an auscultatory diagnosis of isolated mitral valve prolapse. In each patient, the diagnosis of mitral valve prolapse had been confirmed by echocardiography or angiography or both.10-13 Nineteen patients had been followed with a diagnosis of mitral valve prolapse; in seven patients the diagnosis was made during the study. All patients...
had been referred for evaluation of a murmur. The characteristics of the patients as a group are summarized in table 1. No patient had stigmata suggestive of Marfan's syndrome. Fourteen of 26 patients were below the fiftieth percentile for height; 15 of 26 patients were below the fiftieth percentile for weight. Four of 26 had minor thoracic skeletal deformities. Twenty-two patients were entirely asymptomatic; two patients complained of nonspecific chest pain and two patients, one of whom had documented supraventricular tachycardia, complained of palpitations. Of these four patients, two complained of shortness of breath with exertion.

Data Collection

All patients had a standard 12-lead ECG, a maximal treadmill exercise test and a 24-hour ambulatory ECG. The 12-lead ECG was performed using a Hewlett-Packard 1151A electrocardiographic unit with the patient in the supine position. The treadmill exercise tests were performed on a Quinton Instruments Cardio Exercise Treadmill, Model 18-54, using the Bruce protocol and normal pediatric values. Modified lead V₆ was monitored continuously during exercise to completion of the staged test or to termination because of dysrhythmia or exhaustion, and during 10 minutes of recovery by radio-telemetry on a Hittman Medcraft Accuscan Recorder. Samples were recorded for 30 seconds at the end of each minute of exercise and recovery, and any arrhythmias observed were recorded using a 5-second delay system. The 24-hour ambulatory ECGs were recorded using a single-channel Hittman Medcraft Accutape 24 recorder and a modified V₆ system. Patients followed their normal daily routine and kept a record of their activities and any symptoms experienced. No patient received antiarrhythmic medications before data collection. The magnetic tapes containing the 24-hours of ambulatory ECG were processed at 64 times real time. A technician recorded a 1-minute segment during each hour of the record. Dysrhythmias were recorded in their entirety if a specific onset and conclusion could be detected. Samples were also recorded whenever the patient's record indicated the presence of symptoms. In reviewing the results of the treadmill exercise tests and 24-hour ambulatory monitoring, the criteria of Gillette for evaluating the significance of ventricular extrasystoles (VPCs) were used. VPCs were considered serious when they were multifocal or accentuated with exercise or associated with a variable coupling interval, a prolonged resting Q-T interval, or the R-on-T phenomenon.

Table 1. Characteristics of Patient Population (n = 26)

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at diagnosis—4 months to 17 years</td>
<td>(mean 9.6 yrs)</td>
</tr>
<tr>
<td>Age at study—7-18 years</td>
<td>(mean 13.8 yrs)</td>
</tr>
<tr>
<td>Duration of follow-up—1-13.5 years</td>
<td>(mean 4.5 yrs)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>4</td>
</tr>
<tr>
<td>Female</td>
<td>22</td>
</tr>
<tr>
<td>Cardiac exam</td>
<td></td>
</tr>
<tr>
<td>Midsystolic click</td>
<td>17</td>
</tr>
<tr>
<td>Late systolic murmur</td>
<td>15</td>
</tr>
<tr>
<td>Pansystolic murmur</td>
<td>8</td>
</tr>
<tr>
<td>Thoracic skeletal deformity*</td>
<td></td>
</tr>
<tr>
<td>Pectus excavatum</td>
<td>3</td>
</tr>
<tr>
<td>Scoliosis</td>
<td>1</td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>21</td>
</tr>
<tr>
<td>Palpitations</td>
<td>2*</td>
</tr>
<tr>
<td>Chest pain</td>
<td>2</td>
</tr>
<tr>
<td>Dyspnea on exertion</td>
<td>2</td>
</tr>
<tr>
<td>Echocardiogram of mitral valve</td>
<td></td>
</tr>
<tr>
<td>Pansystolic humping</td>
<td>12</td>
</tr>
<tr>
<td>Late systolic dip</td>
<td>14</td>
</tr>
<tr>
<td>Angiography</td>
<td>8</td>
</tr>
</tbody>
</table>

*No patient had stigmata of Marfan's syndrome.
†One patient had infrequent documented episodes of paroxysmal supraventricular tachycardia.

Results

Twelve-lead ECG

The standard 12-lead ECGs were entirely normal in 14 patients. One patient had coronary sinus rhythm. Rare unifocal VPCs were recorded in another patient. In eight patients (31%), ST-T-wave abnormalities were recorded in the inferolateral leads; two of these had mild right-axis deviation. Two of these patients had a pronounced U wave, as did an additional two patients with no other abnormality. No patient had electrocardiographic evidence of chamber hypertrophy.

Treadmill Exercise Test

Maximal treadmill exercise tests were obtained in 24 patients; two patients did not achieve 90% of predicted peak heart rate, but patient effort was felt to be submaximal and their results were not included in the analysis. One test was terminated by the supervising physician because of dysrhythmia. No patient had significant ST-segment depression, although T-wave inversion with exercise was common. No patient developed symptoms during the test. Endurance times for the 23 patients were compared with normal values. Two of 23 patients were below the tenth percentile; seven were in the tenth percentile; four were in the twenty-fifth percentile; six were in the fiftieth percentile; three were in the seventy-fifth percentile; and one was above the ninetieth percentile.

In 17 patients (67%), no dysrhythmias were recorded during exercise or recovery. The dysrhythmias found in the remaining nine patients are...
TABLE 2. Summary of Dysrhythmia Detection

<table>
<thead>
<tr>
<th>Dysrhythmia</th>
<th>12-lead ECG</th>
<th>Treadmill exercise</th>
<th>24-hour ambulatory ECG</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial (n = 12)*</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Junctional rhythm</td>
<td>0</td>
<td>1</td>
<td>7</td>
</tr>
<tr>
<td>Sinus tachycardia</td>
<td>0</td>
<td>-</td>
<td>5</td>
</tr>
<tr>
<td>Atrial premature contraction</td>
<td>0</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Supraventricular tachycardia</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Ventricular (n = 12)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Premature ventricular contraction</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unifocal</td>
<td>1</td>
<td>6</td>
<td>8</td>
</tr>
<tr>
<td>With R on T</td>
<td>0</td>
<td>3†</td>
<td>2</td>
</tr>
<tr>
<td>With variable coupling interval</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Multifocal, ventricular tachycardia</td>
<td>0</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Bigeminy</td>
<td>0</td>
<td>1</td>
<td>4‡</td>
</tr>
</tbody>
</table>

*Four of these patients also had ventricular dysrhythmias. 
†In one of these patients, the R wave fell on the downslope of the T wave at the initiation of a U wave.
‡Two of these patients had unifocal and two had multifocal premature ventricular contractions.

summarized in table 2. One patient had rare atrial premature complexes (APCs) during recovery. One patient whose basic rhythm was coronary sinus bradycardia had junctional rhythm during recovery. Seven patients (27%) had exercise-related ventricular dysrhythmias, and in five (19%), the dysrhythmias were potentially serious. Frequent multifocal VPCs developed in one patient within 1 minute of stage 2 of exercise (fig. 1). Four patients had unifocal VPCs during recovery; however, in one of these the R-on-T phenomenon was present and in another, the R wave fell on the downslope of the T wave at the onset of a U wave (fig. 2). Unifocal VPCs developed in one patient at peak exercise and disappeared during recovery. One patient manifested bigeminy with R-on-T during recovery. In this group of patients, 19% manifested potentially serious ventricular arrhythmias on treadmill exercise testing.

Twenty-four-hour Ambulatory ECG

Adequate records for analysis were obtained in all patients. As shown in normal young adults,17 considerable variation in sinus rate and bradyarrhythmias, particularly during sleep, were common. The fastest rates were sinus in all patients; the range was 87–176 beats/min (mean 142 beats/min). The slowest rates were recorded during sleep in all patients, with a range of 43 (junctional) to 85 beats/min (mean 61 beats/min). T-wave variations were frequently recorded.

In six patients, normal sinus rhythm was recorded throughout the 24-hour period. The dysrhythmias detected in the remaining 20 patients are summarized in table 2. Twelve patients manifested atrial dysrhythmias; in four of these, ventricular ectopic beats were also recorded. Six patients manifested sinus bradycardia with periods of junctional rhythm during sleep. One patient had a single episode of profound sinus bradycardia with junctional escape associated with “faintness” felt to be secondary to sudden orthostatic change. Long runs of junctional rhythm with activity were recorded in one patient (fig. 3). Five patients had an inappropriate sinus tachycardia to as high as 170 beats/min with minimal exertion (figs. 3 and 4). Three patients had APCs. Two brief episodes of ectopic supraventricular tachycardia were recorded without symptoms in one patient.

Ventricular dysrhythmias were recorded in 12 children, including four who had atrial dysrhythmias. Eight patients had unifocal VPCs; of these, two had
R-on-T and one had a variable coupling interval (fig. 4). In four children, frequent multifocal VPCs were recorded; ventricular couplets were recorded several times in one of these patients and R-on-T was recorded repeatedly in two patients (figs. 3 and 5). Runs of ventricular bigeminy were present in two patients with unifocal VPCs and in two with multifocal VPCs. Using the criteria of Gillette, six of 26 patients (23%) manifested potentially serious ventricular dysrhythmias on 24-hour ambulatory ECG monitoring.

**Correlation of Patient Group Characteristics with Noninvasive Assessment**

No patient with serious ventricular dysrhythmias on either exercise testing or ambulatory monitoring was less than 14 years of age. The mean age for the patients with serious ventricular dysrhythmias was 15.6 years (range 14–18 years), as compared with 12.8 years (range 7–17 years) for the remainder of the group ($p < 0.01$). When the patients with ventricular dysrhythmias were grouped, the mean for the fastest heart rate recorded on ambulatory 24-hour ECG was 153 beats/min (range 130–175 beats/min); this differed significantly ($p < 0.01$) from the peak mean heart rate of 137 beats/min (range 87–176 beats/min) for the remainder of the group. There was no significant difference between the sleeping heart rates of the two groups. Two of the four patients with symptoms had serious ventricular dysrhythmias on evaluation. Four patients with serious ventricular dysrhythmias were entirely asymptomatic. Although eight patients had ST-T-wave abnormalities in the inferolateral leads on their 12-lead ECG, only two of these patients manifested a significant dysrhythmia during treadmill exercise testing or ambulatory 24-hour ECG. In all five patients who manifested potentially serious ventricular dysrhythmias during treadmill exercise testing, the dysrhythmias were also detected on ambulatory 24-hour ECG. In one other patient, ventricular dysrhythmia was detected only during ambulatory monitoring.

**Discussion**

In this study of children with mitral valve prolapse, we documented potentially serious ventricular arrhythmias in 19% on treadmill exercise testing and in 23% on continuous 24-hour electrocardiographic monitoring. Our results confirm those in adults, in whom an incidence as high as 75% of potentially life-threatening ventricular dysrhythmias has been recorded. No patient manifested dysrhythmias with treadmill exercise testing that were not present on ambulatory monitoring. In our patients, as in reported adult series, the 24-hour ambulatory ECG was the more sensitive test.

Although 31% of our patients had inferolateral ST-T-wave abnormalities on their 12-lead ECG, only two of these manifested a significant ventricular dysrhythmia on exercise stress testing or ambulatory ECG. This is contrary to reported experience in adults. Four patients had symptoms that might have been related to dysrhythmias, but only two of these patients had dysrhythmias during exercise stress testing or ambulatory monitoring. Four patients with serious dysrhythmias were entirely asymptomatic. Neither clinical symptoms nor ECG findings correlated with the dysrhythmias demonstrated.

**Figure 2.** Representative ECG strips from the treadmill exercise test of an asymptomatic 15-year-old girl revealed no arrhythmias at rest or during exercise. However, between 2 and 4 minutes of recovery, unifocal ventricular premature complexes that fell on the onset of the U wave were recorded.
A. 9:30 A.M.

B. 10:01 A.M.

C. 10:03 A.M.

D. 11:32 A.M.

A. 4:00 P.M.

B. 7 P.M.

C. 9:34 P.M.

D. 6:25 A.M.

E. 8:00 A.M.

F. 1:47 P.M.

A. 2:30 P.M.

B. 5:50 P.M.

C. 11:50 P.M.

suggested that the more elaborate studies are necessary for detecting dysrhythmias.

In this series, the mean age of the patients with serious dysrhythmias was significantly older than that of the remaining patients. Coupled with the lower overall incidence of dysrhythmias in our patients than in adult series, this suggests that the development of serious dysrhythmias may be an age-related phenomenon. Continued follow-up for detecting dysrhythmia in patients with mitral valve prolapse appears to be indicated. The patients with serious ven-

Figure 3. Selected tracings from the 24-hour ambulatory ECG of a 14-year-old girl who complained of palpitations. The tracings were all taken from a 3-hour period when the patient was in school. Pronounced U waves were present (A). Inappropriate sinus tachycardia (B), multifocal ventricular premature complexes in bigeminy with R-on-T (C), and junctional rhythm (D) were recorded.

Figure 4. Representative ECG strips from an asymptomatic girl on 24-hour ambulatory ECG. Although ventricular premature complexes were not frequent, a variable coupling interval (B, C) and bigeminy (F) were recorded. Inappropriate sinus tachycardia (E) and variable T waves were also recorded.

Figure 5. Selected ECG strips from the 24-hour ambulatory ECG of a 17-year-old patient who had a history of nonspecific chest pain. Multifocal ventricular premature complexes with bigeminy and R-on-T were recorded with activity (B) and during sleep (C). The patient was asymptomatic during this 24-hour period.
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Ventricular dysrhythmias had a significantly higher mean fastest heart rate than did the remainder of the group. This finding may be evidence for high adrenergic tone, as documented in adults with mitral valve prolapse.18

With heightened awareness and noninvasive confirmation by echocardiography, the diagnosis of mitral valve prolapse in children is being made with increasing frequency. As in adults, there is poor correlation between symptoms and detected dysrhythmias. The prognostic implications of the dysrhythmias manifested by these patients are not known; reported cases of sudden death suggest that they may be important. In view of the potentially serious dysrhythmias documented in 19% and 23% of our relatively small group by two methods, ambulatory ECG monitoring and exercise testing appear to be important in assessing pediatric patients with mitral valve prolapse.

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