Paroxysmal Supraventricular Tachycardia
in Patients with Mitral Valve Prolapse

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SUMMARY The mechanism of supraventricular tachycardia was evaluated in twelve patients with mitral valve prolapse utilizing standard intracardiac recording and stimulation techniques. Eight patients had normal electrocardiograms, three had a short PR interval and normal QRS (Lown-Ganong-Levine syndrome) and one had Wolff-Parkinson-White type A. Six of the eight patients with normal electrocardiograms were demonstrated to have atrioventricular bypass tracts. Five patients had A-V nodal re-entrant supraventricular tachycardia. In two patients the bypass tract could be demon-

strated to conduct antegrade when the left atrium was paced via the coronary sinus, while in four the bypass tract only conducted retro-

gradely. In all seven patients with atrioventricular bypass tracts, the accessory pathway was left-sided. We conclude that a) supraventricular tachycardia in the mitral valve prolapse syndrome appears related to a high frequency of bypass tracts; b) electrophysiological studies are required to diagnose these bypass tracts; and c) the atrioventricular bypass tracts may be related to the mitral valve ab-

normality since they are always left-sided.

MATERIAL VALVE PROLAPSE has become recognized as the most prevalent cardiac valvular abnormality, affecting perhaps 6% of the population. While ventricular pre-
mature depolarizations are most frequently noted, parox-
ysmal supraventricular tachycardia is probably the most commonly sustained tachyarrhythmia. The present study was undertaken to analyze the mechanisms of supraventricu-
lar tachycardia that occur with mitral valve prolapse and compare them to the mechanisms that are responsible for supraventricular tachycardias in the population at large.

Methods and Material

Electrophysiologic studies were performed in twelve patients with auscultatory and/or echocardiographic find-
ings of mitral valve prolapse in whom paroxysmal supra-
ventricular tachycardia had been documented (table 1). There were seven women and five men, aged from 17 to 63 years. Their resting electrocardiograms demonstrated a short PR interval (≤ .12 sec) with a normal QRS in three, Wolff-Parkinson-White type A in one, and normal in the remaining eight.

Each patient was studied in the postabsorptive nonsedated state after informed consent was obtained. Antiarrhythmic agents were discontinued 48 hours prior to study. Quadrupolar electrode catheters were percutaneously in-
serted through the femoral and/or antecubital veins and positioned under fluoroscopic control in the right atrium, right ventricle, and coronary sinus. One pair of electrodes was used for recording and the other for stimulation. A tripolar catheter was percutaneously introduced through the right femoral vein and positioned across the tricuspid valve to obtain a His bundle electrogram. The intracardiac elec-

trograms were simultaneously recorded with multiple sur-
face electrocardiographic leads, usually I, aVF, and V5, and displayed on a switch-beam oscilloscope (E for M, DR16).

In each patient the mechanism of the tachycardia was determined using intracardiac recordings and analyzing the response to atrial and ventricular pacing and extrastimuli delivered by a custom designed programmable stimulator using an optically isolated constant current source (Bloom Associates). Particular attention was paid to the mode of initiation of supraventricular tachycardia, the atrial activation sequence during the tachycardia, the effects of developing bundle branch block during the tachycardia, and the effects of stimulation (atrial and ventricular) during the tachycardia.

The mechanisms suggested by the electrocardiographic findings included: a) A-V nodal re-entry; b) A-V nodal re-entry associated with the Lown-Ganong-Levine syndrome; and c) atrio-nodal-ventriculo-atrial circus movement re-entry characteristic of the Wolff-Parkinson-White syndrome. Criteria used to define these mechanisms were as follows:

Re-entry Within the A-V Node

1. Dual A-V nodal pathways.
2. Echo zone and initiation of supraventricular tachycardia dependent upon attaining a critical A-H interval.
3. Normal retrograde atrial activation during supra-

ventricular tachycardia.
4. Recording of retrograde atrial electrogram prior to or simultaneous with the onset of the QRS.

Atrio-Nodal-Ventriculo-Atrial Re-entry (Wolff-Parkinson-

White Syndrome)

1. Supraventricular tachycardia dependent on critical A-V interval only after antegrade block in bypass tract.
2. Eccentric retrograde atrial activation sequence during supraventricular tachycardia.
3. Ability to pre-excite the atrium with ventricular premature depolarizations during supraventricular tachycardia at a time when the bundle of His is either refrac-

tory or participating in antegrade conduction.
4. Prolongation of ventriculo-atrial conduction with the development of functional bundle branch block during supraventricular tachycardia.

Results

A-V Nodal Re-entry

Two patients demonstrated typical A-V nodal re-entry. The spontaneous occurrence of paroxysmal supraventricular
Atrio-Nodal-Ventriculo-Atrial Re-entry

Seven patients demonstrated an atrioventricular bypass tract participating in the tachycardia. The bypass tract was manifest in the one patient who demonstrated a Wolff-Parkinson-White type A pattern on the electrocardiogram. In two patients the bypass tract was latent, with the characteristic delta wave and bizarre QRS brought out by coronary sinus pacing (fig. 2). The supraventricular tachycardia in the three patients with manifest or latent pre-excitation was demonstrated to incorporate the accessory pathway in the re-entrant circuit. In four patients (cases 9–12) an atrioventricular bypass tract was concealed during antegrade stimulation studies, but was shown to be operative during supraventricular tachycardia by a) eccentric retrograde atrial activation during the tachycardia (fig. 3); b) prolongation of ventriculo-atrial conduction with the development of left bundle branch block (fig. 4); and c) the ability to prematurely depolarize the atria with ventricular stimulation during the tachycardia (fig. 5). The tachycardia rates ranged from 170–230 beats/min in this group. In the two patients with the fastest rate of supraventricular tachycardia (230/min) the bypass tract was not manifest on the surface electrocardiogram (cases 8 and 11). In all cases with a functioning atrioventricular bypass tract, the location was demonstrated to be left-sided by a retrograde atrial activation pattern with the earliest activation in the left atrium.

**TABLE 1. Patient Data**

<table>
<thead>
<tr>
<th>Patient/Sex/Age</th>
<th>ECG</th>
<th>Rate of SVT (beats/min)</th>
<th>Clinical findings</th>
<th>Echo findings</th>
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</thead>
<tbody>
<tr>
<td>1/M/48</td>
<td>NL</td>
<td>200</td>
<td>MSC, LSM</td>
<td>PLP and ALP</td>
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<td>2/F/17</td>
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<td>150</td>
<td>Mult. MSC, LSM</td>
<td>PLP and ALP</td>
</tr>
<tr>
<td>3/M/40</td>
<td>Short PR, NL QRS</td>
<td>200</td>
<td>LSM</td>
<td>PLP</td>
</tr>
<tr>
<td>4/F/28</td>
<td>Short PR, NL QRS</td>
<td>150</td>
<td>Mult. MSC, LSM</td>
<td>PLP and ALP</td>
</tr>
<tr>
<td>5/F/37</td>
<td>Short PR, NL QRS</td>
<td>162</td>
<td>MSC, LSM</td>
<td>PLP and ALP</td>
</tr>
<tr>
<td>6/F/53</td>
<td>WPW A</td>
<td>180</td>
<td>LSM</td>
<td>PLP</td>
</tr>
<tr>
<td>7/M/35</td>
<td>1° AVB, IVC, LAH</td>
<td>200</td>
<td>early nonejection systolic mmmur diastolic fluttering holosystolic PLP and ALP, with</td>
<td></td>
</tr>
<tr>
<td>8/F/28</td>
<td>NL</td>
<td>230</td>
<td>Mult. MSC, LSM</td>
<td>PLP</td>
</tr>
<tr>
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<td>NL</td>
<td>180</td>
<td>MSC, LSM</td>
<td>PLP</td>
</tr>
<tr>
<td>10/M/63</td>
<td>IMI</td>
<td>170</td>
<td>MSC, LSM</td>
<td>PLP</td>
</tr>
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<td>230</td>
<td>MSC, LSM</td>
<td>PLP</td>
</tr>
<tr>
<td>12/M/39</td>
<td>NL</td>
<td>200</td>
<td>LSM</td>
<td>PLP and ALP</td>
</tr>
</tbody>
</table>

Abbreviations: ALP = anterior leaflet prolapse; AVB = atrio-ventricular block; IMI = inferior myocardial infarction; IVC = intraventricular conduction defect; LSM = late systolic murmur; MSC = mid-systolic click; NL = normal; PLP = posterior leaflet prolapse; Mult. = multiple.

tachycardia is demonstrated in figure 1. Both demonstrated dual A-V nodal pathway response characteristic of patients with A-V nodal re-entry as depicted in figure 2. The tachycardia rate in these patients was 150 and 200 beats/min.

Three patients (cases 3, 4, 5) demonstrated the short PR, normal QRS, abbreviated A-V nodal conduction time, and blunted A-V nodal response to atrial pacing characteristic of the Lown-Ganong-Levine syndrome. The tachycardia rates in these patients were 150, 162 and 200 beats/min. A-H intervals in these patients were 50–55 msec and all showed less A-H prolongation in response to rapid atrial pacing than normal. A dual pathway response was demonstrated in each patient with the tachycardia occurring during slow pathway conduction through the A-V node. The atrial activation sequence during the tachycardia was identical to that seen in typical A-V nodal re-entry and atrial activation appeared prior to or at the onset of ventricular depolarization.

**FIGURE 1. Spontaneous A-V nodal re-entrant supraventricular tachycardia (patient 1).** The panel is organized from top to bottom: surface ECG leads I, aVR, Vs, a high right atrial electrocardiogram (HRA), coronary sinus electrogram (CS), His bundle electrogram (HBE) and time lines (T). Subsequent intracardiac records are similarly organised. The first two beats are sinus with a normal AH 80 msec. The third beat is an atrial premature depolarization which results in A-H prolongation and the development of supraventricular tachycardia. Note during supraventricular tachycardia, atrial activation is earliest in the HBE and begins prior to the inscription of the QRS, a diagnostic feature of A-V nodal re-entrant supraventricular tachycardia.
as recorded in the coronary sinus electrogram. Further support of a left-sided bypass tract was the development of prolongation of ventriculo-atrial conduction and cycle length with the development of left bundle branch block in three patients. In none of our patients with bypass tracts was A-V nodal re-entry present.\textsuperscript{16, 17}

Discussion

Paroxysmal supraventricular tachycardia is probably the most common sustained tachyarrhythmia in patients with the mitral valve prolapse syndrome.\textsuperscript{1, 3}

The surface electrocardiograms were misleading regarding the mechanism of supraventricular tachycardia in that eight of our twelve patients demonstrated no abnormalities suggestive of pre-excitation. Detailed electrophysiologic evaluation demonstrated that only five patients had A-V nodal re-entrant supraventricular tachycardia. In each of these patients a bypass tract in any location, free wall or septal, was excluded by the recording of retrograde atrial activation prior to the ventricular electrogram (fig. 1). In seven

an atrioventricular accessory pathway formed one limb of the re-entrant circuit.

Of the seven patients with atrioventricular bypass tracts only one had electrocardiographic evidence of the Wolff-Parkinson-White syndrome. In two, antegrade pre-excitation was only manifest during atrial pacing and was much more evident when the left atrium (via the coronary sinus) was paced.\textsuperscript{12, 14, 16} Of great interest was the absence of any evidence of antegrade pre-excitation in the remaining four patients with atrioventricular bypass tracts. In these patients the accessory pathway was concealed during antegrade stimulation studies.\textsuperscript{12-18} The participation of these bypass tracts in the supraventricular tachycardia was demonstrated by a) eccentric retrograde atrial activation; b) the ability of a ventricular premature depolarization to pre-excite the atrium during the tachycardia; and c) prolongation of ventriculo-atrial conduction time with the development of left bundle branch block.

In all patients with atrioventricular bypass tracts, the accessory pathway was left-sided. This is intriguing in the light of the known abnormalities of the mitral apparatus.
Gallagher also found only left-sided bypass tracts in seven patients with mitral prolapse and the Wolff-Parkinson-White syndrome. This situation is analogous to Ebstein's anomaly of the tricuspid valve which, when associated with an atrioventricular bypass, is always associated with a type B Wolff-Parkinson-White pattern which is due to a right-sided atrioventricular bypass tract. In both instances the developmental abnormality giving rise to the valvular malformation may be related to the associated electrophysiologic findings.

**Significance of Results**

In 60% of our patients some form of pre-excitation was present. Since a rational approach to therapy of arrhythmias is predicated on the understanding of their underlying mechanisms, it is obviously important that patients with mitral valve prolapse and supraventricular tachycardia be evaluated for the presence of bypass tracts before therapeutic interventions are begun. Drugs such as digitalis and propranolol that may be extremely useful in either A-V nodal re-entrant tachycardia or circus-motion re-entry using an atrioventricular bypass tract, may be dangerous in the presence of antegrade conduction over an atrioventricular bypass tract. Should any of these patients also have atrial fibrillation or atrial flutter with rapid antegrade conduction over the accessory pathway, digitalis may facilitate conduction over the bypass tract and lead to ventricular fibrillation. The absence of electrocardiographic evidence of the Wolff-Parkinson-White syndrome should not be taken as evidence that such an event could not occur. Three months prior to study, case 7, who only demonstrated evidence of pre-excitation with coronary sinus pacing, had an episode of ventricular flutter-fibrillation induced by a bout of atrial flutter-fibrillation with rapid conduction down his accessory pathway in the presence of digitalis. This mechanism may be responsible for some episodes of sudden death in this syndrome.

**Relationship to Common Mechanisms of Supraventricular Tachycardia in the General Population**

The incidence of bypass tracts participating in supraventricular tachycardia associated with mitral valve prolapse is inordinately high. In our experience, A-V nodal re-entry is the mechanism in two-thirds of the episodes of paroxysmal supraventricular tachycardia in the general population while bypass tracts are operative in only 20% of cases. Wu and Akhtar (unpublished data) have similar findings. This increased frequency of bypass tracts should be taken into consideration when therapy is required or has failed previously. Electrophysiologic investigation should probably be undertaken in such patients.

**References**

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