Role of Mahaim Fibers in Cardiac Arrhythmias in Man

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SUMMARY Twelve patients with evidence of Mahaim fibers are reported, six with nodoventricular (NV) fibers and six with fasciculoventricular (FV) fibers. All patients with NV fibers had left bundle branch block morphology, and a sustained reentrant tachycardia with this morphology was proved in each case. In three of the six, atrioventricular dissociation occurred during tachycardia. We postulate that the mechanism of this tachycardia is a macroreentry circuit using the NV fiber for the antegrade limb and the His-Purkinje system with a portion of the atrioventricular node for the retrograde limb. ECGs of patients with FV fibers were varied, suggesting a functional relation to the right or left side of the septum. No direct relationship of FV fibers to observed arrhythmias could be found.

THE ROLE of Mahaim fibers in the genesis of cardiac arrhythmias in man has been controversial since they were first described. The initial report was limited to fibers connecting the His bundle to the septum, but this was soon broadened to include fibers connecting the atrioventricular (AV) node to the septum. Although the existence of these fibers has been confirmed anatomically, their functional significance is unclear. In this report we present electrophysiologic studies in patients with Mahaim fibers that demonstrate that some Mahaim fibers participate in arrhythmias in man.

Methods

Patient Selection

Rhythm disorders associated with Mahaim fibers form a subset of a larger group of disorders known as the preexcitation syndromes. The 12 patients in this study were selected from 308 patients referred to Duke University Medical Center between 1968 and November 1979 for evaluation and treatment of various preexcitation syndromes. Of these, 250 had at least one accessory atroventricular connection, of which conducted only in the retrograde direction; 46 had accelerated conduction in the AV node, including five patients with concealed accessory atroventricular pathways; and 12 had Mahaim fibers, including three with accessory atroventricular pathways.

All patients were admitted to the hospital and placed on continuous telemetry; all cardioactive medications were discontinued. Before cardiac catheterization, all had history and physical examination, routine blood work, ECG, chest x-ray, and two-dimensional echocardiography using a microcavitation technique. All patients gave informed consent before catheterization.

Electrophysiologic Study

All patients were studied in the postabsorptive, nonsedated state using techniques previously described. Multiple electrode catheters were used to record and pace from the right atrium, the left atrium via the coronary sinus, and the right ventricle. Observations were made during overdrive pacing and refractory period determination from all three locations, during tachycardia and after deliberate induction of atrial fibrillation.

Terminology

Recently, Anderson et al. suggested that there are two main anatomic types of Mahaim fibers — nodoventricular (NV) fibers, which arise from the AV node, and fasciculoventricular (FV) fibers, which arise from the His bundle and bundle branches. Because the functional consequences are significant, we have found it useful to consider the two groups separately according to an anatomic and functional classification (fig. 1). Anderson et al. proposed two varieties of NV fibers — those which arise from the transitional zone of the AV junction and those which arise from the deep, compact nodal portion of the AV junction.

NV fibers, depending on level of takeoff relative to the area of physiologic delay, can be associated with either a short or normal PR interval. Ventricular activation in this case results from fusion of impulse propagation via the NV fiber and the normal conduction system, resulting in QRS complexes with varying degrees of anomalous conduction. The PR interval should be normal with isolated FV fibers because they arise from the His bundle and bundle branches, unless another anomaly of AV conduction is present. The
QRS complex associated with FV fibers may be anomalous, depending on the level of takeoff, but variable fusion is not expected and the QRS morphology should be reasonably constant. Extrasystoles arising in the His bundle or pacing of the His bundle in the case of the NV fibers should normalize the QRS. In the case of FV fibers, the anomalous appearance of the QRS should persist unless the extrasystole arises below the takeoff of the FV fiber, as proposed by Pick and Katz from the study of scalar ECGs. We subdivided our patients with Mahaim fibers according to this proposed functional classification (fig. 1) and examined the possible role of Mahaim fibers in the observed arrhythmias.

Description of the ECG morphology resulting from preexcitation due to FV fibers presents a semantic dilemma. An ECG pattern of left bundle branch block (LBBB) can result whenever the left ventricle is activated after the right ventricle. This can be due to relatively early activation of the right ventricle or relatively late activation of the left ventricle. Thus, preexcitation on the right side or conduction delay or block on the left side can result in similar ECG patterns. In our series, NV fibers exclusively resulted in preexcitation of the right ventricle. We described the resulting ECG pattern as left bundle branch block morphology (LBBBM).

Enhanced conduction in the AV node was defined as an abnormality of conduction between the atrium and the His bundle as evidenced by an AH interval in sinus rhythm less than or equal to 60 msec, the lower limit of normal; 1:1 conduction between atrium and His bundle maintained at cycle lengths of right atrial pacing less than 300 msec; and failure of the AH interval to prolong more than 100 msec over the value in sinus rhythm at the shortest cycle length associated with 1:1 conduction. Because atropine or catecholamines can permit rapid conduction in a normal AV node, this definition is functional and does not imply an anatomic substrate.

Results

Twelve patients with Mahaim fibers were studied; six had NV fibers and six had FV fibers. The clinical and electrophysiologic data are presented in tables 1 and 2, respectively.

Nodoventricular Fibers

Studies During Sinus Rhythm/Atrial Pacing

Representative examples of our findings in sinus rhythm/atrial pacing will be presented to facilitate the discussion of group findings. Figure 2 shows the findings in patient 2, a 15-year-old boy who presented...
TABLE 1. Clinical Data

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Associated anomalies</th>
<th>Presenting arrhythmia</th>
<th>Treatment (oral)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>18</td>
<td>F</td>
<td>Ebstein's anomaly, S/P division of Kent bundle</td>
<td>RT with LBBB</td>
<td>Digoxin, 0.25 mg/day</td>
</tr>
<tr>
<td>2</td>
<td>15</td>
<td>M</td>
<td>Ebstein's anomaly</td>
<td>RT with LBBB</td>
<td>Quinidine sulfate, 200 mg q6h Propranolol, 20 mg q6h</td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td>M</td>
<td>None</td>
<td>RT with LBBB</td>
<td>Quinidine gluconate, 330 mg q12h</td>
</tr>
<tr>
<td>4</td>
<td>17</td>
<td>M</td>
<td>Ebstein's anomaly, S/P division of Kent bundle</td>
<td>RT with LBBB</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>14</td>
<td>M</td>
<td>Klippel-Feil syndrome, S/P closure of ASD</td>
<td>RT with LBBB</td>
<td>Quinidine gluconate, 660 mg q12h</td>
</tr>
<tr>
<td>6</td>
<td>34</td>
<td>F</td>
<td>None</td>
<td>RT with LBBB Atrial reentry</td>
<td>Quinidine sulfate, 300 mg q6h</td>
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</table>

Fasciculoventricular fibers

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Associated anomalies</th>
<th>Presenting arrhythmia</th>
<th>Treatment (oral)</th>
</tr>
</thead>
<tbody>
<tr>
<td>7</td>
<td>58</td>
<td>M</td>
<td>EAVN</td>
<td>Atrial fibrillation</td>
<td>Digoxin, 0.25 mg/day Propranolol, 20 mg q6h</td>
</tr>
<tr>
<td>8</td>
<td>15</td>
<td>M</td>
<td>EAVN, S/P division of Kent bundle</td>
<td>Atrial flutter</td>
<td>None (rare attacks)</td>
</tr>
<tr>
<td>9</td>
<td>52</td>
<td>M</td>
<td>EAVN, coronary artery disease</td>
<td>Atrial fibrillation Ventricular fibrillation</td>
<td>Quinidine sulfate, 300 mg q6h</td>
</tr>
<tr>
<td>10</td>
<td>61</td>
<td>F</td>
<td>EAVN, rheumatic heart disease, sick sinus syndrome</td>
<td>Atrial fibrillation</td>
<td>Mitral valve replacement, caryoablation of His bundle</td>
</tr>
<tr>
<td>11*</td>
<td>8</td>
<td>M</td>
<td>EAVN</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>12</td>
<td>15</td>
<td>M</td>
<td>EAVN</td>
<td>Atrial fibrillation Ventricular fibrillation</td>
<td>Quinidine sulfate, 400 mg q6h</td>
</tr>
</tbody>
</table>

*Asymptomatic patient studied because a sister was resuscitated from ventricular fibrillation due to a septal accessory atrioventricular pathway. ECG screening of the family resulted in referral of this patient for study.

Abbreviations: RT = reciprocating tachycardia; LBBB = left bundle branch block; EAVN = enhanced atrioventricular nodal conduction; S/P = status post; ASD = atrial septal defect.

with palpitations. His resting tracing was normal. Atrial pacing when the QRS was normal resulted in prolongation of the AH and AV intervals, displacement of the His deflection into the ventricular electrogram, and increasing degrees of LBBBM as the cycle length of pacing was decreased. Comparable findings were present during the performance of a refractory period from the right atrium. As the coupling interval of the atrial premature depolarization (A3) was gradually reduced (fig. 2, panels 1-4), the A3AH prolonged from 110 to 180 msec, the A3AH prolonged from 145 to 155 msec, the His deflection became displaced into the ventricular electrogram, and the QRS demonstrated fusion, with increasing degrees of LBBBM. A3 blocked at a coupling interval of 320 msec.

These findings are compatible with ventricular fusion due to an accessory pathway arising in the AV node and inserting either in the specialized conducting tissues of the right ventricle or the right ventricular myocardium itself. An alternative explanation would be an accessory AV node or an accessory pathway with "nodal" properties inserting in the right ventricle.

All six patients with NV fibers had normal or prolonged PR intervals. The PA interval was prolonged in two (one with Ebstein's anomaly, one with an atrial septal defect. Fusion was manifested by QRS complexes varying from normal to complete LBBBM, often at comparable cycle lengths. When right atrial pacing was compared with coronary sinus pacing at comparable cycle lengths, the degree of LBBBM was always greater with right atrial pacing. We could not explain this finding on the basis of the AH interval, which suggested that the atrial insertion of the NV fiber was functionally related to the right atrium. In patient 6, the A3V prolonged very little during straight pacing and refractory period determination from the atrium, suggesting an origin in the transitional zone of the AV node. This patient had ventriculatrial dissociation during reciprocating tachycardia (see below), excluding a Kent bundle as the basis for reciprocation.

Our patients with NV fibers typically had LBBBM associated with left-axis deviation during antegrade conduction over the accessory pathway. All previously reported cases also had this finding, which probably indicates that the site of insertion is related to the posteroinferior right ventricle and septum.

The effective refractory period of the NV fiber could be determined in four of six patients. In these four patients, a decrease in the basic cycle length of testing was associated with a shortening of the effective refractory period of the NV fiber.
TABLE 2. Electrophysiologic Data

<table>
<thead>
<tr>
<th>Pt</th>
<th>PR (msec)</th>
<th>PA (msec)</th>
<th>AH (msec)</th>
<th>HV (msec)</th>
<th>QRS duration (sec)</th>
<th>Antegrade Shortest CL with 1:1 conduction (msec)</th>
<th>ERP of Mahaim/CL (msec)</th>
<th>Retrograde Shortest CL with 1:1 conduction (msec)</th>
<th>Retrograde activation sequence</th>
<th>Atrial fibrillation SRR (msec)</th>
<th>ARR (msec)</th>
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</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>NV = 270 AVN &lt; 450</td>
<td>230 (500)</td>
<td>510 Normal</td>
<td>No VA conduction</td>
<td>Atrial flutter with 2:1 block 465</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>0.16</td>
<td>45</td>
<td>80-140</td>
<td>&lt;0-45</td>
<td>0.08-0.16</td>
<td>NV = 400 AVN &lt; 400</td>
<td>291 (450)</td>
<td>300 Normal</td>
<td>No VA conduction</td>
<td>275 450</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>0.17</td>
<td>65</td>
<td>60-110</td>
<td>&lt;0-55</td>
<td>0.08-0.14</td>
<td>NV = 180 AVN &lt; 280</td>
<td>&lt;190 (400)</td>
<td>≤ 300 Normal</td>
<td>No VA conduction</td>
<td>240 322</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>0.14</td>
<td>50</td>
<td>60</td>
<td>&lt;0-40</td>
<td>0.14</td>
<td>NV = 310 AVN &lt; 340</td>
<td>&lt; 260 (500)</td>
<td>No VA conduction</td>
<td>No VA conduction</td>
<td>205 282</td>
<td></td>
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<tr>
<td>4</td>
<td>0.17</td>
<td>30</td>
<td>140-200</td>
<td>&lt;0-55</td>
<td>0.09-0.16</td>
<td>NV = 230 AVN &lt; 300</td>
<td>312 (500)</td>
<td>No VA conduction</td>
<td>No VA conduction</td>
<td>205 282</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>0.17</td>
<td>20</td>
<td>155</td>
<td>&lt;0-55</td>
<td>0.16</td>
<td>NV = 200 AVN &lt; 300</td>
<td>230 (500)</td>
<td>No VA conduction</td>
<td>No VA conduction</td>
<td>205 282</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>0.14</td>
<td>30</td>
<td>70-110</td>
<td>&lt;0-40</td>
<td>0.08-0.16</td>
<td>FV ≤ 300 AVN ≤ 300</td>
<td>355 (600)</td>
<td>380 Normal</td>
<td>290 353</td>
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<tr>
<td>7</td>
<td>0.10</td>
<td>50</td>
<td>40</td>
<td>30</td>
<td>0.10</td>
<td>FV ≤ 280 AVN ≤ 280</td>
<td>&lt;230 (400)</td>
<td>240 Normal</td>
<td>280 395</td>
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<td></td>
</tr>
<tr>
<td>8</td>
<td>0.10</td>
<td>30</td>
<td>15</td>
<td>15</td>
<td>0.11</td>
<td>FV ≤ 220 AVN &lt; 220</td>
<td>260 (400)</td>
<td>300 Normal</td>
<td>205 283</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>0.11</td>
<td>55</td>
<td>25</td>
<td>25</td>
<td>0.10</td>
<td>FV = 290 AVN = 290</td>
<td>&lt;220 (450)</td>
<td>&lt; 240 Normal</td>
<td>260 370</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>0.09</td>
<td>30</td>
<td>30</td>
<td>25</td>
<td>0.12</td>
<td>FV = 210 AVN &lt; 200</td>
<td>&lt;209 (400)</td>
<td>280 Normal</td>
<td>235 286</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>0.07</td>
<td>30</td>
<td>35</td>
<td>15</td>
<td>0.10</td>
<td>FV = 200 AVN = 200</td>
<td>&lt; 250 (400)</td>
<td>&lt; 300 Normal</td>
<td>230 315</td>
<td></td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>0.10</td>
<td>35</td>
<td>25</td>
<td>15</td>
<td>0.12</td>
<td>FV = 200 AVN = 200</td>
<td>&lt; 250 (400)</td>
<td>&lt; 300 Normal</td>
<td>230 315</td>
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</tbody>
</table>

Abbreviations: NV = nodoventricular; CL = cycle length; SRR = shortest RR during AF; FV = fasciculoventricular; ERP = effective refractory period; ARR = average RR during AF; AVN = AV node.

Studies During Ventricular Pacing

Retrograde conduction was absent at all paced cycle lengths in patients 4, 5 and 6. Patients 1, 2 and 3 had retrograde conduction, and the retrograde His deflection always preceded the onset of retrograde atrial depolarization. The retrograde curves were smooth, but a plateau was observed in two of the three patients. The sequence of the retrograde atrial activation was always initiated in the low interatrial septum and spread symmetrically to the lateral right and left atria. The curves were continuous and H₂ always preceded retrograde atrial depolarization.

Studies During Reciprocating Tachycardia

In all six patients with NV fibers, spontaneous tachycardia with LBBBM was documented before study (cycle length 250-400 msec, mean 332 ± 57 msec). In four of six, the same tachycardia could be induced in the laboratory by overdrive atrial pacing (one patient), premature atrial stimulation (two patients), overdrive ventricular pacing (one patient), or premature ventricular stimulation (two patients). In two patients, paroxysms of tachycardia were so frequent that no conclusion could be reached concerning the relationship of the stimulation to the onset or termination of tachycardia. In the two patients in whom ventricular pacing induced tachycardia, minimal prolongation (i.e., 30 msec) of the ventriculoatrial conduction time preceded the onset of tachycardia.

The His deflection never preceded QRS complexes of LBBBM during tachycardia or atrial pacing. An example is shown in figure 3 (patient 2). During a right ventricular refractory period performed at a cycle length of 400 msec, a premature complex at a coupling interval of 310 msec resulted in the onset of reciprocating tachycardia. No His deflection occurred before the onset of ventricular depolarization during tachycardia, and a 1:1 relationship was present between the ventricles and the atria. This finding clearly suggests ventricular tachycardia, especially in patients with ventriculoatrial dissociation. Ventricular tachycardia was excluded by establishing that the LBBBM manifest during tachycardia in every instance could be duplicated by atrial pacing at a variety of pacing cycle lengths.

In patients 2, 3 and 5, a deflection strongly suggestive of origin in the His bundle could be recognized in the ventricular electrogram during tachycardia associated with LBBBM. In the same three patients, a
deflection assumed to arise in the His bundle was also identified during pacing from the right ventricular apex at a cycle length of 400 msec. The interval from the onset of ventricular depolarization to the His deflection during tachycardia and the interval from the onset of ventricular depolarization to the retrograde His deflection during ventricular pacing in three patients were similar: 50 vs 60 msec, 100 vs 95 msec and 70 vs 60 msec (tachycardia vs ventricular pacing). In patients 1, 4 and 6, no retrograde His deflections could be reliably identified. These data support our hypothesis concerning the mechanism of reentry in our patients. It is impossible to say with assurance that the sharp deflections recorded in the ventricular electrogram during tachycardia were retrograde His deflections.

Ventriculoatrial dissociation was present during tachycardia in patients 4, 5 and 6. In patients 1, 2 and 3, a 1:1 ventriculoatrial relation was present. The sequence of atrial activation in the latter three patients was indistinguishable from the expected normal sequence of retrograde conduction via the AV node. The presence of retrograde conduction during tachycardia correlated with the presence of retrograde conduction during ventricular pacing at comparable cycle lengths (table 2).

The presence of a 1:1 ventriculoatrial relationship in three of our patients further suggested the possibility of an antidromic reciprocating tachycardia using a Kent bundle for antegrade conduction and the His bundle and AV node for retrograde conduction. This possibility was excluded because of the relationship of the atrial and ventricular electrograms. The atrial electrogram typically comes immediately before the ventricular electrogram in true antidromic tachycardia; in the cases described here, the long interval for the atrial electrogram to the next ventricular electrogram can be interpreted as a significant AV conduction delay or the apparent time relationship may be spurious. The presence of a long AV conduction time in itself does not exclude a node-like accessory pathway.

Detailed study during sustained reciprocating tachycardia could be obtained in only three of the six patients. In those with 1:1 ventriculoatrial conduction during tachycardia, the tachycardia terminated spontaneously, with or without an accompanying retrograde atrial depolarization.

A single ventricular premature depolarization during reciprocating tachycardia advanced the next cycle in three of three patients (nos. 2, 3, and 5), but retrograde conduction via the His bundle-AV node could not be excluded; termination occurred with a single ventricular premature depolarization in patient 3 and paired ventricular premature depolarizations in patient 5. In patient 2, reciprocating tachycardia could only be terminated by simultaneous AV pacing, which occurred during attempts to pace the His bundle during reciprocating tachycardia.

In two patients, termination of the reciprocating tachycardia was related to manipulation of the His bundle catheter. In one instance, no conduction over the NV fiber was noted for several minutes. In the other patient, maximal preexcitation over the NV fiber was noted immediately after manipulation of the catheter resulted in termination of the tachycardia; pacing of the His bundle at this time showed that catheter-induced right bundle branch block (RBBB) not present earlier in the study had occurred.

Our working hypothesis concerning the mechanism of the tachycardia is shown in figure 4. The NV fiber arises from the AV node and inserts into either the right ventricle or the specialized conduction system of the right ventricle. The antegrade limb of the tachycardia circuit is from the nodoventricular fiber to the right ventricle, while the retrograde limb of the tachy-
cardia circuit proceeds retrograde up the His bundle to the AV node. A limb of the reentry circuit is intranodal.

Further observations concerning this mechanism were made in patient 5, a 14-year-old boy with a corrected atrial septal defect who presented with recurrent palpitations. Tracings recorded during sinus rhythm usually showed a normal PR interval with a conduction defect of the LBBBM type. The intracavitary records during sinus rhythm showed activation of the His bundle after the onset of the QRS complex (fig. 5). Pacing of the His bundle resulted in an RBBB morphology (RBBBM). Both RBBBM and LBBBM could be produced by rapid right atrial pacing. During rapid pacing, reciprocating tachycardia with a cycle length of 250–260 msec was induced, associated with the same LBBBM present during sinus rhythm and atrial pacing. Intracavitary recordings showed ventriculo-atrial dissociation and the His deflection could not be initially identified during tachycardia. With the introduction of two successive premature ventricular complexes, sinus rhythm and a discernible His deflection resumed. During a second induction of this tachycardia, careful positioning of the His catheter revealed a sharp deflection inscribed during the ventricular

Figure 4. Schematic representation of the reentry circuit underlying a reciprocating tachycardia that uses a nodoventricular fiber. The nodoventricular fiber may insert either into the right ventricle or the right bundle branch (RBB). The retrograde return circuit can conceivably be completed by either the RBB or the left bundle branch (LBB). A portion of the reentry loop is confined to the atrioventricular (A-V) node and the atrium does not form a necessary link in the reentry loop.

Figure 5. Recordings include standard ECG leads I, II, III, V₁ and V₆ and bipolar electrograms from the right atrium (RA), right ventricle (RV), His bundle (HBE) and proximal and distal coronary sinus (PCS and DCS). (A) The result of sudden right atrial (RA) pacing at a cycle length of 300 msec in patient 5, who had a nodoventricular fiber. One-to-one ativoventricular conduction is present with a prolonged AH interval. Each ventricular response is preceded by a His deflection at an interval of 60 msec and the surface QRS manifests a right bundle branch block morphology. On discontinuation of pacing, sinus rhythm resumed with a normal PR and a surface ECG suggestive of left bundle branch block morphology; the His deflection now occurs after the onset of the surface QRS. (B) Right atrial pacing has again been instituted in the same patient at a cycle length of 300 msec. The His deflection is now displaced into the ventricular electrogram and the surface ECG shows left bundle branch block morphology. On discontinuation of pacing, the surface ECG morphology remains unchanged and the His deflection occurs after the onset of the QRS complex.
Electrogram suggestive of a His bundle electrogram (fig. 6). Shortly thereafter, rapid right atrial pacing again induced a tachycardia with a cycle length of 250–260 msec. This time, exclusive conduction with RBBBM was observed. Each ventricular complex was preceded by a His deflection at an HV interval of 60 msec, and ventriculoatrial dissociation was again present (fig. 7). We attempted unsuccessfully to interrupt this tachycardia with single ventricular premature depolarizations. Therefore, two successive ventricular premature depolarizations were introduced (fig. 8). The first of these stimuli was delivered after the onset of the His deflection; the second, before the onset of the next His depolarization, but the HH interval was exactly the same as the preceding one, suggesting that the stimulus did not reach the His bundle. Nonetheless, reciprocating tachycardia terminated.

During a restudy on digoxin, single premature ventricular depolarizations introduced when the His bundle should have been refractory consistently terminated tachycardia. This sequence of events was reproducible, suggesting that reciprocating tachycardia could be terminated by entry of the ventricular stimuli into the AV node over the NV fiber independent of conduction over the His bundle. During reciprocating tachycardia, conduction was either exclusively LBBBM or RBBBM. No fusion or morphologic
variation of the two forms was noted, as might have been expected had this tachycardia represented reentry in the AV node with conduction to the NV fiber as a "bystander phenomenon." We believe the tachycardia associated with RBBBM was caused by reversal of the tachycardia circuit. This patient always exhibited RBBB with His extrasystoles or during junctional rhythm before catheterization, suggesting an additional abnormality of conduction of the distribution of the right bundle branch. Thus, during tachycardia with RBBBM, conduction presumably proceeded antegrade over the His bundle and left bundle branch to the ventricle with retrograde conduction via the Mahaim fiber to the AV node without atrial participation. The first tachycardia in this patient clearly mimicked ventricular tachycardia, whereas the second type of tachycardia suggested a junctional tachycardia.

Atrial fibrillation was induced in every patient and was associated with predominant conduction over the NV fiber, and frequent fusion of beats was observed.

The Effect of Impulse Formation in the Region of the His Bundle

Impulse formation in the His bundle (below the takeoff of the NV fiber) achieved by spontaneous His extrasystoles or His bundle pacing was observed in five of six patients and invariably resulted in either a normal or RBBB QRS morphology.

Associated Anomalies

Three of the six patients had Ebstein's anomaly. Two of these three also had Wolff-Parkinson-White syndrome due to an accessory AV pathway, which was surgically divided before the postoperative study of the residual NV fiber. In the remaining patient (number 2), the auscultatory and echocardiographic manifestations of the Ebstein's malformation (delayed tricuspid closure) were masked by preexcitation of the right ventricle via the NV fiber. Nevertheless, two-dimensional echocardiography verified displacement of the tricuspid valve onto the lower septum and a "forme fruste" type of Ebstein's anomaly was verified by angiography. Patient 5 had undergone closure of a secundum atrial septal and had Klippel-Feil syndrome as well.

Intraoperative Findings

During surgery on the two patients with Wolff-Parkinson-White syndrome, we mapped the sequence of the ventricular activation during NV conduction. These maps, reported elsewhere, showed that the earliest ventricular activation was over the anterior right ventricle distal to the AV groove and encompassing a greater area of epicardial breakthrough than normally encountered. This finding is in contrast to what might have been expected had the findings been caused by a slowly conducting accessory pathway of the classic atrioventricular type.

Treatment

Five of the six patients with NV fibers had frequent symptomatic tachycardias necessitating treatment. Quinidine prolonged the refractory period and decreased the conduction in the NV fiber (three of three patients); digitalis had no effect on the NV fiber (one patient); propranolol caused no change in the properties of the NV fiber in one patient and in a second patient appeared to decrease conduction and prolong refractoriness in the AV node proximal to the takeoff of the NV fiber. Four of the five patients with NV fibers were finally treated successfully with quinidine (combined with propranolol in one) and one was treated with digitalis. The final patient was asymptomatic and required no treatment.

Fasciculoventricular Fibers

Recordings During Sinus Rhythm/Atrial Pacing

During sinus rhythm, the PR interval was abbreviated in five of six patients (0.07–0.11 second, mean 0.09 ± 0.01 second) because of associated enhanced conduction in the AV node. The QRS was slightly prolonged (0.10–0.12 second, mean 0.11 ± 0.01 second), with slurring of the initial forces that suggested a delta wave. The varying QRS morphologies suggested incomplete RBBB in three and incomplete LBBB in three. An example is given in figure 9, which shows a short PR and slight slurring in the initial forces most notable in V2 and V4.

Intracavitary recording in patient 11 (fig. 10) showed abbreviation of the AH interval (35 msec) and the HV interval (15 msec). Introduction of the catheter resulted in RBBB in this and all succeeding tracings from this patient. No fusion was observed in sinus rhythm or with atrial pacing from the right atrium or coronary sinus. When the paced cycle length was decreased from 400 to 200 msec, 1:1 AV conduction persisted down to a cycle length of 220 msec, the AH interval prolonged from 35 to 90 msec, the QRS remained constant in morphology (fig. 11) and the HV

FIGURE 9. Recording during sinus rhythm in patient 5, who had enhanced conduction in the atrioventricular node and a fasciculoventricular fiber. The 12-lead ECG shows a short PR interval and slurring of the initial forces of the QRS complex.
refractory period curves were smooth in all patients except for patient 11, who had an abrupt increase in the ventriculoatrial intervals at early coupling intervals; however, no reciprocating tachycardia could be induced.

**Studies During Observed Arrhythmias**

No observed arrhythmia could be causally related to the presence of FV fiber in this group. In five of the six patients, atrial fibrillation with a rapid ventricular response due to enhanced conduction of the AV node was documented. The QRS complexes were predominantly preexcited during atrial fibrillation, and fusion complexes were not observed. We believe that the presence of the FV fiber was coincidental. Enhanced conduction in the AV node, in concert with an FV fiber, mimicked Wolff-Parkinson-White syndrome in these symptomatic patients, prompting their referral. Ventricular fibrillation was documented in two of these patients (nos. 9 and 12) and was preceded by atrial fibrillation associated with a rapid ventricular response during which the QRS complexes were narrow and constant in morphology.

**The Effect of Impulse Formation in the Region of the His Bundle**

His bundle extrasystoles were observed in two of the six patients and were associated with the same abbreviated HV intervals as during the sinus rhythm. The surface delta wave was unchanged with these extrasystoles (fig. 12).

In patients with FV fibers, one has to attempt to validate the His bundle potential as being a His bundle and not a right bundle branch potential. Pacing of the His bundle was performed in an additional three
patients and was associated with stimulus-ventricular intervals identical to the HV interval during sinus rhythm. In all cases, a large atrial electrogram was evident when the His bundle was recorded. In a final patient, His bundle pacing could not be achieved nor was spontaneous impulse formation in the His bundle observed.

**Associated Anomalies**

Patient 8 had undergone surgical division of an accessory atrioventricular pathway as treatment for his Wolff-Parkinson-White syndrome. Another patient had associated coronary artery disease, which may have accounted in part for his episode of ventricular fibrillation during a rapid ventricular response associated with atrial fibrillation. One patient had associated rheumatic heart disease with sick sinus syndrome.

**Intraoperative Findings**

One patient underwent intraoperative epicardial mapping during surgical correction of an accessory atrioventricular pathway. After division of the Kent bundle, the ventricle was remapped when the PR interval was still abbreviated and associated with an initial delta wave. The epicardial map showed that the earliest ventricular activation occurred over the anterior right ventricle in an area indistinguishable from that expected from conduction over the normal AV node- His bundle system. Specifically, no area of early activation was detected in the region of the AV grove, as might be expected had the preexcitation been caused by an accessory AV pathway. A second patient underwent surgery for cryoablation of her His bundle because of recurrent episodes of rapid ventricular response due to atrial fibrillation. The details of the latter epicardial map, which has been published, showed that early activation occurred over the area trabecularis of the right ventricle (normal) with no early activation noted in the region of the AV groove. Ablation of the His bundle resulted in complete AV block.

**Treatment**

Treatment was directed to the condition of enhanced conduction in the AV node. The final treatment regimen consisted of oral digoxin (one patient), quinidine sulfate (one patient), quinidine sulfate in combination with propranolol (one patient) and cryoablation of the His bundle with insertion of a permanent pacemaker (one patient). Two patients received no treatment: One was asymptomatic and the other had infrequent episodes of tachycardia.

**Discussion**

In our series, the role of Mahaim fibers in arrhythmias appeared to differ according to the type of fiber, so NV and FV fibers will be discussed separately. The existence of NV fibers has been reported or discussed frequently. In all cases, the ECG pattern during the associated tachyarrhythmias has shown LBBB.

The presence of LBBBM and 1:1 ventriculoatrial association during tachycardia suggests a differential diagnosis of supraventricular tachycardia (atrial, intranodal, or via an accessory ventriculoatrial pathway) with LBBB aberration; ventricular tachycardia; and antidromic reciprocating tachycardia. We excluded supraventricular tachycardia with aberration in each case by showing the absence of a His deflection preceding the QRS. Ventricular tachycardia could exhibit all the features observed: absence of His deflection before the QRS, presence of a retrograde His deflection in three of six, and ventriculoatrial dissociation in three of six. However, in every case, the LBBBM during tachycardia could be reproduced by atrial pacing at various paced cycle
lengths. Finally, the long delay from the atrial electrogram to the ensuing ventricular electrogram during tachycardia would not be compatible with antidromic tachycardia using a typical Kent bundle for antegrade conduction.

The presence of an NV fiber during antegrade pacing studies requires consideration of several additional mechanisms of tachycardia: atrial rhythms with depolarization of the NV fiber as a “bystander”; use of the NV fiber as the antegrade limb of a macroreentrant circuit with retrograde conduction via the His-Purkinje system and a portion of the reentry circuit confined to the AV node; and reentry confined to the AV node with depolarization of the NV fiber as a “bystander.” If ventriculoatrial dissociation can be demonstrated during tachycardia (as in three of our six cases), an atrial mechanism can be excluded. The mere presence of ventriculoatrial dissociation does not argue for active or passive participation of the NV fiber, as a portion of the reentry loop can be confined to the AV node, making atrial participation unnecessary.

One could easily imagine a reversal of this macroreentry loop with normal QRS complexes and persistence of ventriculoatrial dissociation, as in patient 5. Ventriculoatrial dissociation accompanying apparent junctional reentry has been reported, although the possibility of a retrogradely conducting NV fiber was not uniformly excluded in the majority of these reported cases. If reversal of the reentry circuit occurred (normal QRS complexes) and 1:1 retrograde atrial conduction was present, the rhythm could resemble supraventricular tachycardia due to reentry in the AV node. Indeed, participation of the NV fiber that conducts only in the retrograde direction might be a potential mechanism of supraventricular tachycardia, analogous to the situation of the Kent bundles that conduct only in the retrograde direction.

Only a few cases of nodoventricular fibers have been reported in sufficient detail to allow examination of the underlying mechanism. The best study of a patient with NV fibers was reported by Wellens, who performed the first clinical electrophysiologic study of this entity.

In all reported cases, LBBB occurred during spontaneous tachycardia, and atrial pacing successfully reproduced this morphology. A reciprocating mechanism was shown in all of these reported cases. A 1:1 relationship between activation of the ventricles and the atria was the rule in all but one case. In general, studies during atrial pacing and premature atrial stimulation demonstrated ventricular fusion; two areas of physiologic delay were identified, one above and one below the takeoff of an accessory pathway arising from the AV node and inserting in the right ventricle. Most workers agreed that activation of the atria was not required. The association of Ebstein’s anomaly and additional accessory atrioventricular pathways has been reported.

Review of the literature uncovered six well-studied case reports. In four instances, the authors postulated a macroreentrant circuit that used an NV fiber during antegrade conduction to the ventricle with retrograde conduction over the His bundle and a portion of the reentry circuit confined to the AV node. Wellens postulated reentry confined to the AV node for the first beat of tachycardia with maintenance of the tachycardia due to the above described macroreentry circuit. Only Ward and co-workers postulated a reentry circuit completely confined to the AV node.

Several factors in our series appear to favor the participation of the NV fiber in a macroreentry circuit: No fusion was observed during reciprocating tachycardia; one would expect during prolonged observation that if the NV fiber had in fact been a “bystander,” some evidence of fusion or block in the NV fiber might have been observed. Ventriculoatrial dissociation was observed in three of the six patients studied; although the ventriculoatrial dissociation associated with junctional tachycardia has been reported, this is still an unusual finding. In our three patients in whom the His bundle could be recognized in the ventricular electrogram during tachycardia with antegrade conduction over the NV fiber, there was a striking similarity between the interval between ventricular depolarization and the His deflection during tachycardia when compared with the interval between ventricular depolarization and a retrograde His deflection recorded during ventricular pacing. Termination of reciprocating tachycardia temporally related to trauma to the NV fiber in one instance and to the right bundle branch in another instance, both required portions of the postulated macroreentry circuit. After reset of the tachycardia cycle with atrial or ventricular premature depolarization, the ensuing ventriculoatrial interval remained constant in patients with a 1:1 association between ventricular and atrial activation. It is difficult to be certain that the NV fiber participates in the reentry circuit. At present, the diagnosis of reentry confined to the AV node is largely one of exclusion. Certainly, patients with NV fibers are not immune to conventional varieties of reentry, and it is conceivable that patients such as those reported by Ward have two abnormalities by coincidence.

All reported cases of NV fibers, including ours, have exhibited LBBB associated with left-axis deviation. This finding probably relates to a somewhat consistent site of insertion of the NV fiber, probably in the intervenricular septum near the apex or diaphragmatic wall.

We have not encountered a patient with tachyarrhythmias related to an NV fiber sufficiently refractory to justify a surgical ablation of the reentry circuit. Such an intervention would certainly seem to carry a high risk of AV block, but theoretically it would seem possible with the aid of cryosurgery to cool selected areas along the right side of the intervenricular septum just beneath the tricuspid annulus until conduction over the NV fiber ceased. If an NV fiber could be localized in this fashion, it would be ablated by cryosurgery. Similarly, cooling of the proximal right bundle branch during sustained tachycardia intraoperatively could

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form a rationale for permanent ablation of the right bundle branch if tachycardia terminated. This would be a less optimal approach because it would be possible for reentry to continue using the left bundle branch.

Another interesting point is the problem of distinguishing NV fibers from accessory AV fibers of the Kent bundle variety, which exhibit decremental properties. We recently reported decremental properties during retrograde studies of accessory pathways using straight ventricular pacing and ventricular premature depolarizations. The decremental properties in ventriculoatrial accessory pathways were so striking in some of our studies as to prompt us to postulate an accessory AV node operative in the retrograde direction. In a comparable way, one might question whether the NV fibers in this report are in fact accessory AV pathways operating with decremental properties in the antegrade direction. In three of our cases, this possibility appears excluded by the observation of ventriculoatrial dissociation during tachycardia. In two of the remaining patients, very long AV intervals were observed during tachycardia, compared with very short ventriculoatrial intervals. Although this finding is inconsistent with a typical Kent bundle, it does not exclude a node-like accessory pathway. Patient 6 had characteristics that suggested an antegrade accessory AV fiber of the Kent bundle variety. Antegrade conduction and refractoriness over the accessory pathway were abbreviated. Nevertheless, during tachycardia with ventricular complexes identical to those produced by atrial pacing, complete ventriculoatrial dissociation was present. We postulate in this case that the NV fiber arose from the transitional portion of the AV node, accounting for the absence of delay proximal to the takeoff of the NV fiber.

Only a limited number of reports have described the existence of FV fibers. A number of authors have described the combination of functional or anatomic “bypasses” of the AV node in combination with FV fibers, mimicking classic Wolff-Parkinson-White syndrome. In our series, all symptomatic arrhythmias in patients with FV fibers could be ascribed to coincidental abnormalities of conduction at the level of the AV node. Although we could not prove that the FV fiber itself participated in any of the rhythm disorders in our six patients, Bharati et al. observed a patient who had ventricular tachycardia associated with LBBBM. Pathologic examination of the heart revealed an abnormally septated His bundle and it was postulated that this abnormal anatomic disposition of the conducting tissues had resulted in the tachyarrhythmia.

In conclusion, there appear to be functional counterparts to the proposed anatomic subdivision of the Mahaim fibers. In sinus rhythm, NV fibers can mimic the presence of LBBBM, and appear capable of supporting a sustained reciprocating tachycardia. FV fibers can mimic intraventricular conduction defects, but we found no direct relationship between FV fibers and arrhythmias. Finally, the association of Mahaim fibers with Ebstein’s anomaly and with an additional accessory pathway of the Kent bundle variety was confirmed.

Addendum

Since submission of this manuscript, we have studied another patient with two NV Mahaim fibers. This was a 34-year-old man with a 6-year history of recurrent reciprocal tachycardia exhibiting LBBBM with an axis of −30° and 1:1 ventriculoatrial conduction. The ECG in sinus rhythm was invariably normal, but right atrial and coronary sinus pacing duplicated the LBBBM. A new finding was observed during reciprocal tachycardia: Two slightly different forms of LBBBM were seen during reciprocal tachycardia associated with two distinctive V-His intervals (15 and 100 msec). No intermediate QRS morphologies or V-H intervals were observed, and transition from one to the other reciprocal tachycardial reproducibly occurred with single ventricular premature depolarizations. We interpreted this as evidence of two NV fibers. The patient was successfully treated with iv. and then oral verapamil.

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