Transcatheter Ablative Techniques for Treatment of the Permanent Form of Junctional Reciprocating Tachycardia in Young Patients

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Five patients with medically refractory incessant supraventricular tachycardia due to a posterior septal, slowly conducting accessory connection underwent transcatheter closed chest ablative treatment. The tachycardia characteristics were consistent with the permanent form of junctional reciprocating tachycardia. In each patient the ablative attempts resulted in independent interruption of either the anterograde limb (atrioventricular node-His bundle conduction) or the retrograde limb (accessory connection) of the tachycardia circuit. Permanent retrograde pathway ablation was achieved in only one patient and followed separate permanent transcatheter His bundle ablation. In three of the other four patients the ablation attempt caused temporary interruption of retrograde conduction. Each patient had improved control of tachycardia related to the ablation attempt. Of the five patients, four required pacemaker implantation.

With further refinements, selective ablation of the retrograde limb of the tachycardia circuit may be possible. This experience confirms the anatomic independence of the anterograde and retrograde limbs of the tachycardia circuit.

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The permanent form of junctional reciprocating tachycardia is the most common form of incessant supraventricular tachycardia in children (1-6). This type of tachycardia has been variously called the atypical form of atrioventricular (AV) node reentrant tachycardia, the "fast-slow" form of AV node reentrant tachycardia, the childhood form of AV node reentrant tachycardia and Coumel's tachycardia (1-8). This variety in nomenclature reflects past and present uncertainty over the mechanisms involved. The current consensus is that the mechanism involves an AV reentrant circuit, in which the retrograde limb is an extra AV node structure with decremental conduction properties (1,3,4,6,9-11). The exact anatomic characteristics and the origin of the bypass pathway are still unclear.

This report reviews consecutive experience with five patients treated for incessant supraventricular tachycardia using transcatheter ablative techniques. This experience affirms the role of an anatomically separate retrograde limb in pathogenesis and suggests a significant role for transcatheter ablative techniques after failure of medical control of tachycardia.

Methods

Electrocardiographic characteristics. The electrocardiographic characteristics of the permanent form of junctional reciprocating tachycardia were consistent in all five patients. A typical 15 lead electrocardiogram is presented in Figure 1. In many patients with the permanent form of junctional reciprocating tachycardia, there are occasional interruptions in tachycardia due to conduction block in either the anterograde or retrograde limb of the tachycardia circuit. There is then a pause ending with sinus node recovery. Often tachycardia is reinitiated by this impulse. No prolongation of the PR interval occurs before reinitiation. The P' wave (resulting from retrograde atrial activation) is uniformly negative in leads II, III and aVF. This P' frontal plane axis is consistent with activation beginning near the mouth of the coronary sinus or the AV node. The ratio between anterograde AV node conduction time and retrograde accessory pathway conduction time is reflected in the RP' and the P'R intervals. In the permanent form of junctional reciprocating tachycardia, this ratio is greater than 1 (that is, the RP' is
Figure 1. Patient 5. Fifteen lead electrocardiogram recorded from a 10 year old patient with the permanent form of junctional reciprocating tachycardia. The typical findings of this form of tachycardia demonstrated here include a relatively slow tachycardia of 125 beats/min and a frontal P wave axis of $-90^\circ$. Failure of conduction through the retrograde limb of the tachycardia circuit is followed by a sinus impulse that reinitiates tachycardia.

These ratios differ from the more typical forms of paroxysmal atrial tachycardia in which the $RP'/PR$ ratio is usually less than 1. The 24 hour electrocardiogram clearly reveals the incessant nature (in tachycardia, the majority of the time) of the tachycardia.

Electrophysiologic characteristics. Certain electrophysiologic characteristics, when present, support the concept of AV reentry through an anatomically distinct AV pathway and also support the concept that this pathway is the retrograde limb of the tachycardia loop. These characteristics include: 1) a point of earliest retrograde atrial activation during supraventricular tachycardia other than the low septal right atrium (the AV node), and 2) retrograde atrial pre-excitation demonstrated by delivering premature ventricular stimuli into diastole when the His bundle is refractory. The permanent form of junctional reciprocating tachycardia is more specifically confirmed when slow, decremental ventriculoatrial conduction is demonstrated and the earliest point of retrograde endocardial atrial activation is close to or within the mouth of the coronary sinus. When associated with slow decremental retrograde pathway conduction properties, this activation sequence is pathognomonic for the permanent form of junctional reciprocating tachycardia (Fig. 2).

Patient characteristics. Each of the five patients underwent an initial evaluation that included an electrocardiogram at rest, chest roentgenogram, 24 hour ambulatory electrocardiogram and echocardiogram. The clinical data are summarized in Table 1; the patients are listed in chronologic order of presentation and treatment. The tachycardia rate was recorded at the time of presentation. Symptoms included chest pain, palpitation and manifestations of congestive heart failure. Myocardial dysfunction documented by echocardiography included ventricular dilation and decreased shortening fraction. Of the five patients, three (Patients 2 to 4) were referred for evaluation of incessant medically refractory tachycardia. Patient 1 was referred specifically for transcatheter closed chest ablation of the His bundle because of incessant tachycardia. Patient 5 was referred with the diagnosis of myocarditis and sinus tachycardia.

Protocol. The noninvasive evaluation was followed by an electrophysiologic study performed using previously published methods (12). In addition to the standard high right atrial quadripolar catheter, a tripolar His bundle catheter and a quadripolar right ventricular catheter, either a quadripolar, hexapolar or decapolar catheter was positioned from the left arm (basilic vein) and advanced into the coronary sinus for determination of the retrograde atrial activation occurring in the mouth of the coronary sinus or posterior left atrium.
The patients were sedated with meperidine and promethazine. Ketamine was given either when additional medication was required or immediately before the ablation attempt. The patients were not intubated or paralyzed. Arterial pressure and periodic arterial blood gases were monitored throughout the procedure.

The first patient underwent transcatheter His bundle ablation using methods similar to those previously described by Gallagher et al. (13) and Scheinman et al. (14). For this purpose a tripolar electrode catheter (USCI) was positioned across the tricuspid valve in the region of the His bundle. The largest unipolar His bundle electrogram was located and unipolar direct current shocks were delivered between the electrode at the catheter tip (the cathode) and a cutaneous 16 cm² electrode (R2 Corp.) (the anode) that was located posteriorly slightly to the left of the midthoracic spine. The energy was delivered from a standard direct current cardioversion unit connected to the electrode catheter.

In subsequent patients, a similar method was used to accomplish ablation only of the retrograde limb of the tachycardia circuit. Unipolar shocks were delivered to the coronary sinus catheter electrode, which identified the earliest obtainable point of retrograde atrial activation. Either a quadrripolar or a hexapolar USCI 6F standard electrode catheter was used. The most proximal and the most distal electrodes were not used to deliver shocks.

Each patient had a temporary ventricular pacing catheter in place at the time of the ablative attempt. Before each ablative attempt surgical personnel were aware that the procedure was being performed and surgical facilities were prepared to accept the patient immediately, if required. The patients were observed in the pediatric intensive care unit for 24 hours after the procedure. The procedure was explained in detail to the patients and their families and informed consent was obtained.

Results

Electrophysiologic study. The presence of a slowly conducting retrograde extra-AV node pathway with decremental properties was confirmed by electrophysiologic study in each patient. Anterograde pre-excitation could not be documented in any patient. Shortening of the atrial cycle length with premature ventricular stimuli during His bundle refractoriness was demonstrated in the four patients in whom it was sought. In all five patients, tachycardia could be interrupted by either premature atrial or ventricular stimuli, and in all the earliest retrograde atrial activation during tachycardia was very close to or within the mouth of the coronary sinus.

Figure 2. Patient 1. Intracardiac electrographic recording of the interruption and reinitiation of tachycardia in a patient with the permanent form of junctional reciprocating tachycardia. The surface electrocardiographic leads from top to bottom are I, aVF, V₁ and V₅. The intracardiac electrograms from top to bottom are: high right atrium (HRA), five coronary sinus electrograms recorded from a decapolar catheter straddling the coronary sinus ost (CSd = distal coronary sinus; CSP = proximal coronary sinus), two electrograms from a tripolar catheter in the His bundle position (LSRA = low septal right atrium; H = His potential) and a single electrogram recorded from the right ventricular apex (RVA). Intraventricular blood pressure appears at the bottom of the recording. The first three impulses end an episode of permanent junctional reciprocating tachycardia. The earliest retrograde atrial activity (ERAA) (labeled with an arrow) is recorded by the electrode pair at the coronary sinus mouth (CSos) and is marked by line a. Permanent junctional reciprocating tachycardia is then interrupted because of failure of retrograde pathway conduction. After a pause, recovery occurs with the earliest atrial activation recorded at the high right atrium. The atrial activation sequence of sinus rhythm is highlighted by line b. A second sinus impulse is followed by reinitiation of the tachycardia. HISd = distal His bundle; HISp = proximal His bundle.
Table 1. Clinical Features of Five Patients With the Permanent Form of Junctional Reciprocating Tachycardia

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yr) at Ablation &amp; Sex</th>
<th>Age (yr) at Onset</th>
<th>Tachycardia Rate (beats/min)</th>
<th>Symptoms</th>
<th>Myocardial Dysfunction</th>
<th>Drug Treatment</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>27F</td>
<td>7</td>
<td>133</td>
<td>Yes</td>
<td>No</td>
<td>Dig, Ver, Quin, Proc, Prop</td>
</tr>
<tr>
<td>2</td>
<td>8M</td>
<td>Infancy</td>
<td>101</td>
<td>Yes</td>
<td>No</td>
<td>Dig, Ver, Quin</td>
</tr>
<tr>
<td>3</td>
<td>2F</td>
<td>Infancy</td>
<td>153</td>
<td>Yes</td>
<td>No</td>
<td>Dig, Ver, Quin, Proc</td>
</tr>
<tr>
<td>4</td>
<td>3M</td>
<td>Infancy</td>
<td>180</td>
<td>Yes</td>
<td>Yes</td>
<td>Dig, Ver, Quin, Prop</td>
</tr>
<tr>
<td>5</td>
<td>10M</td>
<td>5</td>
<td>130</td>
<td>No</td>
<td>Yes</td>
<td>Dig, Ver, Quin</td>
</tr>
</tbody>
</table>

Dig = digoxin; F = female; M = male; Proc = procainamide; Prop = propranolol; Quin = quinidine; Ver = verapamil.

Transcatheter electrical ablation. Table 2 summarizes the ablation attempts in each patient. In each patient one or more attempts were made to ablate the anterograde or retrograde limb of the tachycardia circuit. In Patient 1 no attempt was made to ablate the retrograde limb. In Patient 4 the catheter used for ablation was positioned in the coronary sinus from the right femoral vein. This resulted in a different orientation of the catheter to both the point of earliest retrograde atrial activation and the AV node. The ablation attempt in this patient resulted in complete anterograde AV block without retrograde block.

Tachycardia reinitiation was attempted while each patient with temporary retrograde block remained in the catheterization laboratory. Of the three patients tested, none had inducible tachycardia at the conclusion of the procedure. Transcatheter electrical ablation of anterograde AV node conduction was accomplished in Patient 5 after two failed attempts at permanent ablation of the retrograde pathway. Persistent retrograde conduction resulted in chronic pacemaker-mediated tachycardia that was resistant to control by acceptable reprogramming. A third attempt at retrograde pathway ablation was successful, permanently eliminating retrograde conduction.

Follow-up. The current status of the patients is summarized in Table 3. Improved control of tachycardia was achieved in all. Tachycardia was eliminated in Patients 1 and 5, but both patients required pacemaker implantation. Patient 4 has a pacemaker for catheter-induced AV block, but has intact retrograde conduction that requires unusual and limiting pacemaker settings for control of pacemaker-mediated tachycardia. To achieve this control, the programmed AV interval was shortened to 50 ms and the atrial refractory period was prolonged to 375 ms, effectively limiting the upper tracking limit to 156 beats/min. Patient 3 has improved tachycardia control while taking medications similar to those she received before the ablative attempt. Presumably this effect is due to electrical modification of the retrograde limb of the tachycardia circuit. In this patient, the 24 hour ambulatory electrocardiogram recorded before the ablative attempt demonstrated 95% tachycardia and 5% sinus rhythm. After the attempted transcatheter electrical ablation, while she was receiving similar medications (digoxin and verapamil), the 24 hour electrocardiogram demonstrated 95% sinus rhythm and 5% tachycardia.

Discussion

The permanent form of junctional reciprocating tachycardia is increasingly recognized as an important problem in young patients. Although it is rare in adults, it is a relatively common form of tachycardia in children and adolescents (1–6). Until its recognition as an entity, it was grouped with atrial ectopic tachycardia or AV node reentry occurring entirely within the AV node. The observation that it is relatively common in young patients suggests that the tachycardia resolves at some point between childhood and adulthood or remains unrecognized or that patients with this entity do not survive. Spontaneous resolution of the per-

Table 2. Electrical Ablation Results in Five Patients

<table>
<thead>
<tr>
<th>Case</th>
<th>Ablation Site</th>
<th>Energy (W-s)</th>
<th>Anterograde Conduction</th>
<th>Retrograde Conduction</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>His bundle</td>
<td>25, 50, 50</td>
<td>Blocked</td>
<td>Persisted</td>
</tr>
<tr>
<td>2</td>
<td>MOCS</td>
<td>25, 25, 50</td>
<td>Persisted</td>
<td>Blocked (temp.)</td>
</tr>
<tr>
<td>3</td>
<td>MOCS</td>
<td>30, 50</td>
<td>Persisted</td>
<td>Blocked (temp.)</td>
</tr>
<tr>
<td>4</td>
<td>MOCS</td>
<td>100</td>
<td>Blocked</td>
<td>Persisted</td>
</tr>
<tr>
<td>5</td>
<td>MOCS</td>
<td>150</td>
<td>Persisted</td>
<td>Blocked (temp.)</td>
</tr>
<tr>
<td></td>
<td>MOCS</td>
<td>200</td>
<td>Persisted</td>
<td>Blocked (temp.)</td>
</tr>
<tr>
<td></td>
<td>His bundle</td>
<td>150, 150, 200, 200</td>
<td>Blocked (perm.)</td>
<td>Persisted</td>
</tr>
<tr>
<td></td>
<td>MOCS</td>
<td>300, 300</td>
<td>Blocked (perm.)</td>
<td></td>
</tr>
</tbody>
</table>

MOCS = mouth of the coronary sinus; perm. = permanent; temp. = temporary.
permanent form of junctional reciprocating tachycardia has not been reported.

Cardiac dysfunction. Chronic tachycardia is associated with cardiac dilation and reduced ventricular function (15,16). After surgical ablation of the tachycardia focus in a patient with incessant tachycardia, ventricular function becomes normal (15). Therefore, aggressive intervention to control tachycardia is justified when necessary to prevent development of irreversible cardiac dysfunction and to control symptoms.

Surgical therapy. Since Cobb et al. (17) first reported successful surgical division of AV accessory connections, several centers have reported successful surgical division in large numbers of patients. Single accessory connections located laterally to the left or to the right of the posterior septum have an excellent prognosis for successful surgical division without the complication of AV block. Division of posterior septal pathways in patients with typical paroxysmal supraventricular tachycardia has proven more precarious. This procedure has occasionally resulted in failure or inadvertent damage to the AV node.

 Until recently, the retrograde pathway in the permanent form of junctional reciprocating tachycardia was thought to be contiguous with the AV node; surgical retrograde pathway ablation without resultant anterograde block was considered impossible. However, Guarnieri et al. (3) reported that in seven of nine patients with the permanent form of junctional reciprocating type tachycardia in whom surgical division of the retrograde pathway was attempted, the procedure was successful and anterograde AV node block was not created. These results not only confirmed the physically independent nature of the pathway, but also demonstrated the possibility of surgical cure. In children, the small size of the critical area is an especially important consideration. The distance between the mouth of the coronary sinus and the AV node conduction tissue is very short and dissection of this area in a small child is technically very difficult. Although two small patients (aged 5 and 6 years, respectively) were included in the study of Guarnieri et al. (11), the surgical approach to the retrograde pathway should probably remain a last resort in small children.

Catheter ablation. Recently, experience with transcatheter treatment of arrhythmias has been accumulated and His bundle ablation for chronic medically uncontrolled supraventricular tachycardia has become an accepted mode of therapy (13,14). Successful transcatheter ablation of the posteroseptal accessory connections of the typical type has been reported (18,19). Although permanent ablation of the retrograde pathway alone in our patients was not achieved, the temporary interruption suggests that it may be possible with further experience. Patient 5 did have permanent ablation of the retrograde pathway, although his AV node conduction had already been interrupted. This patient received the highest energy ablation of any of the patients (300 W-s) without untoward effects. Whether permanent anterograde block would have occurred in addition to permanent retrograde block in this patient is unknown. However, permanent interruption of posterior septal retrograde accessory pathway conduction without anterograde AV node block was achieved by Morady et al. (19) with shocks of similar energy in adults with paroxysmal tachycardia. Further experience is necessary to determine the optimal energy of the shock and the specific delivery method; for example, are unipolar or bipolar shocks more effective and which electrode or electrodes should be used for delivery.

Pacemaker-mediated tachycardia. The possibility of interrupting only the retrograde pathway and leaving the normal AV node conduction intact is especially alluring because the patient would have neither a pacemaker requirement nor tachycardia. Of the four patients in this study who required a pacemaker, three received an AV universal pulse generator and one received a ventricular demand pulse generator. In each patient with an AV universal pulse generator, pacemaker-mediated tachycardia occurred, requiring the full potential of the device’s programmable features for control. Because incessant pacemaker-mediated tachycardia, Patient 5 was returned to the catheterization laboratory for the final attempt at ablation of the retrograde pathway. This attempt was successful and he has had no return of pacemaker-mediated tachycardia with nominal pulse generator settings. These settings now allow AV synchrony and rate responsiveness. The electrophysiologic properties

Table 3. Follow-Up of Five Patients

<table>
<thead>
<tr>
<th>Case</th>
<th>Follow-up Duration (mo)</th>
<th>Tachycardia</th>
<th>Medications</th>
<th>Pacemaker</th>
<th>PMT</th>
<th>Ventricular Dysfunction</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>37</td>
<td>None</td>
<td>None</td>
<td>Yes (VVI)</td>
<td>Rare</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>31</td>
<td>Less</td>
<td>Ver</td>
<td>Yes (DDD)</td>
<td>Rare</td>
<td>Resolved (echo)</td>
</tr>
<tr>
<td>3</td>
<td>26</td>
<td>Less</td>
<td>Dig, Ver</td>
<td>No</td>
<td>Resolved with reprogramming</td>
<td>Resolved (echo)</td>
</tr>
<tr>
<td>4</td>
<td>23</td>
<td>None</td>
<td>None</td>
<td>Yes (DDD)</td>
<td>Resolved</td>
<td>Improved (echo)</td>
</tr>
<tr>
<td>5</td>
<td>19</td>
<td>None</td>
<td>Dig</td>
<td>Yes (DDD)</td>
<td>Resolved</td>
<td>Improved (echo)</td>
</tr>
</tbody>
</table>

*PMT cannot occur with a VVI pacemaker. DDD = fully automatic pacemaker mode; echo = echocardiographically determined; PMT = pacemaker-mediated tachycardia; VVI = permanent ventricular demand pacemaker; other abbreviations as in Table 1.
of the retrograde pathway favor pacemaker-mediated tachycardia. In patients needing AV synchrony and rate responsiveness, careful consideration should be given to ablation of both anterograde and retrograde pathways rather than AV node-His bundle ablation alone.

Conclusions. Transcatheter ablative techniques were used without complication for attempted interruption of the retrograde accessory pathway of the tachycardia circuit in four young patients with the permanent form of junctional reciprocating tachycardia. Three of these patients received ablative stimuli through an electrode catheter located in the coronary sinus from the left arm, and had a 24 to 36 hour interruption of tachycardia. Moreover, immediately after the procedure, tachycardia was not inducible. Each patient now has improved control of tachycardia, although each continues to require a pacemaker or antiarrhythmic medication, or both.

This study has shown that: 1) the pathway of retrograde conduction is anatomically distinct from the AV node His-Purkinje system; 2) temporary and permanent interruption of the retrograde limb by transcatheter electrical techniques is possible; 3) permanent interruption of the retrograde pathway without affecting the anterograde pathway may be possible in children with further refinements of technique; 4) interruption of anterograde conduction without interruption of retrograde conduction may result in intractable pacemaker-mediated tachycardia when an AV universal pacemaker is implanted; 5) the exact positioning of the electrode catheter is important and care should be taken to position the catheter used for delivering shocks at the mouth of the coronary sinus, positioning it from the arm rather than from the femoral vein; and 6) modification of one of the limbs of the tachycardia circuit may occur and improve the response of tachycardia to medical management.

References