Establishing the diagnosis of an atypical atrioventricular (AV) node–dependent long RP supraventricular tachycardia (SVT) can be difficult. Standard diagnostic criteria are lacking and often extrapolated from pacing maneuvers applied to the more common short RP SVT. Long RP SVTs involving concealed nodofascicular (NF) accessory pathways (AP) are particularly rare with descriptions limited to isolated case reports. Prolonged conduction over the slow pathway (SP) of the AV node or a decremental AP after entrainment from the ventricle can produce A-A-V patterns that might be mistaken for atrial tachycardia (AT). In addition, slow AP conduction after entrainment of an atypical orthodromic reciprocating tachycardia (ORT) can generate long postspacing intervals (PPI) that cause misdiagnosis of AV nodal reentrant tachycardia (AVNRT), despite correction for delay in the AV node (cPPI). This study sought to evaluate the electrophysiological features and criteria differentiating the 4 atypical AV node–dependent long RP SVTs: atypical AVNRT, atypical AVNRT with a concealed, slowly conducting, decremental AV AP (also called the permanent form of junctional reciprocating tachycardia [PJRT]), and ORT using a concealed NF AP (also called NF reentrant tachycardia [NFRT]).
Methods

Nineteen patients with 20 symptomatic atypical long RP SVTs underwent diagnostic electrophysiological study. After informed consent, femoral venous access was achieved percutaneously, and multipolar catheters were positioned in the right atrium, His bundle region, right ventricle, and coronary sinus. Programmed stimulation and burst pacing were delivered from the ventricle and atrium to evaluate retrograde and antegrade conduction, respectively, and induce SVT. If SVT was noninducible in the baseline state, isoproterenol was infused and the stimulation protocol was repeated. After tachycardia induction, scanning ventricular premature depolarizations (VPDs) were delivered during the diastolic period of tachycardia, and SVT was entrained from the ventricle at a pacing cycle length of 10 to 50 ms shorter than tachycardia cycle length (TCL). Para-Hisian pacing and entrainment were performed selectively to confirm diagnoses.3,9 Radiofrequency ablation was performed in all patients by targeting either the SP or the ventricle; (2) failure of BBB to affect tachycardia; (3) His-refractory VPDs reset to the atrium or terminate tachycardia with VA block; (4) ability to entrain tachycardia from the ventricle with orthodromic capture of the His bundle; (5) PPI–TCL ≥ 115 ms (or cPPI ≥ 110 ms); (6) ∆VA ≥ 85 ms; (7) ∆HA > 0 ms (SN, 67%; specificity, 100%); and (8) ∆AH > 20 ms (SN, 59%; specificity, 100%).

Diagnostic Criteria for SVT

All tachycardias had a long RP (R>P) interval with earliest atrial activation near the ostium of the coronary sinus. Atrial tachycardia was excluded by the following criteria: (1) spontaneous termination with AV block; (2) termination of tachycardia by VPDs that failed to reach the atrium (VA block), and (3) A-A-V response to entrainment from the ventricle.10 The following criteria established a diagnosis of A-A-V arrhythmias (A VNRT): (1) nonobligatory 1:1 AV relationship (persistence of tachycardia, despite retrograde block to the atrium or antegrade block to the ventricle); (2) failure of bundle-branch block (BBB) to affect tachycardia; (3) failure of BBB to affect tachycardia; (4) inability to entrain tachycardia from the ventricle with orthodromic capture of the His bundle; (5) PPI–TCL ≥ 115 ms (or cPPI ≥ 110 ms); (6) ∆VA ≥ 85 ms; (7) ∆HA > 0 ms, and (8) ∆AH > 40 ms (or paradoxically, AHNVSR < AHNVSVT). His-Refractory VPDs (AHRV < AHNVSR).

Atypical AVNRT/NF AP

(1) nonobligatory 1:1 AV relationship (persistence of tachycardia, despite retrograde block to the atrium or antegrade block to the ventricle); (2) failure of BBB to affect tachycardia; (3) His-refractory VPDs reset the atrium or terminate tachycardia with VA block; and (4) ability to entrain tachycardia from the ventricle with orthodromic capture of the His bundle; (5) PPI–TCL ≥ 115 ms (or cPPI ≥ 110 ms); (6) ∆VA ≥ 85 ms; (7) ∆HA > 0 ms, and (8) ∆AH > 40 ms (or paradoxically, AHNVSVT < AHNVSR).
an AH interval paradoxically shorter than that during sinus rhythm. Para-Hisian pacing was unhelpful in 14 of 19 (74%) patients because either fast pathway (FP) conduction always preempted slower SP/AP conduction during pacing (n=12) or consistent 1:1 conduction over the SP/AP could not be achieved, despite pacing at the slowest rate allowable by sinus rhythm (n=2). Para-Hisian entrainment was successfully performed in only 2 patients and confirmed the established diagnosis.

### Nodal Pathways

The proximal insertion of all 4 nodal APs was the SP of the AV node. In 1 patient, it was the left atrionodal extension of the SP requiring ablation along the posteroseptal mitral annulus. The distal insertion of the nodal APs was fascicular (para-Hisian pacing [n=1]; para-Hisian entrainment [n=1]; Figure 4), ventricular (manifest QRS fusion during entrainment [n=1]; Figure 2), and indeterminate (n=1).

### Ablation

The successful ablation site for all patients with atypical AVNRT with and without a bystander NF AP was the SP of the AV node along the posteroseptum of the right atrium and includes the patient with both atypical AVNRT/NF AP and NFRT. The other patient with NFRT had an AP inserting into the left atrionodal extension of the SP and required ablation along the posteroseptal mitral annulus. All patients with PJRT had successful AP ablation along the posteroseptum of the tricuspid annulus identified by activation mapping during tachycardia.

### Discussion

Compared with atypical AVNRT, patients with ORT (NFRT/PJRT) were younger and predominantly women, although the demographics may be skewed by the small study population. These long RP tachycardias respond differently than their short RP counterparts to pacing maneuvers; and the rare tachycardia associated with a NF AP can be misdiagnosed as PJRT if the upper circuit is not analyzed. Therefore, separate pacing maneuvers in the atrium and ventricle are required to delineate the upper and lower circuit, particularly when a 1:1 AV relationship exists (Figure 6; Table 2).
Entrainment From Ventricle

Although A-A-V responses are generally considered diagnostic of AT, A-A-V patterns were common in our series, particularly for atypical AVNRT with its longer paced VA interval. Pseudo A-A-V patterns occur when decremental conduction over the SP or AP produced long VA intervals that exceed the pacing cycle length so that the first atrial electrogram after entrainment is actually driven by the penultimate pacing stimulus. True A-A-V responses were the result of dual retrograde responses (double fire) with simultaneous conduction over the FP and NF AP or SP occurring only with atypical AVNRT with and without a concealed, bystander NF AP, respectively. This is different from the A-A-V response of AT, which results from retrograde conduction over the AV node followed by the first return beat of AT after pacing. A mechanism to explain dual retrograde (A-A-V) responses during atypical AVNRT is the presence of a large excitable gap with collision between antegrade and retrograde wavefronts in the SP (retrograde limb) of the circuit. The last (n) paced antegrade wavefront conducts completely over the FP to the atrium (first A) and then collides with the previous (n–1) orthodromic wavefront in the SP. The last (n) paced orthodromic wavefront has no antegrade wavefront with which to collide, conducts over the SP to activate the atrium (second A) before conducting antegradely over the FP to the ventricle. With the more common single retrograde (A-V-A) responses, the collision point between antegrade and orthodromic wavefronts is in the FP (antegrade limb). An alternative mechanism is tachycardia termination and subsequent reinitiation. When pacing stops, retrograde conduction occurs over both the FP and SP, the latter reinitiating tachycardia.

Conventional SVT criteria during entrainment from the ventricle establish the lower portion of the tachycardia circuit as macroreentrant involving the His-Purkinje system/ventricle (PJRT/NVRT) or not (AVNRT). A PPI–TCL<115 was specific for ORT but conduction delay over the AP caused PPI–TCL>115 ms in half ORT yielding misdiagnosis of AVNRT as has been observed in other series. A higher cutoff value of 125 ms increased the SN for ORT by 33%, while maintaining 100% SP. The cPPI correctly adjusted the
long PPI during ORT when both retrograde AP and antegrade AV node decrement occurred—the latter from antidromic capture of the His bundle and retrograde concealment into the AV node. However, when substantial delay occurred over the AP, the cPPI could not correct the long PPI and even paradoxically prolonged it because the first return AH became shorter than during SVT. Slow, decremental AP conduction also affected the SN of the $\Delta V_A$ and $\Delta H_A$ criteria for ORT but maintained their high SP. Therefore, any standard criteria positive for ORT ($PPI-TCL<115$ ms, $cPPI<110$ ms, $\Delta V_A<85$ ms, and $\Delta H_A<0$ ms) was diagnostic of ORT, despite discordance among each other which occurred 50% of the time.

**His-Refractory VPDs**

His-refractory VPDs that reset (advance or delay) or terminate tachycardia indicate the presence of an AP but not necessarily its participation in tachycardia. They can reset or terminate atypical AVNRT in the presence of a concealed, bystander NF AP inserting into the retrograde SP. In such a case, the VPD conducts over the NF AP ahead of the AVNRT wavefront and penetrates its excitable gap in the SP after the lower turnaround point of the circuit. Its antidromic wavefront collides with tachycardia, whereas its orthodromic wavefront encounters either relative or absolute distal SP refractoriness delaying or terminating tachycardia, respectively. His-refractory VPDs identified an AP in all patients with ORT and was the only pacing maneuver to diagnose a concealed, bystander NF AP in 3 patients with atypical AVNRT by delaying the atrium and terminating tachycardia with VA block. Although entrainment of atypical AVNRT/NF AP from the ventricle with orthodromic capture of the His bundle is theoretically possible, it was not observed. His-refractory VPDs also determined the degree of decremental conduction over each AP. Severe AP decrement paradoxically delayed the atrium because the degree of VPD prematurity was offset by a greater than or equal degree of AP conduction delay (fully compensatory). Mild AP decrement advanced the atrium because the degree of VPD prematurity was offset by a lesser degree of AP conduction delay (partially compensatory). Paradoxically delay might identify patients who have long PPIs after entrainment independent of tachycardia mechanism.

**Figure 3.** Atypical atrioventricular nodal reentrant tachycardia with a concealed, bystander nodofascicular accessory pathway (AP). **Top**, A His-refractory ventricular premature depolarization paradoxically delays tachycardia by 41 ms (fully compensatory). The $AH_{\text{desp}}$ is very short (38 ms; $AH_{\text{NSR}}=54$ ms). **Bottom**, Entrainment from the ventricle with antidromic capture of the His bundle showing a true A–A–V response (retrograde FP and slow pathway/nodofascicular AP) and long postspacing interval–tachycardia cycle length=135 ms, $\Delta V_A=135$ ms, and $\Delta H_A=39$ ms. AH indicates atrio-His; CS, coronary sinus; FP, fast pathway; HA, His-atrial; HRA, high right atrium; RV, right ventricle; and VA ventriculoatrial.
Other Criteria

The $\Delta$AH criteria differentiates tachycardia circuits whose upper portion is partially extranodal (PJRT) or completely intranodal (NFRT/atypical AVNRT). During PJRT, the AH interval is a true interval reflecting sequential activation of the atrium and His bundle over the AV node and similar to the AH interval when pacing at the TCL. In contrast, during NFRT and atypical AVNRT, the AH interval is a pseudointerval reflecting simultaneous activation of the atrium and His bundle and is, therefore, shorter than the AH interval when pacing at the TCL. The $\Delta$AH was longer for the nodal tachycardias (NFRT/atypical AVNRT) compared with PJRT, and the AH interval was paradoxically shorter for atypical AVNRT/NF AP ($n=2$) and NFRT ($n=1$) than during sinus rhythm. A major limitation of $\Delta$AH criteria, however, is the SN of the AV node to rapid fluctuations in autonomic tone so that comparison of AH intervals between tachycardia and pacing should be done close in time allowing for minimal change in the autonomic state of the patient. For atypical AV node–dependent long RP tachycardias, para-Hisian pacing was generally not useful because (1) retrograde FP conduction consistently preempted SP/AP conduction, (2) SP/AP often exhibited retrograde Wenckebach conduction, despite ventricular pacing at the slowest cycle length allowable by the sinus rate, and (3) an AV nodal response is not diagnostic of pure AV nodal conduction but can also be observed with a NF AP.

Nodal Pathways

The proximal insertion of all 4 nodal APs was determined to be the SP of the AV node by the ability of His-refractory VPDs to perturb the retrograde limb of the circuit during atypical AVNRT and NFRT. A SP insertion can also be identified by the ability of His-refractory VPDs to reset or terminate typical AVNRT in the antegrade limb. Various maneuvers can determine the distal insertion site of concealed nodal APs. An AP response to para-Hisian pacing/entrainment identifies a nodoventricular AP because retrograde conduction is dependent on myocardial capture. An AV nodal response...
indicates a NF AP because retrograde conduction is dependent on His-RB capture. Limited data suggest that manifest fusion during right ventricular entrainment of ORT using a nodal AP is specific to a nodoventricular fiber. Because the circuit for NFRT is contained within the specialized conduction system, ventricular fusion cannot occur with paced complexes that penetrate the excitable gap and entrain tachycardia (analogous to A VNRT). Although this is true when collision between antidromic and orthodromic wavefronts occurs in the AV node or His bundle, it is not when the collision point is in the right bundle distal to the bifurcation of the His bundle and proximal to the take-off of the NF AP. In this case, the His bundle–left bundle–ventricular axis is orthodromically activated and can fuse with paced complexes from the right ventricle.

**Limitations**

The number of patients in our collection is relatively small, and our data should be evaluated in more patients. Furthermore, 1 patient contributed 2 SVTs, which violates the requirement for independent observations. However, it is to our knowledge the only series comparing both bystander NF tachycardias and NFRT providing useful information about these rarely described tachycardias. Accurate diagnosis requires evaluating all available clues from the electrophysiologic study (eg, effect of BBB) as differentiating NFRT from atypical A VNRT/NF AP using entrainment alone can be difficult and potentially misleading in certain situations. If the refractory period of a bystander NF AP is sufficiently short to support 1:1 conduction during entrainment and conduction over the NF AP is faster than over the His-Purkinje system, the pathway for entrainment of atypical AVNRT/NF AP is the same and the PPI can be short. Conversely, severe decremental conduction over a NF AP might generate long PPIs during entrainment of

### Table 1. Electrophysiological Criteria Differentiating the 4 Atypical AV Node–Dependent Long RP SVTs

<table>
<thead>
<tr>
<th></th>
<th>Atypical AVNRT (w/wo NF AP; n=14)</th>
<th>PJRT/NFRT (n=6)</th>
<th>P Value/95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, y</td>
<td>54±19</td>
<td>42±13</td>
<td>P=0.036</td>
</tr>
<tr>
<td>Women</td>
<td>3/14 (21%)</td>
<td>5/6 (83%)</td>
<td>P=0.036</td>
</tr>
<tr>
<td>TCL</td>
<td>429 ms</td>
<td>435 ms</td>
<td>95% CI=−47.5–35.5</td>
</tr>
<tr>
<td>A-A-V pattern</td>
<td>11/14 (79%)</td>
<td>1/6 (17%)</td>
<td>P=0.036</td>
</tr>
<tr>
<td>PPI–TCL</td>
<td>176 ms</td>
<td>118 ms</td>
<td>95% CI=26.3–89.7</td>
</tr>
<tr>
<td>PPI–TCL&lt;115</td>
<td>0/14 (0%)</td>
<td>3/6 (50%)</td>
<td>P=0.029</td>
</tr>
<tr>
<td>PPI–TCL&lt;125</td>
<td>0/14 (0%)</td>
<td>5/6 (83%)</td>
<td>P=0.001</td>
</tr>
<tr>
<td>cPPI</td>
<td>170±34 ms</td>
<td>115±69 ms</td>
<td>95% CI=21.7–88.3</td>
</tr>
<tr>
<td>cPPI&lt;110</td>
<td>0/13 (0%)*</td>
<td>4/6 (67%)</td>
<td>P=0.007</td>
</tr>
<tr>
<td>∆VA</td>
<td>160±34 ms</td>
<td>101±76 ms</td>
<td>95% CI=25.9–42.2</td>
</tr>
<tr>
<td>∆VA&lt;85</td>
<td>0/14 (0%)</td>
<td>4/6 (67%)</td>
<td>P=0.005</td>
</tr>
<tr>
<td>∆HA&lt;0</td>
<td>72±40 ms</td>
<td>-1±58 ms</td>
<td>95% CI=35.6–110</td>
</tr>
<tr>
<td>∆HA&lt;0</td>
<td>0/11 (0%)</td>
<td>4/6 (67%)</td>
<td>P=0.012</td>
</tr>
<tr>
<td>NFRT/Atypical AVNRT</td>
<td>P&lt;0.05</td>
<td>P=0.01</td>
<td></td>
</tr>
<tr>
<td>∆AH</td>
<td>29±19 ms</td>
<td>10±17 ms</td>
<td>95% CI=3.03–35.0</td>
</tr>
</tbody>
</table>

AV indicates atrioventricular; AP, accessory pathway; CI, confidence interval; cPPI, corrected postpacing interval; NF, nodofascicular; NFRT, NF reentrant tachycardia; PJRT, permanent form of junctional reciprocating tachycardia; PPI, postpacing interval; SVT, supraventricular tachycardia; and TCL, tachycardia cycle length.

*PPI could not be calculated in 1 patient because a His bundle deflection was not observed after entrainment.
NF that resemble atypical AVNRT. Entrainment results, therefore, should be corroborated with the other important findings of the study. Entrainment is not possible for patients with only nonsustained tachycardia or whose tachycardia repeatedly terminates with pacing. In such situations, evaluating the response of tachycardia at the beginning of ventricular overdrive pacing can help differentiate ORT from AVNRT but does not distinguish NFRT from PJRT or AVNRT/NF AP from ORT.21,22 Atrial extrastimulation and overdrive pacing were not systematically performed during tachycardia to exclude junctional tachycardia or assess VA linking.23–25 However, because focal junctional tachycardia associated with retrograde conduction over the SP is extremely rare and none of our tachycardias exhibited nonreentrant behavior (eg, warm-up/cool-down phenomenon, initiation after a spontaneous junctional complex), we are confident of our diagnoses. In addition, the value of VA linking in long RP tachycardias is unclear because VA intervals can vary significantly during atypical ORT and AVNRT because of decremental conduction over the AP and SP, respectively. Rather, AT was excluded by classical electrophysiologic criteria.

Conclusions

Diagnosing the atypical AV node–dependent long RP SVT requires separate pacing maneuvers to delineate the upper and lower limbs of the circuit. Long PPIs are common and a PPI–TCL<125 ms seems better than 115 ms for differentiating ORT (PJRT/NFRT) from atypical AVNRT. Other entrainment criteria (ePPI<110 ms, ∆VA<85 ms, and ∆HA<0 ms) are only modestly sensitive but 100% specific for ORT. Differentiating nodal tachycardias (NFRT/atypical AVNRT) from PJRT can be established by ∆AH criteria or the paradoxical finding of AH_{vent} < AH_{NSR}. His-refractory VPDs was the only maneuver to identify a bystander, concealed NF AP during atypical AVNRT.

Disclosures

None.

References


Table 2. Differential Diagnosis of a Long RP Tachycardia Reset (Advanced or Delayed) or Terminated (With VA Block) by His-Refractory VPDs

<table>
<thead>
<tr>
<th></th>
<th>Atypical AVNRT+Bystander NF</th>
<th>NFRT</th>
<th>PJRT</th>
</tr>
</thead>
<tbody>
<tr>
<td>PPI–TCL</td>
<td>&gt;125 ms</td>
<td>&lt;125 ms</td>
<td>&lt;125 ms</td>
</tr>
<tr>
<td>∆AH</td>
<td>&gt;40 ms or AH_{vent} &lt; AH_{NSR}</td>
<td>&gt;40 ms or AH_{vent} &lt; AH_{NSR}</td>
<td>&lt;20 ms</td>
</tr>
</tbody>
</table>

AVNRT indicates atrioventricular nodal reentrant tachycardia; NF, nodofascicular; NFRT, NF reentrant tachycardia; PJRT, permanent form of junctional reciprocating tachycardia; PPI, postsfiglon interval; VT, ventricular tachycardia; TCL, tachycardia cycle length; and VPDs, ventricular premature depolarizations.

CLINICAL PERSPECTIVE

Atypical atrioventricular (AV) node–dependent long RP supraventricular tachycardias (SVTs) are uncommon and can be challenging to diagnose. Standard SVT criteria using pacing maneuvers in both the atrium (ΔAH criterion) and the ventricle (postpacing interval–tachycardia cycle length, corrected postpacing interval, ΔVA, and ΔHA criteria) were used to define the upper and lower circuit of the 4 AV node–dependent long RP SVTs: atypical atrioventricular nodal reentrant tachycardia (AVNRT) with and without a concealed, bystander nodofascicular (NF) accessory pathway (AP) and orthodromic reciprocating tachycardia using a concealed, slowly conducting, decremental atrioventricular (permanent form of junctional reciprocating tachycardia) or NF AP (NF reentrant tachycardia). We found that a postpacing interval–tachycardia cycle length<125 ms was better than 115 ms for differentiating orthodromic reciprocating tachycardia (permanent form of junctional reciprocating tachycardia/NF reentrant tachycardia) from AVNRT; and other entrainment criteria (corrected postpacing interval<110 ms, ΔVA<85 ms, and ΔHA<0 ms) had 67% sensitivity and 100% specificity for orthodromic reciprocating tachycardia. The ΔAH criterion or paradoxically, AH_{SVT}<AH_{NSR} differentiated nodal tachycardias (NF reentrant tachycardia/atypical AVNRT) from permanent form of junctional reciprocating tachycardia. His-refractory VPDs advanced, delayed, or terminated SVT in all AP patients and was the only maneuver to identify a concealed, bystander NF AP during atypical AVNRT. In summary, diagnosis of these unusual SVTs requires separate pacing maneuvers to delineate the upper and lower limbs of the circuit. Because of decremental retrograde AP conduction, standard entrainment criteria from the ventricle are only modestly sensitive but highly specific for orthodromic reciprocating tachycardia. His-refractory VPDs are important to identify bystander APs during atypical AVNRT.
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