Variable Clinical Features and Ablation of Manifest Nodofascicular/Ventricular Pathways

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Background—Manifest nodofascicular/ventricular (NFV) pathways are rare.

Methods and Results—From 2008 to 2013, 4 cases were identified with manifest NFV pathways from 3 centers. The clinical findings and ablation sites are reported. All 4 cases presented with a wide complex tachycardia but with different QRS morphologies. Case 1 showed a left bundle branch block/superior axis, case 2 showed a right bundle branch block/inferior axis, case 3 showed a left bundle branch block/inferior axis, and case 4 showed a narrow QRS tachycardia and a wide complex tachycardia with a left bundle branch block/inferior axis. Three of the 4 tachycardias had atrioventricular dissociation ruling out extranodal accessory pathways, including atriofascicular pathways. Programmed extrastimuli showed evidence of a decremental accessory pathway in 3 of the 4 cases. Coexisting tachycardia mechanisms were seen in 3 of the 4 cases (atrioventricular nodal reentry tachycardia [2] and atrioventricular reentrant tachycardia [1]). Ablation in the slow pathway region eliminated the NFV pathway in 3 (transient in 1) with the other responding to surgical closure of a large atrial septal defect. The NFV pathway was a critical part of the tachycardia circuit in 1 and proved to be a bystander in the other 3 cases.

Conclusions—Manifest NFV pathways presented with variable QRS expression dependent on the ventricular insertion site and often coexisted with other tachycardia mechanisms (atrioventricular nodal reentry tachycardia and atrioventricular reentrant tachycardia). In most cases, the atrial insertion of the pathway was in or near the slow pathway region. The NFV pathways were either critical to the tachycardia circuit or served as bystanders. (Circ Arrhythm Electrophysiol. 2015;8:117-127. DOI: 10.1161/CIRCEP.114.001924.)

Key Words: catheter ablation $\bullet$ electrophysiology $\bullet$ tachycardia, supraventricular

Variant accessory pathways (APs) include atriofascicular, nodofascicular/ventricular (NFV), and fasciculoventricular pathways. Atriofascicular pathways are the most common with NFV pathways occurring rarely. NFV pathways are defined as a connection between the atrioventricular nodal and fascicle or ventricle. Previous reports of NFV pathways have largely described concealed pathways (antegrade conduction compared with the His bundle and retrograde via the pathway), not manifest pathways during tachycardia. The purpose of this report is to describe the electrophysiological and electrocardiographic features of supraventricular tachycardia (SVT) associated with manifest NFV pathways and their sites of successful ablation.

Methods

From 2008 to 2013, 4 cases were identified with manifest NFV pathways from 3 centers. All 4 patients underwent electrophysiology study. An electrophysiology study was performed with catheters placed in the right atrium, right ventricle (RV), His bundle region, and coronary sinus (CS). Specific diagnostic and ablation catheter selection and ablation technique were performed at the discretion of the electrophysiologist. The clinical findings and electrophysiology studies are reported below. Atriofascicular pathways were excluded in this series by dissociating the atrium from the circuit during tachycardia.

Case 1

A 24-year-old man with a long history of intermittent palpitations presented to the emergency room 20 minutes after the onset of sustained palpitations (Figure 1). The ECG revealed a wide complex tachycardia with left bundle branch block (LBBB)/superior axis QRS morphology and a precardial transition at V5. An echocardiogram revealed normal left ventricular size and function, moderate RV enlargement, and a large secundum atrial septal defect. In the electrophysiology study, baseline intervals included an A–H of 90 ms and a H–V of 36 ms. On atrial extrastimulus testing, there was decremental conduction over the pathway, resulting in a longer A–V time and increasing pre-excitation as the A1–A2 coupling interval decreased (Figure 2A). In sinus rhythm, His bundle activation preceded the right bundle potential (recorded in HIS). During tachycardia, the right bundle potential preceded the QRS, and the His bundle potential was buried in the QRS (Figure 2B). Wide complex tachycardia with a short R–P interval...
WHAT IS KNOWN

• Descriptions of fibers that emanate from the atrioventricular node, cross the central fibrous body, and insert into the ventricle or into the fascicles were first described by Mahaim in the 1930s.

• Although these fibers might be innocent bystanders in relationship with arrhythmias of diverse etiology, demonstration that these fibers are an intrinsic part of a tachycardia circuit is a matter of debate.

WHAT THE STUDY ADDS

• We demonstrate in 4 patients that nodofascicular/ventricular fibers may clearly be manifest and critical to the tachycardia circuit and that the pathophysiology is similar to that seen in patients with atriofascicular pathways except that the atrium is clearly not part of the circuit.

• We also point out that patients with nodofascicular/ventricular tracts are frequently associated with atrioventricular nodal reentry and that often both circuits may be operative.

was initiated with premature ventricular contractions and by atrial extrastimuli (Figure 2B). The earliest atrial activation was recorded at the proximal CS. The tachycardia QRS had variable forms and rates. Tachycardia 1 showed a LBBB pattern with a cycle length of 410 ms and VA block. Tachycardia 2 had a cycle length of 288 ms with or without the LBBB pattern and episodes of spontaneous VA block (Figure 2D). During tachycardia 2, there was a spontaneous transition from a narrow complex tachycardia (atrioventricular nodal reentry tachycardia [AVNRT]) to a LBBB at a slightly lower rate (Figure 2D). The initial delay and switch to a different cycle length during different atrial activation sequences during tachycardia suggest a new mechanism with likely retrograde conduction over a nodal pathway with a different break out. Less likely would be a decremental septal pathway because we observed no advancement of the atrial complex during fused ventricular complex as with RV apical pacing. A third narrow complex tachycardia was induced with atrial extrastimuli that showed an A on V configuration. Ventricular overdrive pacing, 10 ms faster than tachycardia cycle length, revealed a VAV response, with post-pacing interval-tachycardia cycle length=170 ms, and a stim-A minus VA of 100 ms consistent with AVNRT. Ablation was not pursued and surgical closure of a large atrial septal defect proved successful in elimination of pre-excitation and tachycardia. The patient has had no further palpitations since the atrial septal defect closure with >3-year follow-up. Given that there was VA block during the tachycardia, with the atrium not part of the circuit, an extranodal AP (including an atriofascicular pathway) was excluded. The evidence suggests that, in this case, the pathway was a nodofascicular because there was no fusion during RV apical pacing, and the tachycardia morphology showed a typical LBBB pattern. The nodofascicular was both an intrinsic part of the tachycardia circuit and a bystander during AVNRT.

Case 2

A 21-year-old man with wide complex tachycardia was referred for an electrophysiology study. An ECG during tachycardia revealed LBBB/inferior QRS axis with a V6 precordial transition and a 2:1 VA relationship (Figure 3). During atrial extrastimulus testing, the H–V interval decreased with decreasing A1–A2 interval, consistent with the presence of a decremental AP (Figure 4A). Tachycardia was initiated with atrial burst pacing. The QRS morphology during tachycardia matched the pre-excited QRS morphology. During tachycardia, VA block was present, which excluded pre-excited atrial tachycardia or an extranodal AP, including an atriofascicular pathway. During tachycardia, cycle length variability was present, and changes in the HIS–HIS (H–H) interval preceded changes in the ventricular response. Ablation in the slow pathway (initially at the level of the CS ostium) resulted in transient suppression of conduction over the pathway and tachycardia induction for 3 to 5 minutes (Figure 4B), but conduction and tachycardia returned. Ablation was terminated.

Figure 1. A wide complex tachycardia with left bundle branch block/superior axis QRS morphology with precordial transition at V5. aVF indicates augmented vector foot; aVL, augmented vector left; and aVR, augmented vector right.
Figure 2. Continued
when lesions were placed close to the compact atrioventricular node. The tachycardia is presently controlled with flecainide therapy. As in the last case, there was VA block during tachycardia with the atrium not being part of the circuit, a manifest NFV pathway was present. In this case, the NFV pathway appeared critical to the tachycardia circuit (manifest nodofascicular/ventricular (NFV) reentry. A slow pathway modification was performed in the right posterior septum resulting in junctional beats (Figures A–E in the Data Supplement). After ablation, 2 residual echo beats identical to the tachycardia QRS morphology were observed, but tachycardia was no longer inducible, whereas before ablation, it was incessant. The residual echoes show that the NFV pathway was a bystander, a slow pathway modification was performed to target the manifest NFV. After slow pathway ablation the decremental NFV was felt to be a bystander, a slow pathway modification was performed to target the manifest NFV pathway (Figure 7D). Preablation adenosine brought out a manifest anteroseptal AP. Because the manifest NFV was felt to be a bystander, a slow pathway modification was performed to target the manifest NFV. After slow pathway ablation (at the level of the CS ostium), decremental conduction was no longer present. The anteroseptal AP was then ablated, and there was no evidence of either pathway. This patient showed evidence of both a manifest NFV pathway and an extranodal anteroseptal AP. Ablation over the slow pathway rendered the manifest NFV inoperable and allowed for ablation of the extranodal septal AP.

Figure 2 Continued. A, Atrial extrastimuli showed evidence of a decremental pathway with prolongation of A–H and shortening of H–V. B, Tachycardia initiated with both atrial extrastimulation and from a premature ventricular contraction (PVC) as seen above. The initiation follows a PVC (possibly from the HIS bundle catheter), which conducts retrograde over His to the node and initiates pre-excited tachycardia. Note that the first beat is also PVC, which shows retrograde conduction over His (distal to prox as shown with arrows) compared with sinus beat (proximal to distal). The tachycardia was readily initiated by PVCs. C, A second tachycardia with a slower cycle length (410 ms) showing VA block, thus excluding an extranodal pathway (including atriofascicular) or atrial tachycardia. This is exclusively nodofascicular/ventricular (NFV) reentry. D, atrioventricular nodal reentry tachycardia (AVNRT) without (beats 1–4) and with NFV pathway activation (beats 5–9). Note that although the atrial interval increases (330 ms) on transition, the R–R interval remains constant, proving that the NFV tract is part of the tachycardia circuit (beats 5–9) because the A–A interval is dependent on the ventricular activation sequence. Note the slight prolongation of the tachycardia cycle length with transition to the pre-excited tachycardia. The narrow tachycardia had all the features of AVNRT.

Case 3
A 72-year-old man with ischemic cardiomyopathy (ejection fraction, 20%), previous coronary artery bypass grafting, and stable New York Heart Association II heart failure presented to the emergency room with palpitations. An ECG showed a wide complex tachycardia with pre-excitation. During atrial extrastimulus testing, there was no evidence of pre-excitation. During atrial overdrive pacing at a rate similar to the tachycardia cycle length, atrioventricular Wenckebach was seen and the H–V interval was 80 ms (Figure 6A). The tachycardia was inducible with ventricular pacing and was incessant. During tachycardia, there was a 2:1 VA relationship, and there was cycle length variability with changes in the H–H interval preceding changes in the ventricular response. The H–V interval was shorter in tachycardia (43 ms) than in sinus rhythm (80 ms). The tachycardia might be because of AVNRT or junctional tachycardia with associated left anterior fascicular block aberrancy. Against these possibilities was the finding of an identical QRS and H–V as recorded during sinus rhythm with changes that during atrial pacing associated with Wenckebach conduction. In addition, careful examination of the HIS electrogram timing during tachycardia showed a distal to proximal activation sequence (Figure 6C), opposite of what was seen in sinus rhythm or rapid atrial pacing. The latter would exclude AVNRT alone or junctional tachycardia and favor the presence of nodofascicular pathway. Ventricular overdrive pacing from the RV apex at 10 ms faster than tachycardia cycle length entrained the tachycardia without fusion and resulted in a postpacing interval-tachycardia cycle length of 225 ms with V–A–V response consistent with AVNRT. In addition, the tachycardia was terminated after the first unfused paced complex again excluding junctional tachycardia or AVNRT alone but does support the presence of a manifest NFV pathway.

A slow pathway modification was performed in the right posterior septum resulting in junctional beats (Figures A–E in the Data Supplement). After ablation, 2 residual echo beats identical to the tachycardia QRS morphology were observed, but tachycardia was no longer inducible, whereas before ablation, it was incessant. The residual echoes show that the NFV pathway was a bystander. Just as in cases 1 and 2, there was VA block during tachycardia, ruling out an extranodal AP, including atriofascicular. The tachycardia was most consistent with AVNRT and a bystander manifest nodofascicular pathway inserting into the left posterior fascicle, explaining the shortening of H–V during tachycardia. The patient remains without arrhythmias after a follow-up of 16 months.

Case 4
A 52-year-old man with a history of Wolff–Parkinson–White syndrome presented with recurrent palpitations. A Holter monitor revealed wide complex tachycardia at ≈300 beats per minute, prompting referral for an electrophysiology study. A baseline ECG showed manifest pre-excitation via a right anteroseptal pathway (Figure 7A). Atrial premature complexes showed decremental conduction (Figure 7B). Parahisian pacing during sinus rhythm confirmed the presence of a septal AP with retrograde conduction. Spontaneous pre-excited junctional beats were also seen and confirmed the presence of a manifest NFV pathway (Figure 7C). Initiation of nonsustained pre-excited SVT was consistent with atrioventricular reentrant tachycardia with the decremental NFV pathway as a bystander (Figure 7D). Preablation adenosine brought out a manifest anteroseptal AP. Because the manifest NFV was felt to be a bystander, a slow pathway modification was performed to target the manifest NFV. After slow pathway ablation and no evidence of either pathway, this patient showed evidence of both a manifest NFV pathway and an extranodal anteroseptal AP. Ablation over the slow pathway rendered the manifest NFV inoperable and allowed for ablation of the extranodal septal AP.
Main Findings
This report summarizes 4 cases of manifest NFV and highlights the variable role of manifest NFV pathways in influencing tachycardia forms and mechanisms. In each of the cases, the manifest NFV played a role either as a critical part or a bystander to the SVT circuit.

Case 1
This patient had a narrow complex tachycardia with features classic for AVNRT and a pre-excited tachycardia at the identical rate with fused QRS complexes. In addition, at times, changes in SVT cycle length were associated with QRS transition, strongly suggesting that the NFV was critical to the circuit.

Case 2
Case 2, similar to case 1, showed decremental pathway conduction with VA block during tachycardia. Ablation over the slow pathway close to the compact node transiently interrupted pathway conduction and tachycardia. In this case, the pathway was integral to the tachycardia circuit because tachycardia induction was never present in the absence of intact pathway conduction. These findings strongly suggest a manifest NFV critical to the circuit, but we cannot exclude chance ablation of the slow pathway with interception of NFV conduction.

Case 3
Case 3 showed evidence of AVNRT and a bystander manifest NFV pathway with insertion into or near the left posterior fascicle. The pathway was a bystander because slow pathway ablation cured the tachycardia, but evidence of the NFV was still present. Evidence of retrograde His activation and ventricular capture after the first unfused ventricular complex excludes AVNRT or junctional tachycardia.

Of note, in cases 1, 2, and 3, all showed VA block during tachycardia. The atrium in all cases was not part of the circuit, ruling out extranodal APs, including an atriofascicular pathway, as the potential diagnosis.

Case 4
Case 4 showed an interesting combination of an NFV pathway with an extranodal AP. The former was a bystander and was ablated with slow pathway ablation, whereas the extranodal anteroseptal AP was ablated at 12 o’clock on the tricuspid annulus. The evidence of an NFV pathway is the junctional...
beats with pre-excitation and the presence of pre-excitation during initiation of the SVT, proving that the NFV must have been a bystander pathway.

**Characteristics of the ablation site**

In all subjects in whom ablation was attempted (cases 2–4), the initial ablation was performed at the level of the CS ostium.
just posterior to the tricuspid valve. If ablation was not successful, the ablation was performed progressively cephalad toward the compact node. In case 1, the large surgical patch suppressed both the tachycardia and the presence of the NFV pattern. The surgical patch resulted in disruption of the slow pathway and contiguous regions of the right atrium. Case 2 had transient elimination of both NFV conduction and tachycardia with ablation lesions placed over the slow pathway region. In this case, the NFV was likely critical to tachycardia maintenance, and the nodal insertions proved to be close to the compact node. In case 3, tachycardia was only inducible from the ventricle and did not show decremental atrioventricular conduction, the NFV inserted near or on the left posterior fascicle (Figure 8). We have previously reported successful ablation of left sided concealed NF pathways. The successful ablation site was high on the septum (Figure ID in the Data Supplement). Similarly, in case 4, the NFV pathway inserted on the slow pathway and was successfully ablated at the midseptal level just inferior to the compact node. The tachycardia was clearly dependent on a separate right anteroseptal AP.
Figure 6. A, During atrial overdrive pacing producing Wenckebach at a rate similar to tachycardia, we observe a right bundle branch block (RBBB)/inferior axis QRS pattern and a H–V of 82 to 86 ms. B, Tachycardia with a RBBB/superior axis. Tachycardia shows shortening of the H–V interval (41 ms). The left anterior fascicular block pattern seen during tachycardia might be related to aberrant conduction over an abnormal HIS Purkinje system during atrioventricular nodal reentry tachycardia (AVNRT) or junctional tachycardia (JT). C, Tachycardia with the HIS electrograms scale increased for better visualization of the HIS timing. The activation sequence of the HIS deflection is distal to proximal opposite to that seen previously in sinus rhythm and excludes AVNRT or JT.
Figure 7. Continued
Our series adds to previous reports by showing that NFV pathways often coexist with other tachycardia mechanisms (especially AVNRT). Grogin et al reported the coexistence of AVNRT and NFV pathway in 2 of 6 patients with decremental conduction along an NFV pathway. Similarly, in their series, ablation in the right posteroseptal region resulted in

**Figure 7 Continued.** A. Baseline ECG with manifest pre-excitation showing a right anteroseptal pathway. B. Atrial premature complexes showed decremental conduction. C. Pre-excited junctional beat (*). Junctional beat showed pre-excitation similar to the pre-excited pattern, thus excluding an extranodal pathway as a cause for this beat. D. Initiation of nonsustained pre-excited tachycardia with different pre-excitation patterns as previously seen, consistent with atrioventricular reentrant tachycardia with the decremental nodofascicular/ventricular pathway as a bystander (antergrade conduction down nodoventricular pathway retrograde over the septal accessory pathway).

**Figure 8.** Proposed circuits. A. Proposed circuit of manifest nodofascicular (NF) pathway for patients 1 and 2: the nodofascicular pathway originates in the posteroseptal space and inserts into the right bundle. B. Proposed circuit of manifest nodofascicular pathway for patient 3: the nodofascicular pathway originates in the left posterior extension and inserts into the left posterior fascicle. C. Proposed circuit of manifest nodoventricular (NV) pathway for patient 4: the nodoventricular pathway originates in the posteroseptal space and inserts into the right ventricular muscle. An extranodal right anteroseptal accessory pathway (AP) is also present. LB indicates left bundle branch; and RB, right bundle branch.
complete ablation of both the slow atrioventricular nodal pathway and the NFV tracts. Others have reported similar success with ablation in the slow pathway region for elimination of NFV pathways. Ho et al reported a series of patients with atypical AVNRT in which 3 patients had a coexistent-concealed NFV pathway. Abbot et al reported a series of 6 patients with coexistent Mahaim and accessory connections. Four patients had NFV pathways, with 2 of them coexisting with the posteroseptal pathway. Our report confirms previous studies and adds several unique features. To our knowledge, AVNRT with a bystander pathway involving the posterior fascicle has not been previously reported and emphasizes the importance of clearly defining the tachycardia mechanism. Similarly, case 4 showed evidence of a bystander NFV with an anteroseptal pathway, which was somewhat masked until the NFV was ablated.

Conclusions

Manifest NFV pathways are rare and may be either a critical part of the SVT circuit or a bystander. It is therefore critical to determine the mechanism critical for tachycardia maintenance because targeting the NFV may not eliminate tachycardia in cases where it is a bystander. In summary, manifest NFV pathways present with variable QRS morphologies that depend on their ventricular insertion sites and often coexist with other tachycardias (AVNRT and atrioventricular reentrant tachycardia). In most but not all cases, the atrial insertion of the manifest NFV is near the slow pathway region.

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Disclosures

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Supplemental Figure A: Tightest programmed APCs. During atrial extrastimuli (tightest programmed APCs) an HV of 80 msec is seen. B: During RF ablation over the slow pathway produced junctional rhythm with the same pattern of QRS as well as the H-V interval seen in sinus or pacing. C: Tachycardia termination after the first un-fused ventricular paced complex, which shows that the ventricle is involved in the circuit. Diagnosis is not compatible with JT or AVNRT alone. D: Final fluoroscopic images (RAO and LAO) showing place of successful termination in the right septum. The ablation catheter was gradually advanced superiorly, toward the tricuspid valve where JT was initiated and tachycardia was no longer inducible. RA: Right Atrium, RV: Right Ventricle. HIS: bundle of HIS, CS: Coronary sinus, ABL: ablation catheter. E: During induction of tachycardia the HIS bundle is within the a broader QRS with RBBB & LAD similar to the patterns seen with the other cases.
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Junctional rhythm results in long HV, confirming tachycardia was not JT
Supplemental Figure C: Tachycardia termination after the first un-fused ventricular paced complex, which shows that the ventricle is involved in the circuit. Diagnosis is not compatible with JT or AVNRT alone.
**Supplemental Figure D:** Final fluoroscopic images (RAO and LAO) showing place of successful termination in the right septum. The ablation catheter was gradually advanced superiorly, toward the tricuspid valve where JT was initiated and tachycardia was no longer inducible. RA: Right Atrium, RV: Right Ventricle. HIS: bundle of HIS, CS: Coronary sinus, ABL: ablation catheter.
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