Anatomic and Electrophysiologic Substrate of the Permanent Form of Junctional Reciprocating Tachycardia

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Data are reported on three patients with the permanent form of junctional reciprocating tachycardia, in whom conduction over a slow accessory pathway was observed after His bundle ablation. Tachycardia was almost incessant and showed a retrograde P wave (P') and RP' interval longer than P'R interval in all patients; during sinus rhythm, the PR interval was normal and there was no evidence of a delta wave. An accessory pathway with a long conduction time located in the posterior pyramidal space provided the retrograde limb of the reentry circuit. After His bundle ablation, the accessory pathway was capable of conducting in both anterograde and retrograde directions with decremental properties in all patients.

Postmortem documentation of the accessory pathway was achieved in one patient. Serial sections revealed an accessory atrioventricular connection composed of ordinary myocardium joining the lower rim of the coronary sinus outlet to the uppermost ventricular muscle. This anomalous atrioventricular connection pursued a sinuous, tortuous path. As a result of changing cross-sectional area, such an accessory pathway might exhibit slow conduction, thus explaining its decremental characteristics.

Methods

Clinical Data

Patient 1 was a 36 year old woman with no evidence of organic heart disease, who had had supraventricular tachycardia for 14 years. Heart rate ranged between 130 and 180 beats/min during tachycardia.

Patient 2 was a 52 year old man with a recent myocardial infarction and a 15 year history of supraventricular tachycardia (130 to 160 beats/min).

Patient 3 was an 18 year old youth with obesity and tachycardia since birth. The tachycardia rate ranged between 160 and 200 beats/min. He developed repetitive episodes of congestive heart failure in the last 2 years; physical examination of the heart showed marked cardiomegaly with a ventricular gallop and a grade 2/6 holosystolic murmur audible at the apex. Slight peripheral edema was present.

In all patients, the tachycardia was almost incessant and unresponsive to pharmacologic treatment.
Electrocardiographic Findings

Sinus rhythm beats showed a normal PR interval and no evidence of a delta wave in all patients. During tachycardia, a retrograde P wave (P') and a P'R interval shorter than the RP' interval were evident (Fig. 1).

The initiation of tachycardia was different in the three patients (Fig. 1):

1) In patient 1, tachycardia occurred without any modification of the PP cycle or PR prolongation.
2) In patient 2, in whom several initiating mechanisms were observed (critical shortening of the PP interval, premature atrial beat with lengthening of the PR interval, junctional escape beat and premature ventricular beat), the tachycardia could also occur without any antecedent event;
3) In patient 3, the first tachycardia beat was preceded by a critical shortening of the PP interval.

Preoperative Electrophysiologic Studies

Electrophysiologic investigation was performed according to the technique described previously (11). Endocardial mapping of the right atrium and coronary sinus during tachycardia in all patients (Fig. 2) showed a septal sequence of retrograde atrial activation, the earliest retrograde electrogram being recorded at the coronary sinus orifice. Premature ventricular stimulation introduced during tachycardia at the time when the His bundle was refractory resulted in shortening of the atrial cycle length without disturbing the sequence of atrial activation. These observations suggested that an accessory pathway was present and that the lower junction of the reentrant circuit was in or below the distal His bundle (12). Earlier programmed premature ventricular stimulation terminated tachycardia without conduction to the atrium, further suggesting that the rhythm was indeed reciprocating. With resumption of sinus rhythm, the AH and HV intervals were normal.

Analysis of retrograde conduction in two patients. In Patient 2 (Fig. 3), ventricular pacing at a cycle length of 360 ms performed during sinus rhythm initially showed a sequence of atrial activation (earliest activity was at the atrial septum recorded by the His bundle catheter) consistent with retrograde conduction over the His bundle-AV node, with gradual lengthening of the ventriculoatrial (VA) interval from 120 to 150 ms. At the end of the retrograde Wencke-
Figure 2. Endocardial mapping of right atrium and coronary sinus during tachycardia. The earliest retrograde electrogram is recorded near the coronary sinus orifice in all three patients. A premature ventricular depolarization (S) (arrow) introduced when the His bundle is refractory results in shortening of the AA interval without disturbing the sequence of atrial activation. DCS = distal coronary sinus electrogram; HBE = His bundle electrogram; PCS = proximal coronary sinus electrogram; Pt = patient; RA = right atrium electrogram; St = ventricular pacing.

In Patient 3, ventricular pacing at a cycle length of 380 ms instituted during reciprocating tachycardia at a cycle length of 410 ms showed 1:1 capture of the atrium with a sequence of atrial activation and a VA interval similar to that observed during spontaneous tachycardia (earliest electrogram in proximal coronary sinus). A decrease to a cycle length of 300 ms showed a long sequence of retrograde Wenckebach phenomenon, followed by a shorter VA conduction time with initiation of retrograde activity in the atrial septum recorded on the His bundle electrogram.

His Bundle Ablation

 Interruption of the His bundle was accomplished using the catheter technique (10,13–16) in Patients 1 and 2. Two shocks (200 and 280 J, delivered energy) were required in
Figure 3. Patient 2. Endocardial mapping of the right atrium and the coronary sinus during ventricular pacing. At a cycle length (CL) of 360 ms (top), the earliest retrograde atrial activation is recorded on the His bundle electrogram; a gradual lengthening of the VA interval is evident. After the fifth QRS complex, a sudden increase of the VA interval simultaneous with a change (earliest electrogram in proximal coronary sinus) in the sequence of atrial activation (arrow) occurs. Pacing at a cycle length of 330 ms (middle) results in a similar pattern, followed by retrograde block (B) and an atrial escape beat (E). The retrograde conduction then resumes with the same atrial activation sequence and VA behavior as seen before the block. In the lower panel, tachycardia induction is shown. It occurs when a sudden increase of VA conduction simultaneous with a change in the retrograde activation sequence (arrow) is achieved. (See text for further details.) Abbreviations as in Figure 2.

Patient 1, whereas in Patient 2, a single electrical discharge (280 J) was sufficient to obtain therapeutic results. Patient 3 underwent surgical ablation of the AV node-His bundle; he was studied before introduction of the catheter technique and details of his operation were previously described (6). After the His bundle ablation, a permanent pacemaker was implanted in all patients.

Results

Postoperative electrophysiologic studies. After ablation of the His bundle, all three patients had no evidence of anterograde or retrograde conduction over the AV node-His bundle, the activation of the ventricles occurring over the anomalous pathway with decremental properties. A large delta wave suggesting a pure pre-excitation pattern associated with decremental properties was present. This was immediately apparent in Patients 1 and 2, but was recognized in retrospect in Patient 3 as a transient phenomenon due to high degree block in the accessory pathway probably related to the extensive surgical damage of the posterior septal area. No spontaneous tachycardia was noted, nor could it be induced by atrial or ventricular pacing. The decremental characteristics of anterograde conduction of the accessory pathway after His bundle ablation was manifested by the presence of transient spontaneous Wenckebach phenomenon in all patients (Fig. 4). Stable 1:1 AV conduction resumed in all three patients after 2 to 3 weeks (Fig. 5) with a long PR interval (340, 320 and 280 ms, respectively) and a QRS configuration suggesting posterior septal pre-excitation (initial forces negative in leads II, III and aVF and positive in leads I, aVL and V1 to V6). The decremental properties of retrograde conduction over the accessory pathway (documented in the preliminary study in two of the three patients) were observed after His bundle ablation in all three patients. Figure 6 shows the pattern of retrograde conduction in Patient 1: ventricular pacing at a cycle length of 380 ms resulted in retrograde Wenckebach phenomenon, the earliest atrial electrogram being recorded near the coronary sinus orifice.
Follow-Up

During the follow-up period (15 months, 5 months and 6 years, respectively), all patients remained free from tachycardia without antiarrhythmic medication. Patient 2 developed a ventricular aneurysm as a result of his myocardial infarction, and died suddenly 5 months after the procedure; pathologic examination of the heart was performed. Persistent conduction over the accessory pathway was only transient in Patient 3, presumably because of extensive surgical dissection of the septal region.

Pathologic Examination in Patient 2

Methods. In addition to routine examination of the heart, a heart fragment was excised, encompassing the right inferomedial atrial and ventricular wall on either side of the AV sulcus, the trigonum fibrosus, the pars membranacea and the upper two-thirds of the ventricular septum (with the AV junctional specialized pathway). The block of tissue was embedded in paraffin and complete serial sections were obtained according to a method previously described (17). Each section was stained with hematoxylin-eosin and Hedinhein's (azan) method.

Postoperative findings. The heart weighed 480 g. The left anterior descending coronary artery showed atherosclerotic obstruction at a point 1.5 cm from its origin; a healed anterior myocardial infarction with aneurysmal dilation was noted. A recent thrombotic occlusion of the distal right coronary artery with subacute posterior apical myocardial infarction was also found. The pacing electrode catheter was completely surrounded by a fibrin sheath at its tip at the apex of the right ventricle.

Serial sections revealed the following findings. Proceeding from the bottom up, a group of tiny atrial fibromuscular bundles were seen to descend into and criss-cross the fat of the AV sulcus, approaching and intermingling with fascicles belonging to the uppermost ventricular muscle through narrow clefts of rudimentary, fenestrated AV fibrous anulus immediately behind the central fibrous body (Fig. 7, top). These findings confirmed an accessory AV connection of direct Kent bundle type, whose ordinary fibers originated from the lower rim of the coronary sinus outlet. No peculiarities of AV nodal specialized tissue were observed in the anomalous pathway. These fibers exhibited spotty interstitial fibrosis, while infringing on the ventricular myocardium through the narrow fissures of the AV anulus. Because of the general orientation of the anastomosing fascicles perpendicular to the cut, the AV muscular contiguity was barely evident on individual sections of these series. In turn, on the ventricular end of the connection, some ventricular bundles bent so sharply as to present parallel to the section plane, whereas more laterally, a patch of pronounced sclerosis dissociated and disrupted the myocardium (Fig. 7, bottom).

The AV node exhibited severe fibrotic scarring (with numerous newly formed capillaries) of its outer layer, with complete interruption of the atrio-AV nodal connections. The upper and penetrating AV bundle was also totally interrupted by scar tissue replacement; the overlying endocardium on the right side was markedly thickened. The bifurcating His bundle at the outlet of the tunnel in the pars membranacea septi was slightly lipomatous but well preserved. The roots of both bundle branches, separated from each other by a fibrous spur (protruding from the wall of the fibrous tunnel), were healthy.
Figure 5. Standard electrocardiograms recorded 2 to 3 weeks after His bundle ablation. In all three patients, stable sinus rhythm (with sinus tachycardia in Patient 3) is present. The PR interval measures 340, 320 and 280 ms, respectively. The QRS configuration suggests posteroseptal pre-excitation. (See text for further details.)

Discussion

The similarity of both clinical and electrophysiologic features before and after His bundle ablation in all three patients and the histopathologic findings in one patient permit us to formulate a unified hypothesis about the structural-functional substrate of the permanent form of junctional reciprocating tachycardia.

An accessory pathway with decremental properties provides the retrograde limb of the reentry circuit. The diagnosis of an accessory AV pathway utilized in retrograde direction was demonstrated by: 1) an abnormal atrial activation sequence during tachycardia with the earliest electrogram recorded near the coronary sinus orifice (18–20); 2) pre-excitation of the atria by a premature ventricular depolarization introduced during reciprocating tachycardia when the His bundle was refractory, without disturbing the sequence of atrial activation (21); and 3) termination of tachycardia by premature ventricular stimulation introduced at intervals that did not result in retrograde depolarization of either the His bundle or the atria. Decremental properties of the accessory pathway were demonstrated by rate-dependent VA conduction during ventricular pacing with the same sequence of atrial activation as observed during tachycardia.

Theoretically, it is conceivable that a nodoventricular fiber provides the retrograde limb of the reentrant circuit in the permanent form of reciprocating tachycardia. In fact, retrograde conduction to the atrium during tachycardia may
be achieved even when the His bundle is refractory, because of longitudinal dissociation within the AV node. If this were the case in our patients, the electrocardiographic pattern observed after His bundle ablation should imply the survival of a slow conducting nodoventricular bypass tract in all three patients. On the contrary, the findings obtained from Patient 2, in whom postmortem examination was performed, are against this theoretical possibility. Indeed, the lesions observed in the AV region, showing severe fibrotic scarring of the AV node with complete interruption of the approach to the AV node and penetrating His bundle, make the persistence of a hypothetical nodoventricular fiber unlikely.

The accessory pathway is located in the posterior pyramidal space. The posterior septal location of the accessory pathway was suggested by the localization of the atrial and ventricular insertions. Thus, the earliest retrograde activity recorded during conduction over the accessory pathway was situated just inside the orifice of the coronary sinus. Electrocardiograms obtained after His bundle ablation showed sinus rhythm with a QRS configuration consistent with a ventricular insertion in the posteroseptal region. This location was confirmed histopathologically in Patient 2.

Figure 7. Patient 2. Photomicrographs taken from serial sections of the right atrioventricular junction (top) and at the level of the ventricular end of the accessory pathway (bottom). Top. Note atrial fascicles passing from the right atrium across the fat of the atrioventricular (AV) sulcus to join the ventricle (V) through gaps in the AV anulus. Bottom. Close to the anastomosing AV accessory fascicles and defective AV sulcus, ventricular fibers change direction and present with pronounced sclerotic replacement. (Hematoxylin-eosin stain, original magnification x 19, reduced by 27%.)

The accessory pathway can conduct in an anterograde manner in the absence of conduction over the AV node-His bundle. Despite the widely held belief that the accessory pathway in patients with the permanent form of junctional reciprocating tachycardia can conduct only in a retrograde direction, we noted pre-excitation with a long PR interval in all patients after ablation of the AV node-His bundle induced by catheter technique or surgery. This phenomenon was only transient in the surgically treated patient (Patient 3), presumably because extensive dissection and freezing of the septal space also damaged the accessory pathway.

The decremental properties of the accessory pathway may be explained by anatomic considerations. As just discussed, the accessory AV pathway was characterized by the pronounced tortuosity of the Kent fibers bridging the AV sulcus, with an oblique course through the AV anulus fenestrations. Spach (22) emphasized the important influence that geometric features (macroscopic and microscopic) can have on the propagation of depolarization in normal and abnormal cardiac muscle. Theoretically, one would expect substantial disorders in axial resistivity to be present along
the course of a fiber that pursued a sinuous, tortuous path. As a result of changing cross-sectional area, such a fiber might predictably exhibit slow conduction similar to that observed in the AV node. The structural and geometric asymmetry of the accessory pathway, as found in Patient 2, may be critical to decremental conduction. Although this case is not representative of all patients with the permanent form of junctional reciprocating tachycardia, one can speculate that directional distortion and partial fibrotic thinning of the accessory pathway may have a significant bearing on impairment in impulse propagation along the accessory pathway itself.

On the basis of our data, it appears reasonable to modify some currently accepted concepts.

"Impedance mismatch" is not responsible for the lack of pre-excitation in sinus rhythm. The absence of pre-excitation during sinus rhythm, although attributed in some cases to fibrosis of Kent bundles or neural lesions, or both (23-25), has generally suggested that an accessory pathway is unable to conduct in the anterograde direction because of the presence of "impedance mismatch" (26). This phenomenon has been ascribed to the presence of a narrow isthmus of muscle joining a broad sheet of atrial myocardium with a three-dimensional mass of ventricular muscle. Thus, the limited wave front in the accessory pathway might be insufficient to bring the larger volume of ventricular cells to threshold during anterograde conduction, but would be sufficient to bring the atrium to threshold during retrograde conduction. If this were true in our patients, we should have observed complete AV block after His bundle ablation instead of anomalous 1:1 conduction. On the contrary (Fig. 8), we believe that repetitive concealed retrograde conduction from the normal conduction system to the ventricular insertion of the accessory pathway must be invoked to explain the absence of pre-excitation during sinus rhythm. With removal of this influence by His bundle ablation, the accessory pathway is capable of manifesting anterograde conduction.

This interpretation is also supported by observations in Patient 1, in whom it was necessary to deliver two shocks to the His bundle (Fig. 9). After the first, which only resulted in reversible damage of the bundle followed by the reappearance of the tachyarrhythmia, it was possible to observe, at the end of a Wenckebach cycle in the accessory pathway, the sudden appearance of conduction over the AV node-His bundle (with right bundle branch block due to the shock). It is reasonable to suggest that in that moment, conduction over the normal pathway (which was previously inhibited as a result of concealed retrograde penetration by the impulse coming from the accessory pathway) was now possible because block in the accessory pathway prevented retrograde concealed conduction into the normal pathway.

Figure 8. Proposed hypothesis (right) to explain the absence of pre-excitation in the permanent form of junctional reciprocating tachycardia as an alternative to the "impedance mismatch" phenomenon (left) (see text for details). AP = accessory pathway; AVN = atrioventricular node; SN = sinus node.
Once ventricular activation occurred by the normal conduction system, an analogous mechanism—namely, retrograde concealed conduction from the normal to the anomalous pathway—prevented activation of the ventricles by the accessory pathway. After the second shock, when the AV node-His bundle was irreversibly damaged and concealed conduction into the accessory pathway no longer occurred, stable ventricular activation through the accessory pathway was present.

Critical shortening of the PP cycle is not the only initiating mechanism. Our study provides new information concerning the initiating mechanism of the tachycardia in the permanent form of junctional reciprocating tachycardia. Initiation of tachycardia in this arrhythmia is commonly associated with acceleration of the sinus rate without PR prolongation (1,6,18). However, our data demonstrate that several mechanisms other than critical shortening of the PP cycle can be responsible for the tachycardia initiation. Thus, we observed initiation of tachycardia (Fig. 1) after:

1) a decrease in atrial refractoriness (occurring spontaneously because of a premature atrial beat or a critical shortening of the PP cycle length;
2) a junctional escape beat; and
3) a premature ventricular beat. These findings are analogous to mechanisms reported (27–29) in manifest and concealed Wolff-Parkinson-White syndrome.

“Fast-slow” intranodal reentry should not be invoked as the underlying mechanism. Our data suggest that an unusual variety of intranodal reentry should be considered unlikely in the permanent form of junctional reciprocating tachycardia. In all our patients, the presence of a slowly conducting accessory pathway was documented. In contrast, in the few cases in which a fast-slow intranodal reentry was invoked (3,5), an accessory pathway utilized in retrograde direction by the tachycardia was not excluded. In fact, our data indicate that the behavior of VA conduction cannot be used to distinguish whether retrograde conduction occurs over an accessory pathway with atypical properties or through the normal AV conduction system. Multiple atrial electrograms must also be recorded to define the pathway of retrograde conduction.
Therapeutic implications. The presence of an accessory pathway with decremental properties as the substrate of the permanent form of junctional reciprocating tachycardia provides two potential therapeutic possibilities: ablation of the normal or the accessory pathway. AV node-His bundle ablation can be accomplished by the closed chest catheter technique, as was performed in two of our patients. Because of the latent capability of the accessory pathway to conduct anterogradely, pacemaker dependence will not necessarily occur. In addition, the decremental properties of the accessory pathway will prevent a rapid ventricular response in the event of atrial fibrillation. Alternatively, surgical dissection of the posterior septal accessory pathway has been reported (30) in cases of the permanent form of junctional reciprocating tachycardia, leaving the AV node-His bundle intact. This possibility has been realized only by open chest surgery to date, but the possible use of the closed chest catheter technique to accomplish the same purpose seems feasible.

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References