Preexcitation Syndromes

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Abstract: The classic electrocardiogram in Wolff-Parkinson-White (WPW) syndrome is characterized by a short PR interval and prolonged QRS duration in the presence of sinus rhythm with initial slurring. The clinical syndrome associated with above electrocardiogram finding and the history of paroxysmal supraventricular tachycardia is referred to as Wolff-Parkinson-White syndrome. Various eponyms describing accessory or anomalous conduction pathways in addition to the normal pathway are collectively referred to as preexcitation syndromes. The latter form and associated eponyms are frequently used in literature despite controversy and disagreements over their actual anatomical existence and electrophysiological significance. This communication highlights inherent deficiencies in the knowledge that has existed since the use of such eponyms began. With the advent of curative ablation, initially surgical, and then catheter based, the knowledge gaps have been mostly filled with better delineation of the anatomic and electrophysiological properties of anomalous atrioventricular pathways. It seems reasonable, therefore, to revisit the clinical and electrophysiologic role of preexcitation syndromes in current practice. (Curr Probl Cardiol 2016;41:99-137.)

Introduction



he classic electrocardiographic (ECG) finding of preexcitation consists of a short PR interval and prolonged ORS (inscribing a "delta" wave causing an initial slurring of the QRS complex) in the presence of sinus rhythm.

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The term Wolff-Parkinson-White (WPW) syndrome consists of the above ECG findings with coexistence of paroxysmal supraventricular tachycardia (PSVT). During the course of the last century, the concept of preexcitation syndrome, with its variants, has fascinated and intrigued physiologists, anatomists, and clinicians. The discovery of several anomalous conduction pathways and the various eponyms used, however, has also created controversies with disagreements over their actual anatomic existence, locations, clinical, and electrophysiological significance. This communication updates the contemporary understanding regarding the various preexcitation syndromes and the corresponding eponyms.

Historical Perspective

In 1883, Gaskell¹ showed that auricular impulse spread to the ventricles by passing over the muscular connections that exist between the 2 parts of the heart. Paladino² described numerous myocardial atrioventricular (AV) connections near the base of AV valves. The above findings were followed by the pioneering work of Tawara,³ who detailed the morphology of the AV bundle and its communication with Purkinje fibers distally and origin in AV node proximally in humans. Kent⁴ reported muscular connections between a rat's atria and ventricles, not only in the septum, but in the right and left lateral walls of the heart. He pointed out that muscular connections were of 2 kinds: (1) direct continuity of the auricular and ventricular musculature at certain points; one of the points he specified was at the junction of the interauricular and interventricular septa of the heart; and (2) an intermediate continuity network of primitive fusiform muscular fibers that are embedded in the fibrous tissue of the (AV) rings of the heart.

In the same year, His⁵ described the AV bundle, which bears his name today, as the sole bridge between the auricular and ventricular myocardium. He put his discovery to the proof of experiment and showed that section of this bundle produced discordance in the contraction of auricle and ventricle. He thus concluded that Gaskell's observation was true the auricular impulses pass to the ventricle by a muscular connection. He also noted the muscular continuity between the auricles and ventricles disappeared during development of mammalian hearts in all places except at 1 point: the junction of the auricular and ventricular septa, the point at which Kent also observed a connection. Keith and Flack⁶ described a ring of primitive conduction tissue encircling the AV junction.

Later in 1913. Kent^{7,8} described the muscular connection between auricle and ventricle in the heart of man is not singular and confined to the AV bundle, but is multiple. He observed one point at which a muscular connection between auricle and ventricle exists, is situated at the right margin of the heart. He thus proposed, for purpose of identification, to refer to the connection described as the "right lateral" connection. Cohn and Fraser⁹ provided the earliest description of 2 patients with PSVT, terminated by vagal stimulation. Both patients had ECG findings of short PR interval, an abnormal QRS, that is, 1 patient with right bundle branch block (RBBB) pattern and the second with slurring of the initial portion of the ORS complex. Subsequently, Kent¹⁰⁻¹² supporting his histologic findings of existence of a right lateral muscular connection between the auricle and the ventricle, provided evidence of its functional importance. He observed in an animal experiment that despite severance of all the structures that connect the auricles to the ventricles with the exception of a strip of tissue on the right lateral aspect of the organ, spontaneous beats arising in the auricle still conducted to the ventricle and evoked ventricular response. The severance described above passed through ventricular septum and the whole left ventricle. He thus concluded that the transmitted beats only could have passed over the conducting path contained in the only part of the AV connection remaining, on the lateral wall on the right side of the heart. Subsequently, he suggested the presence of neuromuscular tissue in the AV rings, similar to neuromuscular spindle, which connected both the auricles and ventricles. Based upon his findings, Kent postulated that there were several specialized muscular conduction connections in the mammalian heart.

Mines¹³ postulated circus movement rhythms on the basis of these multiple muscular connections and predicted their role as a mechanism of PSVT; subsequently, several other cases were reported with publication of reports by Wilson,¹⁴ Wedd,¹⁵ and Hamburger.¹⁶ Controversy surrounded Kent's description of multiple auriculoventricular connections, so-called "Kent bundles." Initially, Lewis,¹⁷ and later several other researchers, were unable to confirm his anatomic-histologic findings of a specific neuro-muscular spindle in the right lateral wall.

Wolff et al¹⁸ described surface ECG findings of short PR interval and RBBB pattern in patients with PSVT.

Holzman and Scherff¹⁹ attributed the ECG abnormalities described by Wolff, Parkinson, and White to an abnormal AV connection bypassing the AV node and preexciting the ventricles and proposed a circus movement involving the multiple AV connections as a mechanism of tachycardia. Finally, Wolferth and Wood²⁰ and Wood et al²¹ provided histologic proof

of muscular connections between the right auricle and right ventricle on autopsy in a patient with WPW syndrome. The following year, Segers et al²² proposed the term "delta wave" for the initial slurred component of the QRS complex.

Ohnell²³ coined the term "preexcitation," as a phenomena whereby, in relation to atrial events, the whole or part of the ventricular muscle is activated earlier by the impulse originating in the atrium than would be expected if the impulse reached the ventricles by way of normal conduction system.

Kent's description of the presence of multiple muscular connections was denied by many investigators. In a study of 22 fetal and newborn hearts, Lev and Lerner did not find any muscular communications outside the normal conduction system.²⁴

Mahaim and Benatt²⁵ phrased "para-specific conduction," a term he used to describe properties of fibers directly connecting the lower portion of the AV node and the ventricular septum or between upper part of the bundle of His or each bundle branch and the ventricular septum or any part of the ventricle. In his communication, he opined that if conduction by Kent's fibers is accepted, it should be regarded as an accessory form of conduction: para-specific conduction. His original description of such conduction tracts have since been recognized historically by an eponym, as "Mahaim fibers." Although Kent's description of the presence of multiple muscular connections were denied by many investigators, it must be stated that another group of anatomists, that is, Anderson et al²⁶⁻²⁸ were able to confirm Kent's description of specialized connections in the right atrial wall only. However, they too were unable to find multiple muscular connections across the AV annulus as postulated by Kent. The controversy and lack of proof of their existence, the original description of multiple muscular connections have been historically recognized by the eponym as "Kent bundle."

Meanwhile, James²⁹ detailed distinct conduction pathways, separate from the AV myocardium, which included pathways connecting right and left atria and internodal tracts connecting the sinus to the AV node. Per his description, a majority of these fibers enter at the superior margin of the AV node; a distinct subset bypasses the upper and central AV node, connecting directly with the lower third of the AV node or the bundle of His. He postulated that conduction over such a bypass tract would result in electrographic finding of a short PR interval, with resultant preexcitation, albeit with normal QRS duration, during sinus rhythm. However, Brechenmacher³⁰ described fibers in a patient with ECG finding of short PR interval and normal QRS duration, which bore no similarity to the ones described by James. Again, notwithstanding the lack of proof of their existence or functional significance, these connections have historically been described with the eponym as "James fibers."³¹

To circumvent the myriad of complexities involving the description of preexcitation syndromes, Anderson et al^{32} proposed a nomenclature suitable to both the anatomist and the clinicians. Central to the above was the concept that AV node is that portion of the cardiac tissue responsible for AV delay. For preexcitation to occur, it is necessary for the delay producing area, be either short-circuited, or modified by anatomic or physiologic changes. The proposed classification defined the following as schematically depicted in Figure 1.

 Accessory AV muscle bundle: pathway connecting the atrial to ventricular myocardium outside the AV node-His-Purkinje system (HPS) (the normal pathway [NP]). These were further subdivided into septal and parietal bundles: right parietal connection was named as



FIG 1. It should be noted that AV-AP show an oblique orientation as it is considered the rule with most AV-AP. AFP, atriofascicular accessory pathway; AVN, atrioventricular node; AVP, atrioventricular accessory pathway; FVM, fasciculoventricular Mahaim, James (fiber, partial or complete AV nodal bypass tract); HB, His bundle; LBB, left bundle branch; LIF, left inferior fascicle; LSF, left superior fascicle; MV, mitral valve; NVM, nodoventricular Mahaim; NFM, nodofascicular Mahaim; RBB, right bundle branch; SF, septal fasicle; SN, sinal atrial node; TV, tricuspid valve.

Type B preexcitation pattern and left parietal connection was named as Type A preexcitation pattern on surface ECG.

- (2) Accessory nodoventricular muscle bundle: pathway connecting the AV node directly to the ventricular myocardium, short-circuiting the distal/ lower part of the AV node-HPS.
- (3) *Atriofascicular bypass tract*: accessory pathway inserting into specialized tissues, producing preexcitation variant of short PR interval with a normal QRS duration.
- (4) *Intranodal bypass tract*: postulated as anatomically small and may not be functioning so as to produce normal delay.
- (5) *Fascicular-ventricular accessory connections*: connecting specialized conduction system to the ventricular myocardium and may excite the ventricle earlier than would be via normal conduction route.

Accessory AV Pathways

WPW remains the most common variety of preexcitation syndrome. Surface ECG appearance is because of the conduction over an accessory AV pathways (AV-AP) connecting atrial and ventricular muscle directly bypassing the NP, that is, AV node and HPS. As mentioned earlier, the

Warren M (Sonny) Jackman, MD, FACC, FHRS: Fig. 1 is a current update in the understanding of the various forms of accessory pathways, including the oblique course of accessory AV pathways. I would suggest a few minor changes. The right bundle branch is generally unbranching and without septal insertions until the septal aspect of the moderator band. We now believe that right atriofascicular accessory pathways (AFP in Fig. 1) represent a duplica-tion of the normal AV conduction system⁵⁷ with an accessory AV node (located just above the lateral or posterolateral right AV groove), connecting to an accessory right bundle which extends (unbranching) along the right ventricular free wall towards the apex and fuses with the distal right bundle branch at the free-wall aspect of the moderator band. The initial ventricular insertions are the same Purkinje connections as the free-wall insertions of the moderator band in the apical region of the RV free wall. The second insertions would be the Purkinje branches of the moderator band in the apical region of the septum. Although 3 above mentions that atriofascicular pathways exhibit a "preexcitation variant of short PR interval with a normal QRS duration," conduction delay in the accessory AV node during sinus rhythm generally allows a normal PR interval with little or no change in the QRS complex. The final suggestion relates to nodofascicular and nodoventricular pathways, but would be very difficult to display on this figure. The response to ablation described for the rare, clinically significant forms of nodofascicular and nodoventricular accessory pathways suggest that they originate from the rightward or leftward inferior extensions of the AV node (the two most common "slow AV nodal pathways"), relatively far from the compact AV node.

anatomical location of such pathways can vary between septal and parietal (free wall) location on the right or left side of the heart.

ECG Findings. The classic ECG in WPW syndrome is characterized by a short PR interval (<120 ms) and a prolonged QRS duration. The initial slurring of the upstroke of the ORS complex, (ie, delta wave), represents the anomalous excitation of the ventricle, with muscle-muscle conduction, bypassing the AV node-HPS axis (the NP). The ECG in WPW syndrome can be misleading and lead to erroneous diagnosis of several other ECGclinical entities.³³⁻³⁵ Type A preexcitation in which anomalous connection is between left atrium and left ventricle, is often misdiagnosed as RBBB. right ventricular hypertrophy and, at times, posterior wall myocardial infarction, as shown in Figure 2. Similarly, Type B preexcitaton is misdiagnosed as left bundle branch block (LBBB), septal or anterior wall myocardial infarction or left ventricular hypertrophy, as shown in Figure 3. A posteroseptal location of the accessory pathway mimics an inferior wall myocardial infarction on surface ECG, as shown in Figure 4. The key difference to distinguish the above mentioned from ventricular preexcitation is the length of PR interval.

Of interest, the original description by Wolff, Parkinson, and White did, in fact, describe the ECG findings as "bundle branch block with short P-R interval" in healthy young people prone to PSVT.



FIG 2. A 12-lead electrocardiogram during sinus rhythm with Type A preexcitation consistent with left free-wall accessory pathway. Note the ventricular hypertrophy pattern that can be confused with a Wolff-Parkinson-White electrocardiogram. A short PR interval with delta wave is diagnostic of the latter. The ECG leads are labeled in this and Figures 3 and 4.



FIG 3. A 12-lead electrocardiogram during sinus rhythm with Type B preexcitation consistent with right free-wall accessory pathway. The QRS orientation may mimic left bundle branch block with other clinical entities. Again, the short PR interval and delta wave are the distinctive feature of ventricular preexcitation.

Clinical Features. The clinical presentation of WPW varies from completely asymptomatic individuals with incidental detection on a routine ECG to sudden cardiac arrest as the first presentation. The overall incidence of WPW syndrome is about 0.1%-0.3%, ³⁶⁻⁴¹ and patients would have some symptoms over time, often presenting with palpitations and narrow complex tachycardia, so-called orthodromic AV reentry³⁶⁻⁴¹ whereby antegrade conduction occurs via the NP and retrograde conduction over the accessory pathway (Figs 5 and 6). Orthodronic AV reentry may have a wide QRS due to aberrant conduction, that is, right or LBBB with or without fascicular block. On occasions it may present with a



FIG 4. A 12-lead electrocardiogram with preexcitation consistent with posteroseptal accessory pathway. The deep QS pattern in inferior leads, (II, III and AVF), could be interpreted as inferior Q-wave myocardial wall infarct. A short PR interval and delta wave give a clue to correct diagnosis.

FIG 5. Induction of orthodromic AV reentry with atrial extra stimulation (S2). Premature atrial stimulus (A) conducts over the accessory pathway (no H before the QRS) producing maximal preexcitation (wide QRS complex). Premature atrial extra stimulus (S₂ (B)) at a shorter coupling (S₁S₂) than in (A), blocks antegradely in the accessory pathway (no ventricular preexcitation), conducts down the NP (AV node-HPS) initiating orthodromic AV reentry (narrow QRS complex). Note H recordings, which now can be identified due to antegrade activation via the atrioventricular node with normal HV interval. Tracings shown from top-bottom are surface electrocardiogram (Leads 1, 2 and V1), right atrial and His bundle recordings (HRA and HB, respectively). See also accompanying schema. Ae, atrial echo beats; AP, accessory pathway; H, HB, His bundle. All numerical values are in millisecond.

wide complex tachycardia, may be due to preexcited antidromic AV reentry,³⁸⁻⁴⁰ where antegrade conduction occurs over the accessory pathway and retrograde conduction over the NP. The ECG during preexcited antidromic tachycardia mimics ventricular tachycardia, often posing a diagnostic challenge. Given the differences in conduction and refractoriness, properties between the NP and the accessory pathway, an antegrade block in the accessory pathway with concomitant conduction over the NP (Fig 5), could induce orthodromic AV reentry. Conversely, in the retrograde direction block in the NP (usually HPS) and simultaneous activation of atria via the AV-AP would be the anticipated mechanism for orthodromic atrioventricular reentrant tachycardia (AVRT) initiation (Fig 6). During orthodromic AV reentry, the surface ECG does not manifest any

FIG 6. Induction of orthodromic AV reentry with ventricular stimulation. (A) Premature ventricular extra stimulus (S₂) (Vp in the schema) conducts retrogradely over the HPS and atrioventricular (AV) node to produceH₂ retrograde H₂) and simultaneously atrial activation (A₂) via the left free-wall accessory. The latter is indicated by the eccentric atrial activation sequence with earliest A₂ recorded in the distal coronary sinus electrograms (CSd). Retrograde H₂ potential visible after the V₂ is due to retrograde right bundle branch block and delayed conduction over the left bundle branch to reach the AV node. (see also accompanying schema). As AV node is penetrated by V₂ (ie, it activates H₂), and A₂ via AV-AP, it could not propagate through the AV node and hence orthodromic AVR is not initiated. (B) Shows premature ventricular extra stimulus at a shorter coupling interval than in (A), blocks in distal His-Purkinje (both RBB and LBB), while still conducts over the left lateral accessory pathway and can now propagate through AV nodes to initiate the tachycardia. Recorded H (HB potential) is due to antegrade activation over NP following A₂. Tracings are the same as Figure 5. Ae, atrial echo beats.

preexcitation during tachycardia. Many patients with orthodromic AV reentry may not show ventricular preexcitation during sinus rhythm either. This is due to the fact that the accessory pathway in such cases either only conducts in the retrograde direction, that is, ventriculoatrial (VA) (unidirectional AV block in AP), is located far from sinus node and the impulse does not reach the accessory pathway or there is intra-atrial conduction delay or block.⁴² The clinical presentation in such individuals is identical to individuals with the classic WPW syndrome albeit with one

exception: there may be an absence of ventricular preexcitation during atrial fibrillation (AFib). However, the absence of preexcitation on surface ECG alone is not always sufficient to exclude anterogradely functioning AV-AP. As shown in Figure 7, pacing close to accessory pathway atrial insertion site would unmask preexcitation, which otherwise was "concealed" on the surface ECG.

Warren M (Sonny) Jackman, MD, FACC, FHRS: Many reserve the term "WPW syndrome" for the combination of ventricular preexcitation and either a documented tachyarrhythmia or symptoms of a tachyarrhythmia. Some individuals with ventricular preexcitation (less that half) never develop symptoms. In that context, the incidence of ventricular preexcitation during sinus rhythm is 0.1-0.3%, and the incidence of WPW syndrome is less. "Orthodromic" and "antidromic" in the description of AV reentrant tachycardia (AVRT) refer to the direction of conduction through the normal AV conduction system (AV node). The term "preexcited AVRT" includes both antidromic AVRT (antegrade conduction over an accessory pathway and retrograde conduction over the AV node) and preexcited AVRT using one accessory pathway for antegrade conduction.

Several other clinical entities can present with a wide complex tachycardia in the setting of WPW syndrome and often pose a diagnostic challenge. The differential diagnosis includes one or more of the following:

- (1) Atrial tachycardia.
- (2) Atrial flutter with 1:1 or variable conduction over the accessory pathway.
- (3) Multiple accessory pathways with antegrade conduction over one pathway and retrograde conduction over a second accessory pathway.
- (4) Ventricular tachycardia.
- (5) Atrial or atrioventricular nodal reentrant tachycardia (AVNRT) with conduction over the NP with BBB.
- (6) AV nodal reentrant tachycardia with conduction over bystander accessory pathway.
- (7) Atrial fibrillation.

AFib remains the most serious clinical presentation in patients with WPW syndrome.³⁹⁻⁴⁹ The overall incidence of AFib in patients with WPW syndrome varies 11.5%-39%.^{35,36} The exact causes of higher incidence of AFib in patients with WPW syndrome is unclear. The most dreaded consequence of AFib in WPW syndrome may be conversion to ventricular

FIG 7. Effect of pacing site on conduction over left-sided accessory pathway. Pacing from right atrium (A) demonstrates no ventricular preexcitation on surface electrocardiogram (ECG) with same HV interval and identical QRS complexes during the first, second and fourth complex). Orthodromic AV reentry is initiated with premature atrial beat (A₂). Pacing from left atrium (B) shows left ventricular free-wall preexcitation on surface electrocardiogram with very short HV interval of 5 ms. (first and second QRS complexes). Orthodromic AV reentry is initiated with premature atrial beat (A₂), due to block antegradely in the accessory pathway (no more ventricular preexcitation) and conduction along the NP with slightly aberrant conduction. The above underscores the significance of the pacing site in unmasking antegrade accessory pathway conduction, which otherwise is "concealed" on surface ECG (ie, same QRS complexes as in (A) and last 4 complexes in (B)). Labeling of intracardiac tracings in this and subsequent figures are similar.

fibrillation (VFib) and resultant sudden cardiac death. In fact, this may be the first and only presentation in some cases. The electrophysiological properties of accessory pathways, that is, short antegrade effective refractory period may result in rapid ventricular rates and degeneration into VFib.⁴⁰⁻⁴² The functional properties of the accessory pathway can be assessed by determination of antegrade conduction via decremental right or left atrial or coronary sinus pacing. Similarly, antegrade and retrograde effective refractory period of the accessory pathway can be determined by programmed atrial and ventricular stimulation, respectively. The antegrade refractory period of the accessory pathway roughly correlates to the shortest R-R interval observed during preexcited AFib. Alternatively, inducing AFib during a study provides a direct and accurate assessment of the conducting properties of the accessory pathway and can be used roughly as a risk stratifying tool in the laboratory.⁴¹

It needs to be emphasized that the functional properties of the accessory pathways might change in the setting of adrenergic stress as encountered in sporting or other activities during daily life, thereby underscoring the fact that risk of VFib because of rapid antegrade conduction over the accessory pathway may be greater than laboratory tested unless challenged with isoproterenol. The risk of VFib is the lowest in patients who demonstrate intermittent preexcitation on surface ECG or during documented spontaneous AFib with controlled ventricular rate. As mentioned earlier, in many patients, the accessory pathway conducts only in the retrograde direction, that is, absence of preexcitation on the surface electrogram and unidirectional antegrade AV block over the AP. Such patients present clinically with orthodromic AV reentry and do not run the risk of rapid ventricular rates during AFib.⁵⁰⁻⁵⁴

The regular preexcited tachycardias commonly encountered in clinical practice include:

- (1) Atrial tachycardia and atrial flutter
- (2) Antidromic AV reentry
- (3) AVNRT and bystander preexcitation
- (4) Preexcited AV reentry using 2 or more accessory pathways.

Atrial Tachycardia or Atrial Flutter

Atrial tachycardia or atrial flutter (with 1:1 or 2:1 conduction) presents as regular wide complex tachycardia. In this category of preexcited tachycardias, the origin can vary from sinus node, that is, sinus tachycardia or, rarely, sinoatrial reentry, to automatic or reentrant atrial tachycardia and atrial flutter.

At the outset, the relationship of atrial to ventricular activity is critical in discerning the driver of the tachycardia. In case the atrial ventricular ratio is

greater than 1:1, the diagnosis is usually obvious. While the surface ECG may provide diagnostic clues, confirmation of the mechanism can be achieved through electrophysiology study.

Antidromic AV Reentry. This form of preexcited tachycardia presents clinically, as a regular wide QRS tachycardia, and can pose a difficulty in differentiating from ventricular tachycardia and occasionally AVNRT with bystander ventricular preexcitation. The antegrade conduction during antidromic AVRT is over a fast-conducting accessory pathway, and retrograde conduction is over the His-Purkinje-AV node axis (ie, the NP).⁵⁵ Preexcited AVRT can be readily distinguished from myocardial VT when the latter is associated with AV dissociation or any degree of VA block because preexcited AVRT has a 1:1 AV relationship.⁵⁶ Patients with myocardial VT and 1:1 AV relationship can pose a diagnostic dilemma.

A diagnosis of antidromic AVRT can be made with the following electrophysiological findings:

- (1) Delivery of a late premature atrial beat near the site of early ventricular activation during tachycardia with resultant advancement of the QRS with the same QRS configuration provides proof that the AV-AP is the antegrade limb of the tachycardia circuit.⁵⁷⁻⁵⁹ When the next atrial complex is reset, it confirms the diagnosis of AVRT. However, the anticipated preexcitation of the next atrial complex does not always occur due to VH or HA prolongation following preexcited QRS and may cause confusion regarding the nature of the reentry circuit.
- (2) The retrograde limb of the preexcited AVRT can be determined by the sequence of atrial activation (ie, NP vs another AP), with the exception of anteroseptal AV-AP. Early extra stimuli from the ventricle if advance the atrial response with minimum and no VA delay suggests AV-AP. On the other hand, when the VA conduction time parallels VH delay it would indicate that NP is the retrograde limb of preexcited AVRT.
- (3) Termination of the tachycardia with premature atrial beats without reaching the AV node confirms that the accessory pathway is an active limb of the tachycardia circuit, as shown in Figure 8.
- (4) Termination of the tachycardia with an early premature ventricular extra stimulus, strongly favors antidromic AV reentry as due to the prematurity of the delivered ventricular extra stimulus may block in the HPS and link by collision with oncoming preexcited impulse would not affect AVNRT. This maneuver, however; does not exclude preexcited AV reentry using other AP.

FIG 8. Effect of premature atrial extra stimulus during antidromic AV reentry. (Values labeled in milliseconds.) A timed atrial premature beat A_2 delivered in CS at a coupling interval of 260 ms terminates the wide QRS tachycardia. Note that the timing of the HRA and atrial deflection on the HB are unchanged. Thus, A_2 could not have entered the AV node and yet it stops the antidromic reentry suggesting an active role of the accessory pathway.

(5) Catheter ablation of the accessory pathway during antidromic AV reentry where the atrial complex is not followed by QRS provides the proof of the accessory pathway's active participation in the tachycardia circuit (Fig 9).^{60,61} On the contrary, continuation of AVNRT postablation of the accessory pathway is indicative of the bystander status of the AP in the tachycardia circuit.

The distinction between preexcited AVRT from AVNRT with bystander preexcitation is aided by the following:

- (A) Conversion of a preexcited tachycardia to a narrow complex by spontaneous or induced ventricular premature complex and no change in the cycle length of the tachycardia documents the bystander role of the AP (Fig 10).
- (B) Similarly, the reversal of H-RB (or H-LB [or both]) sequence and change in the H-RB interval during baseline can clearly distinguish AVNRT with bystander preexcitation vs antidromic AVRT (Fig 11).⁶⁰
- (C) Thus recording the bundle branch potential along with the His recording during baseline and tachycardia can clearly delineate the tachycardia mechanism that is AVRT vs AVNRT (Fig 12).

FIG 9. Radiofrequency catheter ablation as a diagnostic tool. Note termination of antidromic AV reentry where the last recorded complex is atrial, followed by no QRS due to interruption of conduction over the AVAR. (Adapted with permission from Panotopoulos et al.⁹⁷)

(D) The finding of H-A (retrograde His activation followed by atrial signal) interval during ventricular pacing greater than H-A interval during tachycardia favors AVNRT bystander ventricular

FIG 10. AVNRT with bystander ventricular preexcitation. The tracing shows ongoing wide QRS tachycardia and conversion to narrow QRS by a spontaneous ventricular premature complex (VPc) and no change in tachycardia CL of 350 ms. Note marked shortening of HV interval from 150-50 ms. The reason is that this type of tachycardia cannot exist without linking in the HPS from AVNRT impulse with retrograde wave front from preexcited QRS. Both the HV intervals labeled are misleading. The long HV of 150 ms is due to the fact that the impulse activates the retrograde A, which in turn excites the ventricle, that is, H-A-V sequence rather than HV. The H impulse does not produce the subsequent QRS. With the narrow QRS tachycardia the HV looks even shorter on HB electrogram due to retrograde Ae precedes the QRS and the HV is longer when measured from the onset of QRS on surface ECG. (Adapted with permission from Jazayeri.⁶⁰)

FIG 11. Antidromic AV reentry using right-sided AV accessory pathway. Sinus rhythm without preexcitation (A). Note the H-RB sequence with normal HV interval. (B) Wide complex tachycardia (LBBB pattern). Now the RB potentials (bold arrows) follows the local ventricular electrograms. This is due to retrograde right bundle branch block (RBBB) (first-sixth beats). On resolution of the retrograde RBBB block, (seventh-ninth beat), the VA interval shortens, and HB and RB potentials no longer can be identified, with shortening of cycle length of tachycardia. The onset of atrial electrogram on the HB merges with the local V electrogram. See also the accompanying schema. ((A) Adapted with permission from Jazayeri et al.⁶⁰)

Warren M (Sonny) Jackman, MD, FACC, FHRS: I believe the authors meant to say, "Now the RB potential (bold arrows) follows the local ventricular electrogram and *follows* the H potential. This is due to retrograde right bundle branch block (RBBB) (first to sixth beats), with ventricular conduction across the septum to activate the LBB retrogradely followed by retrograde activation of the His bundle and then antegrade activation of the RBB."

preexcitation.^{60,61} Reversal of these values during the tachycardia would suggest preexcited AV reentry rather than AVNRT.⁶⁰⁻⁶²

Warren M (Sonny) Jackman, MD, FACC, FHRS: I would add to section B: "An abrupt increase in the tachycardia cycle length (due to an increase in V-A interval) resulting from retrograde right or left bundle branch block (Fig. 11) confirms participation of that bundle branch in the tachycardia circuit, and therefore antidromic AVRT."

Preexcited AV Reentry Using 2 Accessory Pathways

This form of preexcited AVRT involves the participation of two separate accessory pathways with one forming the antegrade limb and the other retrograde limb of the tachycardia circuit (Fig 13). The exact

FIG 12. Preexcited tachycardia using posteroseptal accessory pathway. The H-RB interval (A) during tachycardia (25 ms) is identical to sinus rhythm (C). During right ventricular pacing (B) retrograde H2 emerges from V2 and reaches H2 via the left bundle, the H-RB interval measures zero. Right bundle potential recording is critical along with HB potential to differentiate antidromic AV reentry From AVNRT with AV-AP as a bystander. During this preexcited tachycardia the H-RB interval of 25 ms is identical to sinus which suggests AVNRT and bystander role of AP. In the event preexcited tachycardia was antidromic AV reentry the H-RB interval would be expected to be zero (as in B), that is, simultaneous activation of H-RB, or H-RB interval would be shorter than sinus rhythm.

prevalence of such tachycardia is unknown, but most likely is as common as tachycardia due to antidromic AV reentry.^{54,60-64}

ECG and Electrophysiological Features. Unique to this type of reentry is the finding of several morphologies of wide QRS tachycardia, in the same patient, posing a significant diagnostic ECG challenge. The obvious differential diagnosis include ventricular tachycardia either myocardial or bundle branch reentry and supraventricular tachycardia with aberrant conduction. Similarly, several retrograde conduction patterns, that is, atrial activation sequences are encountered during electrophysiology study.

The presence of 2 separate accessory pathways can be unmasked by use of standard electrophysiological techniques including atrial pacing, that is, right and left atrial close to the atrial site of AP insertion (Fig 13). This often creates block in the pathway closer to the stimulation site and preexcited AV reentry is initiated from the other pathway. This can be reversed by pacing close to the second pathway atrial insertion (Fig 13). This is also true for ventricular pacing to determine the earliest atrial activation site. Contrary to the belief that coexistence and participation of a

FIG 13. Preexcited AV reentry involving 2 accessory pathways. Sinus rhythm (A) demonstrates preexcitation on surface ECG (first beat), consistent with anteroseptal accessory pathway conduction (very short HV interval on HB recordings). Premature atrial beat (B) (A₂) (third complex) from HRA starts preexcited AV reentry with antegrade conduction over left free-wall accessory pathway and retrograde conduction probably through anteroseptal pathway. Note retrograde activation sequence (dotted perpendicular line) with earliest activation in HB electrograms. Pacing from left atrium (C) (CS) demonstrates preexcitation on surface ECG (first and second beats), consistent with left-sided accessory pathway conduction. Premature atrial beat (A₂) (third beat), from left atrium (CS) initiates circus preexcited AV reentry with the reversal of circuit, that is, antegrade conduction over the anteroseptal pathway and retrograde conduction over left free-wall accessory pathway. Note retrograde activation sequence (dotted perpendicular line) with earliest activation sequence (dotted perpendicular line) with the reversal of circuit, that is, antegrade conduction over the anteroseptal pathway and retrograde conduction over left free-wall accessory pathway. Note retrograde activation sequence (dotted perpendicular line) with earliest activation in coronary sinus electrograms. The above reemphasizes the importance of site of proximity of the pacing site to the accessory pathway atrial insertion site. Antegrade conduction block is noted in the pathway closest to the site of pacing. See also the accompanying schema. (Adapted with permission from Akhtar.⁶³)

septal and a free-wall pathway in such a preexcited AV reentry is rare, such cases have been encountered in clinical practice many times (Fig 13), and antidromic AV reentry with no retrograde HPS delay and fast retrograde AV nodal conduction (NP) is not much different in terms of the proximity of the retrograde limb than anteroseptal accessory pathway. Catheter ablation of the accessory pathway followed by diligent testing to assess residual conduction over any other accessory pathways should be carried out with additional ablation of the same if needed.

In summary, the incidence of preexcited AV reentry with 2 or more accessory pathways in the same patient is probably as common than antidromic AV reentry. Coexistence of an AV-AP with a slow or decrementing conducting, that is, atriofascicular pathway (AFP) as a cause of antidromic reentry should also be considered, with the AFP acting as the antegrade limb and the fast-conducting pathway as the retrograde limb of the tachycardia circuit. However, a reverse sequence of activation with the above combination has not been reported.

Warren M (Sonny) Jackman, MD, FACC, FHRS: For a preexcited AVRT, the retrograde limb (AV node or second accessory pathway) can identified by a late ventricular extra stimulus, delivered close to the base near the site of earliest retrograde atrial activation. If the ventricular extra stimulus (which doesn't advance the timing of the His bundle potential, and therefore doesn't reach the AV node, but advances local ventricular activation close to the site of earliest atrial activation by at least 30 ms) results in a change in the timing of atrial activation (with the identical atrial activation sequence) and resets the tachycardia, retrograde conduction over a second accessory pathway forms the retrograde limb of the circuit (preexcited AV reentrant tachycardia using two accessory pathways). If the timing of atrial activation is not changed until earlier ventricular extra stimuli are delivered which advance the timing of His bundle activation, and the H-A interval remains similar, retrograde conduction during the tachycardia is occurring over the AV node (i.e., antidromic AV reentrant tachycardia).

Mahaim Fibers. A detailed communication, in 1937, Mahaim confirmed the functional importance of "para-specific conduction." He based this observation on the finding of lack of AV block when both bundle branches were destroyed simultaneously. He believed that there must be upper connections between the specific tissue and the musculature of the upper part of the ventricular septum, in a region which does not directly communicate with the Purkinje terminal network.²⁵

These connections, for decades, have been referred with the eponym, "Mahaim fibers" and implicated in the genesis of clinical arrhythmias under the general auspices of preexcitation syndromes, that is, "Mahaimrelated tachycardias." Not only their functional significance and anatomicphysiologic correlation remains controversial, their role in participation in clinical tachycardias has endured the fascination of generations of clinical electrophysiologists.

Anatomical studies detailed a variety of accessory connections involving the AV node and the right ventricle, RBB or His bundle and the fascicles. Wellens⁶⁴ first described electrophysiological findings in patient with an accessory pathway exhibiting unusual properties of decremental conduction and long AV conduction time, correlating his findings to the fibers described by Mahaim. The term nodoventricular (NV) bypass tract has been used to describe a pathway when the retrograde His bundle recording follows the ventricular potential. The term nodofascicular (NF) was used to describe a pathway when the retrograde His bundle potential preceded the

ventricular deflection. At the same time, anatomical finding of an accessory AV node coursing through the right ventricle and located on the lateral aspect of the tricuspid annulus was described.^{57,61-70} In a series of 12 patients, Gallagher et al⁶¹ concluded that there appears to be functional counterparts to the proposed anatomic subdivision of Mahaim fibers. He found that in sinus rhythm, the so-called NV fibers can mimic the presence of LBBB and capable of sustained reciprocating tachycardia of LBBB morphology with a VA block. In addition, the so-called NF fibers can mimic intraventricular conduction defect, but no clinical arrhythmias could be attributed to these fibers. In the same communication, he reported a case with 2 distinct NV fibers, that is, an LBB morphology reciprocating tachycardia characterized by 2 distinct VH intervals.⁶¹ With current understanding the 2 sets of VH intervals observed can be explained by retrograde conduction block in the RBB, and transseptal conduction (Figs 14 and 15)^{53,56,61,71,72} Despite the above finding, the term NV variety of Mahaim fibers remained in use as a mechanism of clinical tachycardia with a LBBB pattern.

Even a more recent publication did not make a convincing case for antegrade conduction along the so-called NV pathways.⁷³ However, retrograde participation of NV pathways leading to a narrow QRS tachycardia has been documented.⁷⁴

FIG 14. Antidromic tachycardia using atriofascicular pathway (AFP). The first complex is sinus beat with no preexcitation. Second complex is spontaneous PVC. Ventricular pacing is initiated (leftward directed bold arrows). Antidromic reentry is initiated with 2 consecutive premature ventricular beats (second and third). Note the location of RB potential (rightward directed bold arrows). Tachycardia has left bundle branch morphology (LBBB), and demonstrates no retrograde RBBB. The next 2 beats demonstrate retrograde RBBB, with resultant prolongation of VH, VA intervals as well as the tachycardia cycle length (also see the top schema). In last 2 beats, the retrograde RBBB resolves and RB potential now again precedes the local ventricular electrogram, with change of axis toward normal. The VH and VA intervals shorten with shortening of tachycardia cycle length (bottom schema). It is noteworthy to approach reversal of proximal and distal RB potential activation sequence from retrograde to antegrade direction best seen with first 2 rightward arrows. Right bundle potential (RB proximal and RB distal)

FIG 15. Antidromic tachycardia using atriofascicular pathway. (A) (bottom panel) sinus rhythm without preexcitation on surface ECG. Note H and RB recordings with H-RB sequence (H precedes RB). Right atrial pacing (B) demonstrates preexcitation via AF pathway on surface ECG (LBBB pattern). Note H and RB recordings RB-H with reversal of sequence RB precedes H. H recording is retrograde activation via RB and also the ventricular recording precedes surface QRS complex. Top panel shows initiation of antidromic reentry using AFP with left atrial pacing (CS). Note reversal of H-RB sequence on the fifth beat, with antegrade block in the AV node and conduction over the AFP and onset of antidromic reentry, and this sequence remains constant. H is activated retrogradely via RB during tachycardia. The above emphasizes the importance of RB recordings in delineating the mechanism of tachycardia using AFP. ((A) and (B) Adapted with permission from Tchou et al⁷⁸ (top panel) and McClelland et al⁵⁷ (panels A and B))

Warren M (Sonny) Jackman, MD, FACC, FHRS: In panels A and B, note the nearly identical electrogram recorded at the apical region of the RV free-wall (RVFWA) during sinus rhythm without preexcitation (A) and during preexcitation (B) due to pacing of the right atrial appendage (RAA). The RVFWA electrogram in each panel shows an initial sharp Purkinje potential (RB) followed by the local ventricular potential, which is the site of earliest ventricular activation during ventricular preexcitation (panel B). The electrogram was recorded at a Purkinje insertion from the apical region of the moderator band. During sinus rhythm, the right bundle along the moderator band activates the Purkinje insertion which activates the ventricle. During preexcitation, the same Purkinje insertion is activated by the long, accessory right bundle portion of the atriofascicular pathway, as this long bundle fuses with the moderator band at this site. These Purkinje branches of the apical portion of the Moderator band for the distal insertion of the right atriofascicular accessory pathway.

Although, surgical interruption of the AV node became the treatment of choice for such patients with the so-called Mahaim tachycardia, Gillete et al⁷⁵ reported decremental conducting accessory connections in the anterior aspect of tricuspid valve producing antidromic tachycardia of left bundle branch morphology in the absence of manifest preexcitation. Although the patients in this series did meet the clinical criteria of NV pathway associated ("Mahaim") tachycardia, the site of surgical cure in all these patients, was remote from the AV node and abolition of the antegrade conduction over the accessory pathway was achieved only via surgical incision along anterior right atrium. Furthermore, the ventricular insertion of these pathways appeared to be deep in the anterior right ventricular myocardium. With the advent of direct current catheter ablation of AV node. Bhandari et al⁷⁶ reported the persistence of preexcitation despite achieving complete AV block in a patient with NV fiber associated tachycardia. Similar finding was reported by Klein et al⁷⁷ in 1988, who observed that despite extensive cryoablation of AV node, the preexcitation persisted, and it disappeared only when cryoablation lesion were moved to the right lateral aspect of the tricuspid annulus, thereby suggesting that the accessory pathway was not connected to the AV node, that is, so-called NV fibers. Simultaneously, in a pivotal study Tchou et al⁷⁸ provided electrophysiological proof that the so-called NV fibers were actually originating in the atrium and not the AV node. The insertion of distal end of the pathway into the right bundle was demonstrated by recording the right bundle and His bundle recordings simultaneously. During atrial pacing with maximal preexcitation, the normal sequence of His bundle and right bundle activation was reversed with the appearance of right bundle electrogram preceding the His bundle activation (Fig 15), that is, during maximal preexcitation, the right bundle and then the His bundle were activated in a retrograde direction. The authors also noted that the right bundle electrograms occurred 5 ms before ventricular activation, indicative of the pathway insertion either into the right bundle and hence the origin of term AFP. This study also demonstrated that it was possible to advance the ventricular depolarization through delivery of a late premature atrial beat during preexcited tachycardia which was delivered in the atrium when the AV node was already activated, providing strong evidence that the accessory pathway was independent of the AV node. The abovementioned studies provided unequivocal evidence that the so-called NV fibers were actually located remote from AV node coursing across the right AV groove after originating from the right atrium. These findings had clinical and therapeutic implications, particularly when the treatment with catheter ablation is contemplated. The latter can also be accomplished by targeting

the so-called accessory pathway potentials along the lateral tricuspid annulus away from the AV node.⁵⁷ This was the turning point in the saga of the so-called Mahaim fiber associated tachycardias for providing insight into the actual mechanism of the antidromic tachycardia, using AFP antegradely. The decrimental conduction behavior of AFP also explained the lack of clarity that existed in prior publications using the eponym of "nodoventricular Mahaim."

With this as a background, the variants of preexcitation syndromes (known by the eponym as Mahaim fiber-related tachycardias) outlined below based upon their location, conduction, electrophysiological properties and clinical relevance.

These include:

- (A) Atriofascicular pathway
- (B) Nodoventricular pathway
- (C) Nodofascicular pathway
- (D) Fasciculoventricular pathway

Due to either unproven or extremely rare existence of antegrade conduction along the (NV) pathway, with only proven retrograde conduction and consequently rare narrow QRS tachycardia, or NV or NF pathways are not detailed here. Fasciculoventricular pathway is not implicated in any clinical tachycardia but can produce subtle initial slurring in the QRS which does not change rate acceleration. Hence, only the AFP and related decremental conducting AV pathways are discussed here (schema in Fig 1 and summary in the Table).

Atriofascicular Pathway

Long described as Mahaim Fibers, AFP are accessory AV connections typically with a long anatomic course and decremental antegrade conduction. These pathways comprise approximately 3% of all the overt accessory pathways.⁷⁹ The prevalence of AFP pathways in general population is 0.5-1:100,000. The most common clinical tachycardias associated with these include:

- (1) Antidromic AV tachycardia using the AFP as the antegrade limb of the tachycardia circuit with decremental properties and retrograde conduction through the NP. In some patients the distal insertion may be in the RV myocardium with or without concomitant RB insertion.⁶⁸
- (2) AVNRT and AFP with bystander conduction

Table. Eponyms

Current nomenclature	Preferred nomenclature	Clinical presentations
Kent Bundle (WPW)	Accessory AV pathway	12-Lead ECG short PR, delta wave (WPW) or normal
		Orthodromic and antidromic AV
		reentry, preexcited AV reentry Bidirectional conduction/rapid conduction
		Atrial and AVNRT with bistandard preexcitation
Mahaim fibers		
Decrimental atriofascicular pathway	Atriofascicular pathway, slow conducting AV-AP	Baseline ECG with little or no preexcitation. Antidromic tachycardia with LBBB pattern.
		No retrograde (VA) conduction via atriofascicular pathway.
Nodoventricular	Nodoventricular pathway	Narrow QRS tachycardia/AV dissociation,
		Wide QRS tachycardia, existence not convincingly proven
Nodofascicular	Nodofascicular pathway	
Fasciculoventricular	Fasciculoventricular	Normal PR, subtle and fixed ventricular preexcitation.
		No clinical tachycardia
James/LGL syndrome	ECG description, that is, short PR, narrow QRS	No clinical tachycardia

ECG Features. Given the long antegrade conduction time associated with AFP, little or no preexcitation is seen during sinus rhythm (Figs 14 and 16) or long atrial paced cycle lengths (Fig 15). Minimal preexcitation is in the range of 0%-30% of the cases.

Sternick et al⁶⁸ noted 2 distinct patterns during sinus rhythm in patients with long conduction time and decremental property. The most common ECG finding was of rS pattern in Lead III. In addition, the above absence of q waves in Lead I was noted. The authors further emphasized the absence of the classic delta wave in all. The ventricular insertion site of such pathways is typically in the right bundle or in its close vicinity. However, in a very small percentage, it may be RV myocardium in the vicinity of the RBB, and these patients were identified by absence of rS pattern in Lead III.

Bardy et al⁸⁰ reported 6 ECG features with high sensitivity (92%) and negative predictive value (9%) in identifying antidromic tachycardia using

FIG 16. Atriofascicular pathway potential recordings. During sinus rhythm recording from tricuspid annulus (Tad). (A) Shows distinct atrial, accessory pathway, HB of ventricular and potentials. Note the accessory pathway potential (AP) follows the atrial potential by 85 ms. During antidromic tachycardia (B), the A-AP interval increased to 125 ms and the AP potential precedes the onset of QRS complex by 65 ms. Note that during tachycardia, the ventricular potential at the tricuspid annulus is recorded 25 ms after the onset of QRS complex. Indicating that ventricular activation began far from the atrial end of the atriofascicular pathway. Retrograde His bundle activation (retro H) occurs after the onset of QRS complex and AP potential. Tracings shown (top-bottom) are ECG leads (L1, 2, and V1), right atrial, His bundle proximal, 2, 3, and distal) and tricuspid annulus proximal-distal. (HRA, HBp, HB2, HB3, HBd, TAp, and Tad). (Adapted with permission from McClelland et al.⁵⁷)

Warren M (Sonny) Jackman, MD, FACC, FHRS: Note the similarity of the electrogram recorded at the lateral tricuspid annulus to a His bundle electrogram, with the AP potential identical to a His bundle potential.⁵⁷ The AP potential is generated by the proximal portion of the accessory right bundle branch component of the right atriofascicular pathway, and might be considered an "accessory H potential." In panel B, the increase in interval between the local atrial potential and AP potential (or accessory H potential) to 125 ms is due to delay in the accessory AV node portion of the right atriofascicular pathway. The decremental conduction properties of these pathways always occur in the accessory AV node, proximal to the accessory H potential.⁵⁷ Conduction along the accessory right bundle component is rapid and without decremental properties. During antidromic AVRT, the retrograde His bundle potential (Retro H) is typically recorded within 30 ms of the onset of the QRS due to retrograde conduction along the moderator band and septal right bundle branch synchronous with activation of the apical region of the RV free-wall by the Purkinje insertions from the atriofascicular pathway-monerator band connection.

an AFP pathways. These tachycardias are wide complex with LBBB morphology:

- (1) a QRS axis between 06° and 75° (left axis deviation),
- (2) a QRS duration of 0.15 s or less,

- (3) A monophasic R wave in Lead I,
- (4) a rS pattern in Lead V1,
- (5) transition in precordial leads from a predominant positive QRS complex greater than V4, and
- (6) A tachycardia cycle length between 220 and 450 ms.

The above mentioned criteria have been used to identify preexcited tachycardia owing to antegrade conduction over a decrementing conducting AP. Of interest is the fact that Bardy et al⁸⁰ had referred to it as NV Mahaim, later reclassified as AFP by elucidation of electrophysiological mechanism by Tchou et al⁷⁸ and Klein et al.⁷⁷ It should be noted that in about a third of patients, a QS pattern is seen in Lead V1 during tachycardia. The wide range of QRS axis (06-75°, mean = -31°) during tachycardia is due to variation in the site of AFP insertion in the RBB, right ventricle or both.

Electrophysiological Features. As a majority of patients with AFP have no or minimal preexcitation at baseline, the AH and HV intervals at baseline are essentially normal (Fig 16). Recording of the right bundle (RB) potential is critical and cannot be over emphasized as shown in Figs 14-16.^{57,72,78} During incremental atrial pacing, there is prolongation of AH interval, coupled with decreasing HV interval: as progressive AV nodal delay is encountered, the His bundle (HB) recording merges into the ventricular electrogram and during maximal preexcitation, that is, exclusive conduction over AFP and the HB recording is inscribed after the RB recording (Figs 14 and 15). Expressed differently there is reversal of H-RB during sinus to RB-H during the tachycardia (Figs 14-16.) The response to atrial pacing is qualitatively similar but quantitatively different than the AV node. The conduction delay in the AV node is greater thus exposing the preexcitation. At stable maximal preexcitation there is a constant V-H (ie, retrograde) relationship without further changes despite shortening of the atrial pacing cycle length.

The tachycardia with wide QRS complex is typically induced by introduction of premature atrial beats during sinus rhythm, base atrial rhythm or bursts of atrial pacing (Figs 14 and 15). The QRS configuration

is typically that of a LBBB, representative of right-sided AFP with insertion into the RB or in its close vicinity with ventricular activation directly or via the HPS. A RBBB morphology tachycardia is rarely induced which would be reflective of left-sided AFP. The tachycardia is initiated by inherent delay in antegrade conduction over the AFP, allowing for recovery of conduction over the HPS-AV node. Similarly, during programmed ventricular stimulation retrograde block in the AFP and VA conduction over the NP facilitates the initiation of tachycardia, although antidromic using AFP is relatively easy to induce during ventricular pacing.

During antidromic tachycardia related to right-sided AV pathway, there is a short V-H interval due to early activation of the RBB or local ventricular myocardium. A retrograde block in the RB above the insertion site of the AFP would prolong VH > VA due to transseptal conduction and HB activation through the LBB system (Figs 14 and 15).^{71,78} Of note, the H-A interval is equal to H-A interval during right ventricular pacing at the cycle length of the tachycardia.

During retrograde impulse conduction via the right side, that is, RB >His > atria, the VA interval is short and characteristically the onset of atrial electrogram on HB electrogram cannot be clearly separated from the local ventricular electrogram (Figs 14 and 15). This is in contrast to antidromic reentry using AV-AP, in which the onset of a low atrial electrogram is usually identifiable and separated from the local ventricular electrogram, the reason being that during antidromic (or orthodromic) AV reentry the impulse must traverse the ventricular myocardium to reach the NP or AP, respectively, plus conduction time within the AP, and only then can the atria be activated, which would prolong the VA interval. This observation is only true for atrial deflection on the HB electrogram and applies to all fast-conducting AV-APS, regardless of the AP location.⁸¹ AFP-mediated antidromic reentry produces long PR or short RP as opposed to antidromic AV reentry where PR or RP intervals are closer to each other, albeit concomitant antegrade or retrograde BBB could change these relationships.

A unique feature noted in AFP-mediated antidromic reentry is the finding of RB potential preceding the HB and the QRS complex in the absence of retrograde RBBB (Figs 14 and 15). Such a finding is rarely seen in antidromic reentry mediated via fast-conducting, that is, AV-AP or any other reentrant tachycardia with a LBBB pattern. The proof of active participation of the AFP in the reciprocating tachycardia is provided by advancement of the ventricular potential by delivery of a late atrial premature beat during the period of AV nodal refractoriness.⁷⁸ It must

be noted, however, that failure to advance the right ventricular potential, with introduction of timed atrial premature beats, does not exclude the role of the AFP as an active participant in the tachycardia circuit.⁸² In such cases, delineation of lack of preexcitation during incremental atrial pacing and noninducibility of tachycardia postablation, offers the best proof of the existence of AFP as an integral part of the tachycardia circuit. Less commonly, a long V-H interval may be encountered during preexcited tachycardia using AFP as the antegrade limb due to spontaneous or induced transient retrograde block RBBB, with resultant increase in the tachycardia cycle length (Figs 14 and 15).^{71,78}

Rarely, left-sided pathways with decremental antegrade conduction properties may be encountered.

The other tachycardias related to AFP include the following:

- (1) AVNRT with bystander conduction over the AFP
- (2) Automatic tachycardia arising from the AFP
- (3) Atrial fibrillation
- (4) Nonreentrant preexcited tachycardia

AVNRT With Bystander Conduction Over AFP

The common variety of AVNRT is found in less than 10% of patients with AFP-related tachycardias. The clinical and ECG presentation can be indistinguishable from antidromic tachycardia from AVNRT with bystander preexcitation via AFP. During electrophysiology study, a block in the AFP is achieved with using premature atrial or ventricular extra stimuli, evidence of a narrow QRS tachycardia with identical cycle length would suggest that possibility. In addition, finding of fusion beats during induced tachycardia provides another clue to the mechanism of tachycardia. The finding of extremely short retrograde RB-A (and RB-H) interval favors antidromic reentry, as compared to slow-fast AVNRT with bystander AF where the H would precede the RB potential.

The proof of bystander conduction over the AFP lies in ablation of the same with persistence of inducibility of common AVNRT of the same cycle length.

Automatic Tachycardia Associated With AFP

Patients with AFP can occasionally present with repetitive, nonsustained bouts of automatic tachycardia.^{83,84} The clinical presentation in such patients varies from symptoms of palpitations and noninvasive ECG monitoring reveals isolated, repetitive wide complex beats and nonsustained wide QRS complex tachycardia often resembling accelerated idioventricular rhythm. During electrophysiology study in such cases, no V-A conduction is noted, and a sustained tachycardia is not inducible. However, during atrial pacing, exact morphology of the wide complex beats and nonsustained tachycardia can be replicated and ablation along the tricuspid annulus targeting the pathway, that is, Mahaim potential, can successfully abolish such enhanced automaticity arising from the AFP and provide symptom relief.⁸⁵ Of note, a similar phenomenon of enhanced automaticity is observed during catheter ablation of the AFP during sinus rhythm. The automatic rhythm observed during the application of radio-frequency (RF) current, whether slow or fast, is of identical morphology as the induced tachycardia and is also considered by some as a marker of successful ablation site.⁸⁵ Complete abolition of this automatic rhythm during the course of RF lesion application is indicative of long-term, successful outcome.^{83,84}

AFib in Association With AFP. The overall incidence of AFib in patients with AFP is low (<2%) and is much higher amongst patients with AV-AP syndrome (about 32%). The most common mechanism in patients with WPW syndrome remains degeneration of regular reciprocating tachycardia into AFib. In a patient with preexcited AFib, successful ablation of AFP resulted in noninducibility of atrial fibrillation.⁸⁶ The exact reason for low incidence of AFib in association with AFP remains unclear.

Nonreentrant Preexcited Tachycardia Associated With AFP

Very much akin to nonreentrant tachycardia involving the AV node, socalled 1:2 response,⁸⁷ during which simultaneous conduction over slow and fast pathway is noted, a similar phenomenon has been reported involving AFP with resultant incessant tachycardia with 1:2 (P: QRS).⁸⁸ This patient did exhibit absence of V-A conduction and ablation of the AFP was successful in abolition of dual conduction.

Enhanced AV Node Conduction (Lown, Ganong, and Levine Syndrome)

Lown et al⁸⁹ described a clinical syndrome of short PR interval, normal QRS duration and paroxysmal rapid heart action. Patients with such EEG findings have either an abbreviated or normal AV nodal refractory period and enhanced AV nodal conduction manifest by sub-optimal prolongation of AH interval with increasing pacing rate. This syndrome of enhanced AV nodal conduction, historically referred to as Lown, Ganong, and Levine

(LGL) syndrome, has remained an enigma to clinicians for several decades primarily due to lack of anatomic-physiologic correlation. James postulated the presence of such posterior internodal tract as possible explanation for preexcitation due to this intranodal bypass.⁹⁰ The anatomic proof of such tracts was disputed by Truex and Smythe⁹¹ and Meredith and Titus⁹² who found the fibers described by James, in fact, directly passed into the base of tricuspid valve rather than extending toward the AV node bundle and do not function as bypass tracts. Anderson et al⁹³ were unable to find any fibers connecting the transitional cell zone directly to the nodal-bundle junction. Longitudinal dissociation of the AV node has been also postulated, but never proven anatomically. It must be emphasized here that to date no electrophysiological significance of the abovementioned morphologic findings have ever been proven.

In summary, the eponymous use of James fibers as a cause of preexcitation should be relegated to historical import only. Current evidence mitigates against the existence of any clinical or electrophysiological basis for LGL syndrome.

Therapy Options

For decades, pharmacologic therapy remained a mainstay of treatment of patients with AV-AP-mediated tachycardia. Surgical interruption was recommended for those refractory to drug therapy.^{40,54,77,94,95} The success of catheter ablation in the treatment of supraventricular tachycardias in the last 25 years has made this modality the treatment of choice for all patients today and with AV and AF accessory pathways.

Catheter ablation of the latter is typically guided by mapping along the mitral and tricuspid for AV-AP and the latter for AF-AP. The energy source has gone through evolution from direct current and now to RF and sometimes cryoablation.^{57,66,69,95-98} In patients with overt preexcitation either the atrial or ventricular (Fig 9) insertion sites are targeted during sinus rhythm. At times the AP potential guides the location of ablation (Fig 17). Where overt preexcitation is not present which is often the case (concealed AV-AP) only the atrial site guided by ventricular pacing or orthodromic AV reentry is accessible for targeting. This is only practical in patients with AV-AP as patients with AFP do not have VA conduction. AP potential may be used as a guide. Atrial and ventricular pacing before and after pharmacologic agents such as isoproterenol and/or adenosine are routinely employed to unmask residual conduction in the AP. Detailed discussion of various aspects of AP ablation (catheter mapping, type of

FIG 17. Automaticity in atriofascicular pathway. Upon application of RF current, an accelerated rhythm (third-sixth QRS complexes (marked with rightward arrows) is seen immediately upon start of RF current (white arrow). The QRS morphology of the accelerated rhythm is identical to the fully preexcited complexes (beats 1 and 2) with early ventricular activation at the right ventricular apex (RV) and early retrograde His bundle activation (retro H), suggesting an automatic rhythm originating from the accessory pathway produced by RF energy. After the accelerated rhythm, antegrade conduction over the accessory pathway is absent (seventh beat). The next complex is sinus with no preexcitation, A H potential clearly preceding the QRS. Tracings shown (top-bottom) are ECG (leads 1, 2 and V1), right atrial appendage, His bundle proximal-distal and right ventricular (RAA, HBp, HBd and RV). (Adapted with permission from McClelland et al.⁵⁷)

catheters, energy sources, etc) are beyond the scope of this communication and simply referenced above.

Conclusion

Based upon experience it is evident that most of the preexcitation syndromes are the result of antegrade conduction over AV-AP that course across the right and left AV annuli and directly inserting into the myocardium. The preferred name for such connections is AV-AP which would suffice for routine use and avoid confusion by using multiple terminologies and the eponym Kent bundle should be reserved for historical purposes only.

It has also become clear that the so-called Mahaim tachycardia, that is, NVPs long accepted as variants of preexcitation are, in fact, also APs with atrial insertion site along the anterolateral margin of the tricuspid annulus, remote from the AV node and distal insertion site in the RBB and/or occasionally in close proximity to distal RBB. Such pathways exhibit slow antegrade unidirectional conduction (AV and no VA) exhibit minimal or no preexcitation on the baseline surface ECG and LBBB morphology during antidromic reentry. The preferred term for such anomalous

connections is AFP and the use of "nodoventricular" should be avoided and reserved for historical interest only. This can obviously change with more evidence. At this time the eponyms of James fibers and LGL syndrome seem clinically irrelevant due to no association with tachycardia and the ECG pattern when noted should simply be described.

In summary, besides the abovementioned anomalous conduction connections in the human heart, the presence of some more cannot be excluded. The electrophysiology community needs to continue to maintain an inquisitive approach to this fascinating subject.

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