Coexistence of Functional Kent and Mahaim-type Tracts in the Pre-excitation Syndrome

Demonstration by Catheter Techniques and Epicardial Mapping

By Andrew M. Tonkin, M.R.A.C.P., Fortune A. Dugan, M.D.,
Robert H. Svenson, M.D., Will C. Sealy, M.D.,
Andrew G. Wallace, M.D., and John J. Gallagher, M.D.

SUMMARY
An unusual patient with Ebstein’s anomaly of the tricuspid valve and the Wolff-Parkinson-White syndrome was recognized for many years. Elucidation of the underlying mechanisms has recently been greatly enhanced by preoperative and intraoperative electrophysiological studies. It appears that most cases of classical Wolff-Parkinson-White syndrome are due to conduction over accessory (Kent) connections between the atrium and ventricle which bypass the atrioventricular node (AVN). Conduction over fibers coursing between the atrium and the lower part of the AVN (James tracts) is one of several postulated mechanisms for those cases in which tachyarrhythmias are associated with a short P-R interval and normal QRS complex (the Lown-Ganong-Levine syndrome). It has been proposed that AVN-ventricle or bundle of His-ventricle bypass (Mahaim fibers) may explain the rare forms of pre-excitation associated with a normal P-R interval.

The present case is reported because of the coexistence of Kent and Mahaim tracts. The Kent bundle was surgically divided with good result. The value of His bundle pacing in the recognition of Mahaim connections is illustrated.

Case Report
J. W. is a 20-year-old male college student with a history of paroxysmal tachycardias from early childhood. He has been otherwise asymptomatic. Wolff-Parkinson-White syndrome (type B) was first diagnosed in 1970 after a tachyarrhythmia culminated in syncope. After cardioversion to sinus rhythm his electrocardiogram showed a P-R interval of 0.10 sec, a negative delta wave in leads II, III, and aVf, and a QRS duration of 0.13 sec (fig. 1). A systolic murmur and scratchy diastolic murmur were audible at the lower left sternal border and a clinical diagnosis of Ebstein’s anomaly of the tricuspid valve made. Despite digoxin, 0.25 mg, p.o. daily, propranolol hydrochloride, 10 mg, p.o., q.i.d., and quinidine sulphate, 200 mg, p.o., q.i.d., the patient continued to have arrhythmias every few weeks with which he experienced chest pain, dyspnea, and presyncope, and he was referred for further study.

Preoperative Electrophysiological Study
On January 28, 1974, an electrophysiological study was performed. In the postabsorptive, nonsedated state, catheters were positioned across the tricuspid valve (to record activity from the bundle of His) and in the right ventricle, coronary sinus (allowing registration of activity and stimulation of the left atrium) and lateral right atrium. In sinus rhythm at a cycle length of 800 msec, the recordings demonstrated electrical activity from the bundle of His occurring 30...
msec after the onset of the delta wave on the surface electrocardiogram (fig. 2), confirming the presence of pre-excitation. Decreasing cycle lengths during right atrial pacing increased the degree of pre-excitation, but this did not occur in left atrial pacing, suggesting that the accessory pathway was on the right side.

The antegrade effective refractory period of the accessory Kent connection (at a basic cycle length during right atrial pacing of 600 msec), determined by the extrastimulus method, was 260 msec. Atrial premature beats at coupling intervals less than 260 msec initiated tachycardias. These tachycardias were of two different types (fig. 3). One tachycardia had a cycle length of 370 msec, with normal intraventricular conduction (QRS duration: 0.08 sec) and a frontal plane axis of +100°. The second tachycardia had a cycle length of 410 msec, with ventricular complexes of left bundle branch block (LBBB) configuration (QRS duration: 0.15 sec), and frontal plane axis of −60°. The slowing of the cycle length during tachycardia with LBBB configuration initially suggested supraventricular tachycardia with aberration, with slowing due to participation of a left-sided accessory path. However, during tachycardia with LBBB configuration, the onset of ventricular activation on the surface electrocardiogram preceded His bundle activity by 35 msec. In contrast, during tachycardia associated with normal conduction, antegrade His deflections were noted with H-V of 50 msec. During both forms of tachycardia (and during right ventricular pacing) the earliest site of retrograde atrial activation was the lower lateral right atrium, followed successively by the medial right atrium (measured from the His bundle catheter) and the proximal coronary sinus. This eccentric sequence of retrograde atrial activation suggested that ventriculo-atrial conduction in both types of tachycardia was via an accessory pathway in the posterior right ventricular free wall. Both tachycardias could be terminated by appropriately timed single atrial and ventricular premature beats.

Pacing the bundle of His at a cycle length of 400 msec (approximating that of the tachycardia associated with LBBB configuration) with a stimulus–ventricular interval of 50 msec was associated with 1:1 normal intraventricular conduction (QRS duration: 70 msec). There was therefore no evidence of organic bundle branch disease. During right ventricular pacing at cycle lengths decreasing to 400 msec, the interval between the stimulus and atrial electrograms remained constant.

It was concluded that an anomalous Kent connection passed between the right atrium and right ventricle. The etiology of the LBBB tachycardia was less certain, but three possibilities were entertained: 1) a re-entrant rhythm utilizing Mahaim fibers passing...
from the AVN to the region of the right bundle branch system, 2) antegrade conduction over a second Kent connection, or 3) ventricular tachycardia.

Intraoperative Electrophysiological Study

Because of the patient's previous failure to respond to antiarrhythmic agents, he underwent cardiac surgery on February 4, 1974. At operation, bipolar and unipolar potentials were recorded from the epicardium of the exposed heart. During pacing of the right atrial appendage, the earliest area of ventricular activation was the posterior surface of the right ventricle approximately 2 cm to the right of the posterior descending artery (fig. 4). Activation spread radially from this site and was recorded 120 msec later in the normal site of epicardial breakthrough in the area trabecularis on the anterior paraseptal surface of the right ventricle.10 During episodes of supraventricular tachycardia, the earliest area of retrograde atrial activation was also posterior and to the right of the septum, adjacent to the A-V ring. Atrial mapping during right ventricular epicardial pacing showed that this site of earliest atrial activation (70 msec later than the adjacent ventricular epicardium) was located opposite the point of breakthrough on the ventricles in sinus rhythm (fig. 5).

The patient was then placed on cardiopulmonary bypass. After a right atriotomy, mild Ebstein's malformation of the tricuspid valve was noted. A 3 cm incision, centered about the earliest areas of epicardial activation, was then made in the annulus of the tricuspid valve (fig. 6). The patient was then defibrillated. In

**Figure 4**
Intraoperative epicardial activation data during pacing of the right atrial appendage. All values are corrected to the onset of the delta wave and Q wave during anomalous and normal conduction, respectively. Before division of the accessory Kent bundle (upper panel), epicardial breakthrough occurred at the base of the right ventricle, 22 msec before the onset of the delta wave. After division of the accessory path, normal breakthrough occurred on the anterior right ventricle, 29 msec after the QRS onset (lower panel). ⊙ = site of pacing electrode.
sinus rhythm, the P-R interval was 0.22 sec, the ventricular complexes in V1 had an RSR' morphology, and pre-excitation was no longer evident. Epicardial mapping during right atrial pacing demonstrated a normal ventricular activation sequence (fig. 4). Prior to closure of the chest, the patient had a paroxysmal tachycardia at a cycle length of 470 msec with LBBB configuration identical to that previously noted (fig. 7). Epicardial mapping demonstrated breakthrough 30 msec after the onset of the QRS near the area trabecularis of the mid-anterior right ventricle. This effectively excluded antegrade conduction over a second Kent bundle as the etiology of the QRS aberration. In addition, approximately 40 msec delay in propagation over the left ventricle (as compared to the activation in sinus rhythm after division of the Kent bundle) was noted, consistent with the surface electrocardiographic features of LBBB. Simultaneous occurrence of atrial and ventricular activity suggested that the tachycardia may have originated in the A-V junction.

The postoperative course was complicated by the postcardiotomy syndrome. With this, an arrhythmia of the same cycle length and ventricular activation pattern as the operative tachycardia was noted, but with additional evidence of A-V dissociation. This tachycardia too was thought to originate in the A-V junction, but with retrograde conduction block (in contrast to the intraoperative arrhythmia). The patient has had no further evidence of tachyarrhythmias during follow-up.

Postoperative Electrophysiological Study

A further electrophysiological study was performed on July 22, 1974. At this time, the surface electrocardiogram still showed no evidence of pre-excitation. In sinus rhythm, with a cycle length of 810 msec, the A-H and H-V intervals were 140 msec and 55 msec respectively, and a normal QRS morphology was present. At a cycle length in sinus rhythm equal to or less than 720 msec, the conduction pattern changed. An LBBB pattern became evident on the surface electrocardiogram, the onset of the QRS complex preceding His bundle activity. Fusion beats of intermediate QRS morphology and with variable H-V intervals were observed during left atrial pacing at a
cycle length of 750 msec (fig. 8). As the cycle length of both right and left atrial pacing decreased from 700 to 320 msec, 1:1 antegrade atioventricular conduction was sustained, the QRS complexes retaining LBBB configuration and His bundle activity following the QRS onset. As the cycle length of atrial pacing decreased over this range, the interval between the stimulus and the onset of ventricular activation lengthened from 285 msec to 405 msec. The fact that the conduction pattern of the arrhythmia was identical to the preoperative LBBB tachycardia during sinus rhythm and atrial pacing at comparable cycle length suggested that supraventricular tachycardia was much more likely as an explanation of the preoperative LBBB arrhythmia than right ventricular tachycardia.

Pacing from the region of the bundle of His at cycle lengths of 600, 500, and 400 msec was also associated with 1:1 conduction. However, as in the preoperative study, His bundle pacing (stimulus-V interval of 50 msec) was associated with normal intraventricular conduction with no evidence of LBBB (fig. 9). In contrast to the preoperative study, there was no evidence of retrograde His activity nor retrograde V-A conduction during pacing from the right ventricular apex at cycle lengths from 700 to 300 msec. Arrhythmias could not be induced by scanning diastole with premature beats during pacing of either the right atrium or right ventricle. The findings in this postoperative study could be totally explained in terms of conduction over either the normal pathway (normal conduction pattern) or Mahaim fibers passing from the AVN to the right bundle branch system (LBBB pattern) or both (ventricular fusion).

**Discussion**

Surgical therapy is unnecessary for the majority of patients with Wolff-Parkinson-White syndrome. However, should surgery be considered, electrophysiological evaluation of the mechanism producing preexcitation is a necessary prelude. It is important in this preoperative evaluation to recognize multiple accessory pathways, should they exist. In this patient, a tentative preoperative diagnosis of two anomalous conduction paths was made, both of which apparently participated in re-entrant arrhythmias.

**Figure 7**

Epicardial map during tachycardia (cycle length 470 msec) noted during surgery. This arrhythmia occurred after division of the Kent bundle by incision in the A-V ring.
The following observations suggested that an A-V bypass of the Kent type passed between the right atrium and right ventricle:

1) The onset of ventricular activation preceded the recording of electrical activity from the bundle of His.15 The degree of this pre-excitation was selectively increased by right, but not left atrial pacing.2

2) During both forms of supraventricular tachycardia and during right ventricular pacing, the earliest retrograde intracavitary electrogram was registered from an abnormal site in the lateral right atrium rather than the medial right atrium or coronary sinus.

3) Preoperatively, the relative constancy of ventriculo-atrial conduction time during right ventricular pacing also suggested that retrograde conduction was over an accessory pathway rather than through the A-V node.13 This preoperative diagnosis of a right-sided Kent connection was substantiated by the observations during epicardial mapping, and the abolition of pre-excitation and the absence of postoperative arrhythmias (other than tachycardias possibly originating in the A-V junction in the immediate perioperative period) after surgical incision at the site appear to definitely prove its location. This location of the accessory Kent bundle concurs with the previous findings in patients with Wolff-Parkinson-White syndrome who had a similar electrocardiographic pattern.13-15 Division of this Kent bundle was undertaken because the retrograde atrial activation sequence suggested its participation in both forms of preoperative tachycardia.

Even after surgical division of the Kent bundle, during sinus rhythm or atrial pacing at cycle lengths less than 700 msec, electrical activity from the bundle of His was displaced and appeared within QRS complexes which had LBBB configuration associated with a long P-R interval. Additionally, ventricular fusion beats of intermediate morphology (between that of normal intraventricular conduction and LBBB) and with variable H-V intervals were observed. These two observations excluded rate-related LBBB and strongly suggested the presence of a second accessory pathway. Slow conduction through a second Kent bundle16 ("slow" because of the long P-R interval in sinus rhythm) could be excluded as a classical LBBB pattern but no delta wave was observed, and the intraoperative map during tachycardia with LBBB configuration demonstrated no pre-excitation at the A-V groove. This latter finding has invariably been present with pre-excitation due to Kent bundles; even 11 septal accessory pathways mapped to date at this institution demonstrated pre-excitation at the A-V groove. The normalization of ventricular complexes with His bundle pacing at cycle lengths associated with LBBB during atrial pacing excludes a His-right ventricle Mahaim connection. The observations appear fully explicable by the existence of a Mahaim-type connection coursing from the lower end of the AVN to the region of the right bundle branch bypassing the normal His-Purkinje system. We believe that the absence of a delta wave with conduction over these Mahaim fibers can be explained by the presence of fibers that insert into the right bundle branch system rather than the ventricular myocardium.

Mahaim and Winston17 originally described fibers connecting the penetrating portion of the bundle of His and the upper part of the ventricular septum. These fibers, present in normal man, were invoked in paraspecific septal conduction. Since their original description, the term "Mahaim fibers" has been extended to include fibers passing from the lower part of the AVN to the adjacent ventricular myocardium.15, 19 Clues to conduction over Mahaim fibers have been said to include the presence of a normal or prolonged P-R interval with a delta wave5, 6 and the retention of pre-excitation with beats of A-V junctional origin6, 18 or after the development of disturbances of normal A-V conduction.18 It is also possible that some cases of apparent "supernormal" conduction in an anomalous
Kent bundle may in fact be due to preferential conduction via Mahaim fibers passing from one of two upper intranodal pathways. Failure to increase the degree of pre-excitation by atrial stimulation has been cited as another clue to conduction over Mahaim fibers. This may be due instead to total pre-excitation initially (and lack of fusion with conduction spreading from the normal path) or to failure to provide optimal input into a Kent bundle because stimulation occurs at a distance from its location. In fact, very few cases of Mahaim conduction have been reported. A detailed anatomic study has documented coexistent James and numerous Mahaim fibers passing to the basal right ventricular septum in a patient with classic WPW type B, and the presence of Mahaim fibers has been postulated in other reports. However, in none of these cases has Mahaim-type conduction been definitely proved by electrophysiological methods, including His bundle pacing.

This present case illustrates the special importance of pacing the region of the bundle of His to aid in determining the participation of Mahaim fibers. This has been reported once previously. In Coumel's case, demonstration of pre-excitation during electrical stimulation of the His bundle was regarded as evidence that the point of take-off of the Mahaim bypass was at or below this level. This is the first report of normalization of intraventricular conduction during pacing of the His bundle as evidence for conduction over fibers emanating from the AVN.

The ability preoperatively to initiate and terminate tachycardias by a single premature beat suggested re-entry (after functional dissociation of conduction pathways) as the underlying mechanism. The atrial activation sequence suggested that retrograde conduction during the two re-entrant tachycardias was always over the Kent bundle. The retrograde conduction properties of this bundle appeared to be such that ventriculo-atrial conduction could be sustained at rates comparable to those of the two forms of atrial tachycardia. Antegrade conduction during tachycardia apparently could occur over two different paths (fig. 10). One path was that of the normal atrioventricular conduction via the AVN and His-Purkinje system.

Figure 10

Schematic drawing of proposed pathways of circus movement tachycardias. The left panel shows the tachycardia with normal intraventricular conduction (NORMAL). Antegrade and retrograde conduction were, respectively, via the His-Purkinje system (HPS) and right-sided Kent connection (K). The middle panel shows the proposed pathway for the preoperative tachycardia with ventricular complexes of LBBB-type (LBBB). Antegrade conduction was thought to be via Mahaim fibers (M) passing from the atrioventricular node (AVN) to the region of the right bundle branch (RBB) and retrograde conduction over the Kent bundle. The right panel shows the possible mechanism for the LBBB-type tachycardia after division of the Kent bundle, with antegrade and retrograde conduction via the Mahaim fibers and His-Purkinje system, respectively.
system (tachycardia with normal intraventricular conduction). An alternate path was via the AVN, then Mahaim fibers to the right bundle branch system (tachycardia with LBBB). The onset of ventricular depolarization preceded the His bundle deflection in the latter mechanism presumably because the take-off of the Mahaim connection allowed partial bypass of the area of AVN delay. Preoperatively, tachycardias were therefore induced when an atrial premature beat was blocked in the Kent bundle but conducted slowly through either or both of the other two paths. A third possible mechanism of tachycardia was a re-entry loop with antegrade conduction via the Mahaim fibers and retrograde conduction via the His-Purkinje system. The intraoperative and postoperative tachycardias may have been due to this mechanism.

The potential for reciprocation involving the normal and Mahaim tracts would appear to exist postoperatively. Despite this theoretical possibility, no such arrhythmias have been manifested clinically since the perioperative period, nor could they be induced by the extrastimulus technique. Possible explanations for the absence of arrhythmias in the presence of an accessory pathway include similar conduction properties of the paths or alteration of the functional properties of the Mahaim path by operative damage.

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