The Wolff-Parkinson-White Syndrome

Problems in Evaluation and Surgical Therapy

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SUMMARY
Two patients with WPW syndrome underwent surgery to ablate accessory conduction pathways. Endocardial and epicardial mapping in both patients had indicated an area of early right ventricular depolarization. Surgical transection of the areas of early depolarization failed in both cases to normalize the electrocardiogram. In the first patient, additional resection in the area of the A-V node failed to produce heart block and the ECG remained abnormal. However, the paroxysmal tachycardia ceased, and she has remained asymptomatic and active 12 months after surgery. In the second patient, as the A-V node was about to be sectioned, pressure and procaine near the A-V node caused the ECG to normalize transiently and after resection permanently. Microscopic study of this tissue showed "P cells." Postoperatively the patient demonstrated normal A-V nodal function. He was discharged with a normal ECG but expired soon after discharge. Postmortem examination of the heart demonstrated the A-V node and bundle of His plus the location of the resection adjacent to the bundle of His. These two cases illustrate disparities between electrophysiologic mapping and actual site of the accessory conduction pathway. In one of the cases an accessory bundle was demonstrated histologically.

Additional Indexing Words:
Accessory bundle
Epicardial mapping
Surgical transection
Endocardial mapping

Episodes of supraventricular tachycardia associated with the Wolff-Parkinson-White syndrome on occasion, may severely limit the patient's activity, produce profound hemodynamic effects, and rarely may be life threatening. Recent reports by Burchell and colleagues and Cobb and colleagues have indicated that surgical interruption of anomalous conduction pathways between the atrium and ventricle eliminates the delta wave, may prevent retrograde conduction from the ventricle to the atrium, and eliminates the episodes of supraventricular tachycardia. These reports plus the data of Durrer and Roos and Roos and associates also suggest that the location of the anomalous conduction pathway could be identified by epicardial mapping prior to surgical intervention. This report describes the diagnostic studies and surgical therapy of two patients with the Wolff-Parkinson-White syndrome and incapacitating supraventricular tachycardia refractory to medical therapy.

Methods
Because a single, precise location for the aberrant conduction pathway has not been established and since it is frequently difficult to classify the type of WPW (A or B), it is essential that a method be available for accurately
localizing the area or areas of aberrant conduction in a specific patient prior to undertaking any surgical procedure.

Since neither patient was typically type A or B as described by Rosenbaum and co-workers,8 the initial step was to try to identify an area of early ventricular depolarization by transvenous endocardial mapping of the right ventricle. In both patients, a Zucker unipolar multipurpose catheter-tip electrode referred to a Wilson's central terminal was passed into the right atrium and right ventricle under fluoroscopic control. Normal sinus rhythm and aberrant conduction were present throughout both procedures. Approximately 20 tracings systematically obtained from representative areas in the right atrium and right ventricle were compared to a simultaneously recorded external reference lead. Tracings were recorded on a Honeywell 1508 visicorder at 100 to 200 mm/sec paper speed. Frequency response of the recording system was 0.1 to 3,000 Hz.

Previous methods used to follow electrical activity over the endocardial or epicardial surface of the heart have related the depolarization wave (that is, major intrinsicoid deflection) obtained from the exploring electrode to a fixed point on an independent reference lead.4-6 In this study, the interval between the onset of the intrinsicoid deflection in the exploring lead and the peak of the R wave in the reference lead was measured by the unaided eye to the nearest 5 msec. With this method of tracing analysis, an increase in measured interval indicates an earlier time of depolarization while a short interval corresponds with late depolarization. At the suspected location of aberrant conduction, the exploring intrinsicoid deflection corresponded to the delta wave in the reference lead.

It has been assumed that an area of early depolarization corresponded to an aberrant accelerated conduction pathway. Durrer9 has described isochronic lines over the endocardial surface of the right ventricle and indicated that the area of earliest electrical excitation normally

![Figure 1](https://example.com/image)

**Figure 1**
Standard 12-lead electrocardiogram before and after surgery. (Upper panel) Case 1. No change occurred in the P-R interval, QRS complex, or axis after surgery. (Lower panel) Case 2. The P-R interval increased to 0.16 sec, the QRS decreased to 0.08 sec, the delta wave was eliminated, and the axis shifted 60° to the right after surgery.
WPW SYNDROME

**Figure 2**
Results of endocardial mapping in case 1 (M.F.). Twenty recordings were obtained throughout the right atrium, ventricle, and pulmonary outflow tract. The cardiac silhouette on the left contains labeled selected catheter positions marked 1 to 4. The corresponding endocardial tracings are seen on the right. The solid vertical lines originate from the peak of the R wave in reference lead aV1. The dotted line indicates the onset of the isochronic deflection in the exploring lead. The time between the solid and dotted lines is listed in milliseconds. The longer period indicates earlier ventricular depolarization. The earliest ventricular depolarization in this patient was area 3.

 occurs over the lower one third of the interventricular septum followed by a progressive spread of excitation over the endocardial surface up to the atrioventricular ring. We felt that endocardial mapping utilizing an exploring endocardial lead and external reference lead similar to that previously described for epicardial mapping might enable us to demonstrate an interruption in this isochronic line immediately below the tricuspid annulus and thereby localize the area of aberrant conduction.

After an area of early ventricular depolarization was located by endocardial mapping, the patients underwent thoracotomy and epicardial mapping. A grid was established over the right atrium and ventricle, marking off areas approximately 1 cm square. A tracing was then obtained from each of approximately 40 squares, utilizing a unipolar electrode referred to a Wilson's central terminal. The technic of tracing analysis was the same as that used for endocardial mapping. The interval between the onset of the isochronic deflection in the epicardial tracing and the peak of the R wave in the simultaneously recorded reference electrocardiogram was measured utilizing a straight edge and the unaided eye. This was done on line in the operating room. When the location of the early ventricular depolarization was confirmed by epicardial mapping, the region was sectioned; the incisions utilized extended through the entire thickness of the ventricular and atrial walls.

**Figure 3**
Results of epicardial mapping in case 1 (M.F.). Forty recordings were obtained from the pericardial surfaces of the right atrium and right ventricle. On the left, the drawing of the heart with a few anatomic landmarks contains numbers which refer to representative tracings on the right. The two dashed lines indicate the extent of the transatrial and ventricular incisions and their relationship to the right coronary artery. The representative epicardial tracings on the right have the same reference system as that previously described in figure 2. The earliest ventricular depolarization occurred in area 4. This corresponds well with the data from endocardial mapping.

**Report of Cases**

**Case 1**

M.F., a 50-year-old white woman, had experienced progressive increase in the frequency of episodes of paroxysmal atrial tachycardia for the past 9 years. She had been hospitalized 17 times in the past 7 years and six times in the past 3 months. The latter episodes were extremely refractory to extensive drug therapy, including digitalis, quinidine, procainamide (Pronestyl), reserpine, propranolol, and atrial or ventricular pacing. In fact she developed atrial tachycardia while receiving quinidine intravenously, 100 mg an hour. The only significant physical findings were moderate kyphoscoliosis and a prominent left ventricular impulse located in the fifth and sixth intercostal spaces. Results of routine laboratory studies and chest x-rays were normal. The resting electrocardiogram demonstrated the
Wolff-Parkinson-White pattern (fig. 1). Repeated episodes of atrial tachycardia were documented. Endocardial mapping demonstrated an area of early depolarization along the lateral wall of the right ventricle immediately below the tricuspid annulus (fig. 2). The onset of the intrinsicoid deflection from the exploring electrode in this area occurred 75 msec before the reference R wave while the intrinsicoid deflection obtained from the lower interventricular septum preceded the reference R wave by 50 msec, and the intrinsicoid deflection obtained in pulmonary outflow preceded the reference R wave by only 20 msec.

Epicardial mapping revealed an area of early depolarization similar to that obtained with endocardial mapping. Figure 3, using representative areas, demonstrates the pattern of depolarization obtained over the anterior surface of the right ventricle. The onset of the intrinsicoid deflection from the exploring electrode over the lateral margin of the right ventricle occurred 85 and 100 msec before the reference R wave and occurred simultaneously with the delta wave in the reference tracing, while the intrinsicoid deflection preceded the reference R wave by only 55 msec over the more medial area of the right ventricle.

Light fingertip pressure over the area of early depolarization always interrupted the episodes of nodal tachycardia, which occurred during surgery (fig. 4). After completing the epicardial mapping and demonstrating an area of early depolarization, the patient was placed on cardiopulmonary bypass.

A 1-cm incision was made through the entire thickness of the right ventricular wall in the area of early depolarization immediately below and parallel to the A-V groove. When no electrocardiographic changes were noted, the incision was extended laterally past the right heart margin to the diaphragmatic surface of the right ventricle. It was also extended medially to the region of the pulmonary outflow tract. The incision that separated the right ventricle from the tricuspid valve annulus was 8 cm long. A similar incision was made on the atrial side of the A-V groove. Figure 3 demonstrates the extent of these incisions. The form of the electrocardiogram remained unchanged.

An attempt was then made to produce complete heart block by surgically interrupting the A-V node. A scalpel was used to incise the endocardium and myocardium of the atrial septum approximately 3 to 5 mm above the tricuspid annulus. This incision began at the coronary sinus and passed forward, through, and beyond the site where the A-V node and bundle are usually located. The incision was 2 cm long. The QRS remained unchanged as did the heart rate. Atrial pacing provided a 1:1 response indicating that A-V block had not been accomplished (fig. 5). However, the frequent episodes of atrial and nodal tachycardia noted prior to surgery and during cardiopulmonary bypass ceased. The atrial and ventricular incisions were closed, and perfusion was discontinued.

Postoperative Course

Atrial pacing at a rate of 100 beats/min was required to maintain an adequate cardiac output for the initial 6 postoperative days. After the sixth day, the pacing was stopped and the patient remained in regular sinus rhythm and her original...
WPW SYNDROME

Figure 5

Atrial pacing in case 1 after incisions through suspected aberrant conduction pathway and A-V node. Note 1:1 ventricular response with variable rate of atrial pacing. Vertical time lines = 100 msec. Two nodal escape beats with normal conduction are noted on discontinuing atrial pacing. The normal conduction in these two beats is identical to normal conduction when in nodal tachycardia as noted in figure 4.

WPW pattern (fig. 1). Physical activity was gradually increased to a normal daily routine, and she was discharged on the 15th postoperative day. Twelve months postoperatively she has experienced no episodes of tachycardia and is fully active, although retaining her original WPW pattern.

Comment

Since the delta wave persisted after an extensive surgical incision on both the atrial and ventricular sides of right lateral A-V groove, epicardial and endocardial mapping, utilizing a unipolar exploring electrode and external reference electrocardiogram lead, had not precisely located the aberrant conduction pathway. Ablation of the supraventricular tachycardia by applying light pressure over the right lateral margin of the A-V groove prior to surgical incision suggests that this area did contain an aberrant pathway which was at least in part responsible for retrograde conduction and resulting tachycardia. However, the persistence of the delta wave and documented conduction from the right atrium to the ventricle after extensive surgical incisions along the right A-V groove and through the A-V node indicate that another aberrant pathway must be located in the septum or left heart. Despite not interrupting all of the aberrant pathways, the patient has not experienced any episodes of tachycardia for 12 months, and this observation suggests that the circus movement responsible for her episodes of tachycardia has been interrupted.

Case 2

R.P., a 45-year-old white male, had noted episodes of tachycardia since age 8 years. During the past year, he had experienced daily episodes of tachycardia and had required five hospitalizations. On two of these occasions, he had developed severe pulmonary edema, hypotension, and substernal chest pain. Medical therapy was completely ineffective, and frequent electrical

Figure 6

Results of endocardial mapping in case 2 (R.P.). Twenty recordings were obtained throughout the right atrium, ventricle, and pulmonary outflow tract. The cardiac silhouette on the left contains labeled representative catheter positions marked 1 to 4. The corresponding endocardial tracings are on the right and have the same reference system as previously described. The earliest ventricular depolarization in this patient is in area 2.
Results of epicardial mapping in case 2 (R.P.). Forty recordings were obtained from the epicardial surface of the right atrium and right ventricle. On the left, the drawing of the heart contains numbers which refer to the location of representative tracings on the right. The two dashed lines indicate the extent of the transatrial and ventricular incisions and their relationships to the right coronary artery. Representative epicardial tracings on the right have the same reference system as previously described. The earliest ventricular depolarization occurred in area 2 and corresponded well with data from endocardial mapping.

cardiopulmonary bypass. A 1-cm incision was made through the right ventricular wall in the area of early depolarization. When no electrocardiographic changes were noted, it was extended medially to the pulmonary outflow tract and laterally to the acute margin of the right ventricle. A parallel incision was made in the right atrium, thus separating the tricuspid annulus from the atrium

The initial contact of the exploring electrode to the area of earliest depolarization.

Figure 7 using representative tracings demonstrates the epicardial tracing obtained at the time of thoracotomy. Here the earliest intrinsicoid deflection occurred over the medial part of the right ventricle; it preceded the reference R wave by 85 msec and occurred simultaneously with the delta wave on the reference tracing. As the exploring electrode was moved even more medially up the pulmonary outflow tract and laterally along the right A-V groove, the interval between the intrinsicoid deflection and reference R wave decreased to 45 msec.

After completion of the epicardial mapping and demonstration of an area of early depolarization, the patient was placed on cardiopulmonary bypass. A 1-cm incision was made through the right ventricular wall in the area of early depolarization. When no electrocardiographic changes were noted, it was extended medially to the pulmonary outflow tract and laterally to the acute margin of the right ventricle. A parallel incision was made in the right atrium, thus separating the tricuspid annulus from the atrium

The diagrammatic view of the interior of the right atrium in case 2 demonstrates lateral view prior to opening the right atrium. Numbers indicate the following: (1) The wheal of lidocaine that converted the patient’s WPW complexes to NSR with the rectangular initial biopsy site. (2) The final biopsy site that permanently converted the WPW complexes to NSR. (3) Suture used to approximate the biopsy site. (4) A suture placed at this point produced complete heart block after normal sinus rhythm had resulted from the biopsy at 2. (5) The tricuspid valve. Note the straight line passing through this area as it illustrates the place of the microscopic sections taken at autopsy.
WPW SYNDROME

Injection of procaine just anterior to the site where the A-V node is normally located produced a normal QRS complex; accidentally the aberrant conducting pathway was found.

A 2 by 4-mm fragment of endocardium and underlying atrial septum was excised from this area. Since the procaine was still active, we were uncertain whether the excised fragment contained the aberrant conduction tissue and closure of the atrium was delayed. Fifty minutes later the electrocardiogram reverted to its abnormal configuration. Additional small fragments of tissue were then excised in the area of the procaine wheal that had originally produced a normal QRS complex. As a fragment was excised from area 2 as shown in figure 8, the electrocardiogram again became normal. These excision sites were closed with interrupted sutures. The first suture approximated the tissue satisfactorily. The second suture produced complete heart block. It was removed and the electrocardiogram reverted to normal sinus rhythm (NSR). No additional sutures were used. The atrial and ventricular incisions were closed. Cardiopulmonary bypass was discontinued.

Postoperative Course

In the initial 5 days following surgery, the patient was alternately in regular sinus rhythm and atrial fibrillation. His postoperative atrial fibrillation was managed in a routine way with digitalis therapy and was responsive to carotid sinus massage. This was not the case preoperatively. The QRS complex remained normal, and he experienced no documented episodes of atrial tachycardia. He was discharged on the 21st postoperative day. Twenty-four hours after discharge, lethargy, somnolence, and seizures necessitating hospitalization, developed. Neurosurgical evaluation and craniotomy revealed several large cerebral infarctions, and the patient expired.

Pathology

Autopsy revealed extensive, hemorrhagic infarction of the right cerebral hemisphere with marked cerebral swelling, uncal herniation, and pontine hemorrhage. In addition there were small, acute renal and splenic infarctions. Thromboembolic material was identified in the renal artery at the base of the infarct, but no thromboembolic material could be identified in any of the cerebral vessels. The heart at autopsy was free of thrombi and no source for emboli could be identified.

Sections taken from the right atrium and atrial and ventricular septa demonstrated the normal A-V node and His bundle. Figure 9 shows a microscopic section demonstrating the relationship of the A-V bundle and the incision which normalized the electrocardiogram. The line in

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**Figure 9**

Section of the interatrial septum of the heart removed at autopsy in case 2. The plane of the section is indicated by the straight line in figure 8. M and T identify the mitral and tricuspid valves, respectively. Arrow 1 identifies the suture used to approximate the biopsy site. Arrow 2 points to scar tissue that marks the site of biopsy that converted the patient's WPW complexes to NSR (figure 8-2). The five arrows point to the normal common bundle. Gomori's trichrome stain; low-power magnification, × 5.5.

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Figure 10

A portion of a single myocardial cell from the aberrant conduction pathway is illustrated in this electron micrograph, and its cell membrane is indicated by arrows. This cell differs from ordinary working myocardium in that the mitochondria (M) are randomly scattered in a loose granular matrix of glycogen (G), and the myofilaments are sparse. Tissue fixation was performed in the operating room, probably accounting for the good preservation of cell organelles. × 11,700.
WPW SYNDROME

figure 8 provides orientation of this section. The large arrow 2 indicates an area of scarring from the region of the biopsy which produced the normal electrocardiogram. The A-V bundle is located immediately below this biopsy site in the area where the placing of a suture at the time of surgery resulted in complete heart block.

The electron micrograph of the tissue, taken from the biopsy which normalized the electrocardiogram (fig. 10) demonstrates one of several cells which differ from ordinary working myocardium. The cytoplasm of these cells contains sparse filaments and randomly arranged mitochondria in a matrix of increased glycogen. Ultrastructural study of cells of the human sinus node and human atrioventricular node has demonstrated that these areas have cell populations, which differ from ordinary working myocardium.\textsuperscript{10, 11} The small, round, pale cells with randomly distributed mitochondria and sparse myofilaments in these areas have been designated as "P cells." Although it has been postulated that the "P cells" are the site of pacemaking activity in the sinus and A-V node, the question remains "Are pacemaking cells different from conducting cells?"\textsuperscript{12} The cells identified in the biopsy from this case resemble "P cells," but the evidence indicates that the biopsy site was outside of the A-V node, implying that this was conducting rather than pacemaking tissue. The abnormal conduction tissue would appear to lie toward the right atrium and toward the endocardium from the common bundle. The A-V node was not at the level seen in figure 9, but lay a few millimeters toward the coronary sinus ostium. No evidence of scar tissue indicative of surgical manipulation was present in the A-V node.

Comment

Epicardial and endocardial mapping did not precisely locate the pathway of aberrant conduction. The mapping procedures defined an area of early depolarization over the anterior surface of the right ventricle immediately below the A-V groove and to the right of the septum while in fact an aberrant pathway was defined histologically, in the atrial septum, 3 to 5 mm anterior to the A-V node and common bundle. The electrophysiological significance of this aberrant pathway was documented by the loss of the delta wave following its interruption. It can be speculated, however, that even though epicardial mapping did not precisely localize the aberrant pathway, the pattern of epicardial depolarization did reflect the location of the aberrant fiber. A conical spread of depolarization upward from the aberrant pathway through the septum to the epicardial surface of the right ventricle and then out across the surface of the right ventricle and up onto the surface of the pulmonary outflow tract would produce a pattern of depolarization similar to the one observed in this case.

This case emphasizes the difficulty in locating conduction pathways using surface mapping and also documents an aberrant pathway located in the septum near the A-V node.

Discussion

Aberrant conduction pathways between the right atrium and right ventricle have been demonstrated both anatomically\textsuperscript{2, 12-17} and electrically\textsuperscript{4-6} in the WPW syndrome. A circus movement between the right atrium and right ventricle through the normal A-V node and aberrant conduction pathway has been postulated as the mechanism responsible for episodes of atrial tachycardia in this syndrome.\textsuperscript{18, 19} If this is true, interruption of the circle, either at the A-V node or through the aberrant conduction pathways should prevent the circus movement and eliminate the episodes of refractory atrial tachycardia. In addition, if an isolated aberrant conduction bundle is a prerequisite for this syndrome and responsible for early depolarization of the upper part of the right ventricle, interrupting it should ablate the delta wave in addition to eliminating refractory atrial tachycardia.

Recently, two different surgical approaches have been utilized in an attempt to prevent intractable episodes of atrial tachycardia in the WPW syndrome. Dreifus and his colleagues\textsuperscript{10} reported the control of recurrent tachycardia in one patient following surgical ligation of the A-V bundle. Although the delta wave persisted because the aberrant pathway remained intact, atrial tachycardia was eliminated, presumably because the circus movement had been interrupted. Following the initial but transient success of Burchell and associates,\textsuperscript{4} Cobb and his colleagues\textsuperscript{5} reported the successful surgical interruption of the
bundle of Kent in a patient with type B WPW syndrome. Following a 5 to 6-cm incision through the anterior surface of the right ventricular wall immediately below and parallel to the A-V ring, the delta wave disappeared, and the P-R interval and QRS duration returned to normal. Eight weeks after surgery, the electrocardiogram remained normal, and they reported no episodes of tachycardia.

Epicardial mapping was used by both Burchell and associates and Cobb and colleagues to locate the aberrant pathway. Since the final incision reported by Cobb's group was 5 to 6 cm long, it seems unlikely that the precise location of the aberrant pathway had been defined by epicardial mapping. In the case reported by Burchell and associates, procaine was injected into the area which epicardial mapping had indicated as the location of aberrant conduction with immediate normalizing of the electrocardiogram. This was followed by a 1-cm incision in the atrium above this area. However, shortly after surgery, the abnormal WPW pattern returned, suggesting that epicardial mapping had not been precise in localizing the aberrant conduction pathway to a 1-cm area.

There may be several reasons why epicardial mapping has not proven entirely satisfactory. Epicardial fat, in some instances, may prevent the obtaining of action potentials directly from the myocardium. In addition, the exploring electrode measures only the depolarization wave as it moves across the epicardial surface, and the recorded action potentials may reflect electrical activity some distance away from the electrode rather than directly under the electrode as illustrated in case 2. Thirdly, even though an area of aberrant conduction should be localized by this technic, there is no reason to assume that it is the only aberrant pathway in that specific patient. If this is true, it may be extremely difficult to get accurate data concerning the location and function of areas of aberrant conduction from either endocardial or epicardial mapping. Final evaluation of this technic must await more case experience.

It is apparent that a more thorough search for the location of these pathways must be developed before a surgical response can be accurately predicted. Although endocardial mapping as described in this report is a very crude technic, the close correlations between it and epicardial mapping in cases 1 and 2 suggest that it may provide some useful information in typing cases preoperatively, since theoretically type A WPW syndrome should be located in the posterior wall of the left ventricle and type B in the lateral wall of the right ventricle. Studies in and around the A-V node are indicated in those cases which are neither type A nor B.

The most pertinent observation in this report may be that neither patient experienced episodes of refractory atrial tachycardia following surgery suggesting that retrograde conduction following premature ventricular contractions and the subsequent circus movement had been interrupted. In case 1, this must have occurred at the A-V node since the aberrant pathway persisted after surgery. In case 2, the interruption occurred through the aberrant pathway located in the septum but outside the normal A-V node since the delta wave was eliminated and normal A-V nodal function was preserved.

If one is faced with a critically ill patient and is unable to locate any aberrant pathway, transection of the A-V node as proposed by Dreifus and associates and done in case 1 is probably reasonable. However, if there is more than one aberrant pathway in that specific patient, the potential for circus movement between the atrium and ventricle still exists, and refractory tachycardias may not be eliminated.

Another potential postoperative complication in patients who have undergone transection of the A-V node is complete heart block. If, for any reason, the persistent aberrant pathway stops functioning, transmission of the atrial impulse to the ventricle will be interrupted with resulting heart block and possibly ventricular asystole. Edmonds and associates have recently documented this complication.
Although the precise anatomy and electrophysiology of the WPW syndrome is at present not clear, the response in the two cases described in this report indicates that surgical interruption of the circus movement either at the aberrant pathway or A-V node may be practical. It is also apparent that this type of experimental therapeutics can be justified only in the critically ill patient who is completely informed and only after thorough study.

References


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