Corrected Transposition with Severe Intracardiac Deformities with Wolff-Parkinson-White Syndrome in a Child

Electrophysiologic Investigation and Surgical Correction

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SUMMARY This is a report of a 10-year-old child who underwent surgery for complex congenital heart disease consisting of corrected transposition of the great vessels, ventricular septal defect, patent ductus arteriosus, severe left-sided atrioventricular (AV) valve insufficiency (Ebstein's deformity) and Wolff-Parkinson-White syndrome. The site of his accessory AV connection was localized preoperatively at a left anterolateral site by isopotential body surface maps and by intracardiac electrophysiologic studies. He successfully underwent surgery for closure of the ventricular septal defect, ligation of the patent ductus arteriosus, replacement of the left-sided AV valve, and interruption of the accessory AV pathway. Unavoidable complete AV block acquired at surgery required subsequent permanent pacemaker therapy.

CONGENITALLY CORRECTED transposition is an uncommon cardiac anomaly and the incidence of associated intracardiac defects is high; there is a reported association with the Wolff-Parkinson-White (WPW) syndrome. Recently, Kugler et al. suggested that surgery for congenital heart disease in the presence of WPW should include provisions for division of the accessory pathway due to the occurrence of debilitating tachycardia in the immediate postoperative period, which can be a fatal complication. We report a 10-year-old child with congenitally corrected transposition (LTGV) who also had a ventricular septal defect (VSD), patent ductus arteriosus (PDA), left-sided atrioventricular (AV) valve insufficiency and WPW. Recurrent bouts of supraventricular tachycardia (SVT) had occurred earlier in life, but later, recurrent atrial flutter-fibrillation and chronic heart failure required surgery. This report details the electrophysiologic evaluation and surgical management, which included closure of the VSD, ligation of the PDA, replacement of the left AV valve, and interruption of the left anterolateral accessory pathway.

Case Report

This 10-year-old boy was first seen at Duke University Medical Center in October 1978. At 2 months of age he had heart failure and underwent cardiac catheterization, which demonstrated LTGV, VSD and PDA. He also had WPW. In the first year of life he began to have SVT and this was moderately well controlled on digoxin and propranolol. However, he continued to have recurrent bouts of SVT and required frequent hospitalization.

He underwent cardiac catheterization again at age 6 years, with the additional findings of left AV valve insufficiency and pulmonary hypertension to systemic levels with elevated pulmonary vascular resistance (12 mm Hg/l/Min/m²). Because of the elevated pulmonary vascular resistance, surgery was considered inad-
visable. He continued to have recurrent bouts of SVT. Quinidine sulfate was added to his antiarrhythmic drug regimen, but the arrhythmia control was still unsatisfactory, and frequent hospitalization for cardioversion was still necessary.

At the time of his first visit to Duke Hospital he was doing poorly. He had 10-15 episodes of tachycardia per month and occasionally, they persisted for as long as 24 hours. He could not attend school. His medications included digoxin (0.125 mg twice a day), propranolol (12.5 mg four times a day), quinidine sulfate (50 mg four times a day), furosemide (40 mg twice a day) and potassium chloride. Physical examination showed him to be small for his age. The precordium was active. There was a grade III/VI harsh holosystolic murmur heard best at the apex, but it radiated well to the left lower sternal border. The second heart sound was loud and single. The chest x-ray is shown in figure 1. The ECG showed the classic features of WPW (fig. 2).

Cardiac Catheterization

The patient underwent a repeat cardiac catheterization in November 1978. The anatomic findings were similar to those obtained in 1974, i.e., they showed corrected transposition. Pulmonary artery hypertension was at systemic levels with elevated pulmonary vascular resistance (13.6 mm Hg/l/min/m²). Severe left AV valve insufficiency with an elevated pulmonary artery wedge pressure of 20 mm

![Figure 1](image1.png)

**Figure 1.** Preoperative chest roentgenogram. Note the characteristic features of corrected transposition including the general cardiac and left atrial enlargement, the gently sloping ascending aortic shadow in the upper left cardiac silhouette and the absent main pulmonary artery segment.

![Figure 2](image2.png)

**Figure 2.** Electrocardiogram showing preexcitation with a short PR interval, delta wave and prolongation of the QRS.
Hg and a systemic ventricular end-diastolic pressure of 15–20 mm Hg was present. Additionally, a PDA and a VSD were present with a moderate left-to-right shunt (Qp/Qs 1.88:1 by Fick method); the aortic saturation was 95% and the hemoglobin was 14.4 g%. During the catheterization, the patient had several episodes of atrial flutter-fibrillation documented by recording with an intracardiac electrode catheter; the QRS complexes were widened, and the shortest and average RR intervals were 210 msec and 320 msec, respectively. The episodes of atrial flutter-fibrillation, though short, produced hypotension and acute respiratory distress with coughing and production of frothy sputum. All episodes ceased spontaneously.

The presence of the two levels of cardiac shunting and the severe left AV valve insufficiency made it difficult to evaluate the calculated pulmonary vascular resistance. Since the values of the pulmonary vascular resistance calculations from the two catheterizations in 1974 and 1978 were essentially the same, we felt the patient had shown no evidence of progression of his pulmonary vascular obstructive disease. However, his clinical course had deteriorated markedly and on this basis, we felt he should have ligation of his PDA, closure of his VSD and replacement of the AV valve. The presence of WPW was a complicating feature not so much because of reentrant tachycardia involving the accessory pathway, but because of the rapid ventricular response during atrial flutter-fibrillation. We were concerned that this would be a problem after surgery. Therefore, we contemplated the surgery with the plan of localizing and interrupting the accessory pathway.

**Body Surface Map**

The technique for producing accurate body surface maps has been described previously. Figure 3 shows the body surface map at one instant early in the ST-T wave. Based on the experimental studies of Spach et al., the pattern during the ST-T wave localized the site of earliest repolarization as an anterolateral site on the left-sided ventricle, a location subsequently confirmed by intracardiac electrode catheter studies and by epicardial mapping at surgery.

**Electrophysiologic Study**

After sedation with morphine, four electrode catheters were inserted percutaneously and positioned in the high right atrium, in the coronary sinus, across the right-sided AV valve for His bundle recording and in the apex of the right-sided ventricle. The patient had not taken antiarrhythmic drugs for 48 hours.

With the catheter across the right AV valve, we could record a rapid deflection, which occurred after the onset of the surface QRS and demonstrated preexcitation. However, when we paced at this site, the stimulus to onset of QRS interval was 35 msec; the QRS duration was 95 msec; and lead V1 showed a QS pattern. We were uncertain if we were recording from the His bundle or the right-sided bundle branch. The high right atrial catheter was used to explore, and on a pullback from the descending aorta, the patent ductus and the main pulmonary artery, a rapid deflection preceding the QRS was recorded in the subpulmonic region during a preexcited beat and during a His extrasystole (fig. 4). We concluded that we were record-

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**Figure 3.** ST-T-wave isopotential body surface map. The instant shown occurs 179 msec after onset of QRS as indicated by the tic mark on the reference electrogram below. Potentials in microvolts X 10 are shown at 100 sites on the anterior torso and 50 on the back (total = 150). The right most column on the back is duplicated on the left. The plus (+) and minus (−) signs denote the maximum and minimum. The positive (solid) and negative (dashed) isopotential lines are drawn automatically using a logarithmic scale. The thick dashed line represents the zero potential line. Based on the experimental studies of Spach et al., a left lateral site of ventricular preexcitation was predicted.
ing the His bundle electrogram in the subpulmonic region; however, the catheter was unstable in this position. For the rest of the study we used the more stable recording of the bundle branch potential recorded across the annulus of the right AV valve instead of the His bundle potential recorded in the subpulmonic region.

Antegrade and retrograde refractory period determinations were evaluated using the extrastimulus technique. Atrial stimulation with progressively more prematurity resulted in more preexcitation. We could not evaluate the antegrade refractory period of the AV node or the accessory pathway, which were limited by refactoriness of the atrium (240 msec at cycle length 500 msec). Retrograde pacing and premature stimulation always showed eccentric atrial activation with the earliest area at the most distal coronary sinus recording site. A definite statement about retrograde conduction through the AV node could not be made. It was difficult to induce supraventricular tachycardia, but when it could be induced (fig. 5), the QRS normalized and the retrograde atrial activation sequence showed earliest activity in the distal coronary sinus. Atrial flutter with 1:1 ventricular capture at a cycle length of 210 msec was produced by incremental atrial pacing; all QRS complexes were preexcited.

Based on this study we concluded: 1) an accessory AV pathway was present at a left anterolateral site; 2) it participated in SVT; 3) atrial flutter-fibrillation was present with rapid ventricular response; and, 4) antegrade conduction in the AV node was present.

Epicardial Mapping

At the time of surgery, the heart was exposed through bilateral transverse incisions through the fourth intercostal space on the right and the fifth intercostal space on the left. The external anatomy of the heart revealed the general relationship of the great arteries and the ventricles to be that of corrected transposition with ventricular inversion. The epicardial map was performed as previously described.8 Before institution of cardiopulmonary bypass and hypothermia, epicardial mapping was performed with the patient in sinus rhythm. This showed that initial ventricular activation occurred on the anterolateral left-sided ventricle at the base of the left atrial appendage (fig. 6), i.e., the site predicted by the body surface map and the intracardiac electrophysiologic study. Atrial mapping during ventricular pacing confirmed this site. Additionally, a transition of electrograms between atrial and ventricular electrograms were noted over a distance of 2 cm in the region of the left AV groove, confirming the presence of an atrialized portion of the ventricle.

Figure 4. Intracardiac electrograms showing His and bundle branch potentials. Recordings are from standard leads I and V1, apex of right-sided ventricle (RV), subpulmonic region (HBE), across right-sided atrioventricular valve (BBE), and distal coronary sinus (CS). A preexcited beat followed by a His extrasystole is shown. During the His extrasystole the QRS normalizes and in the subpulmonic region is preceded by a rapid deflection (H). When the recording is across the right-sided AV valve, the rapid deflection occurs later. The catheter in the subpulmonic region was unstable and subsequent recordings use BBE for HBE.

Figure 5. Intracardiac electrograms of supraventricular tachycardia. Format is same as in figure 4 except for bipolar recording from high right atrium (RA). The deflection labeled H is probably recorded from a bundle branch (see figure 4). The QRS duration is normal and the retrograde sequence of atrial activation shows the earliest activity in the distal coronary sinus.
Meticulous recordings were then made over the AV groove in the region of preexcitation and a discrete electrogram was localized that was felt to represent a recording from the accessory pathway. However, since the recording site was so close to a coronary artery, we felt that blunt dissection rather than cryoablation would be the procedure of choice for interruption of the accessory pathway.

**Surgery**

After institution of cardiopulmonary bypass and hypothermia, the left atrium was opened and inspection of the left AV valve revealed an Ebstein's deformity of the valve. The method of pathway interruption described for Ebstein's anomaly was used. From the epicardial surface, the coronary artery was reflected downward, exposing the junction between the atrium and the thin, atrialized ventricle, which measured about 2 cm. The atrium was divided through to the endocardium just above the insertion of the atrium into the annulus fibrosus for a distance of 1.5 cm on each side of the earliest point of atrial and ventricular activation.

An additional incision was made in the left atrium, and a 2-cm VSD was identified beneath the septal leaflet of the left AV valve. The valve was removed and the VSD was closed with a Teflon felt patch. A 29-mm Hancock prosthetic valve was positioned and sutured in the left AV valve annulus. Finally, the PDA was suture ligated. The His bundle was not mapped because of time limitation and because inspection of the VSD from the left side revealed no alternative to its closure.

After completion of cardiopulmonary bypass and rewarming, atrial and ventricular pacing demonstrated complete antegrade and retrograde AV block. A permanent pacemaker with an epicardial electrode was implanted.

In the immediate postoperative period the peak systolic pressures in the pulmonary artery and radial artery were 45 mm Hg and 110 mm Hg, respectively. The mean left atrial pressure was 12 mm Hg, and an indocyanine green dye curve was normal. The patient was taken to the recovery room in excellent condition.

The patient was discharged on the ninth postoperative day and has done well in the 9 months since surgery. He has returned to school full time. He continues to have third-degree AV block, which is treated with a pacemaker, but he requires no medication.

**Discussion**

This is the first patient we know of with corrected transposition and severe intracardiac deformities who has successfully undergone interruption of the AV pathway as well as extensive intracardiac corrective surgery. A recent report demonstrates that the presence of WPW in children undergoing corrective surgery for congenital cardiac defects can be fatal. Our patient illustrates some of the difficulties involved in the preoperative electrophysiologic evaluation of patients with severely malformed hearts.

Since previously reported techniques for predicting the site of ventricular preexcitation using electrocardiography, vectorcardiography, and body surface mapping have been based on the analysis of delta waves in older patients with structurally normal hearts, we elected to make our initial estimation of the
location of the site of preexcitation using body surface maps of the early ST-T wave based on the experimental studies of Spach et al. Based on the similarity of the patient's body surface map during the early ST-T wave to that obtained from a chimpanzee with an ectopic stimulus on the left lateral ventricle, we predicted a left lateral location for the accessory pathway. Thus, to confirm this site with intracardiac electrode studies it was mandatory that recordings be obtained from the His bundle and multiple atrial sites including the left atrium, achieved in our patient by recording from the distal coronary sinus. The left lateral location of the accessory pathway was subsequently confirmed by epicardial mapping and surgical ablation of the accessory pathway.

Our patient illustrates several anatomic variants in patients with corrected transposition that can frustrate electrophysiologic evaluation. The subpulmonic location of the His bundle electrogram during electrode catheter studies in our patient was consistent with the location described by Waldo et al., Kupersmith et al., and Anderson et al., and different from the location during catheter studies of Wolff et al. and Gillette et al. We did not map the His bundle during surgery because of time limitation and because inspection of the VSD was from the left side and revealed no alternative to its closure. Additionally, Waldo et al. demonstrated absence of the ostium of the coronary sinus in two of five patients with corrected transposition. Fortunately, it was present in our patient.

Superficially, it might seem advantageous for a patient with corrected transposition to have an accessory AV pathway, because conduction defects in the regular pathway are common and spontaneous complete AV block occurs frequently. Vanetti et al. reported a case of a child with normally related great vessels and WPW who acquired AV block during surgery for subvalvular aortic stenosis. They saw the acquisition of AV block as a reason to leave the accessory pathway intact, but they did insert a demand pacemaker as well. Our patient acquired AV block during surgery, but because of the patient's documented atrial flutter-fibrillation with rapid ventricular response using the accessory pathway, we were committed to interruption of the accessory pathway.

Our patient illustrates the interrelationship between isopotential body surface mapping, intracardiac electrophysiologic studies and epicardial mapping in the electrophysiologic evaluation of WPW even in the presence of quite complex congenital heart disease. In this patient we could evaluate our correct preoperative interpretation of the electrophysiologic techniques by the surgical result, and we successfully carried out all surgical procedures to correct the numerous anatomic, hemodynamic and electrophysiologic defects.

References
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Circulation. 1980;61:1256-1261
doi: 10.1161/01.CIR.61.6.1256

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