to its abolition during intravenous tocainide administration. It seems likely that acceleration of VT rate was induced by tocainide, since in both of our patients their episodic VT had been at constant frequency during observation for 1 month. Although both of these patients had severe underlying heart disease with chronic potentially life-threatening recurrent VT, VF occurred only once in each individual and in each instance in close proximity to initiation of tocainide therapy. Furthermore, VF has not reoccurred since tocainide was discontinued. There were no apparent metabolic or electrolyte abnormalities which might have explained their worsening of ventricular tachyarrhythmias during tocainide. Other antiarrhythmic drugs had been discontinued sufficiently prior to VF to make such change in therapy an important factor. Serum digoxin concentrations were at levels unlikely to be associated with toxic arrhythmias. While a potential interaction between tocainide and digoxin might have resulted in VF, such an event has not been observed with lidocaine, a drug with similar structure and electrophysiologic effects as tocainide. While it is not possible to exclude the causal relation of their severe underlying cardiac disease processes or disturbed pharmacokinetics due to related organ system dysfunction, it is not unreasonable to suspect that tocainide was responsible for initiation of VF in these two patients.

Unfortunately, tocainide blood levels were not available in both patients. Tocainide is principally removed (45%) by excretion in the urine in unchanged form and 23% is metabolized by carboxylation in the liver. Since our first patient had severe congestive heart failure, impaired excretion might have resulted in toxic blood levels. However, our second patient had normal renal and hepatic function. Based on previous experience with the pharmacokinetics of this highly bioavailable oral drug,[4, 6] it is quite unlikely that our second patient had toxic serum levels. Furthermore, signs and symptoms of CNS or gastrointestinal toxicity were absent in both patients, and these side effects usually occur with high tocainide blood levels.[4, 6] Based on these two untoward experiences, we now believe that it is prudent to initiate tocainide therapy in a monitored hospital setting. The drug has proved useful in patients with refractory ventricular tachyarrhythmias and we continue to find tocainide salutary in such patients.

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Tricuspid atresia and the Wolff-Parkinson-White Syndrome: Evaluation methodology and successful surgical treatment of the combined disorders

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The occurrence of tricuspid atresia and Wolff-Parkinson-White syndrome in a single patient is rare, occurring in only one among 349 patients with tricuspid atresia included in three recently published reviews.[1-3] Despite the reported uncommon coexistence of these two complex lesions, recent advances in their individual surgical management as well as the hemodynamic deterioration which frequently accompanies paroxysmal supraventricular tachycardia (PSVT) prompted us to report the successful division of a right-sided posterior bypass tract and the creation of a valveless right atrial to right ventricular conduit in an 18-year-old girl with tricuspid atresia and the pre-excitation syndrome.

At 3 months of age the patient demonstrated during cardiac catheterization tricuspid atresia with normally related great arteries; a right Blalock-Taussig anastomosis was performed 2 years later. At 5 years of age suspected anomalous atrioventricular conduction without PSVT was observed transiently on her electrocardiogram (Fig. 1), followed by her first episode of PSVT at age 12 years. Six years later because of increasing polycythemia, exercise intolerance, and recurrent symptomatic PSVT, the patient underwent cardiac catheterization and electrophysiologic study. Pulmonary arterial pressure was 22/12 mm Hg. Electrophysiologic study (Figs. 2 and 3) demonstrated a right-sided bypass tract supporting the tachycardia retrogradely, with an antegrade effective refractory period of greater than 450 msec. Following these studies, atrial and ventricular epicardial mapping (Fig. 4), division of the right-sided bypass tract, and a modified Fontan procedure[4] were performed (Fig. 5). Following surgery both 24-hour Holter recording and postoperative programmed atrial extrastimuli delivered through transtho-

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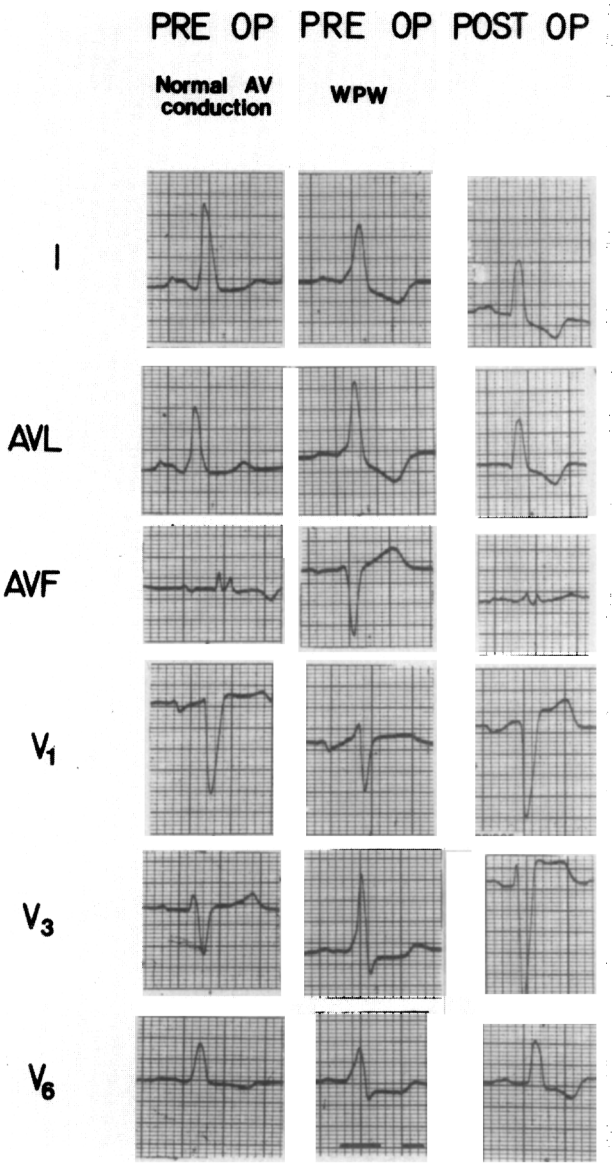

Fig. 1. Selected leads from the surface ECG prior to operation during sinus rhythm and either normal atrioventricular conduction *(left-hand column)* or suspected anomalous (WPW) conduction *(middle column)*. Leads 1 and V_3 exhibit small delta waves and a frontal plane QRS axis shift to a superior orientation. Postoperative selected leads *(right-hand column)* indicate a return to normal atrioventricular conduction. (Paper speed = 50 mm/sec; 10 mm defection equals 1 mV.) *AV* = atrioventricular; *WPW* = Wolff-Parkinson-White syndrome.

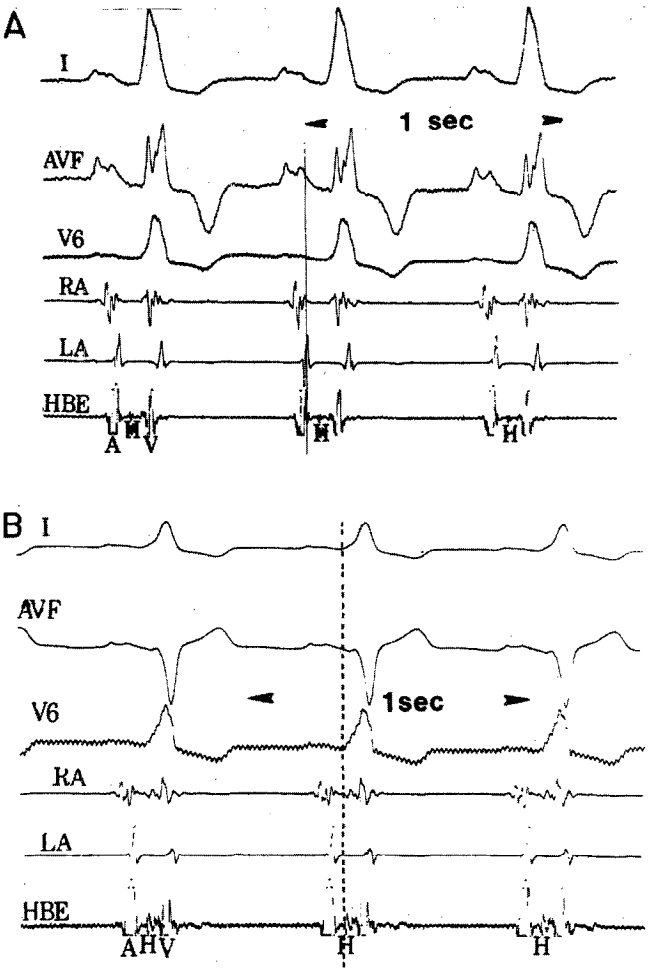

Fig. 2. *Panel A* shows tracings obtained at electrophysiologic study during sinus rhythm and normal atrioventricular conduction. *Panel B* shows tracings obtained at electrophysiologic study during sinus rhythm and anomalous atrioventricular conduction. Note the simultaneous activation of the His bundle *(H)* and ventricle (QRS complex) indicated by the *dotted vertical line*. All electrograms at cardiac catheterization and surgery were recorded through bipolar electrodes (1 to 10 mm apart) connected to an Electronics for Medicine V1205A amplifier. The signals were filtered through a band width of 30 to 250 Hz, monitored on an Electronics for Medicine (VR12) switched-beamed oscilloscope and recorded on photographic paper moving at 100 to 150 mm/sec. 1, AVF, V_6 = standard surface ECG leads; *A* = atrial electrogram; *HBE* = His bundle tracing; *RA* = high right atrial tracing; H = His bundle electrogram; *LA* = left atrial tracing; *V* = ventricular electrograms.

racic temporary atrial electrode wires failed to demonstrate tachycardia. One year following surgery the patient is improved without discernible cyanosis or tachyarrhythmias.

Despite the constraints imposed by the pathologic anatomy of tricuspid atresia, the electrophysiologic study proved to be important in establishing existence of an anomalous conduction pathway (Figs. 2, *A* and *B*), in localizing the origin of the bypass tract on the right side (Fig. 3), and in defining the risk of rapid ventricular rates should atrial flutter or fibrillation develop.[5, 6] The demonstration of ventricular excitation occurring simultaneously with activation of the His bundle (Figs. 2,*A* and *B*) during periods of suspected anomalous conduction proved the presence of the bypass tract.[5] The minor shortening of the PR interval observed during suspected anomalous

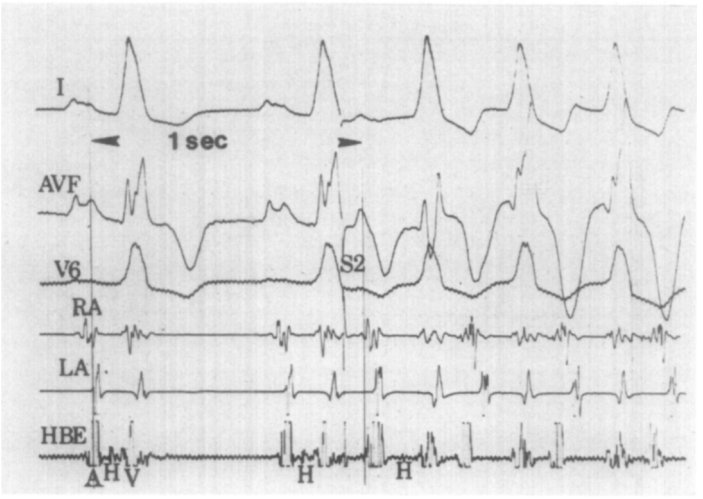

Fig. 3. Tracings obtained at electrophysiologic study demonstrating the initiation of paroxysmal supraventricular tachycardia (PSVT) by a single atrial premature stimulus (S_2) coupled (300 msec) to sinus rhythm. Note that during PSVT the right atrium is activated before the left atrium, indicating a right-side bypass tract. A smooth atrioventricular nodal conduction curve and absence of longitudinal dissociation militated against atrioventricular nodal reentry as the mechanism of this tachyarrhythmia. Note normal QRS morphology during the tachycardia, indicating the antegrade conduction during the tachycardia is through the normal atrioventricular pathway and retrograde conduction is through the bypass tract.

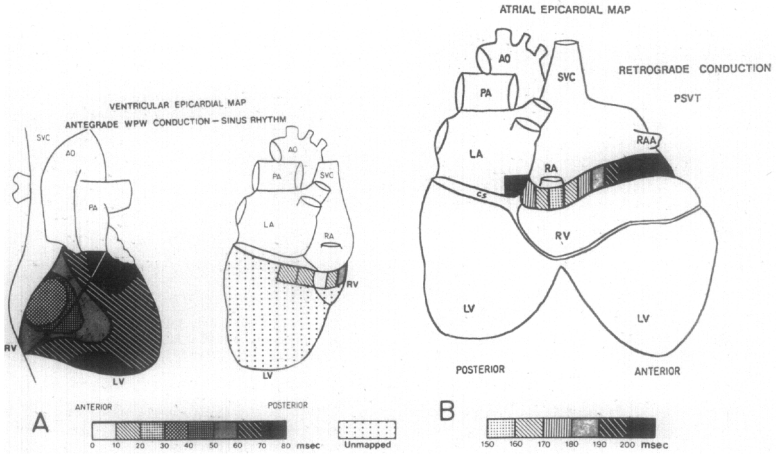

Fig. 4. In *Panel A,* the ventricular epicardial activation sequence during antegrade anomalous (WPW) conduction is marked by 10 msec intervals. The earliest activation of the ventricles occurred on the posterior diaphragmatic surface of the small right ventricle just to the right of the crux. Thus, the ventricular end of the anomalous pathway was identified. Anterior epicardial activation during anomalous conduction occurred earliest (30 to 40 msec) over the diminutive right ventricle, demonstrating that during anomalous conduction the ventricles were activated by both the normal and anomalous pathways, and that the surface ECG with a superior frontal plane axis reflected fusion of these two activation wave fronts. Complete mapping of the posterior ventricular epicardium was not carried out to conserve operating time. Mercator projection of the heart is shown in *Panel B.* Atrial epicardial mapping parallel to the right-sided atrioventricular groove was accomplished during supraventricular tachycardia. This figure *(Panel B)* demonstrates a retrograde right atrial epicardial activation sequence. The retrograde atrial activation time was measured from a reference ventricular electrogram to the atrial electrogram recorded through the hand-held exploring electrode prode. Intervals are 10 msec, beginning at 150 msec. The earliest atrial activation site during PSVT was located on the posterior right atrium slightly to the right of the earliest ventricular activation site obtained during anomalous antegrade conduction, thereby locating the atrial end of the bypass tract and suggesting slight obliquity to its course. *LA* = left atrium; *PA* = pulmonary artery; *RAA* = right atrial appendage; *SVC* = superior vena cava; *LV* = left ventricle; *RA* = right atrium; *RV* = right ventricle; *WPW* = Wolff-Parkinson-White syndrome.

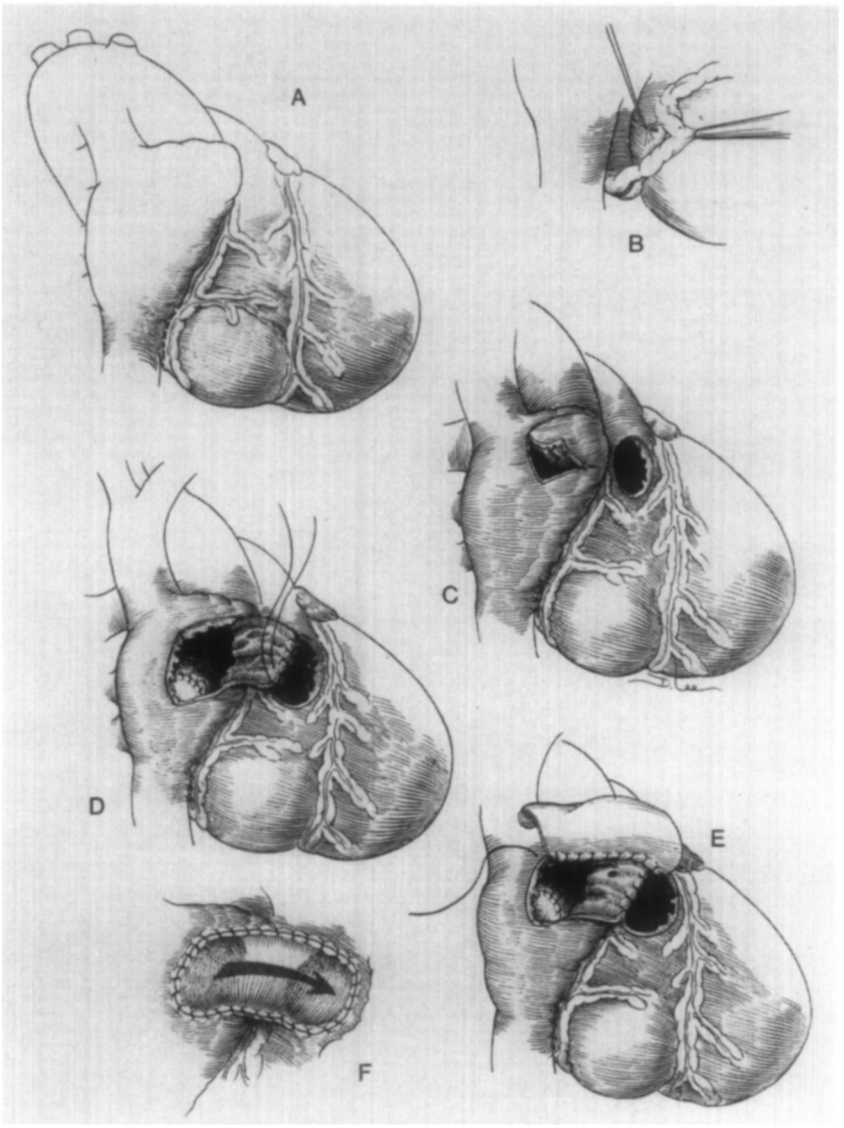

Fig. 5. *Diagram A* shows the lordotic view of the heart (the view is as if one is looking at the heart from the diaphragm). The "Ebstein-like" pouch projects inferiorly from the diaphragmatic surface; this tissue generated a ventricular electrogram but communicated with the right atrium, not the right ventricular cavity above it. The *dotted line* indicates the site of the epicardial incision. In *Diagram B,* the right coronary fat pad was dissected away from the atrioventricular groove through an incision in the epicardium extending from the insertion of the inferior vena cava to the crux of the heart posteriorly. By sharp and blunt dissection under the elevated fat pad the atrium was separated from the ventricle along this entire length, exposing the roof of the coronary sinus and posterior portion of the ventricular septum. A through-and-through incision was then made in the atrialized ventricle at the point of separation from the ventricular septum and coronary sinus to insure complete division of all fibers between the atrium and ventricle. The epicardial and endocardial incisions were then closed. *Diagrams C* through *F* illustrates the modified Fontan technique. The pulmonary valve was normal in size; therefore it was incorporated into the repair. The atrial septal defect was closed with a Dacron patch. A 5 cm longitudinal incision was made in the right ventricular infundibulum. A 5- by 5-cm flap of right atrial appendage was folded back and sutured to the right edge of the ventriculotomy to form the floor of the conduit *(Diagrams C and D).* Then a 7-by-9 cm rectangle of pericardium was sutured to the edge of the atrial flap, and to the left edge of the ventriculotomy to constitute the roof of the conduit *(Diagrams E* and *F).* Upon completion of the procedure, the patient was in sinus rhythm without anomalous atrioventricular conduction (Fig. 1, *right-hand panel*).

conduction was explained by finding a delay in intra-atrial conduction (PA interval 53 msec). Finally, the antegrade effective refractory period in the anomalous pathway of greater than 450 msec (sinus cycle length 740 msec) reduced the risk of the initiation of either ventricular tachycardia or ventricular fibrillation.

The finding of the Ebstein-like defect at surgery (Fig. 5,*A*) raises the question of the anatomy of the primary lesion—Ebstein's anomaly or tricuspid atresia. The major classification of tricuspid atresia is dependent upon the associated lesions.[1] More recently several authors have classified tricuspid atresia by the type of obstruction of the tricuspid valve.[7] Our patient's anatomy conforms to the Ebstein type of tricuspid atresia,[7] whereas the pathophysiology, dependent upon the associated lesions, fits Edward's type 1B.[1] Despite the "Ebstein-like" anatomy, severe hypoplasia of the right ventricle necessitated the Fontan operation. (Figs. 5,*C* to *F*). The surgical approach to the identification (Fig. 4) and division (Fig. 5,*B*) of the bypass tract was similar to previously described techniques in patients with posterior right-sided pathways with Ebstein's anomaly.[8] The epicardial mapping procedure yielded precise localization of the accessory pathway (Fig. 4), allowing accurate and limited dissection (Fig. 5,*B*).

This experience underscores the importance of defining the mechanism of supraventricular tachycardias in patients with tricuspid atresia. Furthermore, patients with tachyarrhythmias and other forms of congenital heart disease should be evaluated electrophysiologically, since their tachyarrhythmia may depend upon anomalous atrioventricular pathways and therefore may be amenable to surgical treatment at the time of anatomic repair.[9-10]

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Romano-Ward prolonged QT syndrome with intermittent T wave alternans and atrioventricular block

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A 2½-year-old female presented with six episodes of generalized convulsions over a period of 6 months. There were no other cardiac symptoms and no history of sudden death in the family. Physical examination was normal. Electrocardiograms showed QT prolongation as a constant feature and intermittently T wave alternans was recorded (Fig. 1). On two occasions postconvulsions, electrocardiograms showed 2:1 atrioventricular block (Fig. 2). Although intravenous atropine caused disappearance of T wave alternans with decrease in QT interval, no response was induced by carotid sinus stimulation. All other relevant investigations performed including an audiogram were normal. The patient is now asymptomatic with institution of phenytoin and phenobarbital therapy. The QT interval continues to remain prolonged.

In 1957 Jervell and Lange-Nielsen[1] jointly reported the peculiar syndrome of QT prolongation with deafness. The syndrome without deafness has been termed the Romano-Ward syndrome. Syncopal episodes in these syndromes have been documented to be due to ventricular fibrillation[2] or less frequently to ventricular asystole.[3] In the Romano-Ward syndrome, the QT interval prolongation represents a stable feature, while T wave alternans is episodic and appears related to changes in activity of the sympathetic nervous system.[4] In our patient T wave alternans was present at slower heart rates, but when the heart rate was increased by a vagolytic agent, T wave alternans disappeared. Similar findings have been reported by Lopez et al.[5] The presence of intermittent atrioventricular block is also an unusual feature in our case. This finding is consistent with the patient reported by Crawford et al.,[6] in whom atrioventricular nodal disease was progressive.

No consistently effective therapy is available. Although propranolol is considered to be the most effective drug for preventing ventricular tachyarrhythmias, the agent is unsafe in the presence of atrioventricular nodal disease because of its action of depressing atrioventricular conduction. Phenytoin (diphenylhydantoin) has been successfully employed, as in our patient, because of the agent's inhibitory effect on central nervous system dis-

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