JACC Vol. 17, No. 1
January 1991:174–81

Electrophysiologic Findings After Fontan Repair of Functional Single Ventricle

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Cardiac arrhythmias are well recognized sequelae of the Fontan operation for complex congenital anomalies. In this study the electrophysiologic effects of the Fontan procedure were evaluated in 30 patients who underwent cardiac catheterization with electrophysiologic study 1.9 ± 1.3 years (mean ± SD) after modified Fontan repair for functional single ventricle. Abnormalities of sinus node or ectopic pacemaker automaticity were detected in 50% (15 patients) by determination of a prolonged corrected sinus node or pacemaker recovery time. Total sinoatrial conduction time was prolonged in 50% of the patients with normal sinus rhythm. Sinus node or ectopic atrial pacemaker function was entirely normal in only 43% of patients. The predominant atrial rhythm was normal sinus in 70% and ectopic atrial or junctional in 30%. Abnormalities of atrial effective and functional refractory periods were noted in 43% of patients and were most pronounced at faster paced cycle lengths. Atrial endocardial catheter mapping revealed intraatrial conduction delays between adjacent sites in 76% of the patients tested and in eight of nine patients with inducible intraatrial reentry.

Programmed atrial stimulation induced nonsustained supraventricular arrhythmias in 10% of the 30 patients and sustained arrhythmias in 27%. Intraatrial reentry was the most common inducible arrhythmia and was present in seven of the eight patients with sustained and two of the three patients with nonsustained atrial arrhythmias. Atrioventricular conduction abnormalities were noted in 10% (three patients). No patient had inducible ventricular arrhythmias with programmed ventricular stimulation.

The electrophysiologic findings after Fontan repair include abnormal sinus node function, prolonged atrial refractoriness, delayed intraatrial conduction and inducible atrial arrhythmias. The predictive value of electrophysiologic testing in this group of patients is unknown. Further study and continued clinical follow-up are essential to determine which patients will be at risk for the development of late life-threatening arrhythmias.

(J Am Coll Cardiol 1991;17:174–81)

Cardiac arrhythmias are well recognized sequelae of the Fontan operation for complex congenital cardiac anomalies. Reported abnormalities include sinus node dysfunction, atrioventricular (AV) block, ectopic atrial rhythm, accelerated junctional rhythm, supraventricular tachycardia and ventricular arrhythmia (1–8). Supraventricular tachycardia with rapid ventricular response may be associated with compromised cardiac output and a contributory cause of death (9,10), especially in the perioperative period (2,11–13). Reported risk factors for the development of supraventricular tachycardia after Fontan repair have included P wave abnormalities on the preoperative electrocardiogram (ECG), elevated pulmonary artery pressure, right atrial enlargement and low aortic oxygen saturation (11,14). Ventricular arrhythmias have been described after this repair (1,2,4,8, 15,16). Sudden death, presumed to be secondary to arrhythmias, has been reported (incidence rate 3% in large series) (7,17–19).

In this study we describe the electrophysiologic findings in 30 patients who underwent the Fontan procedure for repair of functional single ventricle.

Methods

Study patients. The study group included 30 patients who underwent electrophysiologic study between October 1984 and December 1988 at the Children's Hospital of Philadelphia after modified Fontan repair; 20 patients were male and 10 female. The age at surgery ranged from 1.7 to 17.7 years (mean ± SD 7.2 ± 4.1, median 6.4). Electrophysiologic studies were performed 7 months to 6.2 years after surgery (mean 1.9 ± 1.3 years). The cardiac anatomy before operation included tricuspid atresia in 11 patients, single left ventricle in 6, heterotaxy syndrome in 5, mitral atresia in 3, single right ventricle in 2 and other complex lesions in 3.

Preoperative and postoperative arrhythmias. Four patients had preoperative arrhythmias: complete congenital heart block in one patient, frequent ventricular premature

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Manuscript received December 26, 1989; revised manuscript received August 15, 1990, accepted August 27, 1990.

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0735-1097/91/$3.50

beats in two patients and atrial flutter in one patient. Five patients had early (<14 days) postoperative arrhythmias: preexisting complete heart block in one patient and new arrhythmias in four patients, including supraventricular tachycardia in two, atrial flutter in one patient and ventricular tachycardia associated with a perioperative myocardial infarction in one. Eight patients had late (>14 days) postoperative arrhythmias: persistence or recurrence of preoperative arrhythmias in three patients, including complete heart block in one patient, frequent ventricular premature beats in one and atrial flutter in one, new late postoperative arrhythmias occurred in five patients, including supraventricular tachycardia in two and atrial flutter in three. Nine patients (30%) had new early or late postoperative arrhythmias.

Surgical procedures. Twenty-seven patients had corrective surgery or a palliative procedure before the Fontan repair. Forty-three procedures were performed, including a systemic to pulmonary artery shunt in 20 patients, atrial septostomy or septectomy in 13, pulmonary artery banding in 7, coarctation repair in 1 patient, the Rastelli procedure in 1 and Norwood palliation for hypoplastic left heart syndrome in 1.

Five basic modifications of the Fontan repair were utilized and varied according to the preoperative anatomy. In 28 of the 30 patients the Fontan procedure was performed by the same surgeon; in 2, the operation was performed at other institutions. Modifications included direct anastomosis of the right atrium to the right pulmonary artery in 11 patients, intraatrial Gore-Tex baffle of the systemic venous return from the inferior vena cava to the junction of the superior vena cava-right atrium-right pulmonary artery anastomosis in 11, intraatrial tube conduit from the inferior vena cava to the superior vena cava-right pulmonary artery connection in 3, conduit from the right atrium to the right ventricular outflow tract in 2, direct connection of the right atrium to the right ventricular outflow tract in 2 and intraatrial Gore-Tex baffle of the pulmonary venous return to the right atrioventricular (AV) valve in 1.

Electrophysiologic studies. Electrophysiologic studies were performed at the time of scheduled postoperative cardiac catheterization in 25 patients. Five additional patients were studied specifically for evaluation of recurrent or symptomatic arrhythmias noted 11 to 74 months (mean 34.8 ± 26.6, median 31) after surgery. Twenty of the patients were receiving digoxin at the time of evaluation; arrhythmia was the primary indication for initiation of digoxin therapy in seven patients, including all five patients studied for recurrent arrhythmias and two patients whose arrhythmia was adequately controlled with this drug. Two patients with recurrent arrhythmias were receiving quinidine or propranolol in addition to digoxin.

Written informed consent was obtained from all patients and the study protocol was approved by the institutional review board of our hospital. Electrophysiologic studies were performed with patients in the sedated, postabsorptive state after administration of a combination of oral meperidine (3 mg/kg body weight) and pentobarbital (4 mg/kg) or intramuscular morphine sulfate (0.15 mg/kg) and pentobarbital (4 mg/kg) according to our previously described methods (20).

Electrophysiologic study protocol. The following protocol was used to evaluate study patients: 1) basal recordings of the spontaneous cycle length, AH and HV intervals; 2) atrial pacing at multiple cycle lengths from 600 to 250 ms; 3) single premature atrial stimulation during basal rhythm and two paced cycle lengths; 4) double premature atrial stimulation at one paced cycle length; 5) triple premature atrial stimulation at one paced cycle length if there was a clinical history of arrhythmia but no arrhythmia was induced by steps 2, 3 and 4; 6) ventricular pacing at multiple cycle lengths from 600 to 250 ms; 7) single and double premature ventricular stimulation at two paced cycle lengths; and 8) catheter endocardial mapping of the right atrium during basal and induced rhythms.

Endocardial catheter mapping. Catheter endocardial mapping was used to determine atrial activation sequences and intraatrial conduction intervals. Right atrial recordings were obtained with an exploring electrode catheter positioned at six sites (high right atrium, mid-lateral, low lateral and low medial right atrium, interatrial septum and AV junction). Either the onset of the P wave on the surface ECG or a stable electrogram was used as the reference. The earliest site of activation was considered to be the origin of the rhythm. Sinus rhythm was defined as an atrial rhythm with earliest activation in either the high or the mid-lateral right atrium near the onset of the surface P wave; ectopic atrial rhythm was defined as an earliest activation site other than the high or mid-lateral right atrium. Junctional rhythm was defined as a rhythm with earliest activation in the His bundle or ventricle followed by or not associated with atrial activation.

Electrocardiographic data. An ECG was obtained on the day before the electrophysiologic study and analyzed. Twenty-four hour Holter ECG recordings were obtained in 27 of the 30 patients within 1 day of electrophysiologic study.

Statistical analysis. All data are expressed as mean values ± SD. The unpaired two-tailed Student's *t* test was used to compare grouped data. Statistical significance was defined as $p < 0.05$.

Results

Cardiac catheterization. Standard hemodynamic and angiographic cardiac catheterization studies were performed in all 30 patients (Table 1). Right atrial pressure was ≥7 mm Hg in all patients and ≥14 mm Hg in 13 patients. A gradient of 2 to 3 mm Hg across the right atrium to the pulmonary artery anastomosis was present in seven patients. No patient had significant residual intracardiac shunting, although six patients had a small residual interatrial baffle leak. One patient with a clinical history of perioperative myocardial infarction had angiographic evidence of left ventricular dysfunction. One

Table 1. Cardiac Catheterization Data in 30 Patients After Fontan Repair

	Mean	SD	Range
Aortic O_2 saturation (%)	93.5	2.7	86 to 97
End-diastolic pressure (mm Hg)	7.3	2.9	2 to 15
Cardiac index (liters/min per m^2)	2.5	0.5	2.0 to 3.6
Right atrial pressure (mm Hg)	13.3	3.4	7 to 23
Gradient from right atrium to pulmonary artery (mm Hg)	0.9	1.0	0 to 3

patient, with L-transposition of the great arteries, ventricular inversion and single left ventricle had a restrictive bulboventricular foramen. No other patient had any significant hemodynamic abnormality that necessitated additional surgery.

Electrocardiographic results. Twenty-two patients (73%) had normal sinus rhythm and 6 (20%) had an ectopic atrial rhythm with a P wave axis >90° or <0°. Two patients had junctional rhythm with P waves following the QRS complexes and one patient had junctional rhythm alternating with an ectopic atrial rhythm. Atrioventricular conduction disturbances included first degree AV block in 11 patients, first and second degree AV block in 1 patient and complete heart block that was present preoperatively in 1 patient. No patient had tachyarrhythmias on a routine rest ECG.

Holter ECG evaluation. Of the 27 Holter recordings analyzed, the predominant rhythm was normal sinus in 17, normal sinus rhythm alternating with an ectopic atrial rhythm in 4, sinus rhythm alternating with junctional rhythm in 3, ectopic atrial rhythm in 1, ectopic atrial rhythm alternating with junctional rhythm in 1 and junctional rhythm in 1. Eight patients (mean age 10.6 ± 4.6 years) had a minimal heart rate ≤50 beats/min; only two of these patients were teenagers. A 2 s pause was seen in one tracing. Atrioventricular conduction abnormalities were noted in three patients: first and second degree AV block in two patients and complete heart block in one patient. Supraventricular tachycardia was present in three patients, one of whom had findings suggestive of an automatic atrial tachycardia; in the other two, the mechanism of the supraventricular tachycardia could not be determined. Five patients had ventricular ectopic complexes; one patient had >400 single premature ventricular complexes/h, three patients had rare ventricular couplets with coupling intervals from 260 to 580 ms and one patient had eight episodes of torsade de pointes determined to be quinidine related.

Atrial activation during electrophysiologic study. Three to 10 surface ECGs were recorded in all patients during electrophysiologic study. The predominant atrial rhythm was sinus in 21 patients (70%) and ectopic atrial in 9 (30%). The origin of the rhythm was confirmed by catheter endocardial mapping in the 25 patients whose type of surgical repair allowed this technique to be utilized. By catheter endocardial mapping, 17 patients (68%) had sinus rhythm and 8 (32%) had ectopic atrial rhythm from the low medial right atrium (n = 4) (Fig. 1), the low lateral right atrium (n = 3) and the right atrial-right pulmonary artery junction (n = 1). The patient shown in Figure 1 had an ectopic atrial rhythm with a 75 ms intraatrial conduction delay between the low lateral and low medial right atrium.

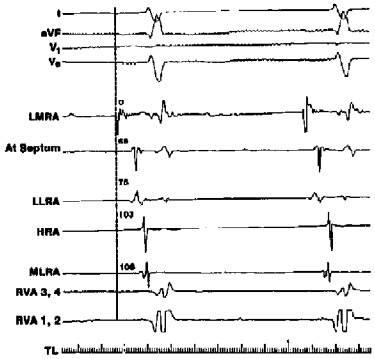

Figure 1. Analog data from which the right atrial activation map was constructed. Electrocardiographic leads I, aVF, V_1 and V_6 are shown with intracardiac electrograms from the low medial right atrium (LMRA), residual atrial (At) septum, low lateral right atrium (LLRA), high right atrium (HRA), midlateral right atrium (MLRA), proximal right ventricular apex (RVA 3,4), distal right ventricular apex (RVA 1,2) and 10 ms time lines (TL). The site of origin of the rhythm, indicated by the dashed vertical line, is the low medial right atrium.

Endocardial catheter mapping was possible in three of the five patients with heterotaxis syndrome; all three patients had an ectopic atrial pacemaker with earliest atrial activation in the low lateral right atrium in one, low medial right atrium in one and right atrial-pulmonary artery junction in one. The surface P wave axis was used in the other two patients with heterotaxis syndrome to determine the origin of the atrial rhythm. As assessed by ECG criteria, one of these patients had normal sinus rhythm and the other a low right atrial rhythm. Therefore, of the five patients with heterotaxis syndrome, four had an ectopic atrial rhythm.

A marked intraatrial conduction delay between adjacent sites, as compared with normal values in our laboratory, was noted in 19 (76%) of the 25 patients in whom atrial maps were obtained. Comparison of the mean atrial activation times in the 17 patients with sinus rhythm with values in 20 normal children (Fig. 2) showed delayed activation of multiple atrial sites.

Sinus node and ectopic atrial pacemaker function. The function of the sinus node or ectopic atrial pacemaker was evaluated by previously described methods (21–24). The mean cycle length of patients with sinus rhythm was 733 ±

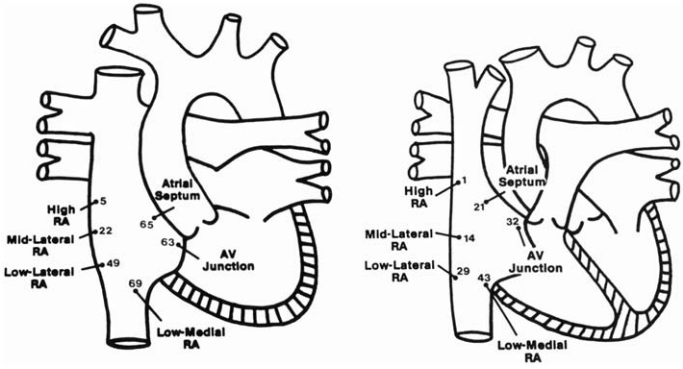

Figure 2. Left, Mean right atrial (RA) activation times (in ms) in 17 patients with normal sinus rhythm after Fontan repair. Right, Mean atrial activation times in 20 normal patients without structural heart disease. Activation of the low lateral right atrium ($p < 0.05$), atrioventricular junction ($p < 0.05$) and low medial right atrium ($p < 0.005$) is delayed in the postoperative patients when compared with that in the normal group.

120 ms versus 779 ± 253 ms in patients with ectopic atrial rhythm (p = NS). Three of nine patients with ectopic atrial rhythm also had periods of junctional escape rhythm. Bradycardia for age with cycle lengths at rest of 1,180 and 1,200 ms was present in two patients (aged 7.4 and 4.9 years, respectively); both patients had an ectopic atrial rhythm, suggesting an escape mechanism.

The electrophysiologic tests of sinus node or ectopic atrial pacemaker function included: 1) corrected sinus node or ectopic atrial pacemaker recovery time; 2) ratio of sinus node recovery time to sinus cycle length or ratio of ectopic atrial pacemaker recovery time to ectopic atrial cycle length; and 3) total sinoatrial conduction time (Fig. 3). Of 21 patients with sinus rhythm during electrophysiologic testing, 11 (52%) had a prolonged corrected sinus node recovery time (>275 ms) and 5 (24%) had an abnormal ratio of sinus node recovery time to sinus cycle length of >150%. The mean corrected sinus node recovery time in the 11 abnormal studies was 441 ± 182 ms and the mean ratio of sinus node recovery time to cycle length in the 5 abnormal studies was 170% ± 22%. Total sinoatrial conduction time, as determined by either the method of Strauss et al. (23) or Narula et al. (24) in 20 patients with normal sinus rhythm, was prolonged to >200 ms in 10 (50%). In the 10 patients with an abnormal sinoatrial conduction time the mean value was 267 ± 111 ms. Of nine patients with ectopic atrial rhythm, four (44%) had a corrected pacemaker recovery time greater than that expected for the sinus node (>275 ms) and three (33%) had a ratio of pacemaker recovery time to cycle length >150%. The corrected pacemaker recovery time >275 ms in the four studies ranged from 320 to 5,380 ms (median 460) and the mean ratio of pacemaker recovery time to cycle length >150% in the three studies was 255% ± 175%. Abnormalities of sinus node conduction or automaticity were detected in 17 patients (57%). Therefore, sinus or ectopic atrial pacemaker function was entirely within the normal limits expected for the sinus node by electrophysiologic testing in only 13 patients (43%). However, to date no study patient has developed sinus node dysfunction of such

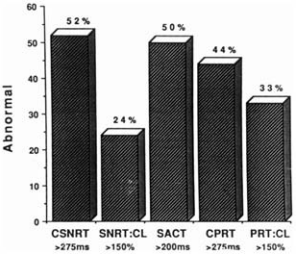

Figure 3. Frequency, after Fontan repair, of abnormalities of corrected sinus node recovery time (CSNRT), the ratio of sinus node recovery time to cycle length (SNRT : CL), total sinoatrial conduction time (SACT), corrected pacemaker recovery time (CPRT) and the ratio of pacemaker recovery time to cycle length (PRT : CL).

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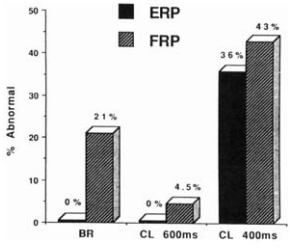

Figure 4. Frequency, after Fontan repair, of abnormalities of atrial effective (ERP) and functional (FRP) refractory periods during basal rhythm (BR) and at paced cycle lengths (CL) of 600 and 400 ms. Note the increased frequency of abnormalities at the shorter paced cycle length of 400 ms.

clinical severity to require pacemaker implantation over a mean postoperative follow-up period of 4.4 ± 1.9 years.

Atrial electrophysiologic function. Atrial effective and functional refractory periods were determined in the study patients and compared with previously reported normal values in children (25). Atrial effective refractory periods were determined in 24 patients during their basal rhythm. No patient had an abnormal effective refractory period during basal rhythm or at a paced cycle length of 600 ms. At a paced cycle length of 400 ms, 10 (36%) of the 28 patients had a prolonged effective refractory period. The functional effective refractory period was prolonged in 5 (21%) of 24 patients during basal rhythm, in 1 (4.5%) of 22 patients at a cycle length of 600 ms and in 12 (43%) of 28 patients at a paced cycle length of 400 ms (Fig. 4). Abnormalities of atrial refractoriness were most common at the shortest paced cycle length of 400 ms.

The normal electrophysiologic response of the atria during pacing is shortening of the atrial refractory period at shorter paced cycle lengths. In 3 (10%) of the 30 patients, this response was reversed, with prolongation of the refractory period at shorter cycle lengths.

Atrial arrhythmias occurred in 11 patients in response to programmed electrical stimulation. Three patients developed nonsustained atrial arrhythmia lasting three to eight beats in response to double premature atrial stimulation. Of these three patients, two had nonsustained intraatrial reentry (cycle lengths 210 and 220 ms with the ECG appearance of atrial flutter) and one patient had supraventricular tachycardia from an undetermined mechanism. The two patients with nonsustained intraatrial reentry also had delayed intraatrial conduction. Two of the three patients with nonsustained inducible atrial arrhythmia had a clinical history of sustained supraventricular tachycardia or atrial flutter before electrophysiologic testing and were taking digoxin at the time of study; their use of this drug may explain the nonsustained nature of the induced tachycardias.

Sustained supraventricular arrhythmias were reproducibly induced in eight other patients by programmed electrical stimulation. Intraatrial reentry with the ECG appearance of atrial flutter but with a cycle length of 200 to 410 ms (mean 262 ± 70, median 250) was induced in seven patients. Delayed intraatrial conduction was present in six of these seven patients. In one additional patient inducible sustained AV reciprocating tachycardia occurred by way of a concealed posteroseptal bypass tract. Five patients with inducible sustained tachycardia had a clinical history of this arrhythmia before the electrophysiologic study.

Patients studied for evaluation of recurrent supraventricular tachycardia. Before electrophysiologic study, nine patients (30%) had a clinical history of previous supraventricular tachycardia (n = 4) or atrial flutter (n = 5). In 5 patients specifically studied for evaluation of such recurrent or symptomatic arrhythmia the arrhythmia was induced in the electrophysiologic laboratory and was sustained in four of the five patients. Of four patients with a clinical history of supraventricular tachycardia (n = 3) or atrial flutter (n = 1) who were asymptomatic while receiving digoxin, two had induction of this same arrhythmia by programmed electric stimulation and two had no arrhythmia induced during drug therapy. Of the 21 patients without a prior history of supraventricular arrhythmia, three (14%) had inducible intraatrial reentry with programmed stimulation. To date, none of these three patients have developed clinical arrhythmia 2.5, 3.5 and 4.5 years, respectively, after electrophysiologic study.

Atrioventricular node and His–Purkinje function. The AH and HV intervals were recorded in 11 patients; 1 of the 11 had a prolonged AH interval (>120 ms); 1 had a prolonged HV interval (>55 ms) and 1 had prolongation of both AH and HV intervals. The paced cycle length producing anterograde AV block was abnormal (>450 ms) in two patients. One additional patient had complete heart block that was present preoperatively. Ventriculoatrial (VA) conduction was present in 15 of the 20 patients who had ventricular stimulation. In these patients the paced cycle length producing retrograde VA block was 367 ± 129 ms.

Ventricular electrophysiologic function. Programmed ventricular stimulation was used to evaluate ventricular electrophysiologic function in 20 patients. No patient developed sustained ventricular arrhythmia. Four patients had single repetitive ventricular responses; two of these four had intraventricular reentry and two had bundle branch reentry. One of the patients with intraventricular reentry had frequent (44) beats/h) premature ventricular complexes on Holter analysis.

Hemodynamic-electrophysiologic correlates. The mean right atrial pressure in patients with sinus node or ectopic atrial pacemaker dysfunction by electrophysiologic testing was 14.0 ± 3.8 mm Hg compared with 12.3 ± 2.9 mm Hg in patients with normal function (p = NS). Similarly, the mean right atrial pressure in patients with abnormalities of atrial refractoriness was not statistically different from that of

patients with normal atrial refractoriness (12.7 ± 2.7 mm Hg compared with 13.2 ± 4.2 mm Hg). However, the mean cardiac index of patients with ventricular ectopic rhythm either on Holter monitoring or by electrophysiologic testing was significantly lower than in patients without ventricular ectopic complexes (2.0 ± 1.0 versus 2.6 ± 0.5 liters/min per m^2) ($p = 0.05$). Other hemodynamic variables such as ventricular end-diastolic pressure, aortic saturation and right atrial pressure were not statistically different between the groups of patients with and without ventricular ectopic complexes.

Discussion

Little electrophysiologic information has been reported previously in patients following Fontan repair. Our study found electrophysiologic abnormalities including sinus node dysfunction, atrioventricular block, prolongation of atrial refractoriness, delayed intraatrial conduction and inducible atrial and supraventricular arrhythmias.

Abnormalities of sinus node function after Fontan repair. Although they were detected in 57% of our 30 patients by electrophysiologic testing, clinical sinus node disease was mild to moderate in severity. This observation may be a function of the timing of the electrophysiologic studies because clinical sinus node dysfunction may be a late progressive occurrence, which has been seen after other types of atrial surgery, such as the Mustard repair for transposition of the great arteries and repair of atrial septal defects (26–29). During the Fontan repair, extensive surgery is performed in the perinodal region, which may result in direct intraoperative damage to the sinus node or in progressive scarring along suture lines with late occurrence of sinus node dysfunction. The mild to moderate sinus node abnormalities detected by electrophysiologic testing in our patients may be an early warning of future clinical problems. Because the predictive value of sinus node testing in this group of patients is currently unknown, close clinical follow-up is indicated to determine which patients, if any, will develop clinically significant sinus node disease.

Abnormalities of atrial refractoriness and conduction after Fontan repair. Delayed intraatrial conduction, described in patients after the Mustard repair for transposition of the great arteries, has been related to interruption of internodal conduction pathways or to extensive scarring in the atrium at suture lines (30–32). Extensive intraatrial surgery is performed during the Fontan repair. Intraoperative interruption of normal conduction pathways may occur, as well as progressive scarring along suture lines, possibly altering the anisotropic properties of the atrial tissue. The delayed conduction to the AV junction, and low lateral and low medial regions of the right atrium after Fontan repair corresponds to the inferior aspect of the intraatrial baffle and is an area predictably affected by this surgical procedure.

Alterations in postoperative hemodynamics, especially right atrial hypertension that is uniformly present after Fontan repair, may produce prolongation of the atrial refractory period. Elevated right atrial pressure, which has been shown to prolong the atrial refractory period in an experimental model (33), may provide a hemodynamic substrate for abnormalities of atrial refractoriness noted in some patients after Fontan repair.

Reentrant supraventricular tachycardias. Although supraventricular tachycardia has been reported to occur frequently after Fontan repair, our study has indicated that intraatrial reentry is the most common mechanism of this supraventricular tachycardia. Prolonged atrial conduction and right atrial enlargement have been reported to increase the incidence of atrial flutter in clinical and experimental models (34,35). The combination of prolonged atrial conduction and right atrial enlargement, which is common after Fontan repair, may increase the probability that reentrant atrial arrhythmias will develop.

The electrophysiologic substrates for the development of reentrant tachycardias are dispersion of refractoriness, areas of slowed conduction with unidirectional block and either an anatomic or a functional circuit. In our study, abnormalities of atrial refractoriness were more frequently noted at the faster paced cycle lengths. Although multiple sites were not tested to determine if dispersion of atrial refractoriness was present, under clinical conditions that increase the heart rate it is possible that affected regions may develop a prolonged atrial refractory period providing areas of slowed conduction and unidirectional block with subsequent development of reentrant arrhythmias.

Comparison of Fontan and Mustard surgical procedures. The electrophysiologic findings after Fontan repair are similar to those reported after Mustard repair for transposition of the great arteries. These abnormalities include sinus node dysfunction, delayed intraatrial conduction, abnormalities of atrial refractoriness and the propensity to develop reentrant atrial arrhythmias (31,32,36). Sinus node dysfunction, as detected by electrophysiologic testing in patients after Mustard repair, has been reported (36) to occur in up to 86% of patients studied, with severe dysfunction present in 27%. In our postoperative patients after Fontan repair, the incidence of sinus node dysfunction was lower (57%) and no patient exhibited severe sinus node dysfunction. The explanation for these differences may relate to the more extensive surgery in the perinodal area required by the Mustard operation, as well as timing of postoperative electrophysiologic testing. Among patients after Mustard repair a higher incidence of severe sinus node dysfunction has been found in patients with than in patients without inducible atrial arrhythmias. Because none of our patients with Fontan repair had severe sinus node disease, this association was not seen in our study. Delayed intraatrial conduction, especially to the low right atrial sites, was noted in 76% of our patients compared with 90% of postoperative patients with Mustard repair (32).

In both surgical procedures suture lines present in the low right atrium result in scar formation and potential areas of

anisotropy, explaining resultant delays in intraatrial conduction to these sites. Abnormalities of atrial refractoriness are less common after Mustard repair (31.36) than after Fontan repair (2% to 20% versus 43%). This difference may be due to the altered hemodynamics after Fontan repair; specifically, right atrial hypertension may result in prolongation of the atrial refractory period and be responsible for the greater frequency of this electrophysiologic abnormality after the Fontan procedure. Finally, the similar combination of delayed intraatrial conduction and abnormalities of atrial refractoriness with resultant unidirectional block may provide the electrophysiologic substrates for the development of reentrant atrial arrhythmias noted after both procedures.

Prevention of postoperative arrhythmias. To date, the optimal surgical approach for avoidance of postoperative arrhythmias after Fontan repair has not been determined. Modifications of the procedure have been reported to diminish the frequency of such arrhythmias but have not abolished them (37,38). In our series none of the different surgical procedures were associated with a greater or lesser incidence of arrhythmias; however, the number of patients in each group was too small to reach statistical significance.

Role of digoxin and other medications. Although cardiac medications were taken at the time of electrophysiologic testing by 20 of the 30 patients in our study, the presence of digoxin was not associated with any consistent abnormality of automaticity, conduction or refractoriness and similar arrhythmias developed in patients receiving and not receiving medications. Although it cannot be determined whether these medications prevented induction of arrhythmias, they did not appear to be responsible for any of the electrophysiologic abnormalities noted.

Predictive value of electrophysiologic testing. The predictive value of electrophysiologic testing is unknown in patients after Fontan repair. In a large series of patients who underwent Mustard repair for transposition of the great arteries (36), 35% of those with clinical episodes of atrial flutter had the arrhythmia documented first in the electrophysiologic laboratory with subsequent development of the spontaneous clinical arrhythmia. Although the electrophysiologic effects after surgery would be expected to be similar in patients after Fontan repair, continued surveillance is necessary to determine which patients will develop clinically significant arrhythmia.

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