Comparison of Anatomic Isthmus Block With the Modified Right Atrial Maze Procedure for Late Atrial Tachycardia in Fontan Patients

Barbara J. Deal, MD; Constantine Mavroudis, MD; Carl L. Backer, MD; Scott H. Buck, MD; Christopher Johnsrude, MD

Background—Late atrial reentry tachycardia (AT) after Fontan repair is common, with limited efficacy of medical therapy in preventing AT recurrence. In this study, two approaches to surgical arrhythmia ablation in patients with refractory AT undergoing Fontan revision are compared: cryoablation of the inferomedial right atrium (RA), and a more extensive modified RA maze procedure designed to eliminate all potential RA reentrant circuits.

Methods and Results—Fontan revision was performed in 23 patients with AT, using inferomedial RA cryoablation (Group 1, n=8) and modified RA maze procedure (Group 2, n=15). There was no difference in age at initial Fontan, age at Fontan revision, age at onset of AT, or number of failed antiarrhythmic medications. Patients underwent preoperative, intraoperative, and postoperative electrophysiological studies. Thirty-eight different tachycardia circuits were induced in preoperative studies with 3 major areas of RA involvement: the lower lateral RA, the atrial septum, and the inferomedial RA. At postoperative electrophysiological study, AT was inducible in 62% of Group 1 patients but only 7% of Group 2 patients (P<0.02). With mean follow-up of 43 months, 5 of 8 patients in Group 1 experienced AT recurrence compared with none in Group 2 (P<0.001). There was no significant difference in length of hospital stay or complication rate comparing the two groups.

Conclusion—Modified RA maze procedure is superior to anatomic isthmus block in treating reentrant AT in postoperative Fontan patients. The modified RA maze has eliminated AT recurrence at mid-term follow-up with low morbidity and mortality. (Circulation. 2002;106:575-579.)

Key Words: atrial tachycardia ■ Fontan operation ■ arrhythmia surgery

The incidence of late atrial tachycardia after the Fontan operation approaches 30% to 50% after 5 years follow-up.1-7 Radiofrequency catheter ablation procedures are acutely successful in 50% to 70% of atrial reentry tachycardia (AT) circuits,8-13 with approximately 50% recurrence within the first 6 months after ablation.12,13 In addition, persistence of hemodynamic abnormalities not addressed during ablation procedures may leave the patient at risk for atrial thrombosis, stroke, and declining ventricular function.

Surgical revision of the Fontan anastomosis has resulted in improved clinical status, but tachycardia recurs in at least 75% of AT patients during short-term follow-up.8,14-21 An earlier study assessing the impact of arrhythmia surgery at the time of Fontan conversion demonstrated that ablation of the inferomedial right atrium (RA) resulted in significant reduction in symptomatic AT.22 However, because a small number of patients with limited RA ablation continued to experience AT, we hypothesized that a modified RA maze procedure designed to isolate all identified tachycardia circuits would eliminate AT recurrence. This retrospective study compares the efficacy of two approaches to arrhythmia ablation in patients with refractory AT undergoing Fontan conversion.

Methods

Between December 1994 and January 2001, 39 patients underwent Fontan conversion with arrhythmia surgery at Children’s Memorial Hospital. Of these patients, 37 had recurrent, refractory atrial tachycardia preoperatively: 23 patients with AT and 14 patients with atrial fibrillation. The 23 patients with AT form the basis for this report; patients in this series were included in our prior reports of surgical techniques and operative outcomes for Fontan conversion.23-24 The initial 8 patients, Group 1, underwent cryoablation limited to the inferomedial RA isthmus (between coronary sinus and inferior vena cava, and right atrioventricular valve when present), based on the hypothesis that ablation of this isthmus was critical to the reentrant circuits. The subsequent 15 patients, Group 2, underwent more extensive RA cryoablation, designed to eliminate all potential RA reentrant circuits. Chart review was performed with the approval of our Institutional Review Board.
tachycardia was not inducible. Early activation, scar potentials, and epicardially in 12 patients; one patient (No. 11) with left atrial anatomy, sequential recordings were obtained from 16 atrial sites. In the initial 10 patients, and due to the risk of air emboli, (CardioMapp, GE Medical System, Prucka Center) was placed over the last 4 patients, a simultaneous epicardial 256-electrode array sock determined using pace-mapping and entrainment techniques. sequence relative to the surface P wave, and critical exit sites were tachycardia. AT circuits were evaluated using atrial activation morphology and cycle length was compared with the clinical 3000, Endocardial Solutions) currently used. Tachycardia P wave for cardiac transplantation (1 in Group 1; 2 in Group 2).

**Patient Characteristics**

<table>
<thead>
<tr>
<th>Diagnosis (n, %)</th>
<th>Isthmus Block (n=8)</th>
<th>Modified Maze (n=15)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>TA</td>
<td>4</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>DILV</td>
<td>2</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>SV</td>
<td>2</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>DORV, mitral atresia</td>
<td>0</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>HLHS</td>
<td>0</td>
<td>2</td>
<td></td>
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<tr>
<td>Hypoplastic RV, PS</td>
<td>0</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Age at 1st Fontan, median</td>
<td>6.25±2.5</td>
<td>6.3±3.9</td>
<td>NS</td>
</tr>
<tr>
<td>Age at SVT onset, mean</td>
<td>11.1±3.4</td>
<td>10.6±7.4</td>
<td>NS</td>
</tr>
<tr>
<td>No. failed medications</td>
<td>2.1±1.4</td>
<td>3.4±1.6</td>
<td>NS</td>
</tr>
<tr>
<td>Catheter results, mean</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RAp, mm Hg</td>
<td>16.9±2.8</td>
<td>14.1±2.3</td>
<td>0.017</td>
</tr>
<tr>
<td>PAp, mm Hg</td>
<td>15.5±4.7</td>
<td>12.3±2.6</td>
<td>0.05</td>
</tr>
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<td>LVEDP, mm Hg</td>
<td>9.0±3.9</td>
<td>7.5±1.7</td>
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<tr>
<td>Cl, L/min−m²</td>
<td>2.1±0.6</td>
<td>2.8±0.7</td>
<td>0.026</td>
</tr>
<tr>
<td>Age at reoperation, mean</td>
<td>15.3±3.8</td>
<td>16.6±7.7</td>
<td>NS</td>
</tr>
</tbody>
</table>

CI indicates cardiac index; DILV, double inlet left ventricle; DORV, double outlet right ventricle; HLHS, hypoplastic left heart syndrome; NS, not significant; PAp, mean pulmonary artery pressure; PS, pulmonary stenosis; RV, right ventricle; RAp, mean right atrial pressure; SV, single ventricle; SVT, supraventricular tachycardia; and TA, tricuspid atresia.

**Electrophysiology Studies**

All patients underwent electrophysiological studies preoperatively, intraoperatively, and postoperatively before hospital discharge. Preoperative intracardiac studies were performed at the time of cardiac catheterization. AT was induced using programmed stimulation in the baseline state, or during isoproterenol infusion at 1 to 2 mcg/min. Contact mapping with a minimum of 2 multipolar electrode catheters was initially performed, with a noncontact mapping system (EnSite 3000, Endocardial Solutions) currently used. Tachycardia P wave morphology and cycle length was compared with the clinical tachycardia. AT circuits were evaluated using atrial activation sequence relative to the surface P wave, and critical exit sites were determined using pace-mapping and entrainment techniques.

Intraoperative activation mapping was performed after cannulation for bypass. A reference electrode was sutured to the superior RA surface, and AT was induced. A hand-held roving electrode was used in the initial 18 patients: using a standardized grid of the atrial anatomy, sequential recordings were obtained from 16 atrial sites. In the last 4 patients, a simultaneous epicardial 256-electrode array sock (CardioMapp, GE Medical System, Prucka Center) was placed over the atrial surface. Activation mapping was performed endocardially in the initial 10 patients, and due to the risk of air emboli, epicardially in 12 patients; one patient (No. 11) with left atrial tachycardia was not inducible. Early activation, scar potentials, and zones of slow conduction were directly compared with anatomic landmarks.

Postoperative studies were performed after chest tube removal and discontinuation of inotropic support. All patients received an atrial pacemaker that was used to deliver programmed stimulation using atrial incremental pacing to a minimum paced cycle length of 200 ms, and atrial extrastimulus testing using single, double, and triple extrastimuli to a minimum interval of 200 ms. Pacing was performed in the baseline state, and using isoproterenol infusion at 1 to 2 mcg/min.

**Ablation Strategy**

Group 1 cryoablation lesions were delivered between the os of the coronary sinus and the inferior vena cava, and between the right atrioventricular groove or annulus and the inferior vena cava orifice (Figure 1). The subsequent 15 patients underwent a modified RA maze procedure (Figure 2) designed to interrupt all potential RA reentrant circuits identified. The modified RA maze procedure consists of the following: (1) an incision from superior vena cava to inferior vena cava, (2) resection of RA appendage, (3) resection of a large portion of the anterior RA wall, including the presumed location of the sinoatrial node and the anterior portion of the superior vena cava, and (4) multiple cryoablation lesions (a) between the coronary sinus os and inferior vena cava, (b) between inferior vena cava and right atrioventricular annulus or groove, (c) from the posterior rim of the newly created atrial septal defect across the crista terminalis to the lateral atrial wall, and (d) between the superior rim of the atrial septal defect to the base of the resected RA appendage. For 3 patients with prior lateral tunnel repairs and tachycardia mapped to the left atrium, the atrial partition was resected, allowing direct exposure of the coronary sinus os within the pulmonary venous atrium. Cryoablation lesions were delivered at −60°C for 90 seconds each, using either multiple overlapping circular lesions (5- or 15-mm probe) or a linear 3-mm lesion (Frigitronics). The time necessary to complete cryoablation lesions was recorded for each

**Figure 1.** Inferomedial right atrial cryoablation sites. Arrows indicate ablation line from coronary sinus (CS) os to inferior vena cava (IVC), and tricuspid valve (TV) annulus or groove to IVC; RAA, right atrial appendage; SVC, superior vena cava.

**Figure 2.** Modified right atrial (RA) maze cryoablation sites. Inferomedial RA cryoablation as in Figure 1, plus line from posterior edge of created atrial septal defect (ASD) to resected edge of lateral atrial wall, and superior rim of ASD to base of resected RA appendage.
patient. Atrial wall thicknesses were measured and recorded after resection.

**Fontan Conversion**

After the arrhythmia surgery, patients underwent Fontan conversion using the surgical technique of Fontan conversion reported earlier.\(^ {22-24} \) Associated surgical procedures included pulmonary arterio-pleasty in 6 patients, atrioventricular valve repair in 4 patients, and valve replacement in 1 patient.

**Pacemaker Implantation**

The initial 10 patients received atrial antitachycardia pacemakers (Intermedics, Intertach II). Due to the benefits of atrial rate-responsiveness and subsequent generator unavailability, 10 subsequent patients received atrial rate-responsive pacemakers and 3 patients dual-chamber pacemakers (Medtronic, Thera SR or DR). Two of these 3 patients had preexisting dual-chamber systems; the third patient had transient atrioventricular dissociation. Transmural endocardial leads (Medtronic model No. 4965) were placed in 15 patients,\(^ {25} \) with epicardial steroid-eluting leads (Medtronic model No. 10295B) implanted in 8. All patients received chronic anticoagulation with warfarin postoperatively.

**Follow-Up**

Patient follow-up consisted of review of symptoms, electrocardiograms, continuous 24-hour electrocardiographic monitoring, and pacemaker analysis during 4 visits in the first postoperative year, and at minimum yearly intervals thereafter. Symptomatic tachycardia recurrence was recorded, including the need for hospitalization or chronic antiarrhythmic therapy.

**Statistical Analysis**

Continuous variables are summarized as mean±standard deviation and categorical variables as frequencies. All surgical and other patient characteristics are compared between the two groups: \( t \) test was used for continuous variables that satisfied the normality assumption and the nonparametric Wilcoxon test was used for data that deviated from normality. Chi-square test or Fisher’s Exact test was used to compare frequency distribution of categorical variables. Time to AT recurrence was described using Kaplan-Meier curves and compared between groups using Log-Rank test. Patient characteristics that differ between the two groups were controlled for in the hazard-free survival models. Univariate analysis was undertaken to test the effect of the various simultaneous interventions. All conclusions were made at 0.05 level of significance.

**Results**

**Patient Characteristics**

The Table summarizes clinical variables of the 2 groups. The median age at Fontan conversion was 10.9 years (range 2.6 to 33.5 years), with a mean postoperative interval of 7.9 years. There was no significant difference in age at initial Fontan, age at onset AT, number of antiarrhythmic medications, and age at Fontan conversion. Group 1 patients had significantly higher mean RA pressures (\( P=0.017 \)) and lower cardiac indices (\( P=0.021 \)), reflecting the more significant hemodynamic compromise in the initial patients undergoing Fontan conversion.

**Preoperative Electrophysiological Studies**

A total of 35 AT circuits were induced, but not all could be adequately mapped due to hemodynamic instability or the transient nature of certain tachycardias; 27 sustained AT circuits were adequately mapped in 22 patients. Critical AT sites were identified in 3 major areas of the RA in 22 circuits: the superior or medial edge of the atrial septum in 9, the lateral RA wall in 8, and the inferomedial RA isthmus in 5 circuits (Figure 3). Exit sites of 3 AT circuits were localized to the left atrium. Only 2 exit sites were localized to the atriopulmonary anastomosis area.

**Intraoperative Atrial Mapping**

Intraoperative studies were limited to activation mapping of 1 to 3 atrial tachycardias, in contrast to more lengthy preoperative studies. Of 10 patients undergoing endocardial mapping, earliest activation was the inferomedial RA in 6, septal in 3, and high lateral RA in 1. Cryoablation in the atrial isthmus terminated AT in 6 patients. Epicardial mapping was performed in 12 patients and was concordant with the dominant clinical and preoperatively mapped AT in 11/13 circuits. In the 4 patients who had undergone prior catheter ablation procedures, there was no visible evidence of ablation lesions within the RA in 3; one patient had isolated punctate 1- to 2-mm areas of dark gray discoloration, thought to be due to ablation. Resected atrial wall thicknesses ranged from 3 to 14 mm, and averaged 8 mm.

**Operative Outcome**

The Fontan conversion consisted of an extracardiac total cavopulmonary anastomosis in 16 patients, a lateral tunnel in 6 patients, and a 1 1/2 ventricular repair in one patient (No. 13). There was no operative or perioperative mortality. Mean hospital stay was 10.2±4.6 days, with mean period of chest tube drainage equalling 9.0±6.0 days. The mean time for completion of cryoablation lesions for isthmus block was 7 minutes, versus 11 minutes for the modified RA maze procedure. Early postoperative AT developed in 1 patient in Group 1 while receiving inotropic support at 3 days postoperatively, and in no patients in Group 2. Complications included prolonged chest tube drainage >7 days in 2/8 Group 1 patients, and 8/15 Group 2 patients (\( P=NS \)), and arrhythmias in 3/15 Group 2 patients (\( P=NS \): early junctional
ectopic tachycardia in 2, and transient complete heart block in 1 patient.

**Postoperative Electrophysiological Testing**

Of Group 1 patients, 62% (5/8 patients) had inducible AT, versus 7% (1/15) of Group 2 patients ($P=0.016$). AT was inducible in the baseline state in 3 patients (using pacing cycle lengths <200 ms in 2 patients), with isoproterenol using atrial incremental pacing in 1 patient, and spontaneously during isoproterenol infusion alone in 2 patients.

**Late Postoperative Tachycardia**

Among Group 1 patients, 62% (5/8) experienced AT recurrence after hospital discharge, versus none among Group 2 patients ($P<0.001$), with median time to AT recurrence being 8 months (range 1.5 to 26 months) (Figure 3). Neither right atrial pressure ($P=0.35$) or cardiac index ($P=0.53$) had an effect on time to AT recurrence. One of 16 patients (6%) with extracardiac Fontan experienced AT recurrence compared with 4/6 (67%) with lateral tunnel Fontan ($P=0.001$). Five of twelve patients (42%) with antitachycardia pacemakers experienced AT recurrence compared with none among those with atrial rate responsive pacemakers ($P=0.035$).

**Follow-Up**

Group 1 mean follow-up duration is 68 months, and 34 months in Group 2 ($P<0.001$). However, the difference in follow-up did not account for the difference in AT recurrence, as the mean time to recurrence was 9.4 months.

Two patients (both in Group 2) have undergone cardiac transplantation for progressive ventricular dysfunction, at 9 and 33 months, respectively; the second patient was initially referred for transplantation before Fontan conversion. Both patients had a diagnosis of double outlet right ventricle.

**Discussion**

These findings demonstrate that the modified RA maze procedure is superior to anatomic isthmus block in treating AT and has essentially eliminated AT recurrences in Fontan conversion patients during midterm follow-up. Arrhythmia surgery can be safely incorporated into the surgical conversion of the Fontan anastomosis, with low mortality and morbidity.

**Comparison to Surgical Conversion Approach**

Several surgical series of Fontan conversion surgery without arrhythmia intervention have now been reported. In a meta-analysis of 60 reported patients, refractory AT was present preoperatively in 60%; during short-term follow-up, AT recurred in 76% of patients. Our prior study assessing the impact of anatomic isthmus ablation during Fontan conversion to intracardiac lateral tunnels showed a significant reduction in tachycardia recurrence when atrial isthmus cryoablation was added to the surgical strategy. However, a percentage of these patients had recurrent tachycardia, terminated by the implanted antitachycardia pacemaker. Due to the multiplicity of AT circuits in these complex patients with multiple prior surgeries, the necessity of addressing all potential RA tachycardia circuits became apparent during our early experience.

Our mapping results indicate 3 primary exit sites of mapped AT circuits: the lateral atrial wall, the perimeter of the atrial septal defect, and the inferomedial RA isthmus. The atrial isthmus is known to be an important part of typical atrial flutter circuits, and in up to 1/3 of atrial circuits in patients with other forms of operated congenital heart disease. The atriotomy site has been shown to be important to the acute initiation of AT in dog models undergoing lateral tunnel incision lines. The superior and medial rims of the atrial septal defect were critical areas in over 1/3 of our patients, with 3 circuits localized to the pulmonary venous partition adjacent to the atrial septal patch. Of note, in our series, the atriopulmonary anastomosis, although providing an anatomic obstacle, did not appear to be a critical part of the AT circuit in most patients. We believe that this modified RA maze effectively addresses all major RA reentrant circuits, while preserving intact anteroinferior to conduction. This surgery makes no attempt to preserve sinus node function, which is impaired preoperatively in many patients. Atrial pacing to avoid even mild degrees of atrial bradycardia is important to prevent the prolongation of atrial refractoriness that may contribute to the development of AT.

**Comparison to Catheter Ablation Procedures**

After acutely successful radiofrequency catheter ablation of AT in Fontan patients, approximately 50% of patients experience recurrence during 6 months of follow-up. Limitations of the catheter technique that are directly addressed by the surgical modified RA maze include restricted catheter access, distorted anatomy, inability to create complete lines of block, and the multiplicity of reentrant circuits. Due to marked atrial hypertrophy and sluggish atrial blood flow, it is extremely difficult to achieve lesions of significant depth and continuity with present transvenous catheter technology. Visible evidence of significant ablation was difficult to identify in our patients and did not extend between anatomic obstacles; patchy ablation lesions may provide increased areas of conduction delay, potentially aggravating the arrhythmia substrate. The majority of Fontan patients with atrial tachycardia have associated hemodynamic abnormalities or marked atrial dilatation, leaving them at risk for atrial thrombosis or decreased exercise tolerance after catheter ablation procedures.

**Limitations**

The primary limitation of this study is the statistical inability to analyze the independent effects of the modified RA maze, the extracardiac lateral tunnel, and the type of pacemaker on tachycardia recurrence. The number of subjects in each subgroup and the number of adverse effects (number of arrhythmia recurrences) were too small for multivariate analysis. Because our initial study demonstrated the improved outcome of isthmus ablation in patients undergoing lateral tunnel revision, the focus of this analysis was the impact of the modified RA maze versus isthmus ablation on arrhythmia recurrence. The statistical analysis was performed comparing...
these two groups; we believe that the clinical relevance of our findings and our analysis warrant this approach.

Limitations of tachycardia mapping included hemodynamic instability, the transient nature of some AT, lack of entrainment studies intraoperatively, and analysis of completion of lines of block could not be safely demonstrated after prolonged cardiac surgery. The minimal amount of atrial incisions or cryoablation lesions needed to effectively prevent AT was not assessed. Finally, the modified RA maze is effective for right atrial macroreentrant tachycardia circuits, and will not address focal atrial tachycardia or atrial fibrillation.

Implications
We believe that performance of multiple ablation procedures in Fontan patients may delay definitive surgery, and persistent tachycardia may allow for progressive decline of ventricular function, clot formation, and development of atrial fibrillation. Due to the high incidence of AT in Fontan patients, the modified RA maze surgery should be incorporated into any planned Fontan conversion surgery. The present modified RA maze surgery addresses all potential right atrial macroreentrant circuits, making preoperative and intraoperative mapping unnecessary in most cases, except to identify focal or left atrial tachycardia. We believe that the extensive intra-atrial suture burden of the intracardiac lateral tunnel contributes to the development of AT. Patients undergoing initial lateral tunnel repairs instead of extracardiac conduits may benefit from strategies to incorporate lines of block, taking into account the 3 sites of reentrant circuits and not being limited to the inferior atriotomy.

References
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