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Fontan conversion with arrhythmia surgery

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Abstract

Objective: Hemodynamic abnormalities and refractory atrial arrhythmias in patients late after the Fontan operation result in significant morbidity and mortality. We reviewed our experience with Fontan conversion and concomitant arrhythmia surgery.

Methods: Between January 1996 and February 2004, 16 patients underwent Fontan conversion and arrhythmia surgery. Mean age at the initial Fontan operation was 5.1 ± 3.5 (range: 2–15) years and mean age at Fontan conversion was 17.0 ± 5.8 (range: 6–30). The initial Fontan operations were atriopulmonary connections in 14 patients, extracardiac lateral tunnel in 1, and intracardiac lateral tunnel in 1. The types of arrhythmia included atrial flutter in 10 patients and atrial fibrillation in 3. Fontan conversion operation was performed with intracardiac lateral tunnel in 5 patients and extracardiac conduit in 11. Arrhythmia surgery included isthmus cryoablation in 10 patients and right-sided maze in 3.

Results: There has been no mortality. At Fontan conversion operation, 7 patients required permanent pacemaker. All patients have improved to New York Heart Association class I or II. With a mean follow-up of 26.9 ± 30.6 (range: 1–87) months, 16 patients had sinus rhythm, 2 patients had transient atrial flutter which was well controlled, and 2 patients required permanent pacemaker during follow-up.

Conclusions: Fontan conversion with concomitant arrhythmia surgery and permanent pacemaker placement is safe, improves New York Heart Association functional class, and has a low incidence of recurrent arrhythmias. In most patients, concomitant permanent pacemakers are needed.

Keywords: Fontan conversion; Arrhythmia surgery

1. Introduction

Fontan first constructed a completely in-series circulation with exclusion of the right ventricle, using the right atrium as a dynamic element, for correction of tricuspid atresia, and a large number of patients with functional single ventricle had variations of atrio pulmonary artery connections (APC) before the present era. Postoperative morbidities after the Fontan operation are related to follow-up duration and include right atrial dilatation, thromboembolism, atrial arrhythmias, circulatory pathway obstructions, coronary sinus hypertension, decreased ventricular function, increasing cyanosis, and protein-losing enteropathy (PLE) [1,2]. Most of these complications occur in the setting of an APC as a result of the increase in right atrial size and pressure.

The increased right atrial flow and suture load that these patients experience from the time of the original Fontan have been felt to be one cause of their atrial arrhythmias. The arrhythmias can further increase right atrial pressure, which leads to a negative cycle that leads to considerable morbidity and mortality [1–3]. Atrial arrhythmias have been reported between 22 and 50% of patients with an APC Fontan [1–3].

The Fontan operation has undergone a number of major modifications [4] leading to the present-day practice of total cavopulmonary artery connections (TCPC) using lateral tunnel [5] or extracardiac [6] techniques. Conversion of an APC Fontan to a TCPC has been attempted to alleviate complications by improving central systemic flow patterns. In recent reviews of patients undergoing Fontan revision by conversion to TCPC, half of the patients received no relief from their arrhythmia or had a new arrhythmia develop after the revision surgery [5].

When the extant group of patients with the APC Fontan operation presents an increasing incidence of complications, reconstructive and arrhythmia ablative techniques might often alleviate the common complications. Our arrhythmia ablative techniques include isthmus cryoablation and
standard right-sided maze procedure to treat atrial reentry tachycardia and atrial fibrillation, respectively. We reviewed our surgical experience with conversion to TCPC and arrhythmia circuit cryoablation.

2. Materials and methods

2.1. Patients’ profile

From January 1996 to February 2004, 16 patients underwent Fontan conversion and arrhythmia surgery. There were 7 male and 9 female patients. The mean/median ages at the initial Fontan operation were 5.1 ± 3.5 (range: 2–15) years and 4 years, respectively. The mean/median ages at the latest Fontan conversion were 17.0 ± 5.8 (range: 6–30) years and 16 years, respectively. The mean time interval between the initial Fontan operation and the eventual conversion was 11.9 ± 3.7 (range: 4–16) years. Table 1 reviews clinical characteristics of our 16 patients, including primary diagnoses, previous Fontan connections, indications for reoperation, components of the Fontan conversion, types of arrhythmia surgery, and outcome. Intracardiac anomalies were tricuspid atresia in 6 patients, right ventricle type univentricular heart in 5, double outlet right ventricle in 3, pulmonary atresia with intact ventricular septum in 1, congenitally corrected transposition of the great arteries in 1. The initial Fontan operations were mostly APCs in 14 patients (87.5%), of whom 6 had undergone modified Blalock-Taussig shunt initially, an intracardiac lateral tunnel with 2.7 mm fenestration in 1 (6.3%) after bidirectional cavopulmonary shunt, and an extracardiac epicardial lateral tunnel (EELT) in 1 (6.3%). All patients except one underwent one-stage Fontan procedure without bidirectional cavopulmonary shunt. Patients with Fontan procedure received either acetylsalicylic acid (ASA) or warfarin permanently in our hospital. Warfarin was medicated in selected cases with risk factors.

Our current indications for conversion were multiple (Table 2; Fig. 1). Preoperative symptoms included palpitation in 12 patients, dyspnea on exertion in 9, cyanosis in 7, chest pain in 4, and edema in 3.

2.2. Fontan conversion

Three types of anatomic connections were present. Fourteen patients had atrial compartmentalization with right atrial-pulmonary artery anastomosis, one had EELT in which stenosis of Fontan pathway has been developed, and one had an intracardiac lateral tunnel in which thromboclusion of lateral tunnel and inferior vena cava has been developed. All of 16 patients were converted to TCPC, of whom 11 had extracardiac and 5 had intracardiac lateral tunnel Fontan operations. In early series, the technique of Fontan conversion was lateral tunnel. Recently the technique of Fontan conversion was extracardiac conduit procedure. A liberal right atrial wall resection was performed and two patients in the entire series had a fenestration.

Careful median sternotomy and relevant dissection were followed by extracorporeal cardiopulmonary bypass and cardioplegic arrest. Standard cardiopulmonary bypass was initiated with aortic and bicaudal cannulation, and patients were cooled to moderate hypothermia and the heart was arrested. A considerable amount of atrial tissue was excised in 6 patients to markedly reduce atrial cavity size. After wide resection of the previously constructed prosthetic atrial septum, cryoablative lesions were placed. Previous right atrial suture lines were excised whenever possible. Associated surgical procedures is described in Table 3. Three patients were present moderate (1) to severe (2) atrioventricular valve regurgitation preoperatively. One patient received valve repair (mild regurgitation after operation) and 2 patients valve replacement (trivial physiologic leakage after operation). A standard total cavopulmonary artery reconstruction was accomplished with takedown of the APC (n = 14), EELT (n = 1), intracardiac lateral tunnel (n = 1). The lateral tunnel was reconstructed in 5 patients, and fenestration was made in 1. After atrial closure, all hearts were deaired and reperfused. The extracardiac type of Fontan revision was carried out using a Gore-Tex vascular graft (W. L. Gore and Associates, Flagstaff, AZ) in 11 patients with the heart beating and patient rewarming. Extensive pulmonary artery reconstruction, when necessary, was performed at this time with Gore-Tex vascular graft, and fenestration was made in 2 cases. Separation from cardiopulmonary bypass is followed by transesophageal echocardiographic assessment. The 2 patients (No. 5, 13 in Table 1) who had fenestration were severe atrioventricular regurgitation and ventricular dysfunction (ventricular end diastolic pressure = 16 and 18 mmHg) preoperatively. Both patients underwent atrioventricular valve replacement, Fontan conversion and arrhythmia surgery. Intraoperative transesophageal echocardiography showed decreased ventricular contractility and high central venous pressure (> 20 mmHg). So we made fenestrations. The dual permanent epicardial pacemaker leads were placed and attached to a generator usually placed in a subcutaneous abdominal pocket in 7 cases (Fig. 2). Patients were usually extubated the following day.

2.3. Arrhythmia surgery and permanent pacemaker implantation

The types of arrhythmia included atrial flutter in 10 patients and atrial fibrillation in 3. We have standardized intraoperative arrhythmia management by performing an isthmus block via cryoablation for patients with atrial reentry tachycardia and a modified right-sided maze procedure for patients with atrial fibrillation. Ten patients had isthmus cryoablation for atrial flutter and three had right-sided maze procedure for atrial fibrillation. In our series, isthmus block was performed via cryoablation at the area between the coronary sinus os and the inferior vena cava os and the right-sided atrioventricular valve (when present). The right atrial cryoablation lesions for modified right-sided maze procedure were made in the fashion described by Mavroudis and associates [7]. A 10-mm linear probe (CCS-200, Cooper Surgical, Shelton, CT, USA) was used, and lesions were laid down for 90 s at −70 °C in our series. Seven of 16 patients had a planned permanent epicardial pacemaker implantation at the time of Fontan conversion consistent with our protocol for management of sinus node
Table 1
The characteristics of patients at Fontan conversion with arrhythmia surgery

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Dx Initial Fontan</th>
<th>Age Initial Fontan</th>
<th>Interval</th>
<th>Age at redo</th>
<th>Indications</th>
<th>Operations</th>
<th>F/U: NYHA Fc, anti-arrhythmic Mx, arrhythmia</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>RV type SV</td>
<td>AP</td>
<td>7.6</td>
<td>7.4</td>
<td>15.1</td>
<td>Huge RA, Thrombosis in RAA, AF</td>
<td>Conversion to LT, Isthmus cryoablation Conversion to LT, RA reduction, Isthmus cryoablation PPM insertion</td>
</tr>
<tr>
<td>2</td>
<td>TA</td>
<td>AP</td>
<td>2.9</td>
<td>3.6</td>
<td>6.6</td>
<td>R→L shunt</td>
<td>PPM insertion Conversion to LT, Angioplasty Conversion to LT, PA angioplasty Isthmus cryoablation</td>
</tr>
<tr>
<td>3</td>
<td>cc-TGA</td>
<td>AP</td>
<td>4.2</td>
<td>9.3</td>
<td>13.5</td>
<td>R→L shunt, AF</td>
<td>Conversion to LT, PA angioplasty Isthmus cryoablation</td>
</tr>
<tr>
<td>4</td>
<td>TA</td>
<td>AP</td>
<td>1.9</td>
<td>8.7</td>
<td>10.6</td>
<td>R→L shunt, AF</td>
<td>Conversion to LT (fene), AVV replacement, RA reduction, PA angioplasty, Rt. sided maze, PPM insertion</td>
</tr>
<tr>
<td>5</td>
<td>RV type SV</td>
<td>AP</td>
<td>3.6</td>
<td>14.9</td>
<td>18.5</td>
<td>Huge atrium, AVVR, Vent. dysfunction, AF</td>
<td>Conversion to LT, Isthmus cryoablation</td>
</tr>
<tr>
<td>6</td>
<td>PAIVS</td>
<td>AP</td>
<td>2.1</td>
<td>12.1</td>
<td>14.2</td>
<td>RPA thrombus, RA dilatation, RLPV compression, AF</td>
<td>Conversion to ECC, RA reduction, Isthmus cryoablation</td>
</tr>
<tr>
<td>7</td>
<td>DORV</td>
<td>AP</td>
<td>15.4</td>
<td>15.2</td>
<td>30.6</td>
<td>PV compression, Huge RA, PA thrombus, AF</td>
<td>Conversion to ECC, RA reduction, Isthmus cryoablation PPM insertion</td>
</tr>
<tr>
<td>8</td>
<td>DORV</td>
<td>AP</td>
<td>7</td>
<td>10.9</td>
<td>17.9</td>
<td>LVOTO, RAE, NYHA class IV, AF</td>
<td>Conversion to ECC, VSD widening, Subaortic m. resection, PA angioplasty, Rt. sided maze, PPM insertion</td>
</tr>
<tr>
<td>9</td>
<td>TA</td>
<td>AP</td>
<td>8.9</td>
<td>16.3</td>
<td>25.2</td>
<td>RA thrombus, RAE, AF</td>
<td>Conversion to ECC, Isthmus cryoablation PPM insertion</td>
</tr>
<tr>
<td>10</td>
<td>RV type SV</td>
<td>AP</td>
<td>5.8</td>
<td>14.1</td>
<td>19.9</td>
<td>PLE, R→L shunt, AF, SN dysfunction</td>
<td>Conversion to ECC, Isthmus cryoablation PPM insertion</td>
</tr>
<tr>
<td>11</td>
<td>TA</td>
<td>AP</td>
<td>2.9</td>
<td>13.6</td>
<td>16.5</td>
<td>RAE, RA thrombus, Rt. PV compression, SN dysfunction, AF</td>
<td>Conversion to ECC, RA reduction, Isthmus cryoablation PPM insertion</td>
</tr>
<tr>
<td>12</td>
<td>RV type SV</td>
<td>AP</td>
<td>5.3</td>
<td>15.6</td>
<td>20.9</td>
<td>RA thrombus, RAE, AVVR, RV dysfunction</td>
<td>Conversion to ECC, AVV repair</td>
</tr>
<tr>
<td>13</td>
<td>RV type SV</td>
<td>EELT</td>
<td>3.3</td>
<td>11</td>
<td>14.3</td>
<td>AVVR, Path stenosis, Vent. dysfunction, AF</td>
<td>Conversion to ECC (fene), AVV replacement, Isthmus cryoablation PPM insertion</td>
</tr>
<tr>
<td>14</td>
<td>TA</td>
<td>AP</td>
<td>5.9</td>
<td>14.9</td>
<td>20.9</td>
<td>Pulmonary TE, RA thrombus, RA dysfunction, AF</td>
<td>Conversion to ECC, RA reduction, PA angioplasty, CS unroofing, Rt. sided maze,</td>
</tr>
<tr>
<td>15</td>
<td>DORV</td>
<td>AP</td>
<td>2.4</td>
<td>15.7</td>
<td>18.1</td>
<td>RAE, Vent. dysfunction, AR, AF</td>
<td>Conversion to ECC, AVV, Isthmus cryoablation</td>
</tr>
<tr>
<td>16</td>
<td>TA</td>
<td>LT (fenes)</td>
<td>2.9</td>
<td>7.9</td>
<td>10.9</td>
<td>Thromboembolus of Fontan pathway</td>
<td>Conversion to ECC, Thromboembolomy in IVC</td>
</tr>
</tbody>
</table>

LT, lateral tunnel; fenes, fenestration; R→L, right to left; AF, atrial flutter; EELT, extracardiac epicardial lateral tunnel; AP, atrio-pulmonary Fontan; AVVR, atrioventricular valve regurgitation; Vent., ventricular; AF, atrial fibrillation; AVV, atrioventricular valve; PPM, permanent pacemaker; RLPV, right lower pulmonary vein; ECC, extracardiac conduit; LVOTO, left ventricular outflow tract obstruction; RA, right atrial enlargement; PLE, protein-losing enteropathy; SN, sinus node.
rhythm, and the lack of venous access to the heart in these patients, which would require obligatory thoracotomy at a future time. Our protocol was to maintain a regular paced atrial rhythm to prevent bradycardia and to decrease atrial extrasystoles, which are substrates for atrial reentry tachycardia.

2.4. Follow-up monitoring

All patients received follow-up on at least a yearly basis with physical examination, echocardiogram, electrocardiogram, continuous 24-h electrocardiographic monitoring, and pacemaker analysis. Clinical status was assessed on the basis of the functional classification of the NYHA. Arrhythmia assessment included a review of clinical history, electrocardiograms, and 24-h continuous Holter monitoring. The average follow-up time was 27.1±30.6 months (range 1–87 months) and was complete. All patients were placed on lifelong warfarin sodium (Coumadin) therapy.

2.5. Statistical analysis

Statistical analyses were performed using SPSS version 10.0 software (SPSS, Inc., Chicago, IL, USA). All results were expressed as mean±standard deviation. The improvement in NYHA class after operation was analyzed by the Wilcoxon signed ranks test, and a value of *P* less than 0.05 was considered statistically significant. The follow-up status of patients was determined by retrospective review of hospital records or by telephone interviews.

3. Results

No perioperative death occurred in a patient undergoing Fontan conversion. Resternotomy was accomplished safely and without morbidity in all patients. Conversion to TCPC resulted in a favorable hemodynamic result. Patients were usually extubated the following day (extubation on postoperative hour 16.1±11.1 h, range 2–44 h, median 13 h). Chest tube output was favorable in most cases (removal of the last chest tube on postoperative day 12.1±7.0 days, range 5–24 days, median 9 days). The average hospital stay was 19.1±6.9 days (range 10–34 days, median 17 days). Four patients had pleural effusion that required thoracic drainage for more than two weeks postoperatively. Two patients had transient seizure without sequelae, one had diaphragmatic palsy (Fig. 2), and one required reexploration for bleeding.

Atrial arrhythmia was not present in any patient at the time of discharge. There was an overall recurrence of atrial arrhythmia in two of sixteen patients (12.5%) with a mean

### Table 2

<table>
<thead>
<tr>
<th>Indications</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial arrhythmia</td>
<td>12 (75.0)</td>
</tr>
<tr>
<td>Thrombus</td>
<td>8 (50.0)</td>
</tr>
<tr>
<td>Ventricular dysfunction</td>
<td>5 (31.3)</td>
</tr>
<tr>
<td>Pulmonary venous compression</td>
<td>3 (18.8)</td>
</tr>
<tr>
<td>AV valve regurgitation</td>
<td>3 (18.8)</td>
</tr>
<tr>
<td>Residual R-L shunt</td>
<td>3 (18.8)</td>
</tr>
<tr>
<td>Pulmonary thromboembolism</td>
<td>1 (6.3)</td>
</tr>
<tr>
<td>Fontan pathway stenosis</td>
<td>1 (6.3)</td>
</tr>
<tr>
<td>LVOTO</td>
<td>1 (6.3)</td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td>1 (6.3)</td>
</tr>
<tr>
<td>Protein-losing enteropathy</td>
<td>1 (6.3)</td>
</tr>
<tr>
<td>Total</td>
<td>16 (100)</td>
</tr>
</tbody>
</table>

AV, atrioventricular; R-L, right to left; LVOTO, left ventricular outflow tract obstruction.

### Table 3

<table>
<thead>
<tr>
<th>Combined procedures with Fontan conversion</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. (%)</td>
</tr>
<tr>
<td>--------------------------------------------</td>
</tr>
<tr>
<td>Thrombectomy</td>
</tr>
<tr>
<td>PPM insertion</td>
</tr>
<tr>
<td>PA angioplasty</td>
</tr>
<tr>
<td>RA reduction plasty</td>
</tr>
<tr>
<td>Fenestration</td>
</tr>
<tr>
<td>AV valve replacement</td>
</tr>
<tr>
<td>AV valve repair</td>
</tr>
<tr>
<td>Aortic valve repair</td>
</tr>
<tr>
<td>VSD widening and subaortic muscle resection</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

PPM, permanent pacemaker; PA, pulmonary artery; RA, right atrium; AV, atrioventricular; VSD, ventricular septal defect.
follow-up of 27.1 ± 30.6 months (range 1–87 months). Atrial flutter recurred 1 month after discharge in one (No. 9 in Table 1) of ten patients undergoing isthmus cryoablation for atrial flutter. It was easily controlled by cardioversion and the patient maintained sinus rhythm for 10 months’ follow-up with anti-arrhythmic medication (sotalol). Of three patients undergoing a modified right-sided maze procedure for atrial fibrillation, atrial flutter had recurred in one (No. 5 in Table 1) 1 month after discharge. It was easily controlled by medication, and the patient maintained paced sinus rhythm with 28 months’ follow-up with anti-arrhythmic medication (sotalol). During follow-up of 28.6 ± 32.2 months, all patients maintained regular rhythm of atrioventricular synchrony, 2 patients remain with antiarrhythmic medications.

Epicardial permanent pacemaker leads were placed in seven patients because of sinus node dysfunction. Four patients with sinus bradycardia had a DDD pacemaker and three had a DDD-R pacemaker placed after arrhythmia circuit cryoablation. During follow-up, two patients needed a permanent DDD and DDD-R pacemaker implantation for management of sinus node dysfunction 6 months and 12 days after operation, respectively.

There was no late death. The mean duration of follow-up in the 16 survivors was 27.1 months (standard deviation 30.6 months, range 1 month to 87 months). The symptoms such as palpitation, dyspnea on exertion, cyanosis, chest pain, and edema were improved. All patients are currently classified NYHA class I or II, with resumption of normal activities, full-time employment, school, or university. No patient has worsening NYHA functional class. There was a significant improvement in NYHA class (P<0.001, Wilcoxon signed ranks test): class I, 13 patients; and class II, 3 patients (Table 4).

The one patient in our series with PLE (No. 10 in Table 1) has shown remarkable improvement. Before operation, she had to receive regular albumin infusion for generalized edema, ascites and dyspnea. She no longer requires heparin and albumin transfusions. Her last stool alpha-1-antitrypsin level was normal. She was in NYHA class III before the operation and improved to NYHA class I after the procedure.

The patient with a history of recurrent pulmonary emboli (No. 14 in Table 1) has seen significant relief from recurrent pulmonary emboli and atrial fibrillation. He has not had a thromboembolic event in the more than 1.5 years since the surgery and his atrial fibrillation is well controlled.

The patient with lateral tunnel (No. 16 in Table 1) had thromboocclusion of Fontan pathway, and took conversion to extracardiac conduit with thromboembolectomy. His exercise tolerance has improved substantially, and follow-up echocardiography showed good Fontan pathway and flow pattern. He had the deficiency of coagulation factor II, VII, IX, X, and protein C, and has been placed on strict, lifelong warfarin sodium (Coumadin) therapy.

The patient with EELT (No. 13 in Table 1) had Fontan pathway stenosis, severe atrioventricular valve regurgitation, and ventricular dysfunction. He was intravenous inotropic dependent preoperatively. He took conversion to extracardiac conduit after take down of EELT and atrioventricular valve replacement. He was in NYHA class IV before operation and improved to NYHA class II after the procedure.

4. Discussion

Although Fontan operation ameliorated the life expectancy of the patients born with functional single ventricle,
Increasing concern exists that APC Fontan patients are at risk for intraatrial thrombi, atrial arrhythmias, hemodynamic derangement, and impaired exercise tolerance \cite{1,2}. Improved modifications of the Fontan operation have resulted in today's accepted practice of TCPC such as lateral tunnel or extracardiac techniques with which hemodynamic profile was relatively well preserved and the incidence of late morbidities related to the unnatural physiology of Fontan procedure was diminished \cite{4,6,8}. For the patients with impaired hemodynamics after APC Fontan connection, hemodynamic and functional improvements are expected with the conversion of their Fontan connection to TCPC. Actually, Fontan conversion has resulted in significant hemodynamic improvement in patients with failing Fontan circulations with acceptable mortality and morbidity \cite{9}. Decreasing tension on the atrial tissue, reducing atrial size, maintaining laminar flow, and allowing the pulmonary veins and coronary sinus to decompress may all be factors that allow cardiac function to improve and arrhythmias to regress. Our selection criteria was that if we find a substrate that can be repaired, such as a valve lesion, an obstructive lesion, or cardiac dysfunction based on arrhythmias, then we prefer to attempt a Fontan conversion \cite{7}. In our cohort, the symptoms such as palpitation, dyspnea on exertion, cyanosis, chest pain, and edema were improved and the NYHA functional class was significantly improved. It is reasonable to regard this clinical and functional improvement resulted from the hemodynamic improvement.

Conversion of an APC Fontan to a TCPC without arrhythmia circuit cryoablation has been shown to result in a very high level of recurrence, with its attendant morbidity \cite{5}. Gandhi’s animal studies suggested that preexisting Fontan intraatrial suture lines provide the substrate for reentry atrial tachycardia and relief of atrial hypertension by means of Fontan conversion will not effectively control atrial arrhythmias unless accompanied by arrhythmia circuit cryoablation \cite{11}. There is considerable data suggesting that interruption of atrial arrhythmia with cryoablation in a failed Fontan, in addition to right atrial reduction and conversion to an extracardiac connection, will result in superior hemodynamics and an improvement in atrial arrhythmia \cite{5,7,11-13}. The extracardiac connection is attractive because it is generally easier to perform and has the added benefit of fewer atrial suture lines that are one of the substrates for atrial tachycardia \cite{13}.

With a combination of preoperative and intraoperative mapping, the Chicago group has identified the anatomic circuits responsible for the atrial arrhythmias \cite{7}. The exact sites for arrhythmia circuit cryoablation are the predominant anatomic critical isthmus in the area between the coronary sinus os, tricuspid anulus, and the inferior vena cava os \cite{14}. Isthmus cryoablation was performed in all patients for atrial flutter, and those patients with atrial fibrillation also received the modified right-sided maze procedure in our cohort. With this strategy, all patients had acutely successful atrial arrhythmia ablation. Although two patients (12.5%) had transient atrial arrhythmia recurrence in immediate postoperative period, their condition was easily controlled. Currently, we choose not to add left atrial cryoablation in atrial fibrillation \cite{15}. Left-sided maze procedure requires opening the left atrium and increasing cardiac ischemic time. Some authors report that a right-sided procedure will effectively treat patients who have congenital heart disease with accompanying atrial fibrillation \cite{16}. In some cases, the critical point of reentry is tissue that has been damaged through hypoxia or stretch rather than an incision. Chronic elevated right atrial pressure resulting in distension, hypertrophy, thickening, and dysplasia of right atrial wall which were developed after APC Fontan also alters the electrical characteristics of the atrial myocardium, increasing its susceptibility to arrhythmogenesis. We think that the right sided maze procedure are sufficient for atrial fibrillation after Fontan operation in selected cases. Even if arrhythmias were not completely eradicated postoperatively, they have become much easier to control.

Sinus node dysfunction may result from right atrial dilatation, right atrial hypertension, and injury to the sinus node or its blood supply after Fontan operation. Late term sinus node dysfunction has also been reported to be progressive, with occurrence rates of almost 10-12% after APC and 20-25% after lateral tunnel type operation \cite{17,18}. A multistaged operative pathway to Fontan reconstruction is associated with a higher early risk of altered sinoatrial node function \cite{19}. Moreover, sinus dysfunction or a slow sinus rate may predispose patients to the development of atrial flutter or atrial fibrillation, which is one of the major causes of late term morbidity and mortality \cite{19}. Atrial tachycardia occurs frequently accompanied by sinus bradycardia because irregular sinus bradycardia can promote more diversity for refractory period of atrial muscle, and then induce the reentry mechanism. Atrial pacemakers prevent bradyarrhythmias, which are caused by the commonly associated sick sinus syndrome. Preventing episodes of bradyarrhythmia through the use of pacemakers significantly decreased supraventricular and ventricular tachyarrhythmias \cite{20}. Preventing bradycardia thus may be an important adjunct to therapy in these patients, supporting the aggressive placement of atrial pacemakers after the cryoablation \cite{20}. The presence of the pacemaker also allows the therapeutic doses of antiarrhythmic agents that can worsen bradycardia (especially sotalol) to be administered safely if some tachycardia persist postoperatively.

In our cohort, permanent DDD epicardial pacemakers were placed in seven patients after arrhythmia circuit cryoablation because of sinus node dysfunction. During follow-up, two patients needed a permanent dual-chamber pacemaker implantation. Postoperatively, all patients with pacemaker therapy have not had inducible atrial tachycardia except one (No. 5 in Table 1) in whom atrial flutter developed, but easily withdrawn. It is often difficult to find a favorable right atrial lead location because of the endocardial fibrosis often present in the right side of the atrium. The left atrium lacks endocardial fibrosis because reconstructive suture lines are rarely placed in this area, and the left atrial appendage is a superior site for the atrial pacemaker yielding favorable eletrophysiologic measurements \cite{21}. Transvenous access for a pacemaker lead would be difficult postoperatively, particularly in patients with an
and permanent pacemaker placement is a safe and efficacious operation in improving the clinical outcome for patients with atrial arrhythmia and low cardiac output. Debilitating arrhythmias are cured, and patients have an improvement in NYHA class. In most patients, concomitant permanent pacemakers are needed. Optimal selection criteria for conversion have yet to be determined, but most patients can be expected to benefit to some extent. The degree of improvement is likely to correspond with the proportion of complications of the previous Fontan connection in a given patient that can be addressed by conversion. If PLE is accompanied by failed Fontan circulation, the improvement of PLE could be expected with the hemodynamic improvement and arrhythmia control after this procedure.

5 Conclusions

Fontan conversion with concomitant arrhythmia surgery and permanent pacemaker placement is a safe and
Dr Kim: These patients were Fontan conversion only.

Dr Edmunds: So the 16 patients were all of the patients that you converted to the Fontan in 8 years?

Dr Kim: Yes. Sixteen patients all conversion Fontan procedure.

Dr Edmunds: All right. And of those 16 patients, 13 had a preoperative atrial arrhythmia?

Dr Kim: Yes.

Dr Edmunds: All right.

Dr F. Haas (Utrecht, The Netherlands): What was the main indication for doing this difficult operation? Was it arrhythmia alone or was it the enlarged right atrium with hemodynamic instability?

Dr Kim: Indication is several and arrhythmia itself is a strong indication. Development of arrhythmia is strong indication. In some patients, intra-atrial thrombus is a strong indication, and the ventricle dysfunction associated with arrhythmia. And the pulmonary venous compression is also an indication.

Dr V. Alexi-Meseshvili: You have quite a high number of patients with thrombosis. So maybe those patients also have coagulation disorders. What is your policy for anticoagulation treatment after the Fontan operation?

The second question concerns the insertion of the pacemaker in the regular Fontan operation. Sometimes it’s quite difficult to find a good place for electrodes. So what are the preferable places for electrode insertions? Do you prefer to put them on the left atrium or on the pulmonary veins?

Dr Kim: We routinely do a staged operation for Fontan procedure, so it is often difficult to find a favorable right atrial lead location because of the endocardial fibrosis in AP connection-type Fontan patients. It is time-consuming procedure. Usually we can find a good site in left atrium. So during Fontan conversion, approach to left atrium is very easy. So we contend that a prophylactic epicardial lead may be helpful even without sinus node dysfunction during Fontan. So we routinely put the left atrial lead in during Fontan conversion.

And the first question, the anticoagulation for Fontan case, we have medication only experience. And if there is a risk factor, we put heparin. And these Fontan conversion patients, all cases, we strongly plan lifelong Coumadin.

Dr V. Hjortdal (Aarhus, Denmark): You have a high number of patients requiring pacemaker. Was it because of an underlying sick sinus node syndrome or was it because of AV block?

Dr Kim: We routinely do a staged operation for Fontan procedure, so sinus node damage is high incidence. And in case of AV node dysfunction, it’s high. And we preoperatively Fontan convert 7 patients with edema and sinus node and AV node dysfunction, and so 7 cases was pacemaker during Fontan procedure. Two patients was doing poorly after post-operatively 7 days, and 8 months later we put the permanent pacemaker. So it is beneficial to Fontan conversion patients, it is reminder that permanent pacemaker is very important for postoperative arrhythmia prevention.
Fontan conversion with arrhythmia surgery
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