Conversion of atriopulmonary or lateral atrial tunnel cavopulmonary anastomosis to extracardiac conduit Fontan modification

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Received 22 September 1998; accepted 16 December 1998

Abstract

Objective: Obstruction of the atriopulmonary anastomosis or the lateral atrial tunnel cavopulmonary anastomosis in the Fontan circulation for univentricular physiology may result in dilation of the right atrium or the right atrial free wall that is incorporated in the lateral atrial tunnel, respectively. Secondary detrimental sequelae may consist of supraventricular dysrhythmias, thromboembolism, right pulmonary vein compression, pleural effusions, and protein-losing enteropathy. Conversion of these Fontan connections to an extracardiac conduit cavopulmonary anastomosis may improve central systemic venous flow patterns and provide clinical improvement in these patients.

Methods: Eighteen patients (7–40 years old) with atriopulmonary anastomosis (n = 15) or obstructed lateral atrial tunnel cavopulmonary anastomosis (n = 3) presented at 5.7–3.9 years with moderate to severe right atrial dilation (n = 15), Fontan pathway obstruction (n = 12), atrial dysrhythmia (n = 13), pleural effusion (n = 8), right atrial thrombus (n = 3), right pulmonary vein compression (n = 3), and protein-losing enteropathy (n = 3). All patients underwent conversion to an extracardiac conduit cavopulmonary anastomosis. Results: Two of the three patients with protein-losing enteropathy died (2/18; 11%) on the 30th and 52nd postoperative days. At a mean follow-up of 19 months, the remaining 16 patients had marked (n = 11) or moderate (n = 5) clinical improvement. The SaO₂ improved from 90.7–5.3% to 96.0–4.1%. None of the patients had obstruction in the systemic venous pathway. In the 13 surviving patients with previous atriopulmonary anastomosis there was a drastic reduction in right atrial size. Four of 13 patients with atrial dysrhythmias converted to sinus rhythm. The right pulmonary vein compression as present in three patients resolved after conversion. Pleural effusions disappeared in four patients.

Conclusions: Conversion to an extracardiac cavopulmonary connection may lead to clinical improvement in patients with atriopulmonary or lateral atrial tunnel Fontan connection associated with specific target conditions such as obstruction, pulmonary vein compression, right atrial enlargement, atrial dysrhythmia, or atrial thrombus. The conversion operation should not be unduly delayed to prevent irreversible deterioration of clinical status with chronic rhythm disturbances or protein-losing enteropathy. The benefit of the conversion operation is questionable in patients with poor clinical condition and protein-losing enteropathy. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Fontan circulation; Univentricular heart; Cavopulmonary connection

I. Introduction

Anastomotic obstruction, right atrial dilation, progressive loss of sinus rhythm, thromboembolism, right pulmonary vein compression, chronic pleural effusions, and protein-losing enteropathy have been noted in survivors of the atriopulmonary anastomosis and, to a lesser extent, the total cavopulmonary connection of the Fontan operation [1–23]. In the absence of irreversible ventricular systolic or diastolic dysfunction and elevated pulmonary vascular resistance, such patients may benefit from conversion of their atriopulmonary or lateral tunnel cavopulmonary Fontan connection to an extracardiac conduit modification. In an attempt to stabilize or improve the clinical status, we have recently converted a number of patients with atriopulmonary or total cavopulmonary anastomosis to an extracardiac conduit modification of the Fontan procedure [7,24,25]. This study reports our experience with this technique.
2. Patients and methods

Since October 1994, 18 patients with atriopulmonary anastomosis (n = 15) or lateral atrial tunnel cavopulmonary connection (n = 3) had conversion to an extracardiac conduit Fontan procedure. Eleven of these patients had undergone their primary Fontan procedure at other institutions. Since 1991, the extracardiac conduit Fontan modification is our procedure of choice for patients who need to undergo a primary Fontan procedure. There were 12 male and six female patients. Table 1 summarizes the diagnoses and clinical characteristics of the patients. The mean age at reoperation was 15.1 ± 6.5 years (range 7 to 40 years). The mean time interval between the primary Fontan operation and the conversion operation was 5.7 ± 3.9 years (range 3 to 29 years). Before the Fontan operation, all patients had undergone one or more previous palliative procedures including bidirectional cavopulmonary anastomosis (n = 3), modified Blalock–Taussig shunt (n = 12), pulmonary artery banding (n = 2), Norwood operation (n = 3), atrial septectomy (n = 6), and resection of aortic coarctation (n = 1).

Indications for operation included New York Heart Association (NYHA) functional class, Fontan pathway obstruction (Fig. 1), moderate to severe right atrial dilation (Fig. 2), atrial dysrhythmia, right atrial thrombus, right pulmonary vein obstruction, chronic pleural effusions, and protein-losing enteropathy (see Table 1). None of the patients had a clinical history of atrial dysrhythmia before the initial Fontan procedure. Four patients had an epicardial pacemaker. Seven patients had a moderately to markedly decreased exercise tolerance (NYHA classes III (n = 4) and IV (n = 3)). Fifteen patients had been on long-term anti-coagulation treatment: ten patients received acetylsalicylic acid and five patients received warfarin. Two of the three patients with intraatrial thrombus were treated with acetylsalicylic acid; the remaining patient had no anti-coagulation treatment.

Preoperative cardiac catheterization was performed in all patients. The hemodynamic and oximetric data are summarized in Table 2. The mean pulmonary vascular resistance was 3.1 ± 1.9 Wood units (range 1.9 to 4.6 units). Twelve patients (nine with an atriopulmonary anastomosis and three with a lateral tunnel cavopulmonary connection) had Fontan pathway obstruction (mean gradient 3.4 ± 3.0 mmHg; range 1 to 10 mmHg). In three patients with an atriopulmonary Fontan connection, thrombus was demonstrated to be adherent to the dilated right atrial free wall. Obstruction of the right upper pulmonary vein, as a result of compression by a markedly dilated right atrium, was diagnosed in three patients, with measured gradients of 3, 5, and 8 mmHg. Two patients had elevated left ventricular end-diastolic pressures but preserved systolic ventricular function. Two patients had moderate left atrioventricular valve regurgitation.

2.1. Operative procedure

Through a repeat median sternotomy the heart and the great vessels, both branch pulmonary arteries, the superior vena cava (SVC), and inferior vena cava (IVC) are dissected and completely mobilized. In selected patients with large, thinned-walled and anterior right atria, initial femoral artery-femoral vein partial cardiopulmonary bypass may be a safe technique to decompress the right atrium for more effective and safe reoperation. The aorta and both venae cavae are cannulated (with the cannula in the IVC placed as low as possible) (Fig. 3A). Alternatively, it may be helpful to cannulate the left brachiocephalic vein and the femoral vein in order to have a free operative field in the crucial areas of the SVC and IVC. In patients with a pressure gradient across a previous superior cavopulmonary anastomosis, the SVC is detached from the pulmonary artery, scar tissue excised, and a wide anastomosis between both structures made. Atrial septectomy with division of the atriopulmonary anastomosis or resection of the intraatrial patch of the lateral tunnel or additional procedures (such as resection of subaortic stenosis or repair of a regurgitant atrioventricular valve) are performed during a brief period of aortic cross-clamping with cardioplegic arrest using moderate hypothermia. In the presence of a large right atrium, as was the case in 15 patients in this series, a reduction atrioplasty is performed. Subsequently, after deairing of the heart and removal of the aortic

Table 1

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Previous operation</th>
<th>NYHA class</th>
<th>Indication for revision</th>
<th>Ventricular function</th>
</tr>
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<tbody>
<tr>
<td>(S,D,D),TA (5)</td>
<td>BCPA (3)</td>
<td>Class I (4)</td>
<td>Right atrial dilation (15)</td>
<td>Normal (11)</td>
</tr>
<tr>
<td>(S,D,D),TA (2)</td>
<td>BT shunt (12)</td>
<td>Class II (7)</td>
<td>Fontan pathway obstruction (12)</td>
<td>Mildly depressed (5)</td>
</tr>
<tr>
<td>(S,D,S),HLHS (3)</td>
<td>PAB (2)</td>
<td>Class III (4)</td>
<td>Atrial dysrhythmia (13)</td>
<td>Moderately depressed (2)</td>
</tr>
<tr>
<td>(S,D,S),PA,VJS (4)</td>
<td>Norwood operation (3)</td>
<td>Class IV (5)</td>
<td>Pleural effusion (8)</td>
<td>–</td>
</tr>
<tr>
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<td>–</td>
<td>RA thrombus (3)</td>
<td>–</td>
</tr>
<tr>
<td>(S,D,D),CA,PA (1)</td>
<td>CoA Repair (1)</td>
<td>–</td>
<td>RPV compression (3)</td>
<td>–</td>
</tr>
<tr>
<td>(S,D,S),Unh AVC (1)</td>
<td>–</td>
<td>–</td>
<td>PLE (3)</td>
<td>–</td>
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</tbody>
</table>

BCPA, bidirectional cavopulmonary anastomosis; BT, modified Blalock–Taussig shunt; CA, common atrium; CoA, coarctation of the aorta; DLV, double-inlet left ventricle; HLHS, hypoplastic left heart syndrome; IJS, intact ventricular septum; PA, pulmonary atresia; PAB, pulmonary artery banding; PLE, protein-losing enteropathy; PS, pulmonary stenosis; RA, right atrial; RPV, right pulmonary vein; (S,D,D), (S,D,S), (S,L,L), segmental situs; TA, tricuspid atresia. Unh AVC, unbalanced atrioventricular canal.
cross-clamp, the remainder of the operation is performed with a beating heart. In patients with a previous atriopulmonary anastomosis, the SVC is divided at the level of the corresponding branch pulmonary artery, the cardiac end is oversewn, and a longitudinal pulmonary arteriotomy is made on the superior surface of the artery. A cavopulmonary anastomosis is constructed with 6-0 polyglyconate suture (Maxon, Davis and Geck, Danbury, CN). The right atrium is clamped at its base just proximal to its junction with the IVC (with avoidance of the coronary sinus) and transected below the clamp, thereby leaving a beveled cuff of atrial tissue attached to the IVC (Fig. 3B). The base of the right atrium is oversewn with two layers of polypropylene suture (Prolene, Ethicon, Somerville, NJ). The incision at the underside of the right pulmonary artery that is created by division of the atriopulmonary anastomosis is extended laterally or centrally, or both, and a 22-mm to 24-mm flexible Gore-Tex tube graft (W.L. Gore and Assoc, Elkton, MD) is cut obliquely and anastomosed end-to-end to the IVC using Gore-Tex suture in a continuous fashion. The upper portion of the conduit is then positioned so that it arches over the right upper pulmonary vein and extends to the incised right pulmonary artery. The conduit is fashioned in such a way that a flap extends laterally onto the right pulmonary artery. In case of central pulmonary artery stenosis, the underside of the pulmonary artery is filleted open centrally and the Gore-Tex conduit cut tangentially so that it acts as an onlay augmentation patch of the pulmonary artery (Fig. 3C). In the presence of a left-sided IVC, the conduit is positioned to the left side of the heart and anastomosed to the underside of the left pulmonary artery (Fig. 3D).

In moderate-risk or high-risk Fontan patients, a communication between the extracardiac conduit and the right atrium is constructed by anastomosing a 4-mm GoreTex tube graft between both structures (see Fig. 3B). An adjustable snare is placed around the graft and passed through an 8F polyethylene tube. If, after weaning from cardiopulmonary bypass, the pressure in the extracardiac conduit is less then 16 mmHg, the communication is closed with hemostatic clips. If the arterial oxygen saturation is less than 85%, the communication is likewise made smaller or closed. If at the end of the operation, based on an elevated pressure in the extracardiac conduit, it is decided that the communication is to be left open, the snare in the 8F tube graft is buried underneath the linea alba for easy retrieval postoperatively. After the completion of the procedure, pressure monitoring lines are placed in the common atrium and in the IVC after having excluded any gradients across the cavopulmonary pathway. Intraoperative transesophageal echocardiographic assessment is performed routinely. The patients are extubated as soon as possible.

Fig. 1. An angiogram of patient with lateral tunnel cavopulmonary anastomosis. Arrows indicate levels of obstruction.

Fig. 2. An angiogram of patient with moderate right atrial enlargement and pressure gradient across the atriopulmonary anastomosis. RA, right atrium.
class I or II. Angiography in three patients demonstrated improvement (Table 3). Thirteen patients are in NYHA class I or II. 

In 17 patients a right-sided extracardiac conduit was constructed; in one patient with heterotaxy syndrome, a left IVC and dextrocardia, a left-sided conduit was inserted. In all three patients with preoperatively diagnosed right atrial thrombus, the thrombus was confirmed to be adherent to the dilated right atrial free wall, where there was the potential for stagnant blood flow. In the three patients with a lateral tunnel cavopulmonary connection, the posterolateral right atrial free wall was dilated secondary to tunnel outflow obstruction. In 12 patients a 4-mm fenestration between the extracardiac conduit and the adjacent atrium was constructed. In two patients this communication was closed intraoperatively and in three patients it was closed on the second (n = 2) or third (n = 1) postoperative day in the intensive care unit. Both patients with atrioventricular valve regurgitation underwent valve repair. The intraoperative SaO2 was 94.8 ± 4.9% at 48 h postoperatively. Intraoperatively measured pressures demonstrated the absence of obstruction between the extracardiac conduit and the pulmonary artery (mean pulmonary artery pressure 14.8 ± 4.8 mmHg). The mean postoperative length of hospital stay in the 16 survivors was 11 ± 4 days. Chest tube output was minimal for 4.1% at 48 h postoperatively. Intraoperatively, the pleural effusions recurred, accompanied by protein-losing enteropathy persisted in both patients. Eventually, the pleural effusions recurred, accompanied by exacerbation of ascites and peripheral edema, ultimately leading to the demise of both patients.

The mean postoperative length of hospital stay in the 16 survivors was 11 ± 4 days. Chest tube output was minimal in ten patients. The last chest tube was removed on postoperative day 8 ± 4. In four of the six surviving patients with chronic pleural effusions, the effusions disappeared. At a mean follow-up of 19 months, the surviving 16 patients had marked (n = 11) or moderate (n = 5) clinical improvement (Table 3). Thirteen patients are in NYHA class I or II. Angiography in three patients demonstrated excellent patency of the extracardiac conduit (Fig. 4). Echocardiography showed non-patency of the extracardiac-to-atrial communication in four of the seven patients who had a patent communication at discharge. In the 13 surviving patients with previous atriopulmonary anastomosis there was a drastic reduction in right atrial size. Four of 13 patients with atrial dysrhythmias converted to sinus rhythm; in two of these this occurred early postoperatively, and in the other two conversion was demonstrated at 3 months postoperatively. None of the three patients with atria dysrhythmias and a prior lateral atrial tunnel converted to sinus rhythm. A pacemaker was inserted in three of the seven surviving patients with persistent atrial dysrhythmias and in the two patients who converted to sinus rhythm late postoperatively. The right pulmonary vein obstruction, as present in three patients, resolved after operation. One patient with persistent protein-losing enteropathy had a moderate improvement in his clinical status (from NYHA class IV to III).

**Table 2**

<table>
<thead>
<tr>
<th>Before conversion</th>
<th>After conversion</th>
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<tbody>
<tr>
<td>SaO2</td>
<td>SaO2</td>
</tr>
<tr>
<td>FP</td>
<td>PAP</td>
</tr>
<tr>
<td>PAP</td>
<td>PVR</td>
</tr>
<tr>
<td>AP</td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>90.7</td>
</tr>
<tr>
<td>SD</td>
<td>16.7</td>
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<td></td>
<td>12.9</td>
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<tr>
<td></td>
<td>3.1</td>
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<td></td>
<td>94.8</td>
</tr>
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<td></td>
<td>14.8</td>
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<td></td>
<td>7.1</td>
</tr>
</tbody>
</table>
| AP, common atrial pressure (mmHg); FP, Fontan pressure (in right atrium or lateral tunnel; mmHg); PAP, pulmonary artery pressure (mmHg); PVR, pulmonary vascular resistance (Wood units); SaO2, percent arterial oxygen saturation; SD, standard deviation.

**3. Results**

In 17 patients a right-sided extracardiac conduit was constructed; in one patient with heterotaxy syndrome, a left IVC and dextrocardia, a left-sided conduit was inserted. In all three patients with preoperatively diagnosed right atrial thrombus, the thrombus was confirmed to be adherent to the dilated right atrial free wall, where there was the potential for stagnant blood flow. In the three patients with a lateral tunnel cavopulmonary connection, the posterolateral right atrial free wall was dilated secondary to tunnel outflow obstruction. In 12 patients a 4-mm fenestration between the extracardiac conduit and the adjacent atrium was constructed. In two patients this communication was closed intraoperatively and in three patients it was closed on the second (n = 2) or third (n = 1) postoperative day in the intensive care unit. Both patients with atrioventricular valve regurgitation underwent valve repair. The intraoperative SaO2 was 94.8 ± 4.9% (see Table 2) with an increase of the SaO2 to 96.0 ± 4.1% at 48 h postoperatively. Intraoperatively measured pressures demonstrated the absence of obstruction between the extracardiac conduit and the pulmonary artery (mean pulmonary artery pressure 14.8 ± 2.6 mmHg). The mean common atrial pressure was 7.1 ± 4.8 mmHg.

Two patients, a 19-year-old female and a 40-year-old male, both with a previous atriopulmonary anastomosis, died on the 30th and 52nd postoperative days, respectively (2/18;11%). In both patients, who had severe protein-losing enteropathy, chronic pleural effusions, ascites and cachexia, the initial postoperative course had been relatively uneventful with the disappearance of the effusions. However, the protein-losing enteropathy persisted in both patients. Eventually, the pleural effusions recurred, accompanied by exacerbation of ascites and peripheral edema, ultimately leading to the demise of both patients.

The mean postoperative length of hospital stay in the 16 survivors was 11 ± 4 days. Chest tube output was minimal in ten patients. The last chest tube was removed on postoperative day 8 ± 4. In four of the six surviving patients with chronic pleural effusions, the effusions disappeared. At a mean follow-up of 19 months, the surviving 16 patients had marked (n = 11) or moderate (n = 5) clinical improvement (Table 3). Thirteen patients are in NYHA class I or II. Angiography in three patients demonstrated excellent patency of the extracardiac conduit (Fig. 4). Echocardiography showed non-patency of the extracardiac-to-atrial communication in four of the seven patients who had a patent communication at discharge. In the 13 surviving patients with previous atriopulmonary anastomosis there was a drastic reduction in right atrial size. Four of 13 patients with atrial dysrhythmias converted to sinus rhythm; in two of these this occurred early postoperatively, and in the other two conversion was demonstrated at 3 months postoperatively. None of the three patients with atria dysrhythmias and a prior lateral atrial tunnel converted to sinus rhythm. A pacemaker was inserted in three of the seven surviving patients with persistent atrial dysrhythmias and in the two patients who converted to sinus rhythm late postoperatively. The right pulmonary vein obstruction, as present in three patients, resolved after operation. One patient with persistent protein-losing enteropathy had a moderate improvement in his clinical status (from NYHA class IV to III).

**4. Discussion**

In the Fontan circulation a single ventricle and two resistors, i.e. systemic and pulmonary, are connected by the systemic arterial, the systemic venous, and the pulmonary venous conduits, respectively (Fig. 5). If there is no pulmonary vascular obstructive disease, subaortic obstruction or atrioventricular valve regurgitation, and ventricular function remains well preserved, the outcome of the Fontan operation is mainly related to the absence of obstruction in the systemic venous conduit and absence of atrial dysrhythmias. To avoid recirculation of both pulmonary and systemic venous return with resultant volume overload of the single ventricle and attendant sequelae, there is a well-established trend to perform a bidirectional superior cavo-pulmonary anastomosis at early age as an intermediate-stage palliative procedure [25,26]. Associated lesions, such as subaortic stenosis, atrioventricular valve regurgitation, or pulmonary artery stenosis are usually corrected at the time of construction of the bidirectional cavo-pulmonary anastomosis. We prefer to complete the Fontan operation using an extracardiac conduit technique at an age of 3.5–4 years. This timing is primarily based on physiologic and practical issues. Delay of completion of the Fontan opera-
tion beyond this age, secondary to a maturational decrease in apportionment of systemic blood flow to the upper versus the lower body segment, is often accompanied by exercise incapacity and excessive cyanosis with attendant sequelae [27,28]. A concomitant practical advantage of completing the Fontan operation at 3.5–4 years of age is the fact that,
empirically, at this age an adult size conduit (22–25 mm) can usually be inserted.

Right atrial dilation, progressive loss of sinus rhythm, thromboembolism, pulmonary vein compression, persistent pleural effusions, ascites and protein-losing enteropathy have been noted in long-term survivors of the atriopulmonary or, less common, the cavopulmonary anastomosis [1–23]. In the vast majority of patients with these complications, there is evidence of obstruction in the systemic venous conduit [4,5,7,12,13]. Such an obstruction leads to dilation of the right atrium in the presence of an atriopulmonary anastomosis or the segment of atrial wall that is incorporated in the lateral tunnel of the cavopulmonary anastomosis. The resulting wall tension predisposes to the development of atrial dysrhythmias. Other causes for the development of dysrhythmias are the presence of atrial suture lines, as present in the lateral atrial tunnel (in particular when a flap of atrial free wall is used) or damage to the sinus node at the time of the completion of the Fontan procedure [29,30].

The turbulent flow in the dilated and non-cylindrical pathway, enhanced by the atrial dysrhythmia, may lead to the stasis of blood and thrombus formation, particularly in an enlarged right atrium proximal to a stenosed atriopulmonary anastomosis. In two other reports on the conversion operation, intraatrial thrombus was present in two of the 15 patients (13.3%) [5,7]. In our series, three patients with a giant right atrium, all after an atriopulmonary Fontan connection, had thrombus in the right atrium. In all cases, the thrombus characteristically was adherent to the massively dilated atrial wall, suggesting that stasis of blood preferentially takes place in areas where eddy currents are formed. In a multicenter study Mertens et al. [23] reported an incidence of 0.6% of intracardiac thrombus among 3029 Fontan patients over a 20-year period. Du Plessis et al. [18] reported a 2.6% incidence of stroke among 645 patients who underwent the Fontan procedure over a 15-year period. In the 16 patients who underwent echocardiography at the time of the revision.

Table 3

<table>
<thead>
<tr>
<th></th>
<th>Before Fontan revision (n)</th>
<th>After Fontan revision (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>NYHA Class I</td>
<td>4</td>
<td>9</td>
</tr>
<tr>
<td>Class II</td>
<td>7</td>
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</tr>
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<td>Class III</td>
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<td>3</td>
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<tr>
<td>Class IV</td>
<td>3</td>
<td>2*</td>
</tr>
<tr>
<td>Sinus rhythm</td>
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<td>9</td>
</tr>
<tr>
<td>Atrial dysrhythmia</td>
<td>13</td>
<td>9</td>
</tr>
<tr>
<td>Right atrial dilation</td>
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<tr>
<td>Fontan pathway obstruction</td>
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<td>0</td>
</tr>
<tr>
<td>Right atrial thrombus</td>
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<td>0</td>
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<td>Right pulmonary vein compression</td>
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<td>0</td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Protein-losing enteropathy</td>
<td>3</td>
<td>3</td>
</tr>
</tbody>
</table>

*Both patients died.
stroke, seven were found to have intracardiac thrombus that in every case was in a position to generate emboli into the systemic circulation (main pulmonary artery stump, on a fenestrated right atrial baffle or lateral tunnel and on the mitral valve). Jahangiri et al. [16] documented the presence of thrombus in the right atrium in ten of the 55 surviving patients (18.2%) after atriopulmonary or cavopulmonary Fontan connections, none of whom received coumadin for more than 8 months postoperatively.

A third effect of chronic obstruction in the systemic venous pathway is the potential for the development of pleural effusions, ascites or protein-losing enteropathy [19–23]. These complications have been shown to be due to impendiment of the parietal pleural venous drainage into the azygos and hemiazygos system and internal mammary veins which empty into the SVC and innominate vein [19]. The inability of the thoracic duct to empty properly in the systemic venous compartment, since the systemic venous pressure continuously exceeds the pressure in the thoracic duct, contributes to lymphatic stasis and the development of ascites and intestinal lymphangiectasis. A second mechanism by which protein-losing enteropathy occurs is the increased lymph production due to the increased pressure in the IVC and portal vein. This situation leads to a loss of albumin, lymphocytes, and immunoglobulins into the gastrointestinal tract, resulting in hypoalbuminemia with generalized edema and eventually immunologic abnormalities.

Conversion of traditional Fontan connections into an extracardiac conduit Fontan modification may ‘break’ the negative vicious circle in which many of these patients are. Specific indications for the conversion operation are not clearly defined as yet. The most consistent clinical improvement can be expected in patients in whom a specific condition or complication is directly addressed, such as right atrial thrombus, pulmonary vein compression, or dysrhythmia refractory to conventional medical or electrophysiologic treatment. Although the presence of other unfavorable conditions such as obstruction at the level of the atriopulmonary anastomosis or lateral tunnel cavopulmonary anastomosis or in the subaortic or bulboventricular foramen area, or atrioventricular valve regurgitation do not, per se, require conversion to an extracardiac Fontan connection, such patients may certainly benefit from the improved systemic venous flow pattern. In our series, maximum benefit of the conversion operation was obtained in patients with NYHA class I, II or III in whom a specific condition was addressed. In 13 patients with a previous atriopulmonary anastomosis and right atrial enlargement there was a drastic reduction in the right atrial size. Fontan pathway obstruction was eliminated in 12 patients, right atrial thrombus was removed in three patients, and pulmonary vein compression was resolved in three patients. Four of the 13 patients with atrial dysrhythmias converted to sinus rhythm. The likelihood of conversion to sinus rhythm was greatest in patients with an atriopulmonary Fontan connection in whom the rhythm disturbance had been of a relatively short duration. Interestingly, in none of the three patients with lateral tunnel cavopulmonary anastomosis, the conversion operation resulted in improvement of the rhythm. One explanation for the observed difference in improvement of atrial dysrhythmias between both groups may relate to the presence of more extensive atrial suture lines and potential trauma to the sinoatrial node in the latter group.

Regardless of the presence of specific target conditions, the indication for the conversion operation may be questionable in patients with deteriorating functional status with associated protein-losing enteropathy and pleural effusions. Two of three patients with protein-losing enteropathy and cachexia in this series died and the third had persistent protein-losing enteropathy despite a moderate improvement of his clinical status. Similarly, of three converted patients with protein-losing enteropathy in other series, one died and the remaining two patients, one of whom was subjected to cardiac transplantation, had persistent protein-losing enteropathy [5,7]. These limited data suggest that such physiologically fragile patients may best be treated by direct orthotopic heart transplantation rather than by conversion to an extracardiac conduit Fontan modification.

The expected benefits in the prevention of thromboembolism after the Fontan operation must be weighed against the risk of life-long anticoagulation. With increased follow-up and using a sensitive diagnostic method like transesophageal echocardiography, the incidence of thromboembolism after Fontan procedures is likely to be greater than 20% [13–18]. Although the question of routine anticoagulation in all patients after a Fontan procedure needs to be clarified by a careful prospective study, available data from the literature [13–18] suggest that long-term anticoagulation should be maintained, particularly in patients in whom there is stagnant flow in the area of the Fontan connection or in whom supraventricular dysrhythmias have been documented. Since these conditions are met in the majority of patients who are subjected to a conversion operation, we believe that most patients who undergo
this operation should be maintained on a regimen of long-term anticoagulation. Our current regimen is to start heparin after 24–48 h postoperatively and to initiate warfarin (international normalized ratio of 3) once the patient is adequately heparinized and starts absorbing by mouth. In patients with excellent to good hemodynamics (including sinus rhythm), after an arbitrary period of 6 months, warfarin may be replaced by acetylsalicylic acid. In patients with less favorable hemodynamics, long-term warfarin medication is indicated. Anticoagulation treatment may be avoided in the rare patient with good hemodynamics in whom only autologous tissue is used for the extracardiac Fontan conduit [31].

References


Appendix A Conference discussion

Mr B. Sethia (Birmingham, UK): One of the criticisms of the extracardiac Fontan type of procedure is that you may cause compression of the right-sided pulmonary veins by the line of the conduit. Can you say more about how you made sure that that wasn’t in fact a problem in your patients. And would you recommend this sort of procedure in much
younger patients in whom, certainly, the anatomy of the conduit may compress the pulmonary veins?

**Dr van Son**: In order to avoid pulmonary vein compression the upper portion of the extracardiac conduit is positioned anteriorly so that it arches over the right upper pulmonary vein. With this technique we have had excellent results.

**Mr Seithia**: That may be so in the short-term, but there are instances of late pulmonary venous compression, and three of your patients had pulmonary venous compression beforehand.

**Dr van Son**: These three patients had atriopulmonary Fontan connections with giant right atria. I believe that completion of the Fontan procedure is best performed at an age of 3–4 years, which timing is based both on physiologic and practical issues. Delay of completion beyond this age, secondary to a maturational decrease in apportionment of systemic blood flow to the upper versus the lower body segment, is often accompanied by exercise incapacity and excessive cyanosis with attendant sequelae. At 3–4 years of age an adult-sized conduit (22–24 mm) can usually be inserted using the technique described before. Adhering to these principles we have not yet encountered a single case of pulmonary vein compression.

**Dr B. Maruszewski** (Warsaw, Poland): To support your data, I want to say that we have already operated on 16 patients with this technique, using 12 direct anastomosis between the mobilized MPA and inferior vena cava, and using now, electively, the aortic homograft in any case when we cannot connect it directly. And there is a trick that we learned to avoid the compression of the right pulmonary veins, which is the mobilization of the pericardium over the pulmonary veins. We recatheterized these patients, the youngest was 11 kg at first surgery, we recatheterized these patients 1–2 years postoperatively, and we haven’t seen any compression of the right pulmonary veins. I think it’s a very good operation.

**Dr van Son**: I am concerned that a direct anastomosis between the main pulmonary artery and the inferior vena cava may lead to tension on this anastomosis and to distortion of the bidirectional cavopulmonary anastomosis with the potential for secondary detrimental sequelae such as obstruction. This may offset the primary goal of the Fontan operation, that is, the creation of maximally streamlined cavopulmonary connections. Did you have enough tissue to bridge the gap between the main pulmonary artery and the inferior vena cava?

**Dr Maruszewski**: Out of 16, in 12 patients we were able to perform direct anastomosis, and in the others we used aortic homograft tube.

**Dr W. Williams** (Toronto, Ontario, Canada): I am concerned about the arrhythmia question, which long-term in the Fontan operation is going to be a very major problem. I was disappointed to see that you had arrhythmia (SVT) in 13 patients preoperatively and nine postoperatively. I wondered if you would consider adding intervention for the arrhythmia specifically, such as cryoblation of the right atrium along the cavaotral junction. Have you given thought to dealing with the arrhythmias more specifically now that conversion to an extracardiac conduit doesn’t seem to be the answer.

**Dr van Son**: Over the past decade, staged management with construction of a bidirectional cavopulmonary anastomosis at early age has led to better candidates for the Fontan operation by drastic reduction of the deleterious effects of long-term ventricular volume overload, repeated palliative procedures and chronic hypoxemia. Completion of the Fontan procedure as an extracardiac modification avoids extensive atrial suture lines as used in the lateral tunnel technique and avoids damage to the sinusotral node, and therefore, I expect that the long-term results of this technique will be considerably better than conventional Fontan modifications in terms of atrial arrhythmia. In our series, four patients with atrio-pulmonary Fontan connections and giant right atria converted to sinus rhythm after the conversion operation. Interestingly, none of the three patients with a previous lateral tunnel cavopulmonary connection converted to sinus rhythm, which may be due to the previously mentioned factors. I believe that the best results in terms of restoration of sinus rhythm can be achieved in patients in whom the loss of sinus rhythm is primarily related to increased right atrial wall tension and in whom the arrhythmia has been of relatively short duration. In patients with long-term arrhythmia, the conversion operation can be combined with intraoperative mapping and selective cryoblation, which can interrupt the arrhythmia circuit in many patients. Remaining atrial arrhythmia can be managed by an insertion of a pacemaker. At last it is worth mentioning that rhythm management in those patients who do not convert to sinus rhythm after the conversion operation is generally much easier due to reduced right atrial wall tension and probably also secondary to reduced coronary sinus pressure.

**Dr M. Masuda** (Fukuoka, Japan): We are also comfortable with using the extracardiac conduit for TCPC. What kind of regimen do you use for anticoagulation? Do you use anticoagulation for your patients with conduit or don’t you anticoagulate them?

**Dr van Son**: I believe that all patients who are converted to an extracardiac conduit Fontan circulation should be anticoagulated. Patients with moderate to poor hemodynamics probably should be managed with long-term coumadin medication. If the hemodynamic situation is good, administration of acetylsalicylic acid is probably satisfactory.

**Dr C. Knott-Craig** (Oklahoma City, OK): I agree with your conclusions. And converting the patients to an extracardiac conduit is something which I have done often, and in fact, extracardiac conduit is often my first choice of technique for a Fontan repair. There is one word of caution though, once you have an extracardiac conduit, it’s very difficult for the interventional cardiologists to gain access to the heart, both in terms of pacing the patient, either perioperatively or late after repair. If you have an atriopulmonary connection or a TCPC, they can always get a wire pacemaker in the heart, and if you develop sick sinus rhythm later, it’s easy to treat. If you have an extracardiac conduit this is not possible.

**Dr van Son**: That is a good point. However, I think that with modern management, that is, early construction of a bidirectional cavopulmonary anastomosis with, if present, concomitant repair of associated intracardiac anomalies, such as atrioventricular valve regurgitation or subaortic stenosis, followed by completion of the Fontan procedure as an extracardiac conduit at 3–4 years of age, that the incidence of residual problems, notably arrhythmia, will be dramatically lower than in the past.

**Dr A. Merozhko** (Tomsk, Russia): I would ask you what is the tactic for your patients now, what is the first step, extracardiac Fontan or some other procedure?

**Dr van Son**: For palliation of ductus-dependent univentricular physiology, it is our policy to construct a 3.5 mm modified Blalock-Taussig shunt or central aortopulmonary shunt. In non-ductus-dependent physiology, we attempt to manage the neonate medically until primary construction of a bidirectional cavopulmonary anastomosis becomes feasible after 5–6 weeks of life. In both categories we generally attempt to construct the bidirectional cavopulmonary anastomosis before the 6th month of life. Associated lesions, if present, are usually corrected at the time of construction of the bidirectional cavopulmonary anastomosis. The Fontan circulation is completed as an extracardiac conduit modification, in order to create a maximally streamlined cavopulmonary connection, which in my mind is the most important factor for the maintenance of a well-functioning circulation.