Intermediate-term outcome following the fontan operation:
a survival, functional and risk-factor analysis

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Abstract

Objective: To investigate survival and risk factors influencing intermediate outcome after the Fontan procedure. Methods: Retrospective analysis of 122 patients operated between April 1991 and September 2002. Poor outcome was defined as late death or poor functional status (intractable supraventricular arrhythmias/NYHA 3–4) necessitating revision surgery. Results: 64 (52%) patients had an intermediate bi-directional cavo-pulmonary shunt (BCPS). 91 (76%) patients had a lateral tunnel total cavo-pulmonary connection, 21 (17%) patients had an atrio-pulmonary connection and 10 (8%) patients had a Kawashima connection. There were 6 (5%) early deaths. Over a median follow up of 54 months (1–133), 12 (10%) patients had had surgical revision for poor functional status. There were 7 (6%) late deaths, 5 of which occurred after revision surgery. Univariate analysis identified older age at operation (>4 years) (P = 0.04), higher postoperative pulmonary artery pressure at 24 h (P = 0.012), arrhythmia postoperatively (P = 0.03) or during follow-up (P = 0.01) and the requirement for anticoagulation during follow-up (P = 0.03) as significant predictors of poorer outcome. Patients who had an intermediate BCPS (P = 0.002) or Norwood Stage 1 (P = 0.05) had a better outcome. Multivariate analysis identified an intermediate Glenn shunt and lower postoperative pulmonary artery pressure as significant predictors of better outcome. Actuarial freedom from death or revision is 93% (±2), 88% (±3), 86% (±4) and 69% (±7) at 1, 5, 7 and 9 years respectively. Actuarial freedom from death or revision for the lateral tunnel group is 92% (±2), 89% (±3), 85% (±5) and 66% (±10) respectively. Conclusion: Atrio-pulmonary connection results in a higher incidence of arrhythmias and failure than the lateral tunnel Fontan. Even in patients with a lateral tunnel Fontan there is a continuing hazard phase in the intermediate term. Mid-term outcome appears to be favorably influenced by an intermediate BCPS.

Keywords: Fontan; Lateral tunnel; Mid-term

1. Introduction

Fontan and Baudet first successfully demonstrated the clinical feasibility of pulmonary artery perfusion without a pumping chamber in 1968, in a patient with tricuspid atresia using a direct atrio-pulmonary connection [1]. However, the incorporation of the entire systemic atrium in the Fontan circulation was associated with a high incidence of supraventricular arrhythmias (SVT) and thrombo-embolic complications [2].

The lateral tunnel Fontan (total cavopulmonary connection) was described by de Leval and associates [2] and Jonas and Castaneda [3] in 1988. Long-term results have reported survival of more than 90% and freedom from new SVT of more than 90% at 10 years [4]. Since the late 1990’s prosthetic tube grafts have been used as an alternative in some centers to accomplish an extracardiac connection between the IVC and the pulmonary artery, the so-called ‘extracardiac Fontan’. Theoretical advantages include the avoidance of aortic cross-clamping and the avoidance of atrial suture lines with the complete exclusion of the atrial wall from high venous pressures. This could potentially result in a lower incidence of atrial arrhythmias. Potential disadvantages include the lack of growth potential of the circumferential conduit and the need for formal anticoagulation. Satisfactory mid-term results have been reported with the extracardiac Fontan [5-8]. However in the absence of randomized evidence the debate over the lateral tunnel versus the extracardiac Fontan remains unresolved.

At our institute we have performed 122 Fontan operations over a 10-year period. In the early part of our experience we employed the direct atrio-pulmonary connection. Since 1995 we have exclusively used a fenestrated lateral tunnel Fontan (n = 91, 75%). The purpose of this study was to review our experience with the Fontan procedure over a 10-year period.
and determine any patient and procedure-related risk factors that might influence intermediate-term outcome.

2. Material and methods

2.1. Definitions

Early death was defined as mortality within 30 days of operation or within the first hospital admission. Poor outcome was defined as late death or poor functional status (New York Heart Association NYHA 3 or 4 and/or intractable supraventricular arrhythmias-SVT) requiring revision surgery.

2.2. Patient characteristics

122 consecutive patients undergoing the Fontan procedure between April 1991 and September 2002 were identified from the databases of the Department of Congenital Heart Disease at Guy's Hospital, London (Table 1). The medical records, pre-operative echocardiographic and cardiac catheterization data, and operative notes were reviewed for each patient.

2.3. Operative technique (lateral tunnel Fontan)

All patients underwent a pre-operative trans-thoracic echocardiogram. All procedures were performed using cardiopulmonary bypass (32 °C) and cardioplegic arrest.

The intermediate bi-directional cavopulmonary shunt (BCPS) was performed using the technique described by Norwood (Hemi-Fontan) [10]. A patch of pulmonary homograft was used to augment the superior vena cava (SVC) to right pulmonary artery anastomosis. The patch was folded inward to close off the junction of the right atrium and the SVC in a circumferential fashion. If a pulmonary arterioplasty is required, it was usually performed at this operation. More recently a longitudinal incision in the inferior circumference of the SVC is anastomosed to a longitudinal incision in the superior surface of the underlying right pulmonary artery. The SVC is left in continuity with the right atrium and a circular patch of Gore-Tex™ (W.L Gore & Associates, Inc, Flagstaff, Arizona, USA) is used to close the SVC-right atrium junction.

At the time of the Fontan operation the right atrium was opened anterior and parallel to the crista terminalis. After ensuring adequate communication between the 2 atria, a Gore-Tex™ patch was inserted so as to direct blood flow from the inferior vena cava (IVC) to the orifice of the SVC or the superior end of the right atrium, incorporating a portion of the lateral wall of the right atrium. In patients who have had an intermediate BCPS the circular patch of Gore-Tex™ the junction of the right atrium and the SVC was excised and the Gore-Tex™ patch was sutured to the medial margins of the junction. Near the IVC the suture line was carried medial to the coronary sinus leaving it to drain into the lateral tunnel. A 4 mm fenestration was created in the Gore-Tex patch using a coronary punch. The margins of the fenestration were marked with 2 liga-clips to facilitate identification at the time of subsequent cardiac catheterization.

2.4. Follow-up

All patients were followed up by a pediatric cardiologist from our institution. All patients receive a physical examination and a trans-thoracic echocardiogram at each follow-up visit. All cardiac catheterization procedures were carried out at Guy's Hospital.

2.5. Statistical analysis

Variables are reported as mean (± SD) or median (range) where necessary. Continuous variables were analyzed using the t-test and categorical variables were analyzed using chi-square test or Fisher’s exact test. Variables with a P < 0.1 in the univariate analysis were entered into a multiple logistic regression model. A P value < 0.05 was considered significant. Actuarial survival and freedom from revision were analyzed using the Kaplan-Meier method. Statistical analysis was performed using the SPSS version 10.0 software package (SPSS, Inc, Chicago, Illinois, USA).

3. Results

3.1. Morphological characteristics

3.1.1. Anatomy

In general patients belonged to one of 5 groups (Table 2). The main anatomical abnormalities are shown in Table 3.

3.1.2. Ventricle

Ventricular function as assessed by echocardiography was good in 112 (92%) patients, moderate in 4 (3%), poor in 1 patient and unknown in 5 (4%) patients. The dominant ventricle was morphologically the left-sided ventricle in 70 (57%) patients, the right-sided ventricle in 50 (41%) patients and indeterminate in 3 (2%) patients.

3.1.3. Valve

18 (15%) patients had a common atrio-ventricular valve (AVV). There was mild atrio-ventricular valve (AVV) regurgitation in 28 (23%) patients and moderate AVV regurgitation in 3 (2%) patients.

Table 2
Morphological grouping (LV: left ventricle, TGA: transposition of great arteries, RV: right ventricle, HLHS: hypoplastic left heart syndrome)

<table>
<thead>
<tr>
<th>Diagnostic group</th>
<th>Patients (n=122)</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV/normally related great arteries</td>
<td>38 (31%)</td>
</tr>
<tr>
<td>LV/TGA</td>
<td>32 (26%)</td>
</tr>
<tr>
<td>RV/Indeterminate ventricle</td>
<td>14 (11%)</td>
</tr>
<tr>
<td>HLHS</td>
<td>19 (16%)</td>
</tr>
<tr>
<td>Heterotaxy</td>
<td>19 (16%)</td>
</tr>
</tbody>
</table>
3.2. Demographic data

There were 80 (66%) males and 42 (44%) females (2:1). The median weight at operation was 18.5 kg (6–116). The mean age at operation was 67 months (G36). The age distribution is shown in Table 4.

3.3. Previous operation

111 (91%) patients had a total of 192 previous procedures. 19 (15%) patients had a Norwood Stage 1 and 22 (18%) had a pulmonary artery band. 54 (44%) patients had one systemic-pulmonary artery shunt, 17 (13%) patients had 2 previous arterial shunts and 2 patients had three previous arterial shunts. 64 (52%) patients had an intermediate BCPS (Table 5).

3.4. Pre-operative investigation

95 (78%) patients had a pre-operative cardiac catheterization. 13 (11%) patients had a preoperative magnetic resonance angiography (MRA). 11 (9%) patients had both investigations and 26 (21%) patients had only a trans-thoracic echocardiogram before their completion Fontan. All patients after 2001 have had either a cardiac catheterization or a magnetic resonance angiography before their Fontan. Mean pulmonary artery pressure at the time of cardiac catheterization was 13.9 mmHg (±4.8). 17 (14%) patients had varying degrees of pulmonary artery distortion identified. 4 (3%) patients required a pulmonary arterioplasty at the time of their Fontan.

3.5. Pre-operative arrhythmia

10 (8%) patients had a history of arrhythmia before their Fontan operation. 6 (5%) patients were taking anti-arrhythmic medication.

3.6. Pre-operative anticoagulation

4 (3%) patients were taking warfarin at the time of admission. All these patients had a documented history of an arterial or venous thrombotic event in the past. 10 (8%) patients were taking aspirin.

3.7. Surgical considerations

91 (75%) patients had a lateral tunnel Fontan. 21 (17%) patients had an atrio-pulmonary connection and 10 (8%) patients with an interrupted IVC and hemi-azygos continuity had a bilateral bi-directional superior cavo-pulmonary anastomosis (Kawashima connection) [9]. Since 1995 we have only performed the lateral tunnel Fontan as our standard technique. 2 surgeons performed all operations over the 10-year period (DA 115, CA 7). 97 (80%) patients had a fenestration. 87 of the 91 patients (96%) with a lateral tunnel Fontan were fenestrated. 59 (65%) of the lateral tunnel procedures and 5 (50%) of the Kawashima connections were preceded by an intermediate BCPS. The median age at BCPS was 64 months (1–360). All the atrio-pulmonary connections were performed as a single-stage Fontan. 15 additional procedures were performed concomitantly with the Fontan operation: atrial septal defect closure (2), Damus-Kaye-Stenzel anastomosis (2), pulmonary artery reconstruction (4), correction of total anomalous pulmonary venous drainage (1), closure of pulmonary valve (1), ventricular septal defect enlargement (2), shunt takedown (2).

3.8. Peri-operative data

The median cardiopulmonary bypass time was 90 min (30-172) and the median cross-clamp time was 49 min (16-121). 6 patients required deep hypothermic circulatory arrest (median 19 min, 11-36). The median duration of ventilation was 8 h (1-48) and the median length of stay in the intensive care was 48 h (24-120). Mean post-operative pulmonary artery pressure at 4 h and 24 h was 14 (±3) and 13 (±3) mmHg respectively. The median requirement of vasoactive drugs was 24 h (12-96). The median hospital stay was 11 days (3-46).

3.9. Morbidity

3.9.1. Post-op arrhythmia

17 (14%) patients had a significant new arrhythmia (lateral tunnel n=13, 14%). The median duration of
arrhythmia was 26 h (1-120). 12 patients required anti-arrhythmic medications. 16 patients had their medication discontinued by 8 weeks. Only 1 patient with atrial flutter from the lateral tunnel group with atrial flutter was still taking sotalol at follow-up. 3 patients (lateral tunnel n=1) required implantation of a permanent pacemaker (PPM).

3.9.2. Post-op anticoagulation

16 (13%) patients were prescribed warfarin post-operatively. The indications for warfarin were SVC/IVC thrombus (3), pulmonary embolism (4), atrial fibrillation (1), ischemic bowel (1), intra-tunnel thrombus (1), sluggish intra-tunnel flow (1) and reduced ventricular function (5). In the lateral tunnel group 9 (10%) patients were prescribed warfarin postoperatively (SVC/IVC thrombus n=3, sluggish intra-tunnel flow n=1, intra-tunnel thrombus n=1, suspected pulmonary embolism n=3, unclear n=1). 46 (38%) patients were discharged on aspirin (36 of 44 patients (82%) operated since 2000).

Other complications are listed in Table 4. 40 patients (33%) made an uneventful recovery.

3.10. Early death (n=6)

There were 6 (5%) early deaths (atrio-pulmonary connection n=2, lateral tunnel n=4). In 4 patients the cause of death was predominantly cardiac (cardiac arrest n=3). 1 patient required extracorporeal membrane oxygenation following a re-sternotomy for bleeding. He subsequently succumbed to complications of acute renal failure. 1 patient died of septic complications.

3.11. Follow-up

Follow-up is complete in 102 of 109 late survivors (94%). Median duration of follow-up is 54 months (1-133).

3.11.1. Functional class

63 (57%) patients are in NYHA 1, 42 (38%) are in NYHA 2 and 5 (5%) are in NYHA 3.

3.11.2. Fenestration Fate (n=97, 80%)

The fenestration is known to be occluded in 40 patients (41%). 9 closed spontaneously and 31 were closed with a device by an interventional cardiologist at a median of 19 months (3-121) after operation. The fate of the fenestration could not be reliably ascertained in the remaining patients.

3.11.3. Arrhythmia

26 (21%) patients developed a new supraventricular arrhythmia during follow-up. This includes 16 of 91 patients (17%) from the lateral tunnel group and 6 of 21 (28%) from the group with an atrio-pulmonary connection. The arrhythmias include supraventricular tachycardia (13), atrial flutter (9), bradycardhythmia (1), atrial fibrillation (1) and nodal rhythm (1). 16 (13%) patients are taking anti-arrhythmic medication at follow-up. 9 (8%) patients have had a radiofrequency ablation and 8 (7%) patients have had permanent pacemaker implantation. In the lateral tunnel group new arrhythmias include atrial flutter (8), supraventricular tachycardia (6), bradycardhythmia (1) and nodal rhythm (1). The latter 2 patients have received a permanent pacemaker. 4 patients have undergone successful radiofrequency ablation and 11 patients were taking anti-arrhythmic medication.

3.11.4. Anticoagulation

Of the 16 (13%) patients who were started on warfarin post-operatively, 12 (9%) patients were still on warfarin at their last follow-up. In addition warfarin was started on 9 (8%) patients during follow-up. The indications for warfarin (n=19, 16%) were arrhythmia (10), intra-cardiac thrombus (4), venous thrombosis (4), pulmonary embolism (2), ischemic bowel (1), right pulmonary artery stent (1) and embolic cerebro-vascular accident (1). There has been only 1 complication (nose bleed) related to warfarin. In addition to the 46 patients who were discharged on aspirin, 19 additional patients were started on aspirin during follow-up (n=65, 56%).

Of the 9 patients in the lateral tunnel group (n=91, 10%) who were prescribed warfarin postoperatively, 5 patients continued to take warfarin at follow-up (SVC/IVC thrombus n=3, sluggish intra-tunnel flow n=1, intra-tunnel thrombus n=1). 8 (n=91, 8%) additional patients were prescribed warfarin during follow-up (atrial flutter n=4, intra-tunnel thrombus n=2, pulmonary embolism n=1, poor ventricular function n=1). Only 2 of these patients were discharged on aspirin. Both these patients received prophylactic warfarin for arrhythmias.

3.11.5. Protein-losing enteropathy (PLE)

1 patient who underwent revision surgery also had PLE. He died during the early post-operative period despite Fontan takedown.

3.12. Poor outcome

3.12.1. Revision surgery

12 (9%) patients have required revision surgery. 10 patients had their Fontan operation before 1995. Preoperative anatomy was heterotaxy (5), tricuspid atresia (4), pulmonary atresia (1), DILV (1) and HLHS (1). 11 patients had good ventricular function before the Fontan procedure. 4 patients had a staged Fontan (intermediate BCPS). 6 patients had a lateral tunnel connection, 3 patients had an atrio-pulmonary connection and 3 patients had a Kawashima connection. The median time to revision was 73 months (20-112).

All patients were in NYHA 3 or 4 or had very dilated right atriums with intractable arrhythmias. Patients with an atrio-pulmonary connection were converted to a lateral tunnel connection. In patients with a previous lateral tunnel, the right atrium was trimmed, a partial right-sided Maze procedure performed and the lateral tunnel re-fashioned. 3 patients with heterotaxy (Kawashima connection) had persistent low saturations despite coil occlusion of collaterals on more than one occasion. In all these patients a lateral tunnel was constructed in an attempt to divert hepatic venous blood to the pulmonary arteries. 1 patient had pulmonary venous obstruction secondary to a fibrous ridge around the intra-atrial baffle and 1 patient had near-total occlusion of his lateral tunnel by thrombus. 1 patient...
had a very dilated right atrium with severe left AVV regurgitation. He underwent a revision of his lateral tunnel with mechanical left AVV replacement.

4 patients died in the early post-operative period (cardiac n=3, massive hemoptysis n=1). The patient with massive hemoptysis had angiographic evidence of significant pulmonary arterio-venous fistulae preoperatively and had undergone an earlier Kawashima connection. 1 patient presented 4 months later with massive pericardial effusion and succumbed to cardiac failure despite drainage.

7 patients are alive and well at a median of 15 months (1-45) after their revision surgery. All patients are in NYHA 1 or 2. 5 patients are in a normal sinus rhythm.

3.12. Late death (n=7)

There have been 7 (5%) late deaths. 5 patients died after revision surgery (see above). 2 patients had a fatal cardiac arrest at home (6 and 14 months post-operatively respectively). Both patients were alive and well until their demise and an arrhythmia is presumed to be the cause of death. Actuarial freedom from death or revision is 93% (±2), 88% (±3), 86% (±4) and 69% (±7) at 1, 5, 7 and 9 years respectively (Fig. 1). Actuarial freedom from death or revision for the lateral tunnel group is 92% (±2), 89% (±3), 85% (±5) and 66% (±10) respectively (Fig. 2).

3.13. Risk factor analysis

Univariate predictors of poor outcome or good outcome are shown in Table 6. Multivariate analysis identified only intermediate BCPS (P=0.002) and pulmonary artery pressure less than 14 mmHg at 4 or 24 h postoperatively (P=0.03) as independent predictors of good outcome.

4. Limitations

The most obvious limitation of our study is its retrospective nature. The incidence of arrhythmias is likely to be underestimated because of their transient and intermittent nature. Therefore only a fraction of them are recorded and receive treatment. Similarly the incidence of thrombo-embolic complications is probably underestimated because only the clinically relevant events receive attention.

Our study represents the evolution over a decade of our experience with the Fontan procedure over a decade. Much of our technique (lateral tunnel with fenestration) and post-operative management is now standardized and the impact of the lessons learned through experience will not be reflected in the intermediate-term outcome of the entire cohort. Only prospective analysis of the patients operated more recently will provide further clarification with regard to long-term mortality and morbidity of the patients with a fenestrated lateral tunnel Fontan.

Caution is also recommended in the interpretation of the risk factor analysis. The cohort is limited and the absolute number of events is small.

5. Discussion

As the number of surviving young adults with a single ventricle continues to increase, there is an increasing urgency to either find adequate and appropriate future therapy, or as a minimum, optimize the Fontan procedure.

Table 6

<table>
<thead>
<tr>
<th>Risk factor</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Poor outcome</td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td>0.66</td>
</tr>
<tr>
<td>Dominant ventricle=left</td>
<td>0.21</td>
</tr>
<tr>
<td>Age &gt;4 years</td>
<td>0.04*</td>
</tr>
<tr>
<td>Common AVV</td>
<td>0.29</td>
</tr>
<tr>
<td>Isomerism</td>
<td>0.16</td>
</tr>
<tr>
<td>Post-operative arrhythmia</td>
<td>0.03*</td>
</tr>
<tr>
<td>PAP &gt;14 mmHg (at 4 or 24 h post-operatively)</td>
<td>0.01*</td>
</tr>
<tr>
<td>Late-onset arrhythmia</td>
<td>0.03*</td>
</tr>
<tr>
<td>Anticoagulation at follow-up</td>
<td>0.16</td>
</tr>
<tr>
<td>Implantation of permanent pacemaker</td>
<td>0.01*</td>
</tr>
<tr>
<td>Good outcome</td>
<td></td>
</tr>
<tr>
<td>Intermediate BCPS</td>
<td>0.002*</td>
</tr>
<tr>
<td>Previous Norwood Stage</td>
<td>0.05*</td>
</tr>
</tbody>
</table>

Fig. 1. Kaplan Meier Freedom from Death or Revision.

Fig. 2. Kaplan Meier Freedom from Death or Revision in the Lateral Tunnel Fontan Patients.

![Graph](image-url)
An improved risk analysis for these patients plays a key role in improving patient selection for the completion Fontan.

Currently in our hospital we elect to perform an intermediate BCPS in all patients between 6-12 months followed by a fenestrated lateral tunnel completion Fontan at 24-36 months of age. The benefits of a staged Fontan procedure have been previously described [10–13] and was further substantiated by multivariate analysis in our series.

Advances in magnetic resonance imaging (MRI) now allow comprehensive 4-dimensional (3-dimensional spatial imaging over time) in children with congenital heart disease. MRI is noninvasive and does not involve biologically harmful ionizing radiation. We have gained considerable experience with MRI over the past few years [14,15] and we believe that MRI together with trans-thoracic echocardiography provides sufficient information pre-operatively to safely undertake the Fontan procedure. More recently, at our institute, there has been a decline in the use of invasive catheterization procedures performed solely for diagnostic indications before the Fontan operation.

There is good evidence that a baffle fenestration of the Fontan circulation is beneficial not only in the high-risk individual [16,17] but also in standard risk Fontan patients (shorter hospital stay, decreased pleural drainage and fewer additional procedures) [18]. We currently fenestrate all total cava-pulmonary connections at our institution and have ceased active post-operative closure due to low oxygen saturation as this has been shown to be significantly associated with a poor outcome [18].

There were six early deaths (5%) in our series (1991-1997: 4, 1997-2002: 2). This is comparable to earlier reports from The Children′s Hospital of Philadelphia, Mayo Clinic and Children′s Hospital, Boston [4,17,19]. This improvement in early outcome is most likely due to the performance of a staged Fontan as well as the use of a fenestration. In addition, improvements in perfusion strategy, anesthesia and intensive care management over the past decade have probably made important contributions.

Early and late onset arrhythmias especially supraventricular arrhythmias remain a challenge after the Fontan operation with a reported early incidence of 15 and 17% at 5 years [20]. In our series, 22 (18%) of the early survivors of the Fontan operation (n = 116) developed supraventricular arrhythmias during follow-up. Patients with an atrio-pulmonary connection had a higher incidence of postoperative arrhythmias as compared to the lateral tunnel group, which is similar to what has previously been reported [21]. We also found that arrhythmias at anytime post-operatively were significantly associated with a poorer outcome. This probably is a reflection of the higher incidence of arrhythmias and poor outcome in patients with an atrio-pulmonary connection in our cohort (see later). Though experience with the extra-cardiac Fontan has resulted in an appreciably lower rate of supraventricular arrhythmias [4–6,8] concerns still remain with regard to longer-term follow up and consequently its application is not uniform. Currently there is no randomized evidence that clearly establishes the superiority of one technique over the other.

Of the 9 patients with a lateral tunnel who received warfarin post-operatively, 5 patients had a documented thrombo-embolic complication (SVC/IVC thrombosis n = 3, intra-tunnel thrombus n = 1). The SVC and IVC thrombosis was undoubtedly related to the peri-operative placement of internal jugular and femoral venous lines. This complication can be potentially avoided by careful attention to low-dose heparin infusion while the lines are in situ or alternatively by using only large-bore peripheral access together with intra-cardiac lines [22]. Of the 8 additional patients who were prescribed warfarin during follow-up only 3 patients developed a primary thrombo-embolic event (intra-tunnel thrombus n = 2, pulmonary embolism n = 1). In the remaining 5 patients the indication for warfarin was primarily prophylactic (atrial flutter n = 4, poor ventricular function n = 1). None of the patients with thrombo-embolic complications had received aspirin postoperatively. In fact only 2 of the 13 patients with a lateral tunnel who were receiving warfarin at follow-up were discharged on aspirin. Thus the potential benefit of warfarin in our patients remains speculative. There is no randomized evidence to support the routine use of warfarin. We believe that regular aspirin confers adequate protection from thrombo-embolism and all our patients, in the absence a documented history of venous or arterial thrombosis, are now routinely prescribed aspirin on postoperative day one and are discharged on aspirin. This feeling is shared by others [22]. Only 56% of our patients were on aspirin at the time of the study. We have since commenced all our remaining patients on aspirin. It must be noted that there is some evidence that the majority of the thrombo-embolic complications occur within the first year and anticoagulation may be indicated only during this period [23].

14 (11%) patients had a poor outcome in the intermediate-term. 12 (10%) required revision surgery, 10 of whom had the Fontan procedure before 1995. Gentles et al. [24] showed a late failure rate of 9% in their series of 500 patients and identified heterotaxy and atriopulmonary type connections as significant risk factors for failure. Stamm et al. [4] had a late failure rate of 6% in their series of 220 patients with a lateral tunnel Fontan. In our series there was a higher incidence of revision surgery in the patients with an atrio-pulmonary connection (3 of 21 patients: 14% versus 6 of 91 patients with a lateral tunnel Fontan: 6%). 5 patients (5%) were in NYHA 3 at the time of the study. Gentles et al. and Stamm et al. reported a mid-term NYHA 3 rate of 8.9 and 6% respectively [4,24]. 4 of these patients received an atrio-pulmonary connection prior to 1995 and all have dilated right atriums with recurrent SVT. Revision surgery is being considered in all these patients. If these patients are included then 7 of 21 patients (33%) with an atrio-pulmonary type connection can be deemed to have had a poor outcome (P = 0.006).

Mavroudis et al. demonstrated that a total cavopulmonary conversion with a modified Maze procedure could be performed with no early mortality [25], and an arrhythmia recurrence rate of 12.5% at a mean of 30 months. 38 of the 40 patients in their series received an extra-cardiac conduit. We had 5 deaths (4 in the early post-operative period) amongst the 12 patients who required revision surgery in our series. All these patients underwent revision surgery at an advanced stage of ventricular dysfunction. Since this study we have followed a policy of early identification of patients with dilated right atriums. These patients are followed-up at
shorter intervals and are referred for consideration of revision surgery when any deterioration of symptoms or an arrhythmia supervenes. We believe that early operation and the use of an extracardiac conduit may represent a better alternative in this group of patients.

The atrio-pulmonary connection appears to have a higher incidence of arrhythmia and failure in the intermediate term. As nearly all these operations were performed before 1995, whether the difference in the outcome is a true reflection of the surgical technique or merely the longer follow up of the atrio-pulmonary connection can only be answered by continuing to prospectively monitor this cohort of patients. Clinicians are often surprised by the ability of these patients to lead fairly normal lives. 95% of the late survivors in our study were in NYHA 1 or 2 at last follow-up. Nevertheless even for patients with a lateral tunnel, the Fontan procedure demonstrates a continuing hazard phase in the intermediate term. Future efforts should concentrate on optimizing patient selection, randomized evaluation of different surgical techniques and early re-intervention of the failing Fontan.

References