Extracardiac versus Intra-Atrial Lateral Tunnel Fontan: Extracardiac is Better

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I have no conflicts of interest to disclose...
(except a personal rule never to debate Dr. Richard Jonas)!
Modifications of the Fontan operation applicable to patients with left atrioventricular valve atresia or single atrioventricular valve

FRANCISCO J. PUGA, M.D., MARIO CHIAVARELLI, M.D., AND DONALD J. HAGLER, M.D.

ABSTRACT Two modifications of the Fontan operation are proposed for prevention of pulmonary venous obstruction in patients with left atrioventricular valve atresia or single atrioventricular valve. In the first technique, a bipedicled flap of right atrial wall is used to separate systemic and pulmonary venous drainages; an atrio pulmonary connection is constructed by use of the right atrial appendage, and the defect in the right atrial wall is repaired with a pericardial patch. In the second technique, the superior vena cava is transected and the cardiac and cephalad ends are anastomosed separately to the pulmonary arterial confluence. A patch is positioned inside the right atrium to separate the systemic from the pulmonary venous drainage. Both techniques achieve unrestricted systemic and pulmonary venous channels. Severe atrial arrhythmias, present in the four patients operated on with the first procedure, were not seen in 12 patients who had repair with the second procedure. 

Total cavopulmonary connection: A logical alternative to atriopulmonary connection for complex Fontan operations

Experimental studies and early clinical experience

To understand better the contribution of a right atrium in a valveless atriopulmonary connection, we performed some basic hydrodynamic studies. Pulsation of a valveless chamber in a simple continuous flow circuit was found to generate turbulence and thereby increase resistance to net forward flow. Measurements of energy losses across nonpulsatile cavities, corners, and stenoses suggested ways in which hydrodynamic designs of the Fontan circulation might be improved. In parallel with these in vitro studies, we have developed a modified approach to Fontan reconstruction that entails exclusion of most or all of the right atrium (total cavopulmonary connection). The operation consists of three parts: (1) end-to-side anastomosis of the superior vena cava to the undivided right pulmonary artery; (2) construction of a composite intraatrial tunnel with the use of the posterior wall of the right atrium; and (3) use of a prosthetic patch to channel the inferior vena cava to the enlarged orifice of the transected superior vena cava that is anastomosed to the main pulmonary artery. The operation was performed in 20 patients between March 1987 and March 1988, of whom 11 had double-inlet ventricles, seven had hypoplastic systemic or pulmonary ventricles, and two had absent right atrioventricular connections. There were two early deaths and one death from late atrial thrombosis (two patients). None of the deaths was related to the actual procedure but rather to increased pulmonary vascular resistance (two patients) or systemic ventricular failure (one patient). Total cavopulmonary connections have the following advantages: (1) They are technically simple and reproducible in any atrioventricular arrangement and are away from the atrioventricular node; (2) most of the right atrial chamber remains at low pressure, which reduces the risk of early or late arrhythmias; (3) reduction of turbulence prevents energy losses and should minimize the risk of atrial thrombosis; (4) postoperative cardiac catheterization performed in 10 patients confirmed these favorable flow patterns with minimal gradients throughout the connections. These encouraging early results support the continuing use of total cavopulmonary connection, at least for patients with a nonhypertrophied right atrium.

Marc R. de Leval, MD (by invitation), Philip Kilner, MD (by invitation), Marc Gewillig, MD (by invitation), and Catherine Bull, MD (by invitation), London, England
Sponsored by Dwight C. McGoon, MD, Rochester, Minn.
The modified Fontan operation for asplenia and polysplenia syndromes

From 1975 through 1986, 49 patients with asplenia (23 patients) or polysplenia (26 patients) syndromes underwent a modified Fontan operation. All patients had anomalous systemic venous return, and 43 (88%) had anomalous pulmonary venous return. The atrioventricular valve anatomy varied: 36 patients had a common atrial chamber and common atrioventricular valve, eight had mitral valve atresia, and five had severe mitral valve hypoplasia. Redirection of systemic venous return was accomplished by (1) atrial baffle (29 patients), (2) intraatrial conduit (19 patients), and (3) extraatrial conduit (one patient). From 1975 through 1986, six of 22 patients (27%) died, and in patients requiring atrioventricular valve repair or replacement, eight of 11 patients (73%) died. Lower mortality was seen in patients with polysplenia (24%) and those receiving an intraatrial conduit (26%), although this decrease also represents the more recent experience. We conclude: (1) The Fontan operation for patients with asplenia or polysplenia syndromes has resulted in a significant mortality; (2) mortality is higher in patients with asplenia; (3) patients with atrioventricular valve insufficiency requiring repair or replacement are at higher risk; and (4) intraatrial conduits seem to offer promise for successfully accomplishing this repair.

Inferior vena cava-pulmonary artery extracardiac conduit

A new form of right heart bypass

From November 1988 to May 1989, four patients underwent total right heart bypass by means of bidirectional cavopulmonary anastomosis and interposition of an extracardiac conduit from the inferior vena cava to the pulmonary artery. All of them had an uneventful postoperative course, and there have been no early or late deaths. We propose this technique as an alternative surgical option in candidates for a Fontan procedure with (1) hypoplasia or atresia or the left atrioventricular valve, (2) common atrioventricular valve, (3) anomalies of systemic and pulmonary venous return, or (4) auricular juxtaposition.

Carlo Marcelletti, MD, Antonio Corno, MD, FACC, Salvatore Giannico, MD, and Bruno Marino, MD, Rome, Italy
Children’s Memorial Hospital
Transition from LT to EC Fontan

Fontan conversion series began

Lateral Tunnel, n=67
Extracardiac, n=113
In the failed atriopulmonary Fontan, atrial exposure to elevated CVP led to atrial distension and arrhythmias.

Our strategy for Fontan conversion has been to use extracardiac Fontan with arrhythmia surgery.

This experience influenced our primary Fontan strategy to favor extracardiac Fontan.
Primary Fontan at Children’s Memorial Hospital

• From 1990-2009, 180 patients underwent primary Fontan
  ✓ Fenestrated lateral tunnel (n = 67)
  ✓ Non-fenestrated extracardiac (n = 113)

• Age range, 9 months - 43 years
  ✓ Median, 3.3 years

• Previous staging operations in 78% (97% since 2000):
  ✓ Bidirectional Glenn (n = 112, 62.2%)
  ✓ Unidirectional Glenn (n = 14, 7.8%)
  ✓ Hemi-Fontan (n = 15, 8.3%)
Surgical technique

- Bicaval CPB – 32°C
- No cross-clamp
- 16-20 mm PTFE graft
- Prophylactic atrial pacing lead
- Early extubation
Results

**CPB time**

- LT: 156.4 min
- EC: 101.7 min

**Xclamp time**

- LT: 56.2 min
- EC: 17.6 min

**p<0.0001**
Length of Stay

All patients:
Mean 15.4 ± 10.7 days
Median 11 days
# Mortality and Takedown

<table>
<thead>
<tr>
<th></th>
<th># Pts</th>
<th>Op Mortality</th>
<th>Takedown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lateral Tunnel</td>
<td>67</td>
<td>2 (3%)</td>
<td>4 (6%)</td>
</tr>
<tr>
<td>Extracardiac</td>
<td>113</td>
<td>1 (1%)</td>
<td>5 (4%)</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td>180</td>
<td><strong>3 (1.7%)</strong></td>
<td><strong>9 (5%)</strong></td>
</tr>
</tbody>
</table>

\[ p = 0.56 \quad p = 0.73 \]
Extracardiac Fontan Procedure at CMH

Conclusions

- Very low operative mortality rate (0.9%)
- EC Fontan takedown (4.4%), not different from LT (6%)
- Significantly lower CPB and aortic cross-clamp times (p<0.001)
- Postoperative LOS same as for LT with fenestration
- Our procedure of choice since 2002
Supporting evidence from the literature on the *superiority of extracardiac Fontan* and reasons to avoid lateral tunnel Fontan

- Hemodynamics
- Arrhythmias
- Operative mortality
- Potential complications
- Late results
Computational fluid dynamic study of flow optimization in realistic models of the total cavopulmonary connections

Tain-Yen Hsia, MSc, Gabriele Dubini, PhD, Giancarlo Pennati, PhD, and Marc de Leval, MD, London, UK.

“Of the four basic TCPC geometries, EC angled toward the left lung achieved the least power loss”

“Atrial wall dilation in the LT results in moderate worsening of flow dynamics”
Fluid dynamic comparison of intra-atrial and extracardiac total cavopulmonary connections

OBJECTIVE: Extracardiac total cavopulmonary connection has recently been introduced as an alternative to intra-atrial procedures. The purpose of this study was to compare the hydrodynamic efficiency of extracardiac and intra-atrial lateral tunnel procedures in total cavopulmonary connections.

METHODS: Intra-atrial lateral tunnel, extracardiac tunnel, and extracardiac conduit with and without caval vein offset were performed on explanted sheep heart preparations and studied in an in vitro flow loop. A rate of fluid energy dissipation analysis was performed on each inlet and outlet of the right side of the heart. Preparations were perfused by using a steady flow blood pump at 4 flow indices (1-6 L/min/m²) with the inferior vena cava carrying 65% of the total venous return.

RESULTS: Fluid power losses were consistently lower for the extracardiac conduit procedure compared with the two tunnel configurations (P < .01). A further reduction in energy dissipation of up to 36% was noted in the extracardiac procedure, with 5 mm offset of the extracardiac conduit toward the distal right pulmonary. The intra-atrial and extracardiac tunnel procedures were least efficient, with losses 73% greater than the optimal extracardiac conduit procedure.

CONCLUSIONS: The extracardiac conduit procedure provides superior hemodynamics compared with the intra-atrial lateral tunnel. This hydrodynamic advantage is markedly enhanced by the use of conduit-superior vena cava offset, particularly at high physiologic flows that are representative of exercise. These data suggest additional rationale for the use of extracardiac conduit procedures for final-stage completion of the Fontan circulation.

Albert C. Lardo, MD, Steven A. Webber, MD, Ingeborg Friehs, MD, Pedro J. del Nido, MD, and Edward G. Cape, PhD, Pittsburgh, PA.
Surgical repair of tricuspid atresia

F. FONTAN and E. BAUDET

Centre de Cardiologie, Université de Bordeaux 11, Hôpital du Tondu, Bordeaux, France

Surgical repair of tricuspid atresia has been carried out in three patients: two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, ‘ventriclized’, to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficient pressure to allow a cava-pulmonary anastomosis.

“One element remains unpredictable—the haemodynamic consequences of an eventual atrial rhythm disturbance such as an atrial fibrillation or flutter.”
Arrhythmias

• Lateral tunnel Fontan requires extensive atrial suture lines
  ✓ Scarring along suture lines
  ✓ Potential damage to SA node and/or blood supply
• Atrial exposure to elevated CVP leads to distension and arrhythmias
• LT Fontan may have the same problems found in atriopulmonary Fontan leading to Fontan conversion
New onset arrhythmias after the extracardiac conduit Fontan operation compared with the intraatrial lateral tunnel procedure: early and midterm results

BACKGROUND: Arrhythmias are one of the main causes of postoperative morbidity superseding Fontan operations. Comparative data on the incidence of sinus node dysfunction after the extracardiac Fontan operation (ECFO) and the intraatrial lateral tunnel Fontan operation (LTFO) are very limited and controversial. The aim of this study was to evaluate whether ECFO decreases the risk of postoperative arrhythmias compared with LTFO.

METHODS: Seventy-four consecutive patients received either an LTFO (n = 29) or an ECFO (n = 45). The rhythm was documented preoperatively and postoperatively with standard electrocardiogram (ECG) recording and ECG monitoring. During follow-up all patients had 2-8 (median 3) standard ECG recordings per year. Additionally, 45 patients (65%) had a Holter ECG at least once a year. Median follow-up post-ECFO was 4.4 years (1.6-7.2) and post-LTFO it was 7.9 years (5.4-11.1). Sinus rhythm persisted in 37 (86%) ECFO patients (38% supraventricular tachyarrhythmias) and in 13 (50%) LTFO patients (38% supraventricular tachyarrhythmias), respectively. Early postoperatively, 10 LTFO patients (34%) and another 3 patients during follow-up required permanent pacemaker implantation due to bradyarrhythmias, but none of the ECFO patients required this.

CONCLUSIONS: Our data suggest that ECFO decreases the incidence of postoperative new onset arrhythmias during early and midterm follow-up compared with LTFO.

Jan Hendrik Nürnberg, MD, Stanislav Ovroutski, MD, Vladimir Alexi-Meskishvili, MD, PhD, Peter Ewert, MD, PhD, Roland Hetzer, MD, PhD, and Peter E. Lange, MD, Berlin, Germany.
Persistence of Sinus Rhythm

LT Fontan is an independent risk factor for new-onset postoperative tachyarrhythmia (p=0.007)

Incision across the cavoatrial junction during LT Fontan is an **independent risk factor** for bradyarrhythmia (p=0.001)

Extracardiac conduit versus lateral tunnel cavopulmonary connections at a single institution: Impact on outcomes

OBJECTIVE: To compare outcomes of extracardiac conduit and lateral tunnel Fontan connections in a single institution over a concurrent time period. METHODS: Between January 1994 and September 1998, 60 extracardiac conduit and 47 lateral tunnel total cavopulmonary connections were performed. Age, sex, and weight did not differ between the 2 groups. Compared with the lateral tunnel group (LT group), patients undergoing the extracardiac conduit procedure (EC group) had a trend to a higher incidence of morphologically right ventricle (EC group 48% vs LT group 32%; P < .09), a higher incidence of isomerism/heterotaxy syndrome (EC group 22% vs LT group 0%; P < .001), worse atrioventricular valve regurgitation (EC group 11% vs LT group 0%; P < .06), and lower McGoon indices (EC group 1.8 +/- 0.5 vs LT group 2.1 +/- 0.5; P < .03). Preoperative transpulmonary gradients, room air oxygen saturations, ejection fractions, ventricular end-diastolic pressure, and pulmonary artery distortion did not differ between groups. RESULTS: Overall operative mortality was 5.6% and did not differ between groups. The LT group had a significantly higher incidence of postoperative sinoatrial node dysfunction (45% vs EC group 15%; P < .007), supraventricular tachycardia (33% vs EC group 8%; P < .0009), and need for temporary postoperative pacing (32% vs 12%; P < .01). Median duration of intensive care unit stay (EC group 2 days, range 1-10 days, vs LT group 2.8 days, range 1-103 days; P < .07) and ventilatory support (EC group 1 day, range 0.25-10 days, vs LT group 1 day, range 0.25-99 days; P < .03) were all longer in the LT group. Median chest tube drainage (EC group 8 days, LT group 9 days) was similar in both groups. Follow-up averaged 2.5 +/- 1.4 years in the EC group and 2.8 +/- 1.9 years in the LT group. There were 2 late deaths. Overall survival is 94% at 1 month, 92% at 1 year, and 92% at 5 years. Late ejection fraction or atrioventricular valve function did not differ between groups. Intermediate follow-up Holter analysis showed a higher incidence of atrial arrhythmias in the LT group (23% vs 7%; P < .02). Multivariable analysis showed that (1) prolonged cardiopulmonary bypass time was the only independent predictor for perioperative mortality, prolonged ventilation and intensive care unit length of stay, and increased time to final removal of chest tube drains and (2) lateral tunnel Fontan connection is an independent predictor of early postoperative and intermediate atrial arrhythmias. CONCLUSIONS: Although patients in the EC group were at higher preoperative risk, their outcomes were comparable with those of the LT group. Use of the extracardiac conduit technique for the modified Fontan operation reduces the risk of early and midterm atrial arrhythmia.

Anthony Azakie, MD, Brian W. McCrindle, MD, Glen Van Arsdell, MD, Lee N. Benson, MD, John Coles, MD, Robert Hamilton, MD, Robert M. Freedom, MD, and William G. Williams, MD, Toronto, Ontario, Canada.
<table>
<thead>
<tr>
<th>Postoperative arrhythmias</th>
<th>EC</th>
<th>n</th>
<th>LT</th>
<th>n</th>
<th><em>P value</em></th>
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<tbody>
<tr>
<td>Temporary pacing</td>
<td>7 (12%)</td>
<td>59</td>
<td>15 (32%)</td>
<td>47</td>
<td>.01</td>
</tr>
<tr>
<td>Junctional</td>
<td>9 (15%)</td>
<td>60</td>
<td>21 (45%)</td>
<td>47</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Supraventricular Tachycardia*</td>
<td>5 (8%)</td>
<td>60</td>
<td>16 (33%)</td>
<td>47</td>
<td>&lt;.001</td>
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*Atrial Flutter, Junctional ectopic tachycardia, Atrial ectopic tachycardia, Supraventricular tachycardia*

<table>
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<tr>
<th>Midterm outcomes</th>
<th>EC</th>
<th>n</th>
<th>LT</th>
<th>n</th>
<th><em>P value</em></th>
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</thead>
<tbody>
<tr>
<td>Follow-up (y)</td>
<td>2.5±1.4</td>
<td>60</td>
<td>2.7±1.9</td>
<td>47</td>
<td>.50</td>
</tr>
<tr>
<td>Atrial arrhythmia*</td>
<td>7 (13%)</td>
<td>54</td>
<td>11 (25%)</td>
<td>43</td>
<td>.02</td>
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*Junctional rhythm, Sinus bradycardia, Supraventricular tachycardia, Pacemaker*
## Extracardiac Fontan at CMH

<table>
<thead>
<tr>
<th>Fontan Type</th>
<th># Pts</th>
<th>Op Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan Conversion</td>
<td>126</td>
<td>2 (1.6%)</td>
</tr>
<tr>
<td>Primary EC Fontan</td>
<td>113</td>
<td>1 (0.9%)</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>239</strong></td>
<td><strong>3 (1.3%)</strong></td>
</tr>
<tr>
<td>Year</td>
<td>Pts</td>
<td>Op Mortality</td>
</tr>
<tr>
<td>------------</td>
<td>-----</td>
<td>--------------</td>
</tr>
<tr>
<td>Backer 2009</td>
<td>239</td>
<td>3 (1.3%)</td>
</tr>
<tr>
<td>Harada et al 2008</td>
<td>72</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Nakano et al 2007</td>
<td>126</td>
<td>1 (0.8%)</td>
</tr>
<tr>
<td>Lee et al 2007</td>
<td>98</td>
<td>2 (2.0%)</td>
</tr>
<tr>
<td>Petrossian et al 2006</td>
<td>285</td>
<td>3 (1.1%)</td>
</tr>
<tr>
<td>TOTAL</td>
<td>820</td>
<td>9 (1.1%)</td>
</tr>
</tbody>
</table>
## LT Fontan Operative Mortality

<table>
<thead>
<tr>
<th>Study</th>
<th>Year</th>
<th># Pts</th>
<th>Op Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Robbers-Visser et al</td>
<td>2010</td>
<td>102</td>
<td>4 (3.9%)</td>
</tr>
<tr>
<td>Brown et al</td>
<td>2010</td>
<td>220</td>
<td>1 (0.5%)</td>
</tr>
<tr>
<td>Backer</td>
<td>2009</td>
<td>67</td>
<td>2 (3.0%)</td>
</tr>
<tr>
<td>Jacobs et al</td>
<td>2008</td>
<td>100</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Lee et al</td>
<td>2007</td>
<td>67</td>
<td>3 (4.5%)</td>
</tr>
<tr>
<td>Nakano et al</td>
<td>2004</td>
<td>88</td>
<td>1 (1.1%)</td>
</tr>
<tr>
<td>Stamm et al</td>
<td>2001</td>
<td>220</td>
<td>12 (5.5%)</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td></td>
<td>864</td>
<td>23 (2.7%)</td>
</tr>
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Anticoagulation and incidence of late cerebrovascular accidents following the Fontan procedure

Thromboembolic events are a well-reported complication following the Fontan procedure, but no previous studies have compared the incidence of thromboembolic events relative to the prophylactic anticoagulation strategy utilized. We examined the time-adjusted incidence of late thromboembolic events relative to chronic anticoagulation strategy. All patients who have undergone Fontan palliation and are followed at our institution were reviewed. All thromboembolic and major bleeding events were recorded and compared among different subgroups (anticoagulant medication utilized, Fontan variant, and the presence of a residual right-to-left shunt). The incidence of late cerebrovascular accidents (CVA) per patient-year was calculated for each subgroup. The records of 132 patients were analyzed (median follow-up, 7.6 years; 1066.5 total patient-years). There were no major bleeding complications. One patient receiving no anticoagulation therapy developed a symptomatic thrombus 6 months after Fontan. Three patients suffered late CVA (range, 3-7 years); 2 were receiving aspirin, and the other received no anticoagulation therapy. All 3 had lateral tunnel Fontan and a residual right-to-left shunt.

The overall incidence of late CVA was 2.3%, with an event rate of 0.28% per patient-year. Late CVA was not related to anticoagulation strategy or time from Fontan but was associated with a residual right-to-left shunt and lateral tunnel-type Fontan palliation (p < 0.001). Regardless of anticoagulation strategy utilized, symptomatic CVA is a rare long-term complication following the Fontan procedure.

C.B. Mahnke, G.J. Boyle, J.E. Janosky, R.D. Siewers, F.A. Pigula, Pittsburgh, PA.
Regarding strokes after single ventricle palliation, Dr. Jonas writes,

“thrombus forming on the pulmonary venous atrial side of the baffle or in the stump of the main pulmonary artery is the source of these emboli.”
Do we need fenestration when performing two-staged total cavopulmonary connection using an extracardiac conduit?

Between August 1999 and December 2007, 72 consecutive patients with single ventricle physiology underwent a modified Fontan procedure after a bidirectional Glenn shunt using an extracardiac polytetrafluoroethylene conduit without fenestration. Nitric oxide gas inhalation was commenced just after cardiopulmonary bypass together with intravenous phosphodiesterase III inhibitor administration. After oral intake was started, pulmonary vascular dilators such as beraprost, sildenafil, bosentan were given orally according to amount of chest drainage and patient’s condition. After discharge, oxygen therapy at home was continued for three months. No hospital death occurred after surgery. All patients were followed up by our institute and follow-up period was 44.2+/−26 (36–106.8) months. One late death occurred during this follow-up period after re-operation. Cardiac catheterization after the Fontan completion showed transpulmonary gradient of 5.9+/−2.4 mmHg, systemic output of 3.4+/−2.1 l/min m². Arterial oxygen saturation (SaO₂) at the latest outpatient visit was 94.4+/−3.8%.

According to our clinical experience with two-staged total cavopulmonary connection using an extracardiac conduit without fenestration, fenestration in the Fontan circuit is not necessary when performing the Fontan completion. Two-staged extracardiac total cavopulmonary connection without fenestration can be satisfactorily completed with the aid of pulmonary vasodilation therapy.

Impact of Fenestration on Event-free Survival

<table>
<thead>
<tr>
<th>Advantages of EC Fontan</th>
<th>EC</th>
<th>LT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Better Fluid Dynamics</td>
<td>++</td>
<td></td>
</tr>
<tr>
<td>Easier Surgical Technique (Teaching)</td>
<td>++</td>
<td></td>
</tr>
<tr>
<td>Avoids Aortic Cross-Clamping</td>
<td>++</td>
<td></td>
</tr>
<tr>
<td>Can be Performed without CPB (West Coast)</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Avoids Extensive RA Suture Lines</td>
<td>+++</td>
<td></td>
</tr>
<tr>
<td>Avoids Elevated RA Pressures (Dilation, Thrombus, Arrhythmias)</td>
<td>+++</td>
<td></td>
</tr>
<tr>
<td>Avoids Intracardiac Prosthetic Material</td>
<td>++</td>
<td></td>
</tr>
<tr>
<td>No Fenestration Needed (lower risk of CVA)</td>
<td>++</td>
<td></td>
</tr>
<tr>
<td>Preferred Technique for Fontan Conversion</td>
<td>+++</td>
<td></td>
</tr>
</tbody>
</table>
Extracardiac Fontan is Better

LT Fontan:
- Inferior hemodynamics
- Higher operative mortality (3%)
- Longer CPB and X-clamp times
- Increased postop arrhythmias
- Risk of CVA with fenestration

EC Fontan:
- Superior hemodynamics
- Very low operative mortality (1%)
- Fewer postoperative arrhythmias
- Rescue procedure of choice for Fontan conversion
Rebuttal
Slides
Extracardiac versus Intra-Atrial Lateral Tunnel Fontan: Extracardiac is Better – Yes, it is!

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Chicago, Illinois
The LT and EC Fontan were first performed in the 1980s.

Lateral tunnel  Extracardiac
Also in the 1980s, there were two new “power cars” on the market:

- DeLorean DMC-12 (American)
- Ferrari F40 (Italian)

The race began…
As time progressed and the cars make their laps...

- Long CPB and cross-clamp times
- Inferior hemodynamics
- Potential for RA dilation
- Increased atrial arrhythmias
- Potential for stroke
- Higher operative mortality (3%)

VS.

- No cross-clamp, shorter CPB
- Superb hemodynamics
- Procedure of choice for Fontan Conversions
- Operative mortality (1%)
The extracardiac conduit Fontan operation using minimal approach extracorporeal circulation: Early and midterm outcomes

OBJECTIVE: Our approach to the extracardiac conduit Fontan operation has evolved over time from full-pump, to partial-pump, to completely off-pump. This study is designed to report our overall experience with the extracardiac conduit Fontan operation and to evaluate the evolution in bypass technique on postoperative outcomes.

METHODS: From September 1992 to April 2005, 285 patients, median age 4.5 years (1.4-44 years), median weight 16 kg (9.4-94 kg), underwent a primary extracardiac conduit Fontan procedure. Early and late outcomes were analyzed for the entire cohort and for 2 patient groups depending on whether an oxygenator was used in the bypass circuit (166 patients; 58%) or not (119 patients; 42%).

RESULTS: Early failure (including death and takedown) occurred in 7 patients (2.5%). Prevalence of new early postoperative sinus node dysfunction necessitating a permanent pacemaker was 0.4%, and that of new tachyarrhythmias necessitating discharge home on a regimen of antiarrhythmia medications was 2.5%. Ten-year actuarial freedom from Fontan failure, new sinus node dysfunction necessitating a permanent pacemaker, and reoperation for conduit thrombosis or stenosis was 90%, 96%, and 98%, respectively. Fenestration rate was lower (P = .001) in the no-oxygenator group (8%) than in the oxygenator group (25%). Patients in the no-oxygenator group had lower intraoperative Fontan pressure (12.0 +/- 2.3 vs 13.5 +/- 2.4 mm Hg, P < .001), common atrial pressure (4.6 +/- 1.8 vs 5.3 +/- 1.8 mm Hg, P = .003), and transpulmonary gradient (7.5 +/- 2.1 vs 8.3 +/- 2.2 mm Hg, P = .013) than did the oxygenator group.

CONCLUSIONS: The extracardiac conduit Fontan operation coupled with minimal use of extracorporeal circulation is associated with favorable intraoperative hemodynamics, low fenestration rate, minimal risk of thrombosis or stenosis, and minimal early and late rhythm disturbance.

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Clinical outcome of 193 extracardiac Fontan patients

The first 15 years

OBJECTIVES: We sought to evaluate the mid-term outcome of hospital survivors with extracardiac Fontan circulation.

BACKGROUND: Few data exist about the mid-term and long-term results of the extracardiac Fontan operation.

METHODS: From November 1988 to November 2003, 221 patients underwent an extracardiac Fontan procedure as primary (9 patients) or secondary (212 patients) palliation, at a mean age of 72.2 months (range 13.1 to 131.3 months). A total of 165 of 193 early survivors underwent programmed noninvasive follow-up evaluations and at least one cardiac catheterization.

RESULTS: The overall survival, including operative deaths, was 85% at 15 years. Freedom from late failure among hospital survivors is 92% at 15 years. A total of 127 of 165 survivors (77%) were in New York Heart Association functional class I. The incidence of late major problems was 24% (42 major problems in 36 of 165 patients): 19 patients had arrhythmias (11%), 5 patients had obstruction of the extracardiac conduit (3%) and 6 of the left pulmonary artery (3.5%), and 5 patients experienced ventricular failure (3%), leading to heart transplantation in 3 patients. Protein-losing enteropathy was found in two patients (1%). The incidence of late reinterventions was 12.7% (21 of 165 patients, including 15 epicardial pacemaker implantations). Four patients died (2.3%), two after heart transplantation.

CONCLUSIONS: After 15 years of follow-up, the overall survival, the functional status, and the cardiopulmonary performance of survivors of the extracardiac Fontan procedure compare favorably with other series of patients who underwent the lateral tunnel approach. The incidence of late deaths, obstructions of the cavopulmonary pathway, reinterventions, and arrhythmias is lower than that reported late after other Fontan-type operations.

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Midterm surgical results of total cavopulmonary connection: Clinical advantages of the extracardiac method

OBJECTIVE: We evaluated the midterm surgical outcomes of intra-atrial lateral tunnel and extracardiac conduit total cavopulmonary connection to clarify the clinical superiority.

METHODS: Patients (n = 167) underwent total cavopulmonary connection (88 with lateral tunnel and 79 with extracardiac conduit) from November 1991 to March 1999. Survival, incidence of reoperation and late complications, exercise tolerance, hemodynamic variables, and plasma concentration of natriuretic peptide type A were compared. In the lateral tunnel group, time-related change in lateral tunnel size was investigated for its relationship to postoperative arrhythmias.

RESULTS: The 8-year survival was 93.2% in the lateral tunnel group and 94.9% in the extracardiac conduit group. Seven reoperations were performed in the lateral tunnel group but none in the extracardiac conduit group. Supraventricular arrhythmias developed in 14 patients (15.9%) in the lateral tunnel group and in 4 patients (5.1%) in the extracardiac conduit group (P = .003). Freedom from cardiac-related events was 72.5% in the lateral tunnel group and 89.8% in the extracardiac conduit group at 8 years (P = .0098). Hemodynamic variables and exercise tolerance were similar in both groups but plasma natriuretic peptide type A concentration, a parameter of atrial wall tension, was higher in the lateral tunnel group. In the lateral tunnel group, intra-atrial tunnel size increased by 19.4% during the 44.2-month interval and the percent increase in tunnel size was an independent predictor of supraventricular arrhythmias.

CONCLUSIONS: The midterm survival, hemodynamic variables, and exercise tolerance were similar and satisfactory in both lateral tunnel and extracardiac conduit groups; however, the incidence of cardiac-related events was significantly less frequent in the extracardiac conduit group. In the lateral tunnel group, careful observation is required to monitor the relationship of the dilating tendency of the intra-atrial lateral tunnel and the development of late complications.
Results of staged total cavopulmonary connection for functionally univentricular hearts; comparison of intra-atrial lateral tunnel and extracardiac conduit

Objectives: This study aims to compare the outcome of the two co-existing modifications of staged total cavopulmonary connection (TCPC) - the intra-atrial lateral tunnel (ILT) and the extracardiac conduit (ECC). Methods: We included 209 patients after staged TCPC (102 ILT and 107 ECC), operated on between 1988 and 2008. Medical and surgical records were reviewed for (1) patient demographics and cardiac anatomy; (2) pre-Fontan procedures; (3) pre-Fontan haemodynamics and cardiac functional status; (4) operative details; (5) postoperative hospital course; (6) follow-up information on arrhythmias and thrombo-embolic events; (7) post-Fontan interventions; and (8) clinical status at last follow-up until June 2008. Results: Median follow-up duration was 4.3 years (interquartile range 1.5-7.4 years). At 6-year follow-up, freedom from Fontan failure (i.e., mortality or re-operations for Fontan failure) was 83% for the ILT and 79% for the ECC groups (p=0.6); freedom from late re-operations (other than re-operations for Fontan failure) was 79% for the ILT and the ECC groups and freedom from arrhythmias was 83% for the ILT, and 92% for the ECC groups (p=0.022). Multivariable Cox regression analysis identified intensive care unit stay and cardiopulmonary bypass time as risk factors for Fontan failure, but they were not strong predictors. Right ventricular morphology was identified as a risk factor for arrhythmias. The occurrence of thrombo-embolic events was low with no difference between the ILT and the ECC groups, and irrespective of the postoperative use of anticoagulant or anti-platelet aggregation therapy. At most recent follow-up, sinus rhythm was present in 70% of patients; in 23% of the patients, ventricular function was found to be moderately or severely impaired at echocardiography. Conclusions: Outcome after staged ILT- and ECC-type Fontan operations is good, with comparable freedom from late re-operations and freedom from Fontan failure at 6-year follow-up. The incidence of arrhythmias was significantly lower in the ECC group. Right ventricular morphology was identified as a risk factor for arrhythmias.

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Freedom from arrhythmias was significantly higher in patients with an ECC, analyzed up to 6 years after Fontan completion.

The somatic growth of autologous vessels in venous pathway after extracardiac total cavopulmonary connection

OBJECTIVE: In a total cavopulmonary connection (TCPC) with an extracardiac conduit, the future development of stenosis in the venous pathway and distortion of the pulmonary artery according to the somatic growth of the patients is a major concern for surgeons and pediatricians. METHODS: Thirty patients who underwent extracardiac TCPC (EC-TCPC) between 1990 and 1998 and who had received at least two postoperative angiograms were enrolled in this study. To evaluate the postoperative change in the anastomosis, the cross-sectional area of the venous root at three different points was measured on the first and second angiograms after the EC-TCPC. Further, to evaluate the somatic growth of the autologous tissue, 12 patients who grew more than 10 cm in height after the completion of the EC-TCPC were selected and the length of three different parts, the diameter of the pulmonary artery, length of the artificial graft, and length between the branching point of the hepatic vein and artificial graft’s anastomotic site to the inferior vena cava were measured. RESULTS: The cross-sectional area at each point did not change during the follow-up, and the pressure gradient across the grafts has not been observed. The diameter of the pulmonary artery and length of the inferior vena cava above the hepatic vein insertion grew similarly during the follow-up. CONCLUSIONS: The mid-term clinical results after the completion of the EC-TCPC, including the somatic growth of the venous pathway, were satisfactory.

“Our results of no distortion of the PA in patients with somatic growth more than 10 cm in height may indicate a growth ability of the autologous venous tissue...our results showed that the PA and IVC were growing”

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Measurements of cross sectional area

- Cross-sectional area unchanged at follow-up
- No focal stenosis
- No incidence of thromboembolism

Measurements of length

- PA diameter and IVC length grew similarly during follow-up
- No deformities of PA

Optimal conduit size of the extracardiac Fontan based on energy loss and flow stagnation

BACKGROUND: In the extracardiac Fontan operation, larger conduits are used when considering the patients' growth rate. However, larger conduits may cause inefficient flow due to turbulence or stagnation, resulting in late problems such as thrombosis or stenosis. Our objective was to reveal the physiologic effects of respiration and exercise using numerical models, based on the energy loss and flow stagnation, and to determine the optimal conduit size. METHODS: For the Fontan operation, conduits from 14 to 22 mm were created based on angiographic data from 17 Fontan patients (mean age, 36.0 months; mean body surface area, 0.53 m\(^2\)). Respiratory-driven flow of the superior and inferior vena cava was determined at rest and during exercise on two levels (0.5 and 1.0 W/kg) by magnetic resonance imaging flow studies. Flow stagnation was defined as the volume of the region where flow velocity was less than 0.01 m/second at both the expiratory and inspiratory phases. BACKWARD FLOW at the expiratory phase was slighted large for smaller conduits (14 mm, 3.1 m\(^2\); 16 mm, 4.199 m\(^2\) during 1.0 W/kg exercise). Stagnation volume at the expiratory phase increased with an increase of conduit size (14 mm, 9.20% vs 22 mm, 33.9% conduit volume at rest).

CONCLUSIONS: Fontan circulation is a low-energy system even during exercise. Larger conduits were proven to have redundant spaces, thus 16 and 18 mm conduits were optimal.

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Has the LT Fontan gone down the same road as…

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EC Fontan wins!
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Extracardiac Fontan

- Superb hemodynamics
- No cross-clamp required
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- Avoids elevated RA pressures
- Very low operative mortality (1%)
- Fewer postoperative arrhythmias
- Lower incidence of CVA
- Procedure of choice for Fontan conversion