Fontan modification for subsequent non-surgical Fontan completion

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Abstract

Objective: Establishment of Fontan circulation in complex univentricular hearts often requires several surgical procedures. We developed a procedure which maintains the advantages of a staged approach, however, during the initial surgery additional preparatory measures are performed to allow subsequent non-surgical Fontan completion. Methods: The operation is a lateral baffle Fontan procedure. The baffle bears multiple perforations to allow the inferior vena cava blood to drain into the systemic atrium. Total cavopulmonary connection is performed as usual and the cardiac end of the superior vena cava is subtotally banded. Formally the operation establishes a bi-directional Glenn physiology. During subsequent catheter intervention the banding of the superior vena cava is dilated and the holes in the baffle are closed with appropriate devices. Results: From April 1994 to December 1995, 18 children having at least two risk factors for Fontan operation received the above described operation. Ages ranged from 3 months to 15 years. Ten patients had one or more previous operations. Bypass time ranged from 86 to 128 min and cross clamp time from 14 to 79 min. O2 saturation after discontinuation of cardiopulmonary bypass was 76% (70–81%). The postoperative recovery of all patients was rapid with early extubation (mean 6 h) and discharge to the ward the morning of the first postoperative day. One patient died. No fluid retention as pericardial, pleural or abdominal fluid effusions occurred. At discharge O2 saturation was 77% (75–82%). In thirteen children successful conversion to total cavopulmonary connection with interventional debanding of the superior vena cava and closure of the fenestrations was performed. After a hospital stay of only a couple of days the children were discharged with normal O2 saturation after Fontan completion. Conclusions: We suggest that this modification of the staged Fontan procedure reduces the need for surgical interventions by applying balloon angioplasty and occluder technology to this unique subset of patients. © 1998 Elsevier Science B.V. All rights reserved

Keywords: Fontan modification; Non-surgical Fontan completion

1. Introduction

The Fontan principle is currently the treatment of choice for functionally univentricular hearts. In patients with risk factors concerning the standard criteria a staged surgical approach reduced the mortality [3]. These staged procedures include neonatal pulmonary artery banding, aorta-pulmonary shunting, or a Norwood procedure, followed by a bi-directional Glenn procedure, which will be completed to Fontan circulation by an intra- or extracardiac conduit, which may be fenestrated or not. Also the so called Hemi–Fontan procedure and its modifications [6] need subsequent surgical conversion to complete Fontan circulation. All these modifications require several operations which appear to have still a certain degree of mortality or morbidity with complications such as effusions which account for increased length of stay in hospital [1]. In our institution a concept has been developed with the combination of a single surgical and a subsequent interventional procedure in order to complete the Fontan circulation.

2. Material and methods

2.1. Patients

From April 1994 to December 1995 a total of 62 opera-
tions for various forms of functionally univentricular hearts were performed. Of these, eighteen patients with two or more risk factors regarding the Choussat’s [2] criteria for the Fontan operation presented at our institution. These risk factors were increased pulmonary pressure above 16 mm Hg, distorted pulmonary arteries, atroventricular (AV) valve regurgitation, long standing arterio-pulmonary (AP) shunts or peripheral pulmonary artery stenosis and are given in detail in Table 1. Ten of them had one to four previous shunt operations.

Preoperative diagnoses in these patients were tricuspid atresia in eight, hypoplastic left heart syndrome in two, pulmonary atresia without ventricular septal defect (VSD) in six and an unbalanced AV canal in two. Preoperative hemodynamic data of the patients are given in Table 2. Patient’s ages ranged from 3 months to 15 years. After having obtained informed consent from the parents, a two-stage univentricular repair combining surgical preparation and interventional completion was performed.

2.2. Surgical technique

The details of the operative procedure have been reported in a previous publication [7]. In brief, during cardiopulmonary bypass and cardioplegic arrest a Goretex baffle with three to seven perforations, according to body weight and inferior vena cava (IVC) diameter was implanted to the lateral wall of the right atrium. The perforations were 5 mm in diameter. After closure of the atriotomy and removal of the aortic crossclamp, total cavopulmonary connection with the heart beating was performed. Before discontinuation of cardiopulmonary bypass subtotal banding of the cardiac end of the superior vena cava was performed. The band was a 3-mm wide Dacron strip. In order to maintain a minimal communication between the superior vena cava and the pulmonary artery, the band was fixed over a 16-gauge catheter and secured with a single 6-0 polypropylene suture. Pulmonary artery distortion was dealt with by extensive freeing of the vessels from surrounding scar tissue, resection with end-to-end anastomosis (n = 1) or patch plasty with Goretex (n = 2). Venous flow from the superior vena cava is directed to the pulmonary arteries (Fig. 1), while blood from the inferior vena cava enters the systemic atrium through the multiple baffle holes (Fig. 2). In terms of physiology a bidirectional Glenn circulation was established with this procedure.

2.3. Interventional completion

Fontan completion was achieved by a transcatheter approach. Technical details have been reported elsewhere [4]. During this procedure the banding was reopened by balloon angioplasty and subsequent insertion of a 12-mm Palmaz iliac stent was performed. The procedure was completed after the holes in the baffle were occluded with persistent duct arteriosus (PDA) occluders and the stent was dilated to a diameter of 14–16 mm.

The procedure was performed in the first two cases under general anesthesia. After realizing that sedation with ketamine and diazepam without intubation of the patient was possible for safe completion, all subsequent procedures in this series were performed under local anesthesia.

During and after completion heparin was given (100 IU/kg) and the antithrombin III blood level was kept at above 90%. Two days later warfarin was usually started and administered for 3–4 months, thereafter only aspirin was given.

3. Results

While performance of the first stage procedure required in the first case an aortic cross clamp time of 79 min, due to construction of the cavopulmonary anastomosis with the heart arrested, clamp time in the consecutive patients could be reduced because in all subsequent cases the cavopulmonary connection was performed with the heart beating.

The median cross clamp time in the whole series was 18 min (range 14–79 min). Bypass time ranged from 80 to 128 min (median time 85 min). Off pump saturation ranged from 70 to 81% with a median of 76%. Only normothermic bypass was used during the procedures and patients were extubated 3.5–16 h postoperatively (median 6 h).

One death occurred after surgery. This was a 4-month-old baby who died after successful conversion from Norwood

### Table 1
Preoperative risk factors in patients undergoing the modified Fontan procedure

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation less than 4 years</td>
<td>11</td>
</tr>
<tr>
<td>Age at operation less than 1 year</td>
<td>3</td>
</tr>
<tr>
<td>Pulmonary artery pressure (PAP) &gt; 16 mm Hg</td>
<td>9</td>
</tr>
<tr>
<td>Left ventricular end diastolic pressure (pLVED) &gt; 12 mm Hg</td>
<td>15</td>
</tr>
<tr>
<td>AV-valve regurgitation</td>
<td>3</td>
</tr>
<tr>
<td>AP-shunt</td>
<td>10</td>
</tr>
<tr>
<td>Distorted pulmonary arteries</td>
<td>3</td>
</tr>
<tr>
<td>Peripheral pulmonary arteries stenosis</td>
<td>2</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>PAP (mm Hg)</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>Postintervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean (Median)</td>
<td>17.5 (10)</td>
<td>8 (5–14)</td>
<td>7 (5–12)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SaO2</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>Postintervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean (Median)</td>
<td>80 (71–88)</td>
<td>77 (75–82)</td>
<td>94 (88–98)</td>
</tr>
</tbody>
</table>
operation to modified Fontan on the 14th postoperative day from respiratory problems after initially excellent recovery.

None of the patients required chest tube drainage for more than 3 days. No classical Fontan complications, like pericardial, pleural or abdominal effusions, renal or hepatic failure or protein losing enteropathy occurred. At discharge from surgery oxygen saturation was 77%. All patients received postoperatively 2–3 mg/kg daily aspirin.

Meanwhile 13 patients have been completed by means of dilatation of the superior vena cava and placement of PDA occluders. Time to completion was between 3 and 11 months postoperatively. No technical complications occurred. After Fontan completion the oxygen saturation reached levels above 90% (Table 2). All patients recovered rapidly and all underwent control angiography (Fig. 3) 3–4 months after completion which showed in all cases a well established Fontan circulation. Minimal residual baffle leaks were found in two patients. Further control during the follow-up period was performed with transthoracic echocardiography. In the intermediate term (more than 40 months) no complications occurred. Holter monitoring showed normal sinus rhythm in all cases without evidence of sinus node dysfunction.

4. Discussion

Univentricular repair with total cavopulmonary connection (Fontan principle) is an established treatment for patients with various forms of functionally univentricular anatomy. Although it remains a palliative procedure, its use in children with univentricular circulation is currently being broadened. More complex forms are now being repaired by this procedure, and the results have improved significantly, with an overall mortality under 10% [1,5,8].

In order to reduce the risk of postoperative complications and to decompress the right atrium, many authors recommend the use of a fenestrated baffle for the Fontan completion with subsequent transcatheter closure [1] of the baffle hole. The fenestrated Fontan carries still a risk of complications, which has been further reduced by the creation of a bi-directional Glenn circulation [2].

Trying to keep this advantage, but on the other hand eliminate also the need and risk for another surgery, we developed a concept of a two-stage procedure in which the second stage was performed non-surgically in the cath-lab [3,6].

The first step in this new concept consists of the creation of a bi-directional Glenn circulation and preparatory measures that allow subsequent transcatheter completion. These include the implantation of a multifenestrated baffle to the lateral wall of the right atrium and the creation of a total cavopulmonary connection with banding the cardiac end of the superior vena cava.
Venous blood flows from the superior vena cava to both pulmonary arteries, while the return from the inferior vena cava enters the systemic atrium through the multiperforated baffle (Fig. 2).

This procedure is performed safely with standard cardiopulmonary bypass with only a short cardioplegic arrest for baffle implantation.

The short period of cardioplegic arrest however does not seem to impair ventricular function or increase mortality or morbidity. Recovery occurred rapidly and generally the postoperative course was uneventful.

Bandung of the superior vena cava is performed as near as possible to the cavopulmonary anastomosis in order to avoid the cavo-atrial junction and creation of a sinus-node dysfunction. Insertion of the stent in this position can be performed safely with placement as high as possible in the superior vena cava. In our series consideration of these points resulted in normal sinus rhythm in all patients.

With this approach, a bi-directional Glenn physiology is achieved which unloads the heart by directing 30–50% of the systemic venous return to the pulmonary artery thus allowing reverse remodeling of the systemic ventricle before completion of the repair [3,4,7].

Transcatheter intervention for Fontan completion was performed several months postoperatively. As this procedure proved to be smooth all transcatheter interventions are now performed under light sedation in local anesthesia.

A dilatation of the stent up to 20 mm in diameter should give also later in life adequate venous return from the inferior vena cava to the pulmonary vasculature. Such a diameter is equal to that of an extracardiac conduit implantation.

Stent implantation to the cavo-atrial junction with subsequent closure of the multiperforated baffle offers the advantage of leaving a residual leak in the baffle converting the bi-directional Glenn physiology, after the surgical procedure, to a fenestrated Fontan circulation with the possibility of occluding this fenestration later. In our series this staged interventional procedure became necessary in two cases.

The uneventful clinical course during and after both stages suggests that morbidity can be further reduced by this modification while a reoperation is avoided. Because only a small part of the right atrium is exposed to high pressures, all advantages of the lateral baffle are maintained.

In conclusion this staged approach for creating a Fontan baffle (Fig. 2).

References


Appendix A. Conference discussion

Mr J. Meoro (Southampton, UK): This is an ingenious idea. What was the time interval between the first and second procedures?

Dr Sidiropoulos: It differs between 3 and 10 months depending on when the children are ready to be done.

Mr J. Stark (London, UK): Did you consider to use the adjustable atrial septal defect instead of your technique? Using adjustable ASD saves money, because coils or umbrellas are expensive. It also gives you greater versatility with regard to the size of the atrial shunt. Is it easy to reduce its size or completely occlude by tightening the snare through a small reopening of the lower part of the incision. This can be done under local ANESTHESIA.

Dr Sidiropoulos: We may discuss it with our cardiologist, maybe it is a valuable option, of course.

Mr Stark: I think it is up to you not the cardiologist what you do at surgery, isn’t it?

Dr Sidiropoulos: Well, not only up to us, because in a highly interactive program as ours, both partners are in charge.

Dr S. Conte (Copenhagen, Denmark): I have been very interested in your modified Fontan procedure since you reported the initial experience in the first seven patients treated with this method. My questions are: as I would expect some shunt through the subtotally banded superior vena cava, did you have any case with severe decreasing of arterial oxygen saturation? Considering the presence of the caval stent and the multiple perfusion, do you give antiocoagulant therapy to your patients after non-surgical Fontan completion? Finally, do you actually recommend this procedure to all patients with single ventricle or do you have some special indications?

Dr Sidiropoulos: May I begin from the latter, it is the easier one. We do not anticoagulate the patients. We give them aspirin, and we continue on aspirin also after interventional completion. We have never seen any thromboembolic events, although I may accept that something like this could occur. The first question about saturation, we have never faced this problem, although it may exist. In this case we probably proceed with interventional Fontan completion.

Dr H. Laks (Los Angeles, CA, USA): A very ingenious idea. One of my concerns is that with the multiple holes there is the danger that the holes will begin to heal in a patient who subsequently demonstrates that the PA pressure is excessively high, and you are then in a situation where your
holes are closing and yet you do not want to complete the Fontan. I am not sure whether one can, with dilating balloons or stents, reopen those holes, but have you been faced with that possibility?

**Dr Sidiropoulos:** We have never faced this problem. Actually our idea is to create such a multifenestration baffle, not only one hole, in case some of the holes could be closed. That is the one question. But we have never faced this problem. The second idea and the second point of view is that if our cardiologists see, through the completion, that it is not possible, or it is maybe for the circulation not better to close all the holes, then they can close with the occluder some of the holes and leave residual shunt to the systemic atrium.

**Dr Laks:** What were the size of your holes?

**Dr Sidiropoulos:** About 5 mm.

**Dr Laks:** Because at least experimentally, and also clinically, the experience has been that 4-mm holes can and will re-endothelialize and close up with time. So I think that that is something that you should consider. We had a similar idea in terms of a late completion of a Fontan with the unidirectional Fontan in which we have an obligatory Glenn shunt to the left PA and then have the inferior vena caval flow going to the right PA with the adjustable ASD left fairly wide open so that you have no inferior vena caval venous hypertension and that gives for a very smooth post-op course. And then in the cath lab, 6 or 12 weeks later, one can, either in stages or depending on the hemodynamics, completely close the adjustable ASD, thus either partially or completely completing the Fontan. So far with the follow up to about 23 months we have not seen fistulae develop in the left PA supplied by the unidirectional Glenn, which was the concern, the absence of the so-called hepatic factor. But with the matching of the blood flow so that you have a smaller left lung perfused with the SVC flow and so far, at least, no fistula formation in those completed Glens. But it is along the same idea. Thank you.

**Dr J. Amato (Chicago, IL, USA):** A very ingenious operation. Similar to Mr. Stark’s suggestion, rather than an adjustable band, we in the past 5 years, rather than perforating the patch, have been making a slit which easily allows a 4-mm probe to enter the baffle with two metallic clips on either side. And on one occasion, when the child had desaturated, we did go back to the cath lab and with the balloon, very simply finding the clips and expanding the hole again, allowing the child to again regain some saturation. So I might add another method of fenestration to you.