Protocols associated with no mortality in 100 consecutive Fontan procedures


The Heart Center for Children, St. Christopher’s Hospital for Children, Drexel University College of Medicine, Erie Avenue at Front Street, Philadelphia, PA 19134, USA

Received 10 September 2007; received in revised form 19 December 2007; accepted 21 December 2007; Available online 1 February 2008

Abstract

Objectives: Results of Fontan’s procedure have improved considerably, but perioperative mortality still occurs, attributed to ventricular dysfunction, stroke, arrhythmia, thromboembolism, and multi-organ dysfunction. Our protocols of operative and intensive care unit management address these potential issues, and have been associated with zero mortality, even with many high-risk candidates. Methods: From 1996 to 2006, all Fontan patients were managed as follows: operative strategy based on aortic and single atrial cannulation, cooling on full-flow bypass, and hypothermic circulatory arrest to create the Fontan pathway. No direct caval cannulation. Use of central venous lines was completely avoided. Fresh whole blood was used for pump prime and for volume restoration. Inotropic and vasodilator therapy was continued for at least 48 h. Aspirin was used exclusively as anti-thrombotic therapy. Postoperative pleural drainage was accomplished with small pigtail catheters. The usual Fontan pathway was by lateral atrial tunnel (84), with extra-cardiac conduit when dictated by anatomy (16). Results: One hundred Fontan operations were performed with no mortality. All patients were extubated by postoperative day 1. Hospital stay was 10±6 days. Complications were: bleeding (1), reintubation (1), emergent fenestration closure (1), pericardial effusion (4), and seizures (1). Risk factors included Fontan connection to one lung (3), diminutive pulmonary arteries (PAs) and unifocalized major aortopulmonary collateral arteries (MAPCAs) (1), discontinuous PAs (3), right ventricle dependent coronaries (3), neonatal pulmonary venous obstruction (3), Trisomy 21 (1), preoperative pacemaker dependence (2), and heterotaxy (10). No candidate was excluded. Conclusions: While many surgeons try to avoid bypass or aortic clamping when performing Fontan operations, the strategies we have employed facilitate safe accomplishment of Fontan’s operation in diverse anatomic groups with multiple risk factors, with avoidance of operative mortality in 100 consecutive cases.

Keywords: Congenital; Fontan; Single ventricle; Univentricular heart

1. Introduction

Over a period of nearly four decades, the operation originally proposed by Francis Fontan for separation of the pulmonary and systemic circulations in patients with tricuspid atresia has been modified extensively, and applied to virtually all forms of congenital heart disease characterized by a functionally univentricular heart. In the early years, patients were selected for modified Fontan procedures based upon rigid selection criteria, and the risk of perioperative mortality remained high. Recently, a more physiologic and rational approach to palliation early in infancy, and staging of reconstruction including removal of the ventricular volume load in the first year of life, have shifted the emphasis from one of patient selection to one of patient preparation. Thus, nearly all patients with non-septatable hearts can theoretically be managed in such a way that they will be good candidates for an eventual modified Fontan operation.

Experience with staged reconstructive surgery for hypoplastic left heart syndrome (HLHS) in the early 1990s [1] taught two very important lessons. The first was that the establishment of the ‘Fontan circulation’ was impacted favorably by dividing the operation into two stages [2]. Early accomplishment of a superior cavopulmonary anastomosis avoided the consequences of long-standing volume loading of the single ventricle, and allowed ventricular remodeling, with normalization of the mass-to-volume ratio and thus diastolic properties of the ventricle before completion of the Fontan procedure. The second lesson was that satisfactory function of the Fontan circulation depended on physiologic principals that were largely independent of the specific underlying anatomic diagnosis.
Despite a general trend of significant improvement in outcomes, most large centers managing patients with complex congenital heart disease report contemporary mortality rates associated with the Fontan operation in the range of 2–7% [3,4]. Perioperative mortality has been attributed to ventricular dysfunction, stroke, thromboembolism, and multi-organ dysfunction. Numerous investigators have sought to identify patient factors and procedural factors associated with increased risk of mortality [5,6]. Many have felt that ventricular hypertrophy, right ventricular morphology of the dominant or single ventricle, atrioventricular valve regurgitation, hypoplasia of the pulmonary arteries (PAs), dependence on a pacemaker prior to Fontan procedure, and heterotaxy syndrome are among the patient factors associated with increased mortality. Some investigators have identified long duration of cardiopulmonary bypass and/or aortic cross-clamping as incremental risk factors. As a consequence, some patients may be denied operations because of patient characteristics identified as risk factors. And, many surgical teams have gravitated to operative techniques that minimize or avoid the use of aortic cross-clamping, and in some instances avoid cardiopulmonary bypass entirely. While excellent results have been reported by some using these techniques, they clearly are not applicable to all patients who may undergo Fontan procedures.

We hypothesized that features essential to the avoidance of perioperative mortality would include strategies to minimize or mitigate the consequences of the inflammatory response to cardiopulmonary bypass, minimize the likelihood of thromboembolic events, maximize cardiac output in the immediate and early postoperative period, and avoid dire complications such as stroke and phrenic nerve injury. Notwithstanding the demonstration by others that in some selected instances the completion of the Fontan circuit may be accomplished without use of cardiopulmonary bypass support, we further hypothesized that the use of a consistent approach to anesthetic management, bypass support, and general conduct of the operation would be associated with the most consistent and best results. This report summarizes our experience with completion Fontan procedures over the past decade.

2. Materials and methods

This study was approved by the Institutional Review Board of Saint Christopher’s Hospital for Children. The records of the Section of Cardiothoracic Surgery were reviewed to find all completion Fontan operations performed from 1996 to 2006. De-identified data including patient factors, operative factors, and outcome were entered into a study database for analysis.

All operations were performed in a consistent fashion. Anesthetic technique was based primarily upon intravenous narcotic (Fentanyl 50 mcg/kg), together with muscle relaxant, and supplementary inhalation agents and small doses of intravenous benzodiazepines. A radial arterial line was placed, as were peripheral intravenous catheters. No central venous lines were used, in order to eliminate one potential source of thromboemboli. Bags of crushed ice were placed around the head, and a cooling blanket was positioned beneath the patient. Repeat sternotomy and limited dissection to expose the ascending aorta and the area corresponding to the appendage of the right-sided atrium were followed by systemic heparinization. The ascending aorta was cannulated for perfusion and a single cannula was placed in the right-sided atrium for venous return to the bypass machine. Dissection and cannulation of the superior and inferior vena cava were avoided completely. Thus no purse-string sutures were placed in the caval veins nor were tourniquets passed around them. This strategy was chosen to minimize the likelihood of phrenic nerve injury and to avoid potential areas of narrowing or flow disturbance at cannulation sites in the caval veins, as these could potentially serve as a nidus for thrombosis. The pump circuit was primed with crystalloid to which whole blood less than 48 h old was added to achieve a hematocrit on bypass of 28%. Flow was established at 150 ml/min per meter square of body surface area. The perfusate was cooled until nasopharyngeal and esophageal temperatures reached 17°C. A hypercarbic scheme of blood gas management was used during cooling, and ventilation was continued to minimize impedance to the flow of superior vena caval blood through the lungs. Adhesions between the right-sided atrium and the adjacent pericardium and pleura were carefully dissected during the cooling phase of cardiopulmonary bypass. When cooling was complete, the aorta was cross-clamped and crystalloid cardioplegia (30 ml/kg) was delivered to the aortic root. The circulation was temporarily discontinued. In instances where there was a particularly large ascending aortic segment, as in some patients who had undergone a previous Norwood Stage I palliation, the cross-clamp was placed upon the innominate artery and left common carotid artery only, or alternatively these vessels were occluded with suture tourniquets. Thus dissection around the aortic root, with potential risk to contiguous structures such as the left phrenic nerve, was avoided. Antegrade cardioplegia was administered in the usual fashion, facilitated by gentle compression of the distal ascending aorta with forceps.

During a brief period of hypothermic circulatory arrest, the total cavopulmonary connection was accomplished. The majority of patients had undergone a previous hemi-Fontan procedure [7], which not only addressed any narrowing or distortion of the central confluence of pulmonary arteries, restoring normal caliber from the point or origin of one upper lobe branch to the other, but sets the stage for a technically straightforward completion of the Fontan by construction of a lateral atrial tunnel. In these cases, the completion Fontan was accomplished using a curved gusset of PTFE, which had been cut from a segment of 10 mm diameter tube graft. In each case, three separate fenestrations were created in the PTFE gusset using a 2.5 mm or 2.7 mm aortic punch. In a small number of patients an extra-cardiac conduit type of completion Fontan (following a previous bidirectional Glenn anastomosis) was performed using a PTFE tube graft. The circumferential conduit was anastomosed to both the divided inferior vena cava and the underside of the pulmonary arteries during a single period of circulatory arrest. After construction of the cavopulmonary connection, bypass was resumed, the cross-clamp released, and the patient rewarmed at full flow, with resumption of ventilation. Two 20 gauge long lines were exteriorized through the chest wall.
and connected respectively to a transducer to monitor atrial pressure and to infusion pumps to deliver inotropic drugs and vasodilators. Upon termination of bypass, the venous cannula was withdrawn from the right-sided atrium and the 20 gauge lines were inserted at the same site through the purse-string suture. Support at the time of separation from bypass typically consisted of dopamine 2–3 mcg/kg min, and dobutamine 2–3 mcg/kg min. In the earlier cases, sodium nitroprusside was used selectively. More recently, milrinone 0.25–0.75 mcg/kg min has been used. After decannulation of the aorta, protamine was administered to reverse heparin. Additional fresh whole blood (less than 48 h old) was administered as necessary to restore hemostasis and augment intravascular volume. Platelet concentrates were not used, nor were aprotinin, epsilon aminocaproic acid, or other pharmacologic agents to promote hemostasis.

The anterior mediastinum was drained routinely. The pleural spaces were drained only if widely opened. From 2002 to 2006, conventional ultrafiltration was performed during the warming phase of cardiopulmonary bypass. Modified ultrafiltration was not used at all.

Exubation of the trachea was achieved as early as possible. In all cases, intravenous inotropic support was continued for 48–72 h, even in the face of excellent perfusion and stable hemodynamics. All patients received aspirin 81 mg daily beginning the evening following surgery. No patient received heparin or warfarin in the postoperative period. Furosemide, spironolactone, and angiotensin converting enzyme inhibitors were given to all patients beginning on the first or second day after operation. Pleural effusions, which were common, were drained using small polyvinyl catheters inserted using Seldinger technique. The ability to seal these catheters with a stopcock and aspirate them periodically facilitated early mobilization of the patients. In cases where pleural drainage persisted with large volumes over many days, fresh frozen plasma was administered to avoid depletion of Protein S, Protein C, and other naturally occurring circulating anticoagulants.

Data are presented as ranges and medians or means, where appropriate.

3. Results

One hundred patients who had undergone previous superior cavopulmonary anastomosis had a completion Fontan procedure between January 1, 1996 and December 2006. In 84 patients who had undergone a prior hemi-Fontan procedure, the completion Fontan was accomplished by creation of a lateral atrial tunnel. In 16 patients who had undergone a prior bidirectional Glenn anastomosis, the completion Fontan was accomplished with use of an extracardiac conduit. The latter choice was based upon specific anatomic considerations, with features such as anomalous pulmonary venous connections or apical caval juxtaposition favoring the Glenn/conduit approach. During this time period, no patient underwent a single stage Fontan procedure, without prior cavopulmonary connection. Anatomic diagnoses are shown in Table 1. Hypoplastic left heart syndrome was the most prevalent diagnosis (37%). The remainder of the cases represent the full spectrum of anomalies characterized by a functionally univentricular heart. Some anomalies were quite rare, such as truncus arteriosus communis with mitral atresia and hypoplastic left ventricle. As in most series of Fontan procedures, a number of patients could alternatively have been considered for biventricular repairs, but were subjected to staged Fontan procedures after consideration of the risks and benefits of both approaches. An example is the group of seven patients with congenitally corrected transposition of the great arteries, ventricular septal defect, and pulmonary atresia [8,9].

Patient age at the time of the completion Fontan procedure ranged from 1.5 years to 15 years. The median age of 26 months is reflective of our general preference to complete the Fontan circuit at about 2 years of age in most patients, and slightly later (in relation to patient size) in those for whom an extra-cardiac conduit technique is chosen. Ninety-eight of the 100 patients were less than 4 years old. Many of the patients had features that would be considered significant risk factors for mortality, or even contraindications to the Fontan procedure in the opinion of some surgeons or cardiologists. These risk factors are shown in Table 2. The three patients with acquired atresia of the left pulmonary artery underwent creation of the Fontan connection to one lung [10,11]. Three additional patients had a history of acquired discontinuity of the right and left pulmonary

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>37</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>11</td>
</tr>
<tr>
<td>Heterotaxy syndrome</td>
<td>10</td>
</tr>
<tr>
<td>Single left ventricle</td>
<td>9</td>
</tr>
<tr>
<td>Complex double outlet right ventricle</td>
<td>8</td>
</tr>
<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
<td>8</td>
</tr>
<tr>
<td>Congenitally corrected TGA with pulmonary atresia</td>
<td>7</td>
</tr>
<tr>
<td>Complex TGA with hypoplastic LV or RV</td>
<td>3</td>
</tr>
<tr>
<td>Malaligned common atrioventricular canal defect</td>
<td>3</td>
</tr>
<tr>
<td>Single right ventricle</td>
<td>2</td>
</tr>
<tr>
<td>Truncus arteriosus communis with mitral atresia</td>
<td>1</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 1

### Significant risk factors among the 100 patients who underwent completion Fontan operations from 1996 to 2006

<table>
<thead>
<tr>
<th>Patient factor</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan connection to one lung</td>
<td>3</td>
</tr>
<tr>
<td>History of discontinuous pulmonary arteries</td>
<td>3</td>
</tr>
<tr>
<td>Pre-Fontan pacemaker dependence</td>
<td>2</td>
</tr>
<tr>
<td>Right ventricular dependent coronary flow</td>
<td>3</td>
</tr>
<tr>
<td>History of neonatal pulmonary vein obstruction</td>
<td>3*</td>
</tr>
<tr>
<td>Pulmonary artery hypoplasia and major aortopulmonary collaterals</td>
<td>1</td>
</tr>
<tr>
<td>Trisomy 21</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary arterio-venous malformations</td>
<td>4</td>
</tr>
<tr>
<td>Heterotaxy syndrome</td>
<td>10</td>
</tr>
</tbody>
</table>

*One patient with hypoplastic left heart syndrome and intact atrial septum, one patient with truncus arteriosus communis with mitral atresia and intact atrial septum, one patient with heterotaxy syndrome and obstructed total anomalous pulmonary venous connection.
arteries and had undergone surgical or interventional procedures to re-establish central continuity of the branch pulmonary arteries prior to the Fontan completion.

Duration of cardiopulmonary bypass was 35–65 min (mean 43 min). Duration of deep hypothermic circulatory arrest was 15–39 min (mean 22 min). No patient was subjected to more than one period of circulatory arrest during the completion Fontan procedure. All patients were extubated on the day of surgery or the day following the operation. Hospital length of stay ranged from 7 days to 45 days (median 11 days). There was no hospital mortality. No patient required takedown of the Fontan connection, and no patient required mechanical circulatory support following the operation. One patient underwent operative closure of a fenestration between an extra-cardiac conduit and the right atrium less than 24 h after the Fontan procedure. One patient underwent re-exploration for bleeding. One patient was reintubated for stridor, and was extubated again following bronchoscopy. Four patients required pericardiotenectomy, and one of them underwent surgical creation of a pleuro-pericardial window. One patient with a history of epilepsy experienced postoperative seizures. One patient required cardioversion for early postoperative atrial flutter. There were no instances of phrenic nerve injury associated with the Fontan procedure. There were no early post-Fontan thromboembolic complications or strokes.

The duration of follow-up ranges from 14 months to 139 months.

During the period of follow-up there has been one death that occurred suddenly in a 6-year-old patient with a previously uncomplicated course after completion Fontan procedure at 2 years of age for transposition of the great arteries with VSD, supero-inferior ventricles with hypoplastic right ventricle. The cause of death is unknown. The patient’s primary cardiologist suspected arrhythmia as the etiology. No autopsy was performed. Three patients have developed protein-losing enteropathy at intervals of 18 months, 35 months, and 77 months after the completion Fontan procedure. All underwent diagnostic cardiac catheterization, with reenfracturation in one, reenfracturation and stent deployment in the pulmonary artery in one, and only medical therapy and nutritional modification in one. All have had favorable responses. In all, five patients have undergone catheterization procedures to deploy stents and/or redilate stents in the pulmonary arteries. One patient with HLHS has undergone surgical tricuspid valvuloplasty. Two patients have undergone transcatheter radiofrequency ablation procedures for atrial arrhythmias. One patient with the diagnosis of pulmonary atresia with intact ventricular septum had truly diminutive pulmonary arteries that supplied less than two thirds of the pulmonary segments. Much of the pulmonary blood flow was accounted for by aortopulmonary collateral arteries. He underwent a unifocalization procedure at the time of his superior cavopulmonary anastomosis. He underwent the completion Fontan procedure at age 2.5. He experienced hemoptysis at age 10. Aspirin therapy was briefly stopped at that time. There have been no other hemorrhagic or other aspirin related complications. Only one patient received warfarin, until a successful ablation procedure for sustained atrial flutter.

4. Discussion

The contemporary approach to patients with functionally univentricular circulation is based upon planning that begins in the neonatal period with physiologically rational palliation, and in most cases then involves early reduction of the volume work of the single ventricle by means of a superior cavopulmonary anastomosis (usually in the first year of life). As such, the completion of the Fontan circulation is the final phase of staged separation of the systemic and pulmonary circulations. The completion of the Fontan procedure as part of a planned, staged approach, rather than out of necessity based upon progressive cyanosis, or deteriorating ventricular function in the face of long-standing volume overload, is a concept that has led to decreased operative mortality at most centers [12]. Nonetheless, operative mortality has not been eliminated completely, and numerous investigators have undertaken analyses to identify incremental risk factors for perioperative mortality. Phenomena or events associated with hospital mortality appear to include ventricular hypertrophy [13], stroke or thromboembolism [14,15], and ventricular dysfunction with a low cardiac output state associated with effusive complications and leading to multi-organ dysfunction. Individual reports have identified some specific anatomic groups [16—18] (including those with right ventricular morphology of the systemic ventricle, those with heterotaxy syndrome, and those with pulmonary atresia and intact ventricular septum with right ventricular dependence of coronary blood flow), unfavorable morphology or physiology of the pulmonary vascular bed (including history of initial presentation with obstructed pulmonary venous return), history of pacemaker requirement which pre-dates the Fontan procedure [3], and Trisomy 21 as risk factors for mortality.

Experience led us to believe that an operative strategy that minimized the likelihood of ventricular dysfunction, low cardiac output, stroke, and thromboembolism would be associated with a very low rate of operative mortality, even for patients with one or multiple risk factors. In the series reported here, ventricular dysfunction and low cardiac output were largely avoided as a result of the following strategies: early reduction of the ventricular volume load by staging Fontan’s procedure with nearly all associated procedures being completed at the time of the superior cavopulmonary anastomosis, minimization of the duration of cardiopulmonary bypass by completing the Fontan pathway during a brief period of hypothermic circulatory arrest, an operative technical approach and blood transfusion strategy that minimizes postoperative bleeding and the effects of component administration on the pulmonary vasculature, and the avoidance of phrenic nerve injury by avoiding circumferential dissection around the caval veins. Despite generally excellent myocardial function at the end of surgery, all patients received inotropic and vasodilator therapy for a minimum of 48 h in order to optimize cardiac output and minimize the likelihood of low-flow related thromboembolism. While some authors have suggested that higher values of pulmonary vascular resistance predict a more difficult postoperative recovery, we have no data to corroborate this hypothesis. Even our patients, who underwent completion of the total cavopulmonary connection to
the right pulmonary artery only, had satisfactory post-
operATIVE courses, as we have described previously [10, 11].
Strategies to prevent thromboembolism include avoidance of
direct caval cannulation, use of trans-thoracic intracardiac
catheters rather than percutaneous central venous lines, and
administration of aspirin to all patients beginning the day of
surgery [19].

Some of the strategies we employed are controversial.
There seems to be a movement toward avoidance of aortic
cross-clamping and even complete avoidance of the use of
cardiopulmonary bypass when feasible in the performance of
Fontan procedures. Certainly there are cases where these
strategies are applicable, and excellent results have been
reported in selected cases [20, 21]. But even enthusiastic
advocates of a ‘no-bypass’ approach encounter cases that
cannot be accomplished without mechanical circulatory
support. Also, the no-bypass techniques require circumfer-
tential dissection and direct cannulation of the caval veins,
putting the right phrenic nerve at risk. We believe that hemi-
diaphragm paresis is a serious unfavorable event in the life of
a patient with Fontan circulation. The important conse-
quences of this complication have been studied and reported
by others [22].

The use of deep hypothermic circulatory arrest (DHCA) is
also considered controversial by some. In our protocol, the
use of this technique results in limitation of the exposure to
bypass to a period of cooling (approximately 15—20 min) and
a period of re-warming (approximately 25 min). And the
duration of circulatory arrest is quite short, certainly within
the range that has been shown to be associated with no
greater risk of adverse neuro-developmental outcome than is
associated with the use of continuous cardiopulmonary bypass.
The 1% incidence of seizures and 0% incidence of
stroke in our experience compare favorably with all
published series. While we have not undertaken a compre-
hensive program of neuro-developmental testing of patients
in this cohort, we did undertake a survey of ‘Health Related
Quality of Life,’ using a standardized pediatric health
assessment tool. This study, performed in 2001, surveyed
all patients operated up to that time, approximately two-
thirds of the total cohort of 100 patients [23]. While the
quality of life assessment of Fontan patients was not on a par
with that of healthy children, the majority of the school age
Fontan patients attended normal schools and were in age
appropriate grades.

Others have published reports of large series of comple-
tion Fontan operations using deep hypothermia with
circulatory arrest with excellent results. In a series of 332
Fontan procedures reported by Gaynor et al. in 2002 [24],
DHCA was used in 319 patients. Mosca et al. [25] reported a
series of 100 consecutive Fontan procedures for HLHS. The
earlier half of the group was supported with continuous
bypass with moderate hypothermia and bicaval cannulation.
The latter half of the group had single atrial cannulation and
DHCA. Mortality was significantly lower in the DHCA group.

There is at the present time no consensus as to the best
strategy to minimize thromboembolic complications early
and late after the Fontan procedure. At the same time, there
is clear evidence that such events can and do occur in some
cases even in the face of heparin or warfarin therapy. Our
surgical strategy, combined with the use of low-dose aspirin
has been associated with very good outcomes, as has been
reported by us previously [19].

One could speculate that the absence of operative
mortality in our 10-year series is related to factors other than
the management strategies discussed here. There is the
possibility, for example, that patients destined for mortality
were ‘selected out’ at the time of superior cavopulmonary
anastomosis. This, however, is not the case. The incidence of
operative mortality associated with the hemi-Fontan proce-
dure and bidirectional Glenn anastomosis during the decade
of the study was 2 of 109 (1.9%). And no patient was denied a
completion Fontan procedure because of the perception of
unacceptable risk. Alternatively, one could speculate that
the patient group was in fact a group of consistently low risk
candidates. This, of course, is not the case as demonstrated
by the diverse anatomic diagnoses (Table 1) and the multiple
risk factors (Table 2). Some patients, in fact, might not have
been considered for completion Fontan procedures at all
centers. This would certainly encompass the patients who
underwent Fontan connection to one lung, a patient with
Trisomy 21, and a patient whose pulmonary vascular bed was
made up in large part by unfocalized aortopulmonary
collateral vessels.

This study is limited by the retrospective nature of the
investigation. Certainly, prospective data collection could
have led to an analysis that would shed more light on factors
associated with perioperative complications and morbidity.
But the fact that the management strategies were largely
constant over a period of 10 years, and were associated with
survival of all patients, is the essential message. Most centers
have achieved low mortality with modified Fontan proce-
dures in the recent past compared to those in earlier eras. We
do not presume to suggest that the methods we have chosen
are the best with respect to achieving low operative
mortality. We suggest only that in a program of moderate
day size and volume, the consistent approach to Fontan
operations presented here enabled us to achieve consistent
survival despite a challenging spectrum of anatomic
diagnoses and patient-related risk factors.

References

[1] Norwood Jr WI, Jacobs ML, Murphy JD. Fontan procedure for hypoplastic
1029—30.
1993;166(5):548—51.
mid JP, Burnett J, Jonas RA, Castañeda AR, Wernovsky G. Fontan opera-
tion in five hundred consecutive patients: factors influencing early and
Williams WG. Interventions associated with minimal Fontan mortality.
factors influencing early and late mortality after total cavopulmonary
Danielson GK, Puga BF, Offord KP. Influence of ventricular morphology on
Norwood Jr WI. Early reduction of the volume work of the single ventricle:
461—2.
Appendix A. Conference discussion

Dr R. Lange (Munich, Germany): I congratulate you on your excellent results, although personally I'm a little bit concerned about why you should make a really simple operation so complicated. The extra-cardiac TCPC in our institution takes 30 to maximally 40 min of normothermic beating heart cardiopulmonary bypass and then it's done. Even if you have good results, we have the same good results, and I cannot quite follow why you would need deep hypothermic arrest in a cardiologically arrested heart.

Dr Jacobs: Well, I understand your concern, and I compliment you on the superb results that you presented.

Your colleague, in the paper that preceded yours, skipped very quickly through a slide that indicated that at the time of the superior cavopulmonary anastomosis, three patients had sustained phrenic nerve pareses, and I don't know if any additional patients did at the time of the completion Fontan. You see, I agree with you. I think that these are technically simple operations. I think they can be performed well in a lot of ways. I don't think that the problems from which Fontan patients succumb are related to the plumbing as much as they are to other factors that impact on the circulation, such as respiratory mechanics, phrenic paresis, and thromboembolic complications. So the avoidance of tourniquets around the cavo, cava cannulation, repeat dissection in those areas are just another approach to the same end product that I think we avoid some of the morbidities.

Dr G. Stellin (Padova, Italy): In our institution we have a very similar experience with no deaths and no complications for a long period of time. However, we have recently experienced two thromboembolic events in two different patients, in one of which after Kawashima operation, where the hepatic veins only are connected to the pulmonary circulation, both the patient survived and I would like to know if you treat differently patients after Kawashima operation and what do you do in order to avoid the thromboembolic events?

Dr Jacobs: Yes, I agree with you. Several years ago, the first 72 of this 100 patient series were the subject of a publication about aspirin anticoagulation and the potential reduction of thromboembolic events. I don't believe that aspirin is a panacea. What I believe is that thromboembolic events can occur in patients who are heparinized, can occur in patients who are receiving warfarin, and so I'm not sure that there is a good rationale to subject the patients to those additional risks.

The question about the Kawashima is very important. There are at least two reports, one a multicase series from the Mayo Clinic, and a second report saying that conduits conveying only hepatic vein effluent put the patient at greater risk for thromboembolism than do cavopulmonary extra-cardiac conduits, and I think it's probably related to the relative size of the conduit and the small amount of flow. I'm not aware of other factors. So one might take a more aggressive approach early postoperatively to a conduit repair after a Kawashima.

Dr Stellin: But what is your anticoagulation policy?

Dr Jacobs: We have used aspirin on all these patients beginning the night of surgery.

Dr Stellin: No heparin?

Dr Jacobs: We have used no heparin and we have used no warfarin, except later on, if a patient has chronic atrial flutter, they will be put on warfarin. I can't tell you whether the use of fresh whole blood and the avoidance of Amicar, the avoidance of aprotinin, the avoidance of platelet concentrates has anything to do with the low incidence of thromboembolism because it's an untestable hypothesis, but I think it's a system that works well for us.

Dr B. Muraszewski (Warsaw, Poland): I have one question and one comment.

The question is, I have noticed you have a series of about 16 extra-cardiac Fontans in your series. Now, did you do them because you changed your policy, or were there special indications to move from your intracardiac tunnel to extra-cardiac tunnel? That's the question.

The comment is, probably due to our relations and your persuasive personality, recently in the last years we have changed in our unit from continuous flow in cardiopulmonary bypass to deep hypothermic circulatory arrest for these patients. It takes us an average of 30 min of deep hypothermic circulatory arrest. How long does it take you? And I agree totally that this is a different operation, because besides this 30 min of circulatory arrest, you save a lot of time and it's much less traumatic surgery, especially as far as the vena cava and the PAs.

Dr Jacobs: I'm happy to hear that you gave consideration to this technique. In addition to our smaller series, in the literature there is a very large series published by Gaynor and Spray from Children's Hospital of Philadelphia of 313 of 330 Fontans performed with hypothermic circulatory arrest, single cannula, and most people are familiar with Dr Mosca's paper where the LHLS Fontan outcomes at Michigan improved dramatically when they went from bicaval cannulation with continuous bypass to single cannulation and circulatory arrest.
To answer your question about the extra-cardiac conduit, my preference is still to do a lateral tunnel, and I think the hemi-Fontan, as Bill Norwood first described it, ideally sets the patient up for a lateral atrial tunnel. We do the extra-cardiac conduit selectively, and that would be cases either of heterotaxy with anomalous pulmonary venous connection; sometimes there’s pulmonary venous connection just at the junction of the superior vena cava and the atrium; sometimes there’s ipsilateral pulmonary venous connection and a conduit might lie on that; or frequently, in heterotaxy or in congenitally corrected transposition with pulmonary atresia, there is what has been given the very complicated name ‘apicocaval juxtaposition’ — the mass of the heart overlies the entrance of the IVC into the right atrium, and I think that’s an awkward arrangement for an intracardiac tunnel and we do a conduit.

Dr Maruszewski: How long was your circulatory arrest?

Dr Jacobs: The average was 22 min. I think that the slide said the range was 15–35 min.