Management of the univentricular connection: are we improving?¹

Andrew D. Cochrane a, Christian P. Brizard a, Daniel J. Penny a, Sune Johansson b, Juan V. Comas c, Torsten Malm b, Tom R. Karl a, *

* Cardiac Surgical Unit, Royal Children's Hospital, Flemington Road, Melbourne, 3052, Australia
b Paediatric Cardiac Surgical Unit, University Hospital, Lund, Sweden
c Cardiac Surgical Unit, Hospital Clinic i Provincial, Barcelona, Spain

Received 2 October 1996; received in revised form 17 March 1997; accepted 25 March 1997

Abstract

Objective: To assess the impact of the bidirectional cavopulmonary shunt, total cavopulmonary connection, and baffle fenestration on outcome of the Fontan operation in our unit. Method: We reviewed 123 bidirectional cavopulmonary shunts and 264 Fontan operations performed from 1980 to 1995. Analysis of pulmonary artery size (right and left main and lower lobe branches) before and after bidirectional cavopulmonary shunt was performed. Outcome of the Fontan operation was analysed in various time periods to assess the effect of prior bidirectional cavopulmonary shunt, use of the lateral tunnel, and fenestration.

Results: Operative risk for the bidirectional cavopulmonary shunt was 4% (CI = 2-10%) with a survival of 89% (CI = 83-95%) at 36 months. Probability of conversion to Fontan operation at 36 months was 49% (CI = 38-61%). Pulmonary artery size (Nakata and lower lobe indices) fell after bidirectional cavopulmonary shunt (P = 0.0006). Fontan risk dropped from 8.5% (1980-1987) to 1.8% (1988-1995) (P=0.02), coinciding with the use of the bidirectional cavopulmonary shunt. There was no further risk reduction after introduction of the lateral tunnel and baffle fenestration, although these comparisons are limited by relatively small numbers. Duration of hospital stay related to pleural effusions was lowest for patients with a fenestrated lateral tunnel operation (P < 0.05).

Conclusion: The bidirectional cavopulmonary shunt is a suboptimal stimulus for pulmonary artery enlargement, but may reduce the risk of Fontan operation in selected children. Fenestrated lateral tunnel operations have reduced the duration of postoperative pleural effusions. © 1997 Elsevier Science B.V.

Keywords: Fontan; Cavopulmonary shunt; TCPC; Univentricular heart

1. Introduction

Over the last decade there have been a number of changes in the management of infants and children undergoing Fontan repair. These changes include the introduction of the total cavopulmonary connection (TCPC) in preference to the atropulmonary connection [4,14], the introduction of various techniques of fenestration [1,9], and the use of staging with a bidirectional cavopulmonary shunt (BCPS) [6].

In our own practise, these changes in surgical management have occurred over a period of 6 years, with the regular incorporation of all of the techniques over the last 3 years. The TCPC was introduced gradually in 1989, and since June 1992 nearly all Fontan procedures have been performed using the TCPC technique. The BCPS was introduced into our practise in late 1988, and the first conversions of a BCPS to a Fontan circulation began in late 1991. Fenestration of the atrial baffle has been a regular part of our practise since June, 1993 (Fig. 1).

The aim of the current study was to assess our results for the Fontan operation over the period from 1980 to
1995, and the pattern of mortality and complications over that time, by comparison of the periods 1980–1987 and 1988–1995. The latter period coincides with our introduction of the BCPS. Since the use of fenestration has been incorporated in our practice more recently, we then divided the second period into two 3 year eras for comparison, 1989–1991 and 1992–1995. Secondly, we reviewed the results of the BCPS and the effect of this procedure on pulmonary artery growth and future Fontan candidacy.

2. Methods and patient population

All patients who underwent the Fontan procedure between 1980 and 1995 and all patients who underwent BCPS from 1988 to 1995 were identified in our surgical database.

2.1. Fontan procedure

In the period from 1980 to 1995, 264 patients underwent a Fontan procedure for various anatomic problems for which the possibility of biventricular repair was excluded. A retrospective follow-up of the hospital and department records was performed, to document the method of repair, use of fenestration, previous BCPS, perioperative mortality, incidence of takedown of the surgical repair, and length of hospital stay (which correlated closely with duration of pleural drainage). Follow-up was also obtained through hospital records or treating cardiologists to document the occurrence of late death or need for cardiac transplantation. This study was divided into two equal time periods to analyse 1980–1987 against 1988–1995, since the introduction of the BCPS commenced in 1988. There were 94 and 170 patients in the two respective time periods. The median age and weight in the two periods were 69.6 months/17 kg and 45 months/18.5 kg (P = 0.11 and 0.13). To assess the additional effects of lateral tunnel and fenestration, we compared the two sequential periods 1989–1991 and 1992–1995, during which 138 patients underwent operation, 66 patients and 72 patients in each respective 3 year period.

2.2. Bidirectional cavopulmonary shunt procedure

Over the period 1988 through 1995, 123 infants and children had a BCPS procedure. The BCPS was the unique source of pulmonary blood flow in all except four patients who had some forward flow through the pulmonary artery maintained. The median age at operation was 17 months (range 3.8 to 197 months). The hospital records of all patients were reviewed to document the primary diagnosis, ventricular morphology, previous source(s) of pulmonary blood flow, presence of pulmonary stenoses or distortion, the presence of sub-aortic stenosis, need for concurrent procedure and the hospital mortality.

One hundred and seven patients of this group (87% of the total) had undergone a total of 148 previous surgical procedures, up to a maximum of five procedures in one patient. The previous primary procedures included—modified Blalock shunt (51 patients), pulmonary artery banding (16 patients), combined coarctation repair and banding (14 patients), Norwood procedure (13 patients) and arterial switch (5 patients).

At the time of BCPS, concurrent procedures included pulmonary artery patching (50 patients), relief of sub-aortic obstruction (19 patients—Damus-Kaye-Stansel anastomosis in 16, VSD enlargement in 3), AV valve repair (5 patients), AV valve replacement (1 patient) and TAPVD repair (4 patients).

In a sub-group of 23 BCPS patients who had undergone follow-up cardiac catheter procedures with pulmonary angiography in our institution prior to Fontan conversion, two or more angiographic studies were available to us for comparison. We compared the pre and post-operative pulmonary artery size before and 15.5 (SD = 6) months after BCPS. The median age at the time of BCPS in this group was 21.9 months (range 3.8 to 193.2 months) and the median weight was 10.1 kg (range 3.9 to 28.9 kg). The dimensions of the right and left main pulmonary arteries were normalised to body surface area to derive the pulmonary artery index using the method of Nakata [12]. The same calculations were performed for right and left lower lobe branches.

2.3. Criteria for Fontan repair

The primary clinical criterion is a congenital cardiac defect which is not amenable to biventricular repair, or has a prohibitive operative risk from attempted biventricular repair. After initial BCPS we like to see evi-
dence of falling saturation or exercise intolerance to warrant progression to a Fontan procedure.

Cardiac catheterization and angiography was performed on all candidates for repair to assess the pulmonary artery pressure, left atrial pressure and left ventricular end-diastolic pressures, pulmonary artery size and anatomy, ventricular function, and to exclude sub-aortic stenosis. Contra-indications to Fontan surgery included ventricular impairment of moderate or severe degree, atrio-ventricular valve regurgitation of moderate or severe degree, elevated left heart pressures, hypoplastic or severely distorted pulmonary arteries, and an elevated pulmonary artery pressure. The level of pulmonary artery pressure considered prohibitive cannot be stated absolutely, but in general a level over 15 mmHg would be considered with great caution. If there was severe desaturation after BCPS, we looked for venous collaterals from the SVC compartment to the IVC, and for evidence of pulmonary arteriovenous malformations.

2.4. Surgical technique

All Fontan procedures were performed on cardiopulmonary bypass, with moderate systemic hypothermia at 28–30°C. Our standard procedure in the earlier time period was an atrio-pulmonary anastomosis, utilising the right atrial appendage in most cases. Augmentation with a pericardial, homograft, or synthetic patch was used when necessary to provide an unobstructed anastomosis. Neosepation with PTFE was performed, leaving both AV valves on the left side. In selected cases with AV valve insufficiency, one AV valve was closed. For patients with tricuspid atresia, simple patch closure of the ASD was employed.

Commencing in 1989, and universally since June 1992, we used a modified TCPC, according to the technique of de Leval et al. [4]. A PTFE baffle was sutured into the right atrium to create a lateral tunnel connecting IVC to SVC. If the atrial septum was restrictive, then an atrial septectomy was performed. After release of the cross-clamp and evacuation of air from the left side of the heart, two cavopulmonary anastomoses were performed, to the superior and inferior aspect of the RPA.

We used fenestration regularly in the latter part of the series. A 4-mm fixed baffle fenestration was performed during a second short period of cardioplegic arrest or induced ventricular fibrillation. Initially it was performed in selected patients with unsatisfactory haemodynamics after attempting to wean from bypass and in 'high risk' patients, but with increasing use it has evolved as a routine part of the procedure.

Post-operative care involved avoidance of positive end-expiratory pressure, early weaning from mechanical ventilation and extubation, inotropic support with dopamine in moderate dose, and aggressive vasodilatation with phenoxybenzamine and nitroglycerin. Tight fluid restriction (50–60% of maintenance) was enforced for several days, and diuretics were used to minimise the tendency to pleural effusion. Patients were anticoagulated with warfarin unless there was major contra-indication, to achieve an INR or 2.5 to 3.

Bidirectional cavopulmonary shunts were performed on cardiopulmonary bypass with the heart beating, usually at 32°C. Bicaval cannulation was generally performed in the latter part of our experience. Other sources of pulmonary blood flow were eliminated, so that systemic to pulmonary shunts and the main pulmonary artery were ligated or divided. A brief period of cardioplegic arrest was used if an atrial septectomy was necessary. An end-to-side anastomosis was performed between the SVC and the RPA, and the cardiac end of the SVC closed directly. Any PA stenoses were relieved by patching with autologous pericardium, and associated subaortic stenosis was relieved by either Damus–Kaye–Stansel anastomosis or direct VSD enlargement. Early post-operative management was similar to that after Fontan repair, except that fluid restriction was less severe. Most patients after BCPS received aspirin at 5 mg/kg/day, but if there was extensive PA patch reconstruction then warfarin was sometimes administered.

2.5. Statistical methods

The comparison of time periods for the Fontan procedure, and the analysis of risk factors and differences in operative techniques were performed with univariate analysis (Fisher exact test). The analysis of the trend in mortality after the Fontan procedure was done by the method of Cuzick [3]. The pulmonary dimensions and Nakata index were compared before and after BCPS by paired analysis, using both parametric and non-parametric techniques (t-test and Wilcoxon signed-rank test). However, since the pulmonary artery data are not normally distributed, only the Wilcoxon result has been used. The Kaplan-Meier method was used for estimation of survival, freedom from Fontan failure, and probability of conversion to Fontan. All proportions are expressed with 95% confidence intervals (CI), using the Greenwood continuity correction.

3. Results

3.1. Fontan operation results

Comparison of the Fontan results for the two periods, 1980–1987 and 1988–1995 (Fig. 2), demonstrated a significant reduction in hospital mortality from 8.5% in the earlier period to 1.8% in the latter period \( (P = 0.02) \). The probability of takedown was 2.1% and 4.1%.
Fontan Operative Mortality by Year (n = 264)

Fig. 2. Mortality after the Fontan procedure from 1980 to 1995. There is a significant negative trend over the whole period ($P = 0.02$). Technical and strategic modifications introduced in each period are noted above each bar. The vertical axis refers to mortality probability. The first two columns refer to atrio-pulmonary anastomoses performed in two eras.

respectively in the two periods, but this difference was not significant ($P = 0.39$). The combined probability of death or takedown (an index of operative failure) was 10.6% in the earlier period versus 5.9% in the later period. This trend did not reach significance ($P = 0.23$).

For the second comparison, (1989–1991 versus 1992–1995), 138 patients were analysed. The differences between the two groups with respect to type of procedure, use of staging, and fenestration are detailed in Fig. 1. In the earlier period, only 3% of patients had been staged via a BCPS, compared to 58% in the latter period. The probability of TCPC repair increased from 41% to 92% in the latter period. Fenestration was used in only one patient in the earlier period, compared to 36% in the later period. There was a similar hospital mortality (2/66 vs 0/72, $P = 0.23$) and takedown risk (1/66 vs 4/72, $P = 0.26$) in the two periods, despite the major changes in surgical technique (Table 1).

The risk of operation for the period 1989–1995, examining the relationship between mortality or takedown and the use of TCPC, staging or fenestration, also demonstrated no effect of these particular tech-

Table 1

<table>
<thead>
<tr>
<th>Index</th>
<th>Change (mm²/m²)</th>
<th>$P$ (Wilcoxon)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right lower lobe</td>
<td>−33.9</td>
<td>0.001</td>
</tr>
<tr>
<td>Left lower lobe</td>
<td>−22.8</td>
<td>0.029</td>
</tr>
<tr>
<td>Both lower lobes</td>
<td>−37.3</td>
<td>0.002</td>
</tr>
<tr>
<td>Nakata</td>
<td>−51.6</td>
<td>0.016</td>
</tr>
</tbody>
</table>

Pulmonary artery changes after BCPS (mean interval = 15.5 ± 6 months). Magnitude of change in PA index did not correlate with age, BSA, interval between measurements, nor change Qp/Qs.

Actuarial freedom from Fontan failure

(early and late death, takedown, transplantation)

Fig. 3. Kaplan-Meier freedom from Fontan failure in two eras.

niques on outcome. However, there was a significant reduction in length of hospitalisation from 16.5 days for a non-fenestrated atrio-pulmonary procedure to 12.7 days for a fenestrated TCPC ($P < 0.05$), reflecting a shorter duration of pleural drainage and fewer recurrent pleural effusions.

Further analysis of the mortality data for the Fontan procedure in 4-year and then 2-year periods (Fig. 2) also demonstrated that there was a significant trend toward improvement over the whole period ($P = 0.02$). The most striking reduction in mortality appears to be in the period 1988–1989, after which the risk has remained very low. The freedom from Fontan failure is plotted in Fig. 3, and includes operative mortality, takedown, late deaths, and cardiac transplantation.

3.2. Bidirectional cavopulmonary shunt

One hundred and twenty-three patients underwent surgery with a hospital mortality of 4% (CI = 2–10%). The survival at 24 months was 90% (CI = 83–93%).

There was no relationship between mortality risk and age at surgery, ventricular morphology, previous source of pulmonary blood flow, subaortic stenosis or need for concurrent procedure (all $P > 0.05$). There were four episodes of caval or pulmonary thrombosis, with one early death and one late death, but the cohort is too small to enable statistical conclusions. Clearly, however, clinical experience indicates that this is a potentially devastating complication. Overall survival was 89% (CI = 81–94%), at 36 months, but the probability of progression to Fontan operation was only 49% (CI = 38% to 61%) (Fig. 4).

3.3. Pulmonary artery growth

In the subset of 23 BCPS patients undergoing detailed PA analysis, follow-up angiography was per-
formed at a mean interval of 15.5 ± 6 months. Although there was an absolute increase in the mean postoperative PA diameter, the Nakata index fell significantly from 320 to 268 mm²/M² (P = 0.0006) (Fig. 5).

The only patients in the series who had an increase in Nakata index had undergone concurrent PA reconstruction for branch stenosis. The same analysis was applied to the lower lobes, after the method of Reddy et al. [16], in order to minimise the possible confounding effect of central distortion from previous bands or shunts. Using normalised cross sections of either the right, left or both lower lobe PAs, a significant reduction in indices was again noted (P = 0.001, 0.028 and 0.002 respectively) (Fig. 6). These changes in Nakata and lower lobe index were not related to age or weight at the time of BCPS, to the interval between the two angiographic assessments, and nor were they related to the magnitude of change in $Q_p/Q_s$. Thus, the BCPS not only failed to effect PA enlargement during this interval, but did not even maintain the preoperative PA indices (Table 1).

4. Discussion

We have demonstrated that there has been a significant reduction in hospital mortality for the Fontan operation between the periods 1980–1987 and 1988–1995, but not of Fontan failure. This coincided with the introduction of the BCPS to our practise, although this may not entirely explain the fall in mortality. Comparison of the two later eras, 1989–1991 and 1992–1995, immediately before and after the introduction of fenestration, demonstrated no difference in mortality or operative failure. There was a significant decrease in both duration of pleural effusions and indirectly, length of hospitalisation. This probably reflects the effect of a modest reduction in right-sided pressures. Secondly, we have demonstrated that the results of the bidirectional cavopulmonary shunt in a large cohort of infants and children with a variety of lesions are satisfactory in the short and intermediate term. Thirdly, although there is an improvement in saturation in the short and intermediate term, there is impaired growth of the central pulmonary arteries after the BCPS, when it is used as the sole source of pulmonary blood flow.

The interpretation of comparisons between different eras and the attempt to associate any difference in results with a particular change in management is fraught with problems. There are often many changes occurring over time in addition to the changes in surgical technique, including changes in the attending surgeons, improvements in anaesthesia, cardiopulmonary bypass and post-operative care, and other changes in overall patient management which may be subtle and unrecognised. Similarly, in the assessment of risk fac-
tors for mortality or adverse outcome after a surgical procedure, particularly the Fontan operation, many of the patients who might be considered for an operation but have adverse risk factors will have been screened by the cardiologist, and may never be seen by the surgeon. A further selection is then imposed by the surgeon, to refuse or defer patients considered high risk, or channel them into another treatment stream. Therefore, if one fails to identify risk factors for adverse outcome, it may be no more than testimony to the good clinical judgement of the clinicians. Furthermore, the increasing acceptance of cardiac transplantation as an alternative to high risk surgery also alters the ability to isolate the risk factors in an analysis of surgical data, by providing another option for primary treatment or for Fontan related complications.

The significant fall in mortality for the Fontan operation in our practise is most readily explained by the introduction of the BCPS into our practise, so that high risk Fontan candidates would have been offered another option. Other factors cannot be excluded, as discussed, including perhaps more careful or more experienced patient selection for surgery.

The introduction of fenestration did not alter mortality, although it may have reduced morbidity. It should be recognised that the second comparison, (1989–1991 and 1992–1995) is limited by the already low mortality in the first period (3%) and by relatively small numbers in the two groups. To demonstrate a significant reduction in mortality in the second period would require no deaths in 250 consecutive patients. The conclusions for the second comparison are therefore somewhat preliminary.

Although there is no significant increase in the number of Fontan procedures taken down in the comparisons, there appears to be a trend for an increase as the mortality declines, so that if we analyse in terms of operative failure (combined death and/or takedown), there is no difference between eras. Familiarity with the BCPS in our unit after 1988 lowered our threshold to reverse the Fontan operation in patients with low cardiac output. The BCPS in this context is more likely to be successful than reversion to an arterial shunt. This change in surgical attitude also may have contributed to the reduction in mortality.

Late results of the Fontan operation have been disappointing in most centres, due to ventricular dysfunction, arrhythmia, protein losing enteropathy, and thromboembolism. Looking at Kaplan-Meier survival in the latter era (Fig. 3), although limited follow up time is available, the curve is roughly parallel to that of the earlier era. This is consistent with a hazard function that is similar over the first 5 years, but with a reduced early mortality. In any case, the widely overlapping 95% confidence limits preclude a statistically significant difference ($P > 0.05$). It is possible that these curves will diverge with longer follow-up time, and that the benefits of current surgical strategies will be realised in the second decade. Many more years of follow-up will be required in order to assess differences in outcome using the modern treatment strategies outlined here. Transplantation may reduce early Fontan mortality by removing high risk candidates from the cohort and improve late survival through application as a ‘rescue’ procedure.

The BCPS is useful for palliation of patients who may eventually be Fontan candidates, but who are currently unsuitable for reasons of young age, high PA pressure, PA distortion, atrioventricular valve regurgitation or ventricular dysfunction. This procedure should have a low mortality if staging by two operations is to be a better alternative than one-stage Fontan operation, and should avoid impairment of pulmonary artery growth, which might have adverse effects on long-term Fontan candidacy and/or outcome. The efficacy of this approach must be assessed in terms of symptomatic benefits compared with alternatives, surgical risk of the BCPS, and influence on the risk associated with a subsequent Fontan operation. The major theoretical advantages of the BCPS are the reduction of volume load on the systemic ventricle, reduction of atrioventricular valve regurgitation, and the protection of the pulmonary vascular bed from high flow or high pressure. However, it has been difficult to prove that the procedure does indeed reduce AV valve regurgitation, or that the reduced volume load provides measurable long-term benefit for ventricular function. There has been good clinical evidence supporting the use of the BCPS as a means to reduce ultimate Fontan risk in various anatomic subsets [7].

The BCPS has an acceptable early outcome, and the systemic oxygenation is maintained or improved in the short and intermediate term. However, in the long term, progressive desaturation will occur in a large proportion of patients, due to the development of venous collaterals from the SVC to the IVC and intrapulmonary arteriovenous shunting and reduction of the SVC/IVC flow ratio with age [2,8,11]. The operation probably provides a limited period of benefit in many patients. In the series of patients reported from Glenn’s own work, two thirds of the cohort required further surgery to improve oxygenation over a mean follow-up of 19 years [8].

If the fall in mortality for the Fontan procedure in the late 1980s in our practise was indeed due to the introduction of the BCPS, then it becomes important to see whether the mortality remains low as those previously ‘high risk’ patients managed by BCPS become eligible for Fontan completion, and whether the use of the BCPS has truly altered the results of surgical intervention and the patient outcome or simply deferred the surgical mortality. So far, the Fontan mortality has
remained very low or zero since these 'staged' patients have begun to undergo completion. However, fewer than 50% of the patients palliated by BCPS have undergone completion by 3 years, and a significant number of those not offered completion remain unsuitable for a Fontan operation. This also reflects an evolving reluctance to convert stable BCPS patients to a Fontan circulation. It is therefore possible that we have improved short-term survival by eliminating 'higher risk' Fontan procedures in favour of the BCPS, but have not yet addressed the question of whether we are providing a better long-term option. The majority of these patients will inevitably require further palliation (e.g. systemic to arterial shunts) due to desaturation if they are not suitable for Fontan completion. It may be that the patients who previously had an adverse outcome from a Fontan operation are now alive but increasingly poorly palliated by the BCPS, remaining at high risk for a Fontan operation.

In this context, if we are to maintain the long-term options for patients receiving a BCPS, the observations regarding PA growth are of some concern, and are in agreement with a previous study [11] which demonstrated a 32% decrease in the Nakata index after BCPS, with no change in the size of the pulmonary artery ipsilateral to the Glenn anastomosis and a decrease in the size of the contralateral pulmonary artery. The mechanisms responsible for PA growth and development have been discussed elsewhere by our group [13]. Other recent work (Reddy et al. [15]) describes 28/112 children progressing to a Fontan operation after BCPS. PA index in patients without an extra source of pulmonary blood flow decreased, while the presence of an extra source of blood flow maintained PA growth. This difference did not affect the results of Fontan conversion in this group. However, in both this study and our own, the statements regarding PA growth are extrapolated from small subgroups, which may not be representative of the changes in the whole group of patients. Uemura et al. [16] also demonstrated preservation of PA size by the maintenance of forward flow from the ventricles. However, other data regarding the effect of extra-pulmonary blood flow are controversial. There are reports of an increased incidence of chylothorax due to higher central venous pressures [5], a higher incidence of pleural effusions and a higher later mortality, which has led some surgeons to routinely eliminate other sources of flow [10].

In conclusion, the mortality from the Fontan operation has decreased significantly in our institution, and our best explanation indicates that this has been due to the introduction of the BCPS, probably by offering a lower risk option. The use of fenestration has reduced postoperative morbidity. The results for the BCPS are acceptable, with satisfactory saturation and palliation in the short term. However, a significant number of patients having undergone BCPS remain unsuitable for Fontan conversion, and their long-term management is unclear. Furthermore, PA growth may be compromised by long-term palliation with the BCPS, although further work is necessary to assess this problem.

Acknowledgements

Some of the operations analysed herein were performed by Mr. Roger Mee, Mr. William Brawn, Professor Shunji Sano and Mr. Ash Pawade, as well as by the authors.

References

in our own practice, especially in the past 18 months, we've had a
who are known to have a smaller proportion of their total venous
return draining through the SVC are unlikely to have the same degree
of pulmonary blood flow, so why would they grow, their flow is going to be less
improved in the long-term survival end result.

And part of the reason for saying that is when we looked histori-
cally at our Fontan series and analyzed the patients who went on
from a cavopulmonary shunt to a Fontan versus those who never did,
for whatever reason, and often the reason was they're inoperable,
they didn't tolerate having the superior vena cava being the only source of pulmonary
blood flow. So my first question is, do you have an age limit for that policy?

The second query I have is about the growth of the pulmonary arteries. The fact that the pulmonary arteries are not growing after your
cavopulmonary shunt may indicate two things: number one, you've successfully cut down on the extraneous source of pulmonary blood flow, so why would they grow, their flow is going to be less than normal. But I would take that as an indication to move ahead with the Fontan earlier rather than later.

And part of the reason for saying that is when we looked histori-
cally at our Fontan series and analyzed the patients who went on
from a cavopulmonary shunt to a Fontan versus those who never did,
for whatever reason, and often the reason was they're inoperable,
there was no difference in late survival. Survival out to 20 years is about 50%, whether or not you complete the Fontan or not.

And we interpreted that to mean that we shouldn't be waiting until the children get in trouble and then converting them to a Fontan. And I sense your message is that you are doing that, you are waiting until they get older and their hematocrits are rising and their ventric-
ular function is failing, at which point you're going to consider a Fontan. And I wonder if that's your policy and would question
whether that's the right approach in the current era. I think we really
don't know what the Fontan is going to do long term for survival,
but I think the policy of waiting indefinitely until the children are in trouble has not been a success. And our hope was that moving ahead,
completing the third stage in the first two years of life, would improve the long-term survival end result.

Dr. T.R. Karl: Firstly, regarding the age cutoff for cavopulmonary
shunt as a unique source of pulmonary blood flow, I would say that
in our own practice, especially in the past 18 months, we've had a
strong tendency to use cavopulmonary shunts exclusively with a
second source of pulmonary blood flow based on the findings of the
study of this initial cohort of patients. And I agree that older patients
may have an arterial oxygen saturation of 80% and yet have a hemoglobin of 21,
and that's because of severe desaturation on exercise. So we are in favor of leaving the additional source of blood flow.

With respect to the timing of the Fontan operation, I think we've
moved toward a policy of not converting stable and cavopulmonary
shunt patients to a Fontan circulation meaning that if we can expect
a hemoglobin saturation following the fenestrated Fontan operation
that's only marginally better than that which has already been
achieved with the cavopulmonary shunt, there really isn't much point,
weighing in the increased long-term risk following the Fontan opera-
tion. We feel that perhaps the clock does start to tick in some respects
when the Fontan is completed.

And in terms of what events should occur to tell us that we need to
move toward a Fontan operation, certainly a fall-off in ventilricular
function is something that's just as likely to occur after a Fontan as
after a cavopulmonary shunt, or perhaps just as unlikely to occur
after a cavopulmonary shunt as after a Fontan, since in either case
we've eliminated the phenomenon of pulmonary recirculation. I think
that the issue of cyanosis is the one that really needs to be addressed.
And the message here is not that we should wait until we have poor
ventricular function and then do a Fontan operation, really it would be quite the opposite.

Dr. H. Laks (Los Angeles, California, USA): I think, first of all, the
conclusion that there has been no improvement in the Fontan could
be debated, no improvement with these modifications. In fact, your
incidence of mortality and takedown dropped from over 10% to
somewhere around 6.9%. And although that may not be statistically
significant, that may be related to the numbers involved rather than
to the fact that there is no change.

Secondly, these are two different time periods, and I think we
would all agree that as we have seen the unfolding of poor early and
late results for patients who are poor candidates for Fontans, our
selection criteria in these time periods have changed so that we are
doing less high risk Fontans, as you have mentioned, and leaving
them with their bidirectional Glenns, and so that would also account
for some difference in the mortality. So I think that you have in fact
suggested quite strongly from this data that there has been an
improvement with these modifications.

The other point I'd like to make is that the bidirectional Glenn, our
practice has always been, and I think your data substantiates, that it
is important to leave an additional source of pulmonary blood flow. I think that the additional source of flow got some bad press when
surgeons were in the habit of taking a patient with a QP/QS of 1.5,
or 1 to 2, from an additional source of flow such as a shunt or a
banded PA and then simply adding a Glenn, ending up with a QP/QS
of 2 to 1 and then seeing pulmonary hypertension and high venous
pressures. If, however, one aims for a QP/QS of somewhere between
1 and 1.3 to 1 from these two sources of pulmonary blood flow, then
you see many advantages. You do not see a volume overload.
You see growth of the pulmonary arteries. And if there is this so-called
hepatic factor, you will have some of your mixed blood going to the
pulmonary circulation and you will also have a response to exercise
which you do not see with a Glenn on its own. As you triple your
inferior vena cava flow, your SVC flow for children playing may
remain exactly the same. And the severe desaturation on exercise
probably accounts for the fact that you may see a patient who at rest
has an arterial oxygen saturation of 80% and yet has a hemoglobin of 21,
and that's because of severe desaturation on activity.

So we are in favor of leaving the additional source of blood flow.
It also probably cuts down on the development of bronchial collateral
flow which can also decrease, and increase the risk when you do the
douential Fontan. It doesn't make any sense, particularly as children
grow older, to have a QP/QS of 0.3 or 0.4 and expect that you will not
give AV fistulae and decreasing size of your pulmonary arteries. So I
congratulate you on really excellent results and on your conclusions.

Dr. T.R. Karl: I don't think I have anything to add. I mean that our
data really does support everything that you said. And the points
about patient selection evolving over this time period are really
the most salient ones. And I would just like to say that Dr Laks was
already performing beautiful Fontan operations in 1981 when I was
his junior resident.

Dr. B. Muraszewski (Warsaw, Poland): Do you believe that looking
into the future, attaining further improvement and development in the
Fontan circulation, should we try to shorten the anastomosis in the
sense of creating the direct extracardiac nonconduit anastomosis in
as many patients as it is anatomically possible?
Dr T.R. Karl: I think that's a good general principle, although there isn't anything in our data to support or refute that. But in general I think it makes sense.

Dr M. Ah. Navabi-Shirazi (Shiraz, Iran): Do you advise separating the blood flow to the lungs if you encounter high SVC pressure after performing Glenn? By separating, I mean the banded PA to the right lung and Glenn to the left lung.

Dr T.R. Karl: We have done this in just a few cases, primarily patients with pulmonary atresia and intact septum. And I don't really have enough experience to advise you one way or another on whether that's a good routine practice in patients in whom absolute pulmonary blood flow is going to be a big worry after this Glenn procedure. I know Dr Laks has done that in many of his patients and perhaps you could talk to him about it.