Equivalent survival following cavopulmonary shunt: with or without the Fontan procedure


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Abstract

Objectives: In 1992, an analysis of our experience with the cavopulmonary shunt (CPS) demonstrated equivalent long-term survival, with or without subsequent conversion to a Fontan circulation. Before 1992 (era 1) intervention was deferred until mandated by clinical deterioration. Since 1992 (era 2), timing of both CPS and Fontan was compressed in an effort to improve survival. Survival following CPS is analyzed to ascertain whether Fontan confers any survival advantage over no further definitive intervention. Methods: From 1962 to 1997 inclusive, 490 patients underwent CPS, excluding those who had a CPS concomitant with a Fontan. In 55 patients the CPS was performed at or after a biventricular repair (BVR), or after a Fontan, and these patients are excluded. The 435 patients remaining followed a surgical protocol which included a subsequent BVR (n = 28), or a subsequent Fontan operation (n = 220), or no further definitive surgery (CPS only, n = 187). Between eras the mean age at surgery decreased for all procedures. Results: Long-term survival 20 years after a CPS in 435 patients is 56 ± 5%. Survival at 20 years among the 220 patients who were subsequently converted to a Fontan circulation is 65 ± 8% compared to 50 ± 11% for the 187 patients who did not have a Fontan. However, most of their survival difference is because all early deaths after a CPS occurred in the non-Fontan group. Multivariable analysis demonstrated that proceeding to a Fontan did have a small survival advantage which was not evident by univariate analysis. Independent risk factors for death, at any time, are a common atrioventricular valve, or pulmonary artery banding. The era had no effect on survival. Conclusions: The single ventricle circulation appears to have a limited durability of, an average, 30–40 years. There is a slight survival advantage in converting patients after a CPS to a Fontan circulation. A marked reduction in age at CPS and at Fontan has, as yet, not improved survival.

Keywords: Cavopulmonary shunt; Fontan; Univentricular heart; Single ventricle; Survival

1. Introduction

Evaluation of the efficacy of the superior vena cava to pulmonary artery shunt (cavopulmonary shunt, CPS) is complicated by the changing objective of the operation. Prior to the advent of the Fontan operation, the CPS was used as a palliative procedure in patients with complex congenital heart disease resulting in a single ventricle circulation. For these patients, a CPS to the right lung and an arterial blood supply from the heart and/or a systemic to pulmonary artery shunt to the contralateral lung provided a 'balanced circulation' and reasonably good long-term definitive palliation. The Fontan/Krützler procedure changed the objective of the CPS to one of 'staging'. A preliminary CPS was used to decrease the risk of a subsequent Fontan operation.

In 1992, an analysis of our experience (unpublished) demonstrated equivalent survival among patients who had a CPS alone compared to those who subsequently underwent a Fontan procedure. The actuarial survival in either group was 55–65% at 20 years after the CPS. These results are similar to the long-term survival after the Fontan procedure reported by other groups [1,2].

Prior to 1992, our protocol for patients with univentricular heart was to defer surgical intervention, either CPS or Fontan, until the patient became symptomatic or was increasingly cyanosed. Subsequent to our analysis in 1992, we changed our patient management protocol to intervene electively.
at an earlier age for both the CPS and subsequent Fontan. The present analysis examines the effect of this change in patient management strategy.

2. Methods

We reviewed all patients undergoing a cavopulmonary shunt at the Hospital for Sick Children and Toronto Congenital Cardiac Centre for Adults from the first patient in July of 1962 until the end of December 1997. Patients who had a CPS concomitant with a Fontan operation are not included in this study. Hospital records, including follow-up clinic reports and correspondence with referring physicians for distant referrals were reviewed. For patients who were lost to follow-up, we attempted contact by telephone with the family and searched government mortality records.

A total of 490 patients were identified. The CPS was performed as part of a biventricular repair (BVR), so-called 1.5 ventricle repair in 39 patients and was performed subsequent to a BVR or after a Fontan operation in 16 patients. These 55 patients are excluded from further analysis.

The remaining 435 patients (Table 1) had a CPS which preceded a Fontan operation \( n = 220 \) or a subsequent BVR \( n = 28 \) or they had no further definitive surgery \( n = 187 \). Mean follow-up of these 435 patients is 6.8 years, and for those discharged from hospital, 7.6 years (range 0.2–33.1 years). Follow-up of the hospital survivors is complete to after January 1, 1997 in 85% of the patients. Six percent have not been followed after January 1, 1996 and are lost to follow-up, although these 19 patients were followed a mean of 11.5 years after a CPS (range 1.5–24.9 years) before being lost to follow-up. The diagnostic categories are listed in Table 2.

Survival analysis (Kaplan–Meier) for all 435 patients after cavopulmonary shunt was performed. Subset analysis before and after 1992, (the year of our change in clinical protocol to intervention at an earlier age) compared outcomes among the three groups of patients with and without a subsequent Fontan or BVR (Table 2).

The age at surgery prior to and after 1992 illustrates the change in patient management protocol. (Table 3). Data are described as frequencies, medians, with ranges, and means \( \pm \) standard deviation. Survival estimates are calculated using Kaplan–Meier estimates and plotted from various entry points; primarily from birth, or from CPS (CPS only group), or from Fontan. Independent risk factors...
were sought in Cox’s proportional hazard modeling. A value of \( P < 0.05 \) was set as the level of statistical significance. Variables entered include era 1 vs. era 2, technique of caval anastomosis, morphology (five types), diagnosis, age at surgery (initial arterial shunt, CPS or Fontan), year of birth, year of CPS, and year of Fontan.

In the survival analysis, all of the early (operative) mortality after a cavopulmonary shunt was charged to the ‘CPS alone’ group because early death precluded a subsequent Fontan or BVR. To overcome this bias, survival analysis was also performed for all early survivors, i.e. survivors greater than 31 days post cavopulmonary shunt (\( n = 407 \)). The clinical relevance of this approach is pragmatic as it addresses the question of prognosis after a successful CPS with regard to whether or not a subsequent Fontan is performed.

The survival curves for patients with a CPS only or CPS and subsequent Fontan operation were plotted from birth, from the CPS, and from CPS excluding early (< 31 days) deaths.

3. Results

During the follow-up period, 111 patients (26%) died including 31 early (less than or equal to 31 days after CPS) and 80 late deaths a mean of 8.9 years after CPS (range 70 days–32.8 years, median 4.7 years). Long term survival after CPS in 435 patients is 76% at 10 years, 57% at 20 years and 54 ± 4.8% at 25 years (Fig. 1). Mean age at CPS is 4.0 ± 6.1 years (range 0.05–49.8 years), but varied significantly between eras (Table 3).

Late death (\( N = 80 \)) occurred at subsequent Fontan operation in 21 patients or at BVR in seven patients. During follow-up, surgical procedures were required in 121 patients including 19 re-operations after a Fontan with six early deaths, and 58 among the 187 patients in whom no further definitive surgery was undertaken, and four after a BVR.

The reasons for not proceeding to a Fontan operation in the patients who have a CPS only are listed in Table 4. Survival from birth of the 407 patients who had either a CPS alone, or a CPS and subsequent Fontan operation is illustrated in Fig. 2, which excludes the BVR group. Survival to age 30 years among patients who have a CPS is 54% (±5%).

For those patients who were subsequently converted to a Fontan circulation (\( n = 220 \)), survival is statistically better than for the 187 patients who have had only a CPS (Fig. 3a). Most of this difference appears to be in the early perioperative period. When the early mortality after a CPS is excluded, then the survival for patients who have a subsequent Fontan operation tends to be better than for those with a CPS alone but this trend is not statistically significant by univariant analysis (\( P = 0.09 \)) (Fig. 3b).

The equivalent survival following a successful CPS whether or not a subsequent Fontan is performed was observed prior to 1992, and persists in the experience after 1992 in spite of a change to a younger age at initial palliation, at CPS and at Fontan (Table 3). The survival of patients with a CPS prior to 1992 is compared to that of patients after 1992 (Fig. 4). Although the Kaplan–Meier analysis shows no statistically important differences in late survival, the earlier conversion from CPS to Fontan may result in a greater number of patients coming to Fontan (Table 1). To date, 56% of the earlier patients and 60% of the later era patients have undergone Fontan. Few, if any, of the surviving early era patients are Fontan candidates whereas many of the 78 surviving patients in the later era will become Fontan candidates (Table 7).

By Cox’s proportional hazard modeling, increased survival from CPS or Fontan or from birth is independently associated with a Fontan protocol. (Table 5) Both pulmonary artery banding and a common AV valve are independent
risk factors for decreased survival, regardless of where the survival curves are started from. These variables were consistently significant when the analysis was repeated for survival from birth, from the date of CPS or Fontan, or from CPS with early deaths excluded. The risk ratios for all models are shown in Table 6.

4. Discussion

The newborn with a univentricular circulation requires a cardiac output greater than normal to maintain adequate oxygenation. Ample evidence demonstrates that earlier conversion of an arterial to a cavopulmonary shunt reduces the volume load on the ventricle [3–5]. These considerations led Norwood to recommend staging of single ventricle patients with a cavopulmonary shunt, using a hemi-Fontan operation, at 6 months with a subsequent Fontan at 12–18 months [6]. He demonstrated an increase in Fontan operative survival with this ‘staging’ approach. Jacobs reported no increase in operative risk for the hemi-Fontan in infants as young as 2 months in their extensive experience [7]. In contrast, our data demonstrate a marked increase in operative risk of 20% in the 50 patients less than 6 months compared to a risk of 1.6% for babies aged 6–12 months (n = 124). The increased risk in patients less than 6 months explains the rise in operative risk for the CPS from the earlier era (6%) to the more recent era (8%).

Although younger age at Fontan was associated with less morbidity in Vogel’s report [8], early survival was not affected. In our series of 435 patients with a CPS, a substantial decrease in the mean age at CPS from 4.6 years to 1.5 years, and at subsequent Fontan operation from 9.8 years to 3.4 years, did not significantly change early or late survival. However the longest follow-up in the more recent younger

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Table 4

The reasons for not proceeding to a Fontan operation are listed for the 187 patients with a CPS only.

<table>
<thead>
<tr>
<th>Reason</th>
<th>N</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan pending (too early)</td>
<td>54</td>
</tr>
<tr>
<td>Died early after CPS</td>
<td>31</td>
</tr>
<tr>
<td>Died late after CPS</td>
<td>29</td>
</tr>
<tr>
<td>Risk of Fontan too high</td>
<td>57</td>
</tr>
<tr>
<td>Patient/family declined</td>
<td>4</td>
</tr>
<tr>
<td>Doing too well</td>
<td>4</td>
</tr>
<tr>
<td>Heart transplant</td>
<td>3</td>
</tr>
<tr>
<td>Unnecessary</td>
<td>1</td>
</tr>
<tr>
<td>Unknown</td>
<td>1</td>
</tr>
<tr>
<td>Lost to follow-up</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>187</td>
</tr>
</tbody>
</table>

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Fig. 3. (A) Survival comparison by univariate analysis for 220 patients who had a subsequent Fontan operation with 187 who had no further definitive surgery. There is a statistically significant difference. Most of the difference in survival appears to be due to the early mortality, all of which is charged to the ‘CPS only’ group. (B) The early postoperative mortality (n = 31) after the CPS has been excluded. There is a trend of better survival among patients who had a subsequent Fontan operation, but by univariate analysis there is no significant difference in survival.

Fig. 4. Kaplan–Meier survival among patients treated at an earlier mean age since 1992 (n = 216) is not different than the 216 managed prior to 1992. Operative risk in the earlier era (6.0%) is not statistically different than in the more recent era (8.3%). The late mortality after a CPS is 23 of 179 patients (12.8%) in the era before 1992 and nine of 196 patients (4.6%) in the recent era. Maximum follow-up in the more recent group is 6 years, at which point the survival in both groups is 80%.
patients is only 6 years at which point the survival is 80%. There may be a later survival advantage which is not evident yet.

The superior caval flow changes with growth. Salim et al. demonstrated that in healthy children superior vena caval flow rose from 49% of total cardiac output in the neonate, to a maximum of 55% at 2.5 years, and declining to an adult level of 35% by age 6.5 years [9]. Therefore maximum benefit of the superior vena cava to pulmonary artery shunt should occur between the age of 6 months to 3 years. An inferior cava to pulmonary artery connection will have important reciprocal effects. A child with an established CPS should have a lesser hemodynamic transition to Fontan’s circulation at 3 years than after 6 years. Indeed older children do not tolerate a CPS as the only source of pulmonary blood flow, and prior to the advent of the Fontan, the ipsi-lateral lung was provided with an arterial shunt as definitive palliation. Our data shows that this ‘balanced circulation’ is capable of very long-term survival.

The assumption that the single ventricle performs optimally with a volume which is appropriate for a normal biventricular heart may not be correct. Dr Pascal Vouhé has suggested that a volume of 1.5 times normal may be more efficient for the single ventricle. The minimal difference in long-term survival among our patients with a CPS only compared to those with a subsequent Fontan suggests that the effect of the diminished preload in the Fontan population is minimal and that there is a finite limit to the durability of the single ventricle circulation. The graph of survival from birth (Fig. 2) excludes those infants who did not survive to a CPS and is therefore inaccurate. There is a sizable attrition in single ventricle patients as has been documented by Franklin and others [10,11]. However the plot of survival from birth is of interest in relating long-term survival to age and does illustrate the present limits of the single ventricle.

Table 6
Risk ratios from the hazard model a

<table>
<thead>
<tr>
<th>Survival from:</th>
<th>Birth</th>
<th>CPS</th>
<th>CPS (exclude early deaths)</th>
<th>CPS or Fontan</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan protocol</td>
<td>0.36</td>
<td>0.35</td>
<td>0.63</td>
<td>0.56</td>
</tr>
<tr>
<td>CPS Age</td>
<td>0.90</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Previous PAB</td>
<td>1.99</td>
<td>2.15</td>
<td>2.00</td>
<td>1.95</td>
</tr>
<tr>
<td>Later era (post 4/92)</td>
<td>1.90</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Common AV connection</td>
<td>2.17</td>
<td>2.32</td>
<td>2.37</td>
<td>2.16</td>
</tr>
</tbody>
</table>

Table 7
Timing of death in 407 Patients after a CPS

<table>
<thead>
<tr>
<th>Number of patients</th>
<th>Interval from CPS to Event (years)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt; 1992</td>
</tr>
<tr>
<td></td>
<td>N = 193</td>
</tr>
</tbody>
</table>
| None             | 127      | 181      | 12.6  | 2.8
| At CPS           | 14       | 18       | 13.0  | 0.9
| Interval         | 29       | 6        | 8.3   | 1.8
| At Fontan a      | 13       | 8        | 10.5  | 2.7
| After Fontan     | 10       | 1        | 11.4  | 2.4

a Number of patients converted to a Fontan operation: < 1992, 102 of 179 (57%) late survivors after a CPS; > 1992, 118 of 196 (60%) late survivors after a CPS.

a The 28 patients who had a subsequent BVR are excluded from this analysis. Variables not selected into model: Date of birth; date of CPS; age at CPS; Previous BT; Norwood; VA connection; vent morphology; isomerism; Glenn; Era.
permanently fenestrated Fontan or alternatively, an inferior cava to pulmonary artery anastomosis without a superior caval connection to the pulmonary circulation, may provide an optimal combination of adequate preload, satisfactory mixing and less resistance [12].

Acknowledgements

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References


Appendix A. Conference discussion

Dr A. Corno (Genolier, Switzerland): What is the practical consequence of this study in your institution? Did you change your policy? What do you do now when you have a patient after a cavopulmonary shunt coming back, do you complete the cavopulmonary connection or not?

Dr Williams: The obvious inferences from this is, is it worth completing a Fontan in a patient who had a successful cavopulmonary shunt? I think the answer to that is probably yes. I think I would phrase it the other way, however. If you have a patient with a stable cavopulmonary shunt who you think is a high risk for a Fontan, I think there is good reason not to go on to a Fontan, because the outcome for those patients without Fontan conversion, at least in terms of survival, is better than one might anticipate. Many of the patients in this series who were followed-up 30 and 35 years were deemed inoperable for a Fontan and had an arteriovenous fistula, and 20 years later are still coping all right. So I don’t think this means we shouldn’t be doing a Fontan operation, but I think it does stress that we have to select patients very carefully and not try to do Fontans on everybody.

Dr D. Di Carlo (Rome, Italy): In our Center, we have adopted enthusiastically the bidirectional cavopulmonary anastomosis and we initially believed that it could represent a form of definitive palliation, an alternative to the more hazardous Fontan procedure, along with the concepts that you just mentioned. We were quite disappointed to see that in many patients the symptomatic improvement did not extend beyond 2 years from surgery and cyanosis gradually reappeared. I wonder, are the patients you referred to as doing very well with a classic or bidirectional Glenn anastomosis possibly also excellent candidates to a Fontan operation? And conversely, are those who have early recurrence of symptoms after a Glenn shunt unlikely to do well whatever surgery is undertaken? In other words, I believe that your results by applying the Fontan operation to patients who were greatly improved after a Glenn shunt would probably be very good as well. And for those who did not fare well with a Glenn shunt, the only alternative would be heart transplantation.

Dr Williams: The question is similar to the first one: were all of the poor risk patients in the cavopulmonary shuntalone group? Not necessarily. Many of them were, but many were clearly not candidates for anything further except for transplant. However when you follow them year after year, they continue to do better than I personally would have expected, at least in terms of survival. But I stress again we had not assessed functional ability.

Dr B. Maruszewski (Warsaw, Poland): With all your experience and longterm results, what are nowadays your indications, your criteria, for doing cavopulmonary shunt in the 1-year-old baby or rather going for complete Fontan? How do you make this decision?

Dr Williams: We use the agreed-upon criteria to a Fontan of ventricular function and PA anatomy and resistance. We try to avoid pulmonary artery bandings. There is not much we can do about children with a common AV valve except, as is general practice, we try to fix the AV valve at the time of a cavopulmonary shunt before going on to a Fontan. Again, the message is if none of that worked and you have a patient with a cavopulmonary shunt that you’re worried about, it does also make sense to go on to a Fontan. But I don’t think it’s changed our criteria per se.