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Original Article

Can we do without routine fenestration in extracardiac total cavopulmonary connections? Report on 84 consecutive patients

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Abstract Fenestration is still widely used in right heart bypass operations. Our study was conducted to assess its need in the most recent modification, the completion of a total cavopulmonary connection with an extracardiac tube.

The extracardiac approach was introduced at our institution in January, 1999. Since June of 2000, no patient had a fenestration. If more than 1 risk factor amongst ventricular function being more than moderately impaired, atrioventricular valvar regurgitation more than moderate, mean pulmonary arterial pressure more than 15 millimetres of mercury, mean atrial pressure higher than 12 millimetres of mercury, pulmonary arterial distortion, or other than sinus rhythm was present preoperatively, the patient was considered a "high risk" candidate. Postoperatively elevated pulmonary arterial pressure higher than 16 millimetres of mercury, prolonged effusions and requirement for drainage longer than 7 days, and death were considered endpoints in the statistical analysis.

Our study group included 84 patients who underwent surgery up to August, 2004. A previous bidirectional cavopulmonary anastomosis had been accomplished in 73 patients at a mean age of 27.01 plus or minus 32.60 months, with a median of 11.5 months, without creating an additional source of flow of blood to the lungs.

At the time of the total cavopulmonary connection, the mean age was 66.4 plus or minus 60.1 months, with a median of 37.1 months, and a range from 17.3 to 251.2 months, with 50 patients being younger than 48 months.

We deemed 16 patients to be at "high risk". These patients were older at the time of bidirectional cavopulmonary anstomosis (p smaller than 0.016), at the time of completion (p smaller than 0.019), and also differed in size at time of completion (p smaller than 0.020). They required a longer time on cardiopulmonary bypass (p smaller than 0.015), and reached higher early postoperative pulmonary arterial pressures after completion (p smaller than 0.025). There were no differences between groups of patients having up to 1 or more risk factors in regard to need for intubation (p smaller than 0.511), pulmonary arterial pressures after extubation (p smaller than 0.817), and duration of chest drainage (p smaller than 0.650). Three patients died, one in the group deemed at high risk. There was no death in the last 38 patients.

We conclude that a total cavopulmonary connection with an extracardiac tube can be performed without fenestration, even if the patients are deemed to be at increased risk. Early staging of patients with functionally univentricular physiology might be one of the keys for these findings.

Keywords: Total cavopulmonary connection; Fontan procedure; functionally univentricular heart

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In January of 1999, we introduced the extracardiac approach for the Fontan procedure at our institution. Gradually, construction of an intracardiac tunnel was abandoned, and from August of 2001, we have performed only extracardiac completions.[1] Overall, until August of 2004, 112 patients have undergone an extracardiac completion. Up to June of 2000, even those patients with an extracardiac connection also received an additional fenestration, which was done at the preference of the individual surgeon. In total, 9 of the patients received a fenestration. In this study, we have focussed only on our cohort of patients undergoing surgery from June 2000, the time we abandoned any fenestration. By focusing selectively on patients who underwent the extracardiac completion without fenestration, we are able to report on a homogenous procedure in our population. In addition, the patients included in the study were operated over a relatively short period of calendar time, specifically 4 years, providing uniformity in the management of pre- and postoperative care. We sought to identify preoperative, operative and postoperative characteristics that might have influenced outcome following completion to an extracardiac cavopulmonary connection without fenestration.

Patients and methods

Our study included all patients seen between June, 2000, and August, 2004, in whom a cavopulmonary pathway was completed with an extracardiac conduit. Retrospective review of their charts was conducted to obtain detailed information about the preoperative, operative, and postoperative clinical course.

The patients were matched to defined preoperative risk factors (Table 1). All patients had preoperative invasive assessment. Pulmonary arterial and atrial pressures were obtained during cardiac catheterisation. At our institution, we have well-established cut-off values for both pulmonary arterial and atrial pressures. Likewise, ventricular function, and atrioventricular valvar regurgitation were assessed by echocardiography. Ventricular function was judged as normal, or mildly, moderately, or severely impaired. Valvar regurgitation was classified as none, trivial, mild, moderate, or severe, taking note of features of the jet with pulsed flow Doppler and in colour Doppler in parasternal short-axis view and apical views. Pulmonary arterial distortion was recorded when review of the cardiac catheterization studies combined with the recorded operative findings demonstrated significant stenoses or distortions in the central pulmonary arteries or markedly hypoplastic central or peripheral pulmonary arteries. These patients required surgical interventions at the site of the pulmonary arteries. A quantitative measure of the size of the central pulmonary arteries was not used. The patients were also divided into those with sinus rhythm, ectopic atrial rhythm, atrioventricular escape rhythm, or pacemaker-dependent rhythm. Atrioventricular nodal function was measured in terms of the PR interval. First degree atrioventricular block was considered present when the PR interval was longer than 150 milliseconds in children younger than 12 years, longer than 180 milliseconds from 10 to 15 years of age, and longer than 200 milliseconds in adolescents and adults older than 15 years. We defined the Mobitz and Wenckebach variants of second degree atrioventricular block, and complete atrioventricular block, on the basis of the electrocardiographic tracings. Tachycardiac events were assessed only if reported in the medical history.

Table 1. Defined preoperative risk factors.

Mean pulmonary arterial pressure higher than 15 millimetres of mercury
Mean atrial pressure higher than 12 millimetres of mercury
Ventricular function more than moderately impaired
Atrioventricular valvar regurgitation more than moderate
Pulmonary arterial distortion
Other than sinus rhythm

Of the patients, 52 had a dominant left ventricle, and 32 a dominant right ventricle. Among the underlying morphologies were tricuspid atresia in 16 patients, pulmonary atresia with intact ventricular septum in 6, double inlet left ventricle in 16, hypoplastic left heart syndrome in 8, and isomerism of the atrial appendages and heterotaxy in 4.

The patients had a mean of 2.18 plus or minus 0.92 operations, with 183 operations in all, the range being from 0 to 5, prior to the total cavopulmonary connection. A previous bidirectional cavopulmonary anastomosis was accomplished in 73 patients, whereas in 11 it was not. Generally, no additional antegrade flow of blood to the lungs remains at the time of bidirectional cavopulmonary anastomosis at our institution.

Only one patient had a primary total cavopulmonary connection, this patient having congenitally corrected transposition with pulmonary valvar stenosis and criss-cross heart, and undergoing surgery at the age of 12 years. In 3 patients, the pulmonary trunk had been banded as an isolated procedure prior to the total cavopulmonary connection, these patients having congenitally corrected transposition with hypoplastic right ventricle; congenitally corrected transposition with pulmonary valvar stenosis; and double inlet left ventricle, discordant ventriculo-arterial connections and obstruction to the outflow tract from the dominant left ventricle. In 7 patients, a systemic-to-pulmonary arterial shunt was followed by a total

cavopulmonary connection. In 10 patients, primary banding was followed by a subsequent bidirectional cavopulmonary anastomosis. In 15 patients, a bidirectional cavopulmonary anastomosis was the initial step prior to total cavopulmonary connection. In 48 patients, initial construction of a systemic-to-pulmonary arterial shunt was followed by a bidirectional cavopulmonary anastomosis, and finally the total cavopulmonary connection. Preoperative characteristics of the patients, and their division according to stratification of risk, are summarized in Table 2. Note that the mean age of the patients without a previous bidirectional cavopulmonary anastomosis was higher, with a mean of 143 months and a range from 47.5 to 227 months.

In the collective of 84 patients, median age at the total cavopulmonary connection was 37.06 months, with a range from 17.3 to 251.2 months, median height was 95 centimetres, with a range from 68 to 180 centimetres, and median age at bidirectional cavopulmonary connection was 10.6 months, with a range from 3.3 to 116.4 months. The patients were seen routinely at intervals of 3 to 6 months by our paediatric cardiologists in their outpatient clinics. Ventricular function, and atrioventricular valvar regurgitation, were assessed by echocardiography, and a 12-lead electrocardiogram was recorded. When assessing these findings, we divided the patients into those with sinus rhythm, ectopic atrial rhythm, atrioventricular escape rhythm, or pacemaker dependent rhythm.

All patients received heparin in the early postoperative period, with the aim of establishing a partial thromboplastin time of 40 to 60 seconds. Oral anticoagulation was eventually commenced, aiming to achieve an international normalized ratio of 2.0–3.0.

Table 2. Patient characteristics according to stratification of risk.

Patient characteristics	Mean ± SD	p value
Age at completion (months)		
0–1 risk factors	59.71 ± 57.32	
⩾2 risk factors	95.06 ± 67.29	0.190
Height at completion (centimetres)		
0–1 risk factors	102.24 ± 24.79	
⩾2 risk factors	120.94 ± 29.65	0.020
Weight at completion (kilograms)		
0–1 risk factors	18.21 ± 12.54	
⩾2 risk factors	25.34 ± 16.03	0.057
Age at partial anastomosis (months)		
0–1 risk factors	20.71 ± 24.53	
⩾2 risk factors	53.56 ± 47.54	0.016
Interval between partial anastomosis and completion (months)		
0–1 risk factors	26.03 ± 26.64	
⩾2 risk factors	35.62 ± 35.98	0.293

Patients, or parents respectively, were generally taught to use a self-testing apparatus (CoaguCheck®; Roche Diagnostics, Mannheim, Germany).

Operative technique

Regardless of any risk stratification, an equal approach was applied. Total cavolpulmonary completion was performed on cardiopulmonary bypass. A nonringed polytetrafluoroethylene graft (GORE-TEX®; W.L. Gore & Assoc., Flagstaff, AZ) was used in all cases. Only tubes of at least 18 millimetres in diameter were implanted. Unless concomitant intracardiac procedures were required, aortic cross-clamping was not used. For additional intracardiac procedures, antegrade crystalloid cardioplegia, at 40 millilitres per kilogram of body weight, was applied and the patients cooled to 28 degree Celsius.

At weaning from bypass, we aimed at a haemoglobin of approximately 10 grams per decilitre, and after routine modified ultrafiltration at a haemoglobin of about 12 grams per decilitre. Transpulmonary gradients and saturations of oxygen were checked, and transoesophageal echocardiography was always performed. A cellsaver, and Aprotinin at 30,000 International Units per kilogram, were used in all cases.

Statistics

Postoperatively elevated pulmonary arterial pressure higher than 16 millimetres of mercury, prolonged effusions longer than 7 days, and death were considered endpoints in the statistical analysis. The day of surgery was counted as the first day for the period of stay on the intensive care, intubation time, and duration of pleural effusion, respectively. Analysis was performed with the Mann–Whitney test. A value less than 0.05 was required for statistical significance. We used the SPSS software package (Version 10.1, SPSS, Inc., Chicago, Illnois). Data are presented as median, means plus or minus the standard error of the mean, or standard deviations, as indicated.

Results

There were no differences between the groups of patients deemed to have up to 1 or more risk factors in regard to requirement for intubation, pulmonary arterial pressures after extubation, and duration of chest drainage, even though the patients deemed to be at high risk required longer time on cardiopulmonary bypass, and initially reached higher early postoperative pulmonary arterial pressures. Operative and postoperative characteristics are depicted in Table 3.

Median cardiopulmonary bypass time for all patients was 71 minutes, with a range from 30 to

187 minutes, and median cross-clamp time was 40 minutes, with a range from 12 to 94 minutes. In 22 patients, the aorta was cross-clamped. The period of cardiopulmonary bypass was 114.5 plus or minus 39.14 in the patients with intracardiac procedures, and 70.8 plus or minus 37.4 in those without. Procedures included valvuloplasty of the atrioventricular valve in 12, closure of an atrioventricular valve in 7, and replacement in 4.

In our collective the mean duration for effusions requiring drainage was 5.55 plus or minus 5.77 days, with a median of 4 days, and a range from 1 to 45 days. Cardiopulmonary bypass of more than 60 minutes was associated with a longer duration of intubation, and higher incidence of pleural effusions. Of 41 patients who were intubated longer than 48 hours, 36 had a cardiopulmonary bypass time of more than 60 minutes. Of the 20 patients who presented with prolonged effusions and drainage requirement longer than 7 days, 14 had a cardiopulmonary bypass time of more than 60 minutes.

Of the 84 patients, 50 were younger than 48 months at time of completion. Of these, the weight was 11.9 plus or minus 2.4 kilogram. The sizes of the extracardiac tubes ranged from 18 to 22 millimetres. A tube of 18 millimetres was placed in 56 patients.

Reoperations were needed in 3 patients. Of these, all were early. In the initial period, after introduction of the extracardiac approach, one patient required shortening of the tube within two days. Another patient developed a tricuspid insufficiency 6 days after completion, which required valvar replacement.

Table 3. Operative and postoperative characteristics.

Patient characteristics	Mean ± SD	p value
Cardiopulmonary bypass		
0–1 risk factors	76.60 ± 37.56	
⩾2 risk factors	106.56 ± 53.34	0.015
X-clamp		
0–1 risk factors	36.00 ± 17.29	
⩾2 risk factors	49.20 ± 31.85	0.432
Pulmonary arterial pressures 3 hours postoperatively		
0–1 risk factors	15.99 ± 2.37	
⩾2 risk factors	17.19 ± 1.47	0.025
Duration of intubation		
0–1 risk factors	62.28 ± 103.74	
⩾2 risk factors	228.81 ± 747.07*	0.511
Pulmonary arterial pressures after extubation		
0–1 risk factors	12.56 ± 1.68	
⩾2 risk factors	12.63 ± 2.09	0.817
Duration of effusions		
0–1 risk factors	5.69 ± 6.13	
⩾2 risk factors	4.94 ± 3.97	0.650

*One patient was intubated for 3024 hours – without this patient mean duration was 42.47 ± 51.94 (p value 0.278).

In one, complete exchange of the tube was necessary after thrombosis because of a coagulopathic disorder within the tube on the ninth postoperative day. Thrombosis occurred without stenoses at the site of the anostomoses.

No late thrombembolic events were observed. So far, no protein losing enteropathy has evolved. Of the entire cohort, 3 patients died, with one belonging to the group considered to be at high risk. There has been no death in our last 38 patients. In 1 instance, death occurred early. This patient had 1 risk factor, not being in sinus rhythm, and was 205.2 months old at time of completion. The underlying diagnosis was double inlet to a dominant left ventricle with a supracardiac totally anomalous pulmonary venous drainage and pulmonary stenosis. The patient developed tricuspid insufficiency after completion, required valvar replacement, and died subsequently in heart failure. In 2 instances, death occurred late. We identified 1 risk factor in one of these, again lack of sinus rhythm, the patient being 68.4-month-old at time of completion. This patient had tricuspid atresia with a right-sided heart, supracardiac totally anomalous pulmonary venous drainage, pulmonary stenosis and isomerism of the right atrial appendages. The patient died after sepsis, multiorgan failure, and peripheral neuropathy with stroke. The other patient had 3 risk factors, namely impaired ventricular function, atrioventricular valvar regurgitation, and lack of sinus rhythm with a pacemaker, and was 229.0 months old at time of completion. The underlying diagnosis was double inlet to a dominant left ventricle through a common atrioventricular valve in a patient with isomerism of the right atrial appendages and right-sided heart. The patient died after sepis, multiorgan failure, and stroke.

Discussion

The extracardiac approach for completion of the Fontan procedure has many potential advantages, simplifying the procedure of completion by avoidance of aortic cross-clamping and giving a shorter duration of cardiopulmonary bypass.[1,2] Furthermore, it possibly provides a more streamlined hydrodynamic connection,[3,4] and is associated with a decreased frequency of arrythmias at mid-term.[5,6]

The concept of a right-to-left "pop-off" communication, and application of fenestration to the Fontan circulation, was introduced in the late 1980s, initially being used for patients deemed to be at increased risk. The idea was to improve cardiac output, at the expense of oxygenation, reduce the systemic venous pressure, and thus improve survival and morbidity following the Fontan procedure.[7–13] It soon became routine at a number of centres, and remains so today.[14–21]

Despite the reported reduction of morbidity and mortality for the total cavopulmonary connection, we wanted to raise the following issues in the current era. Can the cohorts of patients seen today be compared at all with those undergoing surgery in the late 1980s and early 1990s? And therefore, has the advent of the extracardiac approach, and the early staging, likewise not made a routine fenestration irrelevant?

Routine fenestration is discussed controversially.[22–24] Thompson et al.[23] were among the first to raise this question. They stated that a fenestration not only leads to subnormal systemic arterial oxygenation, but that among potential drawbacks are the need to expose the patient to risk, and the costs of subsequent interventions to close the fenestration.

The long-term risks associated with chronic right-to-left shunting, formation of thrombus, and potential paradoxical embolism, have been the major concerns associated with fenestration, especially in adults.[25] In addition, there is the decrease in exercise tolerance, and consequently quality of life, in patients suffering arterial desaturation subsequent to fenestration.[26–28] To date, as far as we are aware, there is only one prospective and randomized trial assessing the clinical utility of fenestration in patients with standard preoperative risk profiles for creation of the Fontan circulation.[29] Whereas, in this trial, fenestration was shown to be associated with clinical benefits, the authors stated that not every patient undergoing Fontan palliation requires fenestration to achieve a good outcome.

Prolonged pleural effusions represent a clinically important cause of morbidity, occurring in just under half the patients.[30] In our cohort, the mean duration of effusions requiring drainage was 5.55 plus or minus 5.77 days, with a median of 4 days. Despite their relative frequency, the basis for the development of pleural effusions following the Fontan procedure remains unexplained.[31] Likewise, we have not found any relation to our defined preoperative risk factors. Not even early postoperative elevated pulmonary arterial pressures in patients considered to be at high risk lead to prolonged requirements for drainage. McGuirk et al.[20] studied 103 patients, in whom lateral total cavopulmonary connection had been created in 19, and an extracardiac total cavopulmonary connection in the remainder, the operations being performed between 1996 and 2001. Amongst these patients, they created 53 fenestrations. Prolonged pleural drainage was needed in two-fifths. Multivariate analysis identified increased postoperative pulmonary arterial pressure and abnormal pulmonary venous drainage as independent risk factors for the prolonged pleural drainage. Both factors are associated with increased pulmonary vascular impedance and raised pulmonary lymphatic pressure, which can produce interstitial pulmonary oedema.[32] It seems likely that these associated haemodynamic abnormalities may account for the increased risk attributable to both increased pulmonary arterial pressure and abnormal pulmonary venous drainage.

Prolonged periods of cardiopulmonary bypass were significantly associated with increased postoperative volumes of pleural drainage in previous studies. Gupta et al.[21] discussed the exposure to inflammatory sequels. We also found that periods of cardiopulmonary bypass in excess of 60 minutes were associated with a longer duration of intubation, and higher incidence of pleural effusions. Gentles et al.[14] report even an increased risk of early death and failure in combination with prolonged periods of cardiopulmonary bypass. They state that every effort should be made to limit the duration of cardiopulmonary bypass when completing the Fontan circulation. If intracardiac procedures are performed at earlier stages, the placement of the extracardiac tube can usually be performed within 35 to 45 minutes of cardiopulmonary bypass. We then routinely use modified ultrafiltration. This has been reported to significantly reduce the incidence of postoperative pleural and pericardial effusions, requirement of blood products, and hospital stay after the Fontan procedure.[33]

It is important to note that the populations of patients undergoing surgery in the late 1980s and early 1990s were, in general, older than they are today. The range of ages reported by Gentles et al.[14] was from 0.3 to 36 years, with a mean of 6.8 plus or minus 5.9 years. The age in the series of patients reported by Cochrane et al.[16] dropped from a median age of 69.6 months over the period from 1980 to 1987 to 45 months between 1988 and 1995. In our cohort, the mean age was 66.4 months plus or minus 60.1, with a median of 37.1 months, and with 50 patients being younger than 48 months at the time of the total cavopulmonary connection. The mean age of the 43 patients undergoing surgery over the last two years dropped to 57.2. plus or minus 59.96 months, with a median of 28 months, and a range from 17.3 to 251.2 months.

We have shown that conduits of at least 18 millimetres diameter can safely be placed in children weighing 10 kilograms, implying that completion of the cavopulmonary connection can be performed at a young age. Our surgical experience, however, is at variance with other studies where also smaller conduits were implanted.[2,6,34,35] Interestingly, Gupta et al.[21] also reported on a large cohort of 100 consecutive patients, where 33 were treated with a conduit smaller than 18 millimetres in diameter. In half of their patients having conduits smaller than 18 millimetres, they encountered persistent pleural

effusions, compared to only one-quarter of patients having conduits larger than 18 millimetres. Our approach, therefore, might have added to the favourable early and intermediate haemodynamic results. To determine whether the approach also serves to avoid reoperations over the longer term will require further investigations and longer follow-up. It is our belief, nonetheless, that routine fenestration is dispensable in the majority of patients treated with an extracardiac connection.

Limitations of the study

Due to the retrospective character of the study, it was unavoidable that our groups contained patients of different ages. As in many other centres with a big case load, many of our patients were transferred having already undergone previous palliative procedures at other institutions. Hence, the impact on the outcome of a timely initial bidirectional cavopulmonary anastomosis, and consecutive cavopulmonary completion, on the outcome cannot be evaluated clearly from our results. It remains a matter of speculation, therefore, if early staging might have been one of the keys in avoiding the need for fenestration. Until August of 2004, a total of 112 patients have undergone an extracardiac cavopulmonary connection. In only 9 did we create a fenestration at the time of completion. It is not possible to reach any meaningful conclusion when seeking to compare patients with or without fenestrations in our own single centre experience.

Our knowledge of the operations creating the Fontan circulation indicates that they remain palliative procedures for patients with functionally single ventricles. A better understanding of the continuing attrition of patients with the Fontan circulation, and a means to develop ways to predict outcomes in these patients, could help in establishing better criterions for selection, and in preventing side-effects. We continue to believe that the risk factors remain at the core of estimating operative outcome.

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