Fontan completion without surgery

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Abstract

Objective: There are several modifications introduced in the preparation for a subsequent non-surgical transcatheter completion of the Fontan procedure. We report our experience with one type of the modification and the short-term results following its implementation. Methods: During bidirectional cavopulmonary connection (BCPC) an intra-atrial lateral tunnel is additionally created, as intended for a Fontan procedure but fenestrated with a 10—14 mm aperture. The cardiac end of the superior vena cava (SVC) is then patched to maintain the physiology of BCPC. During the interventional transcatheter completion procedure, the SVC patch is perforated using radio-frequency (RF) energy, balloon-dilated, and stented as well. The aperture is closed with a device when required. Paired t-test was used to compare data before and after the Fontan completion. Results: From June 2003 to February 2006, 16 patients (9 boys and 7 girls, mean age 12 months) underwent the surgical procedure described. The mean bypass time was 137 min and the mean ischemic time was 77 min. There were no operative deaths. One patient with bilateral SVC required a take down due to recurrent effusions. Ten months later, nine patients underwent completion (mean age 20 months, mean weight 10.6 kg). The stents were dilated to a mean diameter of 14.4 mm. All except one aperture was closed with a device. The mean fluoroscopy time was 41 min. Oxygen saturation increased from 85 to 94% (p = 0.001). Pulmonary artery pressures remained normal (16 mmHg before and 19 mmHg after, p = 0.12). No patients required mechanical ventilation and none developed pleural effusions or arrhythmias. All were discharged from hospital within 6 days of the Fontan completion. Twenty-two months after Fontan, all were well. Echocardiography revealed no gradients across the stents. Two patients had minor leaks across the aperture. One underwent further stent dilatation a year later. Conclusions: Fontan completion without surgery is suitable in patients with single ventricles with lower mortality and morbidity, avoids multiple surgical interventions while maintaining the staged approach and allows for successive dilatation of the Fontan pathway to accommodate for growth. Keywords: Fontan; Cavopulmonary connection; Transcatheter Intervention; Stenting; Non-surgical Fontan; Hybrid procedure

1. Introduction

The Fontan procedure is considered as the final palliation for the majority of patients with univentricular heart physiology as well as many others who are unsuitable for biventricular physiology. Since its introduction in the early 1970s [1], this procedure has undergone multiple modifications including the total cavopulmonary connection and the utilization of an extra-cardiac conduit either with or without fenestration. Recently, a transcatheter technique for the Fontan completion was introduced [2] and since the initial description, there have been several other modifications of the technique. These modifications have all been relatively successful [3–7], however they encompass several fundamental disadvantages. A recent review on this topic illustrated in detail the anatomical, physiological, and technical considerations of this procedure [8]. We report herewith our experience with a related adaptation of the procedure to emphasize the early outcome, ease of application, advantages, and disadvantages as well as future prospects for similar measures.

2. Materials and methods

From June 2003 to February 2006, 16 patients (9 boys and 7 girls, mean age 12 months, mean weight 7.5 kg) with various forms of functionally univentricular heart disease underwent the bidirectional cavopulmonary connection with a modification outlined below. Clinical information, operative records, echocardiographic reports, catheterization data and outpatient charts were examined. We excluded patients with parameters suitable for primary (one-stage) Fontan operation; age above 2 years, mean pulmonary artery pressures below 16 mmHg, ventricular end-diastolic pressure (EDP) less than 8 mmHg and pulmonary vascular resistance.

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(PVR) below 3 Wood units. They all have normal pulmonary artery distribution and, except in one patient, there was normal pulmonary venous connection. For the surgical preparation, all patients were above 3 months old and weighing more than 5 kg. The completion procedure was undertaken when they are above 10 months of age and weighing more than 9 kg. Following the take down in one patient with bilateral SVC, the procedure was offered only to patients with a single right SVC. Approval from the hospital’s Research Advisory Committee (Internal Review Board) was dutifully obtained. Informed consent from parents prior to both stages of the procedure was appropriately acquired.

2.1. Surgical technique

The surgical procedure is essentially a combination of the hemi-Fontan operation and a lateral tunnel Fontan procedure with a fenestration (aperture) larger than the diameter of the inferior vena cava in the medial wall of the tunnel and patch closure of the mouth of the SVC in the right atrium. The construction of the BCPC is carried out under cardiopulmonary bypass during the cooling phase. Cold blood cardioplegic arrest at moderate hypothermia (32°C) is instituted for the establishment of the lateral tunnel. The medial wall of the tunnel is constructed using two-thirds of the circumference of a Gore-Tex vascular tube (W.L. Gore & Assoc. Flagstaff, Arizona) of 14 or 16 mm diameter. The Gore-Tex wall is fenestrated with a single aperture 10–14 mm in diameter, as high as possible on its anterior aspect. The size of the aperture is guided by the preoperative echocardiographic diameter of the inferior vena cava (IVC) at the level of the diaphragm (10 mm if IVC < 8 mm; 12 mm if IVC 8–10 mm; and 14 mm if IVC > 10 mm). The aperture is made bigger than the IVC diameter as we anticipate the former to narrow in time from possible endothelialization or peel deposition on its edges. Next, the orifice of the superior vena cava at its junction within the right atrium is patched with autologous pericardium. The aperture and the patch are positioned as close together as possible. Fig. 1 illustrates the product of this modification where a blind pouch of the cardiac end of the superior vena cava was created directly opposite the bidirectional cavopulmonary anastomosis. This is to facilitate the placement of the RF catheter in the subsequent interventional stage.

2.2. Percutaneous catheter technique

The catheterization is performed under general anesthesia with intravenous Heparin (100 unit/kg) administered. Under transesophageal echocardiographic guidance (Fig. 2), the aperture in the medial wall of the lateral tunnel is sized. Subsequently, the autologous pericardial patch (Fig. 3) separating the lateral tunnel from the right pulmonary artery is pierced with radio-frequency energy. The communication of the lateral tunnel to the pulmonary artery is dilated with PTCA.

Fig. 1. Aperture creation in the PTFE medial wall of the lateral tunnel. SVC, superior vena cava; IVC, inferior vena cava; PTFE, polytetrafluoroethylene (Gore-Tex); PATCH, autologous pericardial patch closing mouth of SVC in right atrium; APERTURE, large fenestration in medial wall of lateral tunnel.

Fig. 2. Transesophageal echocardiography showing unobstructed flow across aperture in the lateral tunnel. ATRIUM, pulmonary venous atrium; TUNNEL, lateral tunnel; APERTURE, aperture within medial wall of tunnel.

Fig. 3. Angiographic image delineating the pericardial patch on the floor of the pouch opposite the bidirectional cavopulmonary connection. SVC, superior vena cava; RPA, right pulmonary artery; PATCH, autologous pericardial patch; APERTURE, aperture in medial wall of lateral tunnel; IVC, inferior vena cava.
balloon followed by a regular balloon. A stent mounted on a balloon is deployed between the upper part of the tunnel and the pulmonary artery. A tunnel angiogram is carried out and should a significant shunt remain through the aperture, a suitable size Amplatzer ASD occlusion device (Amplatzer, AGA Medical Corporation, Golden Valley, MN, USA) is positioned across the aperture (Fig. 4). All patients were administered Coumadin in conjunction with our institutional policy of full oral anti-coagulation for all patients following the modified Fontan procedures.

2.2.1. Statistical analysis

The results are presented as means (and range) for continuous data and as percentiles for categorical data. Paired Student’s t-test and chi-square test were used to compare the values before and after the Fontan. The statistical software used was SPSS for Windows (SPSS, Chicago, IL, USA).

3. Results

Characteristics of the 16 patients are summarized in Table 1. The mean pulmonary artery pressure was 17.8 mmHg and the mean pulmonary vascular resistance was 1.9 Wood Units. The mean Nakata Index was 240 mm²/m² (range 104—380 mm²/m²). The mean bypass time for the surgical preparation was 137 min and the mean ischemic time was 77 min. Four patients underwent additional procedures; a Damus-Stansel-Kay anastomosis in one, atrioventricular valve repair in two patients and repair of total anomalous pulmonary venous connection with atrioventricular valve repair in one. These average bypass and ischemic times were considerably longer than those reported by Konertz et al. [3] (mean bypass time 97 min and mean cross-clamp time 31 min). There were no operative deaths.

One patient with bilateral SVC required a take down due to recurrent effusions. In this patient, the left SVC was thrombosed and there was stenosis of the central pulmonary artery between the two cavopulmonary connections. At the take down operation, the pulmonary artery was reconstructed by patch angioplasty and a left Blalock-Taussig shunt was added, leaving the right BCPC intact. The atrial baffle was removed. The mean oxygen saturation in the other 15 patients at discharge was 80% (69—92%) and was comparable to those reported by others [3,6].

Ten months later (range 5—16 months), nine patients underwent transcatheter Fontan completion (mean age 20 months, mean weight 10.6 kg). Demographics of these patients are shown in Table 2. The mean diameter of the aperture had decreased from 11.8 mm to 7.4 mm; \( p < 0.001 \) (Fig. 2). The mean fluoroscopy time was 41 min (range 27—81 min). The stents across the RF perforated pericardial...
frequent requirements of palliative procedures such as the Blalock-Taussig shunt, pulmonary artery banding, and the Norwood procedure, obligating a substantial cumulative mortality and morbidity risk from the re-operations. It makes good sense therefore to eliminate one of the surgical stages wherever possible in an attempt to trim down the overall risk. This is feasible following the introduction [2] of the transcatheter interventional completion of the Fontan circuit, initially in animals [5,7] and eventually in human subjects [3,4,6,7]. The method was originally described for use in high-risk Fontan candidates; increased pulmonary pressure, distorted pulmonary arteries, atrioventricular valve regurgitation, long standing arterio-pulmonary shunts, and peripheral pulmonary artery stenosis [2,4]. We had similarly excluded patients with parameters ideal for primary Fontan operation. This is reflected in the high mean pulmonary artery pressures, high ventricular EDP, and the need for atrioventricular valve repairs in three patients. There were also two patients who required concomitant complex procedures, namely Damus-Stanels-Kay anastomosis and repair of total anomalous pulmonary venous drainage. The surgical preparations were carried in relatively young patients (mean age 12 months, mean weight 7.5 kg) and although there have been reports of primary Fontan operations in infants [13,14], we prefer to stage them with a BCPC to preserve excellent ventricular hemodynamics [15]. The modification utilized at our institution has several advantages over the previous adaptations. With the creation of the blind pouch (Fig. 1) directly facing the bidirectional cavopulmonary anastomosis, the interventional catheters are easily guided into the pouch thus facilitating the positioning of the RF wire in the ideal location. This is verified by the short procedure and fluoroscopy time for the interventional procedure, as well as the lack of any major complications such as cardiac perforation. The pouch is also capable of growth thereby diminishing the risk of a stenosis in the Fontan pathway. The suture-line of the autologous pericardial patch facilitates anchoring of the stent. This eliminates the need for insertion of a band during the surgical procedure as have been described by earlier modifications [3,4]. The placement of the whole upper segment of the stent within the pouch avoids having a significant protrusion into the right pulmonary artery. We had established the potential for further dilatation of the stent to accommodate for the growth of the child in one of our patients. The use of a Gore-

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Abbreviations: Aperture surg, diameter of aperture at surgical preparation; Aperture cath, diameter of aperture immediately prior to Fontan completion; Grad, gradient across the aperture; Sat, oxygen saturation; PaPr, pulmonary artery pressure.

* Patient numbers in column 1 refer to the same patient numbers in Table 1, and patients numbered 8 and 11–16 have not undergone completion.
Tex medial patch that directs IVC flow into the pulmonary arteries makes stenting at this site unnecessary. This eliminates the need for a marking band around the IVC, as well as the risk of either entrapping the hepatic veins, or of excluding it from the Fontan flow as have been described elsewhere [6]. The cardiac motion and diaphragmatic oscillations furthermore, render this site an unrelentingly moving target for stenting. We position the aperture high up in the tunnel to avoid the risk of the ASD closure device impinging on the atrioventricular valve/s and interfering with its function. Its anterior position minimizes the potential for obstruction of the pulmonary veins.

We chose to create a large aperture to minimize the risk of restriction to ventricular inflow. The application of multiple fenestrations in the lateral tunnel as described by Konertz et al. [3] and Sidiropoulos et al. [4] may run the risk of gradual closure of the small holes with potential reduction in cardiac output. This could be catastrophic or may necessitate an earlier Fontan completion where the patient is not hemodynamically ready. The size of the aperture is critical and we have chosen 10 mm as its lower limit as we have noticed gradual restriction of the flow across the aperture with time. We utilized a single ASD occlusion device for this aperture, and the incidence of residual shunting in our patients is acceptably low.

The use of a covered stent as has been previously described [5—7], could simultaneously dilate the connection between the right atrium and the right pulmonary artery. This simpler approach could bring about equally satisfying output. This could be catastrophic or may necessitate an earlier Fontan completion where the patient is not hemodynamically ready. The size of the aperture is critical and we have chosen 10 mm as its lower limit as we have noticed gradual restriction of the flow across the aperture with time. We utilized a single ASD occlusion device for this aperture, and the incidence of residual shunting in our patients is acceptably low.

Immediate complications are well-known following surgical Fontan completions, with pleural effusions occurring in the range of 13—54% [9,11,12,16,17]. The interventional Fontan completion reduces the incidence of these complications as had been observed in our experience. The need for ICU care is generally averted. The catheterization process allows for immediate peri-Fontan hemodynamic evaluation that undoubtedly helps in the decision to proceed with the Fontan completion, and the hemodynamics following completion can also be instantly assessed. Additionally, aortopulmonary collaterals, which are commonly present in pre-Fontan patients and has significant influence in the incidence of effusions [10,18,19] can be identified and dealt with during the non-surgical Fontan completion.

One may argue that the post-Fontan complications are not eliminated but brought forward to an earlier stage, during the BCPC. This is reasonable as the modifications hereby described entail longer perfusion and ischemic times. Nonetheless, despite this lengthy procedure we have not noticed any significant untoward consequences in our patients. Based on the 100% survival, it may seem intuitive that a more logical approach would be to perform the BCPC without cardiopulmonary bypass and later Fontan operation also without cardiopulmonary bypass. The 'off-pump' Fontan procedure is currently conducted in patients older (mean age 2—9.8 years) and bigger (mean weight 10.8—27.7 kg) than those in our series [20—22].

Undeniably, these lengthy operative times are of concern to us. We foresee nonetheless a possible reduction of bypass time by avoiding cardiopulmonary bypass during the creation of the BCPC. Shortening of ischemic time could also be accomplished by performing the anastomosis between the cardiac end of the SVC and the inferior wall of the right pulmonary artery without aortic cross-clamp as described by Konstantinov et al. [7].

There is however a disadvantage foreseeable, whereby the Fontan has to be completed once the surgical preparation is done. The aperture would eventually be inadequate for the growing child if the Fontan is not completed and may perhaps warrant a surgical take down of the lateral tunnel. We do not foresee that dilating the aperture in the catheterization laboratory is possible, let alone desirable because the Gore-Tex toughens with time and may harbor thrombus that could be released into the systemic circulation.

Another disadvantage is the need for anticoagulation and its attendant problems of inadequate and often difficult control in the very young age group. Investigators had substituted Coumadin with aspirin after 3—4 months [4], or had administered aspirin immediately after the interventional completion [6]. These may diminish the difficulty of this problem to some extent but we continue to prefer the use of Warfarin, as the thromboembolic burden of this new procedure is still unknown. This is even more so in the event of depressed ventricular function later in life. Undeniably, the long-term effects of the early establishment of the lateral tunnel, the impact of growth and the possibility of further stent dilatation as well as the overall outcome of the patients are yet to be discovered as well. A long-term and close follow-up of these patients are essential but we remain confident that this procedure will be beneficial to a large number of patients destined for a multi-staged palliation to establish a Fontan circulation.

5. Conclusions

Fontan completion without surgery is suitable in patients with single ventricles with lower overall risks. It potentially eliminates a final surgical intervention while maintaining the staged approach toward the Fontan completion, and allows for successive dilatations of the Fontan pathway to accommodate for growth. In addition, there are advantages of faster recovery, lower blood transfusion requirements, and a shorter hospitalization. More procedures and a longer follow-up time are needed to demonstrate whether this modification would eventually be the preferred method to complete the Fontan.

References

Appendix A. Conference discussion

Dr C. Pizarro (Wilkinson, USA): You certainly demonstrated that this strategy is applicable to patients, and I think that this technique has been described by a German author many years ago. I have a couple of questions. One is that when you did the bidirectional Glenn did you actually divide the aygoss vein? Because I think that it has been shown that although dilations have been scheduled in the future in all these patients, it is a fact that every time you take a patient to the cath lab to have further dilations you accomplish maybe 2, maybe 3 mm of increased cross section at most, and over time, the area where the stent has been placed, it sort of gets encased in fibrous tissue which is difficult to dilate. If you have not divided the aygoss vein, you would have an alternative route for the blood from the lower compartment to travel towards the branch pulmonary arteries. So, did you divide, the aygoss vein or was left intact?

Dr Sallehuddin: It is our practice and our policy to divide all aygoss veins during bidirectional caval pulmonary shunt. It is a good point that you bring out in the use of the extra amount of caval tissue from the aygoss vein, but the placement of the stent is actually well below where the aygoss vein is. The stent is positioned around the area of the lower end of the superior vena cava at its junction with the right atrium. At that point I think the aygoss is well away and would not be useful in the future if we have left it behind.

Dr Pizarro: And could you comment regarding the leaks that you have had? Are those leaks that relate largely to a lack of seal across the Amplatter device or is it around it? Why are those leaks present? When you close an atrial septal defect with an Amplatter, although you might see a leak initially, those leaks tend to disappear.

Dr Sallehuddin: Those leaks were all around the area of the Amplatter device. I am not quite sure why they were more significant in this group of patients. Most likely the stent that we have to place is quite close to the aperture and there may be poor apposition of the device with the whole rim of the defect. But we have had no leaks other than around the area of the Amplatter device itself.

Dr G. Stellin (Padova, Italy): In your drawing you showed that the anatomy of the atrial stump of the superior vena cava to the inferior aspect of the pulmonary artery was exactly in front of the superior vena cava anastomosis, and this has been demonstrated to be no good in terms of power loss, vortex formation and flow distribution. Is this really the technique that you are using?

Dr Sallehuddin: I am quite aware of the hemodynamic studies that show that there is some benefit of skewing the anatomy of the stump towards the right pulmonary artery and the SVC towards the left pulmonary artery. In this situation we are still early in our experience and we want to make sure it facilitates the interventionalist to find the patch, to perforate it, and further dilate it. But I think with time and with experience, the same concept of skewing the anatomy of the cava to the pulmonary artery can be done, and with increasing experience I think the interventionalist would also easily find the patch.

Dr Corno: The preservation of the ventricular function is very important in these patients for the long-term results. With your approach you avoid the second operation, but for the first stage you need a period of myocardial ischemia. In your experience the average duration of aortic cross clamping was 67 min. The conventional superior vena cava to pulmonary artery anastomosis can be done without any period of myocardial ischemia as well as the second stage with completion of extracardiac Fontan. Can you comment on the advantages with your approach in this regard?

Dr Sallehuddin: Again, this is an early experience and we have combined groups of patients whom we have subjected to Damus-Kaye-Stansel and repair of TAPVD, and those are the patients that require really a long period of aortic cross-clamp. With experience, as has been shown by Konertz in Hannover, who did almost the same kind of reconstruction within the heart, avoiding the use of cross-clamp for the lower anastomosis between the stent and the pulmonary artery; in his publication he could actually reduce the cross-clamp time down to around 30 min, and, in fact, his average time was 31 min.