The Hemi-Fontan operation: A critical overview

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ABSTRACT

The hemi-Fontan (HF) operation is a staging procedure in the journey towards an ultimate Fontan palliation. Although popular in the Western world, it has found limited application in the developing world. In this review we discuss the indications, techniques, merits, and demerits of this procedure along with its present day role in developing world where there is lack of awareness about this operation.

Keywords: Fontan, single ventricle, superior cavopulmonary anastomosis

INTRODUCTION AND HISTORICAL ASPECTS

As early as the 1940s, it was shown that the right ventricle can be excluded from the circulation, still maintaining an adequate pulmonary blood flow.[1] This concept was utilized in the management of children with a single functioning ventricle for whom there was no palliation available and early ventricular dysfunction and death was inevitable. The Fontan operation was first performed for single ventricle palliation in 1968.[2] Although Fontan *et al.*, described this procedure initially for the treatment of tricuspid atresia, in the years that followed, it was found to be useful for all variants of a structurally or functionally univentricular heart.

STAGING A FONTAN: WHY AND HOW?

In hearts with a functioning single ventricle, the same ventricle handles both the systemic and pulmonary circulations, and therefore, it is volume overloaded. This leads to alterations in ventricular geometry and progressive dilatation leading to diastolic dysfunction and failure. Early offloading of the ventricle leads to preservation of the mass to volume ratio and improves the ventricular performance.[3] Fontan circulation is the final target palliation for such a heart in which the

systemic venous return is directed to the pulmonary circulation without an intervening ventricular chamber. This is attained by either directly connecting the vena cavae to the pulmonary arteries (PAs), or by using the right atrium (RA) as an intervening chamber. But a primary Fontan surgery may not be feasible or desirable in all patients due to unfavorable anatomic and physiologic properties. In order to reach the final Fontan state, many patients will require initial staging procedures to balance both the systemic and pulmonary circulations in order to improve the natural history of the disease and to obtain an anatomy and physiology that is favorable for the future completion Fontan. The size of the PAs and the pulmonary vascular resistance have been shown to be critical determinants of the outcome after a Fontan procedure.[4] The staging procedure to be performed initially is dependent on the basic functional anatomy and the pulmonary vascular resistance. A poorly staged Fontan has a higher chance of subsequent failure. Also some reports have stressed on the need for staging all Fontan operations and have shown that the mortality following a primary Fontan is higher than an adequately and appropriately staged Fontan.[5]

All staging procedures aim to direct part of the systemic venous return to the pulmonary circulation, thereby partially off-loading the ventricle and lead to an improvement in the ventricular function and facilitate suitability for future completion Fontan. The choice of the initial palliative procedure depends on many factors. In a child with low pulmonary blood flow and small PAs, a systemic to pulmonary shunt is performed to increase the pulmonary flow as an initial palliation prior to subsequent procedures. But in a systemic to PA shunt, only volume loads the ventricle and may eventually cause PA distortion that can complicate a future Fontan circuit.

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In patients with increased pulmonary blood flow, a PA band is required to restrict the amount of blood flow to the pulmonary circulation and to prevent the development of irreversible pulmonary vascular disease. Apart from the fact that attaining adequate band tightness is a problem, the band can migrate/erode or distort the PAs. In patients with a univentricular heart and a restrictive bulbo-ventricular foramen, placement of a PA band may lead to gradual subaortic stenosis[6] with progressive ventricular hypertrophy that can make these patients poor Fontan candidates. In many such patients where unfavorable primary anatomy contraindicates a band, Lamberti's modification of the Damus–Kaye–Stansel operation or a Norwood strategy may have to be adopted in preparation for a future Fontan.[7]

Other than these short-term palliative procedures, the journey towards a final univentricular palliation comprises of:

a. Bidirectional cavopulmonary anastomosis with preserved antegrade pulmonary flow,
b. Bidirectional cavopulmonary anastomosis with interruption of antegrade pulmonary flow,
c. Hemi-Fontan (HF) procedure,
d. Primary Fontan operation, and
e. Completion Fontan (following an initial staging procedure).

WHAT IS A HF PROCEDURE?

Norwood and Jacobs first described the HF procedure in 1989.[8] It is a staging procedure devised for single ventricle patients in whom primary Fontan operation is not advisable. Since only part of the systemic venous return is directed to the pulmonary circulation and the ventricle is only partially off-loaded, these children are expected to undergo Fontan completion in the future. Hence, all the steps during this procedure should be performed with the future surgery in mind. The primary operation should essentially have all the features to make the future completion Fontan simpler and safer.

A HF operation differs from a bidirectional Glenn operation (BDG) in many aspects. A BDG involves extensive circumferential mobilization and transection of the superior vena cava (SVC) to create an end to side anastomosis between SVC and right PA (RPA). Since a HF procedure preserves the natural SVC-RA confluence, it provides a more physiological flow of blood to the pulmonary circulation. The essential components of a HF procedure [Figure 1] include:

a. Anastomosis between the SVC-RA confluence and the central and branch PAs to direct the SVC blood to the pulmonary circulation,
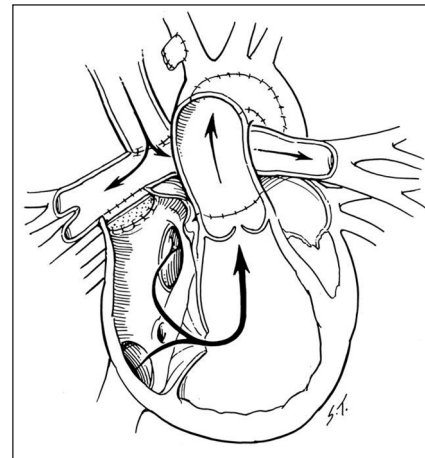b. Homograft patch augmentation of the central and branch PAs,

Figure 1: Cross-sectional view of the HFP. HFP = Hemi-Fontan procedure. Reproduced with permission from: Bando K, *et al.*; Ann Thorac Surg 2000;69:1873-9

c. Interruption of the SVC blood from reaching RA using the homograft dam,
d. Interruption of the antegrade pulmonary blood flow by transecting the main PA and closing its cardiac end and ligating all other sources of pulmonary blood flow, and
e. Additional procedures as atrial septectomy, repair of total anomalous pulmonary venous connection, aortic valve repair, etc., in preparation for future Fontan operation.

The HF procedure directs the SVC blood to pulmonary circulation. The surgery is performed through a median sternotomy incision. Cardiopulmonary bypass (CPB) is established with aortic and right atrial cannulation. The patient is cooled to profound hypothermia and all the extra sources of pulmonary blood flow are ligated. Dissection is carried out to free the SVC, central and branch PAs, and the RA. After cross clamping the aorta, cold blood cardioplegia is delivered through the aortic root and circulation is stopped temporarily. For the performance of a HF, the right atrial cannula is removed and the RA is opened in its superior most portions with the incision extending on to the SVC. The incision in the PA is placed in the central PA and extended to both branches up to the takeoff of the upper lobe branch. At this point, the antegrade pulmonary blood flow is interrupted by transecting the main PA and oversewing the cardiac end. The SVC-RA junction is anastomosed to the right and central PA in a side-to-side fashion. An appropriately sized cryopreserved pulmonary homograft is used to augment the branch PAs as well as to create a dam occluding the flow of blood from the SVC to the RA, after enlarging the size of the SVC-RA junction to the size of the IVC orifice. Once the anastomosis is completed, the heart is deaired, venous cannula is reinserted, and CPB is resumed. Patient is rewarmed, aortic cross clamp is released, and the rest of the operation is completed in the usual manner.

COMPARING HF WITH BDG AS A STAGING PROCEDURE

BDG is a simple procedure that can be performed under short periods of CPB without the need for aortic cross clamping or circulatory arrest. It may even be performed without CPB. But it does not address the problems of PA distortion or stenosis. The SVC has to be circumferentially dissected high up for high cannulation and transection, which increases the chances of phrenic nerve injury. As we oversew the cardiac end of the SVC in a BDG, a future lateral tunnel Fontan may be difficult since dissection of the RA has to be performed to free the cardiac end and that may also entail damage to the artery to the SA node. In contrast, in a HF, the natural SVC-RA confluence is preserved and the anastomosis includes a homograft patch augmentation of the central and branch PAs that corrects any previous PA distortion. Since extensive dissection around the SVC is not required, the chance of phrenic nerve injury and injury to the SA node artery can be avoided. Another advantage is the better fluid dynamics as has been demonstrated in previous computational fluid dynamic models of the univentricular circulation [Figure 2].

ADVANTAGES OF HEMIFONTAN

Better caval offset

Since the natural SVC-RA confluence is preserved in a HF, the blood flowing from the SVC turns posteriorly and enters the pulmonary circulation. The future lateral tunnel Fontan operation directly streams the IVC blood to the PAs, avoiding a competition between two opposing streams of blood. Even though the immediate hemodynamic performance of BDG and a HF are similar,[9] it has been shown that the future lateral tunnel Fontan will have a better performance with less power losses as compared to that following a BDG [Figure 2].

Ease of a future lateral tunnel Fontan operation

In HF, the SVC-RA confluence is enlarged to the size of the IVC opening and anastomosed to the central and branch PAs. Therefore, a future lateral tunnel Fontan completion just requires opening of the RA, excision of the homograft dam occluding the SVC opening, and creation of a lateral tunnel with a patch. No extensive dissection is required and hence procedure time and blood loss can be reduced. An initial HF also permits a percutaneous Fontan completion to be performed by simple perforation of the homograft patch and placement of a stent from IVC towards the PA opening.

Augmentation of the pulmonary arteries

Homograft patch augmentation of the central and branch PAs is an essential part of the HF procedure. This corrects any form of PA distortion or stenosis in the staging procedure itself so that a future completion procedure is performed easily.

Protection of the phrenic nerve

There are less chances of phrenic nerve injury during a HF, as circumferential dissection of the SVC is avoided. Also there are less chances of injury to the artery to SA node, as minimal dissection is needed in the area of the dome of the RA at the time of Fontan completion.

Superior hemodynamics

As a staging procedure, HF offers good hemodynamic performance and can be performed even in early infancy.

LIMITATIONS OF HEMIFONTAN

Despite multiple advantages, the HF has definite problems including:

a. It is technically more difficult to perform as compared to a BDG. Use of a pulmonary homograft, augmentation of the central and branch PAs as well as the anastomosis of the SVC-RA confluence to the PAs needs expertise with a definite learning curve;
b. A future extracardiac Fontan is a difficult option after a HF since the PAs already bear an anastomosis with RA;
c. The suture line through the SVC-RA confluence can damage the sinoatrial nodal artery leading to arrhythmias later if one is not careful at the initial operation; and
d. No large scale prospective data is available to study the results of procedure in detail from the developing world.
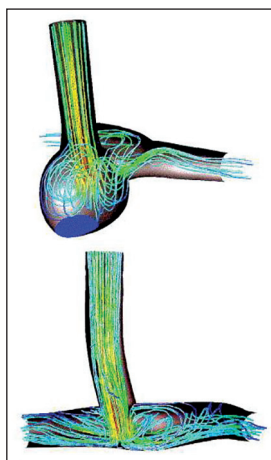

Figure 2: Path lines corresponding to SVC injection for HFP (upper panel) and BDG (lower panel) models. Lines are color coded with reference to velocity magnitude (red for high velocity and blue for low velocity). SVC = Superior vena cava, BDG = Bidirectional Glenn. Reproduced with permission from: Bove EL, *et al*. J Thorac Cardiovasc Surg 2003;126:1040-7

The retrospective data that is available in western world is also based on a heterogeneous patient cohort. A majority of these patients had hypoplastic left heart syndrome (HLHS) as the primary diagnosis and underwent a Stage I Norwood palliation prior to a HF. We are all aware that the patient profile is quite different in the developing world and HLHS is not a very common diagnosis.

Douglas *et al.*,[10] reported their experience of HF on 114 patients with a mean age of 5.4 months after Norwood operation for HLHS. They demonstrated that this procedure can be performed safely and effectively with low operative mortality.(2%) The two independent predictors identified for adverse outcome were poor ventricular systolic function and postoperative PA thrombosis. Younger age and low weight were also determinants of poor outcome. Moreover 95% of the survivors in their series remained good candidates for a later completion Fontan operation. They claim that since 1993 when HF was popularized as a staging procedure, Fontan mortality has been significantly reduced to 2% as against a mortality rate of 21% following BDG as a staging procedure. Fogel *et al.*,[11] studied the systemic venous flow distribution to the branch PAs after completion Fontan operation in patients who underwent HF as an intermediate staging procedure. This study using magnetic resonance imaging showed that the proportion of the total vena caval blood distributed to each branch PA was almost equal. This reinforces the fact that augmentation of the branch and central PAs preserving the natural SVC-RA confluence provides even distribution of pulmonary blood flow and permits better admixture of unsaturated blood in the lungs. Also, as reported by Douville *et al.*,[12] HF was proved to be a safe and well-tolerated procedure even in younger age group and particularly beneficial as a life-saving approach in the event of a take down after a failed Fontan operation.

MODIFICATIONS OF HEMIFONTAN

Partial Fontan operation[13]

The partial Fontan operation[13] is not synonymous with the HF. At best, it can be described as a modification of the HF [Figure 3]. It is being discussed here to clarify the nature of the "partial Fontan" and to discuss its importance in the current era. The SVC-RPA anastomosis is initially performed in an end to side fashion. An incision is placed at the SVC-RA confluence opening into the RA. This opened RA roof with the SVC-RA confluence is anastomosed to the inferior wall of RPA and the central PA creating a blind communication. The wall of PA tends to separate the RA from it. During the future lateral tunnel Fontan, incisions are placed in the anterior aspect of the RPA and the RA. Utilizing both these exposures, the wall of PA is excised and a lateral tunnel is created with a patch to direct the IVC blood to the PA.
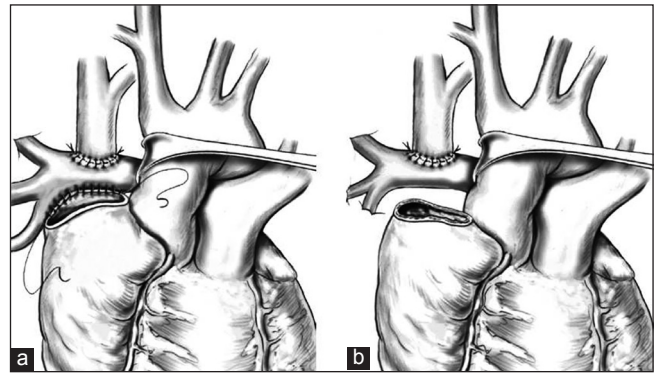

Figure 3: Modified hemi-Fontan operation. (a) The opened roof of the right atrium, including the stump of SVC, is anastomosed in end-to-side fashion to the inferior wall pulmonary artery, creating the blind connection. (b) Bidirectional cavopulmonary anastomosis is centrally located. Incision of medial aspect of cardiac end of SVC is carried horizontally well beyond the junction of SVC and right atrium. IVC = Inferior vena cava, LPA = Left pulmonary artery. Reproduced with permission from: Hraska V. Interact CardioVasc Thorac Surg 2003;2:379-81

A partial Fontan can also be achieved by performing a BDG initially and anastomosing the SVC-RA confluence on the undersurface of RPA (instead of closing as in BDG) and then occluding the anastomosis from within the RA with a patch. However, due to lack of growth potential because of a prosthetic patch, this may lead to anastomotic narrowing and may create difficulties during future completion Fontan.

HEMIFONTAN ON BEATING HEART[14]

The surgery is performed with a median sternotomy and CPB is established with aortic, RA, and innominate vein cannulation. Clamps are applied to the SVC below the SVC-innominate vein junction and above the SVC-RA confluence. SVC is transected in between and the cephalic end is anastomosed to the superior surface of RPA in end to side fashion. The cardiac end of SVC is everted and a patch is sutured inside in order to obstruct the flow and the cardiac end is anastomosed to the undersurface of RPA. The future completion Fontan is facilitated as it requires only excision of the patch and creation of a lateral tunnel to direct the IVC blood to the PA. This operation is thus a form of partial Fontan and not a true HF.

PERCUTANEOUS FONTAN COMPLETION[15]

It is important to add a few lines on the percutaneous Fontan completion [Figure 4] as many centers are increasingly adopting this strategy to avoid a reoperation and CPB. In this approach, the initial surgical procedure consists of a combination of the HF and a lateral tunnel Fontan operation. HF is performed in the standard way. The RA is opened and a prosthetic patch is used to create a tunnel from the IVC to the SVC-RA confluence. An aperture
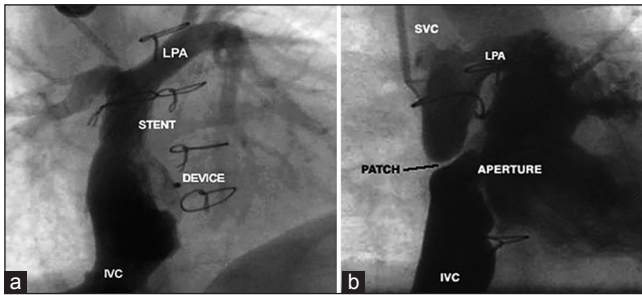
Figure 4: Percutaneous Fontan completion. (a) Angiogram following stent and ASD device placements within the lateral atrial tunnel. ASD = Atrial septal defect, RPA = Right pulmonary artery, STENT = Balloon-mounted bare stent, DEVICE = Amplatzer ASD occlusion device. (b) Angiographic image delineating the pericardial patch on the floor of the pouch opposite to the bidirectional cavopulmonary connection. PATCH = Autologous pericardial patch, APERTURE = Aperture in medial wall of lateral tunnel
Reproduced with permission from: Sallehuddin A, *et al.* Fontan completion without surgery. Eur J Cardiothorac Surg 2007;32:195-201

is made in the superior and medial aspect of the patch that is larger than the opening of the IVC and a patch of autologous pericardium is used to prevent blood from the SVC from reaching the RA. During the Fontan completion, a catheter is passed to the RA through the femoral vein and IVC and the pericardial patch is pierced with radiofrequency energy. The communication of the lateral tunnel to the SVC-RA confluence is enlarged and a stent is placed into it. A suitably sized Amplatzer occluder device is placed to occlude the aperture that was initially created in the superior and medial part of the tunnel at the initial operation. This procedure is claimed to have reduced postoperative morbidity in terms of pleural effusion and reduction in the length of postoperative intensive care. Faster recovery, lower blood transfusion rate, and a shorter hospital stay have been reported as advantages following the percutaneous Fontan completion.

ROLE OF HEMIFONTAN AND ITS MODIFICATIONS IN RESOURCE POOR ENVIRONMENTS

The HF operation was primarily designed as a staging procedure towards a Fontan in younger patients with HLHS who had undergone a previous Norwood Stage I palliation. Often these patients are too small and have PA bifurcation problems that may be quite severe enough owing to the shunt placed at the previous Norwood operation. In such subset of patients, it is undoubtedly superior to a BDG with PA plasty using pericardium as the PAs can be augmented from hilum to hilum, while achieving a good caval offset. Also with the percutaneous Fontan becoming popular, HF is expected to find increasing use.

In India, and the developing world, the HF has not been popular. The reasons are not difficult to understand and include:

a. HLHS is an uncommon substrate for the Fontan operation in the Indian scenario;
b. The patient profile usually consists of tricuspid atresia and various types of double outlet right ventricles and PA bifurcation problems are uncommon;[16,17]
c. The patients are often older and a BDG is a straightforward operation and can often be accomplished without the use of CPB; and
d. Percutaneous Fontan is not yet popular and is not expected to gain widespread acceptance in the near future because of economic reasons unless inexpensive hardware becomes widely available. Also Fontan without CPB has emerged as an inexpensive alternative option.[18]

Given the present scenario, it appears that HF operation, despite its merits is currently not the staging procedure of choice towards an ultimate Fontan, at least in limited resource environments. What the future holds remains to be seen. As the number of patients with HLHS is expected to rise because of better antenatal diagnosis and because of widespread acceptance by the families about the staged univentricular palliation, the HF operation would probably find more widespread acceptance.

CONCLUSION

Reviews of the information available on HF procedure show that this surgical procedure has a definite role in selected circumstances. A HF operation is an attractive option as a staging strategy for single ventricle palliation when the augmentation of the central and branch PAs is desired to address the distorted anatomy. Also HF is considered a superior intermediate staging procedure if a future percutaneous completion Fontan is planned.

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