# Bidirectional inferior vena cava-pulmonary artery shunt: can it be an alternative for older patients presenting single ventricle heart disease in the third world countries?

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**Submitted:** 16 May 2007
**Accepted:** 31 August 2007

Arch Med Sci 2008; 4, 1: 1–6


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## Abstract

The single ventricle cardiac pathologies are commonly treated with total cavopulmonary anastomosis in the current era. The modality is usually performed in two stages and the bidirectional superior vena cava-pulmonary artery shunt constitutes the first stage; after a certain period, the inferior vena cava is connected to the anastomosis. However, especially in relatively older patients, single stage total cavopulmonary anastomosis is performed with various mortality and morbidity rates. This report is a review of an alternative method to the classical order of total cavopulmonary anastomosis, a prior bidirectional inferior vena cava-pulmonary artery shunt as a preparatory operation for total cavopulmonary connection in selected patient groups.

**Key words:** single ventricle, Glenn shunt, bidirectional cavopulmonary connection, Fontan procedure.

## Introduction

Today, single ventricle cardiac pathologies are commonly treated with an initial superior vena cava to pulmonary artery shunt (Glenn shunt) which is followed by a connection of the inferior vena cava to the anastomosis (Fontan procedure) with the aim of decreasing volume overload as well as maintaining ventricular adaptation [1-3]. The main strategy of the surgical planning is based on the blood content of the superior and inferior vena cava depending on the age and body ratios of the children; at younger ages, the superior vena cava is dominant in venous return to the heart; as age increases together with the increasing proportion of the lower half of the body to the upper half, the inferior caval vein is dominant [4].

Diagnosis of patients with single ventricle physiology may be delayed in many countries, especially where primary health care and family medicine concepts are immature; thus the operations might be delayed and performed at older ages. Although total cavopulmonary anastomosis operation in a single surgical session is an alternative treatment for older patients presenting with single ventricle heart disease, increased pulmonary artery flow and pressure following the connection of both vena cavae to the pulmonary artery also increase the risk of operation and mortality and morbidity rates. These postoperative complications are even higher in the developing countries. Hence, many surgeons prefer construction of a Glenn shunt and then plan the Fontan procedure, as the multistage surgical treatment of single ventricle heart patients [1-3, 5]. In the early follow-up period, due to decreased volume of blood return to the heart through the superior vena cava, the expected arterial partial oxygen pressure and the saturation values may not be achieved and the efficacy of the operation decreases in older patients with single ventricle heart disease.

The aim of this article is to evaluate the bidirectional inferior vena cava to pulmonary artery shunt as an alternative to the bidirectional superior vena cava to the pulmonary artery shunt for the first stage palliation of single ventricle heart physiology in specific patient groups.

## Surgical technique

The operation is performed through median sternotomy. Aorta, superior vena cava, inferior vena cava, if it is present persistent left superior vena cava, innominate vein, and pulmonary arteries are prepared. After ligation of the azygos and hemiazygos veins, pressures of pulmonary artery and left atrium are measured. Ratio of pulmonary artery diameter to inferior vena cava should be >1/2. Following administration of 100 U/kg intravenous heparin, an appropriate sized ePTFE graft is anastomosed, with the aid of a partial clamp, to the main pulmonary artery, pulmonary bifurcation, or to the left pulmonary artery, with the aim of leaving the right pulmonary artery untouched for the possible forthcoming superior vena cava anastomosis [6], in an end-to-side fashion with 6/0 polypropylene sutures. Then the graft is clamped at its mid portion. Purse string sutures are placed at the inferior vena cava-right atrium junction and the right atrium and a temporary cavoatrial shunt is placed between the right atrium and inferior vena cava. After clamping the inferior vena cava and right atrium, the inferior vena cava is transected from the inlet of the right atrium. During the clamping period a low dose inotropic agent (i.e. dopamine, 5 μg/kg/min) is initiated, prophylactically, and increased if required. The caudal end of the ePTFE graft is anastomosed to the inferior vena cava and the clamps on the graft and the inferior vena cava are removed [7, 8]. Then the atrial end is oversewn (Figure 1). Temporary shunt cannulae are removed. Pulmonary artery pressure is measured again and a fenestration between the right atrium and the graft either directly or with another small sized ePTFE graft (4-5 mm in diameter) is considered for pressures ≥16 mm Hg.

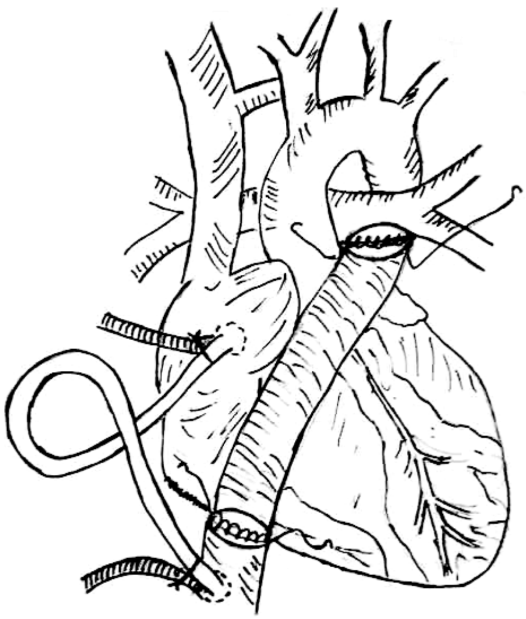

**Figure 1.** Schematic drawing of bidirectional shunt operation between left pulmonary artery and inferior vena cava

In case of systemic venous return anomalies and more than one venous return to the right atrium in the vicinity of the inferior vena cava, a cavoatrial shunt may not be required. In such anomalies, the veins associated with the inferior vena cava must also be connected to the pulmonary artery [9] and eventually direction of all blood flow from the lower half of the body to the pulmonary vascular bed is accomplished.

Another important issue is the maintenance of the antegrade pulmonary flow by transection of the pulmonary artery from the ventricle. This may allow small amounts of low oxygen saturation superior vena cava blood to be directed to the pulmonary vascular bed [10].

The above explained off-pump extra-cardiac procedures may only be possible in cases where there is not a need for an intracardiac intervention; otherwise cardiopulmonary bypass would be inevitable [8]. The size of the atrial septal defect is another important issue and this may be a reason to carry out cardiopulmonary bypass. Sometimes the right atrium may enlarge due to superior vena cava flow. The measures to enlarge the atrial septal defect size in order to decompress the right atrium would aid the enlarged right atrium not to compromise the blood flow in the graft.

## Discussion

The main purpose of the treatment of single ventricle congenital heart pathologies in which biventricular repair could not be achieved is to use maximal power of the ventricle to send all received blood to the aorta. Furthermore, the direction of blood to the lungs without an additional volume load to the ventricle is mandatory.

From that point of view, nowadays, single ventricle pathologies with well developed pulmonary arteries are treated with superior vena cava to pulmonary artery anastomosis in the first stage and then conduction of the inferior vena cava to the anastomosis when the child reaches the desired age, size and weight [1-3]. In this respect, ideally bidirectional Glenn shunt operation is performed on children from the ages of 3-6 months even though in the literature successful applications in younger children are reported [5]. The decrease in the pulmonary vascular resistance and superior vena cava carrying 40-50% of whole body venous return in the early period of life [4] provides excellent palliation for these patients while impeding the progress of cyanosis.

However, with the growth of the child and decrease in blood from the superior vena cava as a consequence the Glenn shunt will result in a gradual decline in the exercise capacity and cyanosis may become apparent again. At this stage, the Fontan procedure is fashioned in order to deliver more blood to the lungs, to decrease cyanosis, and free the single ventricle to deliver all its blood to the aorta. This procedure is best carried out before the child is 4 years of age [11-13] in order to merge the inferior vena cava blood to the pulmonary bed during the time when the inferior vena cava begins to dominate in the systemic venous return [4]; thus the last stage of single ventricle palliation is achieved to provide continuity of life.

Even though the optimal treatment of single ventricle heart patients is Glenn shunt in the early phase and followed by the consequent Fontan procedure [1-3] as described above, due to various reasons unfortunately patients sometimes are not admitted to the clinic at the proper time and under ideal conditions. Many patients may reach older age without any palliation and may be detected with different symptoms [14], especially in third world countries. In these cases, the preliminary bidirectional Glenn shunt and the consequent Fontan procedure usually are not sufficient to decrease cyanosis and improve exercise capacity.

Various alternative treatment modalities may be predicted to increase the pulmonary blood flow in the cases where the child has reached an older age without any palliation. The two commonly applied are: bidirectional Glenn shunt together with Fontan procedure at the same session, or addition of systemic to pulmonary artery shunt to the bidirectional Glenn shunt.

Total cavopulmonary connection at one single stage and several modifications of this application have various risks involved. The most important and serious one is death. As a consequence of being exposed to long-term hypoxia and due to not having any palliative procedures, diminished performance of the heart as well as the body may not tolerate such intervention. Another obstacle to single stage total cavopulmonary anastomosis is a possible drainage difficulty of superior and inferior vena cavae flow because of increased pulmonary vascular resistance which may result from long-term cyanosis [15, 16]. It is a well known fact that the best results are obtained in patients considered for total cavopulmonary anastomosis who meet the Choussat criteria (*Appendix*) [17] and unfortunately these criteria are hardly achieved in older patients with single ventricle heart without preparatory palliative procedures.

Systemic-pulmonary artery shunt in addition to bidirectional Glenn shunt can be applied as another option in the treatment of such patients, as mentioned above. However, the major disadvantage of this modification is early rise in pulmonary vascular resistance in the short run, and this increases the risk of the Fontan procedure. Also, additional volume load due to systemic-pulmonary artery shunt would load the single ventricle and compromise the ventricular functions [18, 19].

When the above patient categories and designed treatment modalities are taken into consideration in the treatment of advanced age single ventricle patients who have not undergone any palliative procedures, alternative treatment options, that would increase the pulmonary blood flow, decrease cyanosis and decrease ventricular workload and mortality as well as mediate drainage of both vena cavae to the pulmonary bed as the last stage, need to be created. This is especially important for physicians receiving advanced age patients with single ventricle hearts in the developing countries.

We proposed the bidirectional inferior vena cava-pulmonary artery shunt first and then addition of the superior vena cava to this anastomosis as the next step, controversial compared to the classical Glenn shunt and its complimentary Fontan procedure. Review of the literature on this subject reveals only a few manuscripts about application of a bidirectional inferior vena cava-pulmonary artery shunt as the first stage palliation treatment of single ventricles [20-22]. Only two cases are described in the most detailed article. Mace et al. describe two cases in which the patients had undergone palliative procedures at younger ages and were in ideal conditions for total cavopulmonary connection at the time of presentation, but increased volume load to the systemic ventricle and distorted pulmonary artery bed were considered as contraindications for total cavopulmonary anastomosis, and bidirectional inferior vena cava-pulmonary artery shunt was therefore performed [20].

Especially in countries where primary health care and family medicine concepts are not satisfactory, for advanced age single ventricle patients surgeons are urged to search for alternative surgical treatment modalities. The described bidirectional inferior vena cava-pulmonary artery shunt as an alternative to total cavopulmonary anastomosis could provide better palliation by reducing the degree of cyanosis and decreasing the systemic ventricle work load, according to the body blood volume haemodynamics. When compared with the superior vena cava, the inferior vena cava carries more than 60% of the total venous return of the body starting from the early ages of life [4]. Also, the operative risk of Fontan procedure as a redo operation following the Glenn shunt could be solved with this method at the first stage and addition of the superior vena cava to that anastomosis could be applied with lower operative risks in the later stages. Moreover, the probability of damage to the sinus node will decrease. As experimentally proven, inferior vena cava pressure is lower when the inferior vena cava alone is connected to the pulmonary artery when compared with total cavopulmonary anastomosis. And this may prevent heart failure, heart failure induced hepatic failure and complications related to elevated renin-angiotensin system in the postoperative period. Absence of the superior vena cava in the pulmonary artery anastomosis will prevent competition of the two venous systems and facilitate drainage of the inferior vena cava to the pulmonary artery. In patients with relatively smaller sized pulmonary arteries, inferior vena cava blood will grow pulmonary arteries better than superior vena cava shunt. The oxygen saturation of the inferior vena cava is higher than the superior vena cava and it seems that it may provide lower oxygenation for the body. However, it is thought that the higher blood amount of the inferior vena cava overcomes the saturation difference. Another advantage of bidirectional inferior vena cava to pulmonary artery shunt is that the blood coming from the lower half of the body is increased during exercise, while there is no change in blood flow from the superior vena cava [20]. Although it has not been proven, theoretically it is believed that there is a hepatic factor that prevents arteriovenous fistula formation in the lungs if it could reach the pulmonary circulation [23]. Bidirectional inferior vena cava-pulmonary artery shunt will provide access of hepatic factor to the lungs. On the other hand, it is very well known that arteriovenous fistulae usually develop in patients, following Glenn shunt procedure, if the antegrade pulmonary artery flow is totally ceased. Risk of venous collateral formation between superior and inferior vena cava after Glenn shunt is also present consequently after this operation. Therefore, systemic venous anomalies should be investigated prior to operation and azygos and hemiazygos veins should be ligated during surgery. Even though a small rise in inferior vena cava pressure will occur following bidirectional inferior vena cava-pulmonary artery shunt, this pressure rise will be much lower when compared with total cavopulmonary anastomosis. Furthermore, the superior vena cava blood return is not compromised and pressure in the vein does not increase and thoracic duct drainage will not be deteriorated. All of these prevent the likelihood of pleural effusion formation or lead to lessened effusion when compared to the total cavopulmonary anastomosis. As frequently seen after total cavopulmonary anastomosis, due to the rise in superior vena cava pressure leading to deterioration in thoracic duct drainage and due to the rise in inferior vena cava pressure leading to increased lymphatic fluid production, ascites formation will also be decreased or be prevented by bidirectional inferior vena cava-pulmonary artery shunt only [20]. From the point of view of surgical technique, the pulmonary and systemic side effects of cardiopulmonary bypass in cyanotic patients can be impeded if the procedure can be completed in experienced hands without cardiopulmonary bypass. This will also lower the risks in the postoperative period [7, 9, 24]. However, the operation can be performed with cardiopulmonary bypass if the cardiopulmonary bypass is inevitable and short-term bypass should be performed without hesitation. Moreover, technically, anastomosis of the graft between the inferior vena cava and main pulmonary artery will facilitate the forthcoming superior vena cava connection if the anastomosis is performed to the main pulmonary artery, pulmonary artery bifurcation or the left pulmonary artery [8]. If the antegrade pulmonary artery flow is not disrupted, this may help the low saturated superior vena cava flow to reach the pulmonary bed in small amounts [10].

In conclusion, especially in late presenting single ventricle patients, there are various risks of single stage total cavopulmonary artery anastomosis, and the postoperative mortality and morbidity can be even higher in developing countries. Therefore, we believe that instead of classical bidirectional Glenn shunt and consequent Fontan procedure, and in order to avoid the risks of single stage total cavopulmonary anastomosis for the palliation of older children presenting with single ventricle heart physiology, bidirectional inferior vena cava-pulmonary artery shunt seems to be an attractive alternative option.

## Acknowledgments

The above explained surgical concept and review of the literature do not represent our experience

since we have not performed any bidirectional inferior vena cava-pulmonary artery shunt operations at our institution. However, we frequently receive advanced age single ventricle patients who have undergone bidirectional Glenn shunt operation rather than single stage total cavopulmonary connection, generally due to the discouraging results of this procedure. These operated patients usually come to the clinic in the early follow-up period with non-disappearing symptoms and we consider the Fontan procedure or various alternatives. Thus, this review article is rather an "opinion" in the light of the review of the literature which we believe may guide physicians in developing countries when they deal with older patients presenting with single ventricle heart physiology.

We would like to thank Omer Ali Sayin, MD for his drawings for the manuscript. Also, we would like to thank Ms. Toni Spring for English language revision.

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## Appendix

### Choussat criteria

1. Age range 4-16 years
2. Sinus rhythm
3. Low pulmonary artery pressure (<15 mm Hg)
4. Low pulmonary vascular resistance (<4 u.m$^2$)
5. Normal venous return
6. Normal right atrial size
7. Well preserved systemic ventricular function (ejection fraction >60%, end-diastolic pressure <12 mm Hg)
8. Appropriate pulmonary artery size
9. No atrioventricular valve regurgitation
10. No distortion in pulmonary arteries or construction before shunt

**Choussat criteria and operations**

| Risk factors | Low | Moderate | High |
|---|---|---|---|
| Main pulmonary artery pressure (mm Hg) | <15 | 15-20 | >20 |
| Pulmonary vascular resistance (woods units) | <2 | 2-3 | >3 |
| Transpulmonary gradient (mm Hg) | <7 | 7-12 | >12 |
| Ejection fraction (%) | >60 | 45-60 | <45 |
| End-diastolic ventricular pressure (mm Hg) | <6 | 6-12 | >12 |
| Outflow tract gradient (mm Hg) | <30 | 30-50 | >50 |
| Atrioventricular valve regurgitation | mild | moderate | severe |
| Operation | F or HF | F or HF | Glenn |

*F – Fontan, HF – Hemi-Fontan*