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## Fontan Surgery: Experience of One Cardiovascular Center

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### 1. Introduction

In order to establish a normal, in-series circulation physiologically, very different from the circulation in parallel which the children are born with single ventricle, doctors Fontan and Baudet (2) and Kreutzer (3) concurrently developed surgical treatment of patients with tricuspid atresia to achieve a passive flow through the pulmonary vascular bed (1).

Management strategies for patients with functional single ventricle required a staged group of procedures where the ultimate goal is to have a single ventricle with a working pressure and volume close to normal as well as normal systemic oxygen saturation (1). It is known that both vascular development and lung maturation are essential for achieving benefits of cavopulmonary connection; the time of surgery has been defined arbitrarily and even more time to transition between partial to total cavopulmonary connection (1).

Single ventricle or univentricular heart anatomically or physiologically characterized by:

- Both atrioventricular valves attached to a single systemic ventricular chamber
- Severe stenosis or atresia of the atrioventricular valves
- There is no separation between the ventricles
- One of the ventricles is hypoplastic or absent

The essential characteristics of hypoplastic left heart syndrome are:

1. Stenosis or atresia of the mitral valve
2. Underdeveloped severely hypoplastic left ventricular
3. Stenosis or aortic valve atresia
4. Small aortic root
5. Ductal dependent for systemic blood flow

Thus, patients with these syndromes will have a parallel circulation in which the systemic and pulmonary circulations will be supplied by "mixed" blood.

The following are the physiological characteristics of hypoplastic left heart syndrome:

- Non-functional left ventricle
- Pulmonary venous return directed to the right atrium through a patent foramen ovale, atrial septal defect or rarely, total anomalous pulmonary venous drainage
- Mixed systemic and pulmonary venous return in right atrium
- The right ventricle supplies the systemic and pulmonary circulation in parallel
- Retrograde blood flow from the ductus arteriosus to the coronary arteries
- In these patients, ductal closure would result an inadequate systemic perfusion and metabolic acidosis with progressive coronary ischemia, and death.

In these patients one treatment option is repair in three stages, as follows:

*The first stage repair* (Norwood operation: Classic or Sano) seeks to:

- Provide systemic circulation: the right ventricle is used to support the systemic circulation
- Ensure non-restrictive egress of the pulmonary venous return to "bypass" the left ventricle, the atrial septal defect is enlarged or created in the absence of (in case it did not exist)
- Create an outflow tract obstruction-free system: the aorta is reconstructed
- Provide a controlled pulmonary blood supply, creating a shunt between the systemic and pulmonary circulations

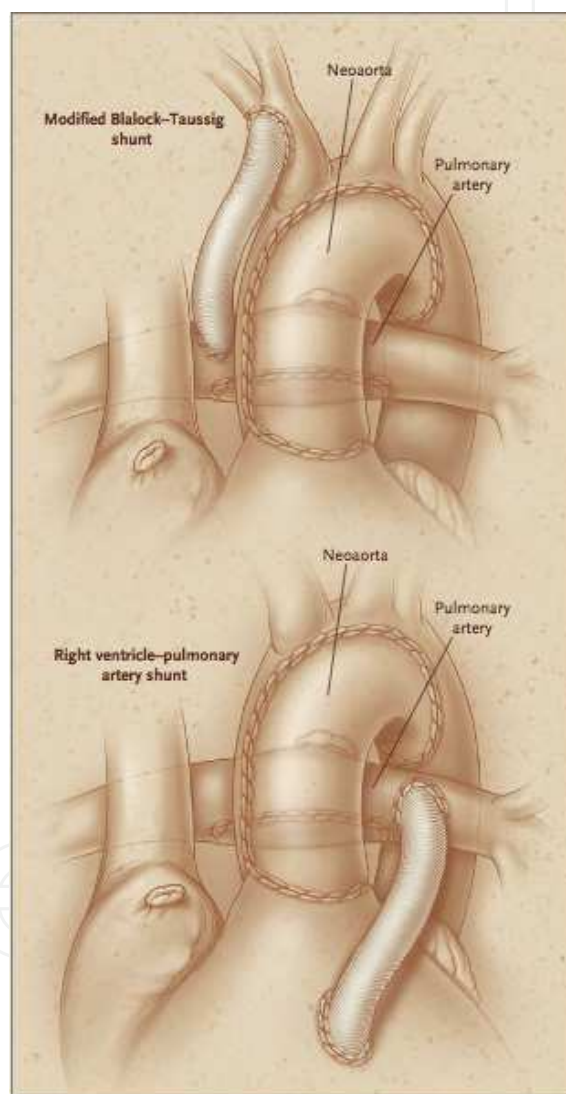


Fig. 1. First Stage: The Norwood procedure with a Modified Blalock-Taussig shunt or a right ventricle – pulmonary artery shunt (Sano)

*The second stage repair* (Glenn operation), which Takes place 6 to 8 months after-the Norwood is looking for:

anastomosing the superior cava to the pulmonary artery, thus direct systemic venous return to the pulmonary artery, begin to create a circulation in series. Fig 2.

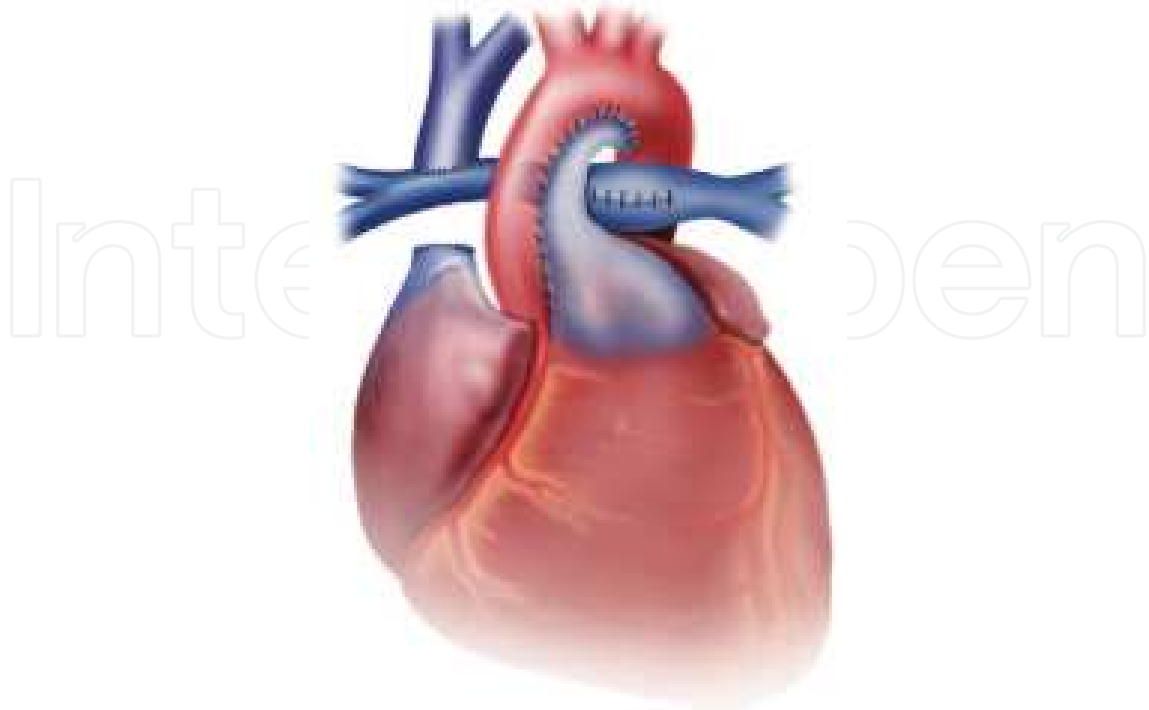


Fig. 2. Second Stage: BT shunt has been replaced with bidirectional Glenn procedure.

*The third stage repair (Fontan operation), which is performed in children 12 kg or 4 years old, looking for:*

anastomosing the inferior cava vein to the pulmonary artery and thus reduces the volume load of the single ventricle and complete the creation of a series circulation (4,5,6). Fig. 3.

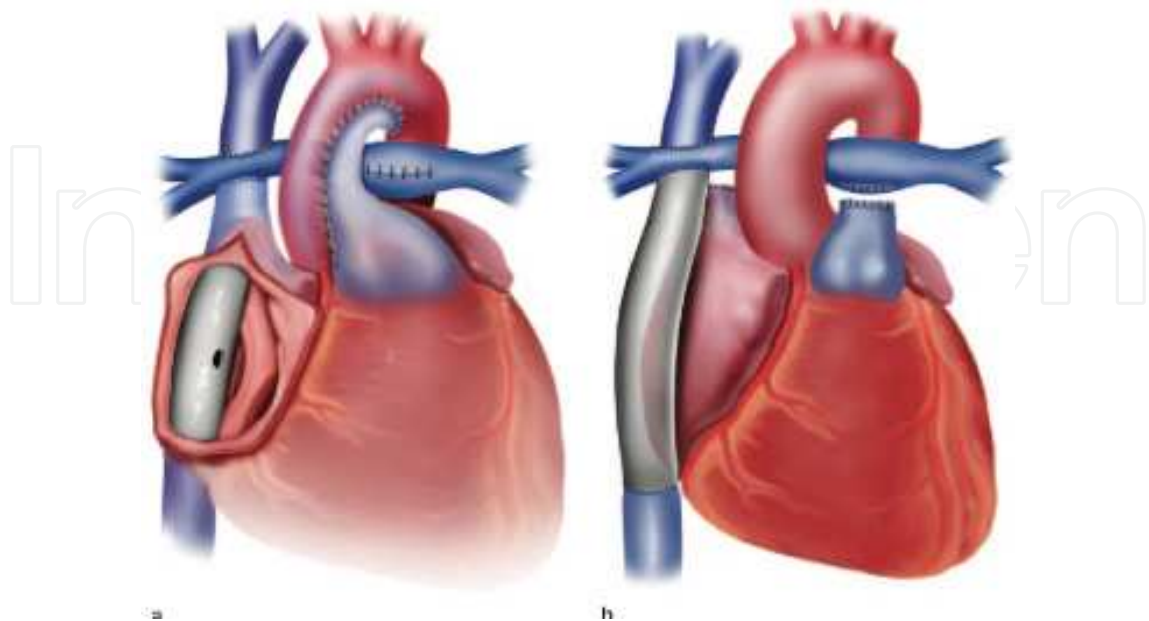


Fig. 3. Third Stage: (a) Fenestrated Fontan with lateral tunnel or (b) Fontan extra-cardiac conduit.

## 2. Surgical technique

All patients were operated through a median sternotomy with extracorporeal circulation and moderate hypothermia. We used two techniques of total cavopulmonary anastomosis, according to anatomical characteristics of each patient. The technique I corresponds to the construction of an intracardiac lateral tunnel constructed with a patch of polytetrafluoroethylene (PTFE) sutured to the lateral aspect of the right atrium, thereby building a tunnel that diverts intracardiac blood from the inferior cava to the right pulmonary artery, thus completing total cavo-pulmonary connection. For these patients we routinely use aortic clamping and cardioplegia with blood. The technique II is interposition of a PTFE tube between the transected inferior cava and pulmonary artery, ipsilateral to the inferior cava vein, also completing total cavo-pulmonary connection. This is known as extracardiac Fontan. Use of aortic clamping and cardioplegia is optional according to surgeon's preference. Both techniques are performed with almost routine fenestration of 4 to 5 mm. In the technique I, a circular punch incision in the PTFE patch is made, so to communicate intracardiac lateral tunnel with atrial mass receiving the pulmonary venous return. In technique II, the fenestration is created by a similar punch incision in the lateral PTFE inner tube and a similar incision on the lateral aspect of the right atrium, proceeding to be anastomosed both holes as Luther-lateral (7,8).

## 3. Type of study

Analytical study of cross-sectional, cohort analysis.

## 4. Population and sample

We included all patients with univentricular hearts who underwent Fontan operation. We reviewed the records of the patients included in the univentricular heart protocol and database service Congenital Cardiac Surgery Cardiovascular Clinic of Santa Maria de Medellin (Colombia), identifying all patients undergoing the Fontan operation with technique total cavopulmonary anastomosis between 1994 and 2010.

We reviewed the medical records, operative reports and echocardiograms.

**Monitoring.** All patients who survived the Fontan surgery underwent clinical and echocardiographic follow-up. We also obtained information about procedures performed after surgery, medication use and functional ability (classified according to New York Heart Association guidelines), interviewing parents and cardiologists caring for for each patient.

## 5. Results

Management strategies in cases of functional single ventricle have come to a group of procedures where the goal is to obtain a ventricular pressure and volume close to normal. Our study demonstrated our experience in the management protocol of univentricular patients in total cavopulmonary connection (Fontan operation). As in other centers, the most commonly used technique has been the fenestrated, extra-cardiac Fontan, which in our study represent 58.9% of the total sample (30 patients) - Table No.1. The hospital stay was 13.6 days on average.

	Frecuency	Porcentage	Cumulative Percentage
Fenestrated Atriopulmonary	2	3.6	3.6
Atriopulmonary not fenestrated	1	1.8	5.4
Fenestrated Extracardiac	30	53.6	58.9
Not Fenestrated extracardiac	16	28.6	87.5
Fenestrated intracardiac	4	7.1	94.6
Not fenestrated intracardiac	3	5.4	100.0
Total	56	100,0	

Table 1. Fontan type

	Frecuency	Percentage	Cumulative Percentage
No	48	85.7	85,7
Yes	8	14.3	100,0
Total	56	100.0	

Table 2. Death

The number of patients who failed was a total of 8; the cause in 4 of them (50% of total deaths) was low output syndrome which was present in the first 72 hours postoperatively, and the remaining 4 patients died after 72 hours after Fontan. These patients had a progressive deterioration associated with symptoms and signs of heart failure, protein-losing enteropathy, persistent chylothorax, affecting directly their functional status and no response to medical management. These data are shown in Table No. 3.

	Frecuency	Percentage	Cumulative Percentage
	48	85.7	85.7
Low Output	4	7.1	92.9
Fontan failed	4	7.1	100.0
Total	56	100.0	

Table 3. Cause of death

The end result is shown in Table No. 4, where 47 of them (84%) were discharged and continued in a functional stage I 9n 82.1% of them (Table No. 5). One patient (1.8%) sample was transferred from the Cardiovascular Clinic to another facility outside the country.

	Frecuency	Percentage	Cumulative Percentage
Discharge	47	84	84
Death	8	14.2	14.2
Transfer	1	1.8	100.0
Total	56	100.0	

Table 4. Cause of discharge

	Frecuency	Percentage	Cumulative Percentage
I	48	85.7	96.4
II	1	1.8	98.2
not evaluated	7	12.5	100.0
Total	56	100.0	

Table 5. Current functional class

Based on this study we conclude that the Fontan operation is a safe with a mortality rate comparable to that reported in previously published large series (Our series 14.3%), which is shown in the table No. 2. The Mayo Clinic experience, shows a overall mortality after Fontan, of 16%, but Many factors may have contributed to decreased early mortality after Fontan. Improved patient selection, younger age at time of operation, refinements in surgical techniques and postoperative management may all have had important roles.

## 6. Discussion

The evolution in the medium and long term outcome of patients operated with the original technique described by Fontan in the 70s demonstrated problems with the incorporation of all the systemic venous atrium in the circuit, characterized primarily by supraventricular arrhythmias and formation of thrombi (9,10,11). This is explained by atrial dilatation and inefficient circulation through this atrium which progressively becomes larger. But, changes in surgical techniques have been made to minimize these problems and improve long-term outcome (9).

At present there are different techniques for completion of the total cavopulmonary diversion: the intracardiac lateral tunnel (technique I) and extracardiac conduit (Technician II), which are the most widely used worldwide and are just two ways that we have employed. We have most often used the intracardiac lateral tunnel and extracardiac conduit including for those patients with abnormal systemic venous return and/or lung disease, which is usually seen in patients with heterotaxy syndrome. Regarding the use or non use of fenestration, this has also been the subject of much discussion, there are groups of surgeons that use the fenestration routinely and others who use it selectively (12,13). We are using this routinely in all our patients because pulmonary vascular resistance is a dynamic phenomenon and therefore it is not always easy to predict its behavior during the postoperative period. It seems safer for the patient having a fenestration that allows you to maintain adequate cardiac output during periods of high pulmonary vascular resistance, plus helps reduce the incidence of prolonged pleural effusions, resulting in decreased hospital length of stay (12,13)

With the passage of time have defined a number of criteria considered important for success in the performance of a Fontan-type surgery. These have included age, single ventricle morphology, anatomy of the pulmonary arteries, the atrioventricular valve function among others (14,15).

Kirklin and colleagues (15) reported 102 patients who had Fontan between 1975 and 1985. They found that age less than 4 years was a risk factor for mortality. Subsequently, the Children's Hospital Boston reviewed 500 patients between 1973 and 1991 and found similar results in relation to age (16). As for the type of functional single ventricle, these techniques

have been implemented for patients classically diagnosed with tricuspid atresia, but now with the increased survival of patients diagnosed with hypoplastic left heart syndrome, the question arises about the reduced ability of right ventricle to be able in time to support adequately the work of a single ventricle physiology to complete the process to the Fontan (3,17,18). This perception has not been recently supported (19,20). Mosca and colleagues show their results in 100 patients with Fontan performed between 1992 and 1998. They found no significant difference in the outcome compared to other types of single ventricle (21). Pizarro subsequently concluded that the Fontan can be performed safely in patients with SHIV making some modifications in surgical technique according to ventricular morphology, the mass ratio - volume and hemodynamic parameters, further suggests that Fontan surgery can be successful at earlier ages avoiding long exposure to hypoxia and risk of paradoxical embolism (1).

Our results regarding operative mortality and mid-term survival are comparable to those reported by other groups. In these studies, the mortality rate varied between 0 and 27% with an average of 10.5% and the 5-year survival varied between 81% and 93%, with an average of 87.5% (9,10,22, 23).

In relation to our surgical protocol in stages, 73.9% of patients had a bidirectional Glenn operation around 6 months of age, preparatory for the Fontan operation. The advantages of this strategy have been previously described (24,25,26).

Regarding the use of anticoagulation, we decided to keep our patients with oral anticoagulation for about 6 months with the objective of preventing thrombosis at the site of fenestration, after this period of time, we defined the need to close the fenestration.

This study determined that the average hospital stay was 13.9 days.

## 7. Conclusions

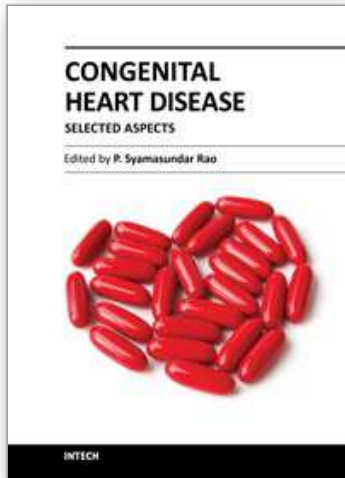
Management strategies in cases of functional single ventricle have come to a group of procedures where the goal is to obtain a ventricular pressure and volume close to normal. This analytical cross-sectional, cohort analysis is meant to show expertise in the management protocol of univentricular patients by total cavopulmonary connection (Fontan operation.) in Cardiovascular Clinic Santa Maria in the city of Medellin. Based on this study we conclude that the Fontan operation is safe with a mortality rate comparable to previously published large series (14.3%); the results are independent of the type of ventricle and the hospital length of stay is short (average hospital stay of 13.9 days). Postoperatively, is that over 90% of patients were in functional class I - II.

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## **Congenital Heart Disease - Selected Aspects**

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There are significant advances in the understanding of the molecular mechanisms of cardiac development and the etiology of congenital heart disease (CHD). However, these have not yet evolved to such a degree so as to be useful in preventing CHD at this time. Developments such as early detection of the neonates with serious heart disease and their rapid transport to tertiary care centers, availability of highly sensitive noninvasive diagnostic tools, advances in neonatal care and anesthesia, progress in transcatheter interventional procedures and extension of complicated surgical procedures to the neonate and infant have advanced to such a degree that almost all congenital cardiac defects can be diagnosed and "corrected". Treatment of the majority of acyanotic and simpler cyanotic heart defects with currently available transcatheter and surgical techniques is feasible, effective and safe. The application of staged total cavo-pulmonary connection (Fontan) has markedly improved the long-term outlook of children who have one functioning ventricle. This book, I hope, will serve as a rich source of information to the physician caring for infants, children and adults with CHD which may help them provide optimal care for their patients.

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