# Risk Factors Affecting the Outcome of the Extra-Cardiac Fontan Operation

**Thesis**

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# ABSTRACT

**Objective:** The purpose of this study is to analyze our experience at Kasr Al Aini Hospital with the Fontan procedure as an eventual palliative procedure for functional single ventricle hearts, focusing primarily on the factors affecting early outcome of this procedure.

**Methods:** From March 2005 till March 2009, 30 patients with functional single ventricle underwent palliation using the Fontan operation. As the study progressed, there was a shift of technique from performing the operation fromon pump to off pump technique.

**Results:** There was no intra or postoperative mortality. Follow up was for a short early period of 30 days postoperative. Outcome as the duration and volume of pleural effusion, and ICU stay were investigated as well their correlation with technique used and preoperative pulmonary artery pressure.

**Conclusion:** The Fontan operation is a safe and effective palliation as an end stage solution to Univentricular hearts. Advances in techniques and progression of learning curves have considerably improved the success rate of this operation. Preoperative mean pulmonary artery pressure remains the most important risk factor affecting outcome. More over the offpump technique has emerged as a safe and effective alternative with improved outcome, shorter ICU stay and shorter duration of pleural effusion

**Keywords:** Fontan, Univentricular heart palliation

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# Contents

Pages

# List of Tables

Page

# List of Figures

**Pages**

Page

**Page**

# INTRODUCTION

## NOMENCLATURE AND ANATOMY

Among pathologists and cardiac anatomists, the debate as to what truly constitutes a single ventricular heart, raged long before the era of surgical therapy for these complicated cardiac anomalies. The debate continues today, and no single system of nomenclature has yet been accepted by all practitioners in the field. There is no escaping the fact that for those of us who treat patients with such anomalies, it is absolutely necessary that we be able to communicate using a nomenclature made up of a precise, and preferably simple terminology ***(Jacobs and Mayer, 2000).***

## Current Classification

For the goal of universality and relative simplicity. The STS congenital heart surgery database committee adopted a descriptive language that incorporates some elements of both the segmental classification and the sequential chamber localization approach, and combined with other familiar systems of classifications as for example those which have evolved with respect of tricuspid atresia.

***Single ventricle heart would encompass:***

1-Hearts with double inlet atrioventricular connection [both double inlet right ventricle (DIRV) and double inlet left ventricle (DILV)].
2-Hearts with absence of one atrioventricular connection (mitral atresia and tricuspid atresia).
3-Hearts with a common atrioventricular valve and only one complete well developed ventricle (unbalanced A-V canal).
4-Hearts with only one fully well developed ventricle and heterotaxia syndrome (single ventricle heterotaxia syndrome).
5-And finally other rare forms of univentricular hearts that do not fit in one of the specific major categories.

Despite the recognition that hypoplastic left heart syndrome is a common form of univentricular heart with a single or dominant ventricle of right ventricular morphology, the current nomenclature and database proposal excluded the hypoplastic left heart syndrome from this group.

Also, it is recognized that a considerable variety of other structural cardiac malformations such as pulmonary atresia with intact ventricular septum, biventricular hearts with straddling atrioventricular valves and some complex forms at double outlet right ventricle may be best managed in a fashion similar to that which is used to treat univentricular hearts ***(Jacobs and Mayer, 2000)***.

## History of the Fontan Procedure

It was in ***1971 when Fontan and Baude***t published their experience using a surgical procedure to restore a physiologic circulation in three patients with tricuspid atresia. Of the three patients, two survived, thus ushering a new era in the treatment of children with complex forms of congenital cardiac disease. This operation itself, however, was the culmination of a series of experimental and clinical observations that underlined the clinical possibility totally to bypass the right side of the heart ***(Fontan 1971)***

As discussed above, it was in 1971, that Fontan and Baudet first reported clinically successful "complete bypass of the right heart" in patients with tricuspid atresia. This was accomplished by the combination of a "classic cavopulmonary anastomosis" between the superior caval vein and right pulmonary artery, and channeling the inferior caval venous flow into the left pulmonary artery by using the proximal end of the right pulmonary artery and right atrium as a conduit, with interposition of an aortic valvar homograft. In light of the more current understanding of the operation, it is interesting to read their speculation as to why their intervention was successful. They suggested that the pump function of a "hypertrophied right atrium, as in tricuspid atresia", coupled with valvar homografts "inserted into the inferior vena cava", and the other at the "exit of the right atrium" to the left lung, provided the necessary "additional work

represented by a pulmonary arterial pressure higher than the left atrial pressure" ***(Fontan 1971).***

The pace of growth and modifications to the original surgical procedure continued to increase, and subsequently other authors published the results of their modifications and experience of the application of Fontan's operation to patients with tricuspid atresia.
In ***1973, Kreutzer et al***. reported their modification using a direct anastomosis of the pulmonary valve to the right atrial appendage, without the concomitant use of a cavopulmonary anastomosis or a valve in the inferior caval vein.

In ***1979, Bjork et al***. published their experience creating a direct and valveless communication between the right atrium and the right ventricle. As experience accumulated with the operation and its variants, it also was applied to other forms of congenital cardiac disease where the anatomy made a biventricular repair impossible.
In ***1976, Yacoub et al***. reported their experience in patients with "single ventricle and pulmonary stenosis".

In the early ***1980s, Norwood et al.*** reported their experience in children with aortic and mitral atresia physiologically "corrected" using the Fontan procedure as part of their surgical management.

When the Fontan procedure was initially described, ***Fontan et al.*** established 10 criterions for selection of patients, which became known as the *"Ten Commandments"*. With time, it became apparent that many of these "commandments" were not true requirements for a successful procedure. Although initially predicated for patients older than 4 years, we now know that it can be done successfully on patients aged 2 years, or even younger. Ideally, the patient can walk or at least crawl. While sinus rhythm is ideal, atrioventricular sequential pacing can be employed if sinus rhythm is not present. Normal caval venous drainage is not necessary, as many creative surgical approaches have now been developed to successfully utilize the Fontan process in patients with abnormalities of venous return. An enlarged right atrium can be managed with a reduction atrioplasty. Elevated pulmonary arterial pressure may be acceptable in states of high flow of blood to the lungs.

The crucial issue is pulmonary vascular resistance. Impairing effects from previous shunts typically do not prohibit the Fontan procedure, and the pulmonary arteries can be rehabilitated either surgically or in the catheterization laboratory prior to the completion Fontan. Thus, in the final analysis, the three crucial issues that remain from these 10 commandments are low pulmonary vascular resistance, satisfactory systemic ventricular function, and competence of the atrioventricular valve(s). ***(Giroud 2006).***