EXPERIENCE OF FONTAN SURGERY AT ARMED FORCES INSTITUTE OF CARDIOLOGY /NATIONAL INSTITUTE OF HEART DISEASES (AFIC-NIHD)

Syed Shahid Nafees Zaidi, Kamal Saleem, Inamullah Khan, Nausheen Bakht, Iftikhar Ahmed

Armed Forces institute of Cardiology Rawalpindi

ABSTRACT

Objective: To assess the overall outcome and success of Fontan surgery at our institute.

Place and Duration of study: AFIC-NIHD Rawalpindi. 01 September 2005 to 31 March 2010.

Patients and Methods: For this retrospective study, institute's cardiac surgery database was used. Patients of single ventricle physiology, who had normal Left Ventricular End-diastolic Pressure and pulmonary artery pressures, were included. Data was analyzed using SPSS version 16.

Results: A total of 34 Fontan procedures were done. The mean age at operation was 4.83±1.37 years. There were 22(64.7%) males and 12(35.3%) females. Twenty five (73.5%) had a staged Fontan (successful previous Bidirectional Glenn's shunt, BDG). Nine (26.5%) were primary Fontan procedures (no successful previous Bidirectional Glenn's shunt, BDG). Thirty two (94.8%) were Extra Cardiac Conduit Fontan (ECCF) and 2(5.8%) were Intra Cardiac Fontan. Mean Bypass time was 132.65±48.44 minutes. Aorta was cross clamped in intracardiac Fontan and its mean time was 43.31±5.85minutes. Fenestration was employed in 14(41.2%) patients. Mean pre-operative oxygen saturations were 77.41±10.27%, which significantly increased to 93.94± 3.96 % post-operatively ($p<0.001$). In-hospital mortality was 2(5.8 %).

Conclusion: Fontan surgery has acceptable morbidity and mortality in our set up.

Keywords: Birectional Glenns Shunt, Fontan, Single ventricle.

INTRODUCTION

Single ventricle is a group of congenital heart defects differing from each other with a common feature of a single ventricle of adequate function and size[1].

These patients undergo reconstructive surgery in stages[2]. This eventually culminates into the final stage of palliation called total cavo-pulmonary anastomosis (TCPC) or the Fontan procedure[3].

There are two types of Fontan procedures depending upon whether a previous Bidirectional Glenns Shunt (BDG) is, or is not done. A patient in whom a previous BDG is not undertaken and Fontan procedure is directly performed is called a "direct/primary Fontan". In the second type of procedure, a BDG is performed previously and the Fontan procedure is called staged Fontan[4,5].

Currently, a primary or staged Fontan operation entails three different technical procedures. These are "Lateral Tunnel Fontan" (LTF), "Extra-Cardiac Conduit Fontan" (ECCF) and "Intracardiac Fontan". In LTF, blood in the inferior vena cava is diverted to pulmonary circulation by anastomosing a small length of intra-atrial Gore-Tex conduit to Bidirectional Glenns Shunt and clamping of aorta is required[6,7] . In ECCF, the pulmonary artery and inferior vena cava are anastomosed by sand wiching a Gore-Tex conduit of age-appropriate size and in this procedure clamping of aorta is not required. Intracardiac Fontan also utilize intra-atrial conduit to deliver systemic venous blood flow to Pulmonary artery.

In majority of the patients, staged Fontan procedure is done. However, selection of a particular surgical procedure is the surgeon's decision and it varies from case to case[8].

Most Fontans do not function efficiently beyond 30 to 40 years. However, improvements in surgical technique and medical care may increase this age significantly[9].

The present study highlights the overall outcome and success of the procedure at our institute. This research work has significance as the authors could not find any published work on the subject, from Pakistan.

Correspondence: Lt Col Syed Shahid Nafees Zaidi, Classified Surgeon, AFIC/NIHD Rawalpindi
Email: nausheenbakht@gmail.com
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PATIENTS AND METHODS

This retrospective study was conducted at Armed Forces Institute of Cardiology-National Institute of Heart Disease (AFIC-NIHD) Rawalpindi Pakistan.

In 2005, AFIC-NIHD collaborated with the International Children's Heart Foundation, (ICHF) USA for the up gradation of its congenital surgery program. Today AFIC-NIHD is the only hospital of Pakistan where Fontan procedure is successfully and regularly done.

Our study subjects were patients operated for single ventricle physiology, in our institute between 01 September 2005 and 31 March 2010.

The study instrument was the institute's cardiac surgery database. Our variables included age, gender, weight, height, disease pattern, staged or primary Fontan (previous or no BDG)s, bypass time, aortic clamp time, pre and post-operative oxygen saturation, Intra-cardiac Fontan or Extra cardiac conduit Fontan (ECCF) done, conduit size, fenestration, ventilation time, Fontan pressures 24 hours post-operatively, cardiac rhythm, Sildenafil use, reopening done, graft thrombosis, duration of inotropes, duration of pleural drainage, ICU stay, in-hospital mortality and 6 months post-operative mortality.

Patients with normal Left Ventricular End-diastolic Pressure and pulmonary artery pressures were included in the study. Cardiac catheterization was performed in all patients.

Statistical Analysis

Data was analyzed using SPSS version 16. For quantitative variables, mean and standard deviation (SD) and for qualitative variables, frequencies along with percentage were used for description of variables. Independent sample's t-test was used for comparison of quantitative variables while chi-square test was used for comparison of qualitative variables between different groups. A two-tailed $p<0.05$ was considered statistically significant.

RESULTS

In this four and a half year study period, a total of 34 patients underwent Fontan procedure. Males were 22(64.7%) and females were 12(35.3%). Demographics are shown in Table. Distribution of cardiac anatomic diagnosis is displayed in Figure.

Twenty five (73.5%) patients had staged Fontan, of these Intra-cardiac were 2 and ECCF were 23. Patients who underwent primary Fontan were 9(26.4%), and all of these were ECCF. Mean Bypass time was 132.65 ± 48.44 minutes. Mean aortic clamp time was 43.31 ± 5.85 minutes. In ECCF the mean size of Gore-Tex conduit was 22.4 ± 2.39 mm.

Fenestration was done in 14(41.2%) patients. Mean pre-operative oxygen saturations were 77.41 ± 10.27 %, which significantly increased to 93.94 ± 6% post-operatively ($p<0.001$). Mean ventilation time was 4.76 ± 3.37 hours. Mean Fontan pressure was 15mm Hg. Ten (29.4%) patients were re-opened; 6 for post-operative bleeding, 1 for graft revision and 3 for blocked pleural drains along with pacing wires readjustment. Inotropes were used in 22(65%) of patients. Pleural drainage continued for 6.21 ±1.9 days. Sinus rhythm was present in 27(79.4%) patients; in 4 cases it was AV nodal while in 3 patients atrial fibrillation was present. Tab Sildenafil was started per-operatively and continued post-operatively in 7(20.65%) patients. 494.12±94.06 ml blood products were used to haemodynamically stabilize the patients. Mean ICU stay was 7.21±1.9 days. One patient developed turbulent flow due to partial thrombus formation on IVC side of ECCF. He was monitored with serial 2D-Echo and treated with IV heparin. Total In- hospital mortality was 2(5.88 %). One death occurred 6 months post operatively due to coagulation disorder and massive upper GI bleeding.

Legend: Complete AVSD=Complete atrio ventricular septal defect; DILV=Double Inlet Left Ventricle; DORV=Double Outlet Right Ventricle; TGA+DILV=Transposition of great arteries and double inlet left ventricle; TGA+DORV= Transposition of great arteries and double outlet Right ventricle; PUL ARESIA=Pulmonary atresia

DISCUSSION

Thirty four patients who underwent Fontan operation had a mean age of 4.8 years.

Table: Demographics of patients

Variables	Minimum	Maximum	Mean	SD
Age(Years)	3.0	7.0	4.831	1.376
Weight (Kg)	9	24	14.62	3.681
Height (CM)	80	113	100.73	8.795

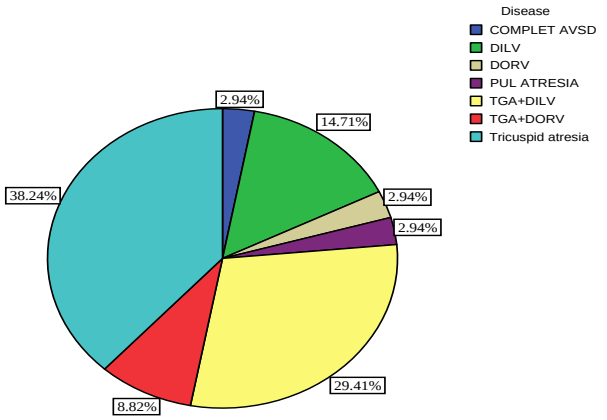

Figure: Distribution of disease pattern in all cases of Fontan operation.

Internationally, Fontan surgery is undertaken at two years of age[1, 2, 3]. In our case, relatively late age surgery is due to late reporting of patients[4,5].

Bypass time and aortic cross clamp time in our study was similar to other contemporary studies[8].

Some researchers have reported good results without using heart lung machine,[9] however, we have used it in all of our cases.

The mean size of Gortex tube graft anastomosed was 22.4 mm which is larger as compared to other studies[10,11,12]. It is due to the older age of our clientele.

In our study, fenestration was done in 41.2% cases but lesser percentages have been quoted by other researches[13,14]. The mean size of fenestration (5.1mm), however, matches with other studies[15].

Post-surgery oxygen saturation increased significantly (p-value <0.001). The increase was less in fenestrated Fontan due to mixing of blood at atrial level.

Mean ventilation time of 4 hours in our study is at par with contemporary studies [16]. Early extubation enhances the haemodynamic stability of our Fontan patients post operatively and improves the patient outcome.

Mean Fontan pressures of 15mmHg in our study is also the same as in other studies[17].

Seven patients who had > 15mmHg Fontan pressures were given Sildenafil per-operatively, which was continued post-operatively for 3 months. Similar results have been reported by others[18]. Mean pleural drainage duration was slightly less than other studies[7,19]. More blood products were used on our patients due to more post-operative blood loss and hence a higher re-opening rate.

Our re-opening rate of 29.4% is higher than others. A strong emphasis on effective haemostatic techniques / protocols, optimum chest physiotherapy, milking of pleural drains will lead to an improvement of results[15].

Mean ICU stay of 7 days is more than what others have reported[7,19,20.] . This is because the procedure is relatively new in our set-up and hope the ICU stay will decrease as more and more volume of this procedure is done in the future

One patient of ECCF, while in hospital, developed partial thrombosis of the inferior vena cava side of the conduit and hepatomegaly. He was treated with intravenous heparin and monitored with serial 2-D Echo. After treatment of 7 days his condition improved, hepatomegaly regressed to near normal levels and he was later discharged. Total in-hospital mortality of two patients (5.8%) is comparable with other studies[4,7,19,21,22].

There are some limitations to the present study. First this study was not a prospective, randomized study and second in more recent patients, there is limited follow-up which may lead to under estimation of long term morbidity and mortality. In addition,further evaluation of a large number of patients with long term follow-up will be necessary.

CONCLUSION

Patients of single ventricle morphology undergo staged reconstructive surgery and usually the first stage is BDG except in few patients depending on their presentation, pulmonary artery band or modified Blalock-Taussig shunt becomes the first stage. If BDG remains successful, they are offered Fontan surgery. The need of the hour is to conduct further research into intracardiac Fontan, LTF and ECCF; fenestrated and non-fenestrated Fontan; and primary versus staged Fontan. Collaboration with International Children Heart Foundation,USA (ICHF) has led to an improvement of our results. We are constantly striving to make them even better.

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