Open Access

Original Article

# Outcome of Early and Late Onset Fontan Operation in Patients with Univentricular Heart Repair

**Shahnawaz Sathio, Shakeel Ahmed, Mehtab Hussain, Nimra Zahid, Saad Ullah Siddiqui, Fazal ur Rehman***

Department of Pediatric Cardiology, National Institute of Cardiovascular Diseases, Karachi Pakistan, *Department of Pediatric Cardiology, Parvez Elahi Institute of Cardiology Bahawalpur

## ABSTRACT

***Objective*:** To determine the outcome of early and late-onset Fontan operation in patients with univentricular heart repair.
***Study Design*:** Quasi-experimental study.
***Place and Duration of the Study*:** Department of Paediatric Cardiology, National Institute of Cardiovascular Diseases, Karachi Pakistan, from Jan 2019 to Dec 2021.
***Methodology*:** During the study period, 121 patients of both genders, up to any age, with functionally univentricular hearts and who planned to undergo Fontan procedures were included. All cases were divided into two groups, Group-A had patients who underwent extra-cardiac Fontan at age ≤6 years, while Group B had patients who underwent extra-cardiac Fontan at age >6 years. Pre-procedure diagnosis, demographic, anatomic and hemodynamic characteristics, post-Fontan procedure morbidity and mortality were noted.
***Results*:** As per inclusion/exclusion criteria, 121 cases underwent Fontan surgery during the study period. There were 61(50.4%) male patients. Overall, the mean age was 7.02±4.6 years. Seventy-six 76(61.2%) cases were in Group-A and 45(38.8%) in Group-B. Mortality (within 30 days post-surgery) was reported in 11(9.1%) patients. Except for the mean duration of chest tube drainage ((0.010), there was no difference in terms of post-Fontan surgery outcomes between both study groups ($p$>0.05).
***Conclusion*:** The age of the patients at the time of the Fontan procedure did not affect the outcomes.

**Keywords:** Fontan surgery, mortality, oxygen saturation, univentricular heart.

***How to Cite This Article:*** *Sathio S, Ahmed S, Hussain M, Zahid N, Siddiqui SU, Rehman FU. Outcome of Early and Late Onset Fontan Operation in Patients with Univentricular Heart Repair. Pak Armed Forces Med J 2024; 74(4): 1007-1010. DOI: https://doi.org/10.51253/pafmj.v74i4.8006*



## INTRODUCTION

The first successful Fontan procedure was reported in 1971, and since then, this procedure has seen many modifications in cases of single-ventricle physiology.[1] Leval and Coworkers introduced total cavopulmonary connection (TCPC) in 1988, which soon became the standard operation approach.[2] The TCPC delivers better venous hemodynamics and is less arrhythmogenic than other contemporary Fontan modifications.[3] Main aims of all the modifications and innovations from the original Fontan surgery have mostly been to reduce the duration between palliative procedures and the actual surgical procedure, the implantation of the extra-cardiac conduit with/without bypass fenestration among the systemic venous reservoir and the atrium receiving the pulmonary veins and the ability to modulate the definitive stepped univentricular model in to two well differentiated surgical steps.[4,5]

The approach to functionally univentricular heart has seen lots of evolution, with the ultimate goal of achieving normal volume and pressure for the univentricular heart and as close as possible to normal oxygen saturation levels in the affected individuals.[6] As Fontan operation is expected to deliver near normal systemic oxygen saturation with a single ventricular physiology, pre-surgery factors like age have been reported to influence outcomes.[7] Contemporary pediatric cardiologists have endorsed opting for cases of relatively younger age for Fontan operation. However, the exact effect of age on early and late outcomes has yet to be described.[8] Among adults, TCPC may carry a higher risk when compared to pediatric age groups, as adults are found to have complications accompanying functional ventricles due to chronic hypoxia, volume overload, and higher venous pressure. Ventricular dysfunction and limitations in exercise capacity.[9] As no local data is available about the outcomes of the Fontan operation, we aimed to report our experience of early and late-onset Fontan operation outcomes in patients with univentricular heart repair. The findings of the present study provide helpful information about the influence of various pre-operative and post-operative characteristics on the outcomes of Fontan operation.

## METHODOLOGY

The quasi-experiment study was conducted at The Department of Pediatric Cardiology, National Institute of Cardiovascular Diseases, Karachi Pakistan,

**Correspondence: Dr Fazal ur Rehman,** Department of Pediatric Cardiology, NICVD, Karachi Pakistan
*Received: 11 Jan 2022; revision received: 15 Jun 2022; accepted: 22 Jun 2022*

from January 2019 to December 2021. Approval from the Institutional Ethical Committee" was obtained (ERC 1021/2021).

**Inclusion Criteria**: Patients of either gender and of any age group, with functionally univentricular hearts and planning to undergo Fontan procedures were included.

**Exclusion Criteria:** Patients with severe anomalies like discontinuation of the pulmonary branches, severe organic mitral insufficiency needing a prosthetic valve, or anomalous pulmonary venous drain were excluded.

Written informed consent were sought from either patients or their parents/guardians. We considered all cases coming to us per inclusion/exclusion criteria for this study during the described study duration. The non-probability consecutive sampling technique was adopted. All cases were divided into two groups. Group-A had patients who underwent extra-cardiac Fontan at age ≤ 6 years, while Group-B had patients who underwent extra-cardiac Fontan at age > six years (Figure).[10]

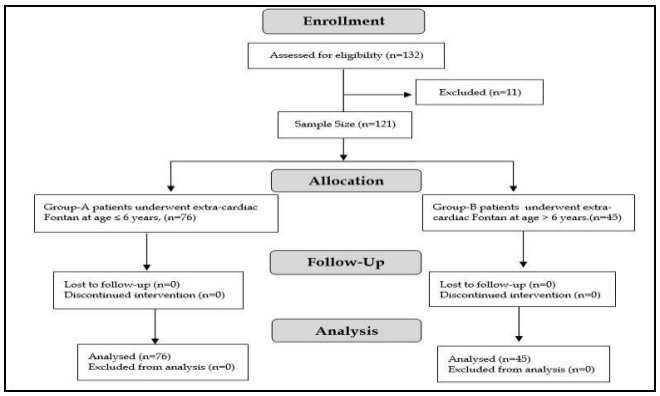


**Figure: Patient Flow Diagram (n=121)**

Hemodynamic criteria for early Fontan completion were pulmonary artery pressure below 15 mmHg, pulmonary vascular resistance below 4 WU*m2 and existence of aortopulmonary collaterals. Increasing cyanosis, decreased physical activity and/or failure to thrive were other significant impetus for early Fontan procedure.[11]

The pre-procedure diagnosis was labelled, while all patients' demographic, anatomic and hemodynamic characteristics were noted. Standard Fontan procedures were done in all cases. Post-procedure, all patients were kept on Warfarin plus Aspirin to have an International Normalized Ratio (INR) between 1.8-2.5. Post-procedure outcomes and morbidity and mortality were noted in all patients and compared between both study groups. A specially designed proforma was used to record all study data.

Data was analyzed using Statistical Package for the Social Sciences (SPSS) SPSS version 26.0. Qualitative data was expressed as frequency and percentages. Mean and standard deviation (SD) were estimated for quantitative data. Independent sample t-test was used to compare quantitative data, while the chi-square test was employed for qualitative data, considering *p* value of <0.05 as significant.

## RESULTS

In a total of 121 cases, Fontan surgery was performed. There were 61(50.4%) males. The mean age was 7.02±4.6 years, and the mean body weight was 20.3±10.8 kg. Mean pulmonary artery pressure was 11.1±1.5 mmHg, whereas mean arterial oxygen saturation (%) at enrollment was 80.3±5.2. There were 76(61.2%) cases who were aged ≤ 6 years so they were placed in Group-A while remaining 45(38.8%) cases belonged to Group-B (Table-I).

**Table-I: Characteristics of the Patients of Study Groups (n=121)**

| Characteristics | | Group-A (n=76) | Group-B (n=45) | *p*-value |
|---|---|---|---|---|
| Gender | Male | 42(55.3%) | 19(42.1%) | 0.166 |
| | Female | 34(44.7%) | 26(57.8%) | |
| Mean Age (years) | | 4.0±1.3 | 12.1±3.7 | <0.001 |
| Mean Body Weight (kg) | | 13.9±3.6 | 30.9±10.5 | <0.001 |
| Single Ventricle Morphology | Right | 23(30.3%) | 7(15.6%) | 0.062 |
| | Left | 41(53.9%) | 24(53.3%) | |
| | Intermediate | 12(15.8%) | 14(31.1%) | |
| Mean Pulmonary Artery Pressure (mmHg) | | 11.2±1.4 | 11.1±1.6 | 0.805 |
| Mean Arterial Oxygen Saturation (%) | | 82.8±3.5 | 80.5±4.9 | 0.004 |
| Diagnosis | Dextrocardia | 8(10.5%) | 4(8.9%) | 0.630 |
| | Double Inlet Left Ventricle | 27(35.5%) | 15(33.3%) | |
| | Double Outlet Right Ventricle | 15(19.7%) | 6(13.3%) | |
| | Mitral Atresia | 4(5.3%) | 1(2.2%) | |
| | Tricuspid Atresia | 8(10.5%) | 9(20.0%) | |
| | Unbalanced Atrioventricular Septal Defect | 13(17.1%) | 8(17.8%) | |
| | Others | 1(1.3%) | 2(4.4%) | |

*Group-A: Early Fontan Surgery (≤6 years); Group-B: Late Fontan Surgery (>6 years)*

The mean post-operative saturation (%) was 92.8±3.1. Overall, the mean post-surgery duration of mechanical ventilation was 1.92±1.0 days. The mean duration of ICU stay was 3.1±1.0 days. Overall, the mean duration of hospitalization was 22.6±5.2 days. In 121 patients, mortality (within 30 days post-surgery)

was reported in 11(9.1%) patients. Table-II shows a comparison of post-Fontan surgery outcomes in both study groups. Except for the mean duration of chest tube drainage ((0.010), there was no difference in terms of post-Fontan surgery outcomes like thrombosis ($p$=0.771), stroke (0.260), chylothorax ($p$=0.854), bleeding ($p$=0.417), serious post-surgery arrhythmias ($p$=0.590), mean saturation ($p$=0.421), mean duration of mechanical ventilation ($p$=0.527), mean ICU stay ($p$=0.472), mean total duration of hospital stay ($p$=0.105) and mortality ($p$=0.475) in between both study groups ($p$>0.05).

**Table-II: Comparison of Post Fontan Surgery Outcomes in Study Groups (n=121)**

| Post Fontan Surgery Outcomes | Group-A (n=76) | Group-B (n=45) | *p*-value |
|---|---|---|---|
| Thrombosis | 8(10.5%) | 4(8.9%) | 0.771 |
| Stroke | 3(3.9%) | 4(8.9%) | 0.260 |
| Chylothorax | 18(23.7%) | 10(22.2%) | 0.854 |
| Bleeding | 4(5.3%) | 1(2.2%) | 0.417 |
| Serious Post-Surgery Arrhythmias | 2(2.6%) | 2(4.4%) | 0.590 |
| Mean Saturation (%) | 93.0±2.9 | 92.5±3.5 | 0.421 |
| Mean Duration of Mechanical Ventilation (days) | 1.96±1.0 | 1.84±1.0 | 0.527 |
| Mean Duration of Chest Tube Drainage (days) | 11.8±2.5 | 13.4±4.4 | 0.010 |
| Mean ICU Stay (days) | 3.2±1.0 | 3.02±1.0 | 0.472 |
| Mean Total duration of Hospital Stay (days) | 23.2±5.1 | 21.6±5.4 | 0.105 |
| Mortality | 8(10.5%) | 3(6.7%) | 0.475 |

*Group-A: Early Fontan Surgery (≤6 years); Group-B: Late Fontan Surgery (>6 years)*

## DISCUSSION

In the present research, we noticed that the most common post-procedure morbidities are thrombosis (10.0%), chylothorax (23.1%) and stroke (5.8%). The types of post-Fontan surgery morbidity and incidence correlate well with other researchers.[12,13] Cazzaniga *et al.* [14] from Spain revealed subaortic stenosis (8%), protein loss syndrome (4%), arrhythmias (33%) and thromboembolism (4%) to be the commonest post-surgery morbidities among cases undergoing Fontan procedures. Another study from the Middle East found thrombosis, stroke, chylothorax, bleeding, pericardial effusion and wound infection to be the commonest short-term post-Fontan procedure morbidities.[10]

There has yet to be a consensus about the best age for Fontan procedure in contemporary literature. Some researchers have propagated early Fontan surgery among children.[15,16] In the present study, we noted a survival rate of 90.9% among cases undergoing the Fontan procedure. Bezuska *et al.* [11] from Lithuania shared their 30-year experience of Fontan surgery, which revealed a survival rate of 94%, which is close to what we observed. A study from the Middle East shared an overall hospital mortality of 7.9%.[10] The literature reports mid-term survival rates between 92-98%, so our findings correlate well with the published literature.[13,17,18] Long-term follow-up data (20-year follow-up) from Spain[14] showed a survival rate of 61%, while past data from Mayo Clinic showed that 16% of the patients died in the 1stmonth following Fontan surgery.[19] Some significant factors affecting the survival of Fontan surgery cases have been labelled as raised central venous pressure and abnormal cardiac functions[20]. However, as we only noticed relatively short-term survival in the present study, we could not establish any linkages between post-Fontan surgery mortality and studied variables. Recent decades have witnessed progress toward better long-term survival rates among patients undergoing Fontan surgery.[21] recent report estimated that cases undergoing Fontan surgery will be doubled in the next two decades, which means that there will be increased demand for medical and non-medical healthcare services for these patients.[22] Stake-holders need to plan strategies to cater to increasing requirements for the care of patients requiring Fontan surgeries. The present study is the first one from Pakistan to show early versus late Fontan surgery outcomes.

## LIMITATIONS OF THE STUDY

We only noted relatively short outcomes in the present study.

## ACKNOWLEDGEMENT

The authors thank Muhammad Aamir (Research Consultant, Bahawalpur, Pakistan) for his volunteer assistance in statistical analysis.

## CONCLUSION

The age of the patients at the time of the Fontan procedure did not affect the outcomes. Multicenter randomized clinical trials with larger data sets and long-term follow-ups can further illuminate the various aspects of pre- and post-Fontan surgery.

**Conflict of Interest:** None.

**Authors Contribution**

Following authors have made substantial contributions to the manuscript as under:

SS & SA: Conception, drafting the manuscript, approval of the final version to be published.

MH & NZ: Data acquisition, data analysis, critical review, approval of the final version to be published.

SUS & FUR: Study design, data interpretation, drafting the manuscript, critical review, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

## REFERENCES


1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971; 26(3): 240–248. https://doi.org/10.1136/thx.26.3.240
2. de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience. J Thorac Cardiovasc Surg 1988; 96(5): 682-695.
3. Balaji S, Gewillig M, Bull C, de Leval MR, Deanfield JE. Arrhythmias after the Fontan procedure. Comparison of total cavopulmonary connection and atriopulmonary connection. Circulation 1991; 84(5 Suppl): III162-67.
4. Kverneland LS, Kramer P, Ovroutski S. Five decades of the Fontan operation: A systematic review of international reports on outcomes after univentricular palliation. Congenit Heart Dis 2018; 13(2): 181-193. https://doi.org/10.1111/chd.12570
5. Caneo LF, Neirotti RA, Turquetto AL, Jatene MB. The Fontan operation is not the end of the road. Arq Bras Cardiol 2016; 106(2): 162-165. https://doi.org/10.5935/abc.20160017
6. Trusty PM, Slesnick TC, Wei ZA, Rossignac J, Kanter KR, Fogel MA, et al. Fontan Surgical Planning: Previous Accomplishments, Current Challenges, and Future Directions. J Cardiovasc Transl Res 2018; 11(2): 133-144. https://doi.org/10.1007/s12265-018-9786-0
7. Fredenburg TB, Johnson TR, Cohen MD. The Fontan procedure: anatomy, complications, and manifestations of failure. Radiographics 2011; 31(2): 453-463. https://doi.org/10.1148/rg.312105027
8. Pizarro C, Mroczek T, Gidding SS, Murphy JD, Norwood WI. Fontan completion in infants. Ann Thorac Surg 2006; 81(6): 2243-2248. https://doi.org/10.1016/j.athoracsur.2006.01.016
9. Veldtman GR, Nishimoto A, Siu S, Freeman M, Fredriksen PM, Gatzoulis MA, et al. The Fontan procedure in adults, Heart 2001; 86(3): 330–335. https://doi.org/10.1136/heart.86.3.330
10. Deraz S, Ismail MF, Jamjoom A. Outcome of early and late onset Fontan operation in patients with univentricular heart repair. Egyptian Heart J 2014; 66(2): 171-176. https://doi.org/10.1016/j.ehj.2013.06.001
11. Bezuska L, Lebetkevicius V, Sudikiene R, Liekiene D, Tarutis V. 30-year experience of Fontan surgery: single-centre's data. J Cardiothorac Surg 2017; 12(1): 67. https://doi.org/10.1186/s13019-017-0634-0
12. Firdouse M, Agarwal A, Chan AK, Mondal T. Thrombosis and thromboembolic complications in Fontan patients: a literature review. Clin Appl Thromb Hemost 2014; 20(5): 484-492. https://doi.org/10.1177/1076029613520464
13. Raj S, Rosenkranz E, Sears B, Swaminathan S. Intermediate-term results after extracardiac conduit Fontan palliation in children and young adults with single ventricle physiology-A single-center experience. Pediatr Cardiol 2016; 37(6): 1111-1118. https://doi.org/10.1007/s00246-016-1402-y
14. Cazzaniga M, Fernández Pineda L, Villagrá F, Pérez De León J, Gómez R, Sánchez P, et al. Single-stage Fontan procedure: early and late outcome in 124 patients. Rev Esp Cardiol 2002; 55(4): 391-412. https://doi.org/10.1016/s0300-8932(02)76619-0
15. Ota N, Fujimoto Y, Murata M, Tosaka Y, Ide Y, Tachi M, et al. Impact of postoperative hemodynamics in patients with functional single ventricle undergoing Fontan completion before weighing 10 kg. Ann Thorac Surg 2012; 94(5): 1570-1577. https://doi.org/10.1016/j.athoracsur.2012.06.022
16. Pundi KN, Johnson JN, Dearani JA, Pundi KN, Li Z, Hinck CA, et al. 40-Year Follow-Up After the Fontan Operation: Long-Term Outcomes of 1,052 Patients. J Am Coll Cardiol 2015; 66(15): 1700-1710. https://doi.org/10.1016/j.jacc.2015.07.065
17. d'Udekem Y, Iyengar AJ, Galati JC, Forsdick V, Weintraub RG, Wheaton GR, et al. Redefining expectations of long-term survival after the Fontan procedure: twenty-five years of follow-up from the entire population of Australia and New Zealand. Circulation 2014; 130(11 Suppl 1): S32-38. https://doi.org/10.1161/CIRCULATIONAHA.113.007764
18. Ono M, Kasnar-Samprec J, Hager A, Cleuziou J, Burri M, Langenbach C, et al. Clinical outcome following total cavopulmonary connection: a 20-year single-centre experience. Eur J Cardiothorac Surg 2016; 50(4): 632-641. https://doi.org/10.1093/ejcts/ezw091
19. Driscoll DJ, Offord KP, Feldt RH, Schaff HV, Puga FJ, Danielson GK. Five- to fifteen-year follow-up after Fontan operation. Circulation 1992; 85(2): 469-496. https://doi.org/10.1161/01.cir.85.2.469
20. Rychik J. The relentless effects of the Fontan paradox. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2016; 19(1): 37-43. https://doi.org/10.1053/j.pcsu.2015.11.006
21. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007; 115(2): 163-172. https://doi.org/10.1161/CIRCULATIONAHA.106.627224
22. Schilling C, Dalziel K, Nunn R, Du Plessis K, Shi WY, Celermajer D, et al. The Fontan epidemic: Population projections from the Australia and New Zealand Fontan Registry. Int J Cardiol 2016; 219: 14-19. https://doi.org/10.1016/j.ijcard.2016.05.035