

# MID-TERM RESULTS OF THE LATERAL TUNNEL OPERATION AT QUEEN ALIA HEART INSTITUTE

*Awni Madani MD\**, *Zuhdi Hanouti MD\**, *Rania Haddadin MD\**, *Abdelafattah Haweleh MD\**,  
*Imad Khreisat MD\**, *Issa Hijazi MD\**, *Mustafa Al-Muhaya MD\**, *Yaser Al-Ghoul MD^*,  
*Yousef Zureqat MD\*\**, *Fakhri Hakim MD\**, *Husam Nesheiwat MD\*\**

## ABSTRACT

**Objective:** To determine midterm results of univentricular repair using intra-atrial lateral tunnel at Queen Alia Heart Institute.

**Methods:** Between January 1999 and January 2005, 159 patients (99 males, 60 females) underwent either a fenestrated or non-fenestrated lateral tunnel (Fontan procedure), for a wide range of complex congenital heart disease with a functional single ventricle at Queen Alia Heart Institute. Median age at operation time was 3.8 years (range 1.5-17 years). Multiple factors were analyzed including: anatomical variations, mean right atrial pressure, pulmonary artery pressure, ventricular end diastolic pressure, aortic saturation, cardiopulmonary bypass time and ischemic time, presence of arrhythmias, pacemaker insertion, thromboembolic complications, early and late mortality.

**Results:** The major anatomic diagnosis was double inlet left ventricle in 72 patients (45.3%) and the least was unbalanced complete atrioventricular septal defect among 8 patients (5%). Dextrocardia was found in 17 patients. The mean right atrial pressure was  $8\pm 3$  mmHg, pulmonary artery pressure  $13\pm 2.6$  mmHg, PVR  $1.9\pm 2$  Woodunits.m<sup>2</sup>, ventricular end diastolic pressure  $14\pm 2.2$  mmHg, aortic saturation  $83\pm 4$  %, cardiopulmonary bypass time  $121\pm 9$  minutes, ischemic time  $47\pm 3$  minutes. Early postoperative supraventricular tachyarrhythmia (SVT) occurred among 10 cases (6.3%) of patients, in whom four needed antiarrhythmic drugs before discharge. Early bradyarrhythmia needing pacemaker in 2 patients (1.26%), whereas 6 patients needed pacemaker at mid term follow-up. There were 10 (6.3%) early deaths and no late deaths occurred after 6 years of follow-up. Seven (4.7%) patients developed protein losing enteropathy as a late complication and 7 more patients (4.7%) developed neurological complications.

**Conclusion:** Proper selection of patients for the lateral tunnel Fontan procedure results in excellent early and mid-term survival and functional outcome with low incidence of complications.

**Key words:** Fontan operation, Lateral tunnel, Single ventricle

JRMS December 2009; 16(3):5-9

## Introduction

Since the first successful Fontan operation for tricuspid atresia that was performed in 1971,<sup>(1,2)</sup>

many modifications have been developed to optimize the early and long-term outcomes of patients with complex cyanotic heart defects with a

From the Departments of:

\*Pediatric Cardiology, Queen Alia Heart Institute, King Hussein Medical Center, KHMC, Amman-Jordan

^Anesthesia, Queen Alia Heart Institute, King Hussein Medical Center, KHMC, Amman-Jordan

\*\*Cardiology Surgery, Queen Alia Heart Institute, King Hussein Medical Center, KHMC

Correspondence should be addressed to Dr. A. Al-Madani, Queen Alia Heart Institute, KHMC, Amman-Jordan, E-mail: [amadani33@hotmail.com](mailto:amadani33@hotmail.com)

Manuscript received June 10, 2006. Accepted January 11, 2007

**Table I. Major anatomic diagnoses**

Double inlet left ventricle	72 patients	45.3%
Tricuspid atresia	63 patients	39.6%
Double outlet right ventricle with small LV	9 patients	5.7%
Pulmonary atresia with intact septum	8 patients	5%
Unbalanced complete atrioventricular septal defect	8 patients	5%

functional single ventricle.<sup>(3,4)</sup> Since the intermediate and long-term results showed that the incorporation of the entire systemic venous atrium in the Fontan circulation was associated with a high incidence of supraventricular tachycardia (SVT) and thrombo-embolic complications,<sup>(2,5)</sup> more recent modifications have focused on the use of intra-atrial communication which was developed by de Leval and associates,<sup>(6)</sup> Jonas, and Castaneda.<sup>(7)</sup> The concept was to achieve a laminar blood flow in a tubular construct and to provide growth potential as well as a source for endothelializations of the synthetic surface.<sup>(8)</sup> This study was conducted to determine midterm results of univentricular repair using intra-atrial lateral tunnel at Queen Alia Heart Institute (QAH).

## Methods

Patients undergoing a total cavopulmonary anastomosis with intra-atrial lateral tunnel at QAH between January 1999 and January 2005 for single ventricle physiology were identified from the filing of the pediatric cardiology department. Patients who underwent extra cardiac tunnel using Contegra were excluded. A total of 159 patients were included in this study for whom the medical records were reviewed for patient personal data, symptoms, echocardiography reports, cardiac catheterization reports, all available ECGs, operative notes, pre- and post-operative hemodynamics. Ninety-nine patients were males (62.3%) and sixty patients were females (37.7%) with a ratio of 1.65:1. The median age at the time of operation was 3.8 years (range 17 months to 17 years). The major anatomic diagnosis was double inlet left ventricle in 72 patients (45.3%) and the least was unbalanced complete AV septal defect in 8 patients (5%) (see Table I).

Dextrocardia was found in 17 patients (10.7%). Eighteen patients (11.5%) had associated anomalies of systemic venous return, whereas 7 patients (4.4%) had associated anomalies of pulmonary

**Table II. Preoperative demographic and hemodynamic data**

Median age	3.8years (range17 months to 17 years)
Male to female ratio	1.65: 1
Mean pulmonary artery pressure	13 ± 2.6 mmHg
Mean pulmonary vascular resistance	1.9± 2mmHg
Mean right atrial pressure	8± 3mmHg
Mean ventricular end diastolic pressure	14± 2.2mmHg
Mean aortic saturation	83± 5%

venous return. None or trace AV valve regurgitation was present in 105 patients (66%), mild AV valve regurgitation in 51 patients (32%) and moderate AV valve regurgitation in 3 patients (2%). One hundred thirty three patients had previously undergone a palliative procedure, as it helps in de-loading the ventricle, improving the pulmonary vascularity and the pulmonary blood flow and improving the ventricular function; 55 of them had a previous systemic pulmonary shunt, 43 patients had bi-directional Glenn shunt, 23 patients had pulmonary artery banding, and 12 patients had coarctation repair. Preoperatively five patients had documented supraventricular tachycardia (SVT). Preoperative demographic and hemodynamic data are shown in Table II.

Patients with increased ventricular end diastolic pressure commonly had increased ventricular volume load with preserved ventricular function. All Fontan (LT) operations were performed via a median sternotomy and utilized cardiopulmonary bypass (CPB) and aortic cross clamping to ensure adequate concomitant intracardiac procedures including atrio-ventricular (AV) valve repair, anomalous pulmonary venous drainage and fenestration of the lateral tunnel which was performed in 32 (20%) patients who met certain criteria.<sup>(9)</sup> These criteria were elevated systolic pulmonary artery pressure > 20mmHg and/ or PVR > 2Wood units/m<sup>2</sup>, abnormal pulmonary artery anatomy and significant AV valve regurgitation.

The fenestra was done using 2.5-4 mm coronary punch. The mean duration of CPB time was 121± 9 minutes, whereas the mean aortic cross clamping time was 47±3 minutes. Postoperatively the mean CVP on the first day post operative was 13.9± 2.5mmHg (range 7- 25 mmHg). Postoperatively, the mean CVP on the first post operative day was 13.9±2.5mmHg (range 7-25mmHg). Bilateral pleural chest tubes and one pericardial were placed

in all patients and were removed once they drained < 2ml/kg/day for each tube. In the first 4 years of the study all patients with fenestration were kept on acetylsalicylic acid (5mg/kg/d), but in the last 2 years they were maintained on coumadin (warfarin) keeping their INR between 2-2.5 as life long anticoagulant in order to prevent thromboembolic complications.

#### **Statistical Analysis:**

Statistical analysis was performed using the CSS software. All results were expressed as mean± standard deviation or median and range. Variables comparing the mortality and the survival group were analyzed using the paired student T test, and the statistical significance was defined as P value < 0.05.

#### **Early Results:**

There were 10 (6.3%) early deaths that occurred in the first 30 days post operatively. The preoperative diagnoses and the cause of death are shown in table III. The most common cause for early death was ventricular dysfunction in seven patients (70%). Significant pleural effusions were drained for longer than 14 days in 28 patients. A total of 21 patients (13.3%) had early postoperative arrhythmias. Ten patients (6.3%) had SVT and 4 of those were discharged on antiarrhythmic medications. Nine patients (5.7%) had atrial junctional rhythm with normal heart rate and 2 patients (5.7%) needed implantation of pacemaker for atrioventricular dissociation before discharge. The rest of the patients (n=148, 93%) had documented normal sinus rhythm before discharge.

#### **Mid-term Results:**

The mean time of follow up was 3.2 years with a range of 1.5-6.5 years, which is a relatively short period of time for such an operation yet there were no intermediate deaths. The fenestration closed spontaneously in 27 patients (84.3%). Neurological symptoms were present in a total of 7 patients, in two of them hemiplegia developed at 5 and 18 months respectively and the other 5 had seizures after the Fontan (LT) operation, all of them had open fenestrations, and were on aspirin. The two patients with hemiplegia found to have protein C and S deficiency for that they were maintained on warfarin. Protein losing enteropathy with persistent or recurrent edema, hypoproteinemia occurred in 7 patients (4.69%), who were treated with parenteral

albumin infusions, diuretics, and angiotensin-converting enzyme inhibitors. Five of them improved, whereas two patients improved after creation of fenestra in the lateral tunnel. Other late reoperations or reinterventions included embolization of systemic-pulmonary arterial collaterals in 5 patients. ECGs recording during follow up were seen in one hundred fourteen (76.5%) of patients, whereas Holter recordings were available for thirty-eight patients (25.5%). SVT was reported in thirteen (8.7%) patients during the follow-up period, 5 of them had it preoperatively and 10 patients had the SVT early post operatively. Twelve patients (8.1%) had atrial junctional rhythm with normal heart rate of which 9 had it early postoperatively. A pacemaker was implanted during follow up period in 6 other patients (4%), for which the major indication was documented symptomatic brady arrhythmia with heart rate less than 50 beats/minute. During midterm follow up, and according to New York Heart Association's (NYHA) functional classes, 141 patients (94.6%) were in class I and II, 8 patients (5.4%) were in class III, 6 of which (75%) actually did not undergo any palliative surgery, and no patients were in class IV.

#### **Discussion**

Many studies emphasize that results of Fontan procedure are steadily improving over the past three decades,<sup>(3,4,10)</sup> however there is considerable late morbidity that is evolving with the longer follow up time. The absence of a ventricular pumping chamber for the pulmonary circulation and the increase in systemic venous pressure are the main haemodynamic features after Fontan procedure.<sup>(11)</sup> Improvements in early survival are in part due to better selection of patients,<sup>(3,4)</sup> patient preparation, optimizing staging of the Fontan procedures,<sup>(3,4,12)</sup> and modifications of the Fontan operation, which included the LT and extra-cardiac connections.<sup>(3,12)</sup>

Our study demonstrates that our results in QAHI with the LT Fontan operation in term of early and midterm complications are excellent and comparable with cardiac centers across the world. The mortality rate in our study is low, and can be attributed to the above mentioned reasons and the selective fenestration of the baffle of the (LT) Fontan operation. The pre operative risk factors that increased the early death or failure of the LT Fontan in our study included: Significant elevation of the mean pulmonary artery pressure (PAP) of mortality

**Table III.** Causes of early mortality

Diagnosis	Mean PAP (mmHg)	Mean VEDP (mmHg)	Death history
1. DORV (small LV)	16	18	Ventricular dysfunction
2. DORV (small LV)	15	17	Ventricular dysfunction
3. DORV (small LV)	18	17	Ventricular dysfunction + (anomalous hepatic veins)
4. Unbalanced A-Vcanal (small LV)	18	20	Ventricular dysfunction (grade II AVVR)
5. DILV	15	15	Sepsis
6. Tricuspid Artesia	17	17	Sepsis
7. Pulmonary atresia & intact septum	20	18	Ventricular dysfunction
8. Unbalanced A-Vcanal (small LV)	15	17	Sepsis
9. DORV (small LV)	16	18	Ventricular dysfunction+(anomalous pulmonary veins)
10. Unbalanced A-Vcanal (small LV)	15	16	Ventricular dysfunction (grade II AVVR)

DORV (double outlet right ventricle), DILV(double inlet left ventricle), AVVR (atrio-ventricular valve regurgitation), PAP( pulmonary artery pressure). VEDP (ventricular end-diastolic pressure)

cases  $16.5 \pm 1.7$  mmHg, the mean ventricular end-diastolic pressure (VEDP)  $17 \pm 1.3$  mmHg with  $p < 0.001$ .

Although the number of patients with right ventricle morphology was relatively small (17 patients out of 159), the death rate was 7 out the 17 patients (44%) which was significantly higher than the mortality cases with left ventricular morphology (3 out of 132 patients) (1.88%) with a  $p=0.003$ . In our analysis also we noticed that the longer stay on CPB ( $127 \pm 4$  vs.  $121 \pm 9$  minutes)  $p= < 0.05$ , the longer the ischemic time ( $51 \pm 5$  vs.  $47 \pm 3$ )  $p= < 0.05$ , the higher the risk of death or Fontan failure, in terms of prolonged ventilatory support and prolonged stay in ICU as was confirmed by many and most studies that were carried on Fontan procedure.<sup>(3)</sup>

Arrhythmias do occur due to the distention of the atrium and the multiple suture lines.<sup>(13)</sup> The incidence of arrhythmias in our study is comparable with other studies ranging from 13.3 to 20.8%; 21 patients (13.3%) had arrhythmia at early postoperative period and 31 (20.8%) patients had it at midterm follow-up, which we believe can be explained by the awareness and meticulous repair of the cardiac surgeons.

Our study had several limitations including; the retrospective nature of the study, the underestimation of the transient and intermittent arrhythmias, the limited availability of serial Holter monitoring studies, the short duration of the follow up mean of 3.2 years (range 2.8-5.1 years).

Despite these limitations we can still conclude that with good selection of patients, the ongoing modifications in Fontan procedure, and the creation of fenestra when needed, provide us with better outcomes. We advise to have more serial ECG, Holter monitoring and electrophysiological studies

for the patients. Long term follow up is still warranted to better evaluate morbidity and mortality of LT Fontan operation.

## References

1. **Burkhart H, Dearani J, Mair D, et al.** The modified Fontan procedure: Early and late results in 132 adult patients. *J Thorac Cardiovasc Surg* 2003; 1252-1258
2. **Stamm C, Frieh I, Mayer J, et al.** Long-term results of the lateral tunnel Fontan operation. *J Thorac Cardiovasc Surg* 2001; 121: 28-41
3. **Azaki A, McCrindle B, Arsdell GV, et al.** Extracardiac conduit versus lateral tunnel cavopulmonary connections at a single institution: Impact on outcomes. *J Thorac Cardiovasc Surg* 2001; 122:1219.
4. **Fontan F, Kirklin JW, Fernandez G, et al.** Outcome after a perfect Fontan operation. *Circulation* 1990; 81: 1520-1536.
5. **Gewilling M, Wyse RK, de Leval MR, Deanfield JE.** Early and late arrhythmias after the Fontan operation: predisposing factors and clinical consequences. *Br Heart J* 1992; 67: 72-79.
6. **de Leval MR, Kilner P, Gewilling 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* 1998; 96: 682-695.
7. **Jonas RA, Castaneda AR.** Modified Fontan procedure: atrial baffle and systemic to pulmonary artery anastomotic techniques. *J Card Surg* 1988; 3: 91-96.
8. **Kumar SP, Rubinstein CS, Simsic JM, et al.** Lateral tunnel versus Extracardiac conduit Fontan procedure: A concurrent comparison. *Elsevier Science Inc* 2003.
9. **Airen B, Sharma R, Choudhary SK, Smruti R. et al.** Univentricular Repair: Is routine fenestration justified. *Elsevier Science Inc* 2000.

10. **Mitchell ME, Ittenbach RF, Gaynor JW, et al.** Intermediate outcomes after the Fontan procedure in the current era. *J Thorac and Cardiovas Surg* 2006; 131:172-180.
11. **Kaulitz R, Luhmer I, Bergmann F, et al.** Sequelae after modified Fontan operation: postoperative haemodynamic data and organ function. *Heart* 1997; 78: 154-159.
12. **Stamm C, Friehs I, Mayer JE, et al.** Long-term results of the lateral tunnel Fontan operation. *J Thorac Cardiovasc Surg* 2001; 121: 28-41.
13. **Bando K, Turrentine MW, Park HJ, et al.** Evolution of the Fontan procedure in a single center. *Elsiever Science Inc* 2000.