

## A retrospective analysis of Fontan procedure and its sequence in tertiary care teaching hospital in southern India

Abhishek Prabhu<sup>1</sup>, Mahadev Damodar Dixit<sup>2</sup>, Mohan Dattatraya Gan<sup>3</sup>, Veeresh Manvi<sup>4</sup>

<sup>1</sup> Postgraduate student, Department of Cardio Vascular and Thoracic Surgery, <sup>2</sup> Professor, Department of Cardio Vascular and Thoracic Surgery, <sup>3</sup> Professor and head, Department of Cardio Vascular and Thoracic Surgery, <sup>4</sup> Paediatric Cardiologist, Jawaharlal Nehru Medical College and Karnataka Lingayat Educational Society's Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi, Karnataka.

Address for correspondence: Dr. Abhishek Prabhu, Postgraduate student, Department of Cardio Vascular and Thoracic Surgery, Jawaharlal Nehru Medical College and Karnataka Lingayat Educational Society's Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi, Karnataka.

### ABSTRACT

**Introduction:** The Fontan operation is usually the final palliative procedure in patients with univentricular heart. The objectives of this study were firstly to describe the clinical and hemodynamic characteristics of a group of patients with univentricular physiology who had previously been palliated with a right modified Blalock Taussig's Shunt, Bidirectional Glenn Shunt and secondly to identify the risk factors that can influence post-operative outcomes after Fontan operation.

**Materials and Methods:** A retrospective analysis of 20 patients who underwent Fontan operation between January 2005 to January 2014 was conducted. Clinical characteristics, catheterization data, type and duration of surgery were reviewed and analysed as predictors of post-operative outcome.

**Results:** We operated 20 cases out of which 14 were

males and 6 were females. The majority of the patients (10) were in the age group of 11-20 years. The diagnosis included double outlet right ventricle (DORV) 5 patients, ventricular septal defect with pulmonary stenosis 5 patients, transposition of great arteries (TGA) 3 patients, tricuspid atresia (TA) 6 patients and double inlet left ventricle with pulmonary stenosis (DILV, PS) 1 patient. Out of these, 4 patients were palliated with right modified Blalock-Taussig (BT) shunt, and 6 patients with Glenn Shunt. The rest were for the direct Fontan procedure. Two patients died during the postoperative period.

**Conclusion:** The performance of hemodynamic study before Fontan operation made it possible to select high risk patients for surgery. Fontan operation showed dramatic improvements in symptoms and saturations.

**Keywords:** Fontan procedure, Univentricular heart, Cardiac catheterization

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### INTRODUCTION

The Fontan operation and its pathway procedures have become the procedure of choice in patients with univentricular heart. In patients born with a single ventricular chamber, the two circulations are in parallel and the systemic and the pulmonary venous blood mix together. In the Fontan circulation, the systemic and pulmonary circulations are divided and are placed in a serial arrangement without the interposition of the ventricle.

The Fontan procedure evolved in the late 1940s with

the recognition that the pulmonary arterial pressure in fish and amphibians was around 25/10 mmHg, and the power acquired for pumping the venous blood through the lungs might be sufficiently supplied by the venous pressure.<sup>1</sup>

Clinically the investigations show the way to the description of various surgical procedures for connecting superior vena cava to right pulmonary artery, called cavopulmonary anastomosis. These procedures were used to treat congenital heart diseases with reduced pulmonary blood flow.

The concept of rerouting systemic venous return directly to the pulmonary arteries for the palliation of congenital heart disease was first described by Fontan and Kreutzer in the early 1970s as an alternate approach for management of tricuspid atresia.<sup>2,3</sup> Since the new description, the evaluation of the “Fontan” has included plentiful surgical advances along with parallel advances in anaesthesia, management, perfusion strategies and intraoperative and postoperative care. In some, these changes have led to substantial reductions in early mortality.<sup>4, 5, 6,7</sup> The objectives of this study were firstly to describe the clinical and hemodynamic characteristics of a group of patients with univentricular physiology who had previously been palliated with a right modified Blalock Taussig's Shunt, Bidirectional Glenn Shunt and secondly to identify risk factors that can influence post-operative outcomes after Fontan operation.

## MATERIALS AND METHODS

This is a retrospective study of 20 patients under going the Fontan operation in our Institution between January-2005 to January-2014. All presented with cyanosis, dyspnoea on exertion and failure to thrive. All were on decongestive therapy. All patients underwent cardiac catheterization prior to the Fontan procedure. Pressures were measured and oximetry was performed in superior vena cava, pulmonary artery, left atrium, systemic ventricle and the aorta. We used these data to calculate the pulmonary flow, systemic and pulmonary vascular resistance based on the published formulas.<sup>8,9</sup> Mean pulmonary arterial pressure was 16mmHg. The pulmonary arteries were confluent. All cases underwent extracardiac Fontan repair on cardiopulmonary bypass using Haemashield graft, diameter ranging from 16-24 mm.

## RESULTS

**Table 1: Age and sex distribution of the patients**

Variables		Number
Age in years	10	8
	11-20	10
	21-30	0
	31-40	0
	41-50	1
	51-60	0
	>60	1
Sex	Male	14
	Female	6

Table 1 shows the age and sex distribution of the patients. The majority of the patients (10) were in the age group of 11-20 years, followed by 8 patients having age less than 10 years. There were 14 males and 6 females.

**Table 2: Baseline diagnosis and previous palliation of the patients**

Variables		Number
Diagnosis	DORV	5
	VSD with PS	5
	TGA	3
	TA	6
	DILV with PS	1
Palliative surgery	BT shunt	4
	Glenn Shunt	6
	Fontan procedure	10

Table 2 describes the baseline diagnosis and previous palliation of the patients. The diagnosis included double outlet right ventricle (DORV) 5 patients, ventricular septal defect with pulmonary stenosis 5 patients, transposition of great arteries (TGA) 3 patients, tricuspid atresia (TA) 6 patients and double inlet left ventricle with pulmonary stenosis (DILV, PS) 1 patient. Out of these, 4 patients were palliated with right modified BT shunt, and 6 patients with Glenn Shunt. The rest were for the direct Fontan procedure.

The mean duration of ventilation was 28 hours. ICU stay was 4 days and all were discharged on the 10<sup>th</sup> postoperative day. The follow up was 90%. Hospital Mortality was two. Six months follow up showed patients with atrial tachycardia responding to amiodarone. Echo showed no conduit thrombosis.

## DISCUSSION

Patients with univentricular physiology who are finally palliated with Fontan surgery form a wide-ranging group. They have different underlying congenital heart diseases and frequently undergo other surgical interventions before the Fontan operation.

The age at which the Fontan procedure is performed is one of the issues which, over the years has been associated to the patient evaluation. Early studies indicated that performing the Fontan operation at a younger age was a major risk factor for mortality.<sup>10,11</sup> Over time, the tendency has been towards performing the intervention earlier without apparent increase in mortality.<sup>12</sup>

The theoretical advantages of earlier intervention include a reduction in the body exposure to cyanosis and its negative effect on cardiac function and partial avoidance of aortopulmonary collateral formation.<sup>13</sup>

Earlier interventions lowered Fontan pressure, however most series have shown higher pressure is associated with increased risk of death and early postoperative failure.<sup>9,11,14</sup>

Extracardiac conduit Fontan is the presently recommended technique for Fontan procedure and that is what we followed.

## CONCLUSION

The performance of hemodynamic study before Fontan operation made it possible to select high risk patients for surgery. Fontan operation showed dramatic improvements in symptoms and saturations.

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