Comparison of outcomes, of patients undergoing the Fontan surgery, before and after the first decade of life.

THESIS

Submitted for the partial fulfillment for the requirement of the degree of

MCh in Cardiovascular and Thoracic Surgery

By

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SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL

SCIENCES AND TECHNOLOGY

THIRUVANANTHAPURAM – 695011, India

Travancore, an erstwhile province of pre-independent India, was ruled by Maharaja Sree Chitra Tirunal Balarama Varma until the country became independent in 1947. The Government of India took over the province after independence and was incorporated into the state of Kerala.

Known for their munificence, the royal family of Travancore considered themselves 'dasas' (servants) of Lord Padmanabha, the reigning deity of Travancore. Interestingly, they wore turban instead of a crown as a mark of respect to the Lord. Their philanthropy finds expression in their countless contributions to the country, then and now.

The Sree Chitra Tirunal Institute for Medical Sciences & Technology (SCTIMST), Thiruvananthapuram is an Institute of National Importance established by an Act of the Indian Parliament. It is an autonomous Institute under the administrative control of the Department of Science and Technology, Government of India.

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DECLARATION

*I hereby declare that this thesis entitled "**Comparison of outcomes, of patients undergoing the Fontan surgery, before and after the first decade of life**" has been prepared by me under the supervision and guidance of **Dr. Sabarinath Menon**, Associate Professor and Professor **Baiju S Dharan.** of the Department of Cardiovascular and Thoracic surgery, SCTIMST, Thiruvananthapuram.*

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INDEX

INTRODUCTION

Patients with functional single-ventricle circulation are among the most complex of those living with congenital heart disease. It is more than 40 years since the first description of the Fontan procedure, and the ideal age to perform the procedure in still unknown. Although most of the centres worldwide are performing the procedure between 2 and 4 years of age, some teams advocate to wait to a later age. The rationale to postpone the procedure is based on the belief that the Fontan circulation provides adequate palliation for a limited time and that postponing this surgery may improve longevity. We therefore intend to retrospectively compare outcomes of patients who have undergone the Fontan procedure in our centre, before and after the first decade of life in order to determine the ideal age to undergo the procedure.

An unoperated single ventricle physiology is characterized by a combination of cyanosis and volume overload. The prolonged duration of the volume overloaded state of the single ventricle prior to surgery often results in profound adverse alterations in myocardial contractility that may impact the functional outcome in Fontan patients. While the initial studies from the single stage surgical era of Fontan procedure indicate that the patients who underwent Fontan repair at an earlier age have better cardiac contractility mechanics and function than the ones who underwent surgery at a later age, the timing of Fontan completion in the contemporary era of staged approach to Fontan repair is controversial.

The long term outcomes of patients who have undergone the Fontan procedure later in life remain even less well defined, in part because of difficulties in obtaining recent follow up in clinical information and presumed lapses in Adult congenital heart diseases care . Current literature shows that significant mortality exists in patients who undergo a Fontan procedure later in life and a standardized medical follow up for this high risk group of patients is difficult. There is now evidence that ventricular function and exercise capacity after Fontan surgery are improved if patients are operated at an earlier age.

REVIEW OF LITERATURE

The Univentricular circulation

A normal circulation has the same design in all mammals: the heart is divided by the septum into two halves, each consisting of an atrium and a ventricle. The right half supports the pulmonary circulation and the left half the systemic circulation. This means that a double pump and two circulations are present, and this is called a biventricular circulation [1]. Some congenital cardiac defects challenge this design. Over a hundred different congenital cardiac defects are described, ranging from 'simple' cardiac defects (for instance an atrial septal defect) to multiple complex congenital cardiac defects. Certain complex congenital cardiac defects have a common denominator: only one ventricle is fully developed and serves both the pulmonary and systemic circulation. The other ventricle is usually underdeveloped or absent. Hence, in these patients, the circulatory anatomy consists of a single pump serving a double circulation. These so-called functional univentricular hearts form 4-5% of the total group of congenital cardiac diseases [2].

This group includes, among others, patients with tricuspid atresia, hypoplastic left heart syndrome, double inlet ventricles, unbalanced atrio ventricular septal defects, and pulmonary atresia with an intact ventricular septum. As a consequence of the functional univentricular morphology, the single ventricle is

volumeoverloaded and continuous mixing of saturated and desaturated blood occurs. Depending on the degree of pulmonary stenosis, these patients have either a pulmonary "overcirculation" and are prone to develop heart failure in early life, or suffer from severe cyanosis and die of profound hypoxemia [3,4] .

The Fontan operation

In 1971/72, both Fontan and Kreutzer in respectively France and Argentina published an exceptional operation to treat the ventricular volume overload or cyanosis of a univentricular heart [5,6]. Their reports described the creation of a direct connection between the right atrium and the pulmonary arteries in patients with tricuspid atresia. This operation created a unique, unphysiologic circulation, in which the functionally single ventricle is used to support the systemic circulation, and the systemic venous return flows "passively", without the help of a ventricular pump, through the pulmonary vascular bed. As a result, the patients are relieved from the chronic cyanosis or ventricular volume-overload, at the expense of living without a subpulmonary ventricle to support the pulmonary blood flow. This unique type of circulation has become known as the Fontan circulation. Over the past decades, the Fontan operation has been subjected to changes in patient selection criteria as well as various modifications in the operative technique. The ten commandments for ideal patient selection, initially proposed by Choussat et al. in 1978, are displayed in table 1 [7].

Table 1. Primary selection criteria for patients with tricuspid atresia for the Fontan operation ('the ten commandments')

1. Minimum age 4 years

2.Sinus rhythm

3.Normal caval drainage

4.Right atrium of normal volume

5.Mean pulmonary artery pressure ≤ 15 mm Hg

6.Pulmonary arterial resistance < 4 U/m2

7.Pulmonary artery to aorta diameter ratio ≥ 0.75

8.Normal ventricular function (ejection fraction > 0.60)

9.Competent left atrioventricular valve

10. No impairing effects of previous shunts

These commandments have gradually been adapted over the years, in particular the age criterion has been modified [7,8]. Furthermore, the classic Fontan operation was performed in patients with tricuspid atresia [6], but gradually more univentricular cardiac defects were referred for a Fontan correction, including double inlet ventricles, pulmonary atresia with intact ventricular septum, hypoplastic left heart syndrome, and other complex congenital cardiac defects that are not suitable for biventricular repair.

These changes in patient selection have resulted in a very heterogeneous group of Fontan patients, in which different cardiac diseases carry different risks and ask for different approaches prior to the Fontan surgery [9-11]. Finally, the surgical techniques have evolved over time. Fontans' classic atriopulmonary connection (APC) involved a valved conduit between atrial tissue and

pulmonary artery. Later on, a direct atriopulmonary connection and a right atrium- to right ventricular connection emerged as a response to the high incidence of valvular dysfunction [12].

In 1987, the first total cavopulmonary connection (TCPC) was performed to provide better streamlining and lower energy loss in the Fontan conduit [13] . The TCPC consisted of an intra-atrial tunnel incorporating the right atrial posterior wall and a Goretex patch or pericardial baffle to tunnel the inferior caval vein to the pulmonary artery. This has become known as the TCPC with a lateral tunnel [13-15]. Furthermore, a lateral tunnel using only autologous tissue, totally created from the right heart auricle, was described [16]. Finally, the TCPC with an extracardiac tunnel was developed, in which the inferior caval flow is directed to the pulmonary artery through a synthetic (PTFE) conduit externally of the heart [17]. The extracardiac tunnel has theoretical advantages in reducing suture lines in the right atrium and relieving it from high systemic pressures, potentially reducing the incidence of arrhythmias [18].

In the early decades of TCPC surgery, the early postoperative mortality was high [19,20]. Rapid changes in ventricular geometry due to volume-unloading during Fontan surgery were thought to alter diastolic function and attribute to the mortality rates. Therefore, a stepwise Fontan procedure was introduced, using a bidirectional Glenn procedure (BDG) to achieve early volume-unloading [21]. With the BDG, the superior caval vein is directly anastomosed to the pulmonary artery. Nowadays, the energetically favorable lateral tunnel and extracardiac conduits are the most commonly applied Fontan techniques. The BDG is usually performed when the patient is around 3 to 6 months of age and can be considered the first stage of the Fontan procedure [21,22].

Characteristics of a Fontan circulation

The common denominator of all Fontan techniques is the pulmonary flow depending on a non-pulsatile driving pressure due to the lack of a sub-pulmonary pump. This so-called passive pulmonary blood flow and the serial (instead of parallel) connection of the pulmonary and systemic circulations result in several adverse characteristics inherent to the Fontan circulation. Firstly, due to the absence of a subpulmonary pump to overcome the pulmonary vascular resistance (PVR), the central venous pressure, together with the muscular and ventilatory pump, becomes the driving force of pulmonary blood flow.

Consequently, central venous pressure increases up to 2 to 4 times of normal [23] and the patients suffer from chronic systemic venous congestion. Secondly, the preload of the single ventricle becomes dependent on the passive pulmonary blood flow [24]. In the Fontan circulation, the ventricular preload is thought to be limited to 60-80% of normal for body surface area, and possibly even less when normalized to ventricular size, with potentially limited ability to increase during exercise [25]. Thirdly, an increased afterload of the heart is caused by the serial arrangement of the systemic and pulmonary circulation, resulting in three resistances: the systemic vascular bed, the Fontan conduit and the pulmonary vascular bed [26,27]. Together, the increased afterload and decreased preload result in a restricted cardiac output, particularly during exercise.

Sequelae of the Fontan circulation

Due to its unphysiologic characteristics, the Fontan circulation is associated with gradual attrition and multiple adverse sequelae. Despite all adjustments in the past decades, the Fontan operation thus remains a palliative procedure and is associated with a reduced life expectancy[8,10,28,29]. This reduced life expectancy is caused by both early/perioperative (within 30 days of Fontan surgery) and late mortality. Various strategies have been tried and tested to improve the early survival after cardiothoracic surgery in general and Fontan patients in particular. In addition to the different operation techniques and changes in patient selection described above, these strategies included changes in the peri-operative management and advances in anesthetic care.

Previous studies investigating survival after the Fontan surgery found that the early and overall survival after the Fontan operation has improved over the past decades, with only a 2-5% early mortality rate in the most recent era[8,10,28-30]. The few reports addressing late mortality show the same trend[29,31] .However, it is important to bear in mind that the more recent Fontan procedures are commonly performed in two stages (first a BDG procedure followed by a Fontan completion). Previous studies on outcome have mostly disregarded the mortality associated with the BDG procedure and the inter-stage period. This might have underestimated the mortality associated with newer Fontan techniques.

With improving early survival rates, long-term sequelae of the abnormal circulation are more commonly observed.These sequelae include (continuation of) impaired exercise tolerance, cardiac dysfunction,restricted pulmonary function,intestinal and hepatic complications and a gradual increase in pulmonary vascular resistance. Eventually, gradual attrition of the circulation can lead to a life-threatening failure of the Fontan circulation.

The peak exercise tolerance, described as peak VO2index, is around 53-65% of predicted in Fontan patients compared to healthy individuals[32-36]. Despite the impaired exercise capacity, most Fontan patients ostensibly live a nearly normal

life and are in New York Heart Association-Functional Class (NYHA-FC) I or II at a mean follow-up of 5 years[10]. Importantly, both NYHA-FC and peak VO2index seem to deteriorate with increasing interval since the Fontan operation[10, 32,34-38]. This has been regarded as indicator of Fontan attrition and associated with increased risk of mortality[35]. The underlying mechanisms for the impaired exercise tolerance in Fontan patients are assumed to be a limited ability to increase cardiac output during exercise and the restricted pulmonary function of Fontan patients.

Secondly, the unphysiologic circumstances of the Fontan circulation affect the cardiac function. The single ventricle in the Fontan circulation is subjected to abnormal loading conditions, including a decreased ventricular preload and increased ventricular afterload. The preload depletion is suggested to be associated with systolic and diastolic ventricular dysfunction, and results in a limited ability to increase stroke volume during exercise[39]. The increased afterload increases workload for the single ventricle and reduces the ventricular efficiency, requiring more power to forward flow[26,27].

Furthermore, Fontan patients are prone to develop arrhythmias due to atrial myocardial scarring from surgery and progressive atrial dilatation when the atrium is exposed to the higher systemic venouspressure[24,34].The progressive atrial dilatation predisposes Fontan patients to atrioventricular valve dysfunction40 and, in combination with the sluggish blood flow and high incidence of coagulation disorders, to a higher incidence of thromboembolic events[41]. Together, the reduced preload, increased afterload, arrhythmias and atrioventricular valve regurgitation all contribute to a limited cardiac output in Fontan patients, leading to potential deterioration over time and eventually Fontan failure[42].

A biomarker to monitor the cardiac function and circulatory performance over time would be extremely valuable in the Fontan circulation. The N-terminal pro natriuretic peptide (NTproBNP), which is released from the cardiac tissue in response to increased wall-stress due to volume- or pressure load, is a potential candidate. In patients with biventricular hearts, NTproBNP is essential in the recognition and monitoring of cardiac dysfunction and congestive heart failure[43]. However, the unique cardiac characteristics in the Fontan circulation hamper the extrapolation of studies on NT-proBNP in biventricular patients to univentricular patients. Therefore, studies aiming at investigating thevalue ofNT-proBNP in the evaluation of cardiac function and circulatory performance of the Fontan circulation are needed.

Thirdly, previous studies have shown that pulmonary function is impaired in almos tall Fontan patients. The forced vital capacity, a measure of the pulmonary volume available for ventilation, is generally 80-90% of normal and the pulmonary diffusion capacity, indicating diffusion capacity for oxygen through the alveolar membrane to the blood vessels, is around 70-80% of normal[44-46.] The reason for the restricted pulmonary function presumably lies in an impaired pulmonary development and repeated thoracic surgeries[45].

Furthermore, patients with a Fontan circulation can develop protein losing enteropathy (PLE), which is a debilitating intestinal complication, occurring with a reported incidence of 3-18% years in late survivors[47-49.] It is caused by a break in the mucosal integrity of the intestinal mucosa, and results in protein loss and nutrient malabsorption[49]. The exact cause of the break in the mucosal integrity is unknown, but inflammation and abnormal elevation in mesenteric vascular resistance due to the chronic systemic venous congestion are thought to play a role[50]. Clinical signs of PLE are ascites, diarrhea, periphereal edema and, when PLE has started prior to adolescence, delayed growth and development. It is one of the most life-threatening complications, with a five-year survival after its onset of less than 60%[48,51.]

Hepatic fibrosis is increasingly recognized in Fontan patients and hepatic cirrhosis is seen in up to 55% of adult Fontan survivors[52-55]. Potential drivers of Fontan associated hepatic disease appear to be the chronic systemic venous congestion, limited cardiac output and thrombo-embolic insults[56,57]. Fontan associated hepatic disease can result in liver failure, gastro-intestinal bleeding, hepatic encephalopathy[52] and even hepatocellular carcinomas, which have an estimated incidence of 1.5-5.0% per year in Fontan patients[53-55]. These adverse hepatic events are associated with significant morbidity and mortality, independent of the time since the Fontan operation[52.] Unfortunately, the evaluation of hepatic fibrosis- and cirrhosis in Fontan patients is difficult, because a liver biopsy, often considered the golden standard[58], is hazardous in Fontan patients due to coagulation disorders and hepatic congestion. One of the potential alternatives for a liver biopsy is the diffusion-weighted magnetic resonance imaging (DWI), which might be promising in Fontan patients due to its ability to distinguish microperfusion components (congestion)from cellular diffusion (associated with hepatic fibrosis/cirrhosis)[59.] However, its relation with hepatic and functional parameters of the Fontan circulation has not yet been described.

Finally, the pulmonary vascular resistance is an important tissue in the Fontan circulation. A low pulmonary vascular resistance (PVR) is essential in the Fontan circulation for the systemic venous return to flow passively through the pulmonary vasculature. Even small increases in PVR will significantly reduce the cardiac preload and cardiac output[8,60,61,] and will further increase the central venous pressure. An increase in pulmonary vascular resistance might result from adverse pulmonary vascular remodeling. There are two potential underlying mechanisms of pulmonary vascular remodeling in the Fontan circulation. The first mechanism includes microemboli in the pulmonary vascular bed which occur with an increased incidence in Fontan patients due to the hypercoagulable state[62.] Secondly, the non-pulsatile pulmonary flow results in changes in circumferential strain and shear stress in the vessel, and is demonstrated to cause endothelial dysfunction, vascular remodeling and an

increase in PVR in animal models[63,64]. Studies exploring the changes in pulmonary vasculature have primarily focused on the peri-operative period, whereas data on the long term changes in pulmonary vasculature are currently lacking.

Despite the restricted exercise tolerance and other adverse sequelae, previous reports have shown that patients with a Fontan operation have a relatively good quality of life[10.] However, they do seem to score lower on physical functioning, mental health and general health than their healthy peers[65-67], and worry about their health, job employment, ability to work and living independently[68]. Previous reports pointed out that worse quality of life in Fontan patients is related to respiratory problems and protein losing enteropathy, but not with exercise tolerance[67,69-71].

One important aspect of quality of life is sexual wellbeing. Previous studies demonstrated patients with congenital cardiac diseases to have increased concerns regarding fertility, inheritability and pregnancy, may experience a broad range of sexual problems and might lag behind in psychosexual development[72,73]. Although these studies have included a variety of congenital cardiac disease, no patients with a Fontan circulation were included. This is unfortunate because the restricted cardiac output and exercise tolerance, the chronic systemic venous congestion, the operations at a young age, frequent hospital visits, previously reported menstrual cycle disorders and fertility problems[74], and impaired life expectancy might affect patients' development and wellbeing.

Eventually, dysfunction of several end-organs can occur and the Fontan patient enters a clinical state called "Fontan failure"[75.] The pathophysiology of Fontan failure is distinct from the classical congestive heart failure in biventricular hearts; In biventricular hearts, failure is mainly characterized by a decreased contractility of the heart[76,77], whereas in univentricular hearts various factors play a role, including hepatic, gastro-intestinal, pulmonary or cardiac

complications. A failing Fontan, with an estimated incidence of 2-13%, has a very poor prognosis and therapeutic options are few[40,78]. Nowadays, three surgical options are embraced worldwide: Fontan takedown[79], Fontan conversion from an atriopulmonary connection to an energetically more favorable connection (i.e. lateral tunnel or extra-cardiac conduit)[80,] and heart transplantation (HTX)[81,82].

A Fontan takedown is usually performed as a bailout option for early Fontan failure, which occurs after 2-6% of the Fontan operations[83,84]. An Australian- and New Zealand series reported a 75% hospital survival after Fontan takedown surgery[85]. Early mortality rates after Fontan conversion or a heart transplantation vary substantially: ranging from 0.9% to 13% for Fontan conversion[80,86], and 4-30% for heart transplantation[87,88]. Very limited data is available on long-term outcome of failing Fontan surgery and there are no reports comparing survival after the different surgical options for the failing Fontan circulation.

AIM OF THE STUDY

1. To retrospectively compare outcomes, of patients who have undergone the Fontan procedure in our centre, before and after the first decade of life.

2. To evaluate factors determining the outcome of the procedure.

MATERIALS AND METHODS

Study group: It is a retrospective comparative study of 53 patients who underwent the Fontan Surgery between January 2013 to December 2017 at Sree Chitra Tirunal institute for medical sciences and technology, Thiruvananthapuram. Review records of all patients were analysed and the cohort was subdivided into two groups of less than 10 years and more than 10 years of age.

OBSERVATIONS AND RESULTS

Demographic details

A total of 53 patients underwent Fontan surgery within the study period. 41.5% (n =22) belonged to the less than 10 year age group(Group A) and 58.5% (n = 31) were more than 10 years of age(Group B) (Fig 1.1 and 1.2).

Males composed 45.3% (n =29) and females 54.7% (n =24) of the study population. The most common morphological diagnosis was Tricuspid atresia (30.2%) followed by Double outlet right ventricle (24.5%) Fig 3.

Pre operative characteristics

7.5 %(n=4) underwent a single stage Fontan surgery without prior palliation (Fig 4.1). 20.8 %(n=11) underwent 2 palliative procedures and 71.7 %(n=38) had 1 palliative procedure prior to the completion Fontan(Fig 4.2). Blalock Taussig shunt was performed in 13.2 %(n=7), bidirectional Glenn shunt in 92.5 %(n= 49) prior to Fontan(Fig 4.3).

The study group had a mean resting oxygen saturation of 79.45% pre-operatively (Fig 5.1). Pre operative ventricular function assessed by echocardiogram showed good ventricular function in 90.6 %(n=48) of the patients, 7.5 %(n=4) had Mild Ventricular dysfunction and 1.9 %(n=1) had severe ventricular dysfunction (Fig5.2).

Atrioventricular valve regurgitation was Nil in 45.3 %(n=24), Mild 43.4%(n=23), Moderate 11.3%(n=6)(Fig 5.3). Pre operative NYHA functional class assessment showed 5.7 %(n=3) to have Class 1, 75.4 %(n=40) Class2 and 18.9 %(n=10) Class 3 symptoms (Fig 6).

Major aortopulmonary collateral (MAPCA) coiling prior to Fontan procedure was performed in 3 patients. 1 patient presented with haemoptysis 1 year following surgery, underwent MAPCA coiling, and subsequently 3 years later had recurrent haemoptysis for which 5 MAPCA'S were coiled. Another patient in the group underwent MAPCA coiling pre and post Fontan procedure. Pumonary

arteriovenous malformation coiling was performed for one patient, 1 month following Fontan.

ADDITIONAL PROCEDURES

Concomittant procedures at the time of Fontan were performed for patients with significant AVVR. 4 patients underwent Tricuspid valve repair and 3 patients had mitral valve repair using posterior mitral annuloplasty technique. 1 patient with prior history of infective endocardidits, had vegetations on the aortic valve and underwent an Aortic valve replacement with a mechanical valve.

Pulmonary artery was augmented with pericardial patch in 8 patients. 5(Left), 1 (bilateral), 1(Right) 1(Confluence).

1 patient underwent radiofrequency ablation pre operative ventricular premature complexes' in the RVOT. During Fontan RVOT ablation was performed.

INTRA OPERATIVE CHARACTERISTICS

Extra cardiac technique was performed in all patients. 4 patients underwent a single stage Fontan with no prior palliation.

TECHNIQUE OF EXTRA CARDIAC FONTAN

Exposure through standard median sternotomy, adhesions are dissected in order to completely mobilize previous SVC and to RPA anastamosis. Pulmonary artery system is dissected from right upper and lower arterial branch junction to pulmonary trunk, working from right to left under the ascending aorta (Fig A)

Patient is heparinised, central portion of pulmonary artery is isolated by placing vascular clamps, on the RPA close to the previous Glenn shunt and on the central LPA. Inferior surface of RPA and central pulmonary artery is incised over an appropriate length to accept the circumference of a sharply beveled 20-mm-diameter PTFE graft tube graft (Fig.A)

Anastamosis between graft and pulmonary artery is performed using nonabsorbable fine monofilament suture and a continous technique. Single clamp

placed on midportion of tube graft, allowing re-establishment of bidirectional SVC- to- pulmonary artery flow. PTFE graft is tailored to appropriate length for end to end graft to IVC (Fig.A)

The ascending aorta and IVC are cannulated, two vascular clamps are placed across the IVC, at the cavoatrial junction and at the level of IVC cannulation. If the femoral vein is used for lower body venous cannulation, the clamp on IVC is placed at the level of the diaphragm. IVC is transected between clamps, IVC remnant on right atrium is oversewn with a nonabsorbable monofilament suture. PTFE tube is connected end to end to diaphragmatic component of IVC using nonabsorbable fine monofilament suture using a continous technique (Fig.B)

Appropriate size fenestration (usually 4mm) is punched in the midpoint of the PTFE conduit. An adjacent incision is made in the right atrial wall. The atrial tissue is sutured to the external wall of the PTFE baffle several millimeters from the fenestration itself (Fig.C).

Fenestration was performed in 75.5% (n=40) and most common size employed was 4mm(39.6%)(n=21) (Fig7.1,7.2). Patients requiring reintervention after surgery were 13.2%(n=7), most commonly for mediastinal bleeding. 7.8%(n=4) had post operative arrhythmias and 1 patient required a permanent pacemaker insertion for complete heart block(Fig 8.3, 8.4). There was no incidence of in-hospital mortality.

POST OPERATIVE CHARACTERISTICS

These outcome variables were assessed during the follow up visit in the outpatient department, one year from the date of surgery. Majority of the patients were in NYHA Class 1 functional class, 60.4 %(n=32) (Fig 9). Post operative echocardiogram showed good LV function in 86.8%(n=46) and Mild AVVR in 50.9%(n=27) (Fig 9, 10).

COMPLICATIONS

Right MCA stroke was diagnosed in 1 patient on post op day 1, he was managed conservatively. 1 patient developed a pseudoaneurysm of the right femoral artery (cannulation site) which was repaired. 1 patient developed acute kidney injury which was managed conservatively.

1 patient who underwent a high risk Fontan with fenestration had significant post operative pleural drainage, cath showed residual LPA stenosis, high PA pressures and multiple MAPCA'S.LPA plasty and MAPCA coiling was peformed in the same setting. The patient presented 3 years following surgery with protein losing enteropathy.

The following re-interventions were performed post Fontan surgery.3 patients were re-explored for mediastinal bleeding. 1 patient underwent decortication for left lung empyema 1 month later. 1 patient was re-explored for high fontan pressures immediate post op, for which the fenestration was enlarged from 4mm to 6mm.

1 patient was readmitted 1 month following surgery for right pleural effusion, treated with inter costal drainage. 1 patient developed atrial tachycardia 2 years post Fontan, treated with direct cardioversion, on follow up continued to have atrial ectopic rhythm.

PATIENT CHARACTERISTICS

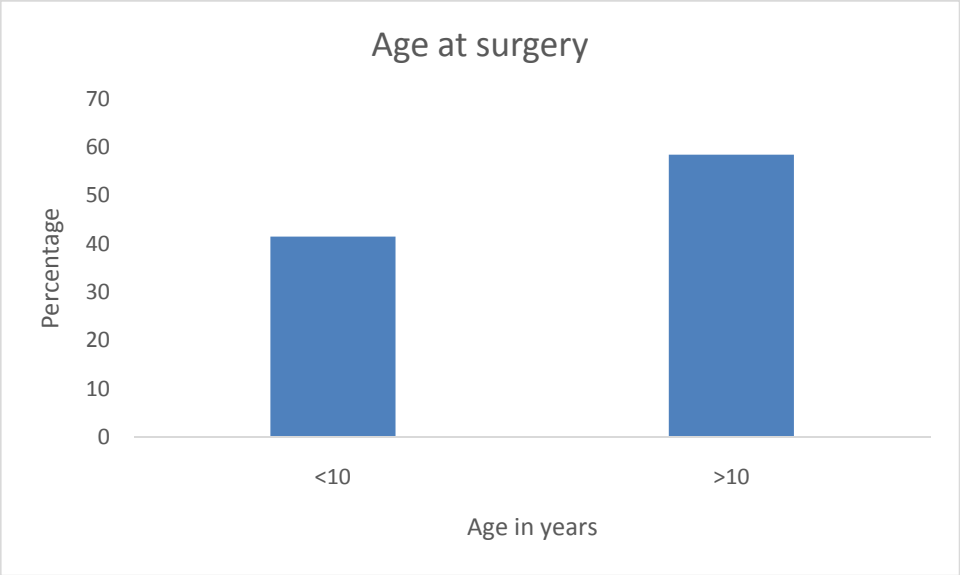

Fig 1.1

Age at surgery	Frequency	Percent
<10	22	41.5
>10	31	58.5
Total	53	100.0

Fig 1.2

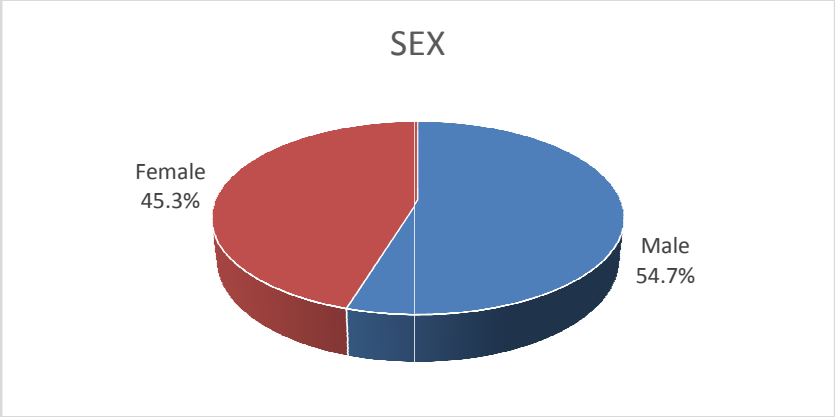

Fig.2

PRIMARY DIAGNOSIS

PRIMARY MORPHOLOGY	Frequency	Percent
DILV	5	9.4
DORV	13	24.5
DTGA	9	17.0
PA	4	7.5
SV	4	7.5
TA	16	30.2
CCTGA	1	1.9
UAVCD	1	1.9
Total	53	100.0

Fig.3

DILV- Double inlet left ventricle, DORV- Double outlet right ventricle, DTGA – Dextroposed Transposition of great arteries ,PA – Pulmonary atresia, SV – Single ventricle of LV or RV morphology, TA- Tricuspid atresia ,CCTGA – Congenitally corrected Transposition of great arteries, UAVCD – Unbalanced Atrioventricular canal defect.

PRE OPERATIVE CHARACTERISTICS

ANY PRIOR PALLIATION	Frequency	Percent
Yes	49	92.5
No	4	7.5
Total	53	100.0

Fig 4.1

NUMBER OF PRIOR PALLIATION	Frequency	Percent
0	4	7.5
1	38	71.7
2	11	20.8
Total	53	100.0

Fig 4.2

PRIOR BT SHUNT	Frequency	Percent
Yes	7	13.2
No	46	86.8
Total	53	100.0

Fig 4.3

	N	O_2 Saturation (%)			
		Minimum	Maximum	Mean	sd
Before surgery	53	52	92	79.45	6.14
After surgery	51	87	98	94.04	2.720

Fig 5.1

PRE OPERATIVE VENTRICULAR FUNCTION ASSESSMENT BASED ON EJECTION FRACTION

VENTRICULAR FUNCTION (PRE OP)	Frequency	Percent
Severe LV dysfunction	1	1.9
Mild LV dysfunction	4	7.5
Good LV function	48	90.6
Total	53	100.0

Fig 5.2

Atrioventricular valve regurgitation assessment on pre operative echocardiogram

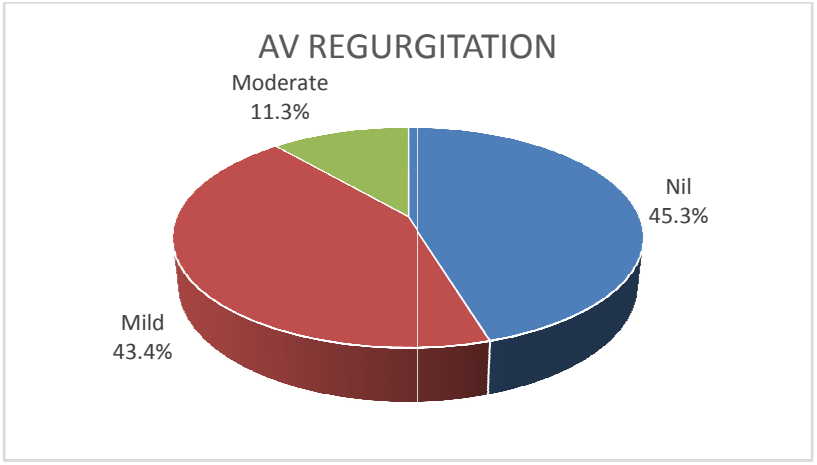

Fig 5.3

Pre operative NYHA class

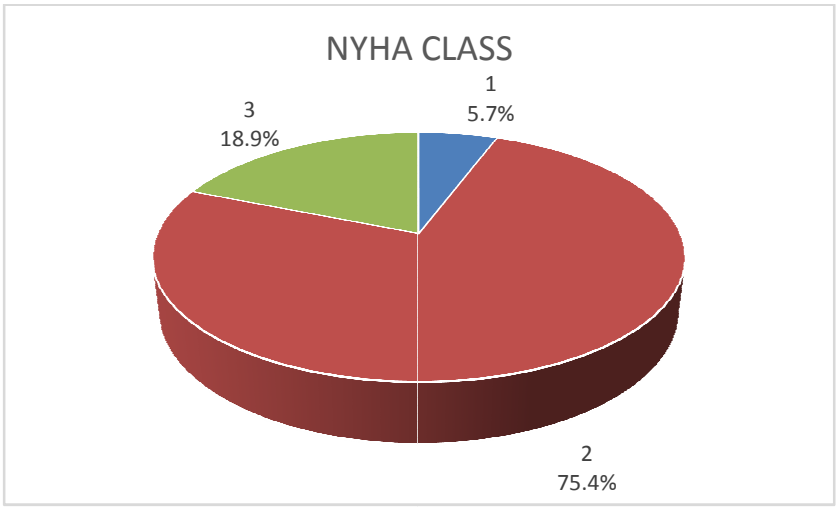

Fig. 6

INTRA OPERATIVE CHARACTERISTICS

FENESTRATION	Frequency	Percent
Yes	40	75.5
No	13	24.5
Total	53	100.0

Fig7.1

FENESTRATION	Frequency	Percent
No	13	24.5
3.50 mm	1	1.9
4.00 mm	21	39.6
4.50 mm	7	13.2
5.00 mm	8	15.1
6.00 mm	3	5.7
Total	53	100.0

Fig7.2

AVVR REPAIR	Frequency	Percent
TRICUSPID	4	7.54
MITRAL	3	5.6
AORTIC VALVE REPLACEMENT	1	1.88
NIL	45	84.9
Total	53	100.0

Fig7.3

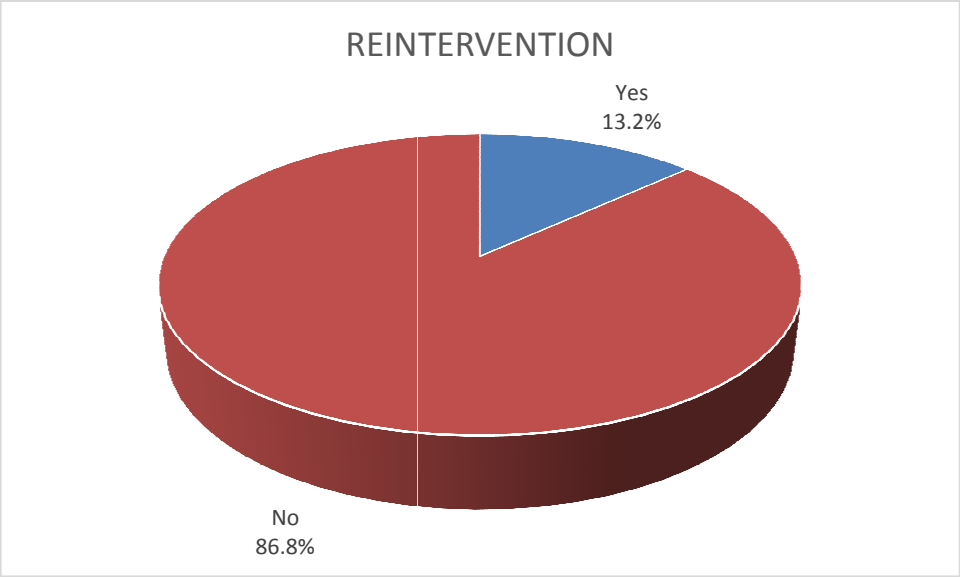

Fig 8.1

IN HOSPITAL MORTALITY	Frequency	Percent
No	53	0.0

Fig 8.2

PERMANENT PACEMAKER INSERTION	Frequency	Percent
Yes	1	2.0
No	50	98.0
Total	51	100.0

Fig 8.3

ARRHYTHMIAS	Frequency	Percent
Yes	4	7.8
No	47	92.2
Total	51	100.0

Fig 8.4

POST OPERATIVE CHARACTERISTICS

NYHA CLASS ON FOLLOW UP AT 1 YEAR	Frequency	Percent
1	32	60.4
2	18	34.0
3	1	1.9
Lost follow up	2	3.8
Total	53	100.0

Fig. 9

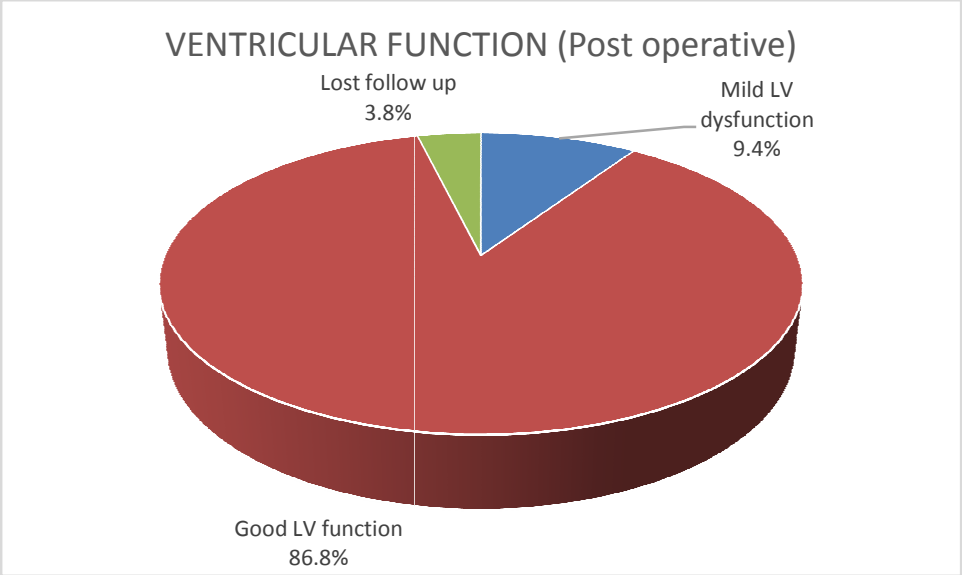

Fig. 10

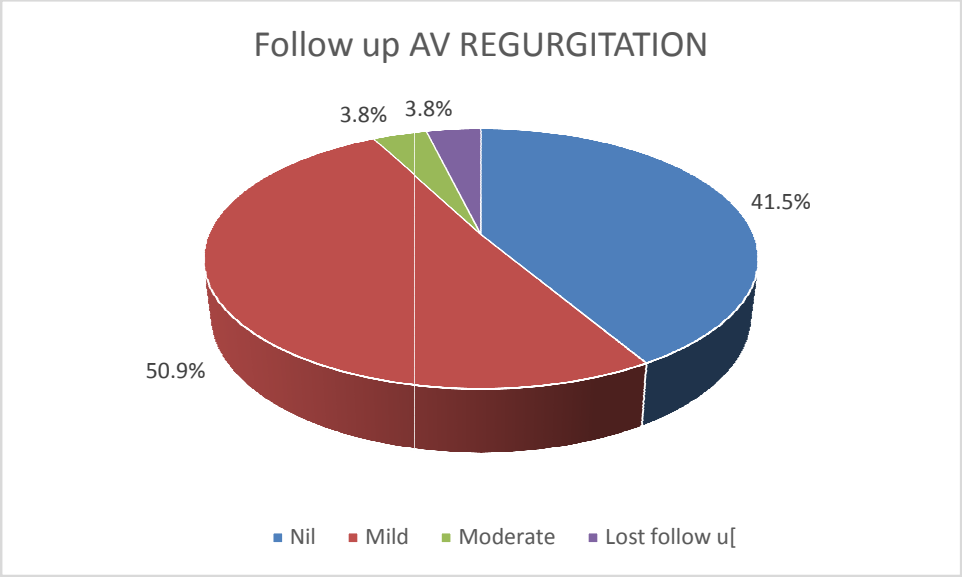

Fig. 11

Follow up AV REGURGITATION	Frequency	Percent
Nil	22	41.5
Mild	27	50.9
Moderate	2	3.8
Lost to follow up	2	3.8
Total	53	100.0

Fig. 12

COMPARISON OF VARIOUS DEMOGRAPHIC, PRE AND POST OP VARIABLES BETWEEN THE TWO AGE GROUPS

	Age				Total	
	<10		>10			
Sex	N	%	N	%	N	%
Male	14	63.6	15	48.4	29	54.7
Female	8	36.4	16	51.6	24	45.3
Total	22	100.0	31	100.0	53	100.0

χ^2 =1.208 df=1 p=0.272

Fig. 13

VENTRICULAR FUNCTION (Pre OP)	Age				Total	
	<10		>10			
	N	%	N	%	N	%
Severe dysfunction	0	0.0	1	3.2	1	1.9
Mild dysfunction	1	4.5	3	9.7	4	7.5
Good function	21	95.5	27	87.1	48	90.6
Total	22	100.0	31	100.0	53	100.0

χ^2 =1.258 df=2 p=0.533

Fig. 14

NYHA class (follow up)	Age				Total	
	<10		>10			
	N	%	N	%	N	%
1	18	81.8	14	48.3	32	62.7
2	4	18.2	14	48.3	18	35.3
3	0	0.0	1	3.4	1	2.0
Total	22	100.0	29	100.0	51	100.0

χ^2 =6.212 df=2 p=0.045

Fig. 15

VENTRICULAR FUNCTION (post OP)	Age				Total	
	<10		>10			
	N	%	N	%	N	%
Mild dysfunction	0	0.0	5	17.2	5	9.8
Good function	22	100.0	24	82.8	46	90.2
Total	22	100.0	29	100.0	51	100.0

χ^2 =4.205 df=1 p=0.040

Fig. 16

Tests of Normality						
	Kolmogorov-Smirnov[a]			Shapiro-Wilk		
	Statistic	df	Sig.	Statistic	df	Sig.
MEAN PA PRESSURE(PRE OP)	.186	42	.001	.944	42	.041
CPB(MIN)	.151	42	.018	.937	42	.022
AOCXCL(MIN)	.198	42	.000	.856	42	.000
INOTROPE SUPPORT DURATION(HOURS)	.127	42	.086	.934	42	.018
TIME TO EXTUBATION(HOURS)	.193	42	.000	.878	42	.000
ICU STAY(HOURS)	.157	42	.010	.893	42	.001
PLEURAL DRAINAGE(DAYS)	.175	42	.002	.751	42	.000
HOSPITAL STAY(DAYS)	.169	42	.004	.824	42	.000
FOLLOW UP 02 SAT(%)	.142	42	.032	.940	42	.030

Fig. 16

Age	MEAN PA PRESSURE			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	12	9	13.25	0.603
>10	12	10	13	

Fig. 17

Age	CPB(MIN)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	162	142.25	213	0.014
>10	215	175	289	

Fig. 18

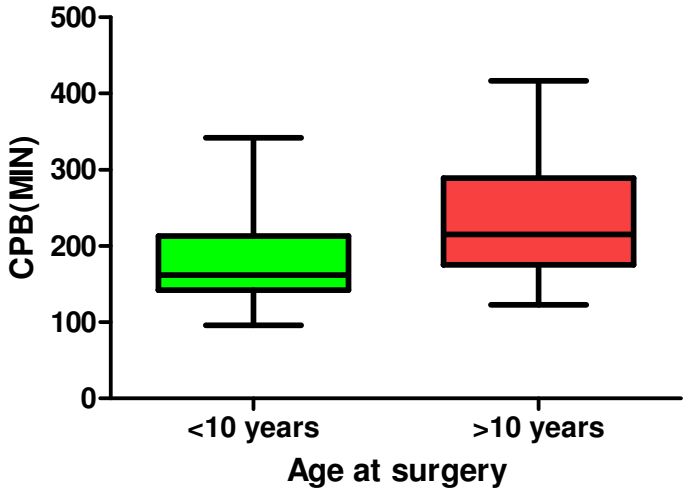

Fig. 19

Age	AOCXCL(MIN)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	41	24	62	0.414
>10	40	30.5	110.5	

Fig. 20

Age	INOTROPE SUPPORT DURATION(HOURS)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	60	48	69	0.229
>10	50	40	66	

Fig. 21

Age	TIME TO EXTUBATION(HOURS)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	6	6	10.75	0.074
>10	8	7	12	

Fig. 22

Age	MEAN PA PRESSURE			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	99.00	83.75	119.25	0.404
>10	96.00	84.00	110.00	

Fig. 23

Age	PLEURAL DRAINAGE(DAYS)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	8	4.75	13.5	0.430
>10	9	6	15	

Fig. 24

Age	HOSPITAL STAY(DAYS)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	15.5	12.75	24	0.338
>10	15	10	21	

Fig. 25

Age	FOLLOW UP 02 SAT			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	95	92.75	96.25	0.204
>10	94	92	95	

Fig. 26

Age	Duration between Diagnosis and Fontan (months)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	10.5	7.0	17.3	0.011
>10	23.0	11.0	59.0	

Fig. 27

Age	Duration between First procedure and Fontan (months)			Mann-Whitney test _ p
	Median	Q1	Q3	
<10	89.0	75.8	98.3	<0.001
>10	156.0	108.0	180.0	

Fig. 28

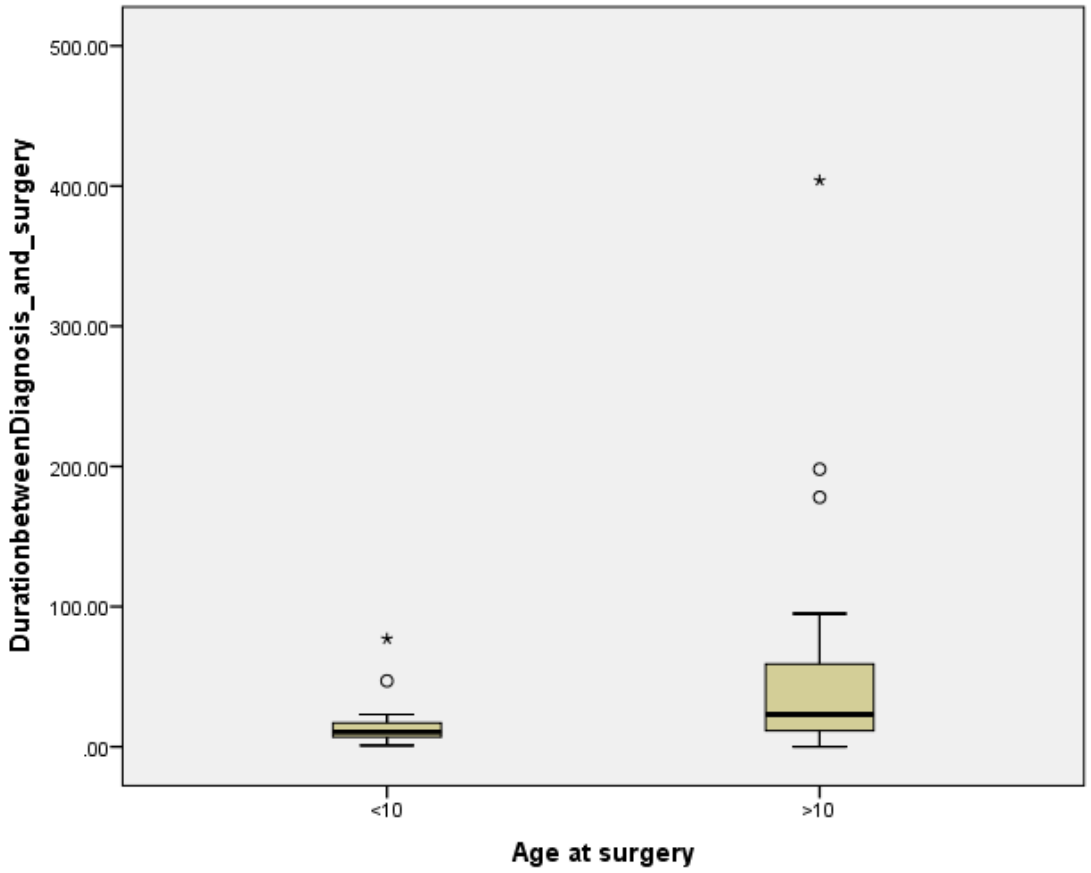

Fig. 29

Box plot diagram describing duration between age at diagnosis and age at surgery, according to age at surgery. The lower and upper end of the whisker represents minimum and maximum duration respectively. Lower border of the box represents 25th percentile and upper border of the box represents 75th percentile. Middle horizontal line represents median duration.

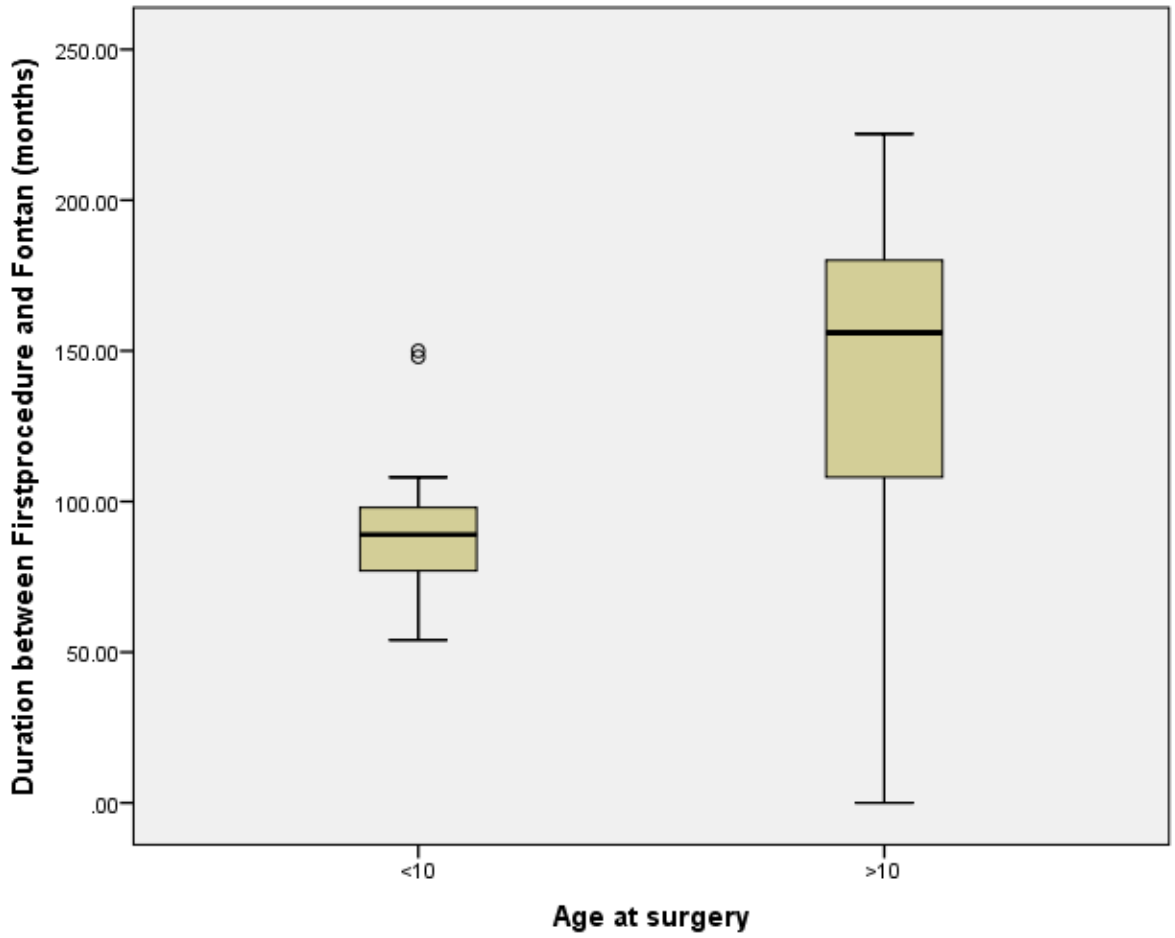

Fig. 30

Box plot diagram describing Duration between First procedure and Fontan According to age at surgery. The lower and upper end of the whisker represents minimum and maximum duration respectively. Lower border of the box represents 25th percentile and upper border of the box represents 75th percentile. Middle horizontal line represents median duration.

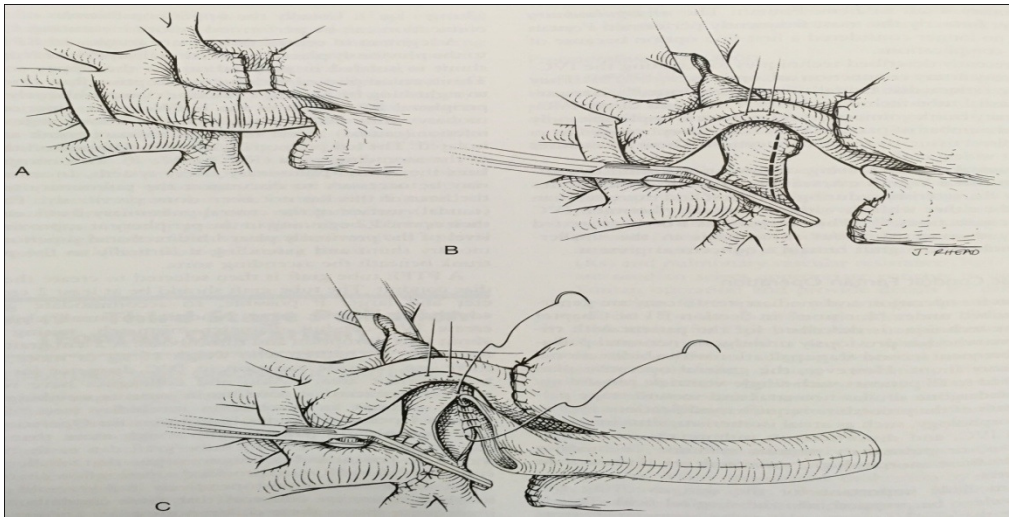

Fig. A

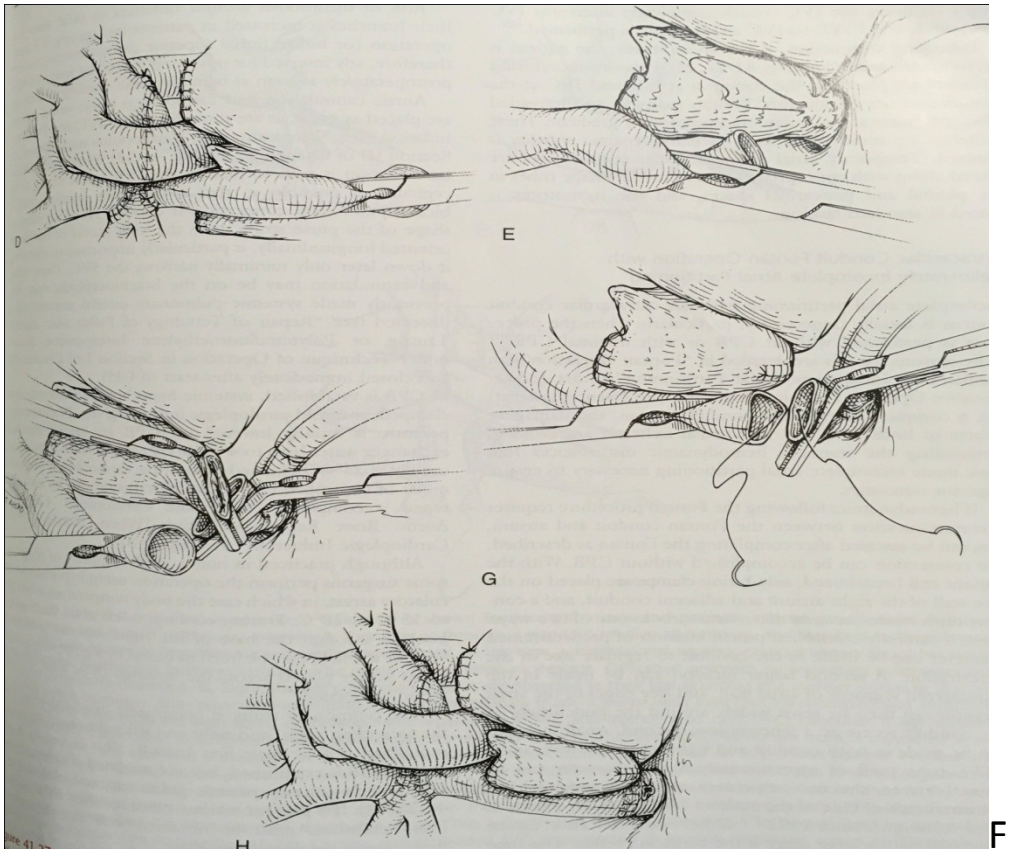

Fig. B

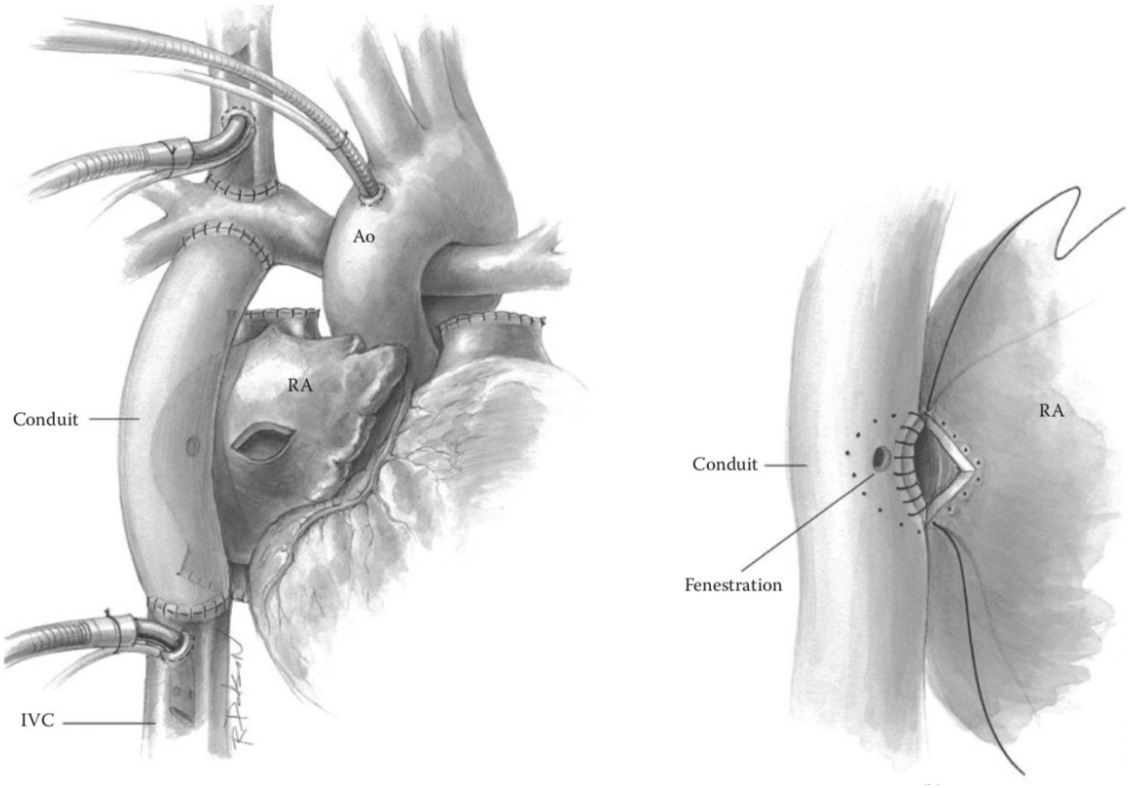

Fig. C

DISCUSSION

As per the current evidence the ideal age to perform Fontan is 4-5 years[89].Unfortunately in most developing nations surgery is delayed most often, beyond first decade. In India the prevalence of diseases of the heart in newborn is 19.14 per 1000 individuals and most children undergo Fontan at 14 years [90].

Delay in Fontan procedure results in sustained exposure to cyanosis and persistent overload (volume) of the single ventricle. This causes ventricular fibrosis and hypertrophy leading to dysfunction both systolic and diastolic. These patients also have increased collaterals and higher pulmonary artery pressures.

In our study population, 5.7 % (n=3) were in NYHA class 1, 75.5 %(n=40) in class 2 and 18.9 %(n=10) in class 3. Post operatively, in group A, 81.8 %(n=18) were in class 1 compared to 48.3 %(n=14) in group B. The poorer functional class in group B was significant ($p<0.045$) (fig 15).

95.5 %(n=21) in group A had good ventricular function, 4.5 %(n=1) mild ventricular dysfunction prior to Fontan procedure. In group B only 87.1 %(n=27) had good ventricular function prior to Fontan procedure. 9.7 %(n=3) had mild dysfunction and 3.2 %(n=1) severe dysfunction. This difference being significant, p=0.533. Post Fontan procedure none in group A had ventricular dysfunction as compared to 17.2 %(n=5) in group B with mild ventricular dysfunction. This was statistically significant, p=0.040(fig. 16).

Lytrivi I and colleagues when studying the bearing of age at completion of Fontan on functional status and ventricular function, showed improvement in function of the ventricles and exercise capacity if patients are operated at an earlier age [91]. Forsdick, Victoria et al following review of their regional experience showed that children operated after 7 years had a higher risk of late mortality and failing of the fontan later [92]. Our results suggest, delay in performing Fontan, would impact the functional status and ventricular function, thus subjecting these patients to higher risk of late Fontan failure.

The mean pre operative PA pressure when compared (Group A=11.2, Group B=11.9) was not significant (fig 23). Duration of CPB was 162 minutes(median) in group A compared to 215 minutes(median) group B, this being significant p=0.014 (fig 18). The prolonged cardiopulmonary bypass times could be due to more time

required for dissection, which is also complicated by presence of numerous collaterals.

We had no incidence of in-hospital mortality. Mortality rates between 7%-14% for Fontan in older age groups have been reported [93]. Low output syndromes, higher pulmonary vascular resistance, and intractable arrhythmia being implicated as causes of death [94.] Mean PA pressure >15mmhg, moderate AVVR, pulmonary artery abnormalities requiring intervention, ejection fraction of <40% have been reported increased mortality risk factors [95].

Concomittant procedures at the time of Fontan were performed for 8 patients with significant AVVR. Pulmonary artery was augmented with pericardial patch in 8 patients. 5(Left), 1 (bilateral), 1(Right) 1(Confluence). 1 patient underwent radiofrequency ablation for pre operative ventricular premature complexes' in the RVOT. During Fontan RVOT ablation was performed. 1 patient who underwent a high risk Fontan presented 3 years later with protein losing enteropathy.

4 patients (all belonging to group B) received a Single stage Fontan. Their outcomes were not different or statistically significant when compared to the staged Fontan group. 75.5 %(n=40) of the patients received a fenestration which could explain the finding of no significant difference in pleural drainage between the two study groups (fig 7.1). Primary Fontan is known to cause prolonged pleural drainage secondary to the sudden increase in pulmonary blood flow culminating in lymphatic congestion.

One single stage Fontan patient had drainage(pleural) for 13 days when compared to mean of 11 days for others. Gates et al had similar pleural effusion rates in adults and children [93]. Talwar et al studied patients who underwent primary fontan after the first decade and recommended staging the Fontan in patients with preoperative ventricular dysfunction, AVVR, preoperative NYHA class 3 and above, mean PA pressure >15mmhg to reduce morbidity and mortality (Early) [96].

Valente et al had survival rates of 83%, 71%, 66% at 5,10,15 years of follow up in adults and adolescents undergoing primary Fontan, with a prolonged ICU stay being a important mortality predictor [97]. In our study we found no difference ICU or hospital stay duration, p = 0.33 (fig.25).

Salazar and colleagues had similar early post operative outcomes with or without fenestration in both the groups. They found, the period to extubation was prolonged in the fenestrated group and concluded that fenestration to be reserved for high risk Fontan only [98]. Our Fontan patients, operated until 2016 received fenestrations and since then fenestration is reserved for highly selective, high risk Fontan procedures.

With age the systemic to collateral flow (pulmonary) increases. This would hinder optimal Fontan circulation. In young adults who present late, surgery may improve their late survival.

LIMITATIONS OF THE STUDY

The overall sample size was relatively small, this complicated the tests of association and also limited the extent of our conclusions.

CONCLUSION

Completion of Fontan procedure should ideally by performed in the first decade of life, preferably between 4-6 years of age. Delay in Fontan procedure beyond the first decade of life results in higher incidence of ventricular dysfunction and poor functional status and could potentially subject them to higher risk of late Fontan failure.

Early recognition and avoiding inter stage delay between the palliative procedures would possibly improve the results of the Fontan procedure. Single stage Fontan in carefully selected patients is safe, with good early results.

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Technical Advisory Committee (Clinical Studies)

SREE CHITRA TIRUNAL INSTITUTE FOR MEDICAL SCIENCES & TECHNOLOGY
THIRUVANANTHAPURAM – 695011, INDIA

TAC Registration No: SCT-/S/2018/733 **Date: 29.05.2018**

***Project title*: COMPARISON OF OUTCOMES OF PATIENTS WHO UNDERWENT THE FONTAN OPERATION, BEFORE AND AFTER THE FIRST DECADE OF LIFE**

Principal Investigator:	
Dr. Pantosh Bajal ,Resident, Department of CVTS, SCTIMST	Degree: MBBS M S
Co-Principal Investigator(s)	
Dr. Sabarinath Menon, Associate Professor, Department of CVTS, SCTIMST	Degree: MCh
Dr. Baiju.S.Dharan, Professor, Department of CVTS, SCTIMST	Degree: MCh
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Dr. Sivasankaran. S , Professor, Department of Cardiology, SCTIMST	Degree: MD DM
Dr. Deepa.S.Kumar, Assistant Professor, Department of Cardiology, SCTIMST	Degree: MD DM
Dr. Arun Gopalakrishnan, Assistant Professor, Department of Cardiology, SCTIMST	Degree: MD DM

Members who participated in the TAC meeting on 19/05/2018

Dr. Rupa Sreedhar (Chairperson)
Dr. Prasantakumar Dash
Dr. Sanjay G
Dr. Krishna Kumar K
Dr. Sankara Sarma P
Dr. Sylaja PN
Dr. Ashalatha. R
Dr. Bijulal S
Dr. Jayadevan ER
Dr. Syam K
Dr. Varghese T. Panicker
Dr. K. Shivakumar (Member Secretary)

Dr. Rupa Sreedhar, Dr. Syam K, Dr. Sylaja PN, Dr. Prasantakumar Dash, Dr. Varghese T. Panicker and Dr. Ashalatha. R stayed away from the proceedings when the projects in which they are involved as investigator were discussed (#736,737, 738, 740, 741,743,744, 746, 749, 752).

Risk Classification of the project (Minimum/ Moderate/ High): Minimum
Requirement of DSMB: No
Recommended members of DSMB: Not applicable

Recommendations of TAC:
Recommended for consideration of IEC in the light of the responses received from the Investigator
The PI may note that there can be no additions / alterations in the documents approved by TAC when they are submitted to the IEC.

Signature of the Member Secretary, TAC (Clinical Studies)

Note for IEC
Copy of the investigator's responses to questions/suggestions from TAC is attached (Appendix-1).

श्री चित्रा तिरुनाल आयुर्विज्ञान और प्रौद्योगिकी संस्थान, त्रिवेन्द्रम
तिरुवनन्तपुरम - ६९५०११, केरल, इंडिया

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Institutional Ethics Committee

(IEC Regn No. ECR/189/Inst/KL/2013/RR-16)

SCT/IEC/1228/JUNE-2018

19.06.2018

Dr. Paritosh Ballal
Resident
Department of CVTS
SCTIMST, Thiruvananthapuram

Dear Dr. Paritosh Ballal,

Thank you for submitting documents related to your proposal titled "COMPARISON OF OUTCOMES OF PATIENTS WHO UNDERWENT THE FONTAN OPERATION, BEFORE AND AFTER THE FIRST DECADE OF LIFE" *(IEC/1228)* to the IEC for review.

List of documents:

1. Covering letter addressed to the Chairman, IEC, SCTIMST with check list
2. TAC Approval Letter
3. IEC Application Form
4. Project Proposal
5. CV of Principal Investigator and Co-Principal Investigators

IEC Recommendations

1. Please envisage appropriate outcome variables and some statistical analysis that will enable comparisons with respect to the two time periods identified.
2. The variables sought could be age at diagnosis, age at surgery and age at various outcomes of interest
3. Page no. 20 – the name of the drug is mis-spelt (Dobutamine)
4. There may be a need for telephonic interview and consent to determine some of the outcomes of interest such as mortality and survival time.
5. Provide TCMC registration for all researchers involved

One set of all the documents including those revised may be submitted. The covering letter should indicate the revisions made.

Sincerely,

Mala Ramanathan
Member Secretary, IEC

Plagiarism Checker X Originality Report

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Remarks: Low Plagiarism Detected - Your Document needs Optional Improvement.

Data collection proforma

Preoperative Variables and Diagnoses

Characteristic

Age (Age at diagnosis)

Sex

Primary Diagnosis

NYHA functional class: Class 1

Class 2

Class 3

Class 4

Atrioventricular valve regurgitation: Present or absent

Ventricular function: Good

Mild LV dysfunction

Moderate LV dysfunction

Severe LV dysfunction

Mean oxygen saturation (%)

Mean haematocrit (%)

Mean PA pressure (mmHg)

Perioperative and Postoperative Variables

Parameter

Age at the time of surgery (years)

CPB (minutes)

Aortic cross clamp (minutes)

Inotropic support - Duration(hours)

Type of operation

Lateral tunnel Fontan (LTF) alone

Fenestrated LTF

Extracardiac Fontan (ECF) alone

Fenestrated ECF

Off pump TCPC

Time to extubation (hours)

Reintubation: Present / Absent

Reintervention

ICU stay (hours)

Hospital stay (days)

Duration of post op pleural drainage (days)

Mortality(in hospital)

Outcome parameters assessed at follow up

(Follow up visit that will assessed for all patients is – follow up visit that occurred 1 year after the Fontan surgery)

Age at follow up visit

NYHA Class: Class 1

Class 2

Class 3

Class 4

Ventricular function (based on ejection fraction (EF) expressed in percentage, assessed by echocardiogram) – Good LV function (EF = >55%)

Mild LV dysfunction (EF= 45-54%)

Moderate LV dysfunction (EF= 30-44%)

Severe LV dysfunction (EF = < 30%)

Atrioventricular valve regurgitation – Present or Absent

Adverse events

Arrhythmia: Yes/ No

Need for PPI: Yes/ No

MASTER CHART

Sl No	Name	Hospital No	SEX	AGE AT DIAGNOSIS(MON	AGE AT SURGERY(YEARS)	AGE AT FOLLOW UP(YEARS)	PRIMARY MORPHOLOGY	ANY PRIOR PALLIATION	PRIOR BTS		NUMBER OF PRIOR PALLIATION	PRIOR AORTIC ARCH INTERVENTION	PRIOR BDG	AGE AT FIRST PROCERDURE(M
1		411061	F	6	11	12	DORV VSD PS AVCD	YES	NO		1	NO	2011	48
2		303394	F	1	8	9	DORV VSD PS	YES		2009	2	PDA STENT 2009	2011	19
3		321919	M	1	22	23	DORV	YES	NO		1	NO	1999	60
4		223853	F	1	13	14	TA	YES	NO		1	NO	2004	5
5		323357	F	1	6	7	DILV	YES	NO	(PA BAND2011+BDG2015)2		NO	2015	6
6		399403	M	1	12	13	DORV VSD PS	YES	NO		1	NO	2008	36
7		232923	F	1	15	16	DILV	YES		2005	2	NO	2008	24
8		275789	M	1	10	11	TA NRGA	YES	NO		1	NO	2009	78
9		291298	M	1	9	10	DORV SV(RV)	YES	NO		1	NO	2009	12
10		9802985	F	4	20	21	DORV VSD PS	YES	NO		1	NO	2012	408
11		343601	F	18	17	18	DORV VSD PS	NO	NO		0	NO	NO	17
13		9908419	M	3	17	18	DTGA VSD PS	YES	NO		1	NO	2004	98
14		214490	F	1	13	14	DTGA VSD PS	YES	NO(BAS AT 2 MONTHS)		2	NO	2004	18
15		237544	M	1	13	14	DTGA VSD PS	YES	NO		1	NO	2005	9
16		306104	M	1	8	9	TA NRGA	YES		2010	2	NO	2011	2
17		289301	M	1	15	16	DORV VSD PS	YES	NO	PA BAND+AS2001+BDG+MPA IN		NO	2011	3
18		268898	M	1	9	10	DTGA VSD PS	YES	NO		1	NO	2008	18
19		270686	M	1	11	12	TA	YES	NO		1	NO	2009	24
20		285442	M	1	8	9	TA	YES	NO		1	NO	2009	12
21		285521	M	2	8	9	DORV VSD PS	YES	NO		1	NO	2009	12
22		202559	F	2	16	17	SV(LV) PS	YES	NO		1	NO	2002	36
23		9809371	M	3	18	20	PA VSD	YES		2001	2	NO	2003	3
24		374560	F	2	19	20	TA	NO	NO		0	NO	NO	18
25		210291	M	3	13	14	PA SV(RV) VSD	YES	NO		1	NO	2003	7
26		260544	F	1	10	11	TA PS	YES	NO		1	NO	2007	18
27		200641	M	1	14	15	DILV PA	YES	NO		1	NO	2002	12
28		259910	F	1	9	10	TA VSD PS NRGA	YES	NO		1	NO	2006	9
29		196297	M	3	14	15	LTGA VSD PS	YES	NO		1	NO	2004	24
30		375137	F	1	17	18	LTGA VSD PS DC	NO	NO		0	NO	NO	17
31		255303	F	168	23	24	DORV VSD PS	YES	NO		1	NO	2006	180
32		260169	M	1	8	9	TA IVS	YES	NO	BDG+AS+PA BAND 1		NO	2007	8
33		9702778	F	1	18	19	LTGA UAVCD	YES	NO		1	NO	2002	60
34		228111	F	1	10	11	SV(LV) VSD SI DC LSVC	YES	NO		1	NO	2004	8
35		239666	M	1	8	9	TA VSD PS	YES	NO		1	NO	2005	6
36		237310	M	5	10	11	PA VSD SI MC	YES	NO		1	NO	2006	21
37		246208	M	2	9	10	SV(LV) PS SS DC MAPCA	YES	NO		1	NO	2006	10
38		224754	M	3	10	11	PA SV(RV)	YES	NO		1	NO	2005	12
39		343601	F	18	18	19	DORV VSD PS	NO	NO		0	NO	NO	216
40		9801987	M	12	18	19	TA NRGA VSD PS	YES	NO		1	NO	2005	96
41		9905307	F	1	18	15	DORV VSD PA	YES	NO		1	NO	2006	60
42		215936	M	1	11	12	SV(LV) PS	YES	NO		1	NO	2006	30
43		188617	F	1	15	16	CCTGA VSD PS DEXTROPO	YES		2001	2	NO	2002	24
44		9700766	M	12	17	18	UAVCD SI SV(RV)	YES		1997	2	NO	1998	20
45		9604829	F	2	17	18	TA NRGA VSD	YES	NO		1	NO	2010	180
46		9800481	F	1	15	16	TA VSD PS	YES	NO		1	NO	2007	96
47		276356	M	1	9	10	TA VSD PS	YES	NO		1	NO	2008	48
48		223885	F	1	9	10	DORV VSD PS	YES	NO		2	NO	2004	14
49		244366	M	1	8	9	DTGA VSD PS	YES	NO		1	NO	2006	24
50		200697	M	1	166	17	DTGA VSD PS MC SI	YES	NO		1	NO	2003	72
51		272410	F	5	7	8	TA VSD PS	YES	NO		1	NO	2007	12
52		224807	F	1	9	10	DILV SV(LV) PS	YES	NO		1	NO	2006	24
53		9903584	M	1	14	15	TA SV(LV) SS DC	YES	NO		1	NO	2002	36
54		279033	M	1	8	9	DILV SV(LV) DTGA	YES		2005	2	NO	2008	3

O2 SAT	NYHA CLASS	AV REGURGITATION	VENTRICULAR FUNCTION	MEAN PA PRESSURE	TYPE OF FONTAN	AGE AT FONTAN	FENESTRATION	CPB(MIN)	AOCXCL(MIN)	INOTROPE SUPPORT DURATION(HOURS)	TIME TO EXTUBATION(HOURS)	REINTUBATION	REINTERVENTION	ICU STAY(HOURS)
79	3	MILD	GOOD	12	ECF	10	NO	289	79	48	9	No	No	96
78	1	MILD	GOOD	12	ECF	8	NO	246	65	72	22	NO	YES-BLEEDING	96
81	2	MOD	MILD	10	ECF	22	NO	152	43	60	12	NO	NO	96
52	3	MILD	SEVERE	15	ECF	13	NO	233	63	44	20	NO	MILRINONE FOR 1 WEEK	576
82	3	NIL	GOOD	14	ECF	6	NO	107	NOT CLAMPED	50	6	NO	NO	144
77	3	MILD	MILD	13	ECF	12	NO	180	76	40	17	NO	NO	132
69	2	MILD	GOOD	10	ECF	15	NO	404	NOT CLAMPED	50	20	NO	NO	96
85	3	MILD	GOOD	14	ECF	11	NO	96	23	40	6	NO	NO	48
78	2	NIL	GOOD	14	ECF	9	NO	275	147	56	4	NO	NO	108
81	3	MILD	GOOD	12	ECF	20	NO	148	12	60	8	NA	No	98
80	2	NIL	GOOD	11	ECF	17	YES 4MM	178	35	35	8	NO	NO	84
73	3	MOD	GOOD	11	ECF	17	NO	265	116	51	5	NO	NO	84
75	2	MOD	GOOD	15	ECF	13	YES 4MM	330	40	48	11	NO	YES	22
82	2	MILD	GOOD	10	ECF	13	NO	175	NOT CLAMPED	64	7	No	NO	90
74	2	NIL	GOOD	15	ECF	8	YES 4MM	148	12	60	5	NO	NO	108
78	2	NIL	GOOD	MPA INTERRUPTED AT	ECF	16	YES 5MM	188	40	22	5	NO	NO	96
82	2	MOD	GOOD	12	ECF	10	YES 4MM	161	41	36	6	NO	NO	84
80	2	MILD	GOOD	12	ECF	11	NO	123	27	41	8	NO	NO	84
82	2	NIL	GOOD	15	ECF	8	YES 4MM	177	44	60	3	NO	NO	72
81	2	NIL	GOOD	12	ECF	8	YES 4MM	123	23	36	4	NO	NO	86
81	3	NIL	GOOD	12	ECF	16	YES 5MM	330	143	96	5	NO	NO	96
84	2	MILD	GOOD	12	ECF	18	YES 6MM	309	NOT CLAMPED	204	192	NO	YES-BLEEDING	288
90	2	NIL	GOOD	16	ECF	20	YES 4.5MM	246	151	50	18	NO	NO	102
72	2	NIL	GOOD	13	ECF	15	YES 4.5MM	164	NOT CLAMPED	20	7	NO	NO	70
82	2	NIL	GOOD	12	ECF	9	YES 4MM	162	59	72	6	NO	NO	110
78	2	MILD	GOOD	9	ECF	14	YES 5MM	125	NOT CLAMPED	36	6	NO	NO	72
76	3	MILD	GOOD	12	ECF	9	YES 5MM	145	30	42	15	NO	NO	96
75	2	NIL	GOOD	15	ECF	17	YES 5MM	240	13	72	8	NO	NO	80
75	2	MILD	GOOD	13	ECF	17	YES 5MM	155	39	52	7	NO	NO	108
78	3	MILD	GOOD	12	ECF	23	YES 5MM	206	30	66	7	NO	NO	96
78	2	NIL	GOOD	12	ECF	8	YES 4MM	147	14	90	8	NO	NO	90
76	2	MOD	GOOD	9	ECF	20	YES 4MM	155	20	46	5	NO	NO	50
72	2	MILD	GOOD	8	ECF	13	YES 4.5MM	190	NOT CLAMPED	67	14	NO	YES-LEFT LUNG DECORTI	168
80	2	NIL	GOOD	9	ECF	13	YES 4MM	254	33	84	10	NO	NO	83
80	2	NIL	GOOD	9	ECF	9	YES 4MM	202	96	99	13	NO	NO	113
84	2	NIL	GOOD	9	ECF	9	YES 4.5MM	198	94	68	6	NO	NO	112
84	1	MILD	GOOD	8	ECF	10	YES 4MM	300	NOT CLAMPED	60	16	NO	YES-BLEEDING	138
77	2	NIL	GOOD	10	ECF	18	YES 4MM	178	35	28	7	NO	NO	84
82	2	NIL	GOOD	12	ECF	18	YES 3.5MM	215	52	36	10	NO	NO	80
82	2	NIL	GOOD	12	ECF	18	YES 4MM	220	143	40	8	NO	NO	120
85	2	NIL	GOOD	14	ECF	11	YES 4MM	205	31	66	4	NO	NO	110
84	2	NIL	GOOD	10	ECF	15	YES 6MM	313	105	46	11	NO	NO	96
84	2	MILD	GOOD	12	ECF	17	YES 4MM	309	3	62	19	NO	NO	82
86	1	MILD	GOOD	9	ECF	17	YES 4.5MM	222	39	43	11	NO	NO	96
82	2	MOD	GOOD	11	ECF	15	YES 4MM	417	145	112	14	NO	NO	120
86	2	NIL	GOOD	12	ECF	9	YES 4MM	162	24	52	6	NO	NO	74
80	2	MILD	GOOD	9	ECF	9	YES 5MM	342	55	68	9	NO	NO	188
86	2	MILD	GOOD	11	ECF	8	YES 4MM	137	26	60	6	NO	NO	100
88	2	NIL	GOOD	12	ECF	16	YES 6MM	177	NOT CLAMPED	110	12	NO	YES-HIGH FONTAN PRES	144
70	2	NIL	MILD	13	ECF	7	YES 4MM	178	35	30	7	NO	NO	80
75	2	MILD	GOOD	13	ECF	9	YES 4MM	112	48	58	6	NO	NO	98
78	2	MILD	MILD	14	ECF	14	YES 4.5MM	253	129	96	9	NO	NO	112
92	2	MILD	GOOD	8	ECF	8	YES 4.5MM	144	62	60	7	NO	NO	168

PLEURAL DRAINAGE(DAYS)	HOSPITAL STAY(DAYS)	IN HOSPITAL MORTALITY	FOLLOW UP 02 SAT	NYHA CLASS	VENTRICULAR FUNCTION	AVVR	FOLLOW UP IN YEARS	AGE AT LAST FOLLOW UP	TELEPHONIC INTERVIEW - MORTALITY	PPI	ARRHYTHMIAS
8	12	No	95	2	Good	Mild	1	11			
5	16	NO	92	2	Good	Mild	1	9			
6	10	NO	92	2	MILD	MILD	1	23			
29	27	NO	NA	NA	NA	NA	NA	NA			
4	15	NO	96	1	GOOD	MILD	1	7			
19	21	NO	94	1	GOOD	MILD	1	13			
4	9	NO	93	2	GOOD	NIL	1	16			
4	8	NO	98	1	GOOD	MILD	1	11			
22	24	NO	97	1	GOOD	NIL	1	10			
8	NA	NO	93	2	MILD	NIL	1	21			
4	10	NO	94	1	GOOD	NIL	3	21			
12	18	NO	98	1	GOOD	NIL	1	18			
47	52	NO	94	3	GOOD	MILD	4	16			
9	NA	NO	93	2	MILD	MILD	2	15			
4	40	NO	95	1	GOOD	NIL	2	10			
10	10	NO	98	1	GOOD	MILD	2	17			
19	28	NO	95	2	GOOD	MILD	2	11			
3	6	NO	91	1	GOOD	NIL	2	13			
4	7	NO	93	1	GOOD	MILD	2	10			
10	13	NO	98	1	GOOD	MILD	2	10			
5	8	NO	96	1	MILD	MOD	2	18		CHB-2012	CHB WITH NARROW
8	19	NO	94	2	GOOD	MILD	2	20			
13	15	NO	90	2	GOOD	NIL	3	22			
20	33	NO	93	1	GOOD	NIL	2	15			
15	16	NO	94	1	GOOD	MILD	3	13			
7	9	NO	NO REC	NO REC	NO REC	NO REC	NO F/U	15			
4	21	NO	96	2	GOOD	NIL	3	12			APRIL 2015 UNDERV
6	13	NO	94	2	GOOD	MILD	3	17			
14		NO	98	1	GOOD	MILD	3	20			
11	16	NO	98	2	GOOD	NIL	3	26			
7	14	NO	94	1	GOOD	NIL	2	11			
19	22	NO	94	2	GOOD	NIL	3	23			
22	50	NO	95	1	GOOD	MILD	4	17			ATRIAL TACHYCARDI
10	13	NO	96	1	GOOD	MILD	4	17			
13	18	NO	98	1	GOOD	NIL	4	13			
5	12	NO	87	1	GOOD	MILD	4	13			
19	26	NO	89	2	GOOD	MOD	4	14			
5	10	NO	93	1	GOOD	NIL	3	22			
4	7	NO	94	1	GOOD	NIL	4	22			
18	22	NO	94	2	GOOD	NIL	3	21			
9	16	NO	90	1	GOOD	MILD	4	15			
11	19	NO	90	1	GOOD	NIL	4	19			
15	19	NO	92	2	GOOD	NIL	4	21			
8	15	NO	98	1	GOOD	MILD	5	22			
21	27	NO	95	1	GOOD	MILD	4	19			
6	10	NO	98	1	GOOD	NIL	5	14			
8	24	NO	95	1	GOOD	NIL	5	14			SR WITH INTERMITT
10	14	NO	94	1	GOOD	MILD	5	14			
8	31	NO	90	2	GOOD	NIL	4	20			
11	22	NO	91	1	GOOD	MILD	5	12			
8	11	NO	95	1	GOOD	MILD	4	13			
10	14	NO	92	2	MILD	MILD	4	18			
8	14	NO	90	1	GOOD	MILD	3	12			

Additional cardiac procedures	REMARKS	YEAR OF SURGERY	LAST FOLLOW UP DATE	POST PROCEDURE FONTAN PRESSURE
MPA INTERRUPTION		2017		
CS CUTBACK+MPA INTERRUPTION		2017		
NIL		2017		
AVR FOR IE+ CONFLUENCE PLASTY	PRE OP AKI AND H/O IE,NO FOLLOW UP RECORDS	2017		
MAPCA COILING PRIOR TO SURGERY		2017		
TV REPAIR		2017		
BDG+ATRIAL SEPTOSTOMY+LPA PLASTY, NOW ECF+ LPA PLASTY	PULMONARY AVM COILING DONE 1 MONTH AFTER SX	2017		
		2017		
BDG+ATRIAL SEPTOSTOMY		2017		
ECF+TV REPAIR		2017		
		2014		
POSTERIOR MITRAL ANNULOPLASTY + P2 PLICATION		2017		
LPA PLASTY+TV REPAIR, RIGHT THORACOTOMY FOR DIAPHRAGM INJUR\	MAPCA COILING DONE IN 2014 PRE FONTAN,POST OP	2015		
BDG+LPA PLASTY		2016		
	POST OP PEUDOANEURSYM AT FEMORAL CANNULATIC	2016		
		2016		
RIGHT AV VALVE ANNULOPLASTY,BDG+AS 2008		2016		
BDG+AS		2016		
BDG+AS+PA BANDING,ECF+ LPA PLASTY		2016		
BDG+AS		2016		
ADDITIONAL LEAD FOR LV PLACED DURING ECF		2016		
	POST OP RIGHT MCA,MCA-PCA INFARCT,POWER IMPR(	2016		
POSTERIOR MITRAL ANNULOPLASTY DONE		2015		
		2015		
		2015		
	HAS NOT COME POST SURGERY, TELEPHONE PENDING	2015		
1MONTH POST SURGERY ADMITTED WITH SEV LVD, TREATED WITH MILI	MAPCA COILING 2011, JULY 2015 DEVELOPED SEVERE I	2015		
1MONTH POST BDG PDA WAS COILED IN 2004, AS ECHO SHOWED LPA SI	CEREBRAL ABSCESS IN 2009 BURR HOLE DRAINAGE DO	2015		
SINGLE STAGE DONE AS PA WAS OF GOOD SIZE.	CRANIOTOMY FOR BRAIN ABSCESS 2014	2015		
		2015		
		2015		
		2015		
LEFT LUNG DECORTICATION FOR LEFT LUNG EMPYEMA ON 1/2/14.	MAPCA AND LIMA COILING 2011 BEFORE SURGERY	2014		
		2014		
		2014		
HAEMOPTYSIS-2015,2MAPCA COILED,HAEMOPTYSIS OCT 2017-5MAPCA	READMITTED 1 MONTH POST FONTAN FOR RIGHT PLEU	2014		
BDG+B/L PLASTY,DURING ECF LAPAROTOMY TO REPAIR IVC TEAR THAT (	IMMEDIATE POST OP MOD AVVR,FENESTRATION GRAI	2014	4/18/2018	
POST GLEN PA WAS 11,HENCE WENT AHEAD WITH S.STAGE,BIVENTRICU .		2014	9/21/2017	
		2014	5/15/2018	
		2014	9/27/2017	22
		2014	5/10/2018	11
		2014	6/4/2018	18
PA PLASTY DONE WITH BDG, RPA PLASTY DONE WITH ECF		2013		
		2013	3/13/2018	
MITRAL ANNULOPLASTY+BPP AUGMENTATION OF PA CONFLUENCE AND LPA ORIGIN		2013	12/13/2017	
		2013	5/25/2018	
BDG WITH MPA INTERRUPTION, MAPCA COILED IN 2010,ECF WITH CONFLUENCE PLASTY DONE,BAS IN 03		2013	2/27/2018	
		2013	1/23/2018	
		2013	2/2/2017	
	UTI POST OP	2013	1/1/2018	
	LEFT DIAPHRAGMATIC PLICATION IN 2013 POST BDG H	2013	8/11/2017	
		2013	12/29/2017	
		2013	4/11/2017	