UNIVERSITÀ DEGLI STUDI DI PADOVA

UNIVERSITA DEGLI STUDI DI PADOVA

Dipartimento di Scienze Cardiologiche, Toraciche e Vascolari

SCUOLA DI DOTTORATO DI RICERCA IN SCIENZE MEDICHE, CLINICHE E SPERIMENTALI
DOTTORATO DI RICERCA IN SCIENZE CARDIOVASCOLARI

CICLO XXV

TESI DI DOTTORATO

ONE AND A HALF VENTRICLE REPAIR: CLINICAL AND ANIMAL STUDY

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RIASSUNTO

La prima descrizione della cosiddetta "correzione a un ventricolo e mezzo", fu pubblicato da Billingsly et al. nel 1989 [1], Questo tipo di correzione è stato poi applicato in altri centri per correggere diverse anomalie congenite cardiache complesse caratterizzati da un ventricolo destro ipoplasico. La logica alla base di questa riparazione è duplice. Da un lato l'obiettivo è di ridurre il flusso di sangue, e quindi il pre-carico in, un ventricolo disfunzionale o ipoplasico destro (RV). D'altra parte la riparazione a un ventricolo e mezzo (1,5 VR) permette di mantenere una separazione completa e fisiologica della circolazione polmonare e sistemica, evitando mixing di sangue e desaturazione. Inoltre rappresenta una valida alternativa alla circolazione secondo Fontan nel contesto di una ipoplasia del ventricolo destro, fattore funzionale, con il vantaggio di fornire un flusso polmonare pulsatile e prevenire l'ipertensione venosa nel distretto della vena cava inferiore [2, 3]. La correzione a un ventricolo e mezzo comprende la connessione della vena cava superiore all'arteria polmonare (shunt cavopolmonare bidirezionale), oltre a completare la chiusura di eventuali comunicazioni intracardiache, o meno alla riparazione di ulteriori difetti cardiaci congeniti associati. Questa metodica può essere applicata ad un ampio spettro di anomalie congenite, purché il ventricolo destro ipoplasico sia funzionalmente valido e pertanto in grado di gestire la portata di sangue della sola cava inferiore [1, 4].

Questo studio è diviso in due parti:

1. Studio clinico 2. Studio sperimentale negli animali.

In questo studio abbiamo valutato i risultati del nostro Istituto a lungo termine e non solo della circolazione polmonare bifocale ma anche l'impatto delle malformazioni cardiache associate. Lo studio sugli animali e stato condotto nei conigli per valutare l'effetto del "pattern" pulsatile del flusso venoso in vena cava superiore e di capire i cambiamenti emodinamici in vena cava superiore, atrio destro, ventricolo destro, arteria polmonare e vena cava inferiore, nell'immediato e dopo stress test formacologico.

A. Studio clinico

Metodi

Tutti i pazienti con correzione in uno ventricolo e mezzo tra il Marzo 1994 e Gennaio 2012 sono stati inclusi. Tutti i dati clinici disponibili e operativi sono stati valutati. I pazienti sono stati suddivisi in: Gruppo A - pazienti con ventricolo destro ipoplasico / disfunzioni e malformazioni delle valvole tricuspide e polmonare, Gruppo B - pazienti con associate malformazioni complesse che coinvolgono altre strutture cardiache.

Risultati

Si tratta di un esame clinico retrospettivo di 18 anni in 24 pazienti consecutivi sottoposti a correzione un ventricolo e mezzo presso il nostro Istituto. L'età media di correzione è stata di 11,9 anni (range 4 mesi - 66,8 anni). Il Z-score medio dell' anello TV era -3,2 (range -6,2 a 3,6). Le resistenze vascolari polmonari medie erano 1,75 U/m2 (range 1,0 - 3,0). Tre pazienti erano affetti da sindrome eterotassica.

Sedici pazienti erano stati sottoposti a precedenti interventi cardiaci, uno di questi aveva una deviazione atriopolmonare totale secondo Fontan.

Le complicanze postoperatorie si sono verificate in 19 pazienti (79%): chilotorace/chylopericardium (n = 7), BAV III (n = 2), aritmie (n = 7), insufficienza renale (n = 4), insufficienza cardiaca (n = 4), cardiaca / versamento pleurico (n = 11), la sindrome di SVC (n = 2), infezione polmonare (n = 3), paralisi emi-diaframma che ha richiesto plastica diaframmatica (n = 1), trombosi SVC con PE (n = 1). Cinque pazienti hanno richiesto l'impianto definitivo di PM. Tutti i pazienti sono sopravissuti e dopo una degenza media di 32 ± 29 giorni. Ad un follow up medio di 8,3 anni (range 1 mese - 17,9 anni, la completezza FU: 96%), ci sono stati 2 morti tardive (1 non cardiaca). Tra i sopravvissuti, lo stato funzionale prevalente era NYHA classe I in 19 pazienti (90,4%). Eventi avversi tardivi si sono verificati in 10 pazienti (43%) tra cui: un nuovo intervento(n = 3), uno di questi era una conversione biventricolare; procedure emodinamiche (n = 6), aritmie (n = 2), eventi neurologici
(n = 1); altre complicazioni (n = 5).

La libertà totale da eventi avversi, la chirurgia e le procedure interventistiche sono state 56,5%, 87% e 73,9% rispettivamente. La libertà da eventi avversi nel gruppo A (n = 12) è stata 83,3%, mentre nel gruppo B (n = 11) è stata pari al 27,3%. L'analisi statistica ha dimostrato che il Gruppo B ha avuto una libertà significativamente più basse di eventi avversi rispetto al gruppo A (p = 0,015).

B. Studio degli animali

Metodi

Esperimento è stato eseguito in conigli. La cura degli animali e stata svolta secondo lo standard stabilito per gli studi sperimentali.

Modello sperimentale: CONIGLIO

Strain: NUOVA ZELANDA

Numero totale Animali: 30

Peso di animali: 4. 5-5,5 kg

Risultati

1. Dopo anastomosi cavopolmonare- la pressione in SVC risultava aumentata ma la pressione in RA invece diminuiva, il che dimostra la diminuzione della pressione nella vena cava inferiore.

2. Dopo lo stress indotto farmacologicamente: la pressione nella vena cava superiore e la frequenza cardiaca aumentavano, mentre la pressione del ventricolo destro, atrio destro e la pressione in arteria polmonare rimanevano simili come in condizione di riposo dopo un'anastomosi cavo polmonare.

3. Ciò significa che la pressione della vena cava superiore è sempre più elevata dopo anastomosi cavapulmonare ma la pressione venosa nella vena cava inferiore è costante e ancora più bassa se confrontato alla pressione preoperatoria.

4. Non abbiamo trovato alcuna modifica della pressione atriale destra, dopo correzione ad un ventricolo e mezzo; anche dopo stress test farmacologico viene dimostrato che il ritorno dalla vena cava inferiore è adeguato e non esiste ipertensione

in vena cava inferiore, (buona tolleranza fisica). Questo è un risultato molto positivo a favore della correzione a un ventricolo e mezzo ventricolo rispetto all' intervento di Fontan.

Conclusione

Si conclude che la correzione 1,5 VR è una scelta sicura e valida per la riparazione chirurgica dell'ipoplasia del ventricolo destro associato o meno ad altre malformazioni cardiache come alternativa alla procedura di Fontan. La maggior parte dei pazienti mostrano un buono stato funzionale al follow-up. Questa riparazione mantiene una bassa pressione nella vena cava inferiore e permette buoni risultati precoci e a medio termine.

SUMMARY

Since the first description of the so called “one and a half ventricle repair”, published by Billingsly et al. in 1989 [1], this type of correction has been applied to several complex congenital cardiac anomalies. The rationale underlying this repair is twofold. On one hand the aim is to reduce the blood flow to, and therefore to downsize the pre-load of, a dysfunctional or hypoplastic right ventricle (RV), considered to be unable to sustain a biventricular correction. On the other hand the one and a half ventricle repair (1.5 VR) permits to maintain a complete and physiological separation of pulmonary and systemic circulations, avoiding blood mixing and desaturation. Furthermore it represents a valid alternative to the Fontan circulation in the setting of a less but still functioning RV, with the advantage to provide a pulsatile pulmonary blood flow and to prevent systemic venous hypertension [2,3]. One and a half ventricle repair consists of bidirectional cavo-pulmonary shunt (BCPS) in addition to complete closure of intracardiac communications, associated or not with repair of other congenital heart defects. It can be appropriate for a wide spectrum of congenital anomalies, provided that the RV is large enough to manage the blood supply from the inferior vena cava. However, feasibility in adult population is being explored [1,4].

This study has divided in two parts:

1. Clinical study 2. Experimental study in animals.

In clinical study we have reviewed our institutional experience with 1.5 VR to evaluate early and long term results and explore the impact of associated cardiac malformations on outcomes of the one and a half ventricle repair. Animal study in rabbits was conducted to evaluate the effect of a pulsatile venous flow pattern in superior vena cava and to understand an immediate & after pharmacologically induced stress test the hemodynamic changes in superior vena cava, right atrium, right ventricle, pulmonary artery and inferior vena cava district.

A. Clinical study

Methods

All patients who underwent one and a half ventricle repair between March 1994 and January 2012 were included. All available clinical and operative data were reviewed. Patients were divided in: Group A - patients with right ventricle hypoplasia/dysfunction and malformations of tricuspid or pulmonary valves; Group B - patients with associated complex malformations involving other cardiac structures.

Results

This is an 18 years retrospective clinical review of 24 consecutive patients who underwent one and a half ventricle repair at our Institution. Mean age at repair was 11.9 years (range 4 months – 66.8 years). Mean TV annulus Z-score was -3.2 (range -6.2 to 3.6). Mean pulmonary vascular resistance was 1.75 U/m2 (range 1.0 to 3.0).

Three patients had heterotaxy syndrome. Sixteen patients underwent previous cardiac operation, one of these had a Fontan operation.

There was no death at operation. Postoperative complications occurred in 19 patients (79 %), and were: chylothorax/chylopericardium (n=7), BAV III (n=2), arrhythmias (n=7), renal failure (n=4), heart failure (n=4), cardiac/pleural effusion (n=11), SVC syndrome (n=2), pulmonary infection (n=3), hemi diaphragm paralysis that required diaphragm plasty (n=1), SVC thrombosis with PE (n=1). Five patients required definitive PM implantation. All patients were discharged home alive and well, after a mean hospital stay of 32 ± 29 days. At a mean follow up of 8.3 years (range 1 month – 17.9 years, FU completeness: 96 %), there were 2 late deaths (1 non cardiac related). Among survivors, functional status was NYHA class I in 19 patients (90.4%). Late adverse events occurred in 10 patients (43 %) including: late reoperation (n=3), one of these was a biventricular conversion; haemodynamic procedures (n=6); arrhythmias (n=2); neurological event (n=1); other complications (n=5).

Overall freedom from adverse events, surgery and interventional procedures was 56.5 %, 87 % and 73,9 % respectively. Freedom from adverse event in Group A (n=12) was 83.3 % while in Group B (n=11) was 27.3 %. Statistical analysis demonstrated that Group B had a significantly lower freedom from adverse events than Group A ($p = 0.015$).

B. Animal Study

Methods

Experiment was performed in experimental Rabbits. Animal care has taken according to established standard for experiment in animals.

Experimental model: RABBIT

Strain: NEW ZEALAND

Total animal number: 30

Weigh of animals:4. 5-5.5 kg

Results

1. After cavapulmonary anastomosis- SVC and PA pressure has increased but RA pressure has decreased, which proves the decrease of the pressure in inferior vena cava district.

2. After pharmacologically induced stress test: increased the pressure in superior vena cava and also increased heart rate but right ventricular, right atrial & pulmonary artery pressures were remained as same as after cavopolmonary anastomosis, as it is in resting condition.

3. That means superior vena cava pressure is always higher after cavapulmonary anastomosis but venous pressure in inferior vena cava district is constant and even lower in comparison with preoperative pressure.

4. We found no any changes of right atrial pressure, after 1.5 ventricular repair and even after pharmacologically induced stress test which proves that the inferior vena cava return is ok and no hypertension in inferior vena cava district even after stress

test(good exercise tolerance), which is a very positive finding in favour of one and a half ventricle repair in comparison to Fontan type procedure.

Conclusion

After having the result of both clinical study and experiment in animals we have concluded that the 1.5 VR is a safe and valid option for surgical repair of hypoplasic, borderline or failing right ventricles as an alternative to Fontan procedure. Most patients show good functional status at follow up. This repair provides a low pressure in the inferior vena cava district and allows good early and mid-term outcomes.

ABBREVIATION

1.5VR = One and a half ventricle repair

ASD = Atrial septal defect

A-V = Atrio- ventricular

AV canal = Atrio ventricular canal

BCPS = Bidirectional cavopulmonary shunt

CHD = Congenital heart disease

CPA = Cavopulmonary anastomosis

CVPA = Cavopulmonary anastomosis

DORV = Double outlet right ventricle

IVC = Inferior vena cava

LPA = Left pulmonary artery

LSVC = Left superior vena cava

MPA = Main pulmonary artery

MRI = Magnetic Resonance Imaging

MUF = Modified ultrafiltration

PA = Pulmonary artery

PAPVR = Partial anomalous pulmonary venous return

PDA = Patent ductus arteriosus

PFO = Patent fossa ovalis

PVR = Pulmonary vascular resistance

RPA = right pulmonary artery

RSVC = Right superior vena cava

RV = Right ventricle

RVOT = Right ventricle outflow tract

RVOTO = Right ventricle outflow tract obstruction

SVC = Superior vena cava

TCPC = Total cavopulmonary anastomosis

TGA = Transposition of the great arteries

TV = Tricuspid valve

CONTENTS

SECTION I

GENERAL OVERVIEW

A. HISTORICAL NOTES

Fig.1 Dr Carlon

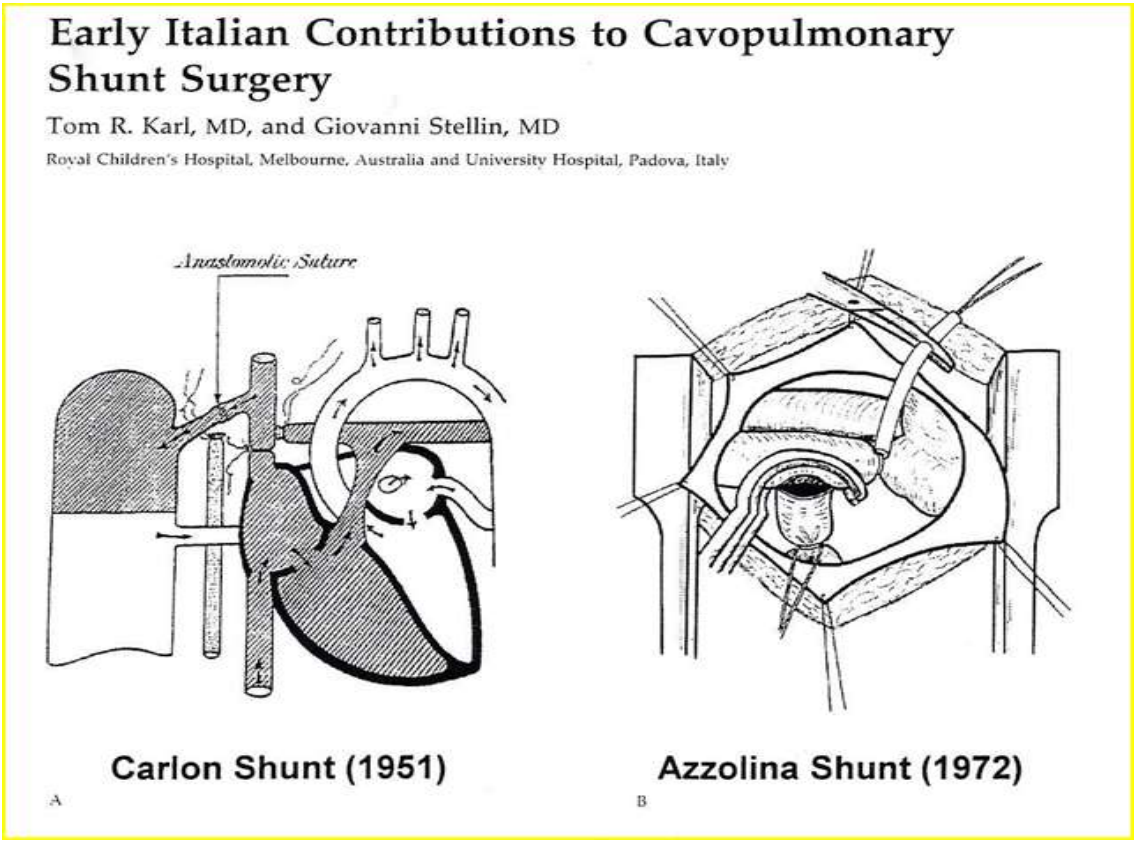


Fig.2: Ann Thorac Surg 1999; 67:1175

Carlon (of Padova) employed his shunt (Fig 2A) in a canine model [5]. His 1951 description noted some of the technical and physiologic advantages of this alternative to the Blalock shunt. The azygos vein was used to connect the right pulmonary artery to the superior vena cava, and the proximal right pulmonary artery and superior vena cava were ligated. Azzolina (of Massa) extended this concept to a bidirectional shunt (Fig 2B), which was employed in 9 patients with tricuspid atresia, the youngest just 5 months of age [6]. The clinical work was carried out between 1968 and 1970.

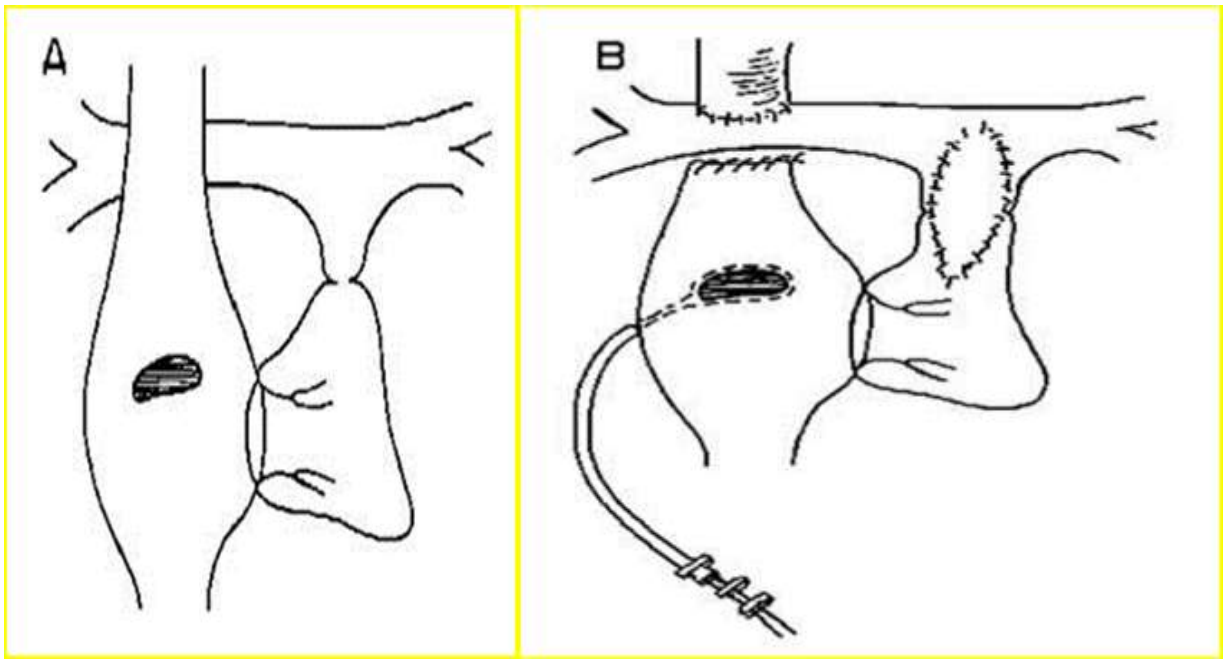


Fig.3: Billingsly AM, Laks H, Boyce SW. J Thorac Cardiovasc Surg 1989; 97:746-54

The Azzolina shunt was performed through a right thoracotomy, and without bypass, using a side-to-side anastomosis. An interesting feature was the use of an adjustable snare on the proximal superior vena cava.

B.INTRODUCTION

Congenital heart disease (CHD) is one of the most common inborn defects, with an extimated incidence of 8/1000 live newborns. Cardiac malformations are currently treated in the pediatric age, with an overall early surgical mortality inferior to 5%.[7] Prior to advent of surgery, less than 20% of children with congenital heart malformations survived to adult life.[8] Since the late '60s, the extraordinary advances in cardiac surgery, intensive care and non-invasive diagnosis have drastically modified the natural history of almost all CHD. Currently, most deaths for CHD occur in adults, who may have had or not repair or any kind of surgery. In fact, approximately 85% of babies born with cardiovascular anomalies can expect to reach adulthood, and with continued improvement in surgical technique, this could increase further in the next future.[9] This improvement has selected a growing number of newborns and infants who have survived through adolescence, until adulthood. [10]

In this contest patients with single ventricle physiology or with borderline right ventricle are still challenging cases for pediatric cardiac surgeons. One and a half ventricle repair, in which intra-cardiac repair is supplemented by bidirectional cavopulmonary anastomosis, may be a valuable concept in management of patients with a failing right ventricular circulation.[11] Patients with borderline right ventricle can be the candidate for one and a half ventricle repair like in case of right ventricular outflow obstruction with right ventricular hypoplasia, straddling tricuspid valve, pulmonary atresia with intact ventricular septum, rare variants of Tetralogy of Fallot, tricuspid valve stenosis with ventricular septal defect and Ebstein's anomaly. In all these above mentioned pathological conditions important is to understand pre operatively, whether the right ventricular function and volume is adequate for one and a half ventricle repair.

There are many different classifications of congenital heart diseases are available in literature. Most of the classifications are based on physiology and pathophysiology of the congenital heart diseases.

Among recent classifications, the most recent one is reported from the International Society for Nomenclature of Pediatric and Congenital Heart Disease.[12] which is a Physiological

classification of congenital heart disease:

1. '**Simple' left-to-right shunt lesions: these cause an increased pulmonary blood flow**

a. Atrial septal defect

b. Ventricular septal defect

c. Atrioventricular septal defect

d. Patent ductus arteriosus

e. Aortopulmonary window

NB The effect of the shunt on right ventricular and respiratory physiology differs depending on the level at which shunting occurs.

2. '**Simple' right-to-left shunt lesions: these cause a reduction in pulmonary blood flow with cyanosis**

a. Tetralogy of Fallot. Consists of right ventricular outflow tractobstruction, right ventricular hypertrophy, ventricular septal defect and an overriding of the aorta

b. Pulmonary atresia

c. Tricuspid atresia

d. Ebstein's anomaly. Consists of downward displacement of an abnormal tricuspid valve into the right ventricular cavity, part of the right ventricle is thus incorporated into the right atrium (atrialized right ventricle), and the remaining right ventricular cavity is malformed

3. Complex shunts: these cause mixing of pulmonary blood flow and systemic blood flow. Cyanosis occurs as a result of complex interactions between systemic vascular resistance and pulmonary vascular resistance

a. Transposition of the great arteries

b. Truncus arteriosus

c. Total anomalous pulmonary venous drainage

d. Double-outlet right ventricle

e. Hypoplastic left heart syndrome

Most of these lesions (except total anomalous pulmonary venous drainage) are examples of a parallel circulation.

4. Obstructive lesions

a. Coarctation of the aorta

b. Interrupted aortic arch

c. Aortic stenosis

d. Pulmonary stenosis

C. PATHOPHYSIOLOGICAL CLASSIFICATION OF CHD

(G. Thiene & C. Frescura –Padova)

Researchers from Padova, group of Professor Gaetano Thiene and Carla Frescura classified congenital heart diseases from pathophysiological point of view in their recent publication. [13]

Fig.4: Prof. Gaetano Thiene

1.CHD with increased pulmonary blood flow (septal defects without pulmonary obstruction and left-to right shunt): Partially anomalous pulmonary venous drainage, ASD, complete AV canal, VSD, Truncus arteriosus.

2. CHD with decreased pulmonary flow (septal defects with pulmonary obstruction and right-to-left shunt): Pulmonary stenosis with ASD, pulmonary stenosis with VSD(Tetralogy of Fallot), tricuspid valve atresia, Ebstein's anomaly, single or double inlet ventricle with pulmonary stenosis.

3. CHD with obstruction to blood progression and no septal defects (no shunt): Pulmonary valve stenosis, Aortic valve stenosis, aortic coarctation.

4. CHD so severe as to be incompatible with postnatal blood circulation:

Ductus dependent (pulmonary atresia, aortic and mitral atresia), interrupted aortic arch, atretic aortic arch, complete TGA, anomalous connection/ obstruction of the pulmonary veins.

5. CHD silent until adult age:

Bicuspid aortic valve, congenital anomalous coronary arteries, wolff- Prkinson – White syndrome, congenitally corrected TGA. According to both of the classifications patients with right ventricular hypoplasia belongs to the group of the patients with decreased pulmonary flow like; Ebstain's anomaly, tricuspid atresia, single or double inlet ventricle with pulmonary stenosis.

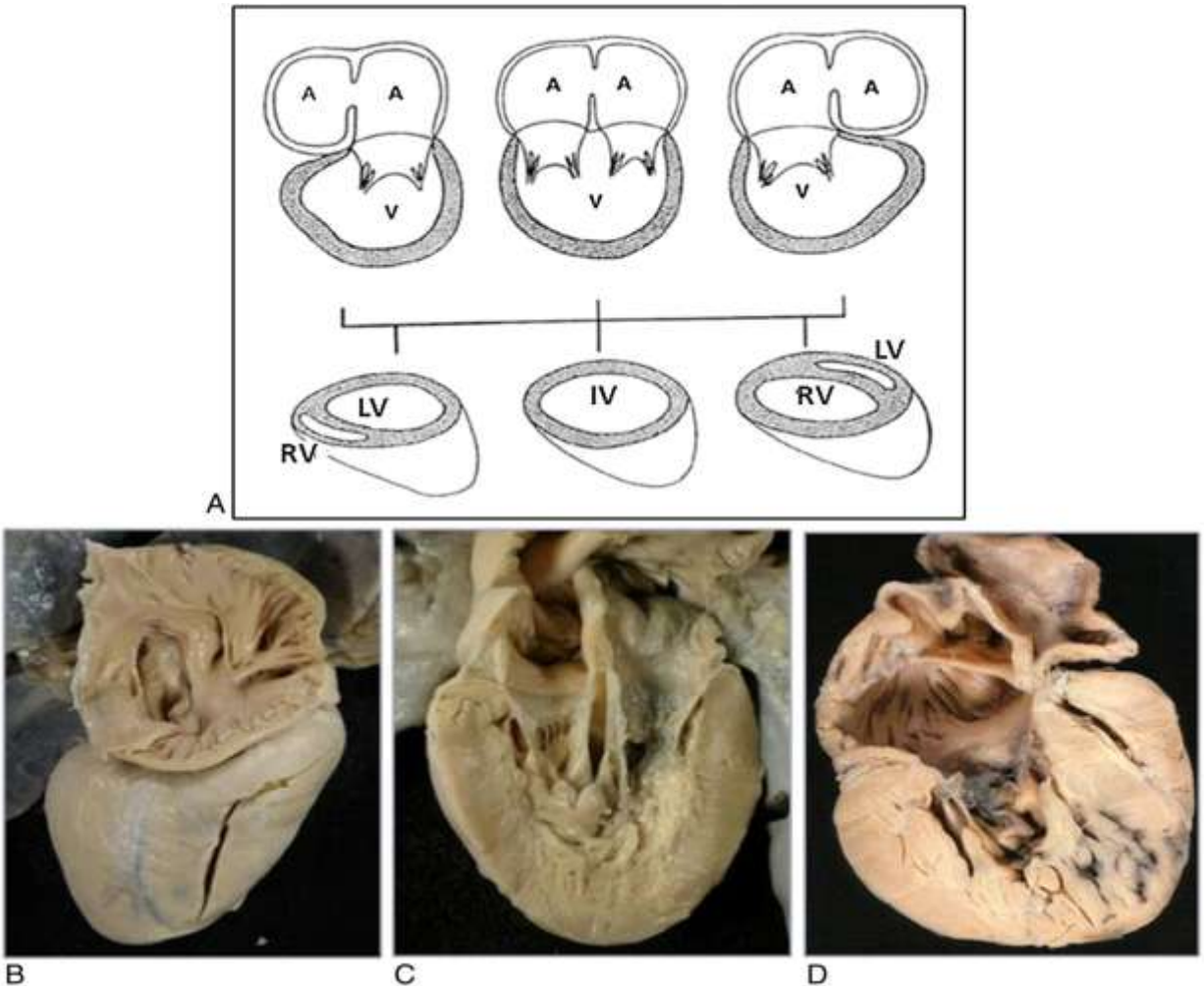


Fig.5: G.Thiene, C Frescura (cardiovascular pathology (2010)

Univentricular AV connection. (A) AV connection is univentricular when both atria drain mostly (N75%) into a single ventricular chamber through two patent valves or a common AV valve (double-inlet connection), or through a single patent valve in case of absent right or left connection. (B) Specimen with absent right AV connection (tricuspid atresia), viewed from the right atrium: no tricuspid valve is present in the atrial floor. (C) Specimen with double-inlet ventricle: both the mitral valve and the tricuspid valve drain into a morphologically left ventricle. (D) Specimen with absent left AV connection (mitral atresia): the mitral valve is absent, and the left cardiac chambers are hypoplastic. A: atrium; V: ventricle; IV: indeterminate ventricle; LV: left ventricle; RV: right ventricle.

Similarly, to have a general concept on pathophysiology of the congenital heart diseases anatomical collection of the congenital cardiac defects at Padova University are valuable. In the book of Prof. G. Thiene and colleagues well described the pathophysiological anatomy of different types of congenital cardiac diseases. According to this concept the first step in the segmental approach to diagnose congenital heart defect is to establish the platform of the heart, namely, the atrial situs and venous drainage.[14] Patients with single heart physiology including right ventricular hypoplasia with heterotaxy syndrome (asplenia or polisplenia) are significantly high risk patients for Fontan type palliation or even for 1.5 ventricle repair, which are also the findings of this study in patients with right ventricular hypoplasia.

In this contest it is considerable to have a general overview on different types of sequential chamber localization to understand the proper anatomy in patients with heterotaxy syndrome.

Sequential chamber localization

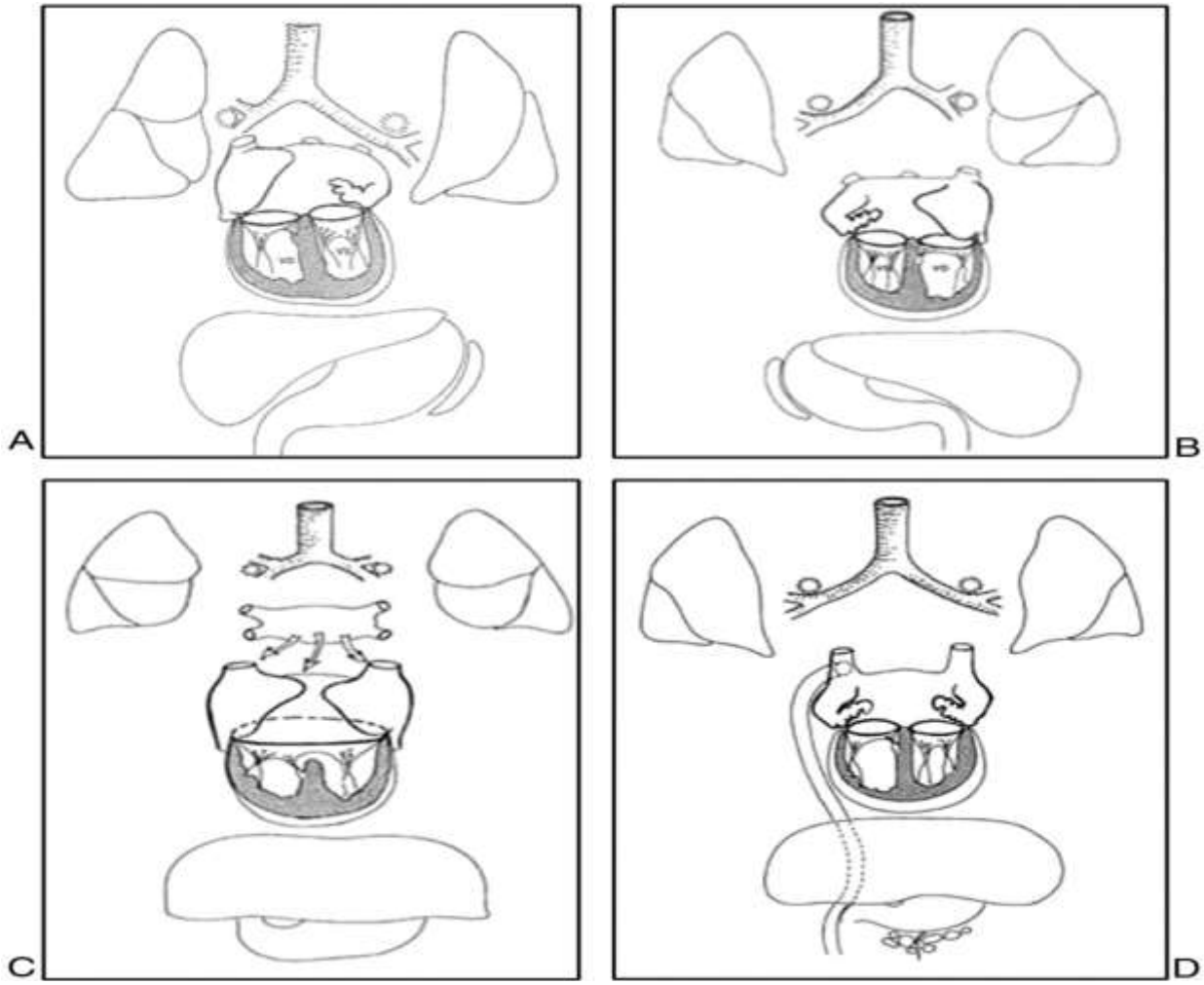


Fig.6: Carla Frescura, Siew Yen Ho, Gaetano Thiene

La collezione anatomica di cardiopatie congenite dell' Universita di Padova 1996

Atrial situs. (A) In situs solitus, the morphologically right atrium is located on the right side, and the morphologically left atrium is located on the left side; the trilobed lung and short bronchus are on the right side; and the bilobed lung and long bronchus are on the left side. (B) Situs inversus is the mirror image of situs solitus, with a reverse position of the atria and lungs. (C) In right atrial isomerism, both atria are of right morphology, the lungs are bilaterally trilobed, and the bronchi are short. Under the diaphragm, there is heterotaxia of abdominal organs, and the spleen tends to be absent (asplenia syndrome). (D) In left atrial isomerism, both atria present a left morphology, and the lungs are bilaterally bilobed with long bronchi. Under the diaphragm, there is heterotaxia of abdominal organs with multiple spleens (polysplenia syndrome).

SECTION II

CONGENITAL HEART DISEASES ASSOCIATED WITH RIGHT VENTRICLE HYPOPLASIA

Congenital heart diseases associated with right ventricle hypoplasia

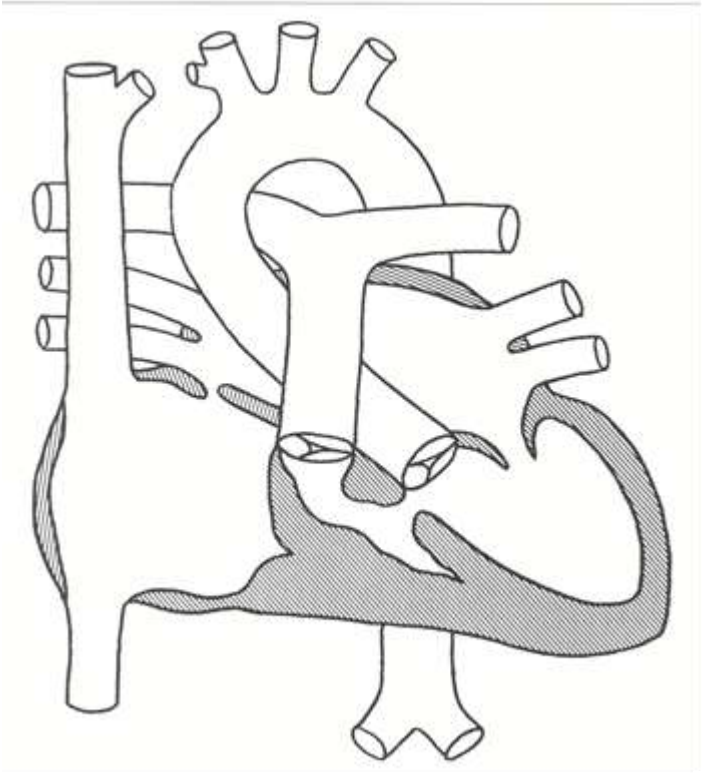


Fig.7: Right ventricle hypoplasia

A hypoplastic but potentially or partially usable right ventricle occurs in a variety of clinical situations. The disease spectrum includes patients with (1)) Ebstein's anomaly; (2) right ventricular outflow tract obstruction and a hypoplastic right ventricle; (3) pulmonary atresia with intact ventricular septum; (4) rare variants of tetralogy of Fallot; (5) tricuspid stenosis with atrial septal defect; (6) inlet ventricular septal defect and a straddling tricuspid valve; and (7) complete atrioventricular canal with (a) extreme left ventricular dominance, (b) tetralogy of Fallot, (c) double outlet of the right ventricle, or (d) pulmonary stenosis. When right ventricular hypoplasia is diagnoses in association with other congenital anomalies, pediatric cardiac surgeons are working to develop a more reliable and optimal surgical techniques to unload the right ventricle and increase the blood flow to the lungs.

Right ventricle hypoplasia can be associated to a wide spectrum of complex congenital cardiac malformations. All these anomalies are characterized by a normal functioning left ventricle, and a hypoplasic right ventricle which is not able to manage the whole cardiac output resulting in a various degree of inadequate blood flow to the lungs and signs of right ventricular failure.

Treatment strategies for these groups of patients include (1) the bidirectional Glenn shunt, (2) a Fontan type of repair, or (3) a biventricular repair with atrial septal fenestration and (4) 1.5 ventricle repair.

A. EBSTAIN'S ANOMALY

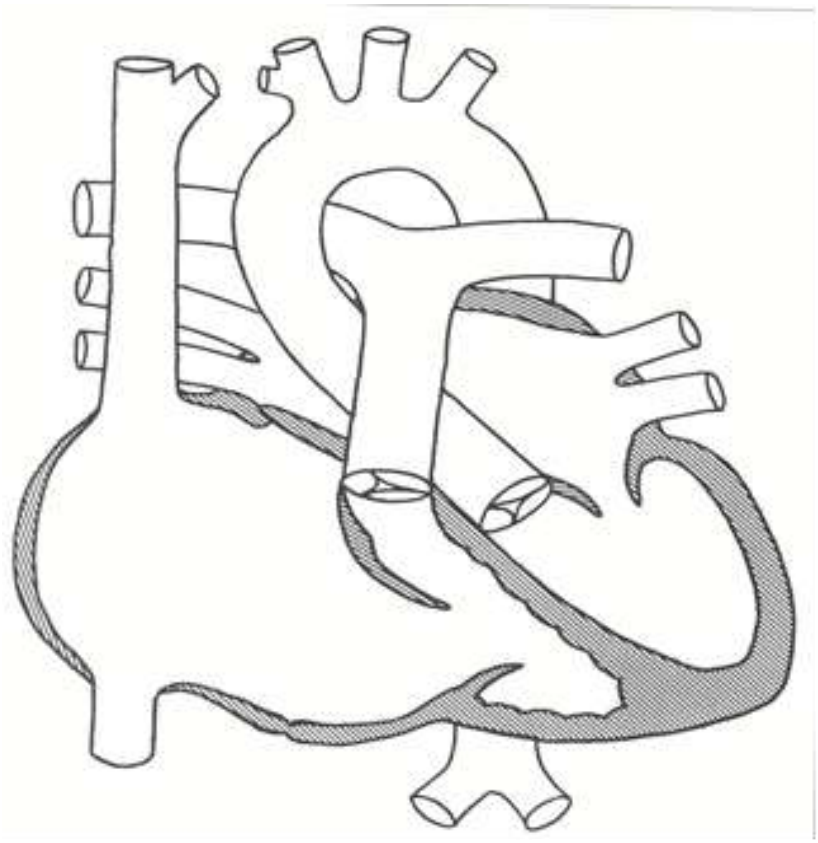


Fig.8: Ebstein's anomaly

Ebstein's anomaly of the tricuspid valve is a rare congenital malformation that accounts for less than 1% of all congenital heart defects [15-26]. The essence of the malformation is the downward displacement of the septal and part or whole of the posterior leaflets into the inlet portion. The apposition of the tricuspid leaflets is displaced towards the infundibulum and the apical trabecular portion of the right ventricle, creating the distal orifice of the tricuspid funnel.

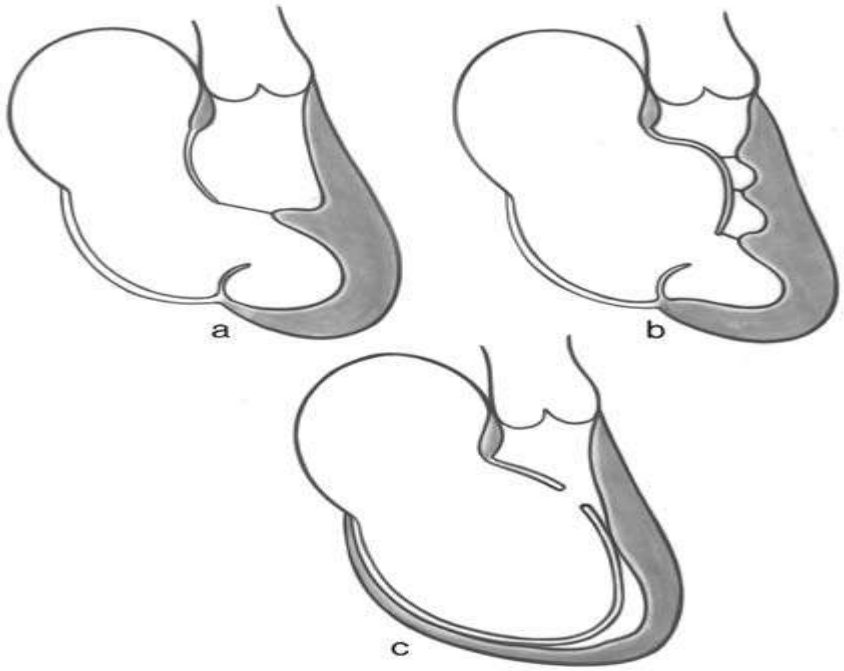
The part of the right ventricle beyond is the functional right ventricle; the part upstream is the atrialized inlet portion of the right ventricle (Figure 8). The tricuspid valve is often incompetent and, at the level of the distal orifice, sometimes stenotic. The anterosuperior leaflet seems to be in continuity with the posterior leaflet, without a clear commissure or attachment to the tricuspid annulus.

Often it is large and thin. Close to the anteroseptal commissure, the leaflet is usually freely mobile but as the observer scans towards the posterior leaflet, the ventricular part of the valve tends to be tethered and then fused to the right ventricular free wall, while the hinge point itself becomes progressively displaced towards the apex of the right ventricle and away from the tricuspid annulus. There is marked obliteration of the interchordal space, almost constituting a veil. Rarely, the normal anterior and medial papillary muscles can be identified; more often, the anterior leaflet is attached directly to the right ventricle cavity [16].

SURGICAL ANATOMY

Ebstein's anomaly has three characteristic features: (a) a downward displacement of the septal and posterior leaflets into the right ventricle; (b) an atrialized portion of the right ventricle between the tricuspid annulus and the attachment of the posterior and septal leaflets (inlet portion); and (c) a malformation of the right ventricle, where the functional right ventricle is small and thin-walled and often has an abnormal systolic function. The trabecular portion is reduced; the infundibular portion may be partially obstructed by excess valvar tissue attachments of the anterior leaflet to the infundibulum. This is best appreciated with magnetic resonance imaging (MRI). Depending on the severity of these malformations, Ebstein's anomaly can be categorized into four types[27.]

(Figure 9): types of Ebstein's anomaly



• Type a. The anomaly is limited to the displacement of the septal leaflet with a small atrialized chamber. The anterior leaflet is large, moves freely and has a free leading edge between the anteroseptal and anterolateral commissures. (illustrated on Figure 9).

• Type b. The anterior leaflet is restricted, with some interchordal space obliteration and attachment of the ventricular surface to the ventricle by fibrous bands. The posterolateral and anteroseptal

commissures are clearly delineated, the latter serving as a communication between the atrialized chamber and the functional right ventricle. The atrialized chamber is large Figure (9.b).

• Type c. The anterior leaflet is partially fused to the infundibulum and the trabecular portion of the ventricular edge of the valve is adherent to the ventricle, in continuity with the adherent septal and posterior leaflets. There is a marked displacement

of the posterior and septal leaflets, which may be severely hypoplastic. The atrialized chamber is large and has a thin, non-contractile wall. The functional right ventricle is small and its contractility may be diminished (Figure 9.c).

• Type d. No anterior or posterior leaflets can be recognized, as they are totally adherent to the ventricular wall. The tricuspid orifice is deeply displaced towards the infundibulum and is often stenotic. The right ventricle is entirely made of the atrialized chamber.

B. RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION AND HYPOPLASTIC RIGHT VENTRICLE

Pulmonary stenosis and pulmonary atresia with intact ventricular septum are classically described as two separate entities. However, there is a consensus that in the neonatal period, critical pulmonary stenosis and pulmonary atresia often have a similar clinical presentation, and hearts with pinhole pulmonary stenosis may indeed exhibit the anatomical features of right ventricular (RV) hypertrophy and cavity hypoplasia commonly seen in pulmonary atresia with intact ventricular septum. Right ventricular outflow tract obstruction (RVOTO) without RV hypertrophy is also included.

Anatomical Substrate:

Pulmonary Valve

In pulmonary atresia, the pulmonary valve is replaced by an imperforate, fibrous membrane. In some cases, prominent commissural ridges converge and meet at the centre of the imperforate valve. This is the form of imperforate valve observed in patients with severe infundibular stenosis or atresia. The other type of imperforate pulmonary valve is characterized by commissural ridges present only at the periphery of the valve, the centre being a smooth fibrous membrane, having the potential to bulge into the pulmonary trunk. This type of valve is usually associated with a lesser degree of right ventricular hypoplasia and with an open infundibulum.[28] In severe pulmonary stenosis, there is commissural fusion at the periphery of the valve. Centrally, there is a smooth fibrous dome with an orifice that is usually central but sometimes eccentric. In less severe forms, two, three or sometimes four leaflets are relatively well formed, with only partial commissural fusion. The valve is usually thickened and may have a myxomatous appearance, particularly in neonates and small infants. Pulmonary stenosis due to mucoid thickening of the valve leaflets without commissural fusion is known as pulmonary valvular dysplasia. The valve ‘‘ring’’ may or may not be hypoplastic. Hypoplastic ring is often seen in the Noonan syndrome. [29]

Pulmonary Arteries

A good-sized main pulmonary artery with confluent branches is the usual situation in pulmonary atresia with intact ventricular septum. Beyond infancy, post stenotic dilatation of the main and proximal left pulmonary arteries is a common finding in patients with pulmonary stenosis. Stenosis of pulmonary arterial branches can occur in isolation or in association with other cardiac malformations. These can be central or peripheral, discrete or tubular, single or multiple. Multiple peripheral pulmonary stenosis are often a part of the rubella syndrome. Right Ventricle and Tricuspid Valve in pulmonary atresia with intact ventricular septum, right ventricular cavity size is variable with different degrees of myocardial hypertrophy and tricuspid valve anomalies. Most patients have both right ventricular hypoplasia and hypertrophy, both of which may be severe. In these hearts, the size of the tricuspid valve annulus has been found to relate to the degree of cavity hypoplasia. In addition, the tricuspid valve leaflets are often thickened and chordae are abnormal in number and attachments. At the other end of the spectrum are those hearts with greatly enlarged right ventricular cavities and a regurgitant dysplastic tricuspid valve, with or without displacement (Ebstein's anomaly). In this rare anomaly the right ventricular wall is usually thinned and, on occasion, may be devoid of myocardium (Uhl's anomaly and pulmonary atresia). In valvar pulmonary stenosis, the obstruction produces concentric hypertrophy of the right ventricle, which may result in secondary infundibular stenosis. There is usually mild to moderate reduction in right ventricular cavity size, which is partly related to the concentric ventricular hypertrophy. Severe right ventricular hypoplasia or enlargement is rare. Isolated subvalvar pulmonary stenosis produced by a large anomalous muscle bands account for 10–20% of patients with pulmonary stenosis and intact ventricular septum. [30]

Right Atrium

An atrial communication is present in all cases of pulmonary atresia with intact ventricular septum and in most neonates with critical pulmonary stenosis. The communication is restrictive in about 5–10% of neonates with pulmonary atresia with intact ventricular septum.[31]

Coronary Arteries

A substantial number of patients with pulmonary atresia with intact ventricular septum and diminutive and hypertensive right ventricle have persistence of right ventricular myocardial sinusoidal coronary artery connections. [31,32] The fistulous communications can connect the hypertensive right ventricle to one or both coronary arteries. Connection to the left anterior descending coronary artery is the commonest. Sometimes the proximal connection between the coronary artery and the aorta is absent, and perfusion of the involved coronary circulation is retrograde from the right ventricle and is dependent on right ventricular hypertension (right ventricular-dependent coronary circulation). Intimal fibromuscular hyperplasia often occurs in the coronary arteries linked with the sinusoids and may be responsible for myocardial ischemia and infarction, involving either the right or the left ventricle or both.

Principles of surgical management

In recent years, a more aggressive approach towards early relief of right ventricular outflow obstruction was adopted. The aim is to provide pulmonary forward flow and stimulate right ventricular growth and enable the right ventricle to serve the pulmonary circulation whenever possible. In addition, there is good evidence that left ventricular function can be compromised by the presence of a hypertrophied, hypertensive right ventricle. [31, 33] We feel that right ventricular decompression should be considered, even if the right ventricle is too small to be incorporated later into a biventricular circulation. However, the right ventricle should not be decompressed in the presence of a right ventricular-dependent coronary circulation. Under these circumstances, the high-pressure right ventricle perfuses portions of the right and left ventricular myocardium with desaturated blood in systole. During diastole, the lower right ventricular diastolic pressure may allow a steal of blood from the coronary arteries into the right ventricle, to the detriment of the coronary circulation. Furthermore, the coronary circulation may be right ventricular-dependent in the case of a proximal discontinuity between one or both coronary arteries and the aorta and/or in the case of peripheral stenosis caused by intimal hyperplasia. Decompression of such ventricles may result in myocardial ischemia and/or infarction in the distribution of the involved coronary arteries. The decision between univentricular and biventricular repair depends on the morphological characteristics of the right ventricle, in particular the size of the right ventricular cavity and the presence or absence of coronary artery fistulae. The adequacy of the hypoplastic right ventricle for biventricular repair is difficult to assess and different methods have been used.

In patients with pulmonary atresia and intact ventricular septum, we have found it useful to classify the hypoplastic, hypertrophied right ventricles into three categories on the basis of the tripartite concept of the right ventricle [34,35] the hearts with all three components present; those with the absence of the trabecular portion; and those with an inlet zone only.

In these hearts, the size of the tricuspid valve annulus was found to relate to the degree of cavity obliteration. Studies of De Leval et al, [36] have suggested that a right ventricle whose tricuspid valve diameter is within 99% (three standard deviations) of mean normal is capable of providing adequate pulmonary blood flow and of being incorporated into a biventricular circulation. Our initial measurements of the tricuspid valve diameter were made on angiocardiograms. Currently they are made on two-dimensional (2D) echocardiograms. (Table 1) gives the mean normal values for a given body weight and their lower 99% confidence limits. The mean normals were derived from the studies of Rowlatt and adjusted by Bull et al.[35] and Hanley et al.[37] suggested using the z value(standard deviation units) of the tricuspid valve diameter.

Table .1: Measurements of valve diameter used in decision making.

Weight (kg)	Angiographically estimated diameter of tricuspid valve (mm)	
	Normal mean*	Lower 99% CL
2	13	5
3	16	8
4	19	11
5	20	12
6	22	14
7	23	15
8	25	1
9	26	18
10	27	19
12	28	20
14	29	21
16	30	22
18	31	23
20	32	24
25	34	26
30	36	28

CL, confidence limit.
∗ Derived from autopsy data of Rowlatt et al. (1963). [38]
Adjusted for angiographic assessment according to Bull et al. (1982). [35]

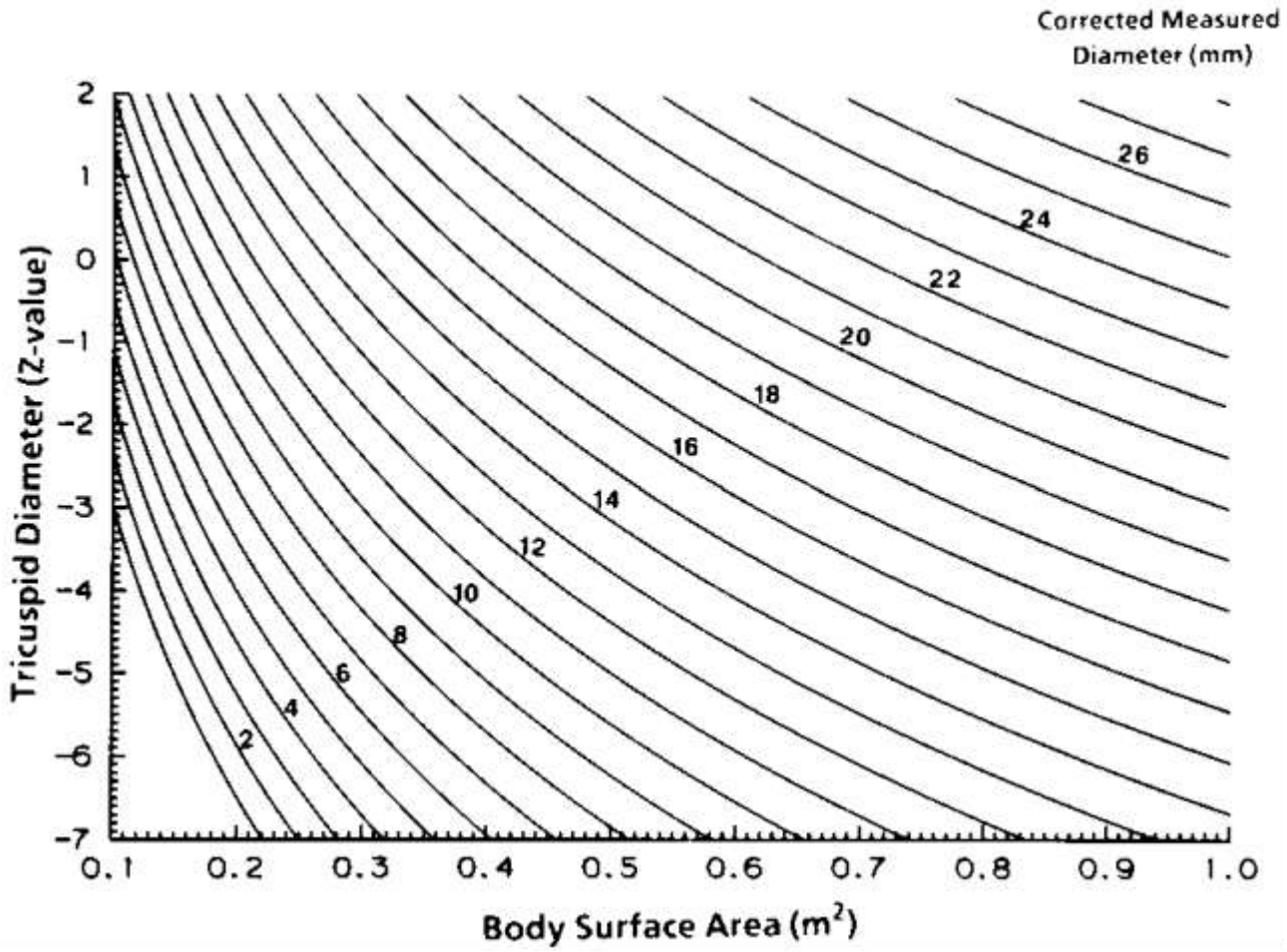


Fig.10. (J. Stark and V. T. Tsang.)

calculated as follows:

$$\text{z value} = \frac{\text{measured diameter} - \text{mean normal diameter}}{\text{standard deviation of mean normal diameter}}$$

The normal diameter is the diameter in a normal individual of the same body surface area. The data for the mean normal diameters and their standard deviations also come from the anatomical data of Rowlatt. A nomogram for use in deriving the z value of the tricuspid valve diameter from the echocardiographically determined diameter and body surface area published by Hanley et al. (1993) [37] is reproduced in Figure 10.

Patients with a three-portion right ventricle, whose tricuspid valve diameter is within three standard deviations of mean normal or whose z value of the tricuspid valve diameter is −1.5 or more, will have valvotomy either by interventional cardiological procedure or by surgery. The majority of these patients will not require a systemic-to pulmonary artery shunt. If there is infundibular stenosis in addition to the valvar obstruction, the patient should receive a trans-annular patch, placed using cardiopulmonary bypass. The patent foramen ovale is usually left open. By and large, in patients with a patent infundibulum whose tricuspid valve diameter is below the 99% (Figure 10).

Confidence limit of mean normal, there is infundibular stenosis in addition to the valvar atresia. These patients are best treated with a trans-annular patch without closure of the patent foramen ovale and with a concomitant systemic-to-pulmonary-artery shunt. In Hanley's protocol, this applies to patients in whom the z value of the tricuspid valve diameter is between −1.5 and −4. In patients without a patent infundibulum or those with a tricuspid diameter that has a z value of −4 or less, and those with significant anomalous coronary connections, a systemic-to-pulmonary artery shunt only is performed. A balloon atrial septostomy should be carried out in these patients if the atrial septal defect is restrictive.

In terms of timing of intervention, patients with right ventricular outflow tract obstruction and intact ventricular septum fall into one of the three categories:

1• The symptomatic neonate, who by and large will receive a palliative treatment.

2 • The child who survived palliation in infancy.

3.• The asymptomatic child with pulmonary stenosis.

Symptomatic Neonate with Critical Pulmonary Stenosis or Pulmonary Atresia Cyanosis is usually present in such infants on the first day of life and rapidly becomes more severe as the ductus arteriosus closes. The suspected diagnosis of critical pulmonary stenosis or pulmonary atresia with intact ventricular septum is easily confirmed non-invasively by 2D echocardiography. In some cases, the diagnosis may already have been made prenatally.[39] Echocardiography must provide the following information: size and type of right ventricular cavity; level of atresia (valvar or

infundibular and valvar); size and competence of the tricuspid valve; size and confluence of the pulmonary arteries; size of the interatrial communication; size of the patent ductus arteriosus; and left ventricular size and function. Cardiac catheterization and angiography are indicated when right ventricular sinusoidal coronary artery connections are suspected. A right ventricular angiogram will disclose connections between the right ventricular myocardial sinusoids and the coronary arteries. If there is retrograde opacification of the aortic root, there is certainly proximal continuity between the aorta and the coronary arteries. When dense opacifications of the coronary circulation from the right ventricle do not lead to visualization of the aortic root, it is necessary to exclude proximal discontinuity between the aorta and the coronary arteries.[31]

A left ventriculography or aortogram is then necessary to study the origin of the coronary arteries and to identify stenosis of the coronary branches linked to the sinusoids. The primary goal of treatment is to maintain an adequate pulmonary blood flow. This ensures survival beyond the neonatal period. In addition, treatment should aim to enable the right ventricle to serve the pulmonary circulation whenever possible, to enlarge a restrictive atrial septal defect, to decompress a hypertensive right ventricle, even if not large enough to serve the pulmonary circulation, and to deal with coronary anomalies if present.

Our current management protocol for neonates with pulmonary atresia and intact ventricular septum is as follows. Prostaglandin E is started, to maintain patency of the ductus arteriosus, as soon as the diagnosis is suspected. For patients with the smallest right ventricles, whose cavity is made of an inlet portion only, a right-modified Blalock–Taussig shunt is our procedure of choice. In patients with a patent infundibulum, relief of right ventricular outflow obstruction can be accomplished using a variety of procedures. In many centers, including our own, percutaneous pulmonary balloon valvuloplasty has replaced open valvotomy for most patients. Using mechanical, laser or radiofrequency energy, the valve is perforated and then dilated with a balloon. Pulmonary regurgitation introduced by the balloon dilatation is generally mild, is usually well tolerated and does not appear to be more severe than that introduced by surgical valvotomy. The prostaglandin

infusion is slowly reduced after the procedure and discontinued over the following few days. Surgical intervention may occasionally still be necessary for severely dysplastic valves that cannot be opened by balloon valvuloplasty. Surgical resection of the valve cusps with/without trans-annular patching may then be required. Under these circumstances the procedure is usually performed via median sternotomy on cardiopulmonary bypass.

Because postoperative right ventricular performance is not easy to predict, it may be difficult to know whether the patent ductus arteriosus and the atrial septal defect should be left patent or closed at the time of the operation. An outflow tract patch inevitably creates pulmonary regurgitation that interferes with right ventricular diastolic function. It may increase the right to- left shunt at atrial level, if the atrial septal defect is left open or causes congestive right ventricular failure, if it is closed. Occasionally, if cardiopulmonary bypass is contraindicated, closed surgical pulmonary valvotomy can be performed. The necessity and the timing of a systemic-to-PA shunt remains controversial in patients presenting with mild to moderate RV hypoplasia and a patent infundibulum . [40]

A smaller tricuspid valve size was the only predictive factor for the need of systemic-to-PA shunt after RV decompression. Management of patients with right ventricular to coronary artery connection remains difficult. Discrete fistulae can sometimes be ligated. Right ventricular thrombo exclusion was advocated [41] in patients with intramural connections, to prevent a steal phenomenon into the right ventricle. Patients with right ventricular-dependent coronary circulation might be inoperable and considered for heart transplantation.

Definitive Repair of Patients with Pulmonary Atresia with Intact Ventricular Septum:

The main strategies for definitive surgery are:[27]

1• Biventricular repair in patients with two adequate ventricles.

2• Single ventricle repair for very small right ventricle and/or right ventricle-dependent coronary circulation.

3 • **One-and-a-half ventricle for varying degrees of RV hypoplasia.**

4 • Transplantation in very high-risk patients for the above strategies.

Study of de Leval et al., (1985) [36] have suggested that a three-portion right ventricle with a tricuspid valve diameter above the lower 99% confidence limit can safely be used to support the pulmonary circulation. This would also apply to patients whose z value of the tricuspid valve diameter is −1.5 or more. Patients who fulfil these criteria, either since birth or as a result of a satisfactory decompression, will undergo radical repair, leading to a biventricular circulation. The operation consists of relieving any residual right ventricular outflow tract obstruction and closing the atrial septal defect and the extracardiac shunt, if present. Some or all of these procedures can be sometimes carried out by interventional cardiac catheterization. Recruitment of the right ventricular cavity by excision of hypertrophic muscle of both the trabecular and infundibular portions may render some ventricles suitable for biventricular repair. [42] An outflow tract patch enlargement or homograft valve placement may be necessary in some patients. If tricuspid valve regurgitation is present, repair of the tricuspid valve is undertaken.

Patients whose decompressed right ventricle has not grown sufficiently to provide a right ventricular dependent pulmonary circulation can benefit from a variety of procedures. If it is believed that the right ventricular cavity is large enough to deal with two thirds of the systemic venous return, the superior vena cava can be connected to the right pulmonary artery (classical Glenn procedure), and the inferior vena caval blood can be diverted to the left pulmonary artery via the right ventricle, following complete relief of any residual right ventricular outflow tract obstruction. The atrial septal defect should ideally be closed to separate completely the systemic and the pulmonary circulations.

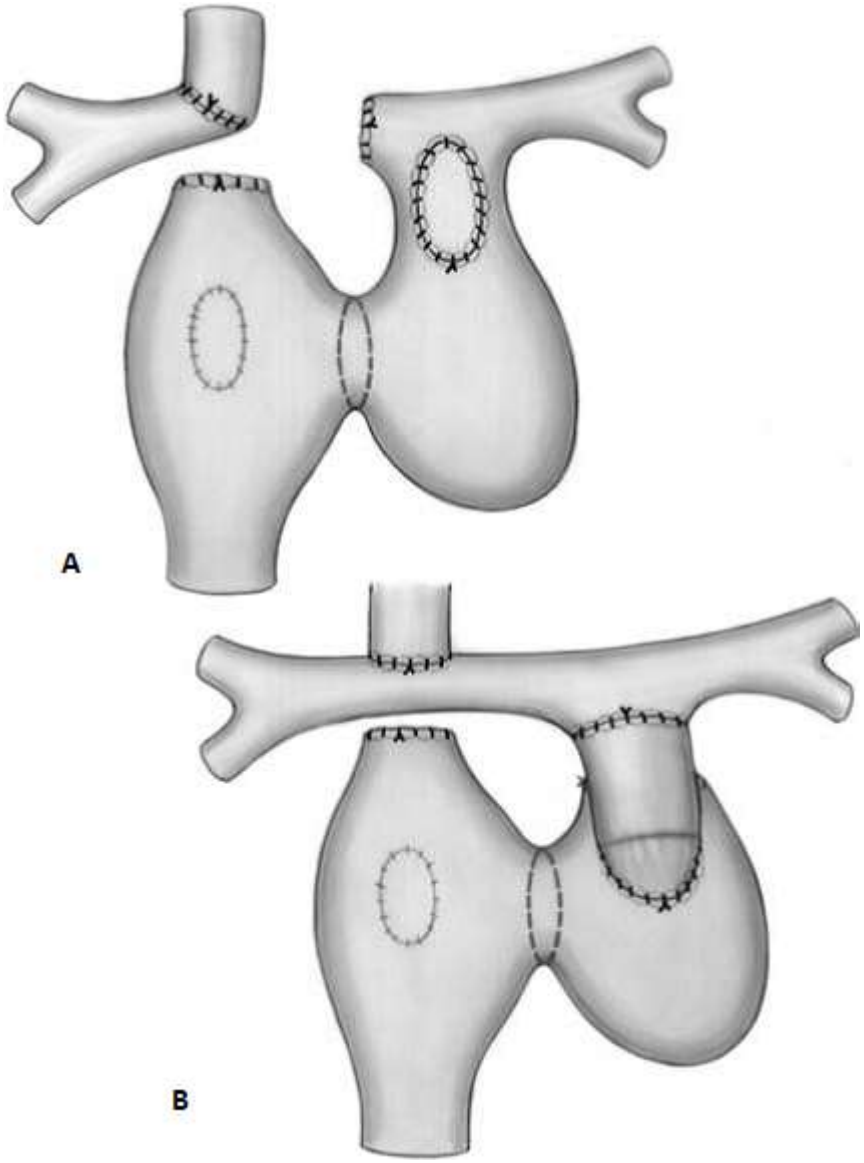


Figure 11. (J. Stark and V. T. Tsang.): A- one and a half ventricle repair with classical Glenn procedure; B- one and a half ventricle repair with bidirectional Glenn procedure.

Because of the known complications, classical Glenn procedure is not advisable.

An alternative approach for the creation of a one and- a-half ventricle circulation, suggested by Billingsley et al. (1989) [1], consists of performing a bidirectional cavopulmonary anastomosis, inserting a homograft in the right ventricular outflow tract, and closing the atrial septal defect (Figure 11).

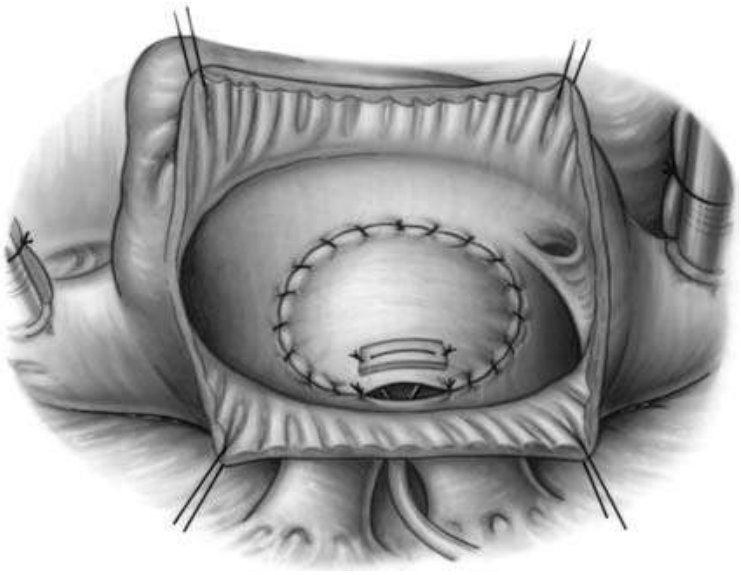


Fig. 12: (J. Stark and V. T. Tsang.): A- one and a half ventricle repair.

The ASD can also be left partially opened, with a small defect created centrally in the patch so that at a later date closure with a device would be possible. Alternatively, an adjustable atrial septal defect can be used [43] (Figure 12). The theoretical concern about one-and-a-half ventricle repair is that in ventricular systole the peak pulmonary arterial pressure can be higher than the superior vena caval pressure, thus producing systemic venous hypertension with retrograde flow in the superior vena cava. Incorporating too small a right ventricle in the circulation can become more of a hindrance than a benefit, and a Fontan type of procedure might be preferable.

Repair of Right Ventricular Outflow Tract Obstructions in Children

There is no unanimity concerning the level of right ventricular systolic pressure at which relief of right ventricular outflow obstruction is indicated. In general, pulmonary stenosis is well tolerated and the risk of sudden death is much lower than with obstruction to the left ventricular outflow. A cooperative study [44] recommended surgery in children with gradients greater than 50 mmHg. The advent of trans-catheter balloon valvuloplasty [45] has reduced considerably the number of patients who require surgical treatment for isolated pulmonary valve stenosis. For balloon valvuloplasty, the combination of right ventricular hypertrophy and a peak velocity of 4m/s would encourage most cardiologists in the UK to intervene.[46]

SECTION III

RIGHT VENTRICLE HYPOPLASIA AND ONE AND A HALF VENTRICLE REPAIR

RIGHT VENTRICLE HYPOPLASIA AND ONE AND A HALF VENTRICLE REPAIR

After experiments on the direct connection of the systemic venous circulation into the pulmonary arterial circulation, Glenn [47] demonstrated the clinical use of a superior vena cava (SVC)-right pulmonary artery (PA) shunt in 1958. Since then, a number of different cavopulmonary shunt for palliation of the cynotic heart diseases have been reported. Also in 1971 Fontan described cavopulmonary circulation as a surgical treatment for tricuspid atresia.[48] However these techniques facing so many postoperative problems and surgeons world widely modifying the techniques of the Glenn and Fontan. Patients after cavopulmonary shunts may have many complications [49, 50] like: pleural effusions, thromboembolism, arrhythmias, protein-losing enteropathy, plastic bronchitis, pancreatitis, liver cirrhosis, nephropathy with microalbuminuria, neurocognitive deficits, arteriovenous malformations and collaterals, reduced heart rate variability, endothelial dysfunction. Many of these complications are related with hypertension in inferior vena cava(IVC).

As continuation of the modification in cavapulmonary shunts, Billingsly et al. In 1989 [1] published the purpose of the so called " one and one half ventricular repair" to achieve a physiological correction by separating the pulmonary circulation from the systemic one, maintaining a pulsatile blood flow in the pulmonary arteries. It was obtained by unloading the insufficient right ventricle(RV). Blood from the SVC was diverted directly to the pulmonery artery as bidirectional cavapulmonary shunt (BCPS) and blood from the inferior vena cava (IVC) was pumped by 30% unloaded RV to the pulmonary artery.[51]

The main goal of the "one and one half ventricular repair" is to maintain a low pressure in the IVC district minimizing the complications related with IVC hypertension after Fontan procedure.[52] Theoretically, participation of the hypoplastic right ventricle in the pulmonary circulation can also restore ventriculoarterial coupling.

G. Stellin and colleagues [51,52] published their study about one and one half ventricle repair and described pulsatile pulmonary blood flow is more physiological than a continuous one, it maintains a low right atrial pressure and improves the systemic oxygen saturation.

Likewise, Shio Kim and colleagues described in their 38 years follow up study, in patient with SVC to PA anastomosis no clinically evident protein losing enteropathy or pulmonary arteriovenous fistula were identified among late survivors. [5] The one and one-half ventricle repair recruits the hypoplastic right ventricle, presumably providing kinetic energy and pulsatility to pulmonary blood flow, while the bidirectional Glenn reduces the volume load of the right ventricle.[52]

Therefore, as an alternative repair for right ventricle hypoplasia with complex congenital malformations to the complete by-pass of the right ventricle (diverting all venous return directly to the lung, according to the principle of Fontan), we believe that there is the possibility, where judged suitable, to utilize the pulsatile pumping capability of a small ventricular chamber, to maintain a more physiological pulsatile flow to the pulmonary vascular bed. This is obtained deviating the blood flow from the superior district of the body directly to the lung circulation by means of a cavopulmonary anastomosis (about 1/3 of the whole cardiac output), leaving to the hypoplasic right ventricle the possibility to manage the venous return from the inferior district of the body (about 2/3 of the whole cardiac output). This, in association to a complete repair of the intracardiac malformation, with a complete division of the systemic from pulmonary blood circulation (one and a half ventricle repair) allow to obtain a bifocal source of pulmonary blood flow (the laminar one from the superior district and the pulsatile one from the inferior district of the body).

The working hypothesis [53,54] for a one and a half ventricle repair has been that the benefits of a pulsatile pulmonary circulation may avoid some of the late complication of the Fontan procedure by maintaining a low pressure in the IVC district and those benefits outweigh the downside risk of having a pulsatile flow in the SVC district.

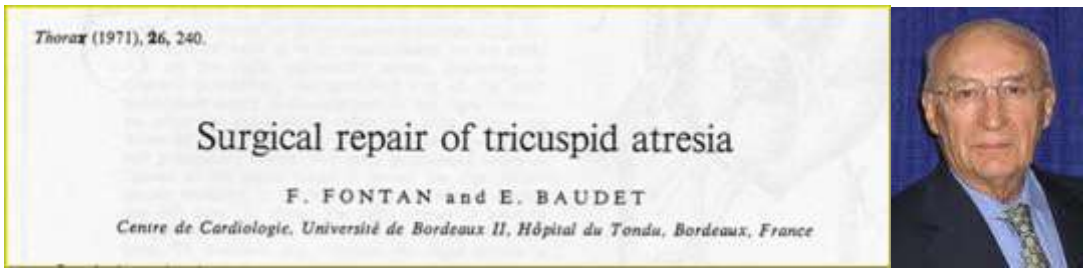
SECTION IV

PALLIATIVE SURGERIES FOR RIGHT VENTRICLE HYPOPLASIA

Palliative surgeries for right ventricle hypoplasia

A. Fontan procedure

Fontan procedure is a palliative surgical procedure used in children with complex congenital heart defects. It involves diverting the venous blood from the right atrium to the pulmonary arteries without passing through the morphologic right ventricle. It was initially described in 1971 by Dr Fontan and colleagues from France as a surgical treatment for tricuspid atresia. Fontan performed the first successful right heart bypass on a patient with tricuspid atresia in 1969.[55]


Thorax (1971), 26, 240.

Surgical repair of tricuspid atresia

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Fig. 13 **Prof. F. Fontan**

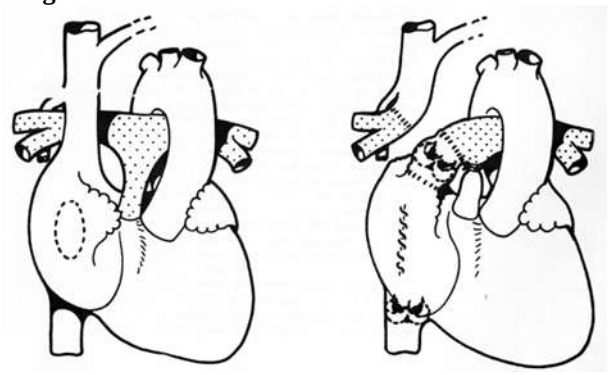


Fig.14: Thorax 1971, F.Fontan and E. Baudet.

It was designed to completely separate the systemic and pulmonary circulations and thereby to eliminate the systemic arterial hypoxemia and LV volume overload that are the hallmarks of this condition. It was recognized at the onset, however, that the elevated systemic venous pressures inevitably present after the Fontan procedure necessary to propel the blood through the lungs in the absence of a "booster pump" on the right side of the circulation might lead to late complications in surgical survivors. Also, functional limitations would likely still exist in these patients, particularly for physical activities that required a substantial increase in cardiac output.

More than 40 years have passed since Fontan's original surgical procedure, and it has undergone many technical modifications, an evolution that still continues, which have resulted in improved survival and a lower complication rate despite expanded application to high-risk patients.

However, despite continued improvements, the incidence of postoperative pleural effusions remains high and is a key determinant of postoperative length-of-stay. [56 - 60] Prolonged peri-operative course has been associated with the late development of protein-losing enteropathy and with decreased long-term survival, suggesting that improvements in peri-operative course might have both short- and long term benefits.[60] Factors previously shown to influence peri-operative morbidity and mortality include: anatomy, atrioventricular valve regurgitation, ventricular function, pulmonary artery pressure (PAP), support times, modified ultrafiltration (MUF), and Fontan type. [61-65] The experience of Fontan procedure for more than 4 decades with different modifications, these days majority of the pediatric cardiac surgeons considers stage approach in Fontan completion. Staged approach in Fontan surgery with recent modification like extracadiac conduit techniques and fenestration demonstrated good short and midterm outcomes. [66]

Staged approach in Fontan palliation

Hemi- Fontan or bidirectional Glenn procedure

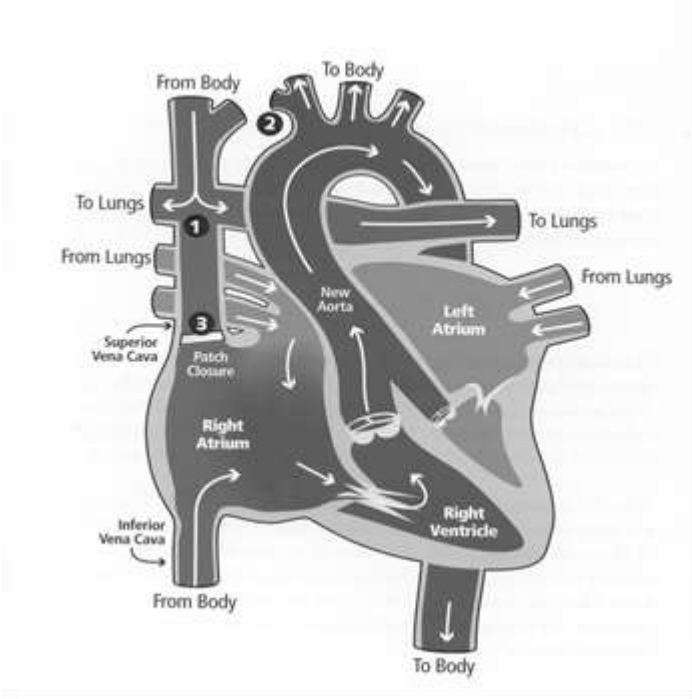


Fig:15. Hemi- Fontan or bidirectional Glenn procedure [67]

Surgical technique

After full median sternotomy the superior vena cava is fully mobilized. Attention must be paid to the right phrenic nerve during dissection. The azygos vein is ligated or divided between ligatures to prevent a run-off from the higher pressure SVC to the lower-pressure IVC. If there is interruption of the IVC with azygos continuation, the azygos vein must be kept patent. The main and right pulmonary arteries are then dissected free from the surrounding tissues. If present, the shunt is also dissected and closed through the posterior pericardium after cardiopulmonary bypass has been established. It is useful to place marking sutures on the SVC and right pulmonary artery before bypass, because after the vessels have been divided it may be difficult to re-establish their normal alignment, and twisting or kinking of the anastomosis may occur.

The aortic cannulation purse-string suture is placed as usual. One purse-string suture is placed on the right atrial appendage, and a second on the SVC near the innominate vein. The aorta is cannulated after full heparinization. A cannula is introduced into the right atrium through the right atrial appendage. Cardiopulmonary bypass is initiated, and the SVC is cannulated with a right angled venous cannula. The operation is performed on the beating heart with mild hypothermia. The cavopulmonary anastomosis is then carried out with a running suture of 6-0 prolene, which is interrupted in two areas to avoid a purse-string effect of the suture line and to maintain a wide anastomosis. The patient is rewarmed, and the bypass is discontinued. Additional sources of blood supply, such as the forward flow across a stenosed pulmonary outflow tract or a left systemico-pulmonary artery shunt, are usually left patent to maintain higher systemic oxygen saturations. In the case of bilateral superior vena cava, it is necessary to do a bilateral bidirectional cavopulmonary anastomosis, unless there is a large bridging vein and one of the two cavae is small. Both cavae must be cannulated separately. The hemi-azygos vein is ligated unless there is hemi-azygos continuation of the IVC, and the left cavopulmonary anastomosis is constructed as described above for the right side. Some surgeons prefer to avoid direct cannulation of the SVC, particularly if the vessel is small, and to perform the cavopulmonary anastomosis during deep hypothermic circulatory arrest. Another technique for the construction of a superior cavopulmonary anastomosis is the use of a temporary shunt between the superior vena cava and the right atrium, thus avoiding the use of cardiopulmonary bypass and its deleterious effects.

Fonatn: Lateral tunnel or intra-cardiac tunnel technique

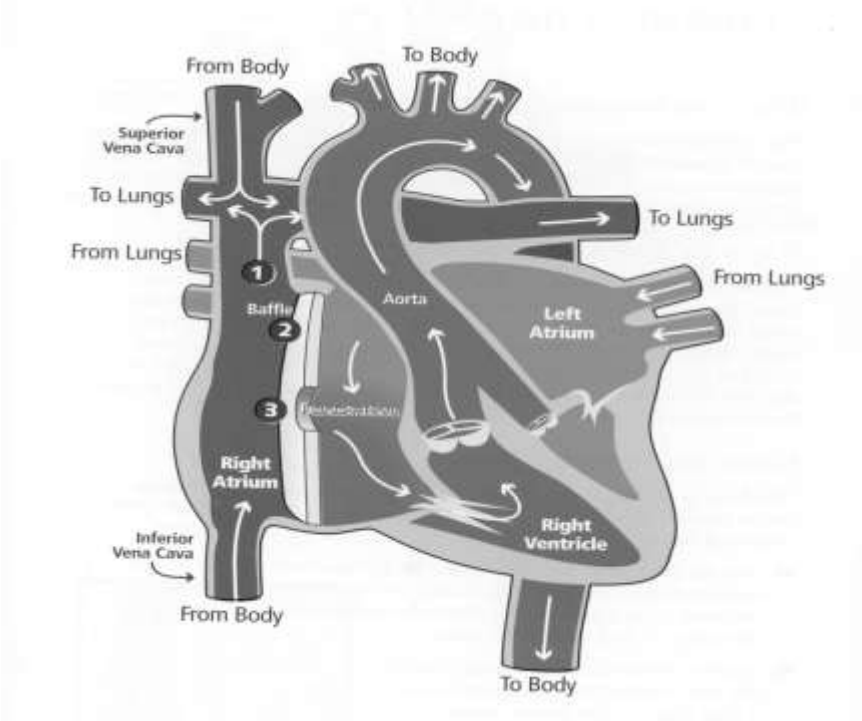


Fig:16. Fonatn: Lateral tunnel or intra-cardiac tunnel technique [67]

Surgical technique

A bidirectional Glenn shunt may already be in place, otherwise the beginning of this operation is similar to that described for bidirectional Glenn procedure using cardiopulmonary bypass. A piece of pericardium is harvested in case of its need as an only patch on the pulmonary artery. The SVC is fully mobilized, taking care not to damage the right phrenic nerve. The azygos vein is also dissected and may be later temporarily occluded with a bulldog clamp during the anastomosis of the SVC to the pulmonary artery. The main pulmonary artery and its bifurcation, the right and left branches, are circumferentially dissected and mobilized beyond the lateral pericardium and their branching. The ligamentum arteriosum is doubly ligated and divided. If present, the systemic-to-pulmonary artery shunts are controlled in the usual manner. A right shunt will be disconnected from the pulmonary artery, as described above. The ascending aorta is cannulated.

The cavae are cannulated with right-angled cannulae superiorly near the innominate vein and inferiorly at the cavoatrial junction. Cardiopulmonary bypass is initiated, and the patient is cooled to 28–30◦C. The dissection of the pulmonary arteries and the shunts is completed if necessary. A snare is snugged around the SVC, or alternatively the vein is controlled with a vascular clamp placed just proximal to the cannula. Another vascular clamp is applied just above the SVC/atrial junction, and the SVC is transected. The bidirectional cavopulmonary anastomosis is then constructed on the beating heart, as described earlier. When the anastomosis is nearly completed, the patient is cooled to 28◦C. The aorta is cross-clamped, and cold cardioplegic solution is infused.

A needle vent is placed in the ascending aorta. The main pulmonary artery is transected. To prevent bleeding and aneurysm formation, the proximal stump is closed with a double row of continuous 5-0 polypropylene suture, reinforced with two small Teflon felt strips, and the pulmonary valve leaflets incorporated in the suture line. If the distal main pulmonary artery is not used for the cavopulmonary connection, it is closed with a patch, so as not to narrow or distort the branch pulmonary arteries. The right atrium is then opened. The atrial septal defect is left open or is sometimes enlarged when the left atrio-ventricular valve is atretic or stenotic, to allow the pulmonary venous return to reach the right atrio-ventricular valve in an unobstructed fashion.

The intra-atrial cavo-caval pathway may consist of the interposition of a conduit of polytetrafluoroethylene (Gore-Tex) between the caval orifices, or the creation of a composite conduit made of the lateral atrial wall and a prosthetic patch, or the construction of an atrial baffle, establishing the continuity between the IVC and SVC.

Conduit insertion.

The right atrium is opened along the crest of the appendage. The bypass flow is reduced to quarter flow. The IVC cannula is clamped and removed and the IVC snare released. A sump sucker is placed into the IVC through the purse-string suture of the cannulation site . This allows good visualization of the IVC orifice from within the right atrium. The length of the atrial conduit is measured between the Eustachian valve and the crista terminalis. The conduit should be as short as possible. A Gore-Tex prosthesis of at least 18mm is then sewn in the IVC at its junction with the right atrium, with a running suture of 4-0 or 5-0 polypropylene. The conduit is then trimmed obliquely and shortened, to be sewn on the prominent ridge made by the crista terminalis in front of the SVC. The right atriotomy is then closed with a running 5-0 Prolene suture.

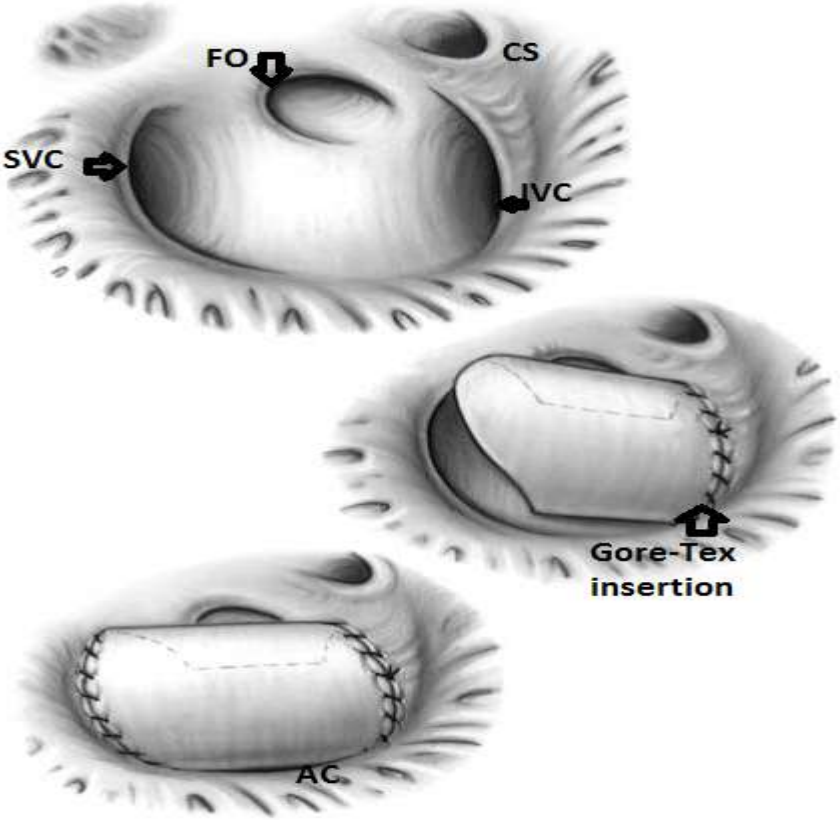


Fig:17- Conduit insertion in lateral tunnel technique [27] . SVC- superior vena cava; IVC- inferior vena cava; FO – fossa ovalis; CS – coronary sinus; AC- after completion of the lateral tunnel technique repair.

When a lateral tunnel is created, a Gore-Tex conduit of at least 16mm in diameter is sized as before and split longitudinally. The baffle is sewn halfway around the junction of the IVC with the right atrium, along the posterior atrial wall, crista terminalis and half-way around the junction with the SVC. Care is taken to avoid the sinus node. The right atriotomy incision is then closed with a running suture of 5-0 polypropylene. The rest of the operation is identical in all cases. The IVC cannula is replaced, and the perfusion flow is increased.

The distal opening of the main pulmonary artery is enlarged, making an incision toward the right pulmonary artery. Alternatively, the anastomosis can be made on the undersurface of the right pulmonary artery only. At initial period of our experience, we used to make an incision on the medial aspect of the SVC to enlarge it to the diameter of the IVC. This does not deal with the restriction produced by the crista terminalis within the right atrium. If this incision was carried out into the right atrial chamber, there would be a risk of damaging the sinus node artery in a significant proportion of patients. We do not enlarge the cardiac end of the SVC. The anastomosis between the cardiac end of the SVC and the pulmonary artery is done with a running suture of 5-0 or 6-0 polypropylene. The suturing starts with the right pulmonary artery, taking wider bites on the pulmonary artery than on the SVC, so as to make up for the discrepancy in the diameter of the two structures. Usually, this anastomosis can be made without a patch. However, it is sometimes necessary to use a small pericardial patch medially . It is a mistake to simply anastomose the transected main pulmonary artery and the transected SVC, as this distorts the pulmonary artery, which has to be brought right-ward and anteriorly. The patient is fully rewarmed, and air is evacuated from the heart. It is particularly important to aspirate the air from that portion of the right atrium that is now part of the systemic circulation. A left atrial line is inserted into the right upper pulmonary vein or directly into the left atrium before cardiopulmonary bypass is discontinued. In patients who have had a prior hemi-Fontan operation, the beginning of the operation proceeds in the same way. Following opening of the right atrium the previously patched SVC orifice is opened.

Placement of the intra-atrial tunnel to complete the total cavopulmonary connection then proceeds as described above.

Extra-cardiac or external tunnel technique with fenestration

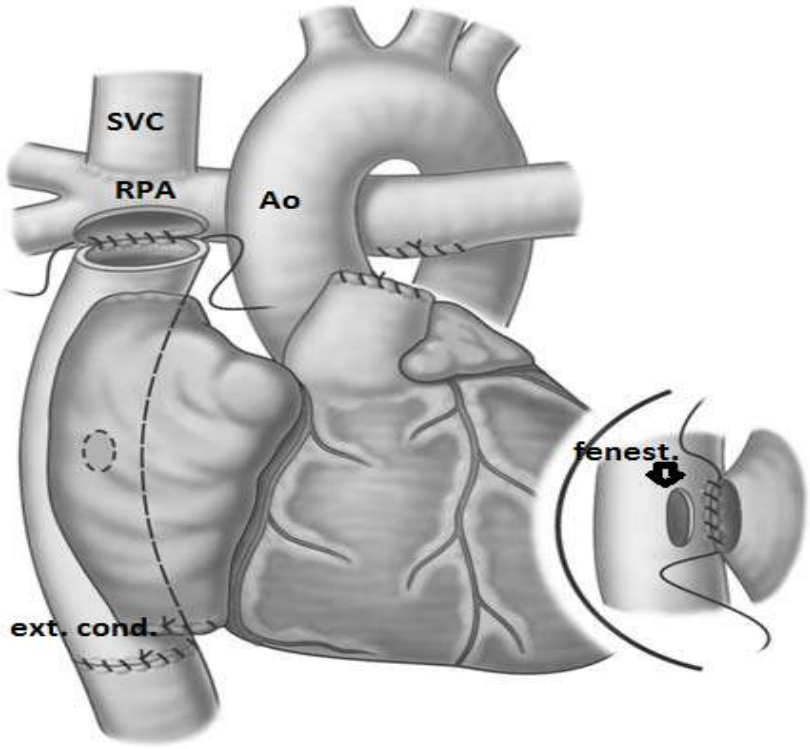


Fig: 18. Fonatn: Extra-cardiac or external tunnel technique with fenestration[27:] SVC- superior vena cava; RPA- right pulmonary artery, Ao- aorta, ext. cond.- external conduit, fenest.- fenestration.

Surgical technique

In selected patients the operation can be performed without exposing the patient to the damaging effects of cardiopulmonary bypass. [68] In patients without a prior SVC-to-pulmonary artery connection, this is performed using a temporary shunt from the SVC to the right atrium. For completion of the extracardiac total cavopulmonary connection (TCPC), the anastomosis between the conduit and undersurface of the pulmonary artery is performed. Next segment of the central pulmonary arteries is isolated by placing two clamps. One is from the medial aspect of the

cavopulmonary shunt obliquely across the right pulmonary artery, thus diverting blood flow from the shunt to the right lung only; care has to be taken not to obstruct the shunt flow, which is necessary to provide systemic oxygenation. The other clamp is placed across the left pulmonary artery.

Following completion of the anastomosis, the clamps are replaced with a single clamp half-way on the conduit and bidirectional cavopulmonary shunt is re-opened. Decompression of the IVC for the distal anastomosis is achieved by constructing a temporary right atrial-to-IVC shunt, by connecting two venous cannulae together.

ADDITIONAL PROCEDURES

Fenestration of the Intra-atrial Baffle

Early mortality after the Fontan procedure can be related to a transient haemodynamic disturbance, such as a postoperative increase in the pulmonary vascular resistance or systemic ventricular dysfunction. Whatever the cause of the failure, systemic venous hypertension is a common denominator that in itself may be responsible for a vicious circle. It causes capillary leakage in the interstitial compartment and in the peritoneal, the pleural, and sometimes the pericardial cavities. Systemic venous hypertension is also responsible for hypertension in the lymphatic system, including the lungs, which may become congested, with further increase in the pulmonary vascular resistance. Immediately after a Fontan procedure, there is usually a reduction in the end-diastolic volume, with significant increase in ventricular wall thickness and stiffness.[69]

This is in the presence of inflow restriction to the left ventricle resulting from the increased pulmonary vascular resistance, another step in the vicious circle. An ingenious and elegant method to prevent this vicious circle was simultaneously developed by the Boston group [70] and the Los Angeles group [71] for patients at increased risk. The technique consists of fenestrating the atrial baffle or conduit or making an adjustable atrial septal defect. Right-to-left shunting through this defect allows the cardiac output to be maintained at the expense of some arterial desaturation and prevents undue elevation of the systemic venous pressure. The size of the interatrial communication

can be calculated from the Gorlin formula. If one-third of the systemic venous return is diverted to the left atrium, the arterial oxygen saturation will be greater than 85%, assuming a mixed venous oxygen saturation of 60%. For a right atrial pressure of 15mmHg and a left atrial pressure of 10 mmHg, and if one-third of the venous return traverses the atrial communication, a defect of 6 mm/m2 is required. In practice, a defect of 4–6mm in diameter unloads the systemic venous circulation while the systemic arterial saturation remains around 85%.

In patients with an intra-atrial baffle, a fenestration is made in the Gore-Tex conduit with a 4–6mm punch. The adjustable atrial septal defect is rarely used any more. If an extracardiac conduit has been used, a 4–6mm hole is cut in the Gore-Tex tube and a slightly larger hole in the opposite free atrial wall. To avoid obstruction of the fenestration by atrial tissue, the atrial wall is anastomosed to the Gore-Tex tube a few millimeters away from the margin of hole in the prosthesis. In a high percentage of cases, the fenestration can be subsequently closed using a trans-catheter technique. In their early experience, Bridges et al [70] were able to close the fenestration in 11 of 19 patients within 20 days of surgery. A test occlusion is first carried out. A rapid fall in the right atrial saturation with test occlusion is an indication to leave the fenestration open. In some of their cases, failure to tolerate test occlusion was attributed to residual distal pulmonary stenosis, which were relieved by transcatheter dilatation; to aortopulmonary collaterals, which were embolized; or to systemic ventricular dysfunction, which improved with time and medical management. We have also adopted this technique of baffle fenestration for high-risk patients. This includes patients who had undergone banding of the pulmonary artery and patients with borderline pulmonary vascular resistance and/or systemic ventricular dysfunction.

Recent development of Fontan procedure

After a successful Fontan operation, their physical performance is most often improved. However, Fontan et al. mentioned in 1990, that the encouraging initial early postoperative results should be re-evaluated for long-term morbidity [72]. Modifications of the Fontan procedure and staging with bidirectional cavopulmonary shunt have extended the indications for this operation to a wide range of congenital heart defects unsuitable for biventricular repair [73].

In spite of significant improvement in outcomes, most of the congenital heart departments managing patients with complex congenital heart disease report contemporary mortality rates associated with the Fontan operation up to 7%. [74] Perioperative mortality has been attributed to ventricular dysfunction, stroke, thromboembolism, and multi-organ dysfunction. Numerous investigators have sought to identify patient factors and procedural factors associated with increased risk of mortality .[75] In patients with hypoplastic right ventricle or with borderline right ventricle one and a half ventricle repair is considerable as an alternative to Fontan procedure.

B. ONE AND A HALF VENTRICLE REPAIR

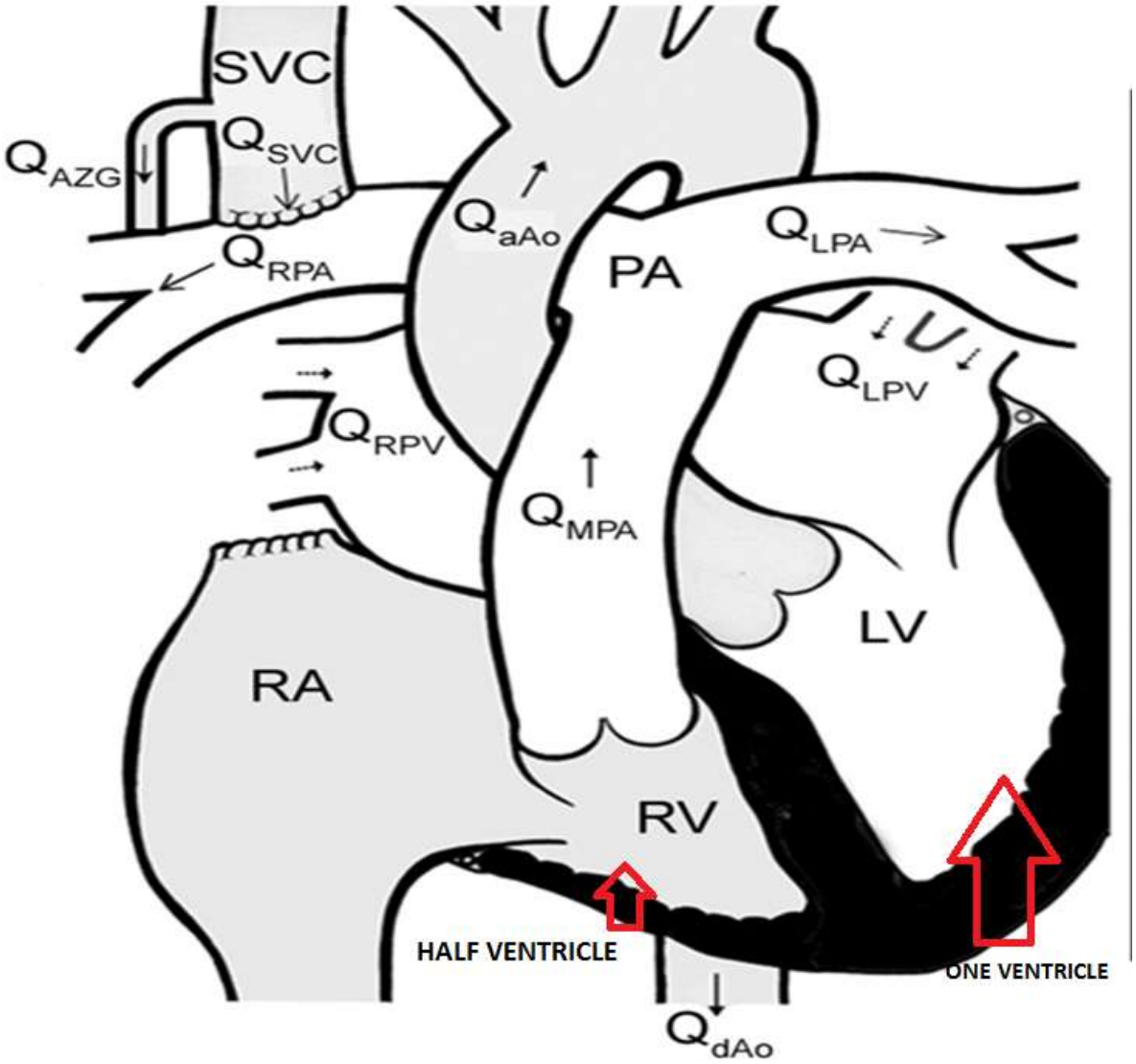


FIG:19. (FROM smilenaire.com, GOOGLE): Q-flow, SVC- superior vena cava, AZG- azygos vein, RPA- right pulmonary artery, LPA- left pulmonary artery, MPA- main pulmonary artery, RV- right ventricle, LV- left ventricle, LPV- left pulmonary vein, RPV- right pulmonary vein, PA- pulmonary artery, Half Ventricle- right ventricle which receives only half of the venous blood of the body which comes from inferior vena cava, blood from superior vena cava goes directly to the RPA. One ventricle means- full or ventricle which receives whole arterial blood; by this way creates the one and a half ventricle.

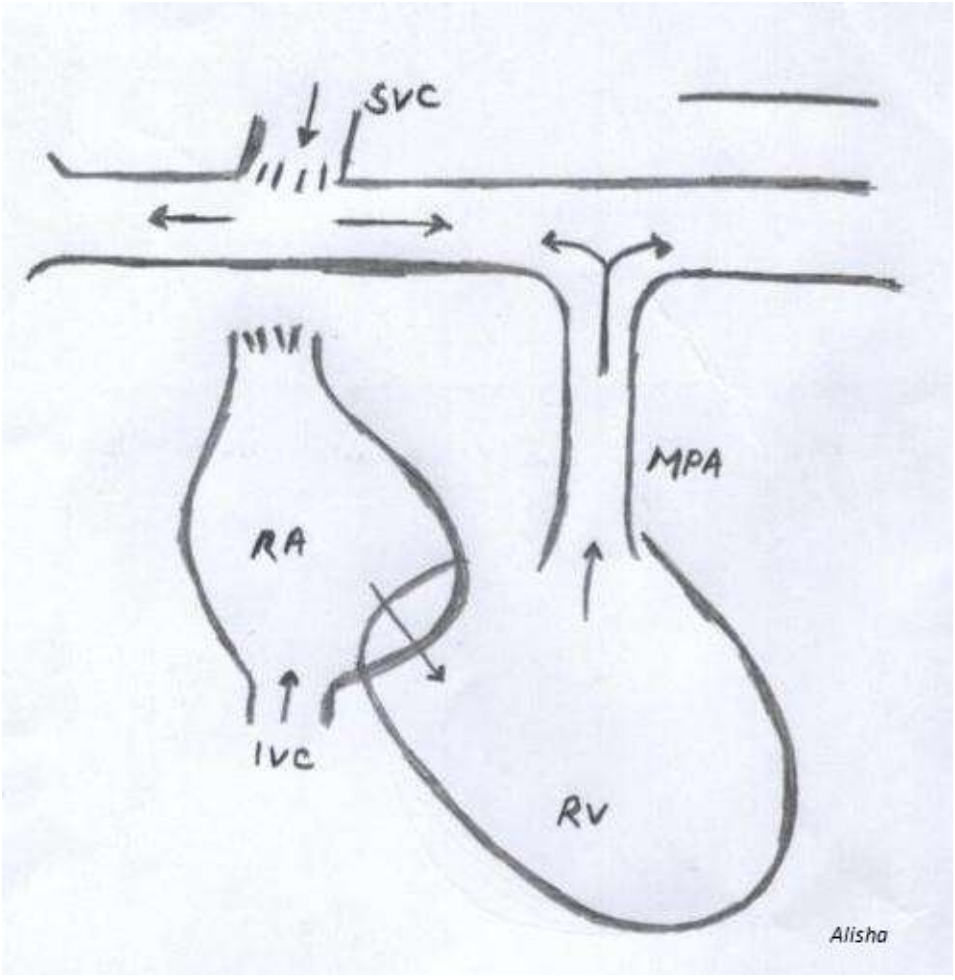


Fig.20: so called "Half ventricle" alone: SVC- superior vena cava, RA- right atrium, IVC- inferior vena cava, RV- right ventricle, MPA- main pulmonary artery: arrow indicates the blood flow.

The one and half ventricle repair consists of a superior cavopulmonary anastomosis combined with the connection of the dysfunctional right ventricle to the pulmonary artery through either a right ventricle outlet tract (RVOT) with the closure of the atrial septal defect in order to allow for desaturated inferior vena cava blood to enter the dysfunctional ventricle and flow to the pulmonary arteries or just direct connection of the SVC to right pulmonary artery and allowing flow of the blood coming from IVC to main pulmonary artery through hypoplastic right ventricle.

The one and a half ventricle repair can be considered the last stage of the functional single ventricle pathway, in that the systemic and pulmonary circulations are entirely separated, and there is no compelling reason to perform a more traditional Fontan operation. A fenestrated one and a half ventricle repair can be performed in patients with significant risk factors, and consists of a superior cavopulmonary anastomosis with connection of the dysfunctional ventricle to the pulmonary arteries, along with a 4-5 mm snared fenestration in the atrial septum.

Since the original contributions of Billingsley et al [1] to the one and a half ventricle concept in patients with hypoplastic right ventricle, the indications for this type of repair have expanded to cover a variety of anomalies that feature a functionally abnormal right ventricle that would not be able to cope with the entire pulmonary circulation.

In recent years, the tendency has been to submit such patients to the so-called "one and a half ventricle repair". In this procedure, the heart is surgically septated. This might include one or more of the following intra-cardiac procedures: atrial septal defect closure, ventricular septal defect closure, atrioventricular canal repair and removal of the Rastelli type left ventricular - aortic valve baffle and the right ventricular - pulmonary artery conduit, thus, creating a superior cavopulmonary shunt. The later component of the procedure eliminates all systemic-pulmonary mixing and provides preload reduction for the limited right heart, thereby, avoiding right heart failure.

Patient selection for the one and a half ventricle repair comes under consideration in two specific clinical border zones. In one, the patient is a borderline candidate for the two ventricle repair, and the surgeon "backs off" to the one and a half ventricle repair to avoid postoperative right heart failure. Examples might include the patient with pulmonary atresia and intact ventricular septum with a right ventricle that is reasonably well developed but demonstrates morphological and/or physiological characteristics. At the other clinical border zone, the patient is clearly not a candidate for the two ventricle repair, and the focus over the course of the patient's life has been toward achieving a Fontan operation; however, the internal morphology of the heart does allow a surgical septation. [76-80]

SECTION V

ORIGINAL STUDY

ORIGINAL STUDY

1. Aim of the study

a. Clinical part

To analyze short term and long term outcomes of the patients who operated by one and a half ventricle repair technique at our institution.

b. Animal model

1. To evaluate the competition of flows at the superior vena cava – right pulmonary artery junction, to understand the hemodynamic interaction of a pulsatile flow in combination to a laminar one.
2. To evaluate the effects of venous hypertension on the superior vena cava districts by evaluating the oxygen saturation.
3. To evaluate the advantages of bifocal pulmonary blood flow (mixed laminar and pulsatile) on pulmonary microvascular circulation in comparison to a whole laminar pulmonary blood flow (Fontan repair).
4. Evaluation of the advantages of the 1.5 ventricle repair on maintaining low pressure in the inferior vena cava district.

2. MATERIAL AND METHODS

a. Clinical study

We retrospectively analyzed our institutional database and reviewed the entire experience with 1.5 VR of congenital heart defects between March 1994 and January 2012. Patients' characteristics are shown in table 2 and diagnosis leading to surgery shown in table 3.

All available clinical and operative data were reviewed. Review of medical records was approved by hospital local committee on clinical investigation. All patients were studied preoperatively by means of a two-dimensional (2D) – echocardiography and Doppler, in order to evaluate TV and RV dimensions and RV function. TV assessment was made by obtaining the larger annulus diameter measured from a standard 2D – Echocardiography apical four chamber view, this was indexed for body surface area and then compared to normal values for body surface area. Cardiac catheterisation was also performed in all, before repair, to assess cardiac pressures and calculate pulmonary vascular resistances (PVR). The preoperative cardiac rhythm was established for all patients by EKG; other clinical data gathered at admission included: venous blood oxygen saturation ($SatO_2$), presence and degree of cyanosis, medical therapy. Any early postoperative adverse events were registered and all patients received a 2D-Echocardiography qualitative evaluation of right and left ventricle contractility; we also evaluated drugs administrated and cardiac rhythm at discharge. Follow-up was completed and updated in all, with a mean of 5.7 years (range 0.2 – 12 years). All the patients were followed with clinical evaluations and 2D-ecocardiography and Doppler. Exercise stress test at cicloergometer was performed in one patient with Ebstein's anomaly.

Patients were divided in: Group A - patients with right ventricle hypoplasia/dysfunction and malformations of tricuspid or pulmonary valves; Group B - patients with associated complex malformations involving other cardiac structures.

Table: 2. Patients' Characteristics

	Total	Simple(groupA)	Complex(groupB)
Pt. NO	**24**	**14**	**10**
Sex(M/F)	**12/12**	**10/4**	**2/8**
Weight	**5.6-83 kg**	**5.6-83kg**	**6.8-18.6 kg**
Age	**4m.-66.8**	**5m-66.8yr**	**7 m-8 yrs**

Table: 3. Diagnosis leading to surgery

Group A(Simple)	group B (Complex)
TV and RV hypoplasia, DIV, Pulmonary valve stenosis	CAVC+TOF+TV-RV hypopl
Ebstein anomaly of TV	inlet VSD+TV straddling and overriding+sub aortic stenosis+aortic arch hypoplasia, DORA
Ebstein anomaly of TV	TV-RV hypopl+RVOTO by fibromuscolar tissue of ostium infundibuli+VSD
TV regurgitation + Dysfunctioning RV	{S,L,L} DORV con DIV subpolmonare, L-TGA, TV straddling II tipo, CoA severea con ipoplasia arco aortico, PDA
TV-RV hypopl	DORV+VSD uncommitted+CAVC+LSVC+PAPVR
PA-IVS+TV-RV hypopl	DORV+VSD subAo+stenosi marcata sottovalvolare polmonare e discontinuità arteria polmonare sx
Ebstein anomaly of TV	TV_RV hypoplasiaTAPVC+TASVR+VSD+PFO
PA-IVS+TV-RV hypopl	inlet VSD+TV straddling and overriding+L-TGA+RV hypoplasia
PA-IVS+TV-RV hypopl, PDA	Truncus con interruzione dell'arco aortico+TV-RV hypopl
PA-IVS+TV-RV hypopl	CAVCp+TV-RV hypopl+destrocardia
inlet VSD+TV straddling and overriding+RV hypoplastic for apical absence	
Ebstein anomaly of TV	
Ebstein anomaly of TV	
inlet VSD+TV straddling+RV hypoplastic for apical absence	

1. Statistical analysis

Overall survival at 18 years for all patients is 85%. Survival is evaluated according to different variables. Survival estimate is 90% at 18 years for group A and 75 % for group B p value = 0.96(Figure 34). Overall freedom from adverse events, surgery and interventional procedures was 56.5 %, 87 % and 73,9 % respectively. Freedom from adverse event in Group A (n=12) was 83.3 % while in Group B (n=11) was 27.3 %. Statistical analysis demonstrated that Group B had a significantly lower freedom from adverse events than Group A (p = 0.015). Overall survival were estimated by the Kaplan-Meier method (figures 34-35). Log-rank test was used for comparisons between groups. Statistical findings were considered significant if the critical level was less than 5% (P-value < 0.05). Statistical analysis was performed using STATA software (Release 10.0 for Windows, StataCorp LP).

Preoperative assessment

All patients were studied preoperatively by means of a two-dimensional (2D) echocardiography and Doppler, in order to evaluate TV and RV dimensions and RV function. TV assessment was made by obtaining the larger annulus diameter measured from a standard 2D – Echocardiography apical four chamber view, this was indexed for body surface area and then compared to normal values. Cardiac catheterisation was also performed in all, before repair, to assess cardiac pressures and calculate pulmonary vascular resistances (PVR). The preoperative cardiac rhythm was established for all patients by EKG; other clinical data gathered at admission included: venous blood oxygen saturation (SatO2), presence and degree of cyanosis, medical therapy.

2. Surgical technique

One and a half ventricle repair was performed by the same surgeon in all patients. Surgical repair was carried out through a midline stenortomy (redo in two patients), superior and inferior vena cava (SVC and IVC) were cannulated directly (SVC next to the origin of the left innominate vein). Mild hypotermic (rectal temperature between 28 and 34 ° C) cardio-pulmonary bypass was instituted in all. Aortic cross-clamping and multiple doses of haematic cardioplegia was employed in all.TV annulus diameters were sized with Hegar's dilators confirming the preoperative echocardiographic values. The three patients with Ebstein's Anomaly showed a various degree of downward dislocation of posterior and septal leaflets and tethering of the anterior leaflet to the RV free wall; all had serious limited functional RV, due to a large atrialized portion. Two of them required TV surgery: one TV repair (Vargas procedure20) and one TV replacement (with bioprosthetic valve Carpentier-Edwards no. 25), this later patient underwent additional annuloplasty of incompetent mitral valve (performed through the ASD II). The remaining two patients presented TV and RV hypoplasia at inspection, one of them required right ventricle – pulmonary artery continuity reconstruction with conduit Contegra no. 16 for pulmonary valve stenosis. Additional procedure were: PFO/ASD II closure, n=4; radiofrequency ablation (RFA) of the IVC sinus, n=1; RFA – Mazè procedure with atrioplasty n=1, this latter patient also had definitive pace-maker implantation.

All patients received end-to-side anastomosis between SVC and right pulmonary artery brunch (BCPS) at beating heart; the azygos vein was ligated in two patients.

3. Postoperative assessment and follow-up

Any early postoperative adverse events were registered and all patients received a 2D-Echocardiography qualitative evaluation of right and left ventricle contractility; we also evaluated drugs administrated and cardiac rhythm at discharge. There were no operative death. Mean ICU stay 6 ± 7 d, median 3 d (range1-29 days). Mean hospital stay : 32± 29 days, media 23 (6-113 days). Among in hospital complications 18 patients had different types of complications like: Pleural effusions in 8 patients, Chylothorx/pericardium in 7, different arrhythmias in 7 patients, acute renal failure 4, Postoperative low cardiac output in 4, Pericardial effusion in 3, Superior vena cava syndrome in 2, complete A-V block in 1,Hemidiafragm paralisys in 1 and pulmonary infection was diagnosed in 1 patient.

Follow-up

Follow up was completed and updated in all, with a mean of 5.7 years (range 0.2 12 years). All the patients were followed with clinical evaluations and 2D-ecocardiography and Doppler. Exercise stress test at cycloergometry was performed in one patient with Ebstein's Anomaly.

Follow up time range 6 months-18 years; mean follow up time is 8.3 years.

Follow up 96% completed 23/24 pts, 1 foreigner patients lost at follow up. At early follow up 7 patients had different 11 complications as shown in table 4.

Table: 4. Early complications

Type of complications	Group A (Simple)	Group B (Complex)
Chylothorax	2	4
LCOs	1	1
Permanent A-V Block		2
Transient A-V Block	1	0
TOTAL	4 complications in 3 pts	7 complications in 4 pts
	3/12 (25%)	4/10 (40%)

Among survivors(22) 19 patients are in NYHA class I. Eleven patients among24(46%) had late complications. Late complications shown in table 5.

Table: 5. Late complications

Type of complications	Simple Anatomy	Complex Anatomy
PLE	0	1
LVOTO + CHF	0	1
Left pulmonary vein occlusive disease	0	1
CVA no cardiac related	1 (dead adult pt after 2 yr from op)	0
Pulmonary hypertension	0	1 (dead after reop.)
TOTAL	1 complication in 1 pt	4 complications in 3 pts
	1/11 (9 %)	3/10 (30%)

During follow up 2 patients died: one patients died during redo surgery; aortic valve replace ment+ RV-PA conduit and left pulmonary artery plasty and immidietely after surgery patients assisted by ECMO and died after few hours of surgery. Another late death non cardiac related, patient with Ebstain's anomaly died due to stroke 2 years after 1.5 ventricle repair.

Three patients had late reoperation:

1. RVOT revision + 2-ventricle-conversion"
2. Subaortic fibrous tissue resection + re-do tunnel IVC to left sided atrium+ Contegra conduit change + RPA enlargement.
3. AVR + RVPA conduit +LPA plasty, ECMO

At late follow up 6 patients had 15 different interventional procedures, shown in table 6.

Table: 6. Transcatheter procedures late after 1.5 ventricle repair

Type of procedure	No.
LPA dilation/stenting	4
MPA dilatation	3
IVC-LA baffle balloon dilatation/ stent	3
RVOT diltation attempts	2
Stent SVC	1
device ASD closure	1
Azygous vein plugging	1

b. ANIMAL STUDY

MATERIAL AND METHODS

This experiment was performed in experimental Rabbits. Animal care has taken according to established standard for experiment in animals.

Experimental model: RABBIT

Strain: NEW ZEALAND

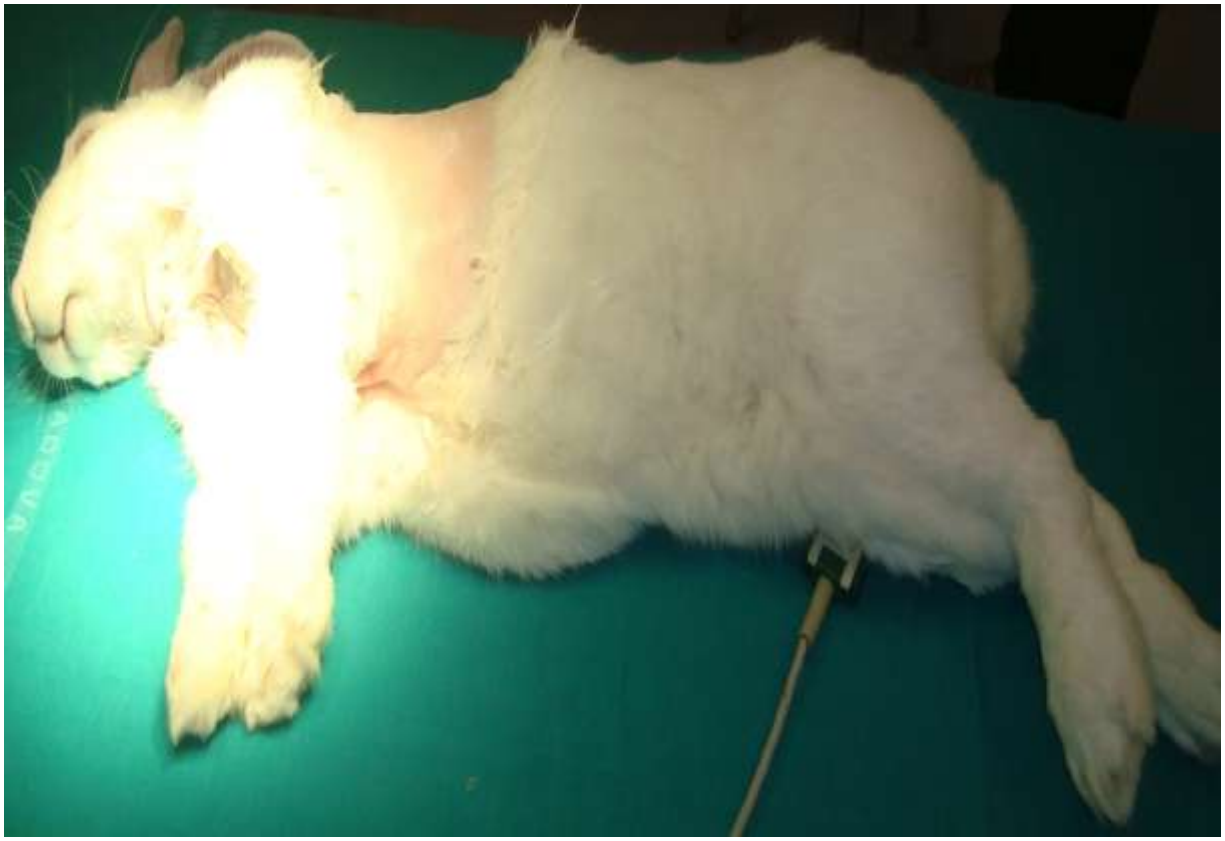


Fig.21. animal prepared for intubation

Total animal number: 30

Weigh of animals: 4.5-5.5 kg

Experimental time period: 24 hours

GROUPS:

Animals were divided in three groups, hemodynamic measurements were taken from all three groups at resting condition and after induced pharmacological stress test with intravenous adrenaline bolus infusion of 0.02mcg/kg/min

1) normal controls 10

Resting condition + after induced pharmacological stress

2) Right BCPS (1.5 v repair) 10

Resting condition + after induced pharmacological stress

3) Right BCPS + LPA closure (CPA alone) 10

Resting condition + after induced pharmacological stress

SVC: superior vena cava, BCPS: bidirectional cavopulmonary shunt, RPA: right pulmonary artery, LPA: left pulmonary artery, 1.5 v repair: one and a half ventricle repair, CPA: cavopulmonary anastomosis.

1. ANESTHESIA

Type: general

Anesthetic Agents [82] Induction: Ketamine 4mg/kg intravenous

Mainteinance: Halothane 2 litre/min, inhalation

Myorlaxant: Pancuromium 0.25mcgr/kg/min, intravenous

2.Vital sign measurements

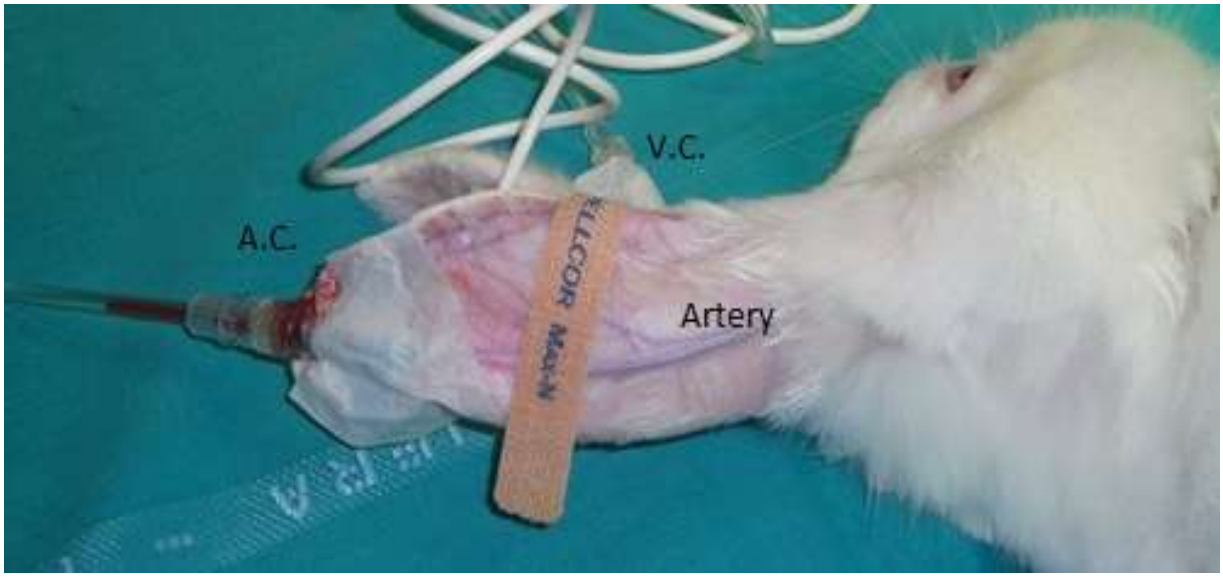


Fig:22. Animal after having the arterial and venous accesses. A.C.- arterial cannula, V.C. – venous cannula.

Fig: 23. Vital sign monitoring

Vital signs were measures as in standard operating room for animal. To administer intravenously fluids and drugs right vein in the right auricle (ear) was cannulated. To measure

the blood pressure during and after performing the procedure artery in the left auricle was cnnulated by 16 fr. IV cannula. To have the intravenous or arterial access in other part of the body found to be difficult. Saturation rate, heart rate, ECG monitoring also was achieved with the use of standard operating room monitor of SIEMENS SC 9000 as shown in fig. 23.

3. **Intubation**: endotracheal

Fig:24. Endo-tracheal intubation of the rabbit through tracheostomy incision

Intubation has performed with direct tracheostomy incision as shown in fig.24. We have used pediatric endotracheal tube- uncuffed tubes of 2.5-3.5 Fr. [82, 83] Uncuffed tube because, to allow as large an internal tube diameter as possible. After intubation, endotracheal tube has fixed with open-weave bandage tied around the connector and then around the back of the rabbit's head then connected with ventilator.

4.Ventilation: mechanical

Fig.25. animal connected to ventilator

Ventilator settings were as follows: [84]

PEEP- 2-3 mm of water.

Tidal volume- 7-10 ml/kg

Respiratory rate- 20-30/min.

We used OLMEDA Excel 210 SE mechanical ventilator, rabbit was connected to the ventilator through endotracheal tube and assisted by Halothane gas with rate of 2litre/min during and after completion of the procedure.

5. SURGICAL TECHNIQUE

Fig: 26. Full animal preparation before surgery

Animal was prepared according to the standard techniques for animals [85-90] and animal care was taken as recommended by the ethical committee. Two technicians, one operating nurse and three surgeons were involved in this procedure. Operation was performed through right lateral thoracotomy in general anesthesia. Duration of the procedure was around 2-3 hours for each rabbit. Rabbit was fully heparinized before making the anastomosis with the dose of 100 IU/kg of heparin. Cavapulmonary anastomosis was done without cardiopulmonary bypass. Anastomosis was made in end to side fashion between superior vena cava and the right pulmonary artery as shown in fig.30. To make this anastomosis two curve Shatinsky clamps were used as showen in fig.28. Monofilament polypropylene (Proline) 6.O suture were used to make the cavapulmonary anastomosis. After completing the anastomosis between superior vena cava and right pulmonary artery blood pressure were measured from superior vena cava, right

pulmonary artery, right atrium and right ventricle. Pressures were measured at resting condition and after pharmacologically induced stress test. Pharmacological stress test was performed by giving intravenously adrenaline bolus 0.02mcg/kg. Rabbits were continuously monitored during and after surgery with standard monitoring techniques as shown in figure 23. After completion of the all measurements animal was sacrificed within 24 hours from surgery as recommended by ethical committee.

Surgical access: right thoracotomy

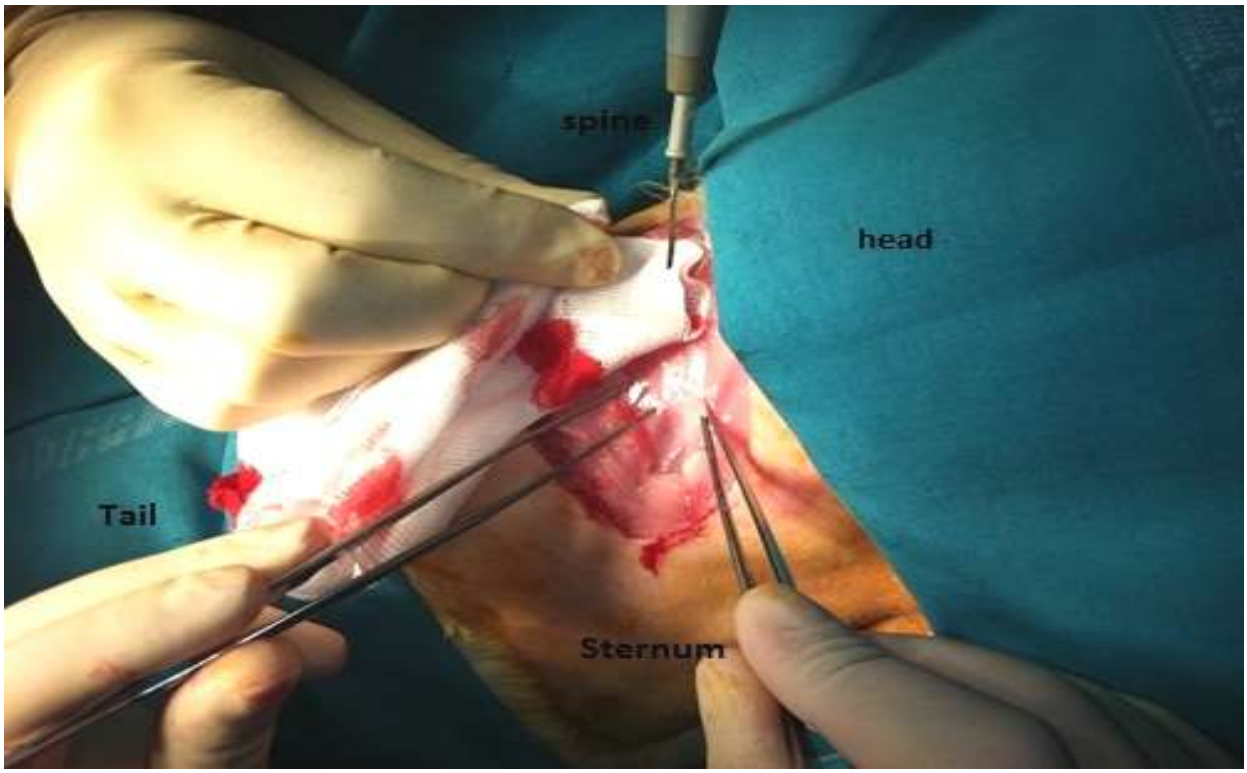


Fig. 27. Right thoracotomy incision in a rabbit

No cardiopulmonary bypass

Partial heparinization: 100 Units/kilograms

Fig:28.A curve Satinsky clamp used during surgery

Fig. 29. During surgical intervention in a rabbit (OR view)

We have divided the experimental animals in four groups for hemodynamic measurments

Group 1: NORMAL CONTROLS

a) pulsatile pulmonary flow

b) no hypertension in the SVC district

c) no hypertension in the IVC district

This group is for normal control and no surgery has been done.

Group 2: after cavopolmonary anastomosis (CVPA)

a) pulsatile pulmonary flow

b) hypertension in the SVC district

c) no hypertension in the IVC district

this group have normal pulsatile pulmonary flow from right ventricle. It has created hypertension in SVC district and found normal pressure as before surgery in IVC district. This group for evaluation of the cerebral metabolic changes due to hypertension in SVC district and evaluation of the blood flow distribution to the lungs.

Group 3: after CVPA with stress test

a) pulsatile pulmonary flow under stress

b) hypertension in the SVC district (BCPS + pulsatile flow)

c) found no Hypertension in the IVC district

in this group excersice test was performed by infusing a bolus of adrenaline 0.08mcg/kg.

In this group we have evaluated mixed laminar and pulsatile blood flow distribution to the lungs and evaluated the indirect signs of the right ventricular failure by measuring the pressure changes in right ventricle and right atrium immediately after giving the adrenaline infusion.

Group 4: after CVPA and clamping of the left pulmonary artery

a) pulsatile pulmonary flow only on LPA

b) no anterograde flow on RPA

b) hypertension in the SVC district

c) found no hypertension in the IVC district

In this group as we close the RPA there won't be any pulsatile flow to right lung but there is anterograde flow to the LPA. There is no hypertension in SVC district because there is no any reverse flow from RPA and found no hypertension in IVC district too.

6. INTRAOPERATIVE EVALUATION

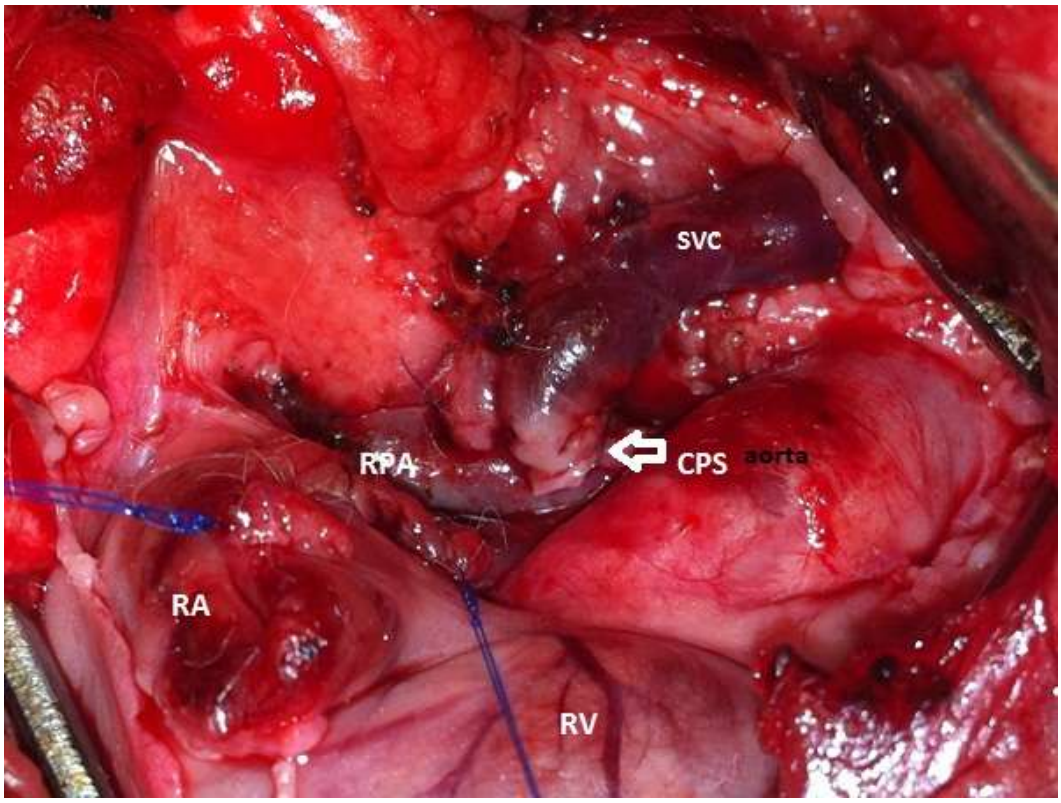


Fig: 30. Intraoperative view of the cavapulmonary anastomosis in a rabbit(1.5 VR); SVC: superior vena cava, RPA: right pulmonary artery, CPS: cavapulmonary shunt(anastomosis between SVC and RPA), RA: right atrium, RV: right ventricle.

Hemodynamical measurements: pressure sampling

(RSVC, RA, RV, MPA, RPA, LPA)

RSVC: right superior vena cava

RA: right atrium

RV: right ventricle

MPA: main pulmonary artery

LPA: left pulmonary artery

Pressure measurement has done by direct puncher to the RSVC, RA, RV, MPA, RPA, LPA.

POSTOPERATIVE TIME (before sacrifice): 24 hours

7. POSTOPERATIVE EVALUATION (before sacrifice)

In post-operative period we have evaluated the effects of venous hypertension on the superior vena cava districts (brain) by evaluating the oxygen saturation.

Likewise, we have analyzed the competition of flows at the superior vena cava, right pulmonary artery junction: to understand the hemodynamic interaction of a pulsatile flow in combination to a laminar one in postoperative period by direct puncher.

We have observed the venous pressure in RA and RV to evaluate the advantages of the 1.5 ventricle repair on maintaining the low pressure in the inferior vena cava district by direct puncher on right atrium and right ventricle.

c. RESULTS

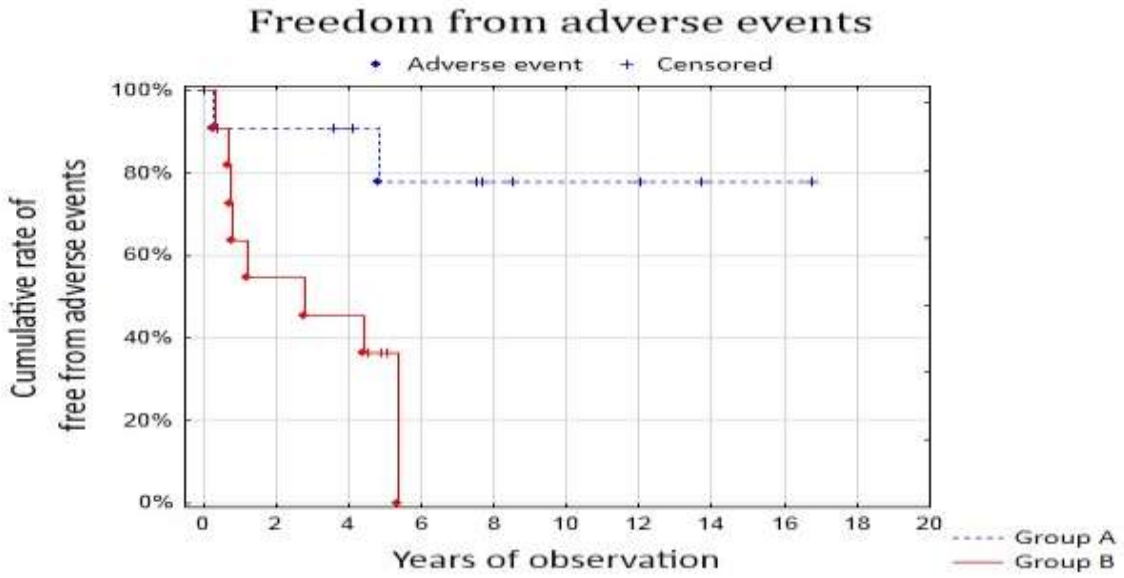
1. Results of clinical study

This is an 18 years retrospective clinical review of 24 consecutive patients who underwent one and a half ventricle repair at our Institution. Mean age at repair was 11.9 years (range 4 months – 66.8 years). Mean TV annulus Z-score was -3.2 (range -6.2 to 3.6). Mean pulmonary vascular resistance was 1.75 U/m2 (range 1.0 to 3.0). Three patients had heterotaxy syndrome. Sixteen patients underwent previous cardiac operation, one of these had a Fontan operation.

There was no death at operation. Postoperative complications occurred in 19 patients (79 %), and were: chylothorax/chylopericardio (n=7), BAV III (n=2), arrhythmias (n=7), renal failure (n=4), heart failure (n=4), cardiac/pleural effusion (n=11), SVC syndrome (n=2), pulmonary infection (n=3), hemidiaphragm paralysis that required diaphragm plasty (n=1), SVC thrombosis with PE (n=1). Five patients required definitive PM implantation. All patients were discharged home alive and well, after a mean hospital stay of 32 ± 29 days. At a mean follow up of 8.3 years (range 1 month – 17.9 years, FU completeness: 96 %), there were 2 late deaths (1 non cardiac related). Among survivors, functional status was NYHA class I in 19 patients (90.4%). Late adverse events occurred in 10 patients (43 %) including: late reoperation (n=3), one of these was a biventricular conversion; haemodynamic procedures (n=6); arrhythmias (n=2); neurological event (n=1); other complications (n=5).

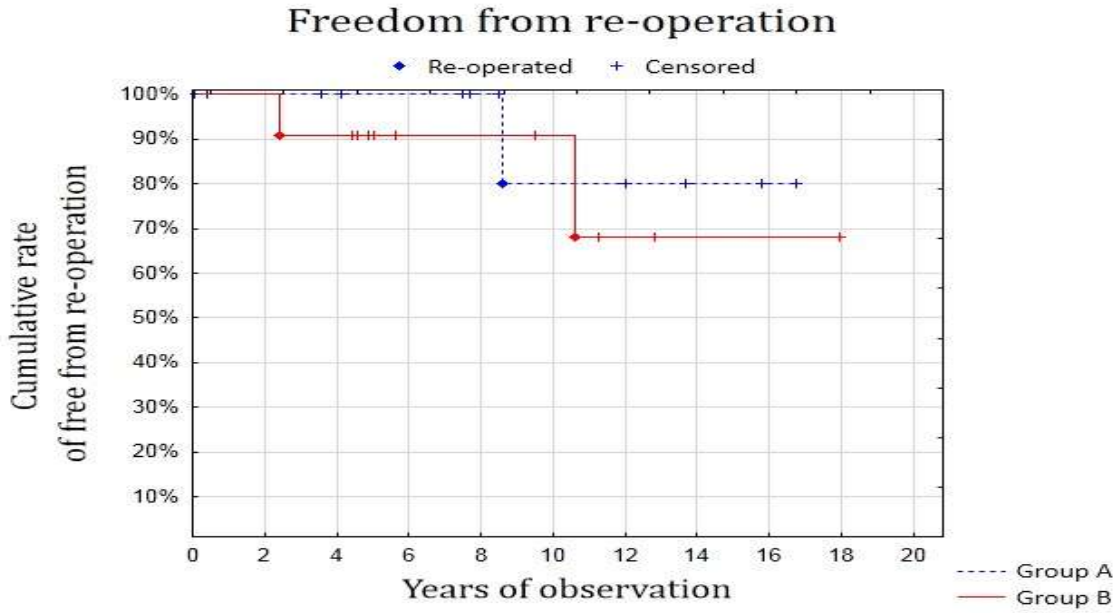
Overall freedom from adverse events, surgery and interventional procedures was 56.5 %, 87 % and 73,9 % respectively. Freedom from adverse event in Group A (n=12) was 83.3 % while in Group B (n=11) was 27.3 %. Statistical analysis demonstrated that Group B had a significantly lower freedom from adverse events than Group A ($p = 0.015$). statistical analysis of the results were shown in figure 31-38.

Fig:31. Freedom from adverse events



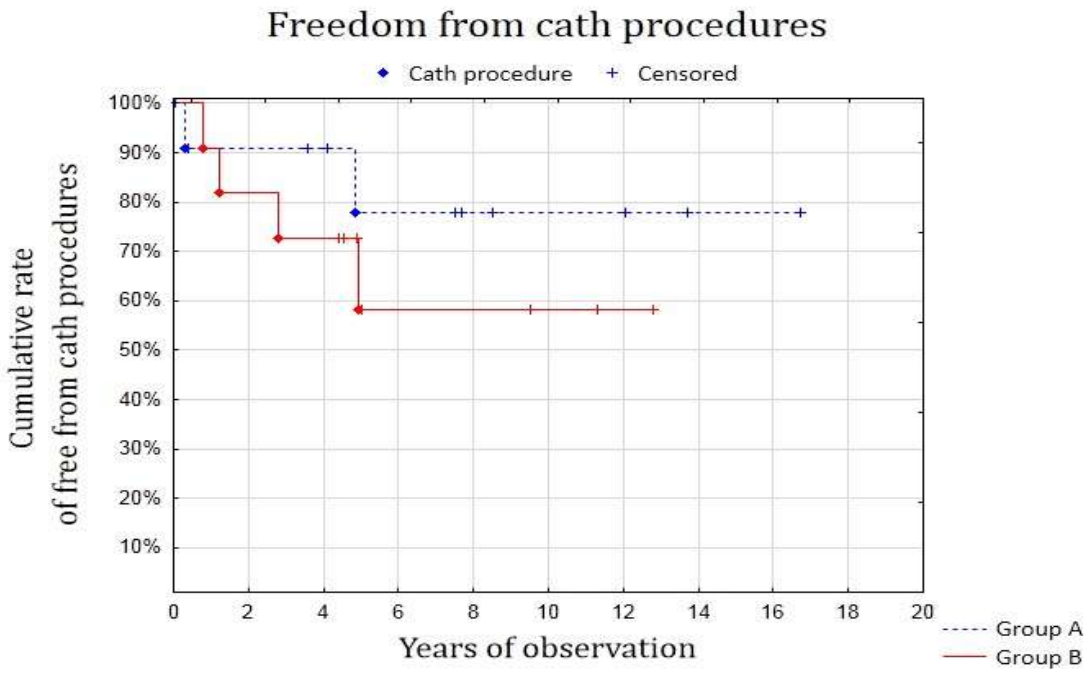
p = 0.015

Fig:32. Freedom from re-operation



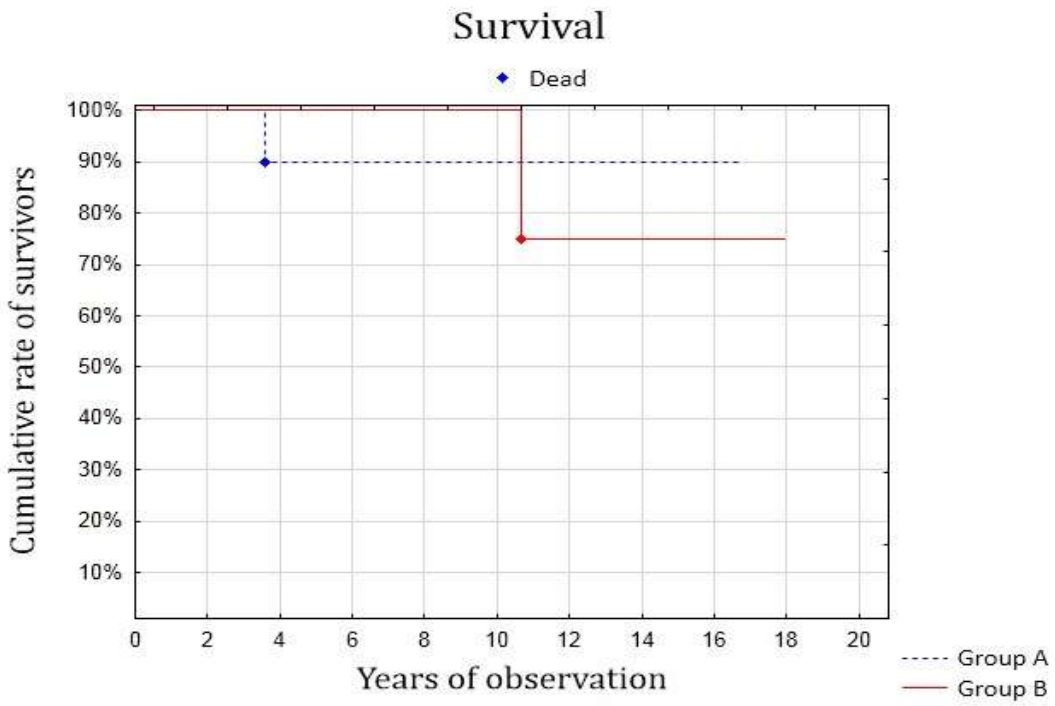
p = 0.57

Fig:33. Freedom from cath. procedures



p = 0.40

Fig:34. Survival rate



p = 0.96

Fig:35. Overall survival

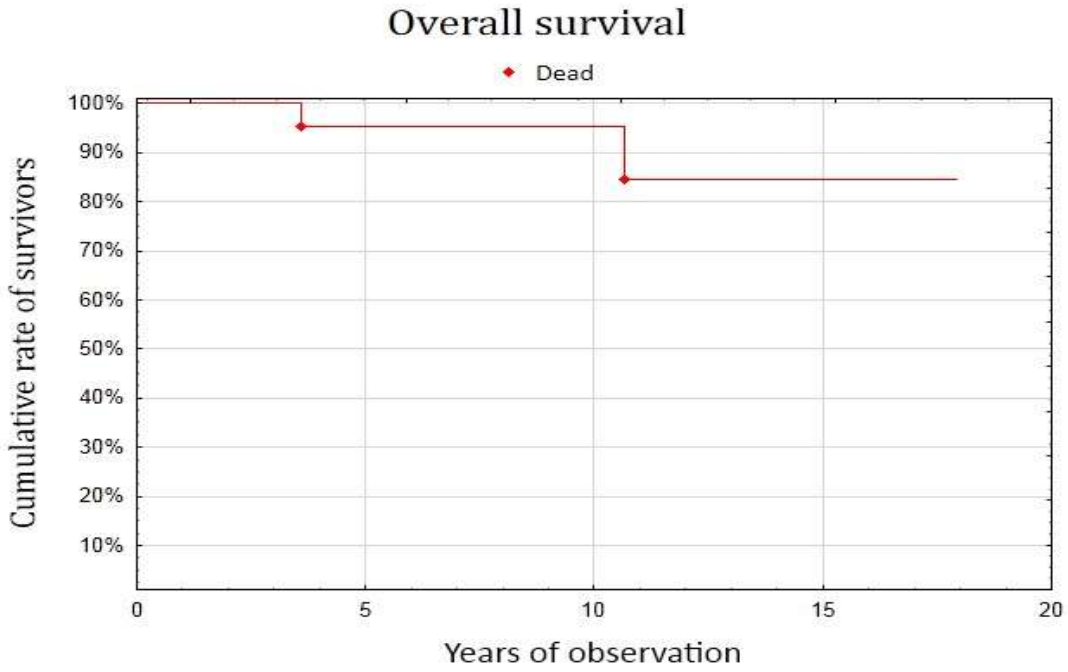


Fig:36. Overall freedom from adverse events

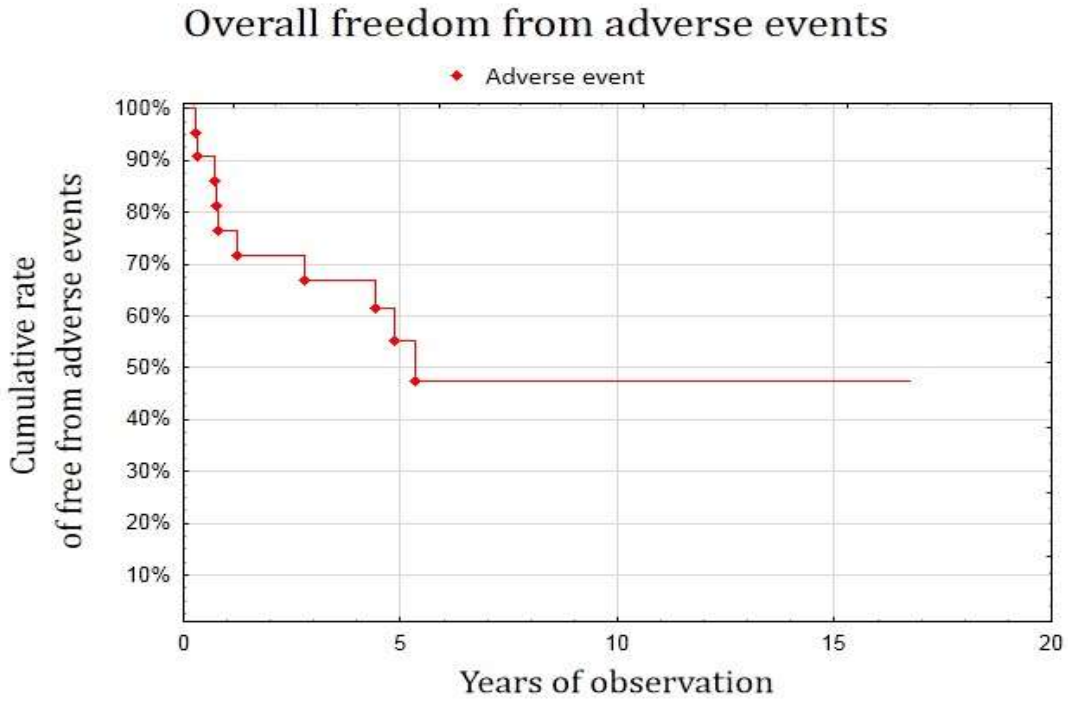


Fig: 37. Overall freedom from re-operation

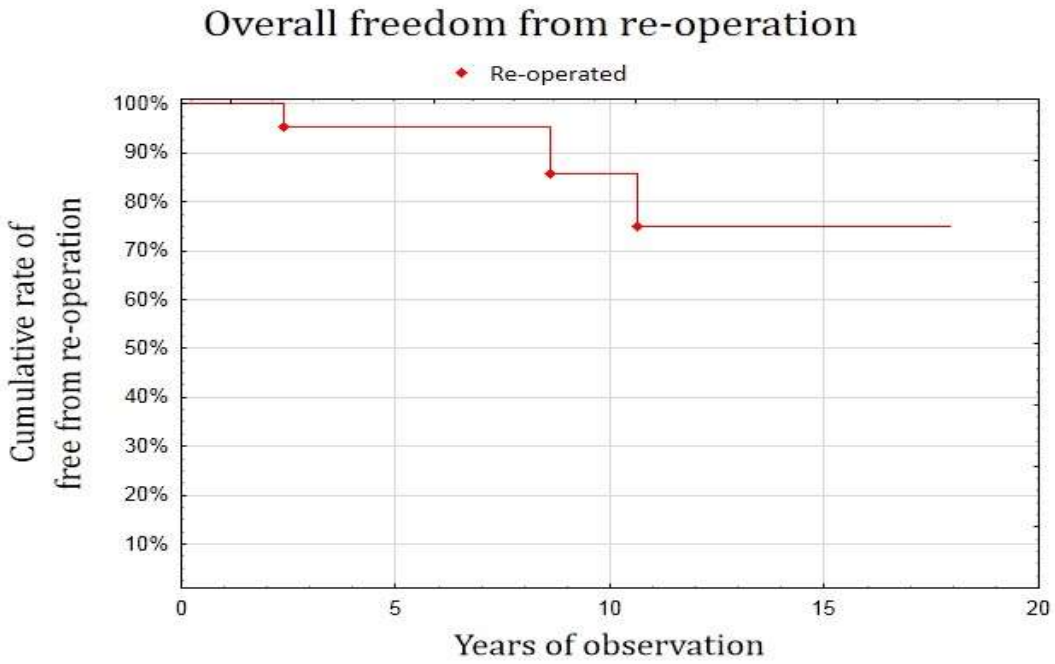
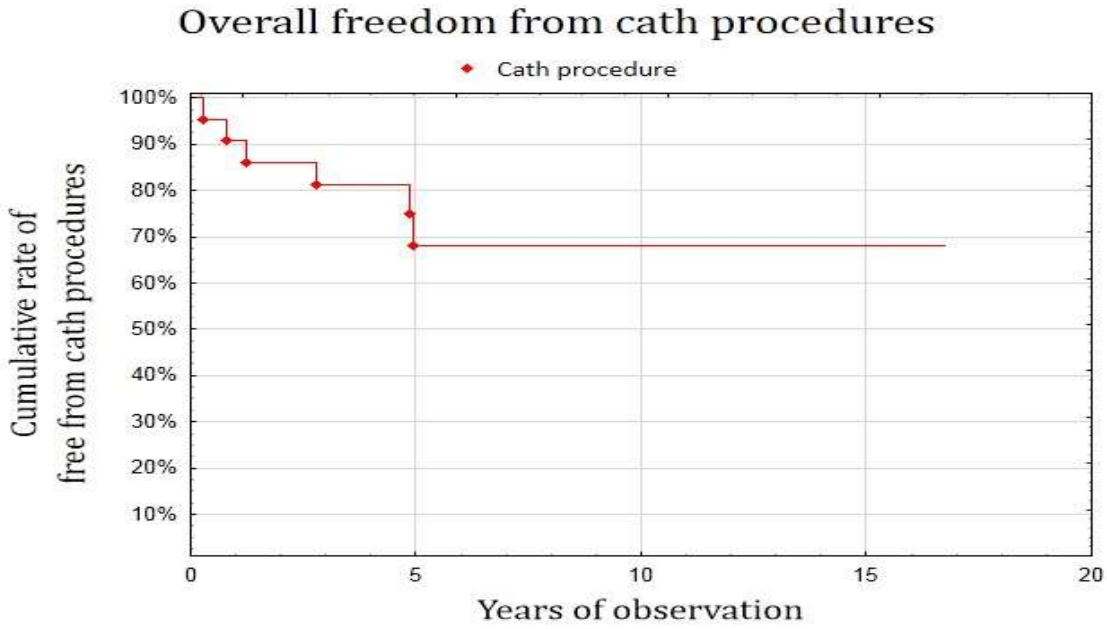


Fig:38. Overall freedom from cath procedure



2. Results of animal Study

This is an animal model study on one and a half ventricle repair which was performed in rabbits. We performed an anastomosis between superior vena cava and right pulmonary artery as a model of one and a half ventricle repair. After performing this cavapulmonary anastomosis we measured the pressures from different cardiac chambers, SVC and pulmonary arteries during resting condition and after induced pharmacological stress test. Likewise, we have created the model of cavapulmonary shunt alone by clamping the right pulmonary artery just proximal to the cavapulmonary anastomosis to allow the blood flow coming from the inferior vena cava only to the left pulmonary artery through right ventricle. Results are shown in table 7-12 and figure 41-45.

Table:7. (A). Control group: (n. 10)

Resting condition

No.	BP	HR	RA	RV	SVC	PA
1	72/40	90	7	14/7	6	14/7
2	90/50	102	8	18/5	5	17/5
3	83/40	86	6	16/4	4	16/4
4	80/50	92	8	18/6	6	17/6
5	94/56	76	7	15/5	6	15/7
6	70/45	106	8	14/4	7	14/4
7	85/42	88	9	13/5	6	13/5
8	90/50	101	6	15/5	4	15/5
9	78/62	105	7	17/6	5	17/7
10	86/65	94	8	20/4	6	20/4

Table:8. Control group

After induced pharmacological stress test

N0	BP	HR	RA	RV	SVC	PA
1	110/90	100	8	15/6	5	15/6
2	115/80	116	9	16/4	6	16/5
3	100/76	100	7	14/3	4	15/4
4	100/80	110	9	15/2	7	15/3
5	120/80	105	8	10/2	6	11/2
6	116/92	120	9	12/3	8	12/4
7	120/80	108	10	14/6	6	14/6
8	110/70	110	7	13/6	5	13/6
9	106/92	116	8	14/4	6	15/4
10	112/96	118	9	14/3	6	15/4

- Median SVC pressure: 1) at rest = 6 mmHg (R 4-7) , 2) after stess: 7 mmHg (R 4-8) > 10%
- Median RA pressure: 1) at rest = 7 mmHg (R 6-9) , 2) after stress: 8 mmHg (R 7-10) >11.1%

Fig: 39. (A) Group: Bidirectional cavapulmonary shunt (BCPS) + pulsatile RV flow (1.5 V model) (n=10)

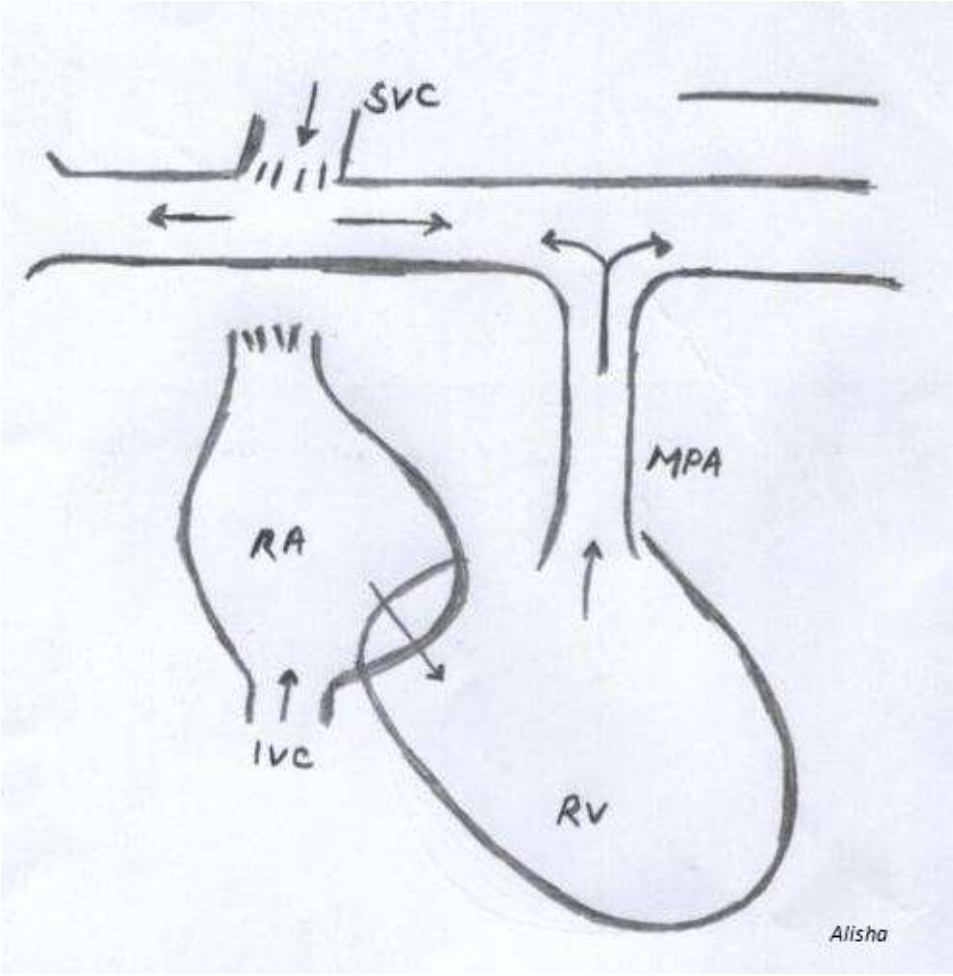


Table:9. One and a half ventricle repair model (n=10)

Resting condition

No.	BP	HR	RA	RV	SVC	PA
1	72/40	90	7	14/7	20	15/7
2	90/50	102	8	18/5	18	18/4
3	83/40	86	5	16/4	22	17/5
4	80/50	92	7	18/6	18	19/6
5	94/56	76	7	15/5	25	15/4
6	70/45	106	6	14/4	20	15/4
7	85/42	88	7	13/5	18	13/4
8	90/50	101	6	15/5	21	15/4
9	78/62	105	7	17/6	20	18/6
10	86/65	94	6	20/4	22	20/5

Table:10. One and a half ventricle repair model (n=10)

After induced pharmacological stress test

N0	BP	HR	RA	RV	SVC	PA
1	110/90	100	7	15/6	32	16/6
2	115/80	116	8	16/4	36	16/4
3	100/76	100	6	14/3	30	14/4
4	100/80	110	8	15/2	32	15/2
5	120/80	105	8	10/2	33	10/2
6	116/92	120	5	12/3	34	12/4
7	120/80	108	9	14/6	28	14/6
8	110/70	110	5	13/6	32	14/6
9	106/92	116	8	14/4	30	14/4
10	112/96	118	6	14/3	30	15/4

- Median SVC pressure: 1) at rest = 20 mmHg (R 18-25) 2) after stess : 32 mmHg (R 28-36) >40%
- Median RA pressure: 1) at rest = 7 mmHg (R 5-8) 2) after stess: 8 mmHg (R 5-9) > 10%

Fig: 40. (C) Group: cavopulmonary shunt alone (no.10)

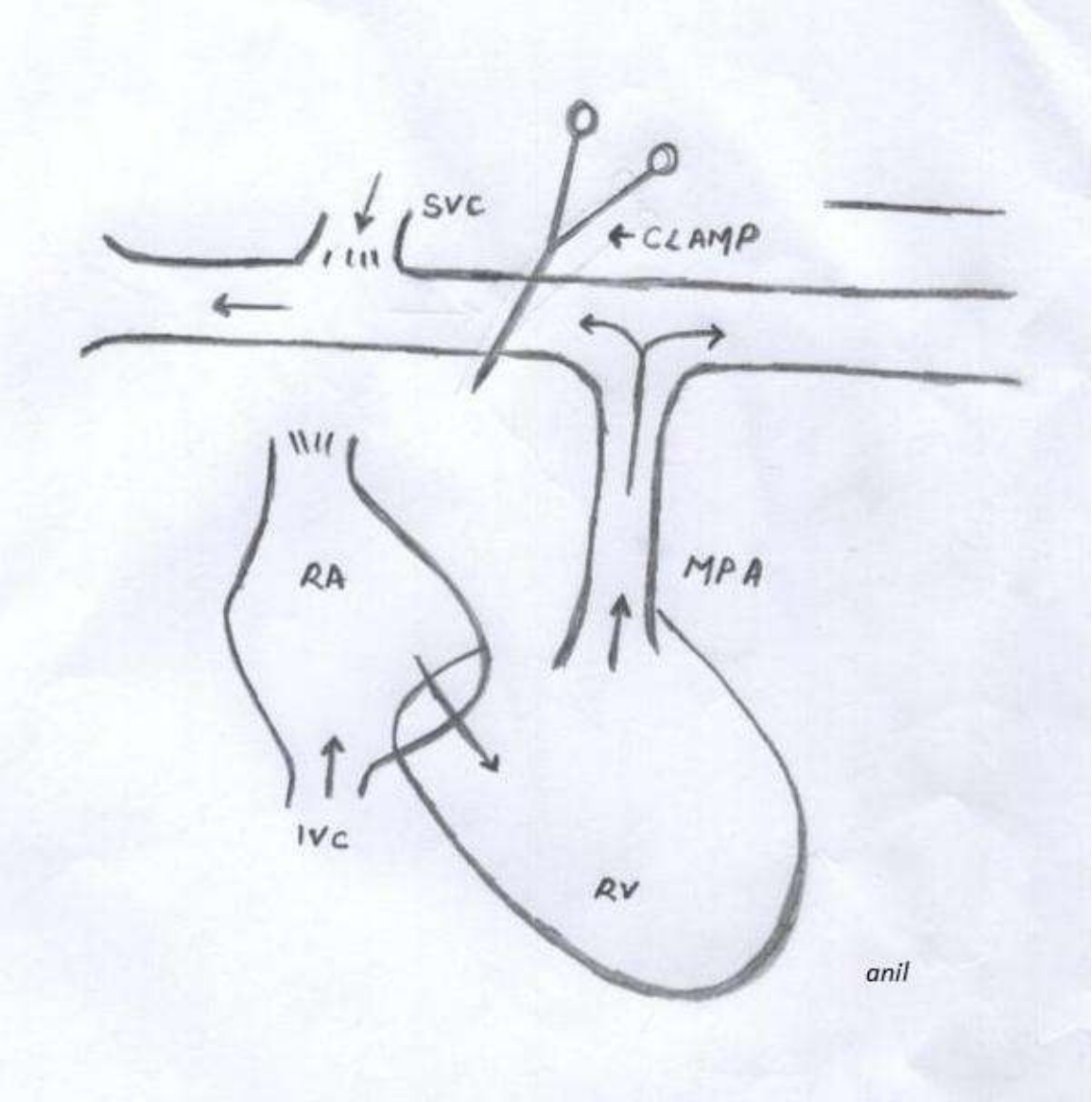


Table:11. cavopulmonary shunt alone (no.10)

Resting condition

No.	BP	HR	RA	RV	SVC	PA
1	72/40	90	7	18/7	8	18/8
2	90/50	102	8	18/5	10	18/5
3	83/40	86	7	20/5	12	19/5
4	80/50	92	8	18/6	14	17/6
5	94/56	76	7	15/5	12	15/5
6	70/45	106	8	18/4	10	18/4
7	85/42	88	9	17/5	12	18/5
8	90/50	101	7	18/5	13	18/5
9	78/62	105	8	17/6	14	18/6
10	86/65	94	9	20/4	11	20/5

Table:12. cavopulmonary shunt alone (no.10)

After induced pharmacological stress test

No	BP	HR	RA	RV	SVC	PA
1	110/90	100	7	15/6	12	20/6
2	115/80	116	9	16/4	14	16/4
3	100/76	100	8	14/3	16	14/3
4	100/80	110	10	15/2	17	15/3
5	120/80	105	9	18/4	14	18/3
6	116/92	120	11	20/4	12	20/4
7	120/80	108	10	18/5	15	18/5
8	110/70	110	8	20/5	16	20/5
9	106/92	116	9	18/6	17	18/6
10	112/96	118	10	22/4	15	23/4

- Median SVC pressure: 1) at rest = 12 mmHg (R 8-14) , 2) after stess test: 15 mmHg (R 12-17) >20%
- Median RA pressure: 1) at rest = 8 mmHg (R 7-9) , 2) after stess test: 9 mmHg (R 7-11) > 11.1%

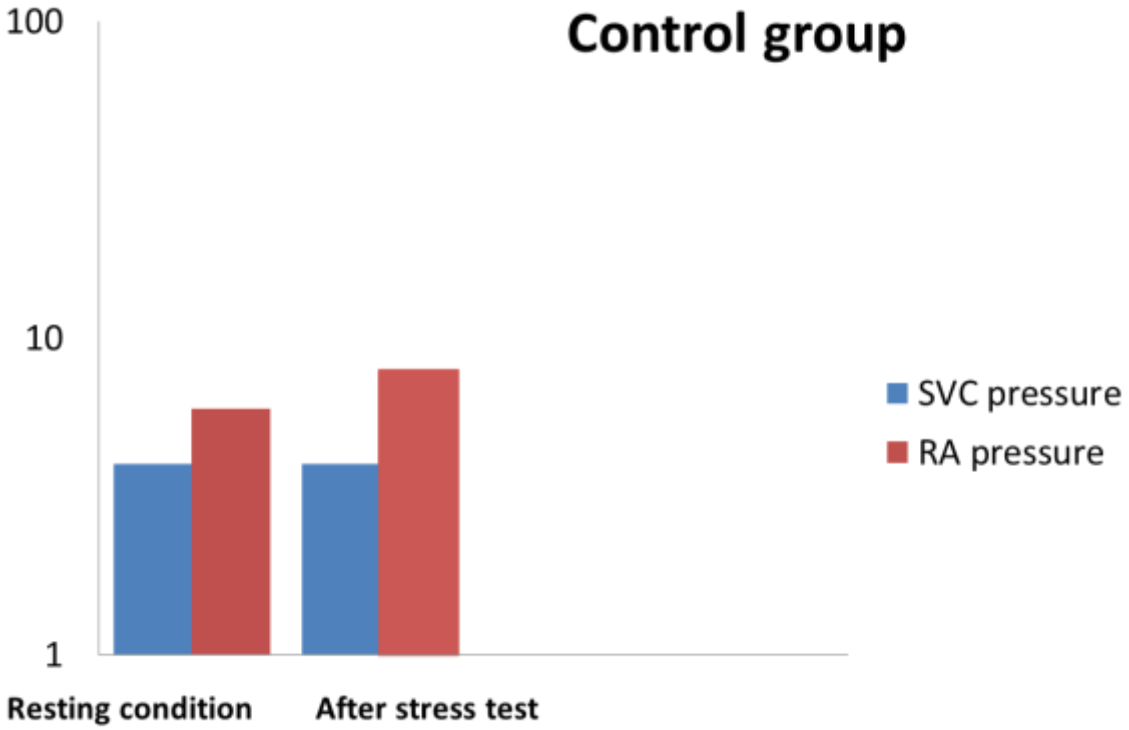


Fig:41. Pressure changes in control group; no any significant changes of blood pressure in SVC and RA after pharmacologically induced stress test

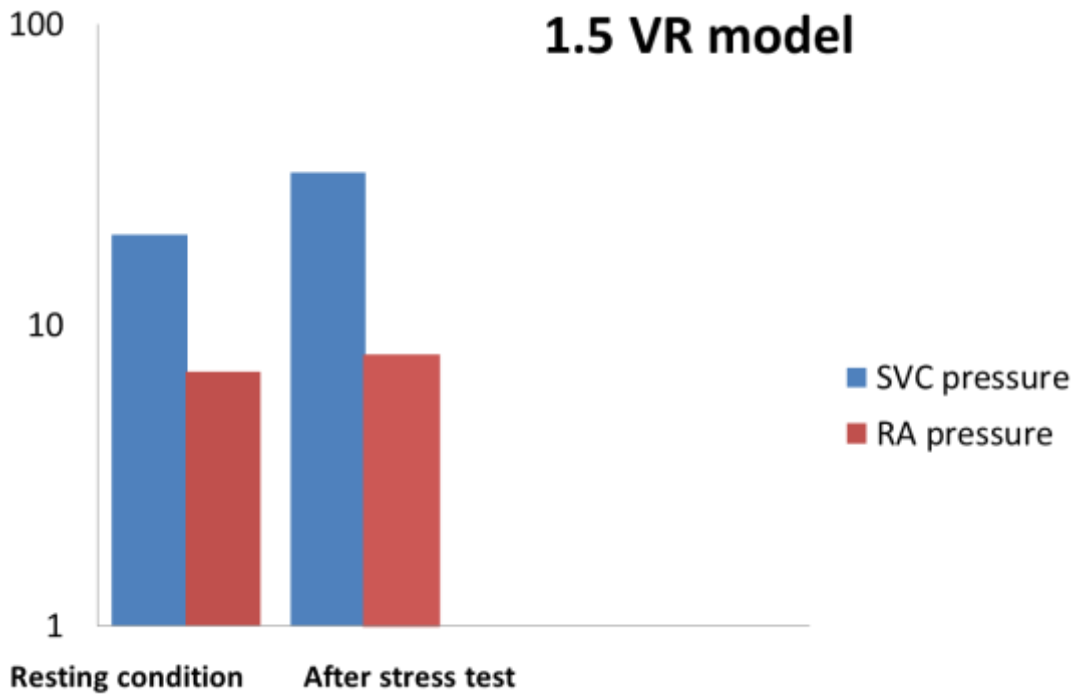


Fig:42. Pressure changes in 1.5 VR group; there is no any significant changes of RA pressure after 1.5 ventricle repair at resting condition and after pharmacologically induced stress test. But after 1.5 VR we found elevated SVC pressure which increased significantly after pharmacologically induced stress test.

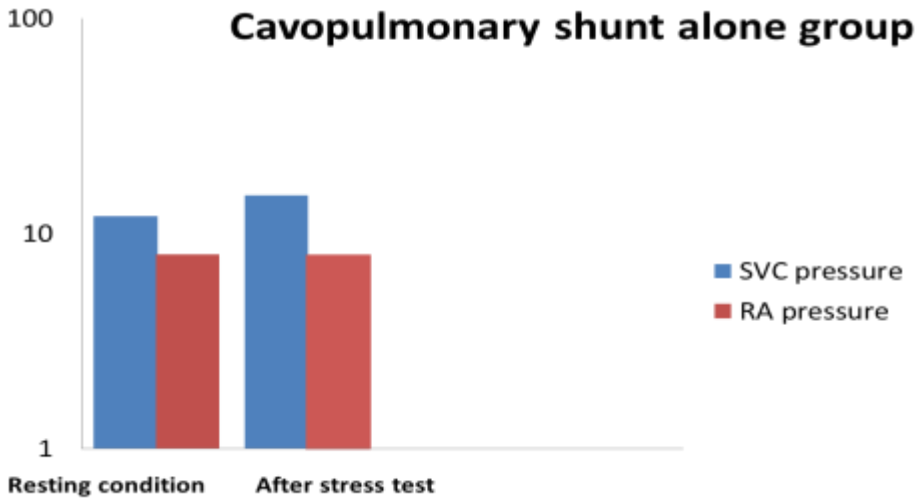


Fig:43. In group of Cavopulmonary shunt alone, found no any significant pressure changes in SVC and RA at resting condition and after pharmacologically induced stress test in comparison with 1.5 VR group.

3. SUMMERY OF ANIMAL STUDY

1. After cavapulmonary anastomosis- SVC and PA pressure has increased but RA pressure has decreased, which proves the decrease of the pressure in inferior vena cava district.

2. After pharmacologically induced stress test: increased the pressure in superior vena cava and also increased heart rate but right ventricular, right atrial & pulmonary artery pressures were remained as same as after cavopolmonary anastomosis, as it is in resting condition.

3. That means superior vena cava pressure is always higher after cavapulmonary anastomosis but venous pressure in inferior vena cava district is constant and even lower in comparison with preoperative pressure.

4. We found no any changes of right atrial pressure, after 1.5 ventricular repair and even after pharmacologically induced stress test which proves that the inferior vena cava return is ok and no hypertension in inferior vena cava district even after stress test(good exercise tolerance), which is a very positive finding in favor of one and a half ventricle repair in comparison to Fontan type procedures.

4. Hemodynamic changes in superior vena cava

1. After bidirectional cavapulmonary anastomosis increased the pressure in superior vena cava in 1.5 ventricle repair group in comparison with group with cavapulmonary shunt alone (> SVC pressure in1.5 VR > CPS alone).

2. At the resting conditions pressure in superior vena cava is higher in 1.5 ventricle repair group then in cavapulmonary shunt group and in control group. (> SVC pressure in 1.5 VR> CPS alone > controls)

3. After stress test in 1.5 VR group, pressure in superior vena cava increased two times more in comparison with cavopilmonary shunt group and control group: (> SVC pressure (1.5 VR >> CPS alone > controls)).

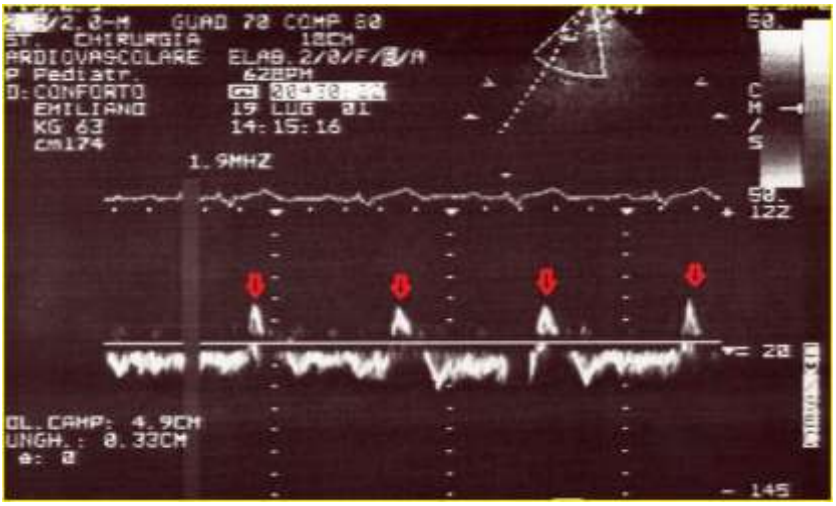


Fig: 44. Doppler from superior vena cava After 1.5 VR (arrows: reverse systolic flow pattern)

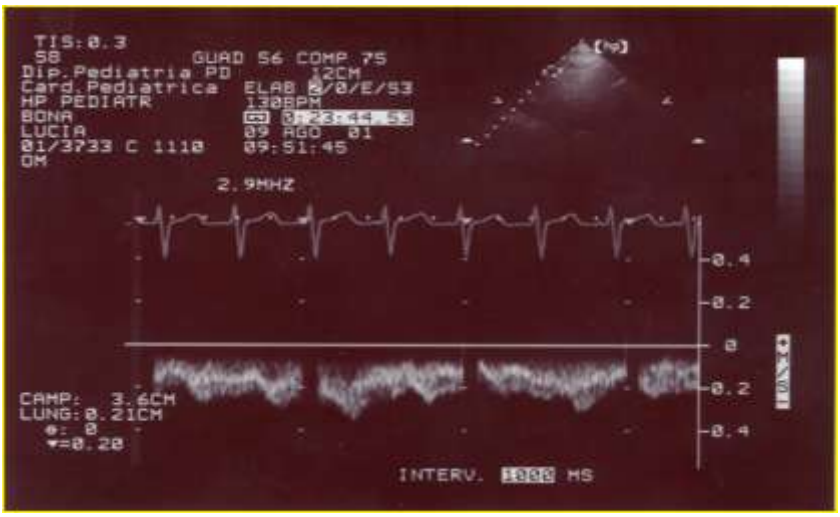


Fig:45. Doppler from superior vena cava after Fontan operation

5.Right atrial pressure (which reflects the pressure in IVC)

1. After cavopulmonary anastomosis right atrial pressure has found to be equal in all three groups: RA pressure (1.5 VR = CPS alone = controls)

2. At resting conditions also right atrial pressure has found to be equal in all three groups: RA pressure (1.5 VR = CPS alone = controls)

3. After stress test also we found the same condition, in all three groups right atrial pressure was found to be equal: RA pressure (1.5 VR = CPS alone = controls)

SECTION VI

STUDY LIMITATION

Study limitation

1. Animal model

2. Normal size RV

3. No long term follow up (clinical+animal)

4. Desirable to have histopathological exams of the cerebral & hepatic tissues to understand long term effect of the 1.5 ventricle repair at tissue level.

SECTION VII

CONCLUSION

CONCLUSION

1. We found significant increase of the SVC pressure after 1.5 ventricle repair & after the pharmacologically induced stress test the pressure in superior vena cava has increased two times more than in resting condition. However, our experience of more than three decades in patients with Fontan type procedure, elevated SVC pressure is well tolerated. Similarly, Brain MRI of all our patients who underwent brain MRI with 1.5 ventricle repair showed good results at midterm fellow up.
2. The 1.5 V repair is a valid option for surgical repair of hypoplastic, failing or borderline right ventricle.
3. This repair provides a low pressure in the inferior vena cava district and allows good early and mid-term outcomes
4. Potential for right ventricle growth restoring the normal circulation. Valid alternative to Heart transplantation in severe right ventricle dysfunction.

SECTION VIII

DISCUSSION

DISCUSSION

Determining optimal repair strategy in a situation where one ventricle is functionally or anatomically marginal in size remains a challenge. The bias that a biventricular repair is always better than a single ventricle repair has been questioned by the finding that operative mortality for a complex biventricular repair can be higher than a single ventricle repair with similar anatomy.[91] Mortality outside of the operative window may also be higher in complex biventricular repairs, as demonstrated by era independent late mortality in the Rastelli operation.[92, 93] For marginal left ventricles, a binary decision regarding the adequacy of the left ventricle is made. For marginal right ventricles, there is the opportunity to incrementally volume unload the right ventricle. [53] As compared with the requirements of a marginal systemic ventricle, the 1.5 ventricle repair, for hypoplasic or failing right ventricle, thus offers the considerable advantage of a variable volume load on the right while largely separating the systemic and pulmonary circulations. [1, 2, 91, 94 - 99]

The surgical management of patients with mild to moderate RV hypoplasia has improved, but the management of those with an RV where is too small to accommodate an entire cardiac output has been less satisfactory. [1, 36, 99, 100] The goals of surgical intervention for this group are to eliminate cyanosis by separating the pulmonary and systemic circulations while avoiding complications related to venous hypertension in inferior vena cava district. These goals are trying to met by using different modifications of Fontan type procedures or by establishing a right ventricle-to-pulmonary artery continuity and using a bidirectional cavopulmonary anastomosis to reduce the volume of systemic venous return that the RV must handle.[1, 36, 101] Both these approaches have disadvantages. In the Fontan approach the main pulmonary artery generally is ligated, leaving a hypertensive RV with the potential to disturb the function of the systemic

ventricle or to cause arrhythmias.[98-105] When a bidirectional cavopulmonary anastomosis is used, the blood flow from the inferior vena cava must pass through the tricuspid valve and RV to the pulmonary artery. Good results have been reported with this approach in patients with an RV volume as low as one third of the left ventricular volume.[71] However, if the RV and/or tricuspid valve are very small, inferior vena cava flow may be restricted. It can be difficult to predict whether the RV and/or tricuspid valve will accept the inferior vena cava blood flow because of difficulties in measuring RV volume and because valvar tricuspid stenosis is often associated with RV hypoplasia.[106] An adjustable atrial septal defect [1, 71] or an atrial septal fenestration may maintain cardiac output and reduce central venous pressure but at the expense of some arterial desaturation. Alternatively, there may also be patients committed to a Fontan-type circulation with its long-term complications [107, 108] who may have benefited from inclusion of the RV into a repair.

For these reasons, we studied alternative operative approach. By anastomosing the superior vena to right pulmonary artery and inferior vena cava blood has left to flow through the right ventricle to pulmonary artery. The hemodynamic consequences of an excessively small tricuspid valve and/or RV are thus avoided by "decompression" of the right atrium via the cavopulmonary anastomosis. If pulmonary vascular resistance is low, this procedure should accommodate the inferior vena cava return through right ventricle without any problem. Increases in inferior vena cava blood flow associated with normal growth and exertion should not result in excessively high right atrial pressure.

By maintaining the RV in the atriopulmonary circulation, RV growth may continue and the pulsatile flow may enhance pulmonary artery growths. In these patients pulsatile flow provides to both lungs. The RV may contribute to pulmonary blood flow, imparting both kinetic energy and pulsatility.

A pulsatile pulmonary flow within pulmonary district may be advantageous in that vascular resistance to pulsatile flow is lower than to nonpulsatile flow, [109, 110] regional pulmonary blood flow may be more uniform, [111] and lymphatic drainage may improve. [112] The role of nonpulsatile blood flow in the development of pulmonary arteriovenous fistulae is more controversial.[113-117] Exclusion of hepatic venous return from the pulmonary circulation may be associated with the formation of such fistulae in patients following a classic Glenn-type cavopulmonary anastomosis .[118, 119] We hypothesize that establishing a communication between the inferior vena cava and the pulmonary arteries may be important in preventing pulmonary arteriovenous malformations. Longer follow-up of patients in this series may shed some light on the question of which etiologic factor; pulsatile flow or hepatic vein origin of flow, is important in the pathogenesis of pulmonary arteriovenous fistulae, since those with the proximal right pulmonary artery interrupted have pulsatile flow in the left lung only, whereas hepatic venous blood reaches both lungs.

Medium-term outcome after this operation is excellent, with no mortality and good functional status in all patient. All patients are acyanotic. Doppler echocardiographic studies demonstrated flow from the right atrium toward the pulmonary artery during atrial systole in all patients thus studied, indicating decompression of the right atrium during that part of the cardiac cycle. Flow from the inferior vena cava to the right atrium during the remainder of the cardiac cycle, forward flow across the tricuspid valve throughout ventricular diastole, and a discrete ventricular systolic flow jet across the RV outflow tract, which were seen in all patients.

There is potential disadvantage to this operative approach; is that increased reverse flow to the superior vena cava but which was proved to be well tolerated by patients with Fontan procedure at midterm follow up. When the RV is very small, left pulmonary artery blood flow may be reduced or, if the RV is large and compliant, right pulmonary

artery flow may be reduced. Reduced pulmonary blood flow may have a detrimental effect on the growth of the pulmonary vascular tree.[120] Unfortunately, reliable assessment of differential lung perfusion is difficult in this group of patients because of the multiple sources of pulmonary blood flow. Preoperative and postoperative branch pulmonary artery sizes were similar, but because the interval between operation and catheterization was relatively short, these measurements may not reflect growth in the more distal pulmonary vascular bed.

In this relatively small group of patients, a continuation of cavopulmonary anastomosis (between superior vena cava and right pulmonary artery)with the inclusion of the hypoplasic RV into the pulmonary circulation showed excellent short- and midterm results. Postoperative right atrial pressures were satisfactory, and there have been no significant atrial arrhythmias.

Our animal study in 30 rabbits also showed well tolerated hemodynamic changes with increased pressure at SVC level with normal right atrial pressure after 1.5 ventricle repair, even after pharmacologically induced stress test. Which is a very positive finding in favor of one and a half ventricle repair in comparison to Fontan type procedure for patients with hypoplasic, failing or borderline right ventricle.

Acknowledgments

1. The author wish to thank Dr. Vladimiro L. Vida for his important support and collaboration for this research project and Dr. Arben Dedja, Mrs Laxmi Shrestha, Mr Silvio Ferron for their valuable cooperation and assistance in animal laboratory.

2. The Author wish to inform that this project has won the prestigious international Award "The Francis Fontan Prize" in the year of 2011 and this study was presented at 26th annual meeting of European Association for Cardio-Thoracic Surgery (EACTS) in Barcelona, Spain.

Fig: 46. Dr. Anil Bhattarai receives his Award from Prof. Pieter Kappetein(left) and Prof. Francis Fontan at 25th EACTS meeting 2011, Lisbon.

3. Special thanks to Prof. Lucio Parenzan, Prof. Giovanni Stellin and Prof. Gaetano Thiene for their scientific support during my PhD in Padova

Fig: 47. From left: Prof. G. Stellin, Dr A. Bhattarai and Prof. L. Parenzan; 25th EACTS Lisbon 2011

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