FONTAN SURGERY EVOLUTION – REVIEW ARTICLE

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Introduction

In the past four decades after its introduction in 1971, the Fontan procedure for univentricular heart has evolved a lot. The single ventricle circulation in many complex congenital heart disease is characterised by two circulations namely the systemic and pulmonary, which are in parallel and are powered by a only single ventricle instead of normal in series circulation powered by two separate ventricle. This anomaly results in hampered quality of life due to constant desaturation of arterial blood both in rest and exercise and chronic heart failure around 4^{th} decade due to considerable volume overload of single ventricle. The palliative Fontan circulation revolves around the idea that only a pressure gradient of 6 mm Hg is needed to propel the blood from pulmonary artery to lungs(1) and right heart can be bypassed for more energy efficient circulation. This palliative surgery connects systemic circulation to pulmonary circulation directly without any intervening ventricle and brings normalisation of arterial saturation and decreases the chronic volume overload of single ventricle.

We will discuss in this review article how the Fontan palliatition has evolved in every aspects and its future course, since its introduction in France in by Francis Fontan.

Indications

The Fontan operation was 1^{st} used for Tricuspid Artesia and hypo plastic right ventricle. Now it is understood Fontan palliation can be applied in any type of single ventricle where the other ventricle is hypo plastic (when the size of the ventricle is less 30% of normal the hypo plastic ventricle cannot contribute adequately to circulation) (2).

Now the indication of Fontan circuit encompasses wide variety of complex congenital malformation like Tricuspid Artesia, pulmonary atresia with intact ventricular septum, double inlet left or right ventricle, complete unbalanced AV canal, Hypo plastic left heart syndrome, DORV with non routable uncommitted VSD. Very complex malformation with high surgical risk of morbidity and mortality and 'high maintenance cases' where there is frequent need for conduit replacement, a Fontan strategy can lower the need for re –intervention (3). So remarkably the Fontan operation has become the most common procedure performed for congenital heart disease after the age of 2 years.

Selection of patients

With the passage of time and experience the initial strict criteria laid down by Choussat and colleagues for patients of Tricuspid Artesia (4) (table 1) has become more flexible and some new criteria has emerged.

Age- successful Fontan operation can be performed 7 months onward and in fact in this era of staged Fontan the preparation for this procedures starts from neonatal age group in the form of shunt, banding, Norwood procedure or stenting of PDA

Sinus rhythm –It is not considered as absolutely necessary criteria for Fontan surgery at present. But presence of arrhythmia or new onset arrhythmia has definite role in early failure of Fontan palliation. Resynchronisation therapy has been found to be effective in univentricular hearts with prolonged QRS and dys-synchronous ventricular contraction (5).

Normal Caval drainage – In interrupted inferior vena cava with azygose connection and single ventricle Kawashima procedure (Bidirectional Glenn procedure used to direct half the body's venous blood flow into the lungs) is a standard palliation at present (6)

Right atrium of normal volume- In modern era of total cavopulmonary connection (TCPC) right atrial morphology has lost its importance

Mean pulmonary artery (PA) pressure ≤ 15 mm Hg- still valued as desirable criteria for Fontan surgery. In modern era, various catheter based procedure can effectively predict the pulmonary artery pressure and also helps in decreasing high pressure by embolisation of artopulmonary collaterarals. Also successful interruption of shunt, Persistent ductus flow and existing native pulmonary flow aids in lowering the pressure intraoperatively. In relatively borderline high PA pressure staged palliation like bidirectional Glenn shunt is applied where the indications are more relaxed compared to Fontan(7).

Pulmonary arterial resistance < 4 Woods unit/m_2-it still remains as an absolute criteria for Fontan palliation

Pulmonary artery diameter –size of pulmonary vessel is a important prognostic criteria for outcome of Fontan operation . It is now better predicted by Mc Goon ratio: the sum of diameter of prebranching left and right pulmonary arteries divided by diameter of descending thoracic aorta above the diaphragm level. A retrospective study by Fontan and colleagues revealed the relation of premature death and early attrition of Fontan circulation in patients with McGoon ratio less than 1.8(8).

Normal Ventricular Function- This is still a important prerequisite criteria for Fontan surgery.But with experience and time it is now under stood that ventricular dysfunction due to previously done shunt surgery or banding is not always an absolute contraindication and they should become an candidate for staged procedure and should be subjected to bidirectional Glenn procedure before proceeding to total cavopulmonary anastomoses.

Some other factors are now considered important for a successful Fontan surgery like a) after repair left atrial pressure (it must be low after surgery) b) a low transpulmonary gradient following surgery c)unobstructed ventricular inflow(no atrioventicular valve stenosis or regurgitation) d)unobstructed outflow (no sub aortic stenosis,no arterial hypertension, no coarctation) e)non restrictive connection from systemic vein to pulmonary artery(fontan connection) f) unobstructed pulmonary venous return (3).

Surgical Evolution

Older circuits- various Atrio pulmonary connection were created between **right atrium** and **pulmonary artery**(figure 1 A). It may be a) anterior atriopulmonary connection with or without inclusion of small hypoplastic right ventricle b) posterior atriopulmonary connection. Different materials were used to create these connections like valved conduits, homograft, patches and direct anastomoses.

But as these patients are followed up it was seen streaming of blood flow from systemic vein to lungs is not satisfactory in these connection giving rise to various complications like thromboembolism, right atrial dialation ,arrhythmias, early attrition and failure resulting in very high reoperation rate –up to 40% in some studies(3) **.**

Most of these older circuits are no longer created, however many patients still survive on such circuits

Total cavopulmonary connection (TCPC)- In 1988 a land mark paper by pioneer surgeon Dr Mark Deleval proved TCPC to be a more logical alternative to atriopulmonary connention.In this operation caval veins are connected to pulmonary artery bypassing not only the hypo plastic ventricle but also the right atrium(9).The superior venacaval flow is directed to pulmonary artery by bidirectional Glenn Shunt and inferior venacaval blood is directed to pulmonary artery by **lateral tunnel technique (** a tubular passage inside the right atrium consisting of a prosthetic baffle and apportion of lateral wall of right atrium) connecting the inferior venacaval orifice to pulmonary artery(Figure-1 B). The advantages of this circuit were a) it could be applied in very small children also as it has growth potential b) to some extent atrial tachyarrhythmia incidence is decreased since only minimal amount of atrial tissue is exposed to high venous pressure.

Still handling and suturing of the right atrium ,reqirement of cardiopulmonary bypass and crossclamp of aorta could not be avoided in this procedure .

To answer these questions **extracardiac conduit** (figure 1C) for connecting the inferior venacava to the pulmonary artery was introduced by Marcelletti in 1990s(10).This connection bypasses the entire right atrum with minimal or no suture line in it and can be performed with minimal or no cardiopulmonary bypass support without crossclamping of aorta but has the disadvantages of flow stasis in conduit with resulting increased incidence of thromboembolism, no growth potential-so can be offered to older children large enough to accept a graft adequate for an adult's inferior vena cava.

Fenestration and subsequent closure- It is the recent modification of Fontan circuit introduced by Castaneda and Bridges in 1990(11). This was achieved by creating a fenestration in the atrial baffle or creating a connection between extra cardiac conduit and right atrial chamber in high risk patients to ward of the stormy postoperative period in the expense of allowing a modest desaturation. This artificially created fenestration can be closed by catheter based technique in a later date. It allows right to left shunt to reduce caval pressure and congestion, increases preload of single ventricle to

increase the cardiac output-thus reducing the mortality and morbidity (i.e. pleural drainage) in post operative period.

Staging of Fontan palliation-

In fact in modern era, the stage approach is preferred whenever a child is selected for univentricular pathway. This staged approach is beneficial in several ways as-

a) It allows the body to adapt to very different hemodynamic conditions
b) Reduces the overall operative mortality & morbidity
c) Allows a better patient selection
d) Keeps room for interim intervention like correction of AV valve regurgitation, correction of Left ventricular out flow obstruction, correction of pulmonary arterial distortion for better preparation for future Fontan pathways.

Staged approach to Fontan Circuit:

A) Neonatal period-

AIM	PROCEDURE
• Remove undue restriction /flow Obstruction from single ventricle to aorta.	Repair of associated coarctation if present Damus –Kaye-Stansel Norwood repair
• A well balanced limited flow to lungs	Pulmonary artery Banding Aorto pulmonary shunt Stent in the duct
• Unrestricted return of blood to ventricle	Rashkind balloon atrial septostomy Open atrial septectomy

B) At the age of 4-12 months-

Bidirectional Glenn shunt –where the superior caval blood is connected to pulmonary artery (bilateral if present). At this stage patient will be subjected to lesser volume overload of single ventricle (as 1/3rd of venous return is directed directly to lungs) and remain slightly cyanotic (as inferior vena caval desaturated blood still mixing in the single ventricle)

C) At 1-5yrs of age-

Patient will be followed up after BD Glenn for saturation, hematocrit and growth of vascular structures. At appropriate time (that may vary from centre to centre and according to process selected i.e. Lateral tunnel vs. extra cardiac conduit) Fontan circuit is completed by connecting Inferior Vena Caval blood return to pulmonary artery either by lateral tunnel technique or extra cardiac conduit with or without fenestration.

Complete Fontal vs. bidirectional Glenn with additional flow-

Sometimes during creating a bidirectional Glenn shunt the limited existing flow through stenosed pulmonary valve or right ventricular outflow tract or a small aortopulmonary shunt or through pulmonary band is kept intact provided this blood flow is controlled without adding much pressure overload (i.e. acceptable Glenn pressure). This creates an opportunity for higher arterial saturation and more growth of pulmonary artery and may be taken as the final palliation. But many children outgrow this procedure and eventually had to be converted to full Fontan (3).

Mortality

Perioperative and early mortality

The mortality has decreased markedly during last 3 decades. The Perioperative mortality has stabilised at around 5%.and in some centre of excellence it has come to nil (12).

The factors that have led the improvements are 1) more energy efficient circulation in the form of lateral tunnel or extra cardiac conduit 2) decrease aortic cross clamp time and cardio pulmonary bypass time with extra cardiac conduit technique 3) Fenestration technique 4) staging of Fontan procedure 4) concomitant clamping of any collateral aorto pulmonary flow (13).

Late mortality

Previously quoted 20 year survival of Fontan patients with atrio pulmonary connection was 65%. Even when the operation was carried out in ideal circumstances there was a progressive regression of functionality of circulation and survival 5, 10 and 15 years after surgery was 86%, 81% and 73% respectively (14).

Mair and colleagues (14) described the risk factors contributing the long term outcome after the Fontan operation was

a) The age of the patient before the operation
b) Existence of previously created palliative surgical procedure
c) The anatomy of complex congenital heart disease i.e. tricuspid atresia is not a risk factor but unbalanced complete AV canal or hypo plastic left heart syndrome is a risk factor
d) Heterotaxy Syndrome
e) Elevated pulmonary artery blood pressure before the time of operation
f) Failure of atrioventricular connection
g) NYHA III/IV before operation

Khairy et al have provided excellent data on mortality in the early Fontan patients after 15 to 20 years (15). They reported 82.6% of early survivors were alive and had not had a cardiac transplant 15 -20 years later.(they have taken into account patients done after 1980).Among the long term survivors 3 most common causes of late deaths are thromboembolism, heart failure, sudden death. According to d'Udkam et al (16) 15 year survivals of the patients having a lateral tunnel was 94%. Although there was a gradual increase in the rate of failure of atriopulmonary connection Fontan after 10 years this trend has not yet observed in lateral tunnel Fontan.

Functional status of the late survivor

Most patients with a Fontan circulation lead a normal life and even indulge in mild to moderate sports activity. More than 90 % of late survivors are in New York Heart Association (NYHA) functional class I and II (3). Most patients can pursue the education and professional carrier like normal people. However with time there is a progressive decline in functional status in some subgroups. Recently Paediatric Heart Network of National Heart, Lung and Blood Institute have conducted a study to delineate the predictors of Functional status of Fontan patients (17). A Fontan functional score was developed in the study using a) Ventricular ejection fraction by Echo cardiogram b) percent predicted maximal oxygen consumption c) child health questionnaire d) brain natriuretic peptide levels. The risk factors which are associated with poor functional score were Time since Fontan, Right ventricular morphology, pre Fontan end diastolic pressure, prefrontal oxygen saturation, parental income. Variables which are not independently associated with poor functional score were surgical centre, age, weight, fenestration, length of hospital stay at time of Fontan operation and post Fontan surgeries and interventions.
Obvious variables that were analysed explained only 18% of Fontan functional score, 82% of the data represented factors still unknown.

Fontan failure or Failing Fontan

Fontan circulation may become "failing" and unbearable because of persistent congestion and oedema due high venous pressure and ventricular failure over time, Low cardiac output with very limited exercise tolerance, relapsing tachycardia, protein losing enteropathy , plastic bronchitis, hypoxemia caused by residual shunt and collateral circulation and thrombo embolic complications. These are complication of Fontan surgery and respond very poorly with conventional treatment with inotropes, vasodilator and diuretics (3).
Frequently a more aggressive approach will be needed to re optimise the circuit, like stenting of stenosis, embolising the collaterals, conversion of atriopulmonary connection to cavopulmonary connection with right atrial maze procedure and atrial reduction plasty.
If this is insufficient fenestration and heart transplantation should be considered.

Future prospect:

In the last few years important progress has been made in the evolution of selection criteria, in the development of surgical techniques and in the design and staging of Fontan circulation. However it appears we still have a long way to go as regards to a complete understanding the physiology and attrition of Fontan circulation. Clinical experience and experimental studies in combination with the cooperation of different fields in medicine and positive sciences are definitely expected to help the evolution even more.

Re-engineering the approach to single ventricle repair

The Fontan paradox exists in all Fontan patients characterised by a systemic venous hypertension (especially in the splanchnic part) with relative pulmonary arterial hypertension. Various proposals are now being investigated for optimisation of flow of in TCPC circuit.

Optimisation of Flow- In a Fontan circulation any potential for pressure loss either in form of anatomic pressure loss across TCPC connection(2-5 mm of Hg)or physiologic pressure loss across the pulmonary capillary bed (5-8 mm Hg) is of critical importance. To decrease the pressure loss due to colliding and recirculation of flow in a four way flow path various optimisation have been proposed. These include either lateral or anteroposterior offset of venacaval axes, vena caval splitting surgery, branched synthetic conduit and flow splitting devices sutured in the midst of TCPC junction(18,19). But these experiments remain to be resolved in clinical practice

Powering the Fontan- according to pioneer researcher and surgeon de Laval the Fontan circulation paradox could be potentially reversed ,if a mechanical assisting device is implanted that could lower the pressure in inferior venacaval by 5 mm of hg while also producing an increase in pulmonary artery pressure by 5 mm of Hg(20). A cavo pulmonary assist pump should serve as low energy input primer of the primary pump (single ventricle) identical to the essential function right ventricle(21). No such pump currently exists. A catheter based intravascular device which utilizes the existing venous path way is an attractive option.

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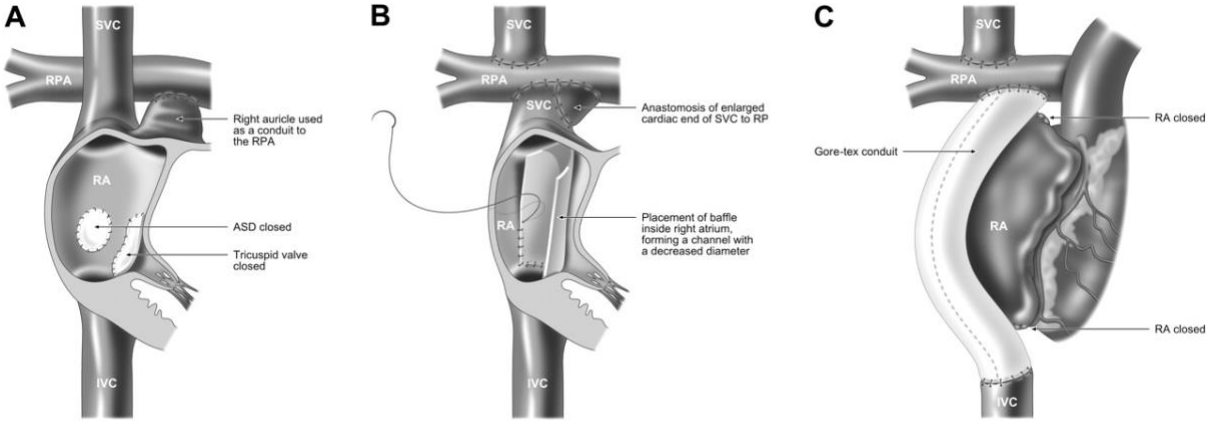
Table-1

Primary selection criteria for patients with tricuspid atresia for the Fontan operation.

1. Minimum age 4 years
2. Sinus rhythm
3. Normal caval drainage
4. Right atrium of normal volume
5. Mean pulmonary artery pressure $\leq$15 mmHg
6. Pulmonary arterial resistance <4U/m2
7. Pulmonary artery to aorta diameter ratio $\geq$0.75
8. Normal ventricular function (ejection fraction >0.6)
9. Competent left atrioventricular valve
10. No impairing effects of previous shunts

Figure-1


A
SVC
RPA
Right auricle used as a conduit to the RPA
RA
ASD closed
Tricuspid valve closed
IVC
Classical Fontan
B
SVC
RPA
SVC
Anastomosis of enlarged cardiac end of SVC to RP
RA
Placement of baffle inside right atrium, forming a channel with a decreased diameter
IVC
Lateral tunnel (intra-atrial baffle)
C
SVC
RPA
RA closed
Gore-tex conduit
RA
RA closed
IVC
Extra-cardiac conduit