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Original Article

SURGICAL MANAGEMENT OF SINGLE VENTRICULAR ANATOMY: EARLY NICVD EXPERIENCE

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Objective: We present early results of surgical management of single ventricular physiology with establishing two separate circulations.

Background: The cavopulmonary anastomosis eliminates the systemic hypoxemia and ventricular volume overload characteristic of single ventricular physiology as well prior forms of palliations.

Methods: we are presenting early results of our surgical managements of univentricular hearts to achieve two separate circulations. Form June 2011 to November 2012 nine patients with single ventricular physiology under went for establishing two parallel circulations at NICVD.

Results: Overall survival was 67%. There was no operative mortality while two patients have early mortality and one has late mortality.

Key words: Single ventricle, congerite hear disease, cardiacsurgeyr.

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INTRODUCTION

A heart with two two atriums but one ventricle always fascinated pediatric cardiac surgeons. An anatomy of challenge for pediatric cardiac surgeons. Chemineau was one who first described a heart composed of 2 auricles but only 1 ventricle in 1699[1]. New England registry reported the incidence of univentricular heart to be 54 cases per million live births[2]. In hypoplastic left heart syndrome alone, the most common form of univentricular heart, a crude median incidence of 2.3 cases per 10 000 live births has been reported[3]. Tricuspid Artesia, the second most common subtype of univentricular heart, is thought to occur less than once for every 10 000 live births[4] and was present in 2.9% and 1.4% of congenital heart disease autopsy[5]. Single ventricular heart always remain a matter of debate for its nomenclature, embryology, hemdynamics and an anatomy with strange anatomy responsible for most creative surgical and interventional approaches in medical history. A number of different terminology used for univentricular heart; "functionally single ventricle," "cor triloculare biatrium (well-formed atrial septum)," "cor biloculare (rudimentary or absent atrial septum)," "common ventricle," and "functionally single ventricle[6,7]. Possible causes of single ventricular hearts include tricuspid atresia, mitral atresia, double inlet RV, double inlet LV, Heterotaxia syndrome[8]. Though hypoplastic left heart syndrome a common form of univentricular physiology not included under the umbrella of single ventricle malformation[9]. Moodie at all reported largest series of unoperated 83 patients with functionally single ventricle with 70% well forms left ventricle were died before the age of 16 years while patients with RV heart morphology have only 50% survival by 4 years[11]. The common causes of mortality were arrhythmias, congestive heart failure, and sudden unexplained death. Surgical options for correction of functionally single ventricular physiology is either palliative procedure like systemic to pulmonary shunt or physiologically corrective procedure restoring parallel systemic and pulmonary circulation. Corrective procedure are always staged resulting in fonton circulation establishing two parallel circulation. However for

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left hypoplastic heart complex we may need to established norwood circulation but finally Fonton circulation will be established.

METHODS

Form June 2011 to November 2012 nine patients with single ventricular physiology under went for establishing two parallel circulations at NICVD. Seven patients had hypoplastic right ventricle while two had hypo plastic left ventricle. These patients underwent bidirectional Glenn or Fonton procedures. Median follow up was 7 months.

Preoperative Doppler/echocardiography

Hypoplastic RV physiology was present in seven patients while left ventricular hypolplasia was found in two patients. Tab 2,3.

Table 1
There were 7 males and two females. Median age was 5 years. Median wt 16.3 kg

S #	Age	Sex	Weight kg	Blood Group	HB%	Urea	Creatnin
1	5	M	19	A	15.9	24	.5
2	9	M	39	O	23	34	.5
3	10	M	14	O	14	23	.8
4	4	M	7.5	A	17	17	.4
5	2	M	10	B	16.4	42	.6
6	2	F	23	O	17	15	.9
7	3	F	28	B	13	15	.4
8	8	M	14	O	23	18	.6
9	8	M		A	16	10	.3

Prior surgical procedures included pulmonary artery banding in one patient and systemic to pulmonary artery shunt was present in two patients.

Operative technique

A median sternotomy is performed and initially the aorta and right atrium are exposed. Heparin is administered to achieve an ACT suitable for cardiopulmonary bypass followed by cannulation of the ascending aorta and placement of a single atrial cannula for systemic venous drainage. Extracorporeal flow is established at 150cc/kg/min. All the patients underwent normothermic cardiopulmonary bypass and all systemic-to-pulmonary artery shunts are occluded. Then either a bidirectional Glenn shunt by creating anastomosis between superior vena cava and right pulmonary artery in end to side fashion or complete Fonton circulation was established as single stage procedure. Fonton circulation was established with superior vena cava anastomosis with right pulmonary artery while inferior vena cava flow is directed to mean pulmonary artery in one case and confluence in three cases by extra cardiac conduit.

Table 2

Diagnosis	Number	%
Tricuspid atresia	4	45
Dorv	3	33
AV canal defect	1	11
Mitral atresia	1	11

Table 3
HR = HYPOPLASTIC RV,PA=PUL ATRESIA,TA=Tricuspid ATRESIA,DOVR= DOUBLE OUTLET RV,MiA=MITRAL ATRESIA

S #	Echo	Surgical procedure
1	HR VSD, DTGA,PS , ASD, NO INOMINATE PER LSVC	BI DIR GLENN
2	TA,ASD,VSD, HR	FONTON
3	VSD,PS, HR,TA	FONTON
4	VSD,PA, R,DORV,DTGA,	FONTON
5	Mi A,VSD,ASD,PS ,DORV	BI DIR GLENN
6	TA,ASD,VSD, HR	BI DIR GLENN LARGE SIZE AZYGUS LIGATED
7	PS,AV CANAL DEFECT, VSD ASD,DORV,DTGA	BI DIR GLENN
8	ASD,VSD,TA,PS	BI DIR GLENN
9	VSD,PS,H R	FONTON

Table 5: Type of Previous Palliation

Blalock-Taussig (modified)	2
Pulmonary artery banding	1

Table 6: Early Outcome

operation	Icu stay	support	complication	Early mortality
Bi glenn56%	75.6	Ad + GTN		2
fonton44%	56.25	Ad + GTN	chylothoracic	0

RESULTS

ICU course showed in table 6. All the patients were shifted in ICU at mechanical ventilation and pharmacological support; adrenaline And GTN infusion. Mean ICU stay was 66.3 h. one patient shifted from ICU to ward but developed chylothoracic; tube thoracostomy done. Once chylothoracic resolved patient is discharged but later readmit with same complication and failed to recover. There were two early deaths due to low output failure.

DISCUSSION

The term "single ventricle anomaly" is purposely non-specific. It is used to describe a group of cardiac defects that may differ quite dramatically from each other but share the common feature that only one of the two ventricles is of adequate functional size. Single ventricular physiology is always remained a challenge for parents and cardiac surgeons. Babies born with single ventricular physiology have very dismal natural history. They died due to arrhythmias, congestive heart failure, and sudden unexplained death if left unoperated. In the normal heart each ventricle does a separate job. The right ventricle pumps blood to the lungs, and the left ventricle pumps blood to the body. In a single ventricle heart, there is only one ventricle large enough to do the normal job of pumping blood to both circulations. These babies have continuous two circulations rather than parallel circulations. surgical treatment of this anatomy is either palliation; that is to improve lung perfusion to maximize oxygenation of blood and corrective surgery; establishing two parallel circulation. General objectives of initial surgical palliation are to provide unobstructed systemic outflow, unobstructed systemic and pulmonary venous return, and controlled pulmonary blood flow.Pre Fonton palliation includes; Pulmonary artery banding, Modified BT shunt, Atrioventricular valve repair, Correction of TAPVC (if present), DKS or Norwood principle, Glenn's shunt: superior cavopulmonary connection .

In patients with unobstructed pulmonary blood flow one may require PA banding to improve systemic flow or division with creation of an aortopulmonary shunt to limit pulmonary blood flow[19]. Pulmonary banding has been associated with adverse outcomes after the Fontan procedure and may result in sub aortic obstruction[12]. While patients with severe pulmonary obstruction or atresia, palliation are achieved with an aortopulmonary shunt, such as a modified Blalock-Taussig shunt or bidirectional cavopulmonary anastomosis (Glenn shunt).

Fontan procedure was first described by Fontan and Baudet for tricuspid atresia using valved conduit between the right atrium and pulmonary artery in 1971[13]. Fonton procedure has undergone multiple modifications to encompass several forms of palliative surgery that divert systemic venous return to the pulmonary artery, usually without interposition of a sub pulmonary ventricle. In 1987, de Leval et al proposed an end-to-side anastomosis of the superior vena cava to the undivided right pulmonary artery, a composite intra-arterial tunnel with the right atrial posterior wall, and a prosthetic patch to channel the inferior vena cava to the transected superior vena cava[14]. Inferior vena cava flow can also be diverted to pulmonary artery via extra cardiac conduits. Fontan pathways may be "fenestrated" by creation of an ASD in the baffle or patch to provide an escape valve that allows right-to-left shunting, which may be beneficial early after the surgical procedure[15]. If

hemodynamics are favorable, these fenestrations can later be closed by a transcatheter approach[16]. Patients with left sided hypoplastic heart my require additional Norwood procedure in neonatal period followed by Fonton to achieve complete Fonton circulation[17,18].

Our data includes 9 patients five patients undergone Glenn while 4 achieved complete Fonton circulation. Two patients undergone Glenn have early mortality during ICU stay. These both failed to extubate and died due to low output failure.

While one of four achieved Fonton circulation developed chylothoracic despite of modified Fonton with fenestration. Though child recovered and discharged but returns back with chylothoracic and fail to resolve this time and child expired.

None of the patients have postoperative rhythm disturbance or thromboembolic episode was observed . While remaining six patient have complete follow up till to date.

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