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One and One half Ventricular Repair with Pulsatile Bi-directional Glenn: Concept, Concerns and Future Directions

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ABSTRACT

The present perspective is a synthesis of 98 published investigations in the setting of one and one half ventricular repair with pulsatile bi-directional Glenn. In this article, we have attempted to address several concerning issues and controversies with reference to morphologic and physiologic criterions for candidate selection, creation of atrial septal fenestration, long-term fate of an unligated azygos vein and free pulmonary regurgitation, the concerns of a pulsatile bi-directional Glenn on superior vena caval blood flow, the growth potential of the pulmonary ventricle and subsequent eligibility for conversion to bi-ventricular repair and long-term functional results. Additionally, this review attempts to address the guidelines for candidate selection for one and one half ventricular repair, bi-ventricular repair and univentricular type of repairs.

Keywords: Biventricular repair, One and one half ventricular repair, Pulsatile bi-directional Glenn, Pulmonary artery banding, Univentricular repair

Introduction (Concept)

The concept of "one and a half ventricular repair" introduced by Billingsley and colleagues in 1980 relates to situations where one ventricle is capable of pumping one half of the circulation while the other ventricle is deemed inadequate and requires off-loading by means of a shunt. The inadequate ventricle is usually assigned the role of pumping the pulmonary circulation. The majority of hearts potentially amenable to this repair will have one large ventricle associated with a smaller and more-or-less rudimentary ventricle [1-5].

In congenital heart disease, ventricles that are surgically separated are said to undergo a "two ventricle repair," whereas ventricles that cannot be separated surgically, commonly undergo the Fontan operation, or "one ventricle repair." Often the morphologic and physiologic characteristics of the right side of the heart are thought to be insufficient to carry a complete cardiac output. In recent years, the tendency has been to subject such patients to the so-called "one and a half ventricle repair." In this procedure the heart is surgically septated [6-10].

A hypoplastic but potentially usable right ventricle (RV) occurs in a variety of clinical situations. The disease spectrum includes patients with i) atrial isomerism complex, ii) isolated congenital right

ventricular hypoplasia, iii) right ventricular outflow tract obstruction with a hypoplastic RV, iv) pulmonary atresia with intact ventricular septum, v) tricuspid stenosis with atrial septal defect (ASD), vi) right ventricle hypoplasia in patients with ventricular septal defect (VSD), tetralogy of Fallot (TOF), complex transposition of the great arteries (TGA) with large VSD, vii) inlet VSD with a straddling tricuspid valve, viii) Ebstein's anomaly (Carpentier's type C and D),ix) complete atrioventricular canal with extreme left ventricle dominance, TOF, double outlet right ventricle (DORV), x) double outlet right ventricle with remote VSD, xi) Uhl's anomaly, xii) right ventricular endomyocardial fibrosis, xiii) hearts with crossed atrioventricular connections, xii) hearts with supero-inferior ventricles. The horizontal ventricular septums of these hearts produce a matrix for supero-inferior ventricles. Usually, the superiorly positioned ventricle is the morphologically right (pulmonary) ventricle and the inferiorly positioned ventricle is the morphologically left ventricle [9-38].

Treatment strategies for these groups of patients include (1) the superior cavopulmonary connection, (2) a univentricular type of repair, or (3) a biventricular repair with atrial septal fenestration. As originally described by Billingsley and colleagues, the one and one-half ventricle repair (1.5 VR) recruits the hypoplastic RV, presumably providing kinetic energy and pulsatility to pulmonary blood flow, while the superior cavopulmonary connection reduces the volume load of the pulmonary ventricle [1,8,10]. However, the ability of the pulmonary ventricle to generate appropriate indexed cardiac output is dependent on morphology, function and pulmonary vascular resistance (PVR) [32,36-38].

In the perioperative period, a 1.5 VR is useful only if morbidity and mortality are less than or equal to those of the corresponding bi-ventricular or univentricular type of repair. Reported mortality has varied from 0% to 12% which compares favorably with the mortality of the Fontan procedure [9,38-46]. Providing hepatic venous blood to both lungs, flexibility to increased pulmonary vascular resistance, and circulation at low venous pressure in the IVC system improves the outcome [46]. Mortality for marginal situations and biventricular repair is not well delineated in the published literature. In certain circumstances it is high [9,12,15, 34-36,41,42,48].

Long-term results of Fontan operation are still debatable with a definite long-term attrition. The Achilles' heel of Fontan circulation is inferior vena caval hypertension. The venous pressure of the splanchnic system is higher than that of the inferior vena caval system because of hepatic sinusoids. Since the systemic venous circulation is in series with the pulmonary circulation, these patients develop chronic hepatic congestion with as cites, pulmonary arteriovenous malformations, plastic bronchitis, arrhythmias, thromboembolism, protein-losing enteropathy and ventricular failure [34-36,42]. Chronic atrial arrhythmias, cyanosis or protein losing enteropathies have not been identified in the short-term and long-term follow-up of 1.5 VR [43-46]. Although most of the isolated reported cases have quoted a functional class of I and II, no cohort data is available on measured exercise capacity and maximal oxygen consumption [9-46].

It has predominantly been used to achieve a septated circulation in the setting of borderline or small right ventricular size – typically in pulmonary atresia with intact septum. The shortcomings of the Fontan procedure have been well documented and the idea of imparting some degree of kinetic energy to pulmonary blood flow is enticing, provided the anatomic substrate allows this approach [47].

Previous reports have largely concentrated on establishing the anatomic criterions for this operation. Right ventricular size is a continuum, ranging from ventricles that are clearly unsuitable for use to those that are nearly normal. Within this range lies the bandwidth of ventricles that are capable of dealing with venous return from the inferior vena cava, but not the entire systemic venous return. Establishing where in this spectrum the right ventricle is satisfactory for the 1.5 VR is probably the predominant concern when considering this approach for a given patient, so the emphasis in previous reports is understandable and appropriate [28,31,36,40,41].

What do we really know about one and one half ventricle repair?

We certainly know that the operation can be accomplished with acceptable surgical mortality, morbidity and midterm functional results. A review of the published literature enunciated in this manuscript only tell us that the procedure can be performed relatively safely and that patients seem to do reasonably well at follow-up [6-8].

What we do not know about one and one-half ventricle repair?

At one end of the clinical spectrum that involves a decision between a two ventricle repair and a 1.5 VR, we do not know what specific criterions should be used for choosing between the two operations, and we do not know whether individuals with right hearts that have concerning morphologic and physiologic characteristics actually are more functional with 1.5 VR repairs or bi-ventricular repairs [6].

Nothing in the literature even remotely addresses the issue of making a qualitative assessment of the right side of the heart; the "difficulty factor" which is being encountered in performing intricate intracardiac repair to divert inferior venacaval blood alone and the safe upper limit of pulmonary artery pressure (PAP) and pulmonary vascular resistance (PVR) that can be used to objectively decide between surgical options. The surgeon is left to make a clinical (subjective) decision between the two operations [6-8]. The literature tells us we can successfully perform the operation, but it does not tell us who the candidates should be.

At the other end of the spectrum, we also do not know whether the 1.5 VR approach results in any benefit over that of the Fontan procedure. Although there are anecdotal case reports of performing 1.5 VR instead of Fontan procedure, no data is available to allow us to make a rational decision as to when the right heart characteristics (size? function?) reach the point at which the benefit is achieved in avoiding the Fontan operation [6-8].

Concerns

The technical aspects of 1.5 VR have previously been enumerated in detail by us as well as other investigators [37,38,49-57]. The safe upper limit of PAP and PVR for patients undergoing 1.5 VR is unknown. One concern regarding 1.5 VR circulation is its safety in the setting of elevated PVR. Raised PVR and/or competitive SVC flow in the setting of pulsatile flow from the right ventricle can lead to high SVC pressures, aneurysmal dilatation of SVC, prolonged pleural effusions and chylothorax, as seen in over 20% of the cases in the published literature [58,59].

The second concern is regarding the group of patients who have a

natural history of developing heart block e.g. corrected transposition of great arteries. The bi-directional Glenn denies access to the heart for pacing procedures – committing them to a series of epicardial pacing procedures [7].

The third concern is the presence of free pulmonary regurgitation. Although the presence of free pulmonary regurgitation could theoretically be helpful in increasing the right ventricular diastolic dimensions, it may lead to a backflow of blood from the superior caval vein to the pulmonary ventricle, thereby compromising the functional efficiency of an already abnormal pulmonary ventricle. Additionally, right ventricular preload is no longer reduced due to backflow of superior vena caval blood to the pulmonary ventricle, thus losing part of its physiologic rationale [9-38,49-59].

The fourth concern is the creation of atrial septal fenestration in cases when the tricuspid valve is between 1/3rd and ½ of normal and post cardiopulmonary bypass (CPB) right atrial pressure is >12mmHg. This approach is fraught with persistence of cyanosis late postoperatively and a definite risk of pulmonary embolism [9-38,49-59].

The fifth concern is the long-term fate of an unligated azygos vein with its dreaded consequences. Some investigators left the azygos vein unligated in all patients undergoing 1.5 ventricular repair because it acts as a safety vent in the event of SVC hypertension or severe right ventricular dysfunction [50,51]. Additionally it was thought to permit blood to flow to the upper compartment during exercise, when the hypoplastic abnormal RV is incapable of handling the increased venous return [50,51]. However, there have been reports of development of a semi-circular circulation in which blood is diverted down and recirculates back to the heart [60].

The sixth concern is the issue of concomitant pulmonary artery banding in the setting of 1.5 VR to prevent superior vena caval syndrome and retrograde flow from the superior caval vein to the right ventricle [37,38,46].

The seventh concern is patients requiring intricate intracardiac repair with prolonged cardiopulmonary bypass (CPB) time for dedicating the inferior vena caval blood alone to the RV resulting in unacceptably high perioperative complications and mortality [37,38].

And the final concern is that the 1.5 VR does not provide the same functional capacity as a true biventricular repair with limited capacity to increase cardiac output during exercise [9-38,49-52]. Although this is not universally proven, but may have contributed to the fact that nearly one-fifth of the patients are in NYHA III/IV at only 3.3 years after this procedure in the series published by Zhang and associates in 2017 [53].

Methods

With these deficiencies in mind, we have analyzed the published literature to identify the described cohort studies of one and one half ventricular repair with pulsatile bi-directional Glenn, atrial septal fenestration, issues of the unligated azygos vein, free pulmonary regurgitation (PR), pulsatile superior venacava (SVC), growth potential of the pulmonary ventricle, and long-term functional results.

The search engines employed were Medline, pubmed, Google scholar, Cochrane database and Embase. The search included literature in all languages. This strategy yielded 98 investigations that provided best answer to these topics. We have then synthesized all these features to outline the rationale, issue of concern and potential future trends of 1.5 VR [1-98].

This review article outlines the pathologic substrates who may be selected for 1.5 VR, the hemodynamic consequences of free PR and a pulsatile bi-directional Glenn. With respect to drawing conclusions from the sum total of the peer-reviewed published literature, based on the morphologic and functional criterions we have attempted to lay down the guidelines for candidate selection for 1.5 VR, biventricular repair and univentricular type of repairs.

Patient selection criterions

In an attempt to address these debatable issues, we have developed a conceptual framework in the context of concerning morphologic characteristics, pathologic substrates and size of the pulmonary/right ventricle. Symptomatic patients with increasing cyanosis or excessive pulmonary blood flow with a hypoplastic but potentially usable RV and those considered borderline for a biventricular repair are generally subjected to 1.5 VR protocol [37,38,54]. Patient selection for the 1.5 VR comes under consideration in specific clinical border zones namely,

a. Simplification of complex procedures [49-52].

b. To limit the risk from a right atrioventricular valve repair in cases of complex straddling and ventricular septation [9-38,49-52].

c. To prolong the conduit survival by reducing venous ventricular preload in patients undergoing Rastelli-type repairs [50,51].

d. To decrease preload to the RV, thus enabling a more aggressive tricuspid valve reconstruction in cases of Ebstein's anomaly [18-21].

The first category includes symptomatic patients with a hypoplastic but potentially usable RV who are borderline candidates for a two-ventricular repair but the surgeon "backs off" to 1.5 VR to avoid postoperative right heart failure. Examples might include the patient with pulmonary atresia and intact ventricular septum with a RV that is reasonably well developed but demonstrates concerning morphologic and/or physiologic characteristics [9].

The second category includes patients who have undergone previous neonatal palliative procedures followed by a later second-stage superior cavopulmonary anastomosis. A subset of these patients with favourable internal morphology might allow a surgical septation. Examples include the patient with a small RV, inlet VSD, and straddling tricuspid valve or the patient with transposition of the great vessels, conoventricular septal defect, small RV and tricuspid valve. The surgeon assesses the "difficulty factor" in achieving intracardiac septation, as well as the size and function of the small right heart, and then decides whether or not to proceed with 1.5 VR [6]. The implicit assumption herein is that avoiding the univentricular pathway is beneficial [6]. The Toronto group performed 1.5 VR with concomitant arterial switch in patients with D-transposition of the great vessels, VSD and a small RV with initial pulmonary artery band [34-36]. Banding was performed to allow reduction of pulmonary vascular resistance [46].

The third category has been in patients with complete transposition of the great arteries with a small left ventricle who underwent an initial atrial septostomy followed by an atrial switch and concomitant superior cavopulmonary anastomosis [34-36].

The fourth category includes patients with bilateral superior caval veins, atrial isomerism and single atrium with concomitant complex cardiac anomalies. A 1.5 VR can avoid a complex infra-atrial baffle, thus simplifying the operation [34-36].

The fifth category includes patients with corrected transposition of the great arteries with left ventricular outflow tract obstruction undergoing an Ilbawi procedure i.e. an atrial switch or Rastelli procedure. A superior cavopulmonary anastomosis in this clinical situation simplifies the operation by eliminating the superior limb of a Mustard operation [40,55,56].

The sixth category includes a select group of patients receiving a Rastelli procedure with a large intracardiac baffle. In addition to compromising a portion of right ventricular volume, the right ventricular function can also be impaired due to ventriculotomy [50,51].

The seventh category includes patients of atrial switch with systemic atrioventricular valve (tricuspid) regurgitation. A 1.5 VR is an attractive alternative for "pressure unloading" of the pulmonary ventricle [30,31]

The eighth category includes patients with corrected transposition of the great arteries with or without a VSD with left ventricular outflow tract obstruction. A 1.5 VR has been performed avoiding an extracardiac left ventricle-to-pulmonary artery conduit or a double switch operation. The native outflow tract to the pulmonary artery was left intact with the presumption that it would lessen the septal shift into the pulmonary ventricle [61-63].

The ninth category includes patients with ventricular septal defect and straddling right atrioventricular valve. Reimplantation of the chordae, valve repair and a superior cavopulmonary anastomosis allows a 1.5 VR to be performed [61].

The tenth category includes patients with Ebstein's anomaly with a small RV, functionally compromised or both (Carpentier class C and D). A 1.5 VR is an acceptable alternative in this subset of patients as it decreases preload to the RV, thus improving the function of the RV and tricuspid valve and enables a more aggressive tricuspid valve reconstruction [18-21,39]. If the right atrial pressure is >12 mmHg or thrice the left atrial pressure after termination of CPB or the tricuspid valve function is inadequate on intraoperative transesophageal echocardiography, addition of a bidirectional Glenn has a favourable impact on arrhythmias and ventricular function [37,38,50,51].

The eleventh category includes patients with an isolated hypoplastic and/or a dysplastic RV. A 1.5 VR reduces the surgical risk for biventricular repair and maintains low right atrial pressure than that of the Fontan [32,64].

The twelfth category is in patients with hypoplastic RV with recurrent untreatable arrhythmias after a Bjork-Fontan procedure in infancy undergoing successful conversion to 1.5 VR [65].

The thirteenth category has been patients with cardiac Ewings sarcoma undergoing tumor debulking with concomitant superior cavopulmonary anastomosis and patients undergoing resection of a metastatic tumour to the RV along with lobectomy for lung cancer [66,67].

The fourteenth category has been patients with Uhl's anomaly and right ventricular endomyocardial fibrosis being subjected to 1.5 ventricular repair protocol [19,29].

The fifteenth category includes those patients requiring conversion to 1.5 VR for acute postoperative right ventricular dysfunction following attempted biventricular repair: "the salvage 1.5 VR". These patients usually have a borderline, dysfunctional and moderately dilated RV [37,38].

Determinants of the ability of the pulmonary ventricle for one and one half ventricular circulatory physiology

There is controversy about what constitutes an appropriate right ventricular size or function with this approach, with different institutions following different protocols [14,15,34-38,65-68].

1. Pulmonary ventricular size

Assessment for a one and one-half ventricle repair: Assessment of pulmonary / right ventricular hypoplasia is usually based on the classification of Milliken and colleagues [69]. According to this classification, the group with severe hypoplasia of the right ventricle is characterized by a tricuspid valve, right ventricular cavity and right ventricular outflow tract (RVOT) that are one-third of normal size or smaller. In patients with moderate right ventricular hypoplasia, the tricuspid valve and the right ventricular cavity size are approximately one to two thirds of normal without an adequate RVOT. In patients with mild right ventricular hypoplasia, the tricuspid valve and right ventricular cavity size are approximately two-thirds of normal, and there is a well-developed RVOT [69].

Tricuspid valve assessment

a) Preoperative echocardiographic assessment: The tricuspid valve can be assessed by preoperative echocardiography. The largest diameter of the tricuspid valve obtained from an apical four-chamber view is indexed to body surface area and compared with the normal values described by King and associates [70-73]. The data for the mean normal diameters and their standard deviations are obtained from the table provided by Rowlatt and colleagues and online resources are used to compute the Z-value of the tricuspid valve from the website (parameterz.blogspot.com) [74,75].

b) Magnetic resonance imaging studies: Multisection gradient-echocardiographic cine-MRI is probably more accurate because it is noninvasive and unaffected by acoustic windows, and it measures ventricular dimensions and functions independently. However, it may overestimate pulmonary regurgitant fraction in patients with combined pulmonary and tricuspid regurgitation. Therefore, we used velocity-encoded cine-MRI for accurate quantification of pulmonary regurgitation [76-84].

c) Intraoperative assessment: Intraoperative transesophageal echocardiography (TEE) gives an accurate estimate of the right ventricular cavity size, function and diameter of the tricuspid valve. The components of the RV are visualized externally on operation table and the tricuspid valve diameter is measured using Hegar's dilators. Indexed tricuspid annular diameter is compared with the published

autopsy data of Rowlatt and colleagues [74]. The "normal" valve sizes published by Rowlatt and colleagues based on autopsy analysis are somewhat smaller than the valve sizes reported by de Leval and collagues [54]. However, the Z values obtained preoperatively by echocardiography in our previous investigations were almost the same as those obtained intraoperatively [37,38].

In many, but not in all patients, the tricuspid annulus directly corresponds with right ventricular volume. A simple method of quantitating the ability of the RV to handle volume is to measure the tricuspid annulus size and plot this measurement on a nomogram to obtain a z value [85]. A z value of 0 indicates a normal size for a given body surface area. Each decrement above or below 0 represents 1 standard deviation away from normal [34-38].

d) Criterions for candidate selection for one and one-half ventricle repair (based on pulmonary ventricular size): Criterions for candidate selection for one and one-half ventricle repair are largely based on cohort reports available in the published literature. Canine animal modeling by Ilbawi and associates has demonstrated a continued favorable hemodynamic response in the pulmonary vasculature when the RV is downsized in the acute setting to as small as 30% of normal [55,56]. This same group has successfully performed 1.5ventricle repairs in the clinical setting with predicted ventricular volumes as small as 30%.

i) Hospital for sick children, Toronto: Von Arsdell and colleagues used a combination of tricuspid valve Z-scores and pulmonary ventricular volume to select patients for a biventricular repair, 1.5 VR or an univentricular type of repairs (Table 1) [34-36]. The smallest RV dimensions that will allow forward passage of the entire systemic venous return without causing excessive right atrial-right ventricular hypertension or low cardiac output postoperatively may vary from patient to patient [8,12,34-38]. Available angiographic limits for the lowest normal right ventricular end diastolic volume have a very wide range (1 SD=86%, 2 SD=68% of predicted body surface area) [85]. It is doubtful that such numerical criterions reliably separate a small normal from a truly hypoplastic RV [68].

ii) All India Institute of Medical Sciences, New Delhi: The selection criterions that were proposed by the corresponding author and was adopted by our group are as follows (Table 2). Patients with moderate or severe RV hypoplasia were considered for 1.5VR. A satisfactory pulmonary artery (PA) size (Z value >(—) 2, McGoon's ratio >2.0, or Nakata index >250 mm/m2) were considered mandatory [37,38,86-91].

The following measurements- tricuspid valve Z-score lower than -4.8, tricuspid valve diameter less than 45% of normal, tricuspid valve to mitral valve ratio of less than 0.54, and right ventricular diastolic volumes less than 45% of predicted normal- were considered as the cutoff points from one-ventricle repair. These values were taken as the lower limits of tricuspid valve size acceptable for inferior vena caval blood alone.

Table 1: Anatomic selection criterionsfor small pulmonary ventricles and a one and one half ventricle repair [36].

Tricuspid z values	Pulmonary ventricular volume	Operation
> -2	>80%	Biventricular repair
< -2	<80%	1½ ventricle repair with cavopulmonary anastomosis
< -2	<50%	1½ ventricle repair, cavopulmonary anastomosis, atrial fenestration, possible delayed fenestration closure
< -10	<30%	Fontan procedure

Table 2: Degree of right ventricle hypoplasia and surgical management of patients [38].

S.No.	Variables / surgical management	RV hypoplasia		
		Mild	Moderate	Severe
1	Tricuspid Z value	≥1.5	-1.5 to -4.8	≤4.8
2	Tricuspid valve diameter (% of normal)	70.5%	45%-70.5%	<45%
3	Tricuspid valve diameter/ mitral valve diameter	0.80	0.54-0.80	<0.54
4	RV diastolic volumes (% of predicted normal)	75%	45%-75%	<45%
5	RV morphology	Tripartite	Bipartite	Unipartite
6	RV cavity and outflow tract	Approximately 2/3 of normal, well-developed RVOT	1/3 to 2/3 normal, RVOT inadequate	<1/3 of normal size, absent RVOT
7.	Definitive operation	Two-ventricle repair	One and one-half ventricular repair	Univentricular repair

RV= Right ventricular, RVOT= Right ventricular outflow tract.

The following measurements- tricuspid valve Z-score greater than -1.5, tricuspid valve diameter 70.5% of normal, tricuspid valve to mitral valve ratio of 0.80 and right ventricular diastolic volumes greater than 75% of predicted normal were considered as the cutoff points from two-ventricle repair.

We limited the use of 1.5 VR to patients with a tricuspid valve Z-score between -1.5 and -4.8 and a tricuspid valve diameter between 45% and 70% of normal. Other authors have expanded the tricuspid disease score to -5 or -6 on a few patients. However, the mean Z scores in those series are similar to the mean Z score in this study, and the reported number of patients is small indeed [42-94]. We would perform a univentricular repair for this group of patients with these extremely low tricuspid valve diameters [37-38].

The morphologic defect may be derived by tricuspid valve Z value which also reflects the corresponding ventricular volume [86]. Some patients have a normal tricuspid Z-value and yet an anatomically small or functionally abnormal RV [55,56]. Therefore, a combination of parameters namely, tricuspid Z-values, tricuspid/mitral valve diameter, RV morphology and RV diastolic volumes are taken into consideration for the decision-making of 1.5 VR, biventricular or an univentricular type of repair [9-46,55,56,86-94].

In the Congenital Heart Surgeons' Society multi-institutional study, the predicted prevalence of biventricular repair at 5 years was less than 30% for a Z value of -3 and less than 50% for a Z value of -2. The authors concluded that patients with a tricuspid valve diameter in this range may be the most suitable candidates for 1.5 VR [88,89].

We agree with Muster and colleagues that absolute sizes of the RV and tricuspid valve Z scores may not be the only factors for operative

success. Right ventricular compliance, tricuspid regurgitation, right ventricular outflow tract, relative pulmonary artery hypoplasia, and pulmonary vascular resistance are important additional considerations in preoperative candidate selection [37,38,67,71,72,86-92].

2. Functional criterions for one and one half ventricle repair

Evaluation of the pulmonary ventricle's ability to handle an appropriate indexed cardiac output is primarily an assessment of both ventricular size and function. These evaluations are somewhat subjective and not completely accurate. However, some guidelines do exist.

Functional analysis of the RV is performed by echocardiographic and/or angiographic measurements and graded as normal or mildly/moderately/severely decreased [37,38]. Visual inspection of ventricular wall thickness and diastolic filling is also made. Thickened ventricles may have good systolic function, but poor filling, dilated ventricles may have good filling, but poor function [37,38]. Patients with poorly functioning RV are considered for 1.5 ventricle repairs when there is dilatation to 120% or more and at least moderate reduction in ventricular function [76-84].

Recently, magnetic resonance imaging (MRI) has been used to quantitate right ventricular systolic and diastolic function as well as volume. This modality may prove useful for future preoperative evaluation of candidates for a 1.5 VR [34-36,76-84].

3. Physiologic criterions based on pulmonary after load

A third important determinant for right ventricular ability to handle volume load is the pulmonary after load. The safe upper limit of PAP and PVR for 1.5 VR is unknown. It seems conceivable that the physiologic criterions for a 1.5 VR may be less stringent than those required for a single ventricle approach. The dismal results of performing 1.5 VR in the setting of high PAP and PVR in our previous investigation suggest that this hypothesis may not be true. One half of the deaths in our previous investigations were due to elevated PVR and the physiological criterions appeared similar to those for Fontan candidates. Therefore, patients with elevated PVR are not candidates for a 1.5 VR unless there is a reparable anatomic cause such as branch pulmonary artery stenosis or left heart obstruction [37,38].

The following physiologic criterions were taken into consideration in our institution for 1.5 VR.

1. Mean PAP less than 15 mm Hg, or less than 20 mm Hg with a net left-to-right shunt.
2. Indexed PVR less than 3.0 Woods units/m^2, and a preoperative Mayo indexless than 4.0.
3. Systemic ventricle end-diastolic pressure less than 12 mm Hg, and ejection fraction greater than 0.45
4. Patients with probability of residual pulmonary arterial hypertension, i.e., raised PVR, multiple ventricular septal defects, and so forth, were contraindications and two-ventricle repair with atrial pop off was considered a better option [37,38].

Specific issues concerning one and one half ventricular repair (Table 3)

Atrial septal fenestration: Atrial septal fenestration in patients undergoing 1.5 VR remains controversial [1,6-10,32,34-38,50,51]. In their experience with upto 38 years of follow-up, the Toronto group has shown that by 10 years of follow-up, only 7.5% of the atrial septal defects remained open and in the remaining they closed spontaneously as the right-sided pressures came down [64]. However, this approach is fraught with the persistence of chronic cyanosis and its complications and a small, but definite risk of paradoxical embolism.

Forty-four of 84 patients in this study (n=10, before 2000 publication; and n=34, after 2000) had concomitant atrial septal fenestration. Four patients in our earlier experience who died early postoperatively of low cardiac output had high right atrial pressure and would probably have benefited from an elective fenestration. Presently, we perform atrial septal fenestration in patients with tricuspid valve between one-third and one-half of normal. With this strategy, the clinical outcome has improved, and no long-term survivors developed pleural effusion, as cites, or protein-losing enteropathy. No patient with fenestration had significant desaturation or a cerebrovascular accident [37,38]. The incidence of perioperative and late supraventricular arrhythmias were 14.3% and 15.9% respectively [37,38].

Presently, if the postoperative right atrial pressure is >12 mmHg, we perform an adjustable inter-atrial fenestration to help balance the pulmonary and systemic circulations to modulate the rise in central venous pressure in the early postoperative period. This will act as a safety vent in the event of high right atrial pressure or a failing 1.5 VR [37,38].

Pulmonary regurgitation: Free pulmonary regurgitation may lead to backflow of blood from the superior vena cava (SVC) to an already abnormal RV which may lead to further reduction in the functional efficiency of an already abnormal RV. Thus, in the presence of free pulmonary regurgitation and regurgitation of the superior vena caval blood back to the pulmonary ventricle, the 1.5 VR loses part of its physiologic rationale, therefore right ventricular preload is no longer reduced. We demonstrated in our previous investigation that the presence of free pulmonary regurgitation could theoretically be helpful in increasing RV diastolic dimensions. Thus, the beneficial vs. harmful effects of 1.5 VR with respect to pulmonary regurgitation is debatable [37,38].

In the settings of corrected transposition of the great arteries with or without a VSD and left ventricular outflow tract obstruction or pulmonary stenosis, and Ebstein's anomaly with right ventricular outflow tract obstruction, Mavroudis and associates have performed intracardiac repair with concomitant superior cavopulmonary anastomosis leaving behind an intact pulmonary outflow tract. The native intact outflow tract retains some degree of outflow obstruction, which may be beneficial in that it would lessen the septal shift towards the pulmonary ventricular side. In an analogous situation in late follow-up of the atrial switch operation, septal shift has been associated with systemic atrioventricular valve (tricuspid) regurgitation. Thus, a 1.5 VR is attractive for "pressure unloading" the pulmonary ventricle [62,63].

Azygos vein- to ligate or not to ligate?: In 1999, Kreutzer and associates recommended a patent azygos vein in all patients undergoing 1.5 VR with the aim of decompressing the SVC or the right atrium in patients with superior vena caval hypertension or severe right ventricular dysfunction, respectively, without oximetric impairment [50,51]. Additionally, it was thought to permit blood to

Table 3: Summary of the published investigations documenting the mid-term and long-term results of one and one half ventricle repair with pulsatile bidirectional superior cavopulmonary anastomosis.

S. No.	Author	Year of publication	No. of patients	Age Mean±SD (range)	Follow-up Mean±SD (range)	Early and late mortality	Results	Recommendations
1.	Chowdhury UK et al[37]	2001	50	3.44±6.15 years (4 months=42 years) <4 years (n=42)	98.6±6 months (8-116 months)	6 (12%) 2 (4.5%)	At 97 months actuarial survival 74% NYHA class I: 88%	Moderate right heart hypoplasia is a safe anatomic category for 1.5 VR. Patients with any probability of postoperative pulmonary artery hypertension, raised pulmonary vascular resistance, relative contraindication for 1.5 VR
2.	Chowdhury UK et al[38]	2005	84 (50 previous series, 34 present series)	47.9±57.3 months (4-504 months) <4 years (n=62)	87.7±56.6 months (12-178 months)	Hospital death (8.8%) Late death (8%)	Actuarial survival 81.9%±0.04% NYHA I/II: 89.8% (n=62)	Physiologic criterions similar to Fontan candidates. 1.5 VR reverses the Fontan paradox; no consequences of pulsatile Glenn. 45% of survivors demonstrated significant growth in tricuspid valve, right ventricle on MRI. None qualified for biventricular repair.
3.	Kreutzer C et al[51]	1999	30	6.7±8.5 years (4 months-40 years)	1-10 years	Hospital death 2 (6.6%) Late death 1 (3.3%)	Actuarial survival at 5 years (90%) NYHA I: 77% (n=21)	Early and intermediate term follow-up results compare favourably with those of the Fontan procedure.
4.	Kim et al[64]	2009	114	3 months-59 years	1 month-38 years	Hospital death (6.1%) Late death (20.2%)	Actuarial survival at 5, 10 and 20 years (83.4%, 80.1% and 69.3%) NYHA I/II: 98.8%	The most common cause of late death was cardiac. The risk of serious complications related to pulsatile superior vena cava was 2.6%.
5.	Lee Yo et al[52]	2011	29	Median 26 months (6 months-26 years)	85.6±44.6 (3 months-190 months) Median: 24 months	Hospital death 4 (13.8%) Late death – nil	Freedom from late reoperation were 80.0±12.6% in group A and 51.4±20.4% in group B NYHA class I (n=21)	Good NYHA without late complications of Fontan pathway, 1.5 VR is a valid alternative to Fontan and biventricular repairs
6.	Numata S et al[49]	2003	13	4±3 years (10 months-9 years)	10±4 years (3-15 years)	Hospital death – nil Late death 1 (7.7%)	Actuarial survival at 10 years (88.9%). Freedom from arrhythmia was 80% and 20% at 10 and 12 years respectively. Exercise testing showed anaerobic threshold values equivalent to those of Fontan and inferior to those who could have undergone biventricular repair	Functional results not hopeful in long-term. Conversion to total cavopulmonary connection may be needed in some patients.
7.	Stellin G et al[32]	2002	8	9.1 years (7 months-35 years)	29.8 months (8 months-7.3 years)	Hospital death – nil Late death – nil	NYHA I (n=8)	1.5 VR reduces the surgical risk of biventricular repair compared to Fontan circulation, reverses the Fontan paradox and improves the systemic arterial oxygen saturation

8.	Arsdell V et al[35]	1996	38	Median 3.5 years (5 months-51 years)	46.3±36.9 months (1-174 months)	Hospital death – nil Late death – nil	No protein losing enteropathy NYHA I (n=22), II (n=8)	Acceptable intermediate term outcomes. Results as a salvage operation unsatisfactory.
9.	Zhang S et al[53]	2017	31	Median 5.4 years (0.75-12 years)	3.3±2.1 years	Hospital death – nil Late death – 1 (3.2%)	NYHA I 83.3% (n=25) NYHA III/IV, n=6 (19.3%) No conduit change	1.5 VR technically feasible in CCTGA, LVOTO, Cardiac malposition
10.	Malhotra SP et al[40]	2011	48	3 years (3.9 months-24 years)	59.2 months (1 month-15 years)	Hospital death – 1 (2%) Late death – Nil	Freedom from reintervention, 1, 5, 10 years (91.5%, 76.8%, 71.8% respectively) 11 (23%) patients required 10 reoperations, 3 interventions	Right ventricle-pulmonary artery conduit longevity improved Prolonged conduit life, reduced baffle / sinus node related complications

CCTGA= Corrected transposition of the great arteries; LVOTO= Left ventricular outflow tract obstruction; 1.5 VR= one and one half ventricular repair; NYHA=New York Heart Association; MRI=Magnetic resonance imaging.

flow to the upper compartment during exercise, when the hypoplastic right heart is incapable of handling the increased systemic venous return [47]. However, Uribe and associates in 2012, using four dimensional flow MRI have demonstrated the development of a semicircular circulation in which blood is diverted down and recirculates back to the heart [60].

In our previous investigations on 84 consecutive patients undergoing 1.5 VR between 1990 and 2003, we had ligated azygos vein in all patients with no adverse hemodynamic consequences. We noted a pressure difference greater than 6 mmHg between the superior vena cava and the right atrium [37,38]. Pressure gradients between the systemic veins could produce collaterals through venovenous shunts [9-46]. No venovenous collaterals or pulmonary arteriovenous malformations had been identified on long-term follow-up. Although we routinely ligate the azygos vein, the issue is debatable and will be apparent on late follow-up. The postoperative hemodynamic data thus compared favorably with the mean SVC and IVC pressure (14 mm Hg) reported after a Fontan operation, and 1.5VR physiology appeared superior to Fontan physiology [37,38].

Pulmonary artery banding: Right pulmonary artery banding has been advocated by Gentles and associates to handle excessive pulsatility in the SVC and to avoid aneurysmal SVC [46]. Retrograde flow from the SVC into the RV in the setting of free pulmonary regurgitation, increases the amount of blood being ejected forward from the RV into the SVC. Banding of the right pulmonary artery at its junction with main pulmonary artery prevents the RV from ejecting into the SVC and allows the SVC blood to flow into right pulmonary artery, thereby creating a hybrid type of pulmonary circulation [37,38,46,50,51,59]. However, some investigators including ourselves have not performed right pulmonary artery banding [37,38,52,59]. In recent years subsequent to our previous reports, seven patients underwent right pulmonary artery banding to decrease the high SVC pressure in operation table with satisfactory results (unpublished).

Growth of the pulmonary chamber, effects of free pulmonary regurgitation on dimensions of the pulmonary ventricle

The impact of pulmonary insufficiency in the setting of 1.5 VR is unclear. In our experience, patients with mild pulmonary regurgitation seem to have a smooth postoperative course [37,38,92-97].

Only 45% of survivors in our previous study demonstrated significant growth of the tricuspid valve and RV. These patients in our previous study were borderline candidates for a biventricular repair and had tripartite RV. However, none have qualified for conversion to biventricular repair so far. The insignificant growth of the tricuspid valve annulus and right ventricle in the great majority is probably because of the existence of dysplastic tricuspid valve and RV. However, long-term effects will be apparent only on longer follow-up. Serial cine-MRI evaluation may be of particular importance in this regard [37,38].

Our study highlighted the importance of quantification of pulmonary regurgitation and quantitative evaluation of right ventricular and left ventricular dimension and function with echocardiography, MRI, or radionuclide studies. Varying ventricular morphologies in the univentricular cohort make the calculation of ejection fraction by conventional angiocardiograms unreliable. Gated blood pool scintigraphy is relatively independent of geometric assumptions, but spatial resolution is poor in small children, and the atrial blood pool cannot be clearly separated from the ventricular region [37,38,76-87].

Multisection gradient-echocardiographic cine-MRI is probably more accurate because it is noninvasive and unaffected by acoustic windows, and it measures ventricular dimensions and functions independently. However, it may overestimate pulmonary regurgitant fraction in patients with combined pulmonary and tricuspid regurgitation. Therefore, we used velocity-encodedcine-MRI for accurate quantification of pulmonary regurgitation [76-87].

Failure of 1.5 ventricular repair and conversion to a univentricular pathway

Conversion from the 1.5 VR circulation to total cavopulmonary connection for a failing 1.5 VR have been reported by some investigators [49,62,63]. Out of 13 patients undergoing the 1.5VR of Numata and associates, conversion to total cavopulmonary connection

with cryoablation of the atrial wall was needed in two patients because of elevated right atrial pressure and atrial arrhythmias subsequent to 1.5VR. They postulated that this conversion to the Fontan operation has the potential to reduce the atrial arrhythmias and reduce the severe congestion of the lower body. However, the latter could still persist after conversion to Fontan circulation [49]. Van Arsdell and associates have reported ASD creation in a patient with high right atrial pressure and low cardiac output syndrome [34-36].

Fontan conversion to one and one half ventricle repair

In 2012, Sharma and colleagues from Mayo Clinic have reported 10 patients undergoing Fontan conversion to 1.5 VR.98Sharma, Hoashi and other investigators concluded that Fontan conversion to one and one half ventricle repair is feasible in selected patients with a failing Fontan circulation with low early mortality. They recommended concomitant arrhythmia surgery as a routine [65,95,98].

Padalino and colleagues have shown that in the setting of a hypoplastic RV and with recurrent untreatable arrhythmias after a Bjork Fontan procedure in infancy, conversion to 1.5VR is an alternative to the Fontan revision [65]. It appears that carefully selected patients initially treated with intent to perform a single ventricle palliation can in some instances be converted to 1.5 ventricle physiology with acceptable outcomes.

Salvage one and one half ventricular repair

Between 1990 and 2003, we performed salvage 1.5 VR on 5 patients to treat acute RV dysfunction following a failed biventricular repair with dismal results. The salvage procedure was performed after 24 hours with 60% operative mortality (n=3) [37,38]. Subsequently, between 2003 and 2017, we have performed 7 more salvage 1.5 VR for a failing biventricular repair on operation table in the same sitting before coming off bypass without any mortality. It appears desirable to plan salvage 1.5 VR on operation table in cases of failing biventricular repair to obtain a satisfactory outcome.

One and one half ventricular repair: Can it restore ventriculo-arterial coupling?

Optimal hemodynamics in normal circulation consists of higher pulmonary artery pressure (mean approximately 15 mmHg) and a lower caval pressure (mean <10 mmHg) to keep the pulmonary vasculature patent. The paradox of the Fontan circulation is that it produces caval and pulmonary artery hypertension [43-46]. Postoperative cardiac catheterization in our previous investigations have revealed a step down in pressure of 5 mmHg in the inferior vena cava and a step up in pressure of 5 mmHg in the pulmonary arteries, thus reversing the "Fontan paradox" and exerting all the potential benefits of a pulsatile circulation in the pulmonary circuit [37,38].

Exploring the Unknowns: Future Directions

Published literature available till date tells us that 1.5 VR can be performed relatively safely and that these patients seem to do relatively well at mid- and long-term follow-up. The literature tells us we can successfully perform the operation but it does not tell us who the candidates should be.

There are anecdotal cases in which attempted biventricular repairs failed decisively in the operating room after weaning from CPB or shortly thereafter and the patients were managed quite effectively by converting them into 1.5 VR. When the 1.5 VR is chosen over the Fontan procedure, even the anecdotal evidence is lacking that the patient benefits. Future publications auditing the above-mentioned issues from retrospective case series of patients undergoing 1.5 VR will add nothing to our understanding of the problems of candidate selection for 1.5 VR [6].

We need to know whether performing 1.5 VR is ever the right thing to do, both when the alternative option is the Fontan procedure and when it is the biventricular repair. We need to define the physiologic and morphologic criterions that can be used in deciding on the optimal procedure for a given patient.

To properly test the hypothesis that 1.5 VR results in superior outcome in selected patients than biventricular repair or a univentricular type of repair, a multi-institutional prospective randomized trial would be necessary and would be the last refuge for those individuals with right hearts that have concerning morphologic and/or physiologic characteristics. Such information will be welcome and noteworthy.

Recommendations

On the basis of the published literature including ours enunciated in the manuscript, we would recommend the following guidelines for candidate selection for 1.5 VR.

1. Patients with moderate right heart hypoplasia will in general have a successful operative outcome after 1.5 VR.
2. Severe right heart hypoplasia with TV Z-score lower than -4.8 and mild right heart hypoplasia with TV Z-score higher than -1.5 would predicate a univentricular type or a biventricular repair respectively.
3. Patients with any probability of residual PAH, i.e. raised PVR, residual VSD and so forth may be considered as having a relative contraindication to 1.5 VR. A biventricular repair with atrial septal fenestration is probably a better option.
4. Patients requiring an intricate intracardiac repair with prolonged cardiopulmonary bypass time for dedicating inferior vena caval blood alone to the RV may be considered a relative contraindication to 1.5 VR in favor of a simpler Fontan repair. Examples include- a) inlet VSD with a straddling tricuspid valve, b) DORV with non-committed VSD not amenable to surgical septation, c) dTGA, conoventricular septal defect with small RV and d) those requiring a Rastelli type of repair with a large intracardiac baffle.
5. Patients with pulmonary atresia and severe right heart hypoplasia or RV dependent coronary circulation should not be subjected to a 1.5 VR protocol. A univentricular-type of repair is a safer option.
6. Atrial septal fenestration should be performed in all patients with severe right heart hypoplasia and in patients with post bypass right atrial pressure of >12mmHg.

Conclusions

Based on the literature and reasoning cited above we conclude that a 1.5 VR can be performed with an acceptable risk provided the anatomy permits and the physiologic criterions appear similar to

Fontan candidates. The operation maintains a low pressure in the IVC compartment and reverses the Fontan paradox with no consequences of the "pulsatile Glenn". Results are unsatisfactory when used in the setting of elevated PVR, and when used as a salvage procedure to treat acute postoperative right ventricular dysfunction. Majority of the published series document favourable late survival following 1.5 VR repair and good functional status without late complications of the Fontan procedure. This procedure appears to be a valid alternative to Fontan and biventricular repairs in selected patients with right ventricular dysfunction or dysplasia.

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