18 Can Surgeons Preserve Right Ventricular Function in Hypoplastic Left Heart Syndrome?

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Abstract

The systemic right ventricle in hypoplastic left heart syndrome commonly shows gradually declining function after initial palliation. It is therefore crucial to optimally preserve myocardial performance. Decisions in the operating room can have long term impact on systemic right ventricular function. For example, the choice of surgical procedures, shunt types, myocardial protection strategies, techniques used to reconstruct the hypoplastic aorta, and tricuspid valve procedures may all play an important role in the preservation of myocardial function. We herein discuss assets and drawbacks of surgical decisions with regard to right ventricular function in hypoplastic left heart syndrome, summarize currently available data, and give a perspective on contemporary and future therapy.

Keywords

HLHS • Systemic right ventricle • Right ventricular function • Norwood operation • Arch obstruction • Recoarctation • Tricuspid valve • Blalock-taussig shunt • Right ventricle to pulmonary artery conduit • Hybrid procedure • Stage II palliation • Fontan completion

Introduction

"It is as if the heart were constituted for the sake of the left ventricle, with some thin heart substance attached to the right side of the septum in order to make the right ventricle" [1]. It was Andreas Vesalius' unprecedented work "De humani corporis fabrica" in 1543 that paved the way for modern cardiac surgery. Exceptionally detailed and accurate, he described distinct anatomic features of both ventricles, and even

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M.K. Friedberg, A.N. Redington (eds.), *Right Ventricular Physiology, Adaptation and Failure in Congenital and Acquired Heart Disease*, https://doi.org/10.1007/978-3-319-67096-6_18

delineated the right ventricle's (RV) close relationship to the lung. For centuries the RV was regarded as less important or even neglectable. 440 years after Vesalius' groundbreaking publication, Norwood et al. laid the foundation of palliation in hypoplastic left heart syndrome (HLHS), bringing the RV back into surgery's focus [2].

The RV is an extraordinary and complex structure. From a developmental standpoint, the RV's crucial role manifests in its function as the systemic ventricle during normal fetal circulation. In HLHS the fetal RV is not only fundamentally different from its left-sided counterpart but also significantly different from the normal RV, as it shows signs of antenatal diastolic dysfunction, lower cardiac output and altered contraction pattern [3–5].

RV dysfunction is a common but poorly understood problem after initial palliation of HLHS. Although some studies failed to link preoperative RV function with early mortality [6–9], parameters that are surrogates of RV performance (e.g. atrioventricular valve regurgitation, postoperative shock, and extracorporeal support) have been clearly identified as risk factors for in-hospital and interstage mortality [9–11]. Acute postoperative failure aside, functional deterioration can occur gradually, late after initial palliation, with a considerable delay and arises from a multitude of causes [12, 13]. Since ventricular function is not only a reflection of myocardial contractility but also the result of preload, afterload, ventricular macroscopic structure, myofiber architecture, valve function, heart rhythm and synchrony, as well as ventricular interdependence and many dynamic processes [14–16], the surgeon's approach in preserving RV function in HLHS has to take all these factors into account. Nevertheless, the surgeon has a limited set of variables, which can be manipulated to prevent RV dysfunction. These variables include the selection of the initial palliative strategy, technique of myocardial protection, technique of aortic reconstruction, and timing and type of tricuspid valve (TV) interventions.

Choice of Surgical Strategy

Obviously, the surgical strategy is one of the key points in single ventricle palliation as it provides the basis for the subsequent course. Many different options are on hand for each palliative step, ranging from timing of procedures to technical surgical aspects.

Stage I

If the patient's condition can be stabilized after birth, timing of stage I surgical palliation in HLHS aims at postpartum maturation and adaptation to extrauterine life on the one hand and avoiding pulmonary overflow or systemic malperfusion on the other hand. Older age has been identified as a risk factor for mortality [17, 18]. Analysis of the Single Ventricle Reconstruction trial did not show an association of age at the Norwood procedure with interstage mortality [9]. At least in the short term, right ventricular function is not necessarily affected by age at stage I [17].

Strategic aspects of stage I comprise the use of a modified Blalock-Taussig (BT) shunt, the implantation of an RV to pulmonary artery (RVPA) conduit, or the placement of an arterial duct stent as part of a hybrid approach.

Most of the studies addressing the type of shunt have a retrospective design and small patient numbers. In contrast, the Single Ventricle Reconstruction Trial is a prospective, randomized, multicenter trial comparing BT-Shunt palliation with RVPA conduit placement in HLHS, and gives valuable insights into the effect on right ventricular function. The ejection fraction after the initial procedure was significantly greater with an RVPA conduit, but insignificantly different at 14 months. RV volumes were lower in the RVPA group before stage II palliation, but again showed no difference at 14 months. Tricuspid valve regurgitation as an indirect measure of RV performance demonstrated no difference after 14 months. In a separate analysis focusing on echocardiographic parameters of ventricular function, RV ejection fraction, percent area

change and myocardial performance index after the Norwood procedure were all significantly higher with an RVPA conduit [19]. Again, no differences in functional ventricular indices could be shown. Indirect parameters of RV function such as TV size and degree of regurgitation were not influenced by shunt type. Follow-up data after 3 years, showed significantly lower pre-Fontan RV ejection fraction in the RVPA conduit group [20]. Furthermore, RVPA conduit patients showed a significant decline in ventricular function from 14 months to the pre-Fontan assessment [21]. Global RV function remained stable irrespective of initial shunt type. This analysis may be confounded by the measurement technique which does not include the right ventricular outflow tract (RVOT). The RVOT could be adversely affected by the RV incision used for placement of the RVPA conduit. Possibly, the deterioration of RV ejection fraction suggests impaired RV remodeling in patients treated with an RVPA conduit. It remains important to see if this decrease is ongoing or plateaus during follow-up [22].

RVPA conduits are considered beneficial for patients at high risk for a Norwood procedure, such as preterm and low-birth-weight patients. Surprisingly, no beneficial effect on survival could be found in this patient group during the interstage period in the SVR trial [9]. Few pathological studies have addressed the impact of the RV incision. The ensuing myocardial changes have been studied on specimens after palliation with either BT shunt or RVPA conduit [23]. Differences in remote myocardial scarring indicating coronary perfusion deficits could not be detected. However, myocardial thinning and scarring was noted at the site of the ventriculotomy. This correlated to previously assessed echocardiography, that detected signs of decreased segmental myocardial deformation at the ventriculotomy and increased deformation at the opposing site in the RVPA group, as a sign of mechanical dyssynchrony [23, 24]. In turn, mechanical dysynchrony has been associated with decreased diminished ventricular function in HLHS.

Several studies highlight the aspects of stage I palliation with regard to hybrid palliation with bilateral pulmonary artery banding and ductal stenting. The advantage of the latter is due to the postponement of major surgery with the use of cardiopulmonary bypass, cardiac and circulatory arrest from the neonatal period to several months of age. In a retrospective analysis of patients at the Hospital for Sick Children, BT shunt, RVPA conduit and hybrid palliation were compared and analyzed beyond stage II palliation [25]. Although diminished ventricular function was associated with mortality, none of the palliative approaches were associated with superior RV function or atrioventricular valve function 3 and 6 months after initial palliation. Other comparisons of hybrid palliation and classic Norwood operations with implantation of a BT shunt could not identify any difference in ventricular function between groups [26, 27]. Therefore, available clinical data shows neither a beneficial nor detrimental effect of hybrid palliation on RV function. Mathematical modeling as a means of theoretically understanding hemodynamic effects in hybrid palliation compared to BT shunt, and RVPA conduit revealed negative effects such as lower RV ejection fraction, lower mechanical efficiency, higher stroke work and higher afterload with reduced coronary and cerebral perfusion in patients after hybrid palliation [28].

In conclusion, a BT shunt seems to have a favorable (or the least detrimental effect) of the three palliative strategies on RV function—with the caveat that long-term comparative data for hybrid palliation is not yet available.

Stage II

The palliated circulation established with the Norwood operation leads to a gradual increase in RV volumes and a decrease in RV function before stage II palliation [29, 30]. After the Norwood procedure the RV has to generate up to 280% of normal cardiac output [31]. After completion of stage II palliation significant decreases in RV volumes were found [30]. Therefore, unloading of the RV at stage II is considered beneficial for preservation of RV function [32, 33].

With regard to timing of stage II, earlier operation offers earlier volume unloading of the systemic ventricle. Available data gives no conclusive statement on the best point in time to proceed to stage II. Although it is feasible to create a cavopulmonary anastomosis as early as 8 weeks, patients may show signs of limited pulmonary blood flow irrespective of preoperatively assessed pulmonary vascular resistance. There is little data to support the notion that early ventricular unloading with a cavopulmonary shunt results in improved RV function. A higher incidence of postoperative cardiac complications, such as cardiopulmonary resuscitation and arrhythmias in patients undergoing cavopulmonary shunt younger than 4 months has been reported [34]. Younger patients had a higher rate of severely impaired RV function pre- and postoperatively. Patients with an RVPA conduit are more susceptible to volume overload than patients with a BT shunt and may especially benefit from an earlier stage II operation [35].

Bidirectional cavopulmonary anastomosis or hemi-Fontan reduces the volume overload of the systemic RV in HLHS, and it seems plausible that this affects RV function positively. Despite a reduction in RV volume, RV ejection fraction and mass-to-volume ratio remained unchanged 14 months after stage II [30]. Assessment of RV functional parameters using two-dimensional speckle tracking detected no changes in global function after stage II [36]. MRI data demonstrated preserved indexed stroke volume and increased RV ejection fraction but decreased RV index [37]. Furthermore, the decreased RV volume did not result in a reduction of TV regurgitation [30]. Although TV annulus size decreases after stage II palliation, TV regurgitation does not demonstrate a commensurate improvement [32, 34, 37–39]. Diverging observations have been reported concerning TV repair at stage II palliation, with some groups advocating a rather conservative approach and other suggesting early intervention [32, 34, 39].

The effects of stage II palliation are therefore not reflected in a substantial improvement of RV function, but significant unloading and RV remodeling. The latter may contribute to improved diastolic dysfunction in the long term [40].

Fontan Completion

Stage III palliation, as the final separation of pulmonary and systemic circulation, can contribute to further ventricular volume unloading [40, 41] through prevention or reduction in systemic-pulmonary collateral development or intrapulmonary arteriovenous malformations. Little is known about special considerations for preservation of myocardial function with regard to type and timing of Fontan completion. Management has been widely derived from mixed single ventricle patient groups.

While it is well-established that an atriopulmonary connection is associated with atrial arrhythmia, sinus node dysfunction and thromboembolic events, there is little data on ventricular function favoring lateral tunnels over extracardiac conduits and vice versa. Extracardiac conduits have the advantage that they can be constructed without cardioplegic arrest and lesser manipulation of atrial tissue. Although this may theoretically be beneficial for the RV, we lack evidence on whether this actually reflects in better preservation of ventricular function. The better exercise capacity seen in extracardiac Fontan patients compared to those palliated with a lateral tunnel may indicate a beneficial effect [42]. Lateral tunnels can increase in size and can therefore be performed earlier than extracardiac conduits. Earlier conversion to a Fontan circulation did not show significant changes with regard to ventricular volumes and ejection fraction, but had a significantly better age-adjusted Tei-index, indicating less ventricular dysfunction.

Myocardium

Adequate myocardial protection is crucial to ensure good long-term function of the hypertrophied systemic ventricle. This can be achieved by ensuring optimal myocardial perfusion, by addressing any inherent anatomical coronary

problems, and by optimal myocardial protection during cardioplegic cardiac arrest.

It is important to consider anatomic features of the native hypoplastic aorta, which functions as a single coronary. Small ascending aortas as seen in aortic atresia subtypes can be technically challenging and pose the risk of pre-coronary stenosis during reconstruction of the aortic arch. On the other hand, thrombus formation in relatively large aortic roots with low flow or stasis has been reported [43, 44]. The surgical technique has to ensure an unobstructed, wide opening to the reconstructed part of the ascending aorta. Common techniques to achieve this surgical objective include longitudinal proximal incision of the native aorta and the corresponding pulmonary artery wall, patch augmentation of the contralateral aortic wall, and division with reattachment of the diminutive ascending aorta into the neoaortic root [45].

Anatomic studies have reported no relevant increase in coronary artery abnormalities in HLHS, but a higher rate of ventriculocoronary connections [43, 46]. Left coronary dominance was higher than in the normal population [43]. Flow reserve in patients with a systemic right ventricle has been examined in other patient populations. For example, patients with transposition of the great arteries after atrial switch have impairment of myocardial flow reserve [47, 48]. Compared to patients with a systemic left ventricle, a disproportionately lower perfusion of the systemic myocardium in HLHS has been reported suggesting higher vulnerability to malperfusion and cardioplegic arrest [49]. Although the systemic RV seems to be susceptible to malperfusion, most of the underlying mechanisms are surgically not modifiable.

Cardioplegia regimes vary between institutions and we lack objective data on advantages and disadvantages in pediatric patients. A survey among members of the Congenital Heart Surgeons' Society illustrated the multitude of cardioplegia techniques used [50]. Nevertheless, no conclusions on optimal cardioplegia and myocardial preservation in HLHS can be drawn from existing data. Strategies have been developed to avoid cardioplegic arrest completely, showing non-inferiority compared to conventional regimes in small series [51].

The use of an RVPA conduit has the theoretical advantage of maintaining higher diastolic arterial pressures and avoiding coronary steal when compared to palliation with a BT shunt. The diastolic runoff seen in patients with a systemic to pulmonary artery shunt reduces coronary perfusion. As discussed above, an RVPA conduit improves early postoperative mortality and this in turn may be the reflection of a better coronary perfusion. Nevertheless, it does not affect long-term outcome and results in decreased RV function after 3 years [20, 52]. Retrograde aortic arch malperfution with impaired coronary flow is a feared complication in hybrid palliation which may be prevented or treated with a reverse BT shunt [53, 54].

Although there may be several important aspects of myocardial protection, there is only little evidence for an optimal strategy with regard to cardioplegia or shunt type.

Aortic Arch

The extensive reconstruction of the aortic arch is one of the key points in palliation of HLHS. The close interrelationship between the systemic RV and the neoaorta, also known as ventriculoarterial coupling, has been studied extensively, showing not only the deleterious effects of recoarctation on ventricular function but also the negative impact of altered mechanical properties of the reconstructed aorta [55–60]. An unobstructed reconstruction, a favorable geometric shape, minimal turbulent flow, and avoidance of recurrence of coarctation are all required to optimize ventricular function.

The right ventricle is sensitive to changes in afterload [15, 61]. Patients with HLHS are particularly vulnerable to increased systemic vascular resistance due to the associated myocardial work and O_2 consumption [62]. Increased afterload leads to RV hypertrophy and functional deterioration. The surgeon faces a wide array of geometric options when reconstructing the aortic arch—and the selection of these options can

have a favorable or unfavorable effect on postoperative RV function. Suboptimal reconstruction, leading to coronary malperfusion, neoaortic obstruction and ventricular failure has been identified as major cause of death [63]. Because of the need to ensure ongoing growth while avoiding arch obstruction, surgically reconstructed aortic diameters are larger than diameters in single ventricle patients with a native aorta [12, 57, 59]. A wide anastomosis and a smooth shape of the aorta ensure maximal reduction in RV workload [64]. Nevertheless a disproportionately large reconstruction may not achieve the intended effects, as an inverse relationship between neoaortic diameter and wave energy could be shown [59].

Acute angles, stenosis, or folds cause turbulence, altered blood flow and pooling in the reconstructed aorta. Many surgical techniques have been developed, ranging from an interdigitating technique to avoid recoarctation, different uses of preshaped or custom-made bovine pericardium, division and reconfiguration of the ascending aorta to an entirely patch-free reconstruction [65–69]. Different techniques of aortic arch reconstruction can result in alterations in flow patterns and subsequent effects on RV performance.

As an example, in techniques avoiding any additional patch material, the pulmonary trunk is anastomosed to the concavity of the arch, shifting the pulmonary trunk closer to the descending aorta. This creates a relatively direct inflow to the aortic arch vessels but a very sharp angle with the descending aorta [66, 70]. Quantification of the impact of arch geometry is relatively difficult to assess. Computational fluid dynamics form an interesting new approach to evaluate the blood flow. Although computational models can only provide an approximation of the biological changes in this setting, they give valuable insights [28, 64, 71–74]. Calculated streamlines, energy loss and wall shear stress show that up to 15% of the energy generated by the systemic RV is lost depending on the neoaortic configuration [64]. To minimize energy loss and wall shear stress, a wide anastomosis and a smooth angle of the reconstructed arch have to be achieved.

It is not only the arch geometry that affects ventricular function, but also the mechanical properties of the arch material including stiffness and distensibility [55, 57, 59]. Most of the neoaortic arch is usually comprised of the augmenting patch. Native tissue can only be found on the outer curvature and its circumferential extent depends on the hypoplastic aorta's original size. Aortic arch growth in HLHS is predominantly due to growth of the native segment with no significant increase of the homograft patch [12].

Correlation of the mechanical properties of patch material and ventricular function is not well studied. Most commonly the reconstruction is performed with pulmonary artery homograft, but bovine pericardium, autologous pericardium and alternative patch material have been used as well. While some report good results with bovine pericardium [67], others consider it a major risk factor for recoarctation [75]. Even the abandonment of any patch material failed to significantly improve the incidence of reinterventions [76]. Wave intensity calculated from MRI data as a reflection of ventricular function, was significantly lower in HLHS patients, although they had no signs of malconfiguration of the reconstructed arch. An increase of arterial elastance in palliated HLHS has been associated with impaired RV diastolic but not systolic function [77]. To date, no surgical approach can significantly improve the unfavorable wall properties of the reconstructed aorta.

The incidence of coarctation of the reconstructed aorta is high with freedom from recoarctation as low as 52% at 5 years [75]. Recoarctation has an unfavorable influence on ventricular performance, RV fractional area change, atrioventricular valve function and systemic-to-pulmonary blood flow balance [56]. An unobstructed aortic arch is therefore essential to optimize and preserve RV function. At the Hospital for Sick Children, we utilize an interdigitating technique [66]. After complete excision of the periductal area, the descending aorta is incised longitudinally at the anterior and posterior wall. The posterior portion of the proximal and distal aortic arch are then anastomosed in a manner similar to an extended end-to-end coarctation repair leav-

ing the anterior portion of the anastomosis open to receive a pulmonary homograft patch. The patch augments the proximal aortic arch and extends distally to the end of the incision which was made in the anterior wall of the descending aorta. This interdigitating technique removes all periductal tissue and creates a wide unobstructed anastomosis with the descending aorta. The anterior and posterior incision thereby lead to a non-circular anastomosis, avoiding circumferential constriction. The technique showed superiority compared to classic and autologous arch reconstruction with regard to recoarctation.

The optimal hypoplastic arch repair has yet to be defined. It is vital to create a neo-aorta that provides mechanical characteristics, geometry and growth as natural as possible. This can be achieved best with wide anastomotic sites, smooth angles, and generous removal of ductal tissue.

Tricuspid Valve

Tricuspid regurgitation (TR) is a risk factor for mortality after the Norwood operation and associated with poor survival [78–81]. Up to 23% of all palliated HLHS patients have to face TV procedures within 10 years [82]. A bimodal time course has been reported with failure relatively early after diagnosis and late failure after more than 5 years [82].

No other pathology unveils its relation to RV function as openly as that of the TV. Therefore, a thorough understanding of TV dysfunction and its surgical therapy cannot be achieved without considering RV characteristics. Changes in RV contraction pattern can already be seen in the fetal circulation in HLHS [3–5], but further adaptive responses have been shown in systemic RVs after birth and the respective palliative steps [83, 84]. The ventricle in single ventricle patients has to generate output that is approximately 2.3 times higher in utero and even up to 2.8 times higher after shunt construction, than in normally developed hearts [31]. The ventricle responds with myocardial hypertrophy, especially of the middle layer, geometric reformation with a septal shift to the left, forming a more cylindrical shape, and a rather circumferential than longitudinal contraction pattern [83–85]. Ventricular dilatation, annulus dilatation, papillary muscle displacement and TV tethering are triggered by these changes and reflect gradual ventricular dysfunction. TV dysfunction may cause further RV functional decline and vice versa.

With regard to ventricular function, surgical strategies have to address any inherent or developing pathology of the TV. The annulus in a normal TV has a complex three-dimensional shape with two anteroposteriorly opposing high and two mediolaterally opposing low points [84, 86]. In patients with functional TR, this geometry deforms and the tricuspid annulus becomes larger and adopts a more circular and flattened shape. Annular dilation occurs in an anteroposterior direction along the free RV wall in patients with two ventricles [86] and in HLHS [84]. Additionally leaflet tethering with papillary muscle displacement and prolapse is associated with significant TR and myocardial dysfunction due to increased myocardial mass, afterload or coronary supply-demand mismatch [82, 84]. It remains inconclusive if ventricular interdependence has a relevant effect on TV function [16, 87, 88]. Most reports confirm the findings of anterior leaflet prolapse, septal leaflet tethering and annular dilatation in HLHS [32, 89–91]. These underlying mechanisms have to be considered in TV repair in HLHS.

TV repair is frequently undertaken for patients with moderate or severe TR. Partial annuloplasty, commissural closure, tricuspid ring annuloplasty, chordal shortening, papillary muscle relocation, and leaflet resuspension have been reported as useful surgical techniques [79, 81, 90, 92, 93]. The necessary combination of valve repair techniques remains individualized for each patient depending on the anatomic and functional contributors to the TR. Success depends on the underlying pathology and surgeon's experience. Posterior leaflet obliteration can be used to form a functional bicuspid valve with favorable results [79, 81]. Because a regurgitation jet along the anteroseptal commissure is found in 68% of patients undergoing TV repair, closure of that

commissure can be utilized [90]. Furthermore papillary muscle relocation forms a promising approach, as it restores ventricular geometry and eliminates an underlying mechanism of insufficient coaptation [90, 94]. RV function in patients with sustained successful TV repair remains better than in those with a failing valve. This can be a result of the repair itself, but may also be biased by a persisting RV function that influences the TV positively [81]. Most likely, both are true and it is impossible to quantify the respective impact.

Results of TV repair in the course of HLHS palliation are fortunately good. Freedom from TV replacement of 97% has been reported [90]. After a median follow-up period of 38.2 months after initial TV intervention 15% of patients showed recurrence of significant TR and 21% had RV dysfunction. TV repair if initially successful, is likely to have lasting results on follow-up [79]. In contrast, even initially successful TV repairs often fail in those patients with persistently poor RV function, highlighting the close interaction of RV and TV. Outcome is significantly associated with TV or RV function respectively [79]. Our experience showed excellent outcome and long-term consistency of TV repair as well [82]. After repair, TR remained stable or declined after a median of 5.1 years of follow-up. RV function was lower directly after the operation but showed non-significant differences compared to patients without the need for TV repair in the long-term. The postoperative decline in RV function can be caused by a preoperatively masked RV dysfunction, that becomes apparent after the operation [89]. The latter along with the reassuring results in this patient group is indicative for a low threshold for TV repair. The reported higher risk with younger and smaller patients should not be interpreted as a reason to delay therapy but as associated with more severe disease [79, 82].

Evolving Concepts

The complexity of HLHS and the impact of staged palliation have led to several attempts to reduce the surgical burden. Intermediate-term results of fetal intervention in a well-selected subset of patients with severe midgestation aortic stenosis have been reported recently, demonstrating the feasibility of biventricular repair [95]. Despite good results in the biventricular group, the concept has to be weighed with the associated risks for the patient and the mother in mind. Furthermore, the biventricular repair strategy preselects less critical patients, as compared to those facing univentricular palliation.

The use of routine postoperative mechanical support shows encouraging results [96]. Facing the risk of low cardiac output or unbalanced pulmonary to systemic perfusion and the possibility of impaired neurological outcome after stage I palliation, the concept aims at initial stabilization. Although this strategy has good hospital survival, it comes with the associated risks of mechanical support and less cost-effectiveness.

At our institution, an interesting new concept was introduced, that created a primary in-series circulation [97, 98]. Since cavopulmonary anastomosis in neonates has deleterious results, due to high pulmonary vascular resistance, palliation delays the connection of the superior caval vein with the pulmonary artery until four to six months of age. To avoid the risks of shunt-dependent in-parallel circulation, the concept of mechanical cavopulmonary assist establishes stable hemodynamics as the mechanical support ensures pulmonary blood flow despite high pulmonary resistance. First promising results still need to find their way from bench to bedside, but the ongoing rapid progress in mechanical circulatory support may open up exciting possibilities in the near future.

Cell based therapies have been developed to reduce RV failure or improve RV function. After years of research on myocardial recovery with autologous stem cells in adults, mainly with ischemic heart diseases, first clinical reports have shown the safety and efficacy of autologous stem cell therapy in children with HLHS [99, 100].

Conclusion

Preserving RV function in HLHS is a challenging task for all involved disciplines. Certainly, the way from the neonate to the adult is paved with endless options and decisions that finally may reflect in myocardial

performance. Nevertheless some of the surgical strategies at hand are well established and can preserve the fragile systemic RV's function. The use of a modified BT-shunt seems to be favorable in the long-term. Earlier conversion to a cavopulmonary shunt may decelerate functional decline. Unobstructed coronary perfusion, wide anastomoses and smooth angles of the reconstructed aorta, as well as techniques to avoid recoarctation are essential for the RV. Early TV repair addressing the underlying mechanisms of regurgitation can further preserve ventricular function.

Although many other aspects of HLHS palliation have been identified as contributors to RV functional decline, we still seek optimal combinations of surgical strategies to preserve RV function and promote patient survival.

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