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Left heart growth and biventricular repair after hybrid palliation

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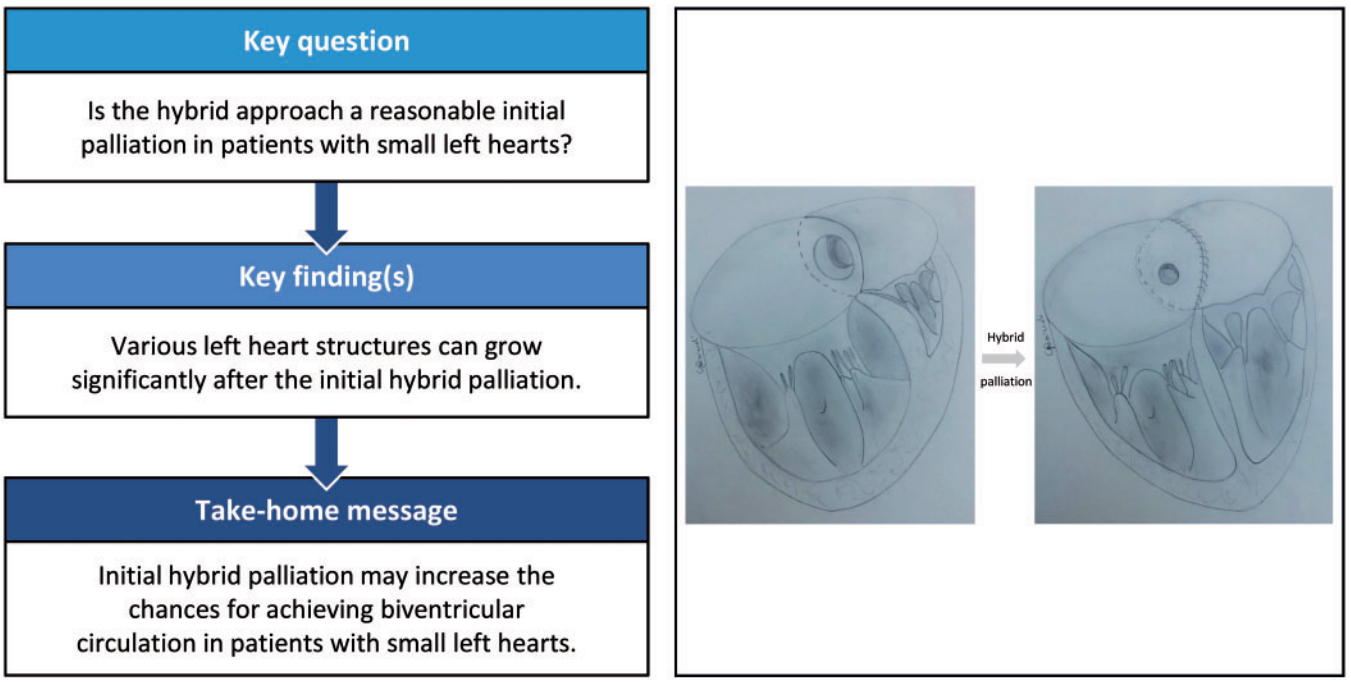

Abstract

OBJECTIVES: We evaluated the outcomes of biventricular repair after initial hybrid palliation performed in small infants with various forms of left ventricle hypoplasia.

METHODS: Between September 2010 and January 2020, a total of 27 patients had biventricular repair after hybrid palliation at a median age of 11 days. Indications for the hybrid approach included growth promotion of the left ventricle outflow tract and/or the aortic valve in 14 patients and that of the left ventricle in 13 patients. Seven reinterventions and 7 reoperations were performed during the interstage period. Significant growth of left ventricle parameters was noted during the median interstage period of 62 days. Sixteen subjects had aortic arch repair, ventricular septal defect closure and relief of subaortic stenosis; 5 patients had the Ross–Konno procedure; 5 patients underwent the Yasui procedure; and 1 patient had unbalanced atrioventricular septal defect and aortic arch repair.

RESULTS: Twenty-three patients (85.2%) are alive at a median follow-up of 3.3 years. Two and 3 patients died early and late after achieving biventricular circulation, respectively. There were 22 reinterventions and 15 reoperations after biventricular repair.

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CONCLUSIONS: Hybrid palliation can stimulate left heart growth in some patients with left ventricle hypoplasia. More patients may eventually achieve biventricular circulation than was initially thought. Additional interventions and operations are foreseeable. Despite ventricular rehabilitation, some patients with borderline left ventricles may develop restrictive physiology.

Keywords: Hybrid palliation • Bilateral pulmonary artery banding • Patent ductus arteriosus stenting • Left ventricle hypoplasia • Biventricular repair • Ventricular recruitment

ABBREVIATIONS AND ACRONYMS

AoV	Aortic valve
ASD	Atrial septal defect
BSA	Body surface area
BVR	Biventricular repair
HLHC	Hypoplastic left heart complex
HLHS	Hypoplastic left heart syndrome
IAA	Interrupted aortic arch
LV	Left ventricle
LVOT	Left ventricle outflow tract
LVEDV	Left ventricular end-diastolic volume
LVEDVi	BSA-indexed LV end-diastolic volume
MV	Mitral valve
PgE1	Prostaglandin E1
PAB	Pulmonary artery banding
PDA	Patent ductus arteriosus
PTFE	Polytetrafluoroethylene
RV–	Right ventricle
SLVR	Staged left ventricle recruitment
SVP	Univentricular pathway
UAVSD	Unbalanced atrioventricular septal defect
VSD	Ventricular septal defect

INTRODUCTION

A hybrid procedure, i.e. bilateral pulmonary artery banding (PAB) with ductus arteriosus (PDA) maintained open with a stent or prostaglandin E1 (PgE1), has been used as an initial palliation in various clinical settings, e.g. as a Norwood alternative for hypoplastic left heart syndrome (HLHS), and for stabilization of critically sick newborns with severe medical conditions and appropriate anatomy (common arterial trunk, aortic arch anomalies).

Successful application of the hybrid procedure in HLHS has broadened the spectrum of indications of this approach to include infants with more complex forms of small left ventricle (LV) who might be candidates for biventricular circulation. Previously, such patients were usually offered a univentricular pathway (SVP) during a newborn period. Despite recent improvements in the clinical outcomes of Fontan patients, their long-term outlooks are rather poor due to considerable morbidity and mortality. In addition, some patients with LV hypoplasia might also have risk factors for SVP, making them less suitable for this treatment pathway. Consequently, an increased effort has been recently made to investigate alternative strategies, possibly leading to biventricular circulation in these patients.

In newborns and small infants with the variants of LV hypoplasia not belonging to the extremes of the spectrum, it is often hard to preoperatively predict whether LV will be adequate to sustain systemic circulation. Decision between SVP and biventricular repair (BVR) has to be done early in life and be based on morphometric and haemodynamic data and clinical assessment. To date, however, there are no validated scoring systems to predict successful long-term outcomes in newborns with borderline LV undergoing BVR. Moreover, it is not easy to predict the growth potential of LV structures beyond a newborn period. Consequently, some patients with biventricular potential might be unnecessarily assigned to a Fontan pathway, while others might experience failing biventricular circulation and thus require high-risk conversion to univentricular circulation. The hybrid approach is an alternative to a complex neonatal procedure, which can defer final decision on surgical strategy in patients with borderline LV according to the observed growth of left heart structures. We present the outcomes of patients with small left hearts who have received BVR after previous hybrid palliation.

PATIENTS AND METHODS

This is a single-centre, retrospective analysis of all borderline LV patients considered potential candidates for achieving biventricular circulation, who had undergone the hybrid palliation at our institution in the period between September 2010 and December 2019. In 2018, we presented an initial study analysing the outcomes of 30 patients undergoing the hybrid palliation as a bridge to BVR [1]. When compared with the previous research, the current study enrolled additional patients with borderline left hearts and excluded critical newborns and infants receiving the hybrid palliation as a salvage procedure. Patient clinical and echocardiographic data were collected from individual hospital records.

The primary end points of the study were clinical outcomes (survival, need for reoperation, need for reintervention, and need for crossover to SVP). The secondary end points of the study were changes in left heart parameters as measured by echocardiography. 2 D-echo measurements made before the hybrid procedure, at the time of BVR and at the last follow-up were included in the analysis. Aortic valve (AoV) diameter, LV outflow tract (LVOT) diameter, mitral valve (MV) diameter and left ventricular end-diastolic volume (LVEDV) were measured in the parasternal long-axis view. The Z-score was defined as the degree of standard deviation from the mean diameter for a normal population as indexed to body surface area (BSA), and it was calculated with the Z-scores of cardiac structures as reported by Pettersen *et al.* [2]. BSA was calculated from body weight and height using the Haycock method [3]. LVEDV was measured from the apical 4-chamber view. LV-volumes were calculated according to the modified Simpson's rule indexed to BSA [4]. The AoV and LVOT data were obviously not included in the analysis during the follow-up in the subjects who had received Yasui or Ross-Konno procedure at the time of BVR.

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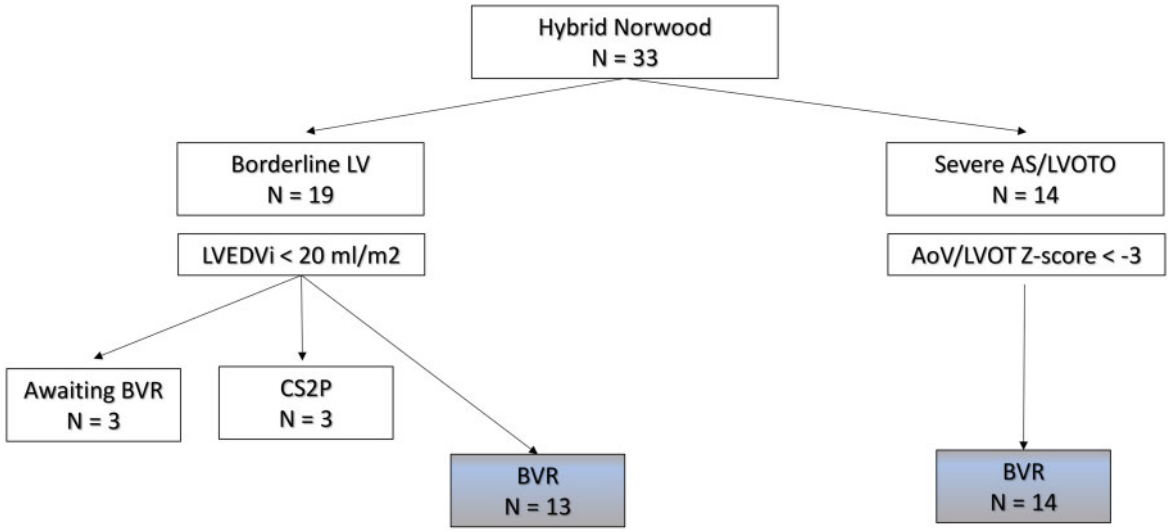

Figure 1. Patient clinical flow chart.
AoV: aortic valve; AS: aortic stenosis; BVR: biventricular repair; CS2P: comprehensive stage 2 procedure; LV: left ventricle; LVEDVi: BSA-indexed left ventricle end-diastolic volume; LVOT: left ventricle outflow tract; LVOTO: left ventricle outflow tract obstruction.

In the study period, 33 patients with potential biventricular outcome underwent an initial hybrid palliation. Patient inclusion was based upon echocardiographic assessment of left heart morphometric parameters using the following criteria, i.e. borderline LV hypoplasia was defined as BSA-indexed LV end-diastolic volume (LVEDVi) ≤ 20 ml/m2, and small LVOT and/or AoV was defined as Z-score of LVOT and/or AoV smaller than -3. In the presence of severe MV hypoplasia (Z-score < -4), severe MV dysplasia with abnormal subvalvar apparatus and/or extensive endocardiac fibroelastosis, the subjects would receive a univentricular treatment strategy. Based on the indication of the hybrid approach, the patients were allocated to the following two groups: a) borderline LV group including 19 patients with planned promotion of LV growth, and b) severe aortic stenosis (AS) and/or LVOT obstruction (LVOTO) group including 14 patients with planned LVOT and/or AoV growth (Figure 1).

The hybrid procedure involves bilateral pulmonary artery banding via median sternotomy without the use of cardiopulmonary bypass. The bands made of PTFE rings with a diameter of 3.5–4 mm were placed around both pulmonary artery branches and were tightened to achieve a diastolic flow with velocities around 3 m/s and flow extension into diastole using an epicardiac or a transoesophageal echo [1]. The PDA was stented and maintained open with PgE1 in 23 and 10 patients, respectively.

Twenty-three patients were discharged home, and they entered a home monitoring programme with daily reporting of oral intake, weight, heart rate and oxygen saturation by parents. The subjects with major changes in trends of the observed parameters needed immediate clinical attention. Otherwise, the patients were seen every week, and the dimensions of left heart structures were assessed by echocardiography [1]. Ten patients stayed in a hospital until BVR due to serious medical conditions or a need for reoperation or reintervention. Three patients underwent a comprehensive stage II procedure because of inadequate LV growth to achieve biventricular circulation. Three patients have been still awaiting BVR. The remaining 27 patients underwent BVR, and they represent the study cohort in this analysis (Table 1).

Statistical analysis

Continuous variables are expressed as a median and a range. Categorical variables are shown as numbers and percentages. Early mortality was defined as death within 30 days after surgery or during the same hospital admission. The differences between the study groups were analysed using the Mann–Whitney *U*-test. The growth of LV structures among the hybrid procedure, BVR and last follow-up was tested using the Wilcoxon signed rank test. Survival and freedom from reintervention was estimated using the Kaplan–Meier method. Difference in survival between the study groups was analysed by the log-rank test. The statistical analyses were made using IBM SPSS statistics for Windows, version 24.0 (IBM Corp., Armonk, NY, USA). A *P*-value of < 0.05 (two-sided) was considered statistically significant.

RESULTS

Initial HYBRID palliation

The patients' median age was 11 days (range: 2–52 days), and median body weight was 3.5 kg (range: 1.8–4.5 kg). The hybrid palliation was usually performed a few weeks after initial presentation to observe whether LV parameters are catching up in an infant without heart or other organ failure. The underlying diagnoses in the study groups are shown in Figure 2. Fifteen patients had hypoplastic left heart complex (HLHC), 9 patients had interrupted aortic arch (IAA) combined with ventricular septal defect (VSD) and severe LVOTO, 2 patients had critical aortic stenosis, and 1 patient had unbalanced atrioventricular septal defect (UAVSD) with IAA. The vast majority of the patients had multi-level left heart obstruction and/or hypoplasia.

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Table 1. Patient demographics [1]

Pt	Dg	Age (d)	BW (kg)	MV (Z-score)	LVOT (Z-score)	LVEDVi (ml/m^2)	Rationale for hybrid Norwood	BVR type
1	MS, AS, AA hypoplasia	14	2.9	-3.7	-3,4	14	LV growth promotion	AA repair, PA debanding, MVP
2	MV dysplasia, AS, VSD, AA hypoplasia	16	3.5	-2,0	-7,0	17	LV growth promotion	AA repair, VSD closure, PA debanding
3	MS, AS, VSD, AA hypoplasia	21	2.4	-2,2	-6,0	20	LV growth promotion	AA repair, VSD closure, LVOTO relief, PA debanding
4	AS, VSD, AA hypoplasia, Down syndrome	8	3.2	-2,0	-3,0	12	LV growth promotion	AA repair, VSD closure, PA debanding
5	IAA type B, VSD multiple, LVOTO, di George syndrome	26	4.5	-1,1	-6,0	15	LV growth promotion	Yasui procedure, PA debanding
6	MV dysplasia, AS, AA hypoplasia	27	4.5	-1,9	-6,6	20	LV growth promotion	AA repair, PA debanding
7	IAA type B, VSD, LVOTO, di George syndrome	20	3.5	-4,0	-5,0	13	LV growth promotion	AA repair, VSD closure, PA debanding
8	MS, AS, AA hypoplasia	18	4	-1,3	-1,3	17	LV growth promotion	AA repair, PA debanding
9	IAA type B, VSD, LVOTO, Charge syndrome	5	2.9	-2,4	-6,7	20	LV growth promotion	AA repair, VSD closure, LVOTO relief, PA debanding
10	MS, AS, VSD, AA hypoplasia	10	3.5	-2,5	-4,6	12	LV growth promotion	AA repair, VSD closure, PA debanding
11	MS, AA hypoplasia, VSD, ASD	2	4	-2.4	-6	19	LV growth promotion	AA repair, VSD closure, PA debanding, LVOTO repair
12	Borderline LV, AA hypoplasia	11	3.5	-2	-2	20	LV growth promotion	AA repair, ASD closure, PA debanding
13	UAVSD, IAA type B	6	3.5	0.0	-3.6	11	LV growth promotion	AVSD repair, AA repair, PA debanding
14	IAA type B, VSD, LVOTO, di George syndrome, a. lusoria	5	3.2	-1,8	-7,0	27	AoV/LVOT growth promotion	Yasui procedure, PA debanding
15	IAA type B, VSD, LVOTO, a. lusoria	35	2.4	0,2	-9,0	31	AoV/LVOT growth promotion	Ross-Konno procedure, AA repair, PA debanding
16	IAA type B, VSD, LVOTO	13	3.9	-1,6	-7,0	23	AoV/LVOT growth promotion	Yasui procedure, PA debanding
17	IAA type A, VSD, LVOTO	16	3.6	-0,5	-7,0	33	AoV/LVOT growth promotion	Ross-Konno procedure, AA repair, PA debanding
18	IAA type B, VSD, LVOTO, a. lusoria	27	2.8	0,4	-7,0	39	AoV/LVOT growth promotion	Yasui procedure, PA debanding
19	IAA type B, VSD, a. lusoria	9	3	-1,7	-3,1	25	AoV/LVOT growth promotion	Yasui procedure, PA debanding
20	AS, AA hypoplasia	11	1.8	-1,8	-6,7	23	AoV/LVOT growth promotion	Ross-Konno procedure, AA repair, PA debanding
21	AS, multiple VSD, AA hypoplasia	22	4	0,0	-3,7	46	AoV/LVOT growth promotion	AA repair, VSD closure, PA debanding
22	AS, VSD, AA hypoplasia	8	3.2	-0.9	-3.1	40	AoV/LVOT growth promotion	AA repair, VSD closure, PA debanding
23	AS, AA hypoplasia	52	2.4	-0,2	-2,5	82	AoV/LVOT growth promotion	Ross-Konno procedure, PA debanding
24	MS, AS, multiple VSD, AA hypoplasia	5	3	-3,0	-3,6	31	AoV/LVOT growth promotion	AA repair, PA debanding
25	AS, VSD, AA hypoplasia	8	4	0,1	-3,0	38	AoV/LVOT growth promotion	AA repair, VSD closure, PA debanding
26	AA hypoplasia, VSD, LVOTO, ASD	6	3,5	-0.2	-3.3	27	AoV/LVOT growth promotion	AA repair, VSD closure, ASD closure, PA debanding
27	Critical AS, S/P BD, AI, AA hypoplasia, ASD	11	3	-2.4	-3.4	25	AoV/LVOT growth	Ross-Konno, PA debanding, ASD closure

AA: aortic arch; AoV: aortic valve; AS: aortic stenosis; ASD: atrial septal defect; ASO: arterial switch operation; AVSD: atrioventricular septal defect; BD: balloon dilatation; BW: body weight; BVR: biventricular repair; d: days; Dg: diagnosis; IAA: interrupted aortic arch; LV: left ventricle; LVEDVi: BSA-indexed left ventricle end-diastolic volume; LVOT: left ventricle outflow tract; LVOTO: left ventricle outflow tract obstruction; MR: mitral regurgitation; MS: mitral stenosis; MV: mitral valve; MVP: mitral valve plasty; PA: pulmonary artery; PAB: pulmonary artery banding; Pt: patient; S/P: status post; VSD: ventricular septal defect; TrV: truncal valve; UAVSD: unbalanced atrioventricular septal defect.

Interstage PERIOD

The median length of interstage period, i.e. between the initial hybrid palliation and BVR was 62 days (range: 10–349 days). While it was not statistically significantly different between the borderline LV and the severe AS and/or LVOTO groups: 53 days vs 65 days, $P = 0.72$, a statistically significant difference was observed between no stent and stent groups (42 days vs 89 days; $P = 0.027$).

The atrial septal defect (ASD) was left deliberately restrictive to improve LV filling and potentially stimulate the growth of underdeveloped left heart structures.

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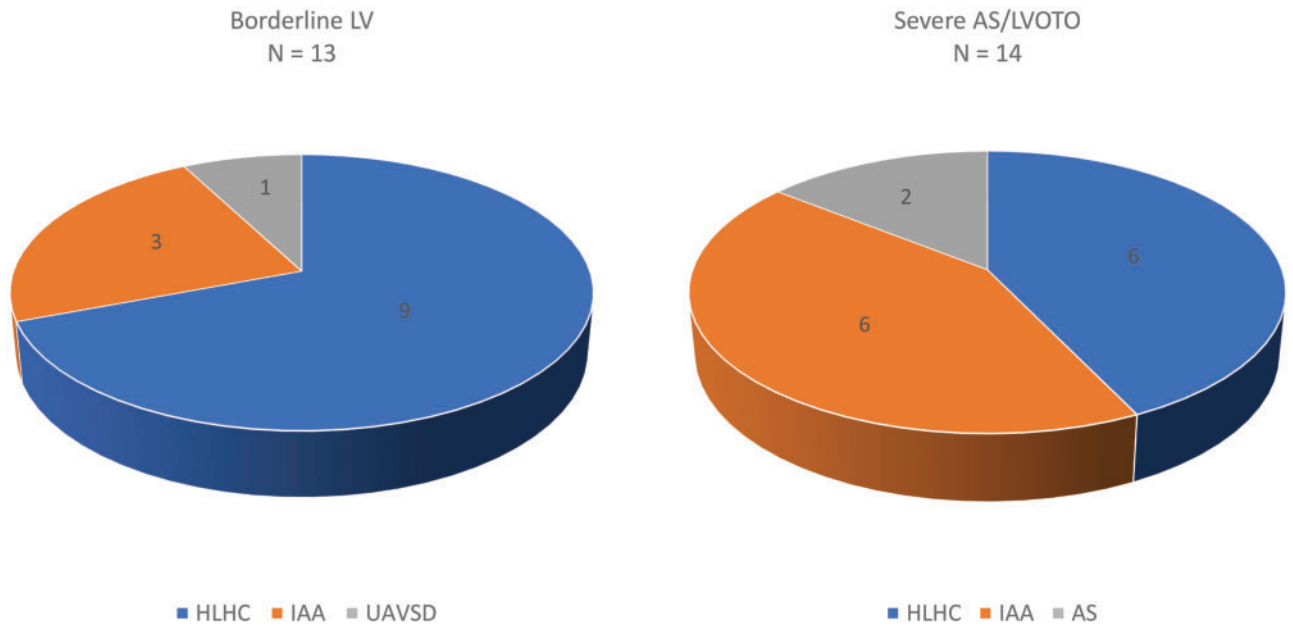

Figure 2. Diagnoses in study groups.
AS: aortic stenosis; HLHC: hypoplastic left heart complex; IAA: interrupted aortic arch; LV: left ventricle; LVOTO: left ventricle outflow tract obstruction; UAVSD: unbalanced atrioventricular septal defect.

Reinterventions. Seven catheter-based interventions including PDA stent balloon dilatation or over-stenting, $N = 6$; and re-balloon atrial septostomy, $N = 1$; were performed during the interstage period.

Reoperations. There were 7 surgical procedures during the interstage interval including PAB loosening, $N = 4$; ASD size reduction because of inadequate interatrial flow restriction, $N = 2$; and extraction of a migrated PDA stent, $N = 1$.

Left heart growth. In the borderline LV group, a significant increase in LVEDVi ($P = 0.0017$) and AoV/LVOT values ($P = 0.005$) was observed during the interstage period. In the severe AS and/or LVOTO group, a similarly significant increase in AoV/LVOT ($P = 0.004$) and LVEDVi values ($P = 0.025$) was noted during the interstage period (Figures 3 and 4).

Biventricular repair

At the time of BVR, the median age was 83 days (range: 15–371 days), and the median body weight was 4.5 kg (range: 2.8–7.0 kg). The decision on BVR and its timing was based upon echocardiographic demonstration of the growth of LVOT and/or AoV (Z-score > -2.5) and that of LV (LVEDVi > 20 ml/m^2). BVR was aimed at radical relief of any obstructive lesions in LV inflow and/or outflow. Sixteen patients underwent conventional aortic arch repair +/- VSD closure +/- LVOTO relief (subaortic membrane resection in 2 patients), 5 patients received Ross–Konno procedure (including aortic arch repair in 3 patients), 5 patients underwent Yasui procedure, and 1 patient had UAVSD and aortic arch repair. Concomitant mitral valvuloplasty was performed in 2 patients. A smaller ASD was left open in 2 patients at risk of developing LV dysfunction.

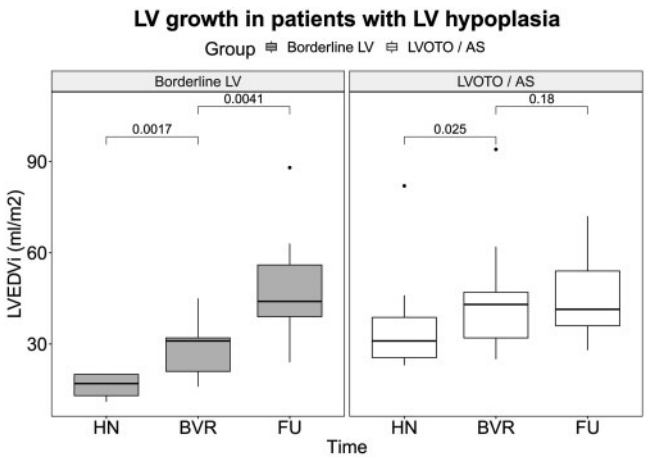

Figure 3. LV growth in patients with LV hypoplasia.
AS: aortic stenosis; BVR: biventricular repair; FU: follow-up; HN: hybrid palliation; LV: left ventricle; LVEDVi: BSA-based left ventricle end-diastolic volume; LVOTO: left ventricle outflow tract obstruction.

Survival. There are 23 survivors among 27 patients who underwent BVR after initial hybrid palliation, suggesting an overall survival of 85.2% (Figure 5). Two deaths occurred early after Ross–Konno and aortic arch repair due to lung bleeding and extracorporeal membrane oxygenation complications. There were two late deaths after BVR. One patient with UAVSD and IAA received a staged repair of aortic arch and UAVSD. Because of residual LVOTO and left atrioventricular valve leakage, he later underwent a Ross–Konno and left atrioventricular valvuloplasty. Subsequently, he developed intractable right ventricle (RV) failure and right atrioventricular valve leakage not responding to valvuloplasty and subsequently to valve replacement. Another patient with IAA, VSD and LVOTO died late after a Yasui procedure due to respiratory infection, pulmonary hypertension and RV failure. No statistically significant difference in survival was

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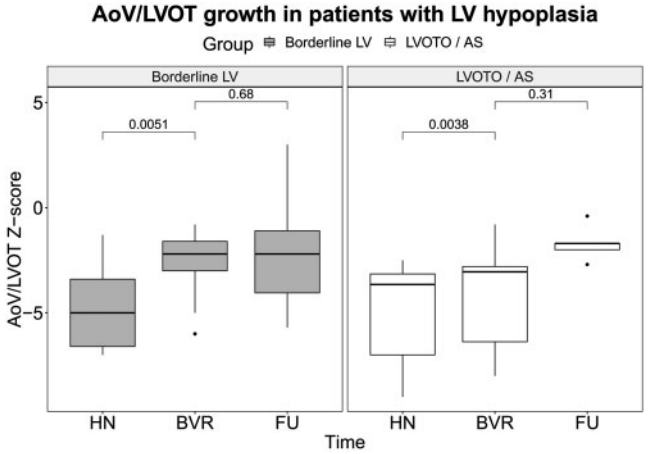

Figure 4. AoV/LVOT growth in patients with LV hypoplasia.
AoV: aortic valve; AS: aortic stenosis; BVR: biventricular repair; FU: follow-up; HN: hybrid palliation; LV: left ventricle; LVOT: left ventricle outflow tract; LVOTO: left ventricle outflow tract obstruction.

observed between the borderline LV and the severe AS/LVOTO group ($P = 0.25$).

Follow-up

Median follow-up is 3.3 years (range: 0.21 – 7.4 years). All 23 survivors have no clinical symptoms and preserved LV function except for 2 patients with pre-existing LV dysfunction.

Reinterventions. Twenty-two reinterventions (balloon dilatation or stenting of aortic arch, $N = 13$; balloon dilatation or stenting of pulmonary artery, $N = 8$; balloon dilatation of pulmonary valve (PV), $N = 1$) were required after BVR. The overall freedom from reintervention after BVR is shown in Figure 6. There was a significant difference in freedom from reintervention between the study groups (borderline LV group better, $P = 0.035$).

Reoperations. The BVR survivors received a total of 15 additional surgical procedures (LVOTO relief, $N = 5$ incl. 2 Ross–Konno procedures; TV repair or replacement, $N = 3$; residual VSD closure, $N = 3$; PV replacement, $N = 2$; mitral valvuloplasty, $N = 1$; and pulmonary artery plasty, $N = 1$. The overall freedom from reoperation after BVR is shown in Figure 7. No significant difference in freedom from reoperation was observed between the study groups ($P = 0.65$).

Crossover to SVP. None of the BVR survivors has to be switched to SVP during the follow-up.

Left heart growth. The borderline LV group showed a significant left heart growth as evidenced by increasing LVEDVi values during follow-up ($P = 0.004$).

DISCUSSION

The management of infants with borderline left heart structures is controversial and varies among institutions. A crucial decision on pursuing SVP vs BVR has to be made early in life in order to

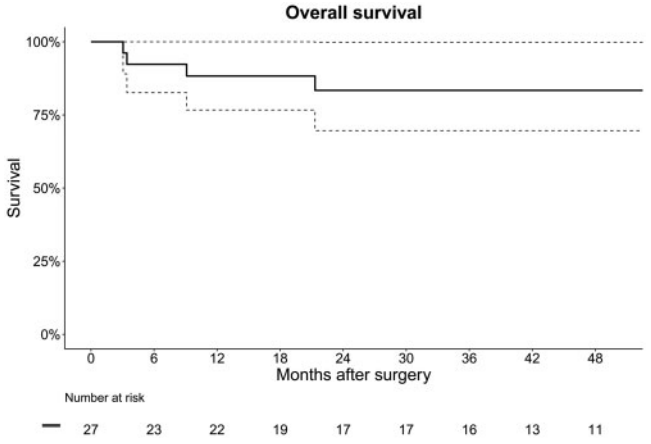

Figure 5. Overall survival.

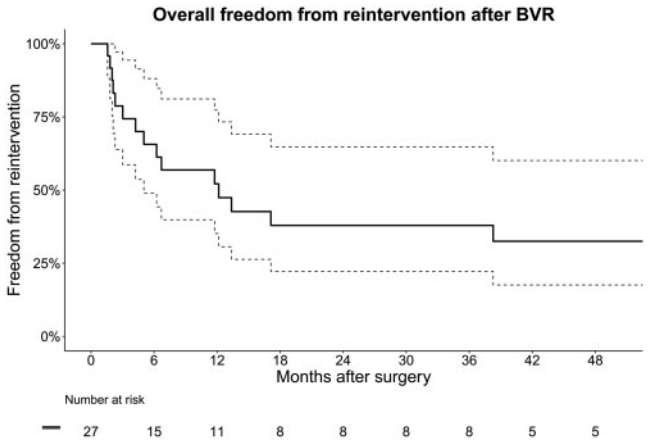

Figure 6. Overall freedom from reintervention after biventricular repair.

achieve optimal outcomes in this patient population. Well-known poor long-term outcomes of patients with SVP have led to a clinical bias favouring BVR in infants with borderline LV hypoplasia. Several predictive scoring systems have been suggested to assist with selection of suitable patients for biventricular circulation. However, these predictors are not uniformly accurate and reliable in small infants with LV hypoplasia. As a result, improper commitment of a newborn with a borderline but inadequate LV size and/or function to biventricular circulation may be associated with poor early and late outcomes [5].

Two surgical approaches have been suggested for possible LV recruitment in subjects with borderline LV hypoplasia and uncertain feasibility of primary early repair. With both strategies, an interatrial communication is left deliberately restrictive to promote the growth of left heart structures in order to possibly achieve successful biventricular circulation.

Emani *et al.* recommend a staged LV recruitment (SLVR), i.e. initial Norwood procedure with a radical relief of obstructive lesions affecting LV inflow and outflow and resection of endocardial fibroelastosis at the same stage or more often during subsequent stages of SVP before a gradual loading of the hypoplastic ventricle will potentially stimulate LV remodelling. Decision regarding biventricular conversion can be delayed even for several years until adequate LV growth and function have been demonstrated by means of cardiac MRI, echocardiography and catheterization. The patients with persistent LV hypoplasia or LV

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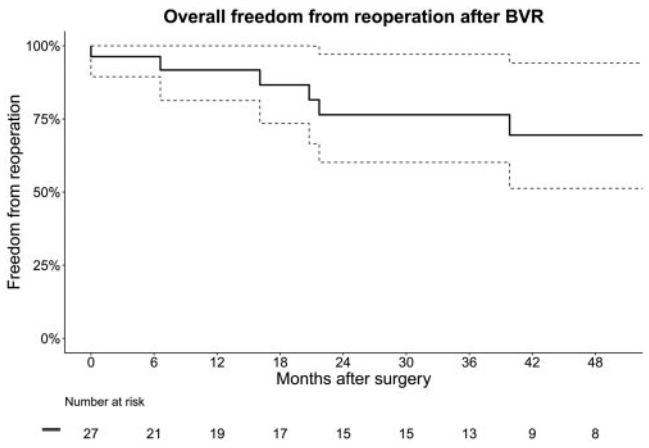

Figure 7. Overall freedom from reoperation after biventricular repair.

dysfunction will remain with SVP. These authors conducted two studies including the 51 patients with HLHS variants and UAVSD [6], and 21 patients with double outlet right ventricle or UAVSD and hypoplastic ventricles [7], respectively.

Other authors suggest an initial hybrid palliation during an early infant period to be followed by a BVR in suitable candidates after demonstration of the growth of underdeveloped left heart structures. Yerebakan *et al.* published the long-term results of BVR after initial hybrid procedure in 40 patients with hypoplastic left heart variants. The authors reported almost doubled number of patients suitable for BVR at the end of the interstage period compared with that at initial presentation. Up to 85% of the patients remained with biventricular circulation during a median follow-up of 7.9 years. Thirty percent of patients required reoperations particularly for LVOTO, and 10% of patients required reinterventions aimed at aortic arch or pulmonary artery [8]. In 2018, we presented an initial study analysing the outcomes of 30 patients undergoing the hybrid palliation as a bridge to BVR. Our findings suggested that this strategy can be safely used in small infants with borderline left hearts, possibly leading to a higher proportion of subjects achieving BVR [1]. The current study enrolled additional patients with borderline left hearts and excluded critical newborns and infants receiving the hybrid palliation as a salvage procedure. No further interstage catheter-based interventions and reoperations were needed suggesting overcoming an initial learning curve. When compared with our previous study, we also noted an increasing proportion of patients undergoing conventional aortic arch repair with LVOTO relief and/or VSD closure despite initial severe AS/LVOTO. We think the long-term outcomes of conventional aortic arch repair and VSD closure in patients with concomitant severe LVOTO are superior to those of Ross–Konno or Yasui procedure due to higher risk of reinterventions and reoperations [9], although larger and longer controlled studies are needed to prove this assumption. Up to 70% of patients from the original cohort with uncertain feasibility of BVR have maintained clinically stable biventricular circulation during median follow-up of 3.3 years. Two early deaths occurred after Ross–Konno and aortic arch repair. Mair *et al.* also suggested high-risk of mortality in small infants undergoing Ross–Konno procedure with aortic arch repair [10]. We observed two late deaths due to pulmonary hypertension and RV failure probably as a result of restrictive LV physiology. In two patients at risk for LV dysfunction, we have left an ASD open at the time of BVR to mitigate possible development of left atrial hypertension in the future. Other authors have also expressed concerns over LV dysfunction as a result of aggressive pursuit of BVR in some patients with small left hearts which can limit the long-term advantage of achieving biventricular circulation [6, 11].

In our opinion, the main advantage of the hybrid approach over SLVR lies in avoidance of major open-heart surgery during a neonatal period and that of complex LV rehabilitation procedure in subjects who might not be candidates for BVR in future. However, we do acknowledge that the patients undergoing SLVR had more severe forms of borderline LV hypoplasia including endocardiac fibroelastosis compared to our study cohort.

In accordance with others [8], reoperations and reinterventions were common after BVR. In our analysis, we were unable to identify any clear-cut echocardiographic criterion that would guarantee the long-term success of BVR or directly affect the risk for reoperation or reintervention after BVR. Larger, long-term, prospective, controlled studies are required to demonstrate whether strategies aimed at LV rehabilitation provide long-term advantage over SVP in patients with smaller left hearts.

Limitations

This was a retrospective study including a non-randomized, non-controlled and highly selective cohort of patients with various diagnoses. Therefore, meaningful statistic comparisons between the patient groups are limited.

CONCLUSION

Hybrid approach is a safe, effective and reproducible initial palliative alternative to a major surgery in a small infant with borderline LV hypoplasia and/or obstruction. It allows for deferred decision on the definitive surgical approach, i.e. SVP vs BVR, based on potential growth of left heart structures. As a result, more patients may eventually receive biventricular circulation than it was thought at initial presentation. Additional operations and interventions are common during both interstage period and follow-up. Finally, a word of caution: despite ventricular rehabilitation, some patients with borderline LV hypoplasia can develop restrictive LV cardiomyopathy and pulmonary hypertension.

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