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# A staged decompression of right ventricle allows growth of right ventricle and subsequent biventricular repair in patients with pulmonary atresia and intact ventricular septum†

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## Abstract

**OBJECTIVES**: To achieve the growth of right-sided heart structures, our choice of the first palliation for patients with pulmonary atresia and intact ventricular septum (PA-IVS) includes a modified Blalock–Taussig shunt (BTS) with pulmonary valvotomy. We sought to analyse the impact of the first palliation on the growth of right-sided heart structures and factors associated with a choice of definitive surgical procedure.

**METHODS**: Fifty patients with PA-IVS who underwent a staged surgical approach from 1991 to 2012 were retrospectively reviewed.

**RESULTS**: Right ventricular (RV)-coronary artery fistulas were seen in 42% of patients at the time of birth. All 50 patients had a modified BTS with pulmonary valvotomy. Six patients died after the first palliation or inter-stage. Thirty patients achieved a biventricular repair (BVR group), 6 patients had a 1 + 1/2 ventricular repair (1 + 1/2V group) and 5 patients had a Fontan completion (Fontan group). After modified BTS with pulmonary valvotomy, tricuspid valve *z*-score did not increase in any of the group (BVR: pre −2.79 vs post −2.24, 1 + 1/2V: pre −5.25 vs post −6.69, Fontan: pre −6.82 vs post −7.94). Normalized RV end-diastolic volume increased only in BVR group after modified BTS with pulmonary valvotomy (BVR: pre 32% vs post 64%, 1 + 1/2V: pre 43% vs post 42%, Fontan: pre 29% vs post 32%). Major RV-coronary artery fistula was a strong factor with proceeding single-ventricle palliation [BVR: 4/30 (13%) patients, 1 + 1/2V: 1/6 (17%) and Fontan: 4/5 (80%)].

**CONCLUSIONS**: Tricuspid valve growth was not obtained by modified BTS with pulmonary valvotomy; therefore, tricuspid valve size at birth appeared to be a predictor for achieving BVR. Proportionate RV growth was seen only in patients who achieved BVR. However, RV growth was not seen in patients having 1 + 1/2 ventricular repair. Major RV-coronary artery fistula was a strong predictor for proceeding single-ventricle palliation.

**Keywords:** Pulmonary atresia and intact ventricular septum • Right ventricle • Biventricular repair

## INTRODUCTION

Biventricular repair (BVR) may offer superior long-term outcome in patients with pulmonary atresia and intact ventricular septum (PA-IVS); however, there is a considerable risk of late deterioration of exercise capacity and functional status, even in patients with adequate size of right ventricle (RV) [1–3]. This entity has been largely treated with single-ventricular repair with reasonable early survival [4–6]. In patients with RV-dependent coronary circulation (RVDCC) and severe hypoplasia of RV and tricuspid valve (TV), surgical decisions are made to perform single-ventricle repair. However, there is still a controversial and institutional bias if patients present with borderline RV and TV.

Given poor long-term outcome of single-ventricle repair, our institutional strategy is to aim for BVR [7, 8]. Our bias towards BVR is based on previous reports showing potential RV growth when the continuity between RV and pulmonary artery was created [9, 10]. Our initial palliation consists of surgical pulmonary valvotomy and placement of a modified Blalock–Taussig shunt (BTS). Catheterization is performed to assess the size of RV and TV, RV pressure and RV-coronary fistula. Percutaneous pulmonary balloon valvotomy is then performed to aim for further anatomical growth and RV decompression. We sought to analyse the impact of the palliation on the growth of right-sided heart and factors associated with a choice of definitive surgical procedure.

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## MATERIALS AND METHODS

The Research Ethics Board at Okayama University Hospital approved this retrospective study and patient consent was waived. From March 1991 to November 2012, 50 neonates with PA-IVS who were managed at Okayama University Hospital were included. Six patients with critical pulmonary stenosis were also included in the study. Patients who had an initial surgery or catheter intervention at other institutions were excluded from the study. All but 2 patients underwent cardiac catheterization before initial palliation and 23 (46%) were diagnosed as having RV-coronary artery fistula. RVDCC was identified in 2 patients. Balloon atrioseptostomy was performed in 35 (70%) patients. The median age at balloon atrioseptostomy was 12 [interquartile range (IQR), 8.5–23.5] days.

### Patient management

The institutional decision-making algorithm is shown in Fig. 1.

***Initial surgical palliation.*** Transarterial pulmonary valvotomy and placement of modified BTS via left thoracotomy were performed without the use of cardiopulmonary bypass [7]. The pulmonary valve was penetrated with a knife about 2 mm. It was further opened with a 3- or 4-mm Hegar dilator through the incision in the main pulmonary artery only if a patient had a minor RV-coronary artery fistulas. Patients with a major RV-coronary artery fistula and RVDCC are not suitable for this as the sudden decrease in RV pressure poses a risk for coronary ischaemia. We aim for equal or supra-systemic RV pressure (RVP/LVP > 1.0) for patients with a major RV-coronary artery fistula and RVDCC and less than systemic RV pressure (RVP/LVP < 1.0) for patients with a minor RV-coronary artery fistula. A 3.5- or 4-mm polytetrafluoroethylene tube (Gore-Tex tube; W. L. Gore & Associates, Inc., Flagstaff, AZ, USA) was anastomosed to the left sub-clavian artery and the incision made on the main pulmonary artery. The ductus arteriosus was then ligated.

***Multistaged palliative procedures.*** All patients who survived the initial surgical palliation underwent diagnostic cardiac catheterization. At this stage, an RV to left ventricular pressure (RVP/LVP) ratio greater than 1.0 was an indication for balloon pulmonary valvotomy ($n = 19$). For patients with major RV-coronary artery fistulas, balloon pulmonary valvotomy was performed to achieve an RVP/LVP ratio of 1.0 or slightly less. RVP/LVP was kept greater than 1.0 in patients with RVDCC. For patients with RV end-diastolic volume (RVEDV) of less than 50% of the predicted normal value estimated at the last cardiac catheterization, we performed an 'RV overhaul' procedure ($n = 21$) [7]. The RV overhaul procedure included repeat pulmonary valvotomy, transatrial and transpulmonary resection of the hypertrophied infundibular muscle, and adjustment of an interatrial communication. To increase blood flow through the TV, we partially closed the atrial septal defect (ASD), keeping right atrial pressure of less than 15 mmHg and maintained a gradient across the ASD of less than 10 mmHg.

***Definitive repair.*** We generally consider a TV $z$-score of larger than −3 as the indication for BVR. If the TV $z$-score is less than −8, we consider single-ventricle repair is favourable. Patient having the TV $z$-score of between −8 and −3 is considered a candidate for one-and-a-half ventricular repair. The BVR included construction of an unobstructed RV outflow and closure of the intra-atrial communication. We used various techniques including pulmonary

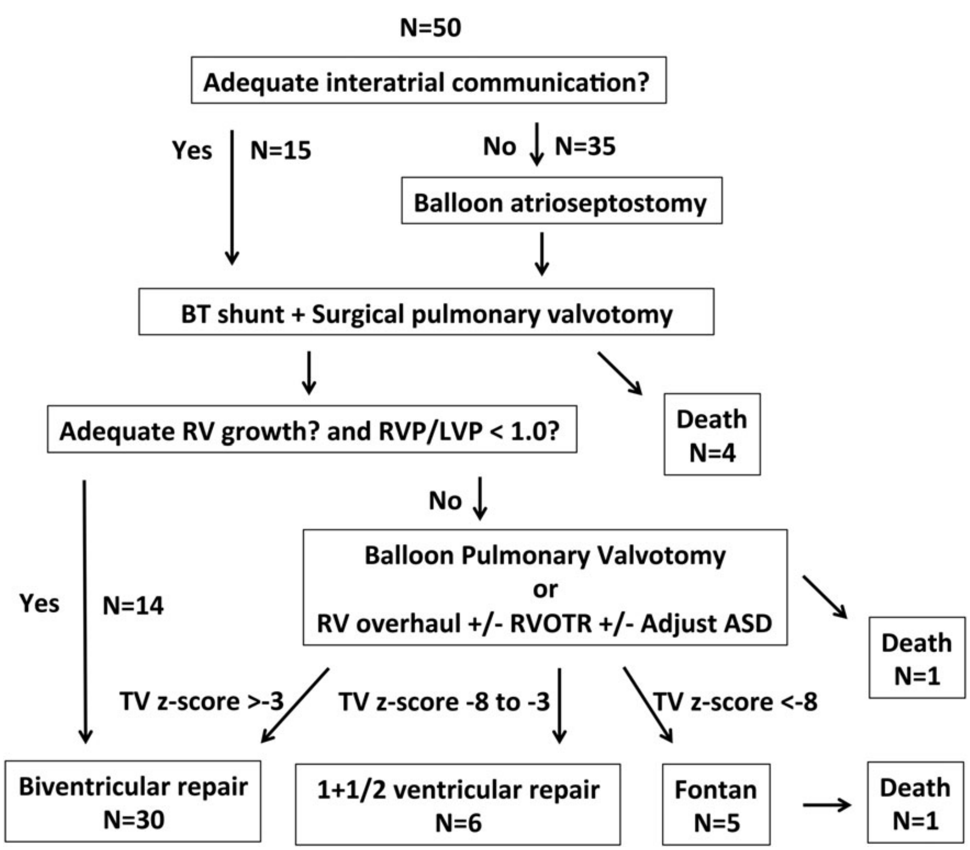


**Figure 1:** The decision-making algorithm for patients with PA-IVS at Okayama University Hospital. BT: Blalock–Taussig; RV: right ventricular; RVP: RV pressure; LVP: LV pressure; ASD: atrial septal defect; TV: tricuspid valve; PA-IVS: pulmonary atresia and intact ventricular septum; RVOTR: right ventricular outflow tract reconstruction.

valve repair, resection of obstructing infundibular muscle, excision of hypertrophied RV muscle to enlarge the cavity and monocuspid transannular patching to achieve a normal-sized outflow tract. During BVR or one-and-a-half ventricle repair with a marginal TV diameter, we used an adjustable stitch to close the interatrial communication for a judgement after weaning from cardiopulmonary bypass. We left a small fenestration during closure of the interatrial septum in 15 patients.

## Statistical analysis

Continuous data are presented as median (IQR). The level of statistical significance was set at $P \leq 0.05$. The paired Student's *t*-test was used for comparing continuous variables for the same patient. Differences between two groups were analysed by unpaired *t*-test. Differences between the groups were analysed by one-way analysis of variance (ANOVA) and *post hoc* test. Freedom from death was analysed using Kaplan–Meier analysis and a log-rank test. Statistical analysis was performed using SPSS version 21.0 (IBM Corporation, Armonk, NY, USA) statistical software.

# RESULTS

## Echocardiographic and catheterization findings before initial palliation

The median body weight at birth was 2.8 (2.4–3.1) kg. The median TV *z*-score measured by echocardiogram was −3.66 (−6.82 to −0.77), including 6 patients having less than −8 of the TV *z*-score. RVP/LVP obtained from catheterization was 1.31 (1.17–1.60). RVEDV was 32% (25–49%) of the predicted normal value. Major RV-coronary artery fistula was noted in 16 and 5 patients had a minor RV-coronary artery fistula. RVDCC was seen in 2 patients.

## Initial palliation

All 50 patients underwent surgical pulmonary valvotomy and modified BTS at a median age of 26 (15.0–37.8) days. There was 1 hospital death due to sudden haemodynamic collapse during surgery in a patient having a major RV-coronary artery fistula. There were 3 inter-stage deaths (arrhythmia in 1, sudden death in 1 and intracranial haemorrhage in 1). A total of 46 patients were evaluated by cardiac catheterization after 9.0 (5.0–21.0) months of initial surgery. RVEDV significantly increased from 32 to 49% of the predicted normal value ($P = 0.03$) (Fig. 2A). RVP/LVP decreased significantly from 1.31 before initial palliation to 1.10 ($P = 0.01$). In contrast, TV *z*-score significantly decreased from −3.66 to −4.57 ($P = 0.04$) (Fig. 2B).

## Multistaged procedure

A total of 44 patients underwent secondary intervention. Twenty-six patients underwent surgical procedure, including BVR in 14, RV overhaul in 4, bidirectional cavopulmonary shunt in 4, one-and-a-half ventricular repair in 3 and BT shunt in 1. Balloon pulmonary valvotomy was performed in 18 patients. There was 1 hospital death due to arrhythmia after RV overhaul. Repeated balloon pulmonary valvotomy was performed in 5 patients to achieve the TV and RV growth before the definitive repair.

## Definitive repair and clinical outcome

Latest status is shown in Fig. 1. Thirty out of 50 (60%) patients achieved BVR and 6 patients underwent one-and-a-half ventricular repair, so 72% of patients were able to avoid single-ventricle repair. Five patients had Fontan procedure with 1 late death due to heart failure. The median follow-up period was 111 (48–191) months. The overall survival was 87.8, 87.8 and 87.8% at 1, 5 and 10 years, respectively. Survival was significantly lower in patients who had single-ventricle repair compared with those who had BVR or one-and-a-half ventricular repair (100% in BVR, 100% in one-and-a-half ventricular repair and 75% in single-ventricle repair at 10 years, $P = 0.01$). There were three reoperations in the patients who had BVR. Two patients had one-and-a-half ventricular repair by bidirectional cavopulmonary shunt for liver congestion. One patient had a TV repair for severe tricuspid regurgitation. There were no patients who had arrhythmia.

## Factors associated with tricuspid and right ventricle growth

RVEDV-predicted normal value at birth was 32% in BVR group, 43% in one-and-a half ventricular repair group and 29% in single-ventricle group ($P = 0.41$). Proportionate RV growth was seen only in patients who achieved BVR (32% of RVEDV predicted normal

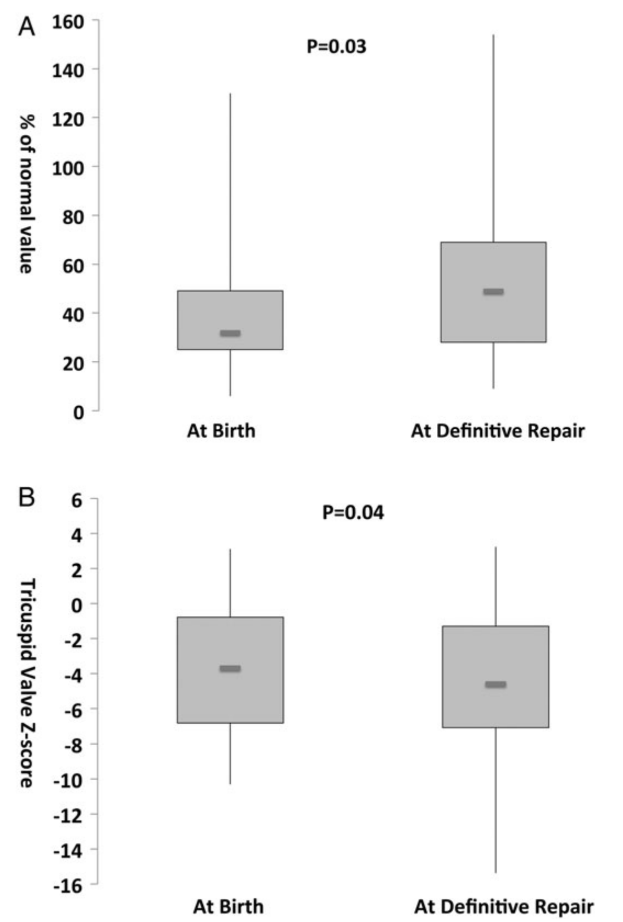


**Figure 2:** Change in the size of right ventricle and tricuspid valve before and after palliation. (**A**) Right ventricular end-diastolic volume-predicted normal value significantly increased after palliation. (**B**) Tricuspid valve *z*-score significantly decreased.

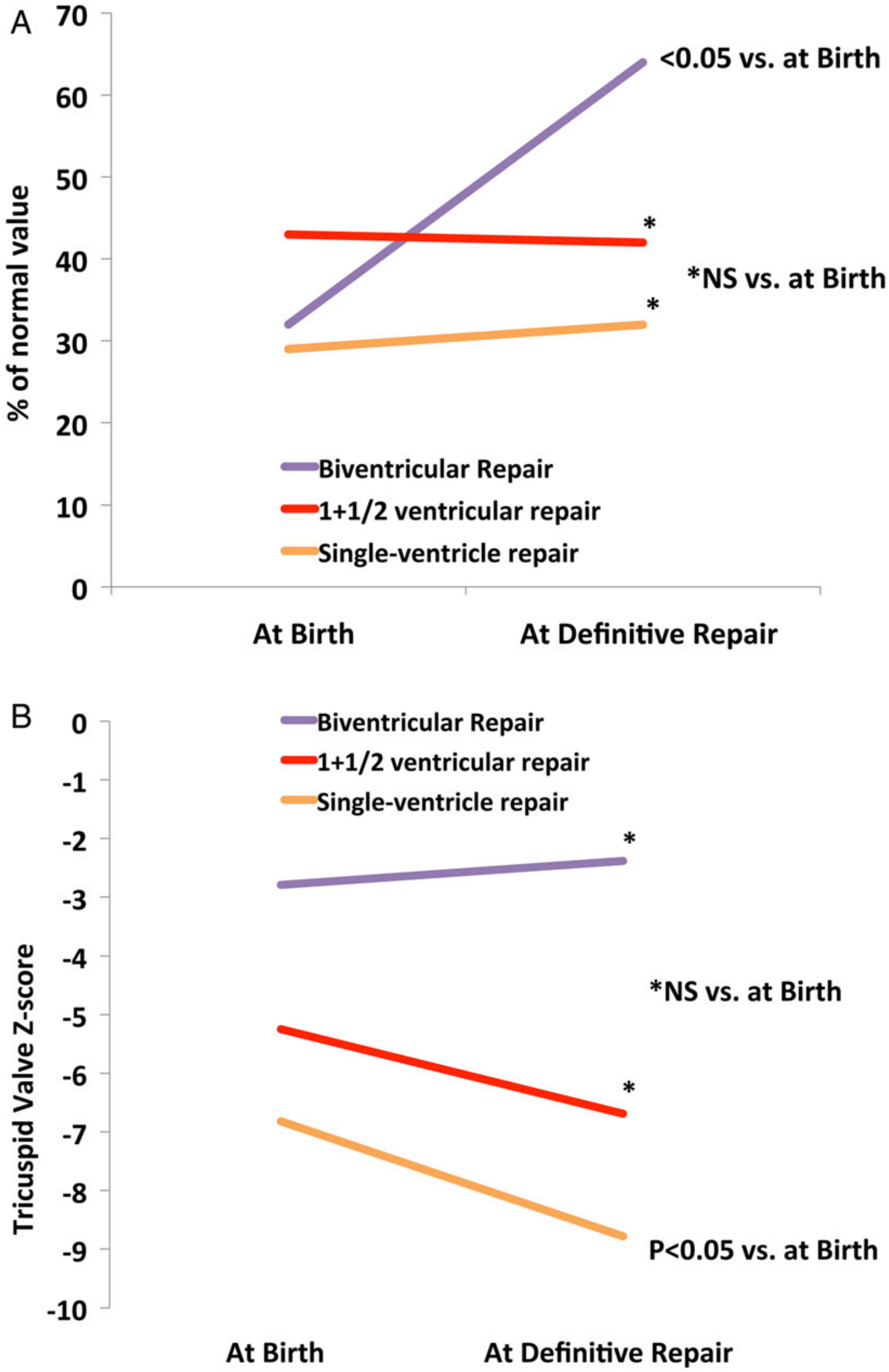


**Figure 3:** Change in the size of right ventricle and tricuspid valve stratified by the type of repair. (**A**) Right ventricular end-diastolic volume-predicted normal value significantly increased only in patients for which biventricular repair was achieved. (**B**) Proportionate tricuspid valve growth was not observed in any type of repair.

value at birth vs 64% at the time of definitive repair, $P < 0.01$) (Fig. 3A). However, RV growth was not seen in patients having one-and-a-half ventricular repair or single-ventricle repair [43–42% in one-and-a-half ventricular repair ($P = 0.99$) and 29–32% in single-ventricle repair ($P = 0.95$)].

TV *z*-score at birth was −2.79 in the BVR group, −5.25 in the one-and-a half ventricular repair group and −6.82 in the single-ventricle repair group ($P = 0.04$). In contrast to RV growth, TV did not grow in BVR (−2.79 of TV *z*-score at birth vs −2.38 at the time of definitive repair, $P = 0.33$) and one-an-a-half ventricular repair (−5.25 vs −6.69, $P = 0.20$) (Fig. 3B). TV size was significantly decreased in single-ventricle patients (−6.82 to −8.78, $P = 0.02$). In 23 patients who had RV-coronary artery fistula at birth, RV-coronary artery fistula was regressed after palliation in 12 (52%) patients, including 8 patients who had these fistulas completely disappeared.

## DISCUSSION

Approximately 60% of patients achieved BVR with our surgical management. Together with patients having 1 + 1/2 ventricular repair, more than 70% of patients could avoid a single-ventricle repair. This study analysed the anatomical factors that influenced surgical choice in patients with PA-IVS. TV growth was not obtained by modified BTS with surgical pulmonary valvotomy; therefore, TV size at birth appeared to be a determinant for achieving BVR. Proportionate RV growth was seen in patients who had undergone BVR. However, RV growth was not seen in patients having 1 + 1/2 ventricular repair or single-ventricle repair. Another key finding was that RV-coronary artery fistula was regressed after RV decompression with our surgical strategy.

## Strategy for biventricular repair

Surgical strategy varies between institutions. Most institutions prefer single-ventricle repair versus BVR when presented with hypoplastic RV and RV-coronary artery fistula [4]. The data analysed in this study were based on patients who had been treated with our consistent surgical strategy aiming for biventricular repair in the last 20 years. Overall survival and freedom from reoperation were excellent, indicating adequacy of surgical decision-making criteria. With our strategy, BVR was achieved in 30 (60%) out of 50 patients, which is higher than previous reports [4, 11, 12].

Our surgical management is different from that previously reported. As an initial palliation, we prefer surgical pulmonary valvotomy and modified BTS without cardiopulmonary bypass. Recently, transcatheter pulmonary valve perforation has been considered a safe and effective treatment and has gained popularity [13, 14]. Hybrid strategy by performing pulmonary valve perforation via RV approach also provides an excellent result [15]. Our initial palliation has two advantages. Firstly, a left thoracotomy approach makes both surgical pulmonary valvotomy and modified BTS feasible during the same procedure without cardiopulmonary bypass. Secondly, pulmonary valvotomy can be performed under direct vision and enables control of valvotomy size. The size of the perforation is of prime importance when major RV-coronary artery fistula or RVDCC present as sudden decrease in RV pressure leads to coronary ischaemia and subsequent lethal arrhythmia and haemodynamic collapse. We aim for an RVP/LVP of 1.0 for those who have major RV-coronary fistula and we think RVDCC is not a contraindication for RV decompression as long as RVP is kept greater than 1.0.

Staged RV decompression is another key management, especially for RV-coronary artery fistula. After initial palliation, catheter pulmonary valvotomy is performed if RVP/LVP is greater than 1.0 and disproportionate RV growth was observed. With staged RV decompression, we have achieved regression of RV-coronary fistula in more than half of patients. Even if the exact mechanism is unknown, we speculate that gradual decrease in RV pressure changed the direction and amount of shunt between RV and the coronary artery, resulting in regression of fistula. Coronary ischaemia is one of the causes of mortality even after single-ventricle repair. We believe that regression of RV-coronary artery fistula attenuates long-term outcome in this entity.

Bryant *et al.* reported their surgical strategy to achieve biventricular physiology by performing RV sinus myectomy [16]. RV sinus myectomy makes more space in the RV and it makes it possible to achieve BVR in half of the patients [16]. Our RV overhaul procedure is similar in terms of resection of hypertrophied muscle to make more space in the RV cavity. A difference is that we do not resect all muscles as we think this decreases RV contraction. Bryant *et al.* showed RV growth after RV sinus myectomy, but RV function has not been well investigated. We had 2 patients out of 30 with a one-and-a-half ventricular repair after achieving BVR for liver congestion. Restrictive RV physiology might be an issue for those who had a borderline biventricular physiology with hypoplastic hypertrophied RV [17, 18]. Further follow-up will be necessary to see whether BVR is the best treatment for all patients.

## Factors associated with the feasibility of biventricular repair

Our study clearly showed that the TV size at birth is the major discriminator of feasibility of BVR. TV *z*-score at birth for those who achieved BVR was about −3 when compared with the ones who had one-and-a-half ventricular repair or single-ventricle repair around −6. During the staged palliation, TV did not grow even in patients who achieved BVR with RV volume increased. Even with high achievement of BVR shown by Bryant *et al.*, no TV growth was demonstrated, which may indicate the discrepancy in growth between TV and RV [16]. Interatrial communication under high RVP may increase right-to-left shunt, resulting in decrease in blood flow through TV.

In contrast to TV, RV volume significantly increased during palliation in patients having BVR. This proportionate RV growth is a consistent finding from previous reports [9, 10]. Experimental study showed ventricular enlargement is the result of ventricular volume overload resulting from tricuspid regurgitation [19]. Normally, patients with PA-IVS have tricuspid stenosis, but not significant tricuspid regurgitation, which does not appear to contribute to RV growth. One of the possibilities is that pulmonary regurgitation occurs after pulmonary valvotomy that can result in RV volume overload.

## Type of repair and long-term outcome

For decades BVR has been generally considered as a best surgical treatment in patients with PA-IVS. A recent clinical study that focused on the RV function after BVR demonstrated that exercise capacity and cardiac reserve function decreased with age when examining the teenage BVR patient [2]. This RV deterioration can be explained by decreased RV diastolic dysfunction caused by RV restrictive physiology with or without myocardial fibrosis [2, 20]. Sanghavi *et al.* showed a consistent finding of decreased exercise capacity with age, but there was no significant difference between BVR and single-ventricle repair [3]. A multicentre prospective study also showed reduced late functional health status and exercise capacity regardless of the type of repair [1]. Although these clinical studies provide useful clinical status information that can alter surgical decision-making for BVR, we need to carefully interpret the data as a wide spectrum of morphology of PA-IVS and institutional bias towards a different type of repair exist. We had 2 patients who had liver congestion, requiring a one-and-a-half ventricular repair, which can be explained with RV diastolic dysfunction, even without symptoms. Future assessment for clinical status will enable validation of our surgical management.

A substantially high mortality rate following single-ventricle repair has been well documented, especially in patients with RVDCC [5, 4]. RVDCC is found in ∼30% of patients with PA-IVS and patients undergo single-ventricular repair [6, 21]. Presence of RVDCC is considered as contraindication for BVR due to concern of potential coronary ischaemia. Our surgical choice for RVDCC is single-ventricle repair as suggested, but initial palliation is different from what others show. RV decompression is considered as a contraindication in patients with RVDCC [22]. However, RV decompression is performed with surgical pulmonary valvotomy with a careful monitoring. We think it would be safe to decompress RV as long as RVP is kept equal to LVP. Arrhythmia with LV dysfunction is an important issue with the existence of suprasystemic RVP, even in patients having single-ventricle repair. Therefore, lesser RVP, as long as not compromising coronary perfusion, may attenuate long-term outcome in patients having RVDCC. This study showed a regression of RV-coronary artery fistula was observed in approximately half of the patients who initially presented with it. Even though the exact mechanisms are unknown, we believe regression of RV-coronary artery fistula will help to avoid coronary-related mortality/morbidity in the long term.

## Study limitations

This study is limited by its small patient cohort and retrospective nature. The surgical decision-making was made on the basis of a single-centre policy. Long-term influence of RV-coronary fistulas on arrhythmia and RV function is not known and the effect of the multistaged RV decompression strategy on the long-term outcome should be assessed.

## CONCLUSIONS

With our surgical management that consists of multistaged RV decompression, 60% of patients achieved BVR. Together with patients having one-and-a-half ventricular repair, more than 70% could avoid single-ventricle repair. TV growth was not obtained by modified BTS with pulmonary valvotomy and following staged RV decompression; therefore, TV size at birth appeared to be a predictor for achieving BVR. Proportionate RV growth was seen only in patients having BVR. However, RV growth was not seen in patients having one-and-a-half ventricular repair. RV-coronary artery fistula was regressed in about half of the patients and we believe this will contribute to long-term outcome.

**Conflict of interest:** none declared.

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