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Comparison of Right Ventricular-Pulmonary Artery shunt and Glenn procedure for the Norwood Operation in patients with Hypoplastic Left Heart Syndrome

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Article

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Abstract

Objectives. The Norwood procedure with a right ventricular-pulmonary artery (RV-PA) shunt for hypoplastic left heart syndrome (HLHS) has been associated with improved postoperative hemodynamics and outcome. This study aimed to evaluate the effectiveness of the Norwood procedure with an RV-PA shunt, and to compare the effect of the Norwood with an RV-PA shunt to the bidirectional Glenn anastomosis (BDG) and the total cavopulmonary connection (TCPC).

Methods. This is a retrospective chart review. Between January 2004 and July 2021, 36 patients with HLHS and its variants underwent BDG: 15 patients (6.5 ± 2.3 months) underwent Norwood with an RV-PA shunt (group S); 21 patients (4.0 ± 4.1 months) underwent Norwood with BDG (group G). Nine group S (24.5 ± 6.6 months) and 17 group G (41.0 ± 15.0 months) patients underwent TCPC.

Results. Post-BDG pressure in the superior vena cava (SVC) was significantly lower in group S (13 ± 2 mmHg) than in G (18 ± 3 mmHg) ($p < 0.01$). Three patients in group S and 19 in group G underwent catheter intervention for pulmonary artery within 30 days after BDG ($p < 0.01$). The percentage of right ventricular end-diastolic volumewas significantly different (group S, 142 ± 41%; group G, 91 ± 28% ($p < 0.01$)).

Conclusions. The Norwood procedure with an RV-PA shunt enabled maintenance of low pressure in SVC and avoidance of percutaneous intervention following BDG. We recommend monitoring the changes in myocardial function post-TCPC.

Introduction

The modification of the Norwood procedure with a right ventricular-pulmonary artery (RV-PA) shunt as a substitute for the modified Blalock-Taussig shunt as the source of pulmonary blood flow was first reported by Norwood and colleagues. This technique was reintroduced by Kishimoto and colleagues as a means to ameliorate short and intermediate term challenges in the care of the Norwood patient, perioperative instability, and inter stage death [1]. The RV-PA shunt may improve perioperative stability and inter stage mortality by decreasing the amount of diastolic runoff into the pulmonary vascular bed, resulting in a higher systemic diastolic pressure and improved coronary blood flow. The RV-PA shunt has greater flow pulsatility in the pulmonary artery (PA), promoting better PA growth over time. However, a potential drawback of the RV-PA shunt includes long-term effects of the right ventriculotomy on myocardial function [2–4]. In contrast, the Norwood procedure with bidirectional Glenn anastomosis (BDG) performed at around two months after bilateral pulmonary artery banding (bPAB) is a useful alternative for hypoplastic left heart syndrome (HLHS), and can lead to a volume reduction earlier than the Norwood with an RV-PA shunt after bPAB [3, 5]. Since 2004, we have performed bPAB as the first-stage palliation in HLHS patients and in those with its variants, followed by the Norwood procedure with an RV-PA shunt at one month, or the Norwood with BDG at 3 months. The purpose of this study was to evaluate the effectiveness of the Norwood procedure with an RV-PA shunt. Our purpose also included a comparison of late hemodynamics and operative outcomes at the time of the BDG and the total cavopulmonary connection (TCPC) operations in patients undergoing the Norwood with an RV-PA shunt at one month, versus the Norwood with BDG performed at 3 months of age.

Materials And Methods

Between January 2004 and July 2021, 36 patients with HLHS and variants achieved BDG operation in a single institution. Perioperative diagnosis is shown in Table 1. There were no statistical differences between the groups for operative characteristics. We provided follow-up for the patients with HLHS treated in our center between January 2004 and July 2021. A total of 38 patients with HLHS and variants underwent bPAB directly after birth.

Table 1
Perioperative and Operative Characteristics

Anatomical Diagnosis	group S (N = 15)	group G (N = 21)	p value
AA/MA	1 (7%)	3 (14%)	0.14
AS/MS	4 (27%)	4 (19%)	0.69
AA/MS	3 (20%)	1 (5%)	0.28
MA	3 (20%)	0 (0%)	0.05
AS	0 (0%)	2 (10%)	0.51
CoA/IAA	7 (47%)	12 (57%)	0.09
Unbalanced AVSD	0 (0%)	2 (10%)	0.51
DORV/d-TGA	4 (27%)	0 (0%)	0.14

All study participants’ parents provided informed consent. The study design was exempt from ethics review board approval.

Operations

Group S included nine girls and eight boys. In group S, 17 patients underwent the Norwood with an RV-PA shunt first with a 5-mm ringed PTFE graft. Two patients died of heart failure in hospital after the Norwood with an RV-PA shunt. A total of 15 patients underwent BDG about three months later. Nine patients achieved TCPC completion and the other six patients were waiting for TCPC. (Fig. 1) All patients in group S underwent the BDG operation at a median age of 6.5 months (range, 3 to 6.8 months). At the time of TCPC, group S patients had a mean age of 24.5 months (range, 15.0 to 35.0 months), and a median weight of 10.1 kg (range, 7.6 to 13.5 kg).

Group G included 10 girls and 11 boys. In group G, 21 patients underwent the Norwood with BDG, but two patients were taken down BDG, one of which underwent the RV-PA shunt and died of sepsis in hospital, the other underwent modified Blalock-Taussig shunt and was transferred to another hospital because of family's relocating. Three patients died of heart failure in hospital and the other 17 patients achieved TCPC. At the time of TCPC, they had a mean age of 41 months (range, 20 to 74 months) and a median weight of 12.0 kg (range, 8.1 to 15.8 kg).

Mortality after BDG and before TCPC was 0% in group S and 9.5% in group G. The post-TCPC follow-up period was shorter in group S than in group G (group S: 38 ± 11 months, group G: 101 ± 23) ($p < 0.01$). No deaths occurred in group S after TCPC (mortality was 0%) and 1 patient died from myocardial infarction in group G (mortality was 5.5%).

The medical records, including echocardiographic records, electrocardiograms, cardiac catheterization reports, surgical notes, and perfusion reports were retrospectively reviewed. Complete hemodynamic and angiographic studies were performed in all patients pre and post BDG, pre TCPC, and for one year after TCPC.

The reports of these studies were reviewed to obtain information on qualitative right ventricular (RV) function, degree of tricuspid regurgitation, central or branch PA stenosis, RV-PA conduit stenosis, and the need for catheter-based or surgical intervention. A significant stenosis was defined as one that required intervention. All cardiac catheterization images were reviewed to obtain left PA and right PA measurements before their first branch point. The Nakata index (PA index) was calculated by the sum of the cross-sectional area (CSA) of right and left PA is divided by the body surface area of the patient. Echocardiography was performed in the cardiac ward. The degree of regurgitation was graded on an ordinal scale from 0 to 4 based on available transthoracic echocardiographic reports (0 = no regurgitation, 1 = trace or trivial, 2 = mild, 3 = moderate, and 4 = severe). The degree of regurgitation was estimated 1 year after TCPC operation.

Surgical Management

The TCPC was performed when the patient could stand up and walk, and body length was over 70 cm. We calculated the cardiac return in the patients who underwent TCPC on-pump arrest. The cardiac return indicated how many collateral vessels grew before the TCPC was performed.

In the postoperative period, mechanical ventilation was continued until the patient's hemodynamic and respiratory parameters were stable. Our strategy was to leave the chest tube in place until daily drainage was less than 2 ml/kg. The oxygen saturation from pulse oximetry at discharge from the hospital was recorded for all patients.

Statistical Analysis

Statistical analysis was performed using SPSS version 18.0 (SPSS Inc., Chicago, IL). Frequencies were presented as an absolute number and percentage. Continuous data are presented as mean ± SD, or median value with range, as appropriate. Mean values were compared using Student's *t*-test.

Perioperative diagnoses were compared using the χ^2-test. The Fisher exact test was used for categorical data. For all tests, *p* values of less than 0.05 were considered as significant.

Results

Table 2 shows the perioperative variables at the time of BDG and TCPC. There were fewer patients who needed PTPA within 30 days after the BDG operation in group S than in group G (13% in group S / 91% in group G) ($p < 0.01$). The age at the TCPC procedure was younger in group S than in group G (24 ± 6 months in group S / 41 ± 15 months in group G) ($p = 0.02$). Cardiac return tended to be lower in group S than in group G (22.4 ± 5.9% in group S / 30.5 ± 6.1% in group G) ($p = 0.06$). The pleural drainage time and mediastinal drainage time were much longer in group G than in group S (pleural drainage time; group S: 7 ± 3 days, group G: 23 ± 12 days) ($p < 0.01$) (mediastinal drainage time; group S: 6 ± 2 days, group G: 14 ± 8 days) ($p < 0.02$).

Table 2
Comparison of perioperative variables for BDG and TCPC

Variable	group S	group G	*p* Value
Age at BDG (months)	6 ± 2.4	4 ± 4	0.18
Body weight at BDG (kg)	5.1 ± 1.2	4.8 ± 1.1	0.91
PA plasty at BDG(%)	6(40)	5(24)	0.27
PTPA after BDG (%)	3(13)	19(91)	< 0.01
Age at TCPC (months)	24 ± 6	41 ± 15	0.02
Operative weight at TCPC (kg)	10.0 ± 1.6	12.0 ± 2.2	0.07
Cardiac return (%)	22 ± 6	31 ± 6	0.06
Mechanical ventilation (day)	0 ± 1	4 ± 47	0.21
Pleural drainage (day)	7 ± 3	23 ± 12	< 0.01
Chylothorax, n (%)	1 (11)	7 (41)	0.12
PTPA after TCPC, n (%)	3 (19)	8 (47)	0.68

Figure 2 shows the change of pressure in the superior vena cava (SVC). Post BDG, the pressure in the SVC was significantly lower in group S than in group G (group S 13 ± 2 mmHg, group G 18 ± 3 mmHg) ($p < 0.01$). However, the difference in pressure in the SVC was not significant between group S and group G before TCPC (group S 11 ± 1.9 mmHg, group G 12 ± 1.5 mmHg) ($p = 0.18$), nor one year after TCPC (group S 11.1 ± 2.2 mmHg, group G 11.8 ± 2.5 mmHg,) ($p = 0.40$).

Figure 3 shows the growth of the pulmonary artery. After BDG, the two groups did not differ significantly with respect to the right or left pulmonary artery diameter One year after TCPC, the two groups did not differ significantly with respect to the left or right pulmonary artery diameter.

Figure 4 shows the perioperative RV function. One year after TCPC, the right ventricular end-diastolic volume (RVEDV) was larger in group S than in group G (group S 136 ± 43.3% of normal, group G 90.5 ± 28.3% of normal) ($p < 0.01$). All patients had less than mild tricuspid valve regurgitation, and the degree of regurgitation was no different between the groups 1 year after TCPC operation ($p = 0.45$).

Discussion

It has been generally reported that the outcome of the Norwood operation with an RV-PA shunt for HLHS may be better than the Norwood with modified Blalock-Taussig shunt [4, 6, 7]; therefore, we performed the primary Norwood operation with an RV-PA shunt instead of a Blalock-Taussig shunt. However, it is also reported that the primary Norwood operation requires CPB at the neonate stage, which might lead to long-term neurodevelopmental dysfunctions [8]. In addition, the patients are in a status of pulmonary high flow, which leads to heart failure until the BDG is performed, and the subsequent failure of BDG or Fontan surgery [9].

In 2004, we changed our strategy to perform bPAB soon after birth, and the Norwood with BDG about three months after. However, after changing our strategy, we performed PTPA after Norwood with BDG for almost all patients because of the possible occurrence of pulmonary artery stenosis at the debanding site. Additionally, the venous pressure of the SVC at Norwood with BDG was high, which might lead to a high risk of stroke or BDG takedown.

In 2012, we changed our strategy to perform bilateral PA banding soon after birth, then Norwood with an RV-PA shunt about one month later, and the BDG at about 6 months of age. Using this strategy, we considered that the risk of immature of pulmonary artery will be reduced since this strategy could shorten the interval of bilateral PA banding and supply the pulsatile flow from the RV-PA shunt to reduce the risk of immature of pulmonary artery.

In order to assess growth of the pulmonary artery, we focused on the diameter of the bilateral pulmonary artery, PA index, the ratio of PTPA after BDG, and the trend of the venous pressure of SVC. We found no difference in the diameter of the bilateral pulmonary artery and the PA index between groups S and G. However, the ratio of PTPA after BDG was lower in group S than G, and the venous pressure of SVC at the time of BDG was lower in group S than G. These findings might support our assumption that shortening the bPAB period and supplying the pulsatile flow from the RV-PA shunt could bring up the pulmonary artery and reduce the influence of th banding site.

In order to assess RV function, we focused on the RVEDV, EF, CI, RVEDP, TR, and B-type natriuretic peptide (BNP). Because the BDG leads to volume reduction for the RV, the timing of the BDG should be earlier to prevent RV volume overload. In this study, we showed that the timing of BDG in group S was later than in group G, therefore, the RVEDV was higher in group S than G peri TCPC and one year after TCPC. We revealed that there were no differences in RVEF, CI, RVEDP, and BNP between group S and G, which indicated that there was little influence on the RV function at one year after TCPC; however, we need to consider that the myocardial function could be affected.

In group S, the patients were younger at the time of TCPC and the interval from BDG and TCPC was shorter than in group G; therefore, the group S patients were in the condition of low saturation for a shorter period than group G. Consequently, the hepatic factor could be supplied to the lung earlier in group S than in group G, which could reduce the risk of formation of collateral vessels and pulmonary arteriovenous fistulas. The fact that RVEDP after TCPC and cardiac return at the TCPC tended to be lower in group S than in group G supports our prediction. The duration of pleural drainage time was shorter in group S than G, with a lower incidence of chylothorax in group S than G. This indicates that we were able to perform the TCPC in good condition in group Scompared to group G

Limitations

This study has several limitations. First, it was a retrospective review of HLHS in a single center. Second, our small sample size limited our ability to perform statistical adjustments. Third, in this study, all patients underwent bPAB at first palliation. The optimal operative approach for performance of a primary Norwood in the neonatal period or perform the bilateral PAB before the Norwood remains unsettled.

Conclusion

Our results suggest that the Norwood with an RV-PA shunt enabled the maintenance of low pressure in the SVC and avoided PTPA following BDG. The influence on myocardial function post-TCPC needs to be closely followed.

Declarations

Statements and Declarations

All study participants’ parents provided informed consent. The study design was exempt from ethics review board approval.

Competing Interests and Funding

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Figures

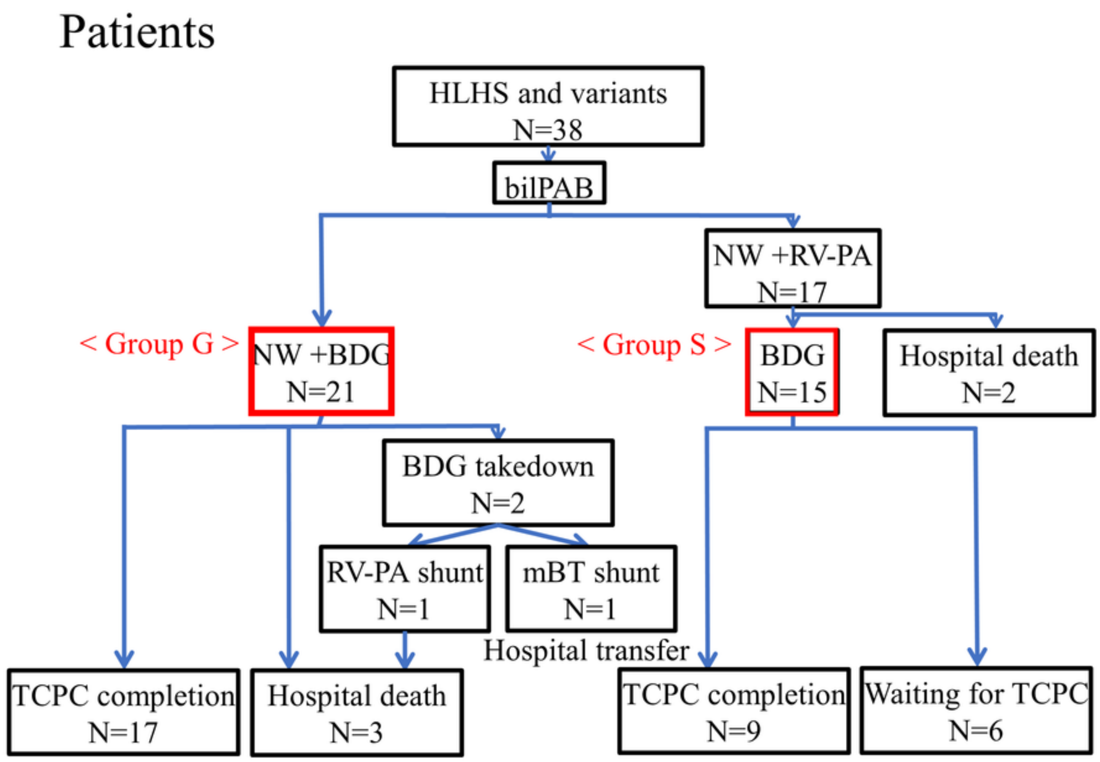

Figure 1

Flow chart of patient outcomes during the treatment period

Flow chart that includes follow-up of the patients with hypoplastic left heart syndrome that were treated in the center between January 2004 and July 2018. The follow-up period is through July 2018.

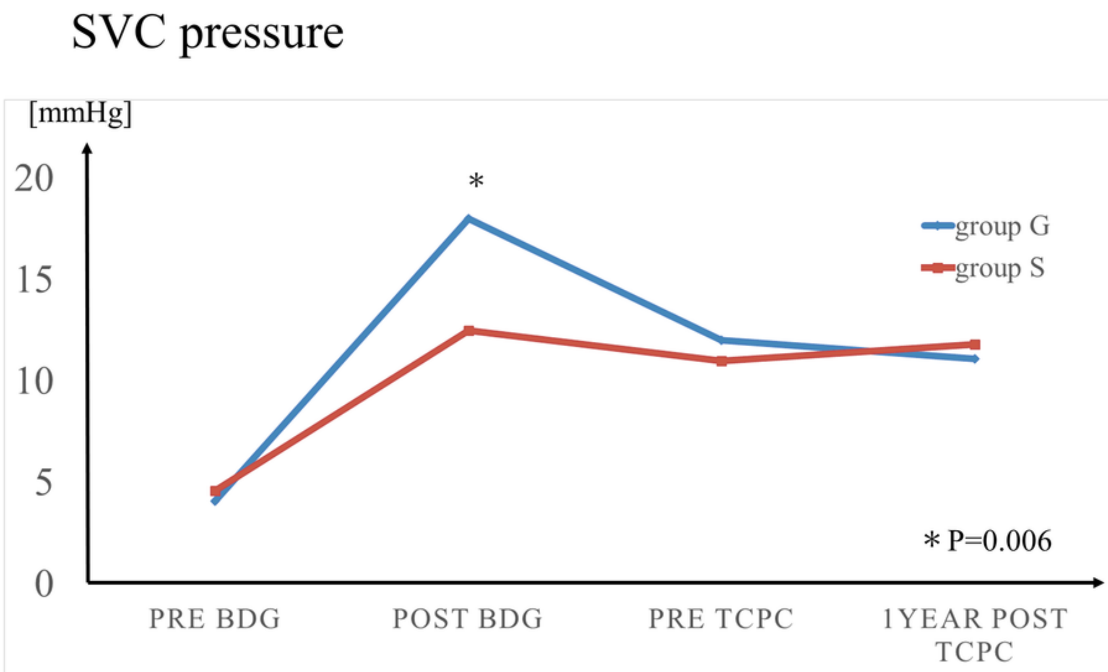

Figure 2

Superior vena cava pressure pre-and-post BGG, and pre-and-post TCPC

Post BDG, the pressure in the SVC is significantly lower in group S than in group G (group S 13 ± 2 mmHg, group G 18 ± 3 mmHg) ($p < 0.01$). However, the pressure in the SVC is not significantly different between groups G and S before TCPC (group S 11 ± 1.9 mmHg, group G 12 ± 1.5 mmHg) ($p = 0.18$) and one year after TCPC (group G 11.8 ± 2.5 mmHg, group S 11.1 ± 2.2 mmHg) ($p = 0.4$)

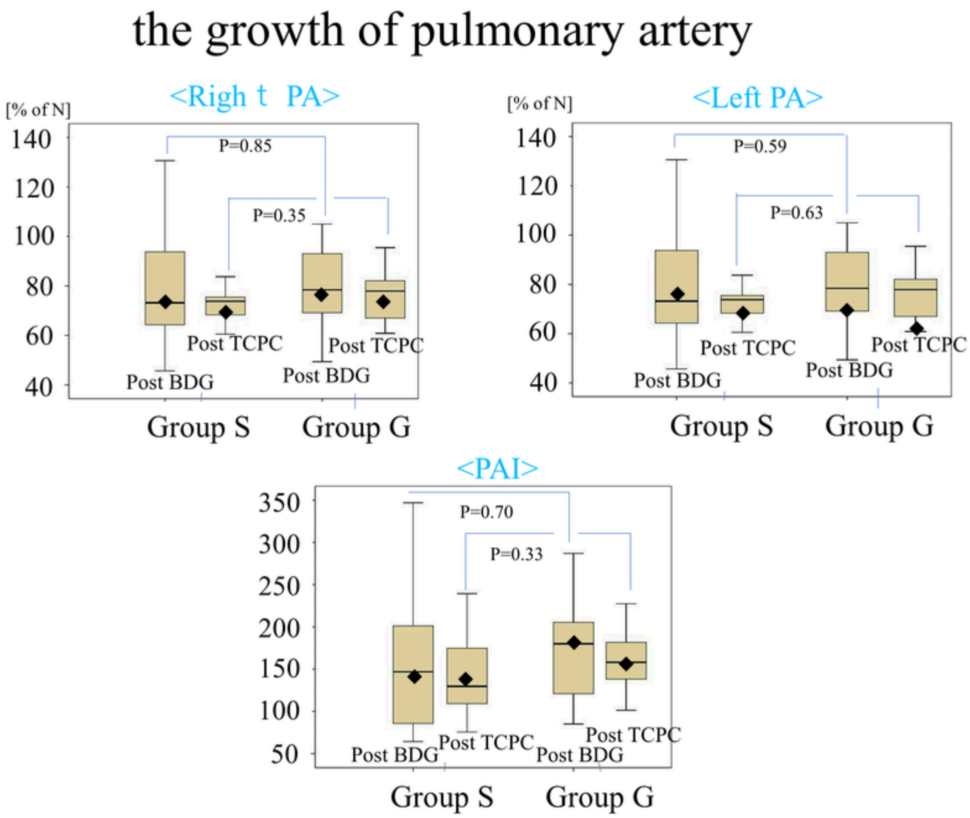

Figure 3

Pulmonary artery growth

At the BDG, the two groups do not differ significantly with respect to the right or left pulmonary artery diameter (right: group S 81 ± 14% of normal, group G 88 ± 24% of normal (p = 0.85), left: group S 89 ± 24% of normal, group G 103 ± 25% of normal (p = 0.59), nor PA index (group S 154 ± 56 mm^2/BSA, group G 199 ± 85 mm^2/BSA) (p = 0.70). One year after TCPC, the two groups do not differ significantly with respect to the right or left pulmonary artery diameter (right: group S 70 ± 10% of normal, group G 74 ± 12% of normal (p = 0.35), left: group S 65 ± 20% of normal, group G 60 ± 15% of normal (p = 0.63), nor PA index (group S 146 ± 54.4 mm^2/BSA, group G 159 ± 41.3 mm^2/BSA) (p = 0.33). Diamond mark indicates the mean value.

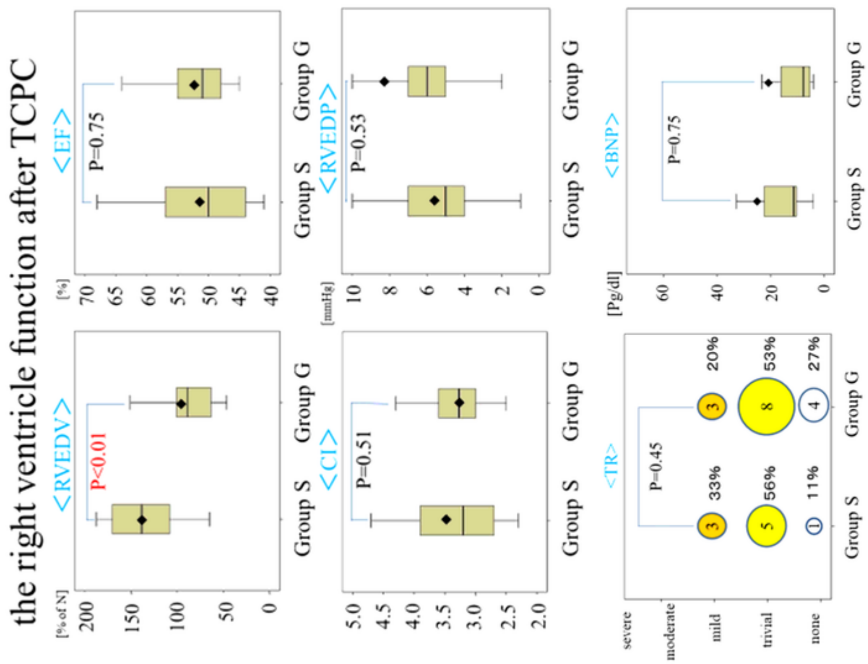

Figure 4

Right ventricular function after TCPC

One year after TCPC, right ventricular end-diastolic volume (RVEDV) is larger in group S than in group G (group S 136 ± 43.3% of normal, group G 90.5 ± 28.3% of normal) ($p < 0.01$). However, the two group do not differ significantly with respect to the ejection fraction (EF) (group S 52.3 ± 8.7%, group G 53.8 ± 11%) ($p = 0.75$), cardiac index (CI) (group S 3.5 ± 1.1, group G 3.3 ± 0.6)($p = 0.51$), right ventricular end-diastolic pressure (RVEDP) (group S 5.6 ± 2.3, group G 8.9 ± 13.2) ($p = 0.53$), and BNP (group S 27.4 ± 53 pg/dl, group G 21.3 ± 18.2 pg/dl) ($p = 0.75$). All patients are less than mild tricuspid valve regurgitation and the degree of regurgitation was no different between groups S and G at 1 year after TCPC operation ($p = 0.45$). Diamond mark indicates the mean value.