

## Cardiology

**KEYWORDS:** Hypoplastic left heart syndrome, Norwood operation, Hemi-Fontan procedure

**THE "DOUBLE DUNK" TECHNIQUE FOR A RIGHT VENTRICLE TO PULMONARY ARTERY CONDUIT FOR THE NORWOOD PROCEDURE REDUCES THE UNINTENDED SHUNT-RELATED EVENTS.**



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**ABSTRACT:**

**Background:** We describe how several modifications employed in the RVPAC for the NP may influence the interstage course, surgical or catheter-based unintended interventions and pulmonary arteries development in the HLHS cohort of patients.

**Methods:** We performed a retrospective analysis of three groups of non-selected, consecutive neonates who underwent the NP between 2011 and 2014, with different RVPAC surgical techniques employed: Group I - the left RVPAC with distal homograft cuff [N=32], Group II - the right RVPAC with distal homograft cuff [N=28], Group III - the "double dunk" right reinforced RVPAC [N=41].

**Results:** There was no intergroup difference in terms of age, weight, prevalence of aortic atresia, diameter of the ascending aorta, deep hypothermic circulatory arrest time and hospital mortality rate (9.3 vs. 14.2 vs. 7.3%, respectively) between the groups. There was a significant reduction in the numbers of catheter-based interventions during the interstage period in the third group (34 vs. 25 vs. 0%, respectively,  $p < 0.05$ ) and/or concomitant surgical interventions (17.2 vs. 4.1 vs. 2.6%, respectively). The diameter of the pulmonary arteries was the most homogenous in the third group.

**Conclusion:** The modified strategy of using the "double dunk", right reinforced RVPAC during the Norwood procedures for HLHS significantly reduces the number of catheter-based and surgical unintended shunt-related reinterventions during the interstage period. This strategy allows for a more homogenous development of pulmonary arteries before the second surgical stage.

**Introduction**

The renaissance of the right ventricle-pulmonary artery conduit (RVPAC) concept as the source of pulmonary blood flow during

the stage I Norwood procedure (NP) suggested originally by Norwood and popularized by Japanese surgeons has positively affected the early, interim, and long term mortality rates in children with hypoplastic left heart syndrome (HLHS). Potential hemodynamic advantages have been also partially explained and confirmed by computer simulations. There is also evidence that initial experience revealed an increased rate of unintended RVPAC shunt-related interventions. In our experience, similar observations and better understanding of their nature have led to several modifications aimed at eliminating this phenomenon. The objective of this study is to present our institutional experience demonstrating in what way changes in the technique of the RVPAC implantation may affect the results and modify the necessity of unintended catheter-based and surgical reinterventions after the NP.

**Materials and Methods**

The Hospital Institutional Review Board approved the study and due to its retrospective nature, the obligation to obtain informed consent was waived. The inception cohort of 101 consecutive neonates with HLHS diagnosed and operated in the Department of Pediatric Cardiac Surgery, Jagiellonian University Medical College, Krakow, Poland, between 2011 and 2014 were enrolled. The available parameters from patient records were analyzed [Table 1].

**Surgical technique****Technique of the RVPAC implantation**

The total cohort of the examined patients was divided into three groups according to the technique of RVPAC implantation during the NP. All the RVPAC were 5 mm in diameter, except 3 patients in Group III, in whom 6 mm shunts were employed when their body weight was above 3.5 kg. The implantation of the proximal aspect of RVPAC was done after choosing the major coronary weak area in the RV outflow tract; however, some distance had to be preserved to avoid damaging the pulmonary artery valve.

**Group I**

In Group I, commencing with the retracting suture, the ventriculotomy was made initially with a blade and subsequently with a 5 mm punch. After the initial experience with severe proximal stenosis in the past era, the ventriculotomy had the shape of truncated cone, with a wider diameter in the inner layer of the free wall of the right ventricle, i.e. a more extended ventriculotomy was employed. The 5 mm polytetrafluoroethylene tube was sutured perpendicularly to the surface of the right ventricle; the graft was located to the left of the ascending aorta and the distal RVPAC part was sutured to the pulmonary confluence after previous augmentation with the homograft cuff, prepared shortly before operation. Usually, the distal anastomosis was done during the cooling phase. The length of the shunt was approximately 35 mm.

### Group II

In the second group, the technique of implantation of the proximal and distal aspect of the RVPAC was identical as in Group I; however, the graft was located to the right of the ascending aorta. The decision on the site of location of the shunt with respect to the ascending "neoaorta" was at the discretion of the surgeons and there were few cases overlapping in time between Group I and II. Some anatomical configurations and the relationship between the reconstructed ascending aorta and the pulmonary confluence at that stage of project pending were thought to be inconvenient for the rightward location. The mean length of the rightward RVPAC was approximately 55 mm. (Figure 1.)

### Group III

In the third group, commercially available 5 mm shunts reinforced with rings were employed, located to the right of the ascending aorta. The Group III consists of 41 consecutive patients operated on most recently. During the rewarming phase, the limited cylindrical shape ventriculotomy was created with a 4.5 mm punch (smaller than in the previous groups). The proximal aspects of the conduit were inserted into the free wall of the right ventricle. The polytetrafluoroethylene membrane was cut sharply at the level of the first ring. For proximal fixation of the conduit, three 5-0 polypropylene sutures were introduced through the full thickness of the right ventricle (including the endocardium) and fixed only to the 3rd ring of the shunt without perforation of the shunt membrane. It means, that some part of shunt was dunked into the right ventricle at the distance equal to the thickness of the right ventricle. After tightening the sutures, one of them was used as superficial, hemostatic suture tied exclusively to the third ring, located at the level of epicardium. The distal aspect of the shunt was sutured directly to the anterior wall of the pulmonary confluence, following previous direct suturing of the pulmonary trunk opening. When the RVPAC was anastomosed directly to the PA's, an approximately 1.5 mm-long shunt membrane margin was left behind the last ring as the protruding, mechanical barrier, working as a stent and preventing the tissue ingrowth into the conduit (distal "dunk" technique). In neonates with low birth weight and a small diameter of the pulmonary confluence, the distal shunt was augmented with a homograft patch and implanted into the anterior aspect of the native pulmonary confluence. In all patients with a right location of the RVPAC (Group II and III), a free whetstone-like patch obtained from the polytetrafluoroethylene vascular shunt was left in the midline, covering the free wall of the right ventricle and the shunt crossing at some point the midline of the sternum. (Figure 2.) (Figure 3.) (Figure 4.)

### Norwood procedure

The NP was performed using deep hypothermia and circulatory

arrest. The technique of reconstruction of the aorta was persistently constant throughout the entire study. Our current preference was to reconstruct the aorta with a pulmonary homograft patch. Routine septectomy was performed through atrial cannulation place. The right atrium was not opened except in 8 cases, in which various forms of tricuspid repair were performed, 2 patients with anomalous pulmonary vein connection and 2 patients with intact interatrial septum. The hematocrit value was maintained around 30%. As soon as the postoperative bleeding has limited the continuous heparin infusion in dose 10 U/kg/h was initiated and escalated into 25 U/kg/h within the hours, than maintained as long as the central venous line was in place. When the enteral feeding become possible (usually the first postoperative day) low dose aspirin 2 mg/kg/day was started.

### The second stage operation

The hemi-Fontan procedure was the method of choice during the second stage. The children were not routinely subjected to catheterization before the second stage, except in the situation when some interventions needed to be performed or echocardiographic assessment was not diagnostic.

The pulmonary arteries diameter included in the analysis was assessed by echocardiography; the measurements included the proximal aspect (at the site of anastomosis with the conduit) and the distal segment, immediately before pulmonary artery bifurcation. The accepted values represent mean values of three measurements performed by the same cardiologist (AM) [Table 2].

### Statistical methods

The data were analyzed using the statistical software package Statistica 10 (Statsoft Inc., Tulsa, OK, USA). Standard descriptive statistics were used for the characterization of the groups. Continuous variables were expressed as means (standard deviations), and comparative analyses were performed employing the Tukey multiple comparisons of means, or the Wilcoxon signed-rank test. Nelson-Aalen analysis of cumulative hazard function of reinterventions was performed. The probability value of less than 0.05 was taken to represent a statistically significant difference between the groups.

### Results

We present the cohort of patients with HLHS divided into three groups according to various strategies of the RVPAC implantation. There was no significant difference between the groups in terms of age, weight, diameter of the ascending aorta, prevalence of aortic atresia and concomitant diagnosis at the time of the NP.

Shunt related catheter-based and surgical interventions During the inter-stage period, the patients underwent catheter-based interventional and operative procedures.

In Group I [N=32], 10 patients underwent interventional catheterizations. The interventions were mostly related to the proximal aspects of the RVPAC, except 1 case, when balloon angioplasty of the distal shunt and proximal aspects of the pulmonary arteries was performed. In 2 cases ballooning of proximal aspect was effective, whereas in another 3 cases, stent implantation into the proximal aspect of the RVPAC followed the ballooning. In 4 cases, balloon angioplasty of the proximal aspect of the RVPAC was ineffective and the patients were referred for surgical shunt exchange. Surgical interventions consisted of RVPAC exchange with concomitant pulmonary artery angioplasty. In all the patients except 1 weighing 4.9 kg, when a 6 mm shunt was applied, a 5 mm RVPAC was used for exchange.

In Group II [N=28], there were 6 catheter-based and 1 surgical intervention. In 2 cases, the proximal part of the RVPA was stented, in another 2 cases the midportion of the conduit, and in 2 cases, angioplasty involving only the proximal aspect of the PA's was employed. In 1 patient, the proximal parts of the PA's were surgically reconstructed at 2.1 months of age due to persistent hypoxia.

In Group III [N=41], no patients required PA interventions. One patient demonstrated significant tricuspid regurgitation and hypoxemia. When the baby was 2 months old and weighed 3.9 kg, it underwent interstage intervention consisting of partial annuloplasty and the RVPac exchange from 5 to 6 mm.

Therefore, there was a significant reduction in the numbers of catheter-based interventions during the interstage period in the third group (34 vs. 25 vs. 0%, respectively,  $p < 0.05$ ) and/or concomitant surgical interventions (17.2 vs. 4.1 vs. 2.6 %, respectively). (Figure 5.)

#### Non-shunt related interventions

In Group I, 4 patients underwent balloon aortoplasty and in another 2, a stent was implanted into the isthmus of the aorta. In Group II, 2 cases of balloon aortoplasty and 2 stent implantations were performed.

In Group III, 1 patient underwent an independent interventional procedure of aortoplasty, whereas in another 3, when the stenosis was borderline, balloon angioplasty was performed during the hemi-Fontan procedure in the hybrid room through the aortic cannula after completing the surgical procedures.

#### Survival rate

The 30-day mortality rate was 9.3% in Group I (3/32), 14.2% in Group II (4/28) and 7.3% in Group III. The cause of death in Group I was ventricular dysfunction in 1 patient. The second patient died due to a sudden RVPA shunt occlusion. The third patient died due to sepsis and multi-organ dysfunction. In Group II, there were 4 deaths; 1 patient presented with severe right ventricle dysfunction and while on ECMO, angiography revealed a huge left coronary artery – left ventricle fistula; 1 patient died suddenly probably due to shunt occlusion, 1 patient with dysplasia and insufficiency of the tricuspid and pulmonary valves demonstrated low cardiac output and eventually multi-organ failure. One patient developed shunt occlusion and died in spite of emergency reoperation. In Group III, there were 3 deaths: 1 involved a patient with the intact interatrial septum; 1 patient with congenital pneumonia was operated at 45 days of age and developed sepsis, and 1 patient died due to ventricular dysfunction, which did not resolve in spite of ECMO.

#### Concomitant procedures

Concomitant procedures during the stage I Norwood: In Group I: 2 patients underwent commissuroplasty of the tricuspid valve, 1 patient underwent aortic valve repair due to significant insufficiency as a consequence of balloon valvuloplasty performed in another institution before the NP. In Group II: total anomalous pulmonary vein connection supracardiac type repair was performed at the time of the NP, commissuroplasty of the tricuspid valve in 2 patients, partial annuloplasty in 2 patients. One patient with the right aortic arch needed the modified Norwood procedure and 1 patient had "situs inversus", so the child underwent the mirror image Norwood procedure. In Group III, there was another patient with "situs inversus", 3 patients had partial annuloplasty of the tricuspid valve and another 3 - commissuroplasty. The chest was left opened or it was reopened within a short time after the NP in 25% of Group I, 21.4 % of Group II and 14.6% of Group III patients ( $p > 0.05$ ).

#### Hemi-Fontan procedure

Hemi-Fontan procedure was the method of choice during the second stage of Fontan pathway in our institution.

In Group I, there was 1 interim death (unknown reason) and 3 patients had the stage II procedures performed at another center. In Group II, there was 1 interim death (shunt related) and 2 patients had stage II operations at another center. There was no interim mortality in Group III.

In total, 25 patients underwent the h-F procedures and 4 the bidirectional Glenn on the left side when the left superior vena cava was present in Group I, 21 patients in Group II (with 2 undergoing the bidirectional left-Glenn) and 38 were subjected to the hemi-Fontan (with 2 – to the bidirectional left-Glenn) in Group III. Patch enlargement of the anastomosis including the proximal aspects of the PA's was an inherent part of the surgery. The patch material was a pulmonary homograft or matrix patches. In 1 patient from Group II and 1 patient from Group III with "situs inversus", the h-F procedures were performed in a "mirror image" fashion. The h-F procedures were performed in deep hypothermia with circulatory arrest in Group I and in 3 patients of Group II. Hypothermic circulatory arrest was not used in Group III. One patient from Group III originally with the intact inter-atrial septum and very small left atrium underwent enlargement of inter-atrial communication employing the "suture-less technique" with a homograft patch. Two patients underwent the rescue hemi-Fontan at the age of 4.5 and 4.8 months because of rapidly progressing cyanosis. In both cases, the interatrial communication was enlarged (1 patient in Group I, 1 patient in Group II); there were no unplanned operations in Group III.

In 1 patient in Group III, in whom the PA's were assessed as hypoplastic during the NP, the proximal PA's were 2 mm in diameter, with 3 to 4 mm at the hilum level at the time of the h-F. The patient underwent the second stage operation with concomitant stenting of both PA's with systemic saturation improvement from 65 to 75% at discharge.

One patient died in Group I and 1 patient in Group II during the h-F procedure. The deaths were not shunt-related. There was a significant difference in the mean diameter of the proximal and distal part of the LPA between Group I and III ( $p < 0.05$ ) (Table 3).

#### Discussion

Our study demonstrates that refinements in surgical technique of the RVPac implantation may significantly reduce the need for catheter-based and surgical shunt-related reinterventions shortly after the Norwood procedure and during the interim period. The new concepts of the RVPac implantation allow for better development of the pulmonary arteries and make the surgical technique more reproducible and straightforward. This is very promising in terms of increasing evidence indicating that the RVPac has a favorable effect on long-term survival and probably no deleterious effect on the RV function and incidence of TV insufficiency in children with HLHS after the NP. The modification considerations include the technique of proximal and distal anastomosis, as well as the location and reinforcement of the shunt. The main conclusion from the SVR trial and 3-year reevaluation of this multi-center, randomized cohort of patients was a higher incidence of unintended shunt-related interventions between the first and second stages of surgical palliation. In our experience, we were able to present what kind of modifications of the RVPac may led to a significant reduction of unintended interventions before the first and second stages.

### Proximal aspect of the RVPAC

The majority of shunt-related reinterventions, especially in Group I, occurred due to stenosis of the proximal aspect of the RVPAC. The mechanisms responsible for stenosis are multifactorial: "neointima" formation, scar migration into the shunt, kinking of the conduit and thrombus formation limiting the blood flow. Some patients were able to develop subshunt stenosis, which was not seen during the operation, but was significant when the turgor and contractility of the right ventricle reached the appropriate value. Additionally, when the RVPAC was located to the right with respect to the ascending aorta (Group II), some stenosis of the midportion of the shunt was also observed. To prevent it, we resorted to the implantation of the RVPAC reinforced with rings. After some initial experience, it was clear that this type of prosthesis may be extremely helpful in managing the proximal anastomotic stenosis. Independently of Tweddell and Hasaniya, we developed our own technique of "dunking" and proximal fixation of the RVPAC with 3 single sutures with full thickness of the wall of the right ventricle, including the endocardium, and the additional superficial purse-string hemostatic suture. The role of 3 single sutures is not only to fix the shunt, but also to keep away the injured myocardium from the lumen of the RVPAC, which - we believe - may prevent the scar tissue migration into the lumen of the shunt. That kind of proximal RVPAC fixation resulted in a significantly smaller, cylinder-like (instead of cone-shaped) ventriculotomy. In our experience, the "dunk" part of the conduit is left in place during the removal of the remainder of the RVPAC. Baird and colleagues suggests that complete removal of the conduit during the second stage palliation may be helpful in preserving the regional RV function. This suggestion needs to be analyzed in the future.

### Switch from the left to right RVPAC

The significant modification of the RVPAC was a transfer of its implantation site from the left to the right side of the ascending aorta. The main disadvantage of the location of the RVPAC on the left side is a difficulty of and a long time needed to reach the pulmonary confluence, the risk of the left phrenic nerve injury and bleeding during the II stage operation. Relocation of the shunt from the left to the right side of the "neo-aorta" resulted in a much easier access to the pulmonary arteries during the second stage, especially when the proximal aspects of the pulmonary arteries were hypoplastic, requiring augmentation of the proximal aspect of both the left and right pulmonary arteries. What seemed important to us when the right RVPAC conduit option was chosen was the fact that the true native confluence was relocated to some degree to the right. The natural course of the LPA is to the left and posteriorly, which makes the segments of the pulmonary artery situated behind the ascending aorta less vulnerable to compression or at least more compliant to preparation and reconstruction. Thus, the potential stenotic segments of the pulmonary arteries are relocated to the right and easier to reconstruct during the second stage if necessary.

### Distal aspect of the RVPAC

The crucial issue regarding the distal RVPAC anastomosis is the stenosis of the proximal aspects of the pulmonary arteries ("bow tie effect"), which in observations and in computer simulations is the result of twisting of the proximal aspects of both the pulmonary arteries. The native pulmonary trunk is directed downward, whereas during the Norwood procedure, suturing the RVPAC to the opening the pulmonary trunk results in a 90 degree twist in the anterior direction because of constraint from the RVPAC polytetrafluoroethylene tube. To

eliminate this torsion effect, we started to employ primary closure of the distal opening of the pulmonary trunk, and the polytetrafluoroethylene tube is sutured directly to a 5 mm hole created with a punch in the anterior aspect of the previous pulmonary trunk. We believe that this modification is crucial in preventing twisting of the proximal PA's. Torsion of the proximal aspect of the PAs reduces directly the diameter of the vessels, but also interferes with the local shear stress, influencing the growth potential. Another crucial point is "dunking" the distal aspect of RVPAC into the pulmonary artery to prevent any tissue migration into the lumen of the shunt. Reinforced shunt also may stent the place of implantation. That modification has been previously published by Mascio and Spray. Choosing the hemi-Fontan procedure as the second stage allows for augmentation of proximal aspects of PA's routinely.

We believe that the outcome changes resulting from the implementation of the new RVPAC technique are so significant that the current cohorts of patients differ enough from previous historical groups to necessitate updating or restarting of pending projects. This will elucidate not only the development of the PA's, but also long-term consequences of the diminished RV ventriculotomy and better hemodynamic performance. So far, higher rate of reinterventions is the main known disadvantage of RVPAC technique versus Blalock-Taussig used in conjunction with the Norwood procedure. Neutralization of this effect with other technical refinements may be a sufficiently strong argument in choosing which sort of pulmonary blood flow pattern should be chosen during the NP. Also the development of PA's need to be reevaluated in most modern cohort of patients after the NP with RVPAC.

### Study limitation

The study has several limitations. The most important is the lack of randomization, and a historical perspective. The learning curve cannot be eliminated in consecutive groups of patients; however, the technique of RVPAC has replaced the BTS during the Norwood procedure in our institution in 2001.

### Conclusions

In conclusion, better understanding and several modifications of the RVPAC implantation introduced in HLHS neonates after the Norwood procedure has allowed for a significant reduction of unintended catheter-based and surgical reinterventions in the interim period; furthermore, when compiled with the second stage technique, these may allow for better preparing the pulmonary arteries to the Fontan completion. Further analyses are necessary for the assessment of this strategy in terms of long-term follow up after the Fontan completion in patients with HLHS.

### Abbreviations:

BTS – Blalock-Taussig shunt  
 h-F – hemi-Fontan procedure  
 HLHS – hypoplastic left heart syndrome  
 LPA – left pulmonary artery  
 NP – Norwood procedure  
 PA's – pulmonary arteries  
 RVPAC – right ventricle-pulmonary artery conduit  
 RPA – right pulmonary artery

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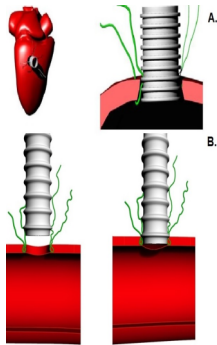
Table I: Patients characteristics at Norwood stage I operation.  
 Table II: Patient characteristics at hemi-Fontan stage II palliation.  
 Table III: Pulmonary arteries diameter assessed before hemi-Fontan operation.



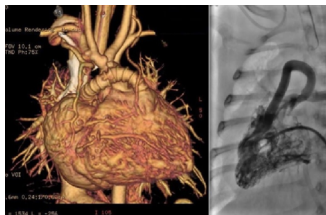
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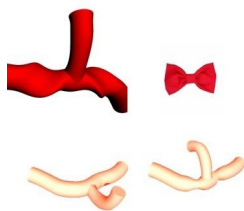
**Figure 1.** CT-angiogram showing the relationship between the ascending aorta and pulmonary arteries after the modified Norwood procedures (left-hand side), when the RVPAC is located to the right with respect to the ascending aorta. Simulation (right-hand side) suggesting that the RPA extending to the right, but also posteriorly, is less vulnerable to potential compression of the ascending aorta, and the proximal aspect of the LPA is more reachable for the reconstruction during the second stage.



**Figure 2.** A. Method of proximal fixation of the RVPAC into the free wall of the right ventricle during the Norwood procedure. B. Technique of “distal dunk” implantation of RVPAC into pulmonary artery. The sutures were placed to the ring and pulmonary wall, while the membrane is protruding into the lumen of pulmonary artery.

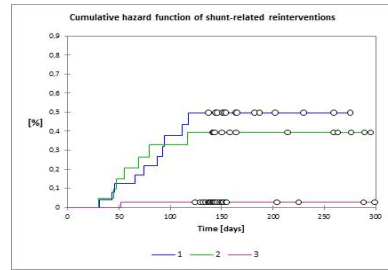


**Figure 3.** Angio-CT (on the left-hand side) demonstrating the right reinforced RVPAC conduit in children with HLHS after the modified NP. The RVPAC reaches the pulmonary confluence from the anterior direction, rather than from the inferior, as in the anatomic position of the pulmonary trunk before the NP. (on the right side) Angiogram of HLHS with “situs inversus” after the modified Norwood procedure with the reinforced “right” (on the left side) RVPAC. The diameter of all the RVPAC is invariable along the conduit.



**Figure 4.** The potential mechanism of the so called “bow tie” effect. Twisting of the proximal aspect of the pulmonary arteries caused by the RVPAC reaching the pulmonary

confluence from the anterior aspect, when anastomosed to the opening of the pulmonary trunk.



**Figure 5.** Nelson-Aalen analysis of cumulative hazard function of RVPAs-related reintervention between the Norwood operation and second stage procedures for HLHS patients. 1 – Group 1 (left RVPAC), 2 – Group 2 (right RVPAC), 3 – Group 3 (right reinforced modified RVPAC). P value =0.001 between I, II and III Group.

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