

Hypoplastic Left Heart Syndrome: the Influence of Surgical Strategy on Outcomes

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Summary

Objectives: To report a surgical strategy for the Norwood procedure in the hypoplastic left heart syndrome (HLHS) that enables short hypothermic circulatory arrest time and aortic arch reconstruction with autologous pericardium patch, and to compare the results of the modified Blalock-Taussig (mBT) shunt with the right ventricle-to-pulmonary artery (RV-PA) conduit procedures as the source of pulmonary blood flow.

Methods: Retrospective study of 71 newborns with HLHS consecutively operated between March, 1999 and February, 2006. One technique for reconstruction of the neo-aorta and two different techniques for reestablishment of the pulmonary blood flow were used: the mBT shunt in the first 37 newborns and RV-PA conduit in the last 34. Cannulation of the ductus arteriosus for arterial perfusion was the main part of the surgical strategy to reduce the hypothermic circulatory arrest time.

Results: In-hospital survival for the entire cohort was 74.64%, or 67.57% and 82.35% for the mBT shunt and RV-PA conduit groups, respectively ($p=0.1808$). Mortality rates between the first and second palliation stages were 40% and 4.4% for the mBT shunt and RV-PA conduit groups, respectively ($p=0.0054$). Hypothermic circulatory arrest times were 45.79 ± 1.99 min and 36.62 ± 1.62 min ($p=0.0012$), respectively. Late coarctation of the aorta occurred in five patients (7.2%).

Conclusions: This surgical strategy resulted in short circulatory arrest time, low mortality and favorable morphology of the neo-aorta, with low incidence of late coarctation of the aorta. The higher rate of survival to first palliation stage with the RV-PA conduit was not significant, but interstage mortality was statistically lower when compared with the modified Blalock-Taussig shunt procedure.

Key words: Mitral valve / abnormalities; aortic valve / abnormalities; mitral valve / surgery; aortic valve / surgery; Norwood procedure

Introduction

The term HLHS was introduced by Noonan and Nadas in 1958 to describe the morphological characteristics of combined mitral and aortic atresia¹. However, the concept of HLHS may be extended to cases in which the mitral and aortic valves are present but where the underdevelopment of the left ventricle makes it unable to maintain the systemic circulation².

The natural history of this syndrome, usually lethal within the first month of life³, was modified with the Norwood procedure, who published the first few successful cases in a series of children operated between 1979 and 1981^{4,5}. This procedure consists of connecting the main pulmonary artery to the aortic arch previously expanded with a polytetrafluoroethylene (PTFE) graft, thus forming a new aorta. Pulmonary perfusion is maintained by a tubular PTFE graft anastomosed in the right subclavian and right pulmonary arteries.

In 2003, Sano et al⁶ published favorable outcomes with a

modified Norwood procedure, using a PTFE graft anastomosed between the right ventricle (RV) and pulmonary artery (PA). Unlike in the systemic-to-pulmonary shunt, flow into the pulmonary artery occurs only during the ventricular systole. This facilitates the postoperative patient management because it prevents the coronary flow reduction caused by the diastolic runoff into the lungs⁶⁻⁸.

The objectives of this study are to report a surgical strategy that aims at reducing times of circulatory arrest and aortic reconstruction with autologous pericardium, and to compare the results of the modified Blalock-Taussig (mBT) shunt and right ventricle-to-pulmonary artery conduit (RV-PA conduit) procedures in the reestablishment of the pulmonary circulation.

Methods

This retrospective study, which excludes the learning period started in 1994, analyzed 71 consecutive newborns operated from 1999 to 2006 divided into two groups: one undergoing the Norwood procedure with modified Blalock-Taussig (mBT) systemic-to-pulmonary shunt, and the other undergoing the Norwood procedure with right ventricle-to-pulmonary artery conduit (RV-PA conduit), as a method to supply the pulmonary

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arterial circulation. This study was approved by the Research Ethics Committee.

Patients' characteristics are shown in Table 1. Only newborns undergoing the Norwood procedure in whom the morphologically right ventricle was the systemic ventricle were included. Therefore, we excluded newborns with anatomic variants that resulted in a morphologically left ventricle as the systemic ventricle when undergoing this procedure. Three newborns to whom surgical treatment was contraindicated due to severe myocardial dysfunction (in two), and severe hypoxia due to intact interatrial septum with subsequent neurological impairment (in one) were also excluded.

The echocardiogram of a patient with HLHS is shown in Figure 1.

Surgical procedure - As of 1999, our surgical strategy has been the same in relation to aortic reconstruction, cardiopulmonary bypass and myocardial protection procedures. However, there were differences regarding the source of pulmonary perfusion, and in the first 37 newborns PTFE grafts anastomosed in the right subclavian and pulmonary arteries (modified Blalock-Taussig) were used. As of 2003, conduit anastomosis in the right ventricle and pulmonary artery (RV-PA Conduit) was adopted and used in the last 34 cases.

Cardiopulmonary bypass - Through a median sternotomy, the ascending aorta, aortic arch, and initial portion of the descending aorta are fully exposed. Cardiopulmonary bypass (CPB) is established with cannulation of the ductus arteriosus for arterial perfusion and cannulation of the right atrial appendix for venous drainage. The arterial cannula is advanced through the ductus arteriosus down to the descending aorta; a tourniquet is tightened around it, thus enabling a great part of the operation to be performed without circulatory arrest.

After proximal section of the ductus arteriosus, the pulmonary artery is divided close to its bifurcation, thus separating the distal stump connected to the pulmonary branches from the proximal stump that will be part of the neo-aorta.

Preparation of the distal pulmonary artery is completed with the suture of the remaining ductus arteriosus and anastomosis of a PTFE conduit after a small transverse plication. The diameter of this conduit which is further connected to the right subclavian artery was 3 or 4 mm (mBT group), and 4 or 5 mm, when connected to the right ventricle (RV-PA Conduit group).

Construction of the neo-aorta - While temperature is

gradually cooled down to 16° C, the ascending aorta is clamped and sectioned, and the cardioplegic solution is infused in the coronary arteries. The proximal portion of the ascending aorta is anastomosed in the lateral side of the pulmonary artery, and the reconstruction of the neo-aorta is started. Only as of this stage is the hypothermic circulatory arrest started, and the arterial cannula removed from the distal portion of the ductus arteriosus. The remaining ductal tissue is totally removed from the aorta, and the resulting opening is extended proximally to the aortic arch, and distally to the descending aorta. A glutaraldehyde-treated autologous pericardial patch is used to enlarge the descending aorta and the aortic arch, which is connected to the pulmonary artery to complete the neo-aorta. The arterial cannula is again placed in the pulmonary artery (neo-aorta). The perfusion is restarted, and the temperature is kept low. CPB is interrupted again for two to three minutes for atrial septal defect enlargement and

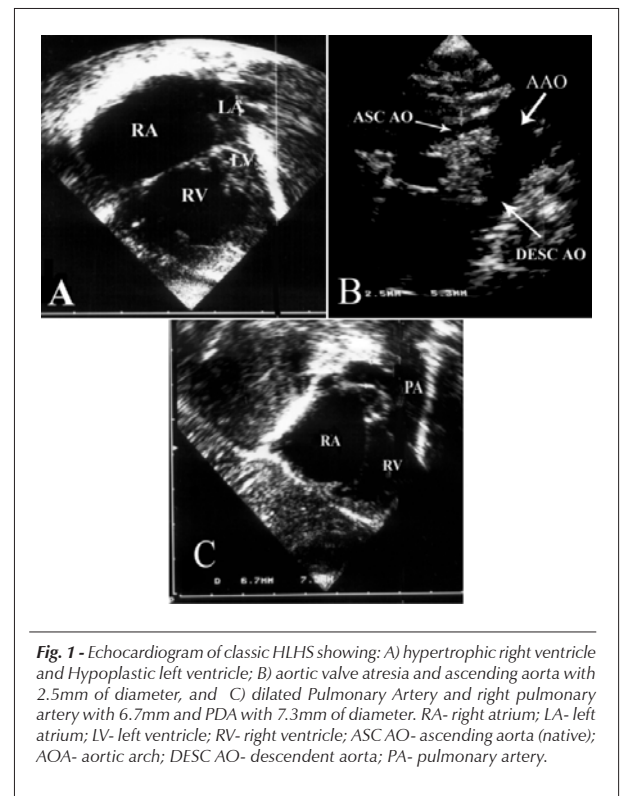


Fig. 1 - Echocardiogram of classic HLHS showing: A) hypertrophic right ventricle and Hypoplastic left ventricle; B) aortic valve atresia and ascending aorta with 2.5mm of diameter, and C) dilated Pulmonary Artery and right pulmonary artery with 6.7mm and PDA with 7.3mm of diameter. RA- right atrium; LA- left atrium; LV- left ventricle; RV- right ventricle; ASC AO- ascending aorta (native); AOA- aortic arch; DESC AO- descendent aorta; PA- pulmonary artery.

Table 1 - Postoperative patients characteristics

	mBT	RV-PA conduit	p
No. of patients	37	34	
Age (days)	9.216 ± 2.07	7.667 ± 1.428	0.5588(NS)
Gender (male)	48%	56%	0.635(NS)
Weight (g)	3101 ± 109	3009 ± 68.5	0.497(NS)
Ascending aorta diameter (mm)	2.713 ± 0.219	2.923 ± 0.220	0.501(NS)

MBT- modified Blalock-Taussig; RV-PA- right ventricle-to-pulmonary artery; NS- non-significant.

tricuspid annuloplasty, when necessary.

Reestablishment of the pulmonary circulation - In the first 37 patients of this series (mBT group), the 3 to 4-mm-diameter PTFE conduit previously sutured to the pulmonary artery was anastomosed in the right subclavian artery or in the brachiocephalic artery. In the RV-PA conduit group, the proximal portion of the PTFE conduit was anastomosed in the RV, where a small ventriculotomy had been previously performed.

Generally, the heart beats return spontaneously by gradually rewarming the patient. The thorax can be kept opened, with sternal retraction, using a latex plaque sutured to the skin, which is further covered with a sterile plastic patch.

Postoperative management - In the postoperative management, we sought to maintain the delicate balance between the vascular resistances (pulmonary and systemic). Ventilation was aimed at achieving a pH of 7.40, PaO₂ of 40 mmHg, and PCO₂ of 40 mmHg (40/40/40 balance). Systemic arterial O₂ saturation was maintained between 75% and 80%.

The inotropes most frequently used were milrinone (0.35 to 0.75 µg/kg/min) and dopamine (5 to 15 µg/kg/min) used in 90% of the patients. No significant difference was observed between the two groups. Epinephrine (0.02 to 0.06 µg/kg/min) or norepinephrine were indicated in approximately 30% of the cases due to significant hypotension (mean blood pressure lower than 40 mmHg).

Routine use of heparin in the postoperative period aimed at preventing PTFE graft thrombosis. Definitive chest closure was performed within 24-48 hours, after cardiac compensation, and was postponed to up to six days in some cases.

Definitions and statistics - In-hospital mortality was defined as every death occurring up to 30 days after surgery, or in children who were never discharged from the ICU after surgery. Interim mortality was that occurring in children who had been discharged from hospital, but who did not reach the second palliation stage. Numeric variables were expressed as mean and standard deviation and compared using the Student's t test. For comparisons of mortality between the groups A and B the Fisher's exact test was used.

Results

Overall survival of the 71 patients with HLHS undergoing the Norwood procedure was 74.64%, that is, 53 survivors.

Images from the two surgical techniques are shown in Figure 2.

The mBT and RV-PA conduit groups were similar in relation to age, weight, gender and diameter of the ascending aorta,

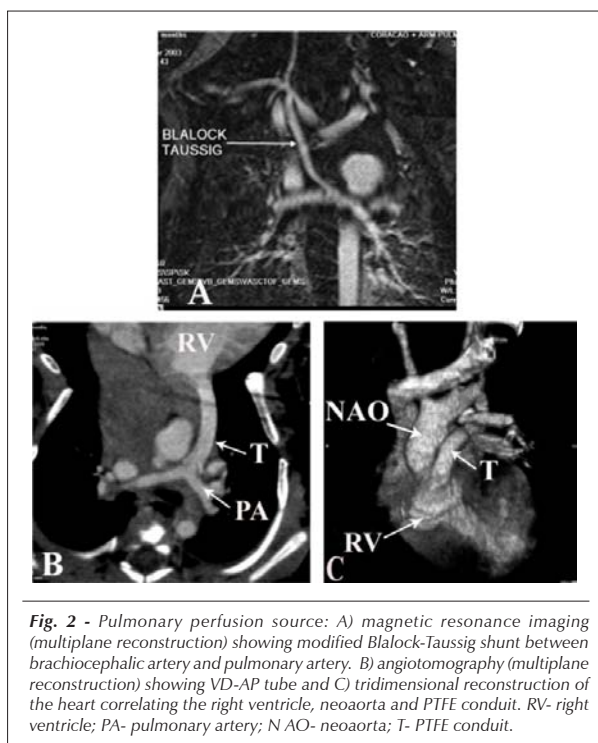


Fig. 2 - Pulmonary perfusion source: A) magnetic resonance imaging (multiplane reconstruction) showing modified Blalock-Taussig shunt between brachiocephalic artery and pulmonary artery. B) angiogram (multiplane reconstruction) showing VD-AP tube and C) tridimensional reconstruction of the heart correlating the right ventricle, neo-aorta and PTFE conduit. RV- right ventricle; PA- pulmonary artery; N AO- neo-aorta; T- PTFE conduit.

as shown in Table 1.

Cardiopulmonary bypass time was shorter in the mBT group because the anastomosis of the PTFE conduit in the subclavian artery was performed during patient rewarming. Hypothermic circulatory arrest time was shorter in the RV-PA Conduit group, as shown in Table 2.

Of the 37 newborns in whom the modified Blalock-Taussig-type systemic pulmonary anastomosis was used, 25 survived (67.57%), and of the 34 newborns undergoing the right ventricle-to-pulmonary artery connection (RV-PA Conduit) as a method to supply the pulmonary artery circulation, 28 survived (82.35%), as shown in Figure 3.

Survival in the period between the first and second palliation stages that corresponds to children undergoing the Glenn procedure is shown in Figure 4. Five patients undergoing the RV-PA Conduit technique await the second-stage procedure. Studies conducted before the second palliation stage show favorable morphology of the aorta in the great majority of patients (Figure 5).

Development of late coarctation of the neo-aorta, which occurred in five patients (7.2%) in the early phase of this series, was resolved with surgical intervention via lateral thoracotomy or balloon-catheter dilatation with favorable outcomes.

Table 2 - Extracorporeal circulation

	mBT	RV-PA Conduit	p
CPB time (min)	114.0±4.39	136.3 ±5.166	0.0017
Circulatory arrest time (min)	45.79 ± 1.99	36.62 ± 1.62	0.0012

MBT- modified Blalock-Taussig; RV-PA- right ventricle-to-pulmonary artery; CPB- cardiopulmonary bypass.

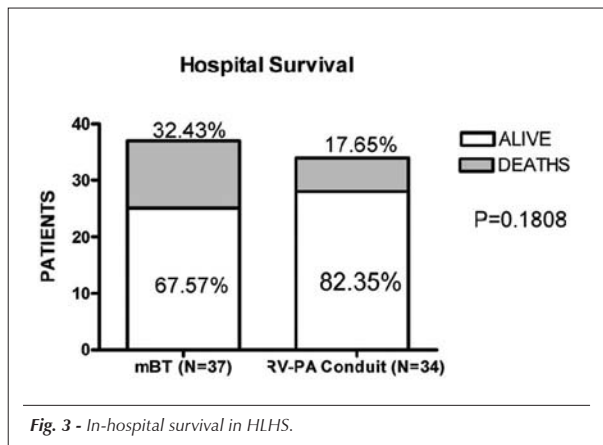


Fig. 3 - In-hospital survival in HLHS.

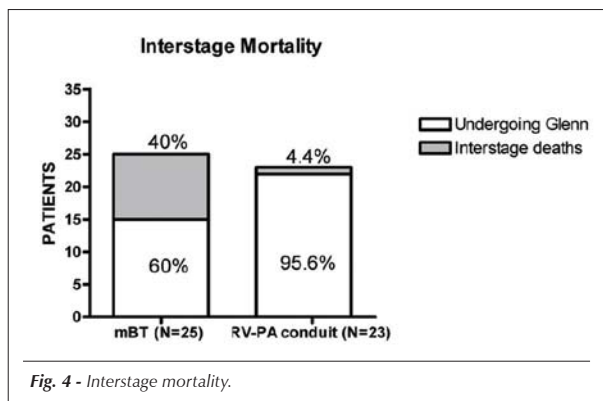


Fig. 4 - Interstage mortality.

Discussion

The favorable overall survival of 74.64% for HLHS patients undergoing the first stage Norwood procedure, or 67.57% in the mBT group in which the modified Blalock-Taussig procedure was used, and 82.35% in the group where the RV-PA conduit was the source of pulmonary perfusion, was achieved after an initial series of 12 newborns operated between 1994 and 1998, who were not included in this study. Of those first 12 patients, only four survived and better outcomes were obtained by reviewing those cases and using concepts from the literature, thus reaching the surgical strategy described here, which enabled satisfactory surgical repair with emphasis on myocardial protection and reduction of circulatory arrest time, in addition to improving the postoperative management. We sought to incorporate the great evolution of concepts and outcomes of the international experience. In the 1980's, few centers performed the Norwood procedure successfully. Publications reported in-hospital survival rates ranging between 42 and 66%, with most of the deaths occurring in the first 24 hours after surgery due to low cardiac output. Long-term survival ranged between 21 and 44%⁹⁻¹¹.

Data from the Congenital Heart Surgeons Society Data Center in a prospective study conducted in 29 institutions showed survival rates of 72%, 60%, and 54% at one month, one year, and 5 years after staged repair, respectively. A total of 710 children with HLHS were analyzed between 1994 and 2000.

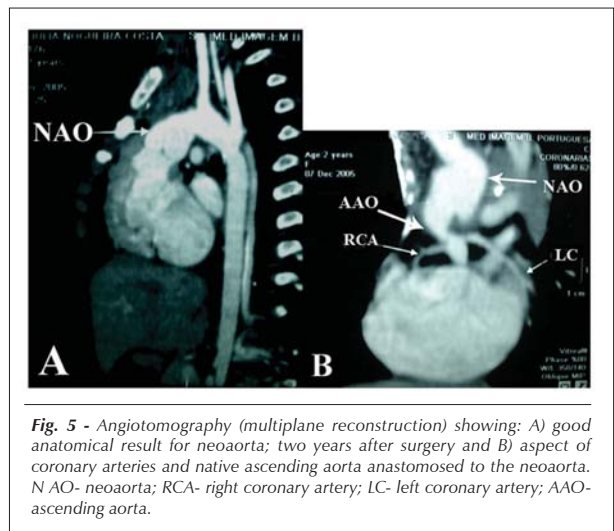


Fig. 5 - Angiotomography (multiplane reconstruction) showing: A) good anatomical result for neoaorta; two years after surgery and B) aspect of coronary arteries and native ascending aorta anastomosed to the neoaorta. NAO- neoaorta; RCA- right coronary artery; LC- left coronary artery; AAO- ascending aorta.

Risk factors for death after the first palliation stage included patient-specific variables (low birth weight, small ascending aorta, and older age at Norwood procedure), institutional variables, and procedural variables (shunt originating from the aorta, long circulatory arrest time, and manipulation of the ascending aorta). Risk factors for death after the second palliation stage included younger age at cavopulmonary shunt and need for atrioventricular valve repair¹¹.

Tweddell et al¹² reported improved in-hospital survival from 53% in the operations performed between 1992 and mid-1996 to 93% in a series of 81 patients undergoing the Norwood procedure with modified Blalock-Taussig shunt between July, 1996 and October, 2001. The use of anti-inflammatory drugs (aprotinine) was shown to improve survival rates by univariate analysis, whereas continuous monitoring of central venous oxygen saturation, use of phenoxybenzamine as vasodilator, and the shorter duration of circulatory arrest were identified as factors of survival rate improvement by multivariate analysis. However, we should point out that this series was not exclusively comprised of newborns with classic HLHS, but included 23% of anatomic variants that resulted in left ventricle as the systemic ventricle.

The team of the Children's Hospital, Boston, conducted a study comparing the outcomes of the first stage Norwood procedure in HLHS with the same procedure applied to other malformations. They demonstrated that operative and one-year survival in the presence of a nonhypoplastic left ventricle (96.3% and 88.9%, respectively) was significantly higher than survival rates in HLHS (64.7% and 52.7%, respectively)¹³.

Although the most recent data reflects a tendency of continuous improvement of results in world centers of excellence in Pediatric Cardiology with the Norwood procedure in HLHS, mortality rates are still high, especially in less experienced institutions. Using data from hospital members of the Child Health Corporation of America, Checchia PA et al¹⁴ studied 1105 newborns from 1988 to 2001 admitted with the diagnosis of HLHS, and observed that 801 had undergone the Norwood procedure, 39 heart transplantation, and the remaining 265 (24%) did not undergo surgical intervention.

The newborns undergoing the Norwood procedure had an in-hospital survival of 68% (546/801). The four institutions with greater number of operations had 78% of survival and the other 25 achieved, on average, a 59% in-hospital survival.

Recent clinical investigations show concern with risks of cognitive, neuromotor, and psychosocial problems that could occur after these operations¹⁵. Preexisting cyanosis, heart failure, and central nervous system abnormalities that may accompany HLHS, as well as extracorporeal circulation and hypothermic circulatory arrest used in the staged procedure may cause neurological damage. Some groups have used selective low-flow cerebral perfusion as an alternative to full circulatory arrest^{12,16}.

The results obtained with the strategy described are similar to those of the international literature, considering that only newborns with HLHS were included in this series. Additionally, the size of the ascending aorta and the presence of tricuspid regurgitation were not exclusion criteria, and the Norwood procedure was contraindicated in less than 5% of the cases. Also, it allows short hypothermic circulatory arrest times, thus probably contributing to the higher survival rate to be accompanied by a lower risk of nervous system damage.

The use of glutaraldehyde-treated autologous pericardium is a new modification in the aortic arch and descending aorta enlargement procedure in HLHS. This was a simple solution that resulted in a favorable morphology for the neo-aorta and low incidence of postoperative coarctation. One of the most frequently employed techniques for aortic arch reconstruction includes the use of pulmonary homografts, but they are expensive, difficult to obtain, and immunologic sensitization may lead to further problems if heart transplantation is required^{12,17,18}.

Direct aortic anastomosis is technically more complex and cannot always be performed, although no difference is observed in relation to homologous grafts as for the need for aortic arch reintervention¹⁹⁻²².

The recent popularization of the RV-PA made the postoperative course more stable, although the impact on survival remains controversial. In our experience, it showed better outcomes in relation to in-hospital survival and interstage mortality, although statistical significance had been observed only in relation to the latter variable, using the Fischer's exact test. Subjectively, the postoperative management was easier. Sano et al⁶ reported survival of 84% (61/73) for patients undergoing the Norwood procedure with this technique in three centers in Japan between 1998 and 2002. They identified surgeons' experience and mechanical ventilation prior to the operation as risk factors. The RV-PA conduit provides forward flow into the pulmonary arteries only during systole, and may have a larger diameter and lower incidence of acute occlusion than mBT, but allows reverse flow during diastole, thus leading to ventricular volume overload. Additionally, the need for right ventriculotomy theoretically increases the risk of cardiac arrhythmias and reduced ventricular function in these patients with single-ventricle physiology. The modified Blalock-Taussig shunt, in turn, provides forward flow through the conduit during all the cardiac cycle, and it is controversial whether it provides greater pulmonary artery growth^{7,23}, but

the lower diastolic pressures in the aorta affect the coronary blood flow^{7,8} and, possibly, the cardiac function due to low subendocardial perfusion.

Sano et al's conclusion that with the RV-PA conduit better outcomes could be achieved by many surgeons was based on a case series report⁶. Other reports showing improvement of survival with the RV-PA conduit also used historical series of patients with mBT as controls^{7,8, 23}.

Studies that showed lack of impact on survival in relation to the type of conduit analyzed very small newborn populations, in addition to comparing non-contemporary groups^{24,25}. Patients with the RV-PA conduit had higher mortality rates and indication for transplant after the second palliation stage. Tabbutt S et al's comparative study with contemporary patients showed no differences in mortality between the RV-PA and mBT groups, but it has the limitation of not being randomized and having some selection biases such as: inclusion of patients with systemic left ventricle, theoretically with better prognosis, only in the mBT group, and preferential inclusion of patients with aortic valve atresia in the RV-PA group²⁶.

Despite the favorable outcomes currently achieved with the first-stage mBT procedure, a 4% to 15% late death risk still persists before the second palliation stage^{11,27-29}, although the use of a home surveillance program could reduce this risk³⁰. The use of a RV-PA conduit reduced interstage mortality among Norwood procedure survivors in a study published by Pizarro, in which 40 out of 46 Blalock-Taussig group patients and 49 out of 50 RV-PA conduit group patients achieved the hemi-Fontan stage with interim mortality rates of 13% and 2%, respectively³¹. However, the groups that showed excellent outcomes with the BT procedure had identical outcomes with Sano's modification. It may indicate that this modification is associated with improvement of outcomes in the groups that had mortality rates less than ideal, perhaps because the RV-PA conduit facilitates postoperative management. Further randomized studies are necessary to corroborate the possible advantages of this procedure.

The major limitation of the present study regarding to the analysis of comparative outcomes between the two procedures is that the groups are not contemporary. Although the groups have similar clinical characteristics and postoperative care, it is undeniable that the last group benefited from the greater team's experience. Also, the practice of keeping the majority of the patients hospitalized until the second palliation stage was progressively adopted, thus benefiting the most recent patients concerning to interstage survival.

Conclusion

The surgical strategy used in the Norwood procedure resulted in short circulatory arrest time, low mortality of newborns with hypoplastic left heart syndrome, and we obtained favorable morphology of the neo-aorta and low incidence of coarctation of the aorta with the use of glutaraldehyde-treated autologous pericardium to enlarge the aortic arch.

The higher survival rate with the right ventricle-to-pulmonary artery conduit was not significant when compared with the modified Blalock-Taussig procedure. However, interstage mortality was statistically lower in the first group.

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