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Hypoplastic left heart syndrome: the report of a surgical strategy and comparative results of Norwood x Norwood-Sano approach

Síndrome do coração esquerdo hipoplásico: estratégia cirúrgica e comparação de resultados com técnicas de Norwood x Sano

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Abstract

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

Method: **A retrospective study was performed of 78 newborns consecutively operated on between March 1999 and June 2006. One technique to reconstruct the neoaorta and two different techniques, to reestablish the pulmonary blood flow, were utilized - mBT shunts in the first 37 newborns and RV-PA conduits in the last 41. Cannulation of the ductus arteriosus for systemic perfusion was the main part of the surgical strategy used to reduce the hypothermic circulatory arrest time.**

Results: **In-hospital survival for the entire cohort was 74.35%, or 67.57% for the mBT shunt and 80.49% for the RV-PA conduit Groups (p=0.21). Hypothermic circulatory arrest times were 45.79 ± 1.99 minutes and 36.8 ± 1.52 minutes (p=0.0012), respectively. Mortality rates between first and second stages were 40% for the mBT shunt and 6.9% for the RV-PA conduit Groups (p=0.007). Late coarctation of the aorta occurred in five patients. A comparison of the actuarial survival curves (Kaplan-Meier) showed that the results were better with the VD-AP conduit (p=0.003).**

Conclusions: **This surgical strategy resulted in a short circulatory arrest time, low mortality and a low incidence of aortic coarctation. Although the higher rate of survival in the first palliation stage with the RV-PA conduit was not significant, the lower interstage mortality and superior medium-term survival in the RV-AP Group were statistically advantageous.**

Descriptors: **Hypoplastic left heart syndrome. Circulatory arrest, deep hypothermia induced. Heart defects, congenital. Aortic valve stenosis.**

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Resumo

Objetivos: **Relatar estratégia cirúrgica na síndrome do coração esquerdo hipoplásico (SCEH), que possibilita tempo curto de parada circulatória hipotérmica e reconstrução do arco aórtico com pericárdio autólogo. Comparar os resultados das técnicas de restabelecimento da circulação pulmonar: anastomose Blalock-Taussig modificado e tubo ventrículo direito para artéria pulmonar.**

Método: **Estudo retrospectivo de 78 neonatos com SCEH, consecutivamente operados entre março de 1999 e junho de 2006. Foi usada a mesma técnica de reconstrução da neoaorta e duas técnicas diferentes de restabelecimento da circulação pulmonar: anastomose BTm, nos primeiros 37 neonatos, e tubo VD-AP, nos últimos 41. A canulação do canal arterial para a perfusão sistêmica foi a parte principal da estratégia cirúrgica para diminuir o tempo de parada circulatória hipotérmica.**

Resultados: **A sobrevida imediata foi de 74,35%, sendo de 67,57% no grupo BTm e de 80,49% no grupo Tubo VD-AP (p=0,21). O tempo de parada circulatória hipotérmica foi de 45,79%±1,99 min e 36,8±1,52 min (p=0,0012) e a mortalidade entre o primeiro e segundo estágios foi de 40% e 6,9%, respectivamente, nos grupos BTm e Tubo VD-AP (p=0,007). Coarctação da aorta ocorreu em cinco pacientes. A comparação das curvas de sobrevida (Kaplan-Meier) mostrou melhor resultado com o Tubo VD-AP (p=0,003).**

Conclusões: **Essa estratégia cirúrgica resultou em tempo curto de parada circulatória, baixa mortalidade e baixa incidência de coarctação aórtica. Embora o melhor resultado imediato com o Tubo VD-AP não tenha sido significante, a menor mortalidade interestágios e a maior sobrevida em médio prazo no grupo VD-AP foram vantagens que atingiram significância estatística.**

Descritores: **Síndrome do coração esquerdo hipoplásico. Parada circulatória induzida por hipotermia profunda. Cardiopatias congênitas. Estenose da valva aórtica.**

INTRODUCTION

The natural history of hypoplastic left heart syndrome (HLHS), which is generally fatal within the first month of life [1], was changed by the ground-breaking work of Norwood et al., who published the first successful cases in a series of children surgically treated between 1979 and 1981 [2,3]. This procedure consists of connecting the pulmonary artery trunk to the previously extended aortic arch, thereby forming a new aorta. Pulmonary perfusion is supplied by a polytetrafluoroethylene (PTFE) tubular graft anastomosed in the right subclavian and right pulmonary arteries.

Reconstruction of the aortic arch, initially performed using PTFE [3], can also be achieved with other prosthetic materials (Dacron, bovine pericardium) or without using additional materials [4,5], however there is a high incidence of re-coarctation [6,7]. Homologous pulmonary grafts are the most commonly used materials for reconstruction however they induce the formation of antibodies [8] and are very expensive.

In 2003, Sano et al. [9] reported good results with a modification of the Norwood technique using a PTFE graft anastomosed between the right ventricle (RV) and the pulmonary artery (PA), allowing blood flow to the pulmonary artery only during ventricular systole. This facilitates treatment of the patient in the postoperative period, because it avoids a drop in the coronary flow to the lung during diastole caused by the “steal phenomenon” [9-11].

The objectives of this study are: 1 – To report a surgical strategy for HLHS that uses autologous pericardium for the reconstruction of the aortic arch and that requires a short duration of hypothermic circulatory arrest and 2 – To compare the immediate and medium-term results of modified Blalock-Taussig anastomosis (mBT or classical Norwood Group) and of the right ventricle to pulmonary artery tube (RV-PA Tube or Norwood-Sano Group) as techniques to reestablish pulmonary circulation.

METHOD

This prospective study, which excluded the development period initiated in 1994, reports on 78 consecutive newborn babies, operated from 1999 to 2006, divided into two groups: one submitted to the mBT-type Norwood operation with systemic-pulmonary anastomosis (classical Norwood) and the other submitted to the Norwood operation utilizing a RV-PA tube (Norwood-Sano) to supply the pulmonary arterial circulation. This research was approved by the Ethics Research Committee.

Echocardiographic data, the immediate mortality and medium-term evolution of the two groups were analyzed, with follow-ups of all patients being carried out.

The patients’ characteristics are illustrated in Table 1. Only newborn babies submitted to the Norwood surgery in whom the right ventricle was the systemic ventricle were included. Thus, anatomical variations, which, after being submitted to the same procedure resulted in the anatomically left ventricle being the systemic ventricle, were not considered. Three newborn babies for whom surgical treatment was contraindicated were also excluded: two due to severe myocardial dysfunction and one for neurological problems due to severe hypoxia caused by entire atrial septum in HLHS.

Operative technique

The surgical strategy of the group from 1999 has been the same in relation to the aorta reconstruction techniques, cardiopulmonary bypass (CPB) and myocardial protection. However, there was a difference in respect to the source of pulmonary perfusion. In the first 37 newborn babies, PTFE grafts were anastomosed in the right subclavian and pulmonary arteries (mBT). From 2003, for the last 41 cases, a tube was anastomosed between the RV and the PA.

Cardiopulmonary bypass: The ascending aorta, the aortic arch and the beginning of the descending aorta are accessed by median sternotomy. CPB is established by cannulation of the arterial canal for arterial perfusion and by cannulation of the left atrial appendage for venous drainage. The arterial cannula is advanced through the arterial canal to the descending aorta and a tourniquet is tightened around it, allowing most of the procedure to be completed without circulatory arrest. After the proximal sectioning of arterial canal, the pulmonary artery is split near to the bifurcation, separating the distal stump connected to the pulmonary branches and the proximal stump, which will be part of the neoaorta. The preparation of the distal pulmonary artery is completed with the suturing of the remaining arterial canal and with the anastomosis of a PTFE tube after a small crosswise plicature. The diameter of this tube, when subsequently connected to the right subclavian artery, was 3 mm or 4 mm (mBT Group or classical Norwood), and 4 mm or 5 mm when connected to the right ventricle (RV-PA Tube Group or Norwood-Sano).

Neoaorta construction: While the temperature is gradually reduced to 16ºC, the ascending aorta is occluded using a clip and sectioned with cardioplegic solution being injected into the coronary artery. The proximal portion of the ascending aorta is anastomosed in the lateral face of the pulmonary artery trunk, initiating neoaorta construction. At this stage the CPB is interrupted and the arterial cannula is removed from the distal end of the arterial canal. The remaining ductal tissue is removed entirely and the resulting opening is extended proximally to the aortic arch and distally to the descending aorta. An autologous pericardium graft treated in glutaraldehyde is used to extend the descending aorta and the aortic arch, which is anastomosed to the pulmonary trunk, completing the neoaorta. The arterial cannula is again placed in the pulmonary trunk (neoaorta). CPB is restarted to maintain the temperature low. Again CPB is interrupted for two to three minutes to enlarge the interatrial communication and perform tricuspid annuloplasty, when necessary.

Restoration of the pulmonary circulation: For patients in the mBT Group, the 3-mm to 4-mm PTFE tube already sutured in the pulmonary artery, is anastomosed in the right subclavian artery or brachiocephalic trunk. In the RV-PA Tube Group, the proximal part of PTFE tube is anastomosed to the RV, where a small ventriculotomy had been performed. In general, the heartbeats returned spontaneously with the gradual heating of the patient. Closure of the chest can be delayed with sternal separation, using a sheet of rubber latex sutured to the skin, which is subsequently covered with a sterile plastic adhesive. Definitive closure may be performed using wire within 24 to 48 hours after cardiac compensation, but in some cases closure needs to be delayed for up to eight days.

The second stage of HSLS consisted of a partial cavo-pulmonary shunt (Glenn operation), which was achieved, in most cases, by ministernotomy, as has been described elsewhere [12].

Pre- and post-operative care

The therapeutic interventions in the preoperative period aimed at keeping the arterial canal patent and achieving a balance between the systemic flow and the pulmonary flow (QP/QS ratio = 1). This balance was identified by O_2 saturation between 75 and 85%, $PaCO_2$ of 40 to 50% and no metabolic acidosis (pH 7.4) as measured by arterial gasometry.

- Prostaglandin infusion: The dose was initiated at 0.05 to 0.1 μg/kg/min, immediately after diagnosis, aiming at maintaining the arterial canal open;
- Ventilation: FIO_2 low;
- Guarantee adequate caloric energy supply;
- Inotropic support;
- Evaluate dysfunction of other organs;
- Use glycocorticoids if patients are unstable (Dopamine > 5 μg/kg/min), the serum level of corticoids should be checked and the therapy begun when low. Hydrocortisone: 1mg/kg/dose every 6 hours
- Diuretics: Reduce the ventricular volume overload, which is useful to diminish cardiac edema and prepare the child for surgery.

Postoperative management tried to maintain the delicate balance between the vascular resistances (pulmonary and systemic). Ventilation aimed at obtaining a pH of 7.4, PaO_2 of 40 mmHg and PCO_2 of 40 mmHg (balance 40/40/40), as well as systemic arterial O_2 saturation between 75% and 80%. The most commonly used vasoactive agents were milrinone (0.35 to 0.75 μg/kg/min) and dopamine (5 to 15 μg/kg/min), which were employed in 90% of cases without statistically significant differences between the two groups. The use of epinephrine (0.02 to 0.06 μg/kg/min) or norepinephrine was indicated when there was significant hypotension (mean arterial pressure less that 40 mmHg), which occurred in around 30% of cases. The routine use of heparin in the postoperative period aimed at avoiding thrombosis of the PTFE graft.

Definition and statistics

Hospital mortality was defined as all deaths occurring within 30 days after surgery or in children that were never discharged from the Intensive Care Unit (ICU) after surgery. The interstage mortality was that that occurred in children who survived the hospital period, but who did not survive long enough to perform the second stage of the surgery. Statistical analysis was achieved using the GraphPad Prism 4.0 computer software. A level of significance was established for a p-value < 0.05. Numerical variables were expressed as means ± standard deviation and compared using the Student t-test; Fishers exact test was used to compare the mortality between Groups A and B. Medium-term survival was analyzed using Kaplan-Meier actuarial survival curve with the Log Rank Test being used to compare the curves of the two groups.

RESULTS

The immediate survival rate of the 78 patients with HSLS submitted to Norwood surgery was 74.35%, with 67.57% in the mBT Group and 80.49% in the RV-PA Tube Group (p=0.21). The mBT and RV-PA Tube Groups were similar in relation to age, weight, gender and diameter of the ascending aorta, as is illustrated in Table 1.

Figure 1 shows images resulting from the two surgical techniques used.

CPB duration was less in the mBT Group, as anastomosis of the PTFE tube in the subclavian artery was performed during the reheating of the patient. The duration of hypothermic circulatory arrest was shorter for the RV-PA Tube Group, as is shown in Table 2. Figure 2 shows the immediate survival. Of the 37 newborn babies in whom mBT-type systemic-pulmonary anastomosis was performed, 25 (67.57%) survived, and of the 41 infants submitted to the right ventricle-pulmonary artery connection (RV-PA tube), 33 (80.49%) survived.

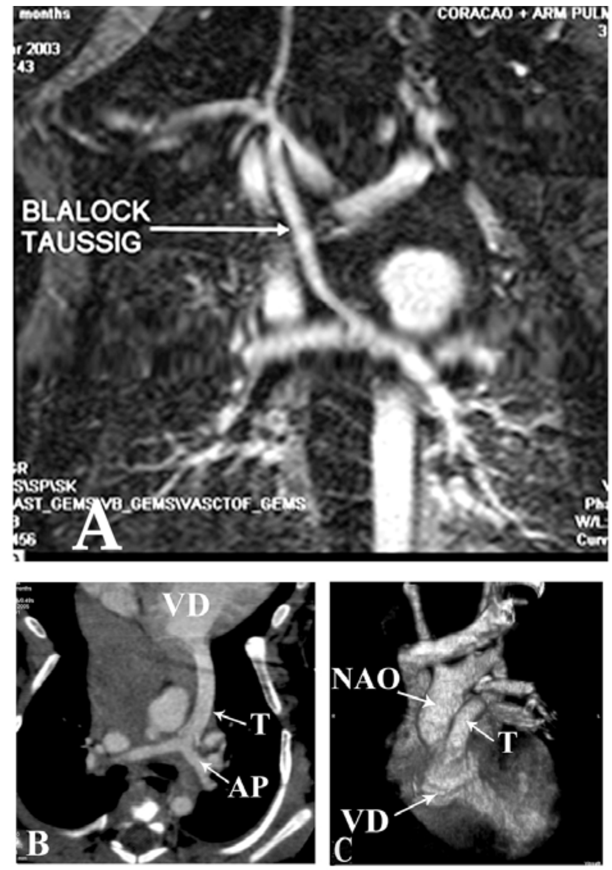

Fig. 1 – Sources of pulmonary perfusion: A: Magnetic resonance angiography (multiplanar reconstruction) showing modified Blalock-Taussing anastomosis connecting the brachiocephalic artery to the pulmonary artery, B: Tomography angiography with multiplanar reconstruction showing the right ventricle tube to the pulmonary artery and C: 3-dimensional reconstruction of the heart correlating the right ventricle, the neoaorta and the PTFE tube. Legend: RV = right ventricle, PA = pulmonary artery, NAO = neoaorta and PTFE T-shaped tube.

Table 1. Pre-operative characteristics of patients

	mBT	RV-PA Tube	p
Patients (n)	37	41	
Gender (men)	48%	56%	
weight (g)	3101 ± 109	3055 ± 63.84	0.698 (NS)
Age (days)	9.22 ± 2.07	8.30 ± 1.26	0.702 (NS)
Diameter of Ao Asc. (mm)	2.713± 0.219	3.137±0.199	0.170 (NS)

mBT = Modified Blalock-Taussig; RV-PA = right ventricle to pulmonary artery; Ao Asc = ascending aorta; NS = non-significant

Table 2. Cardiopulmonary bypass

	mBT	RV-PA tube	p
CPB duration (minutes)	114.0 + 4.39	135.8 + 5.18	0.0017
HCA duration (minutes)	45.79 + 1.99	36.8 + 1.52	0.0012

mBT = Modified Blalock-Taussig; RV-PA = right ventricle to pulmonary artery; CPB = cardiopulmonary bypass; HCA = hypothermic circulatory arrest

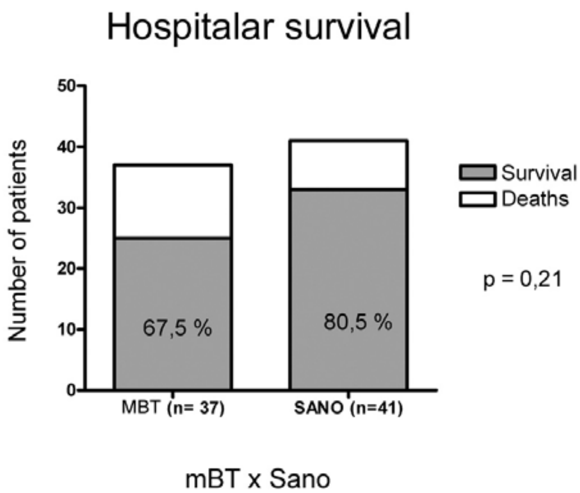

Fig. 2 – Hospital survival in hypoplastic left heart syndrome – mBT = Blalock-Taussing modified

Survival of the period between the first and second stages, which corresponds to the children submitted to the Glenn operation, is shown in Figure 3. Four patients of the Norwood-Sano Group are still waiting to complete the second stage.

Studies performed immediately before the second stage, using magnetic resonance angiography or tomography angiography of the heart and great vessels, demonstrated good aorta morphology in most patients (Figure 4).

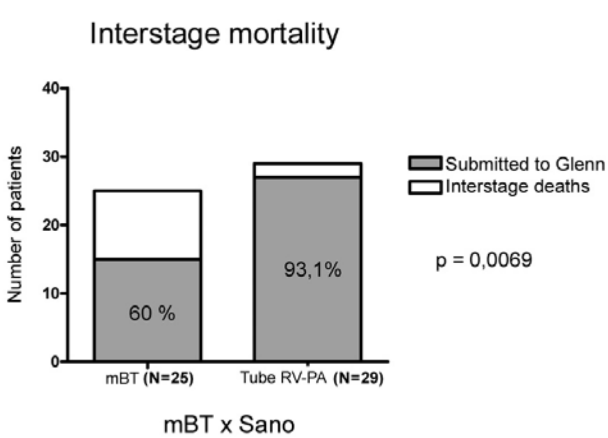

Fig. 3 – Interstage mortality – mBT = modified Blalock-Taussing

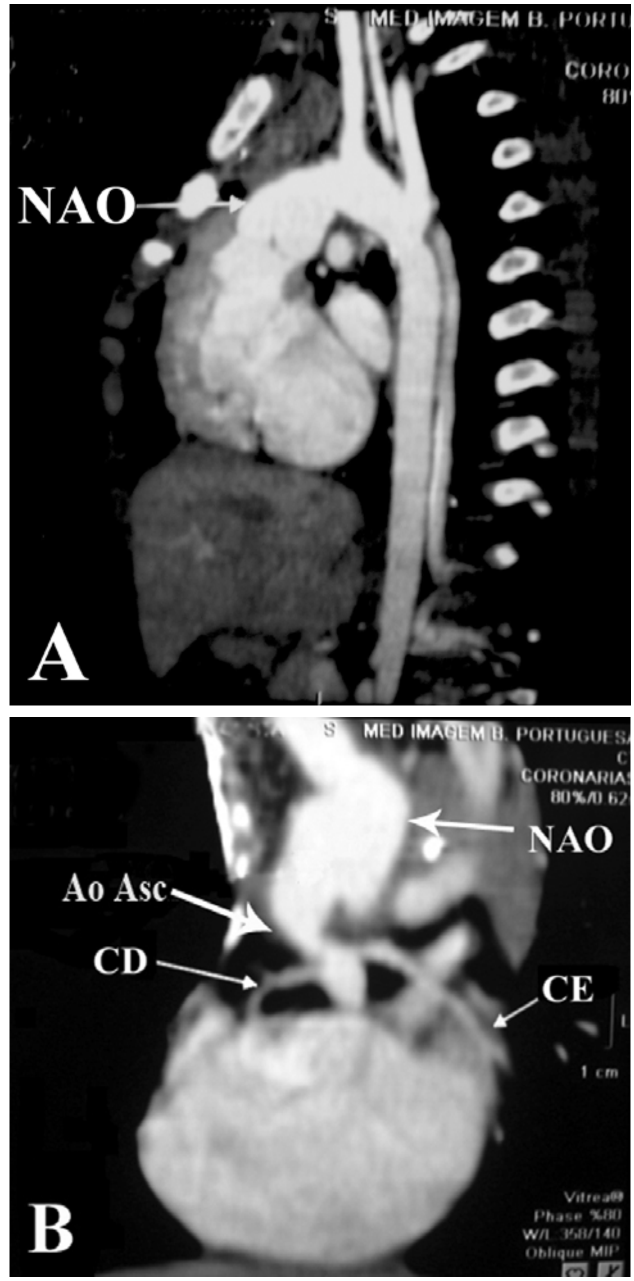

Fig. 4 – Tomography angiography: A: Neoaorta with good anatomic result 2 years after surgery and B: Aspect of coronary arteries and native aorta anastomosed to the neoaorta. Legend: NAO = neoaorta, RC = right coronary, LC = left coronary and AscAo = native ascending artery

Five (6.41%) patients, in the initial phase of this series, evolved with neoaorta coarctation, which was solved by dilatation using a balloon catheter (for two patients after the second stage) or through surgical intervention by thoracotomy (for two patients after the second stage, and one patient after the third stage), giving good results. The comparison of the Kaplan-Meier survival curve (Figure 5) showed better results using the RV-PA tube (p=0.003) in the medium term. Eleven patients have already been submitted to the third stage.

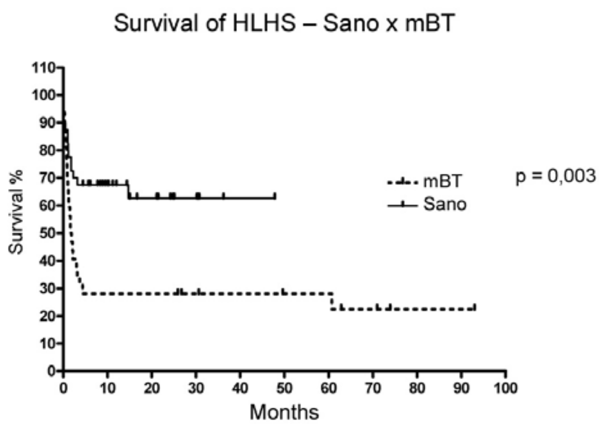

Fig. 5 – long term survival curve – mBT = modified Blalock-Taussig

DISCUSSION

The good immediate survival of 74.35% of patients with HSLS submitted to the first stage of the Norwood operation, with 67.57% in the mBT Group and 80.49% in the RV-PA Tube Group, was obtained after an initial series of 12 newborn babies operated between 1994 and 1998 but not included in this study. Only four of the first 12 patients survived, with an improvement in the results occurring after reviewing these cases and studying published concepts. From this experience the surgical strategy described herein was developed which allowed good surgical correction with myocardial protection and a shorter duration of circulatory arrest, as well as improvement in the postoperative care. We tried to incorporate the evolution in the concepts and results of international experience. In the 1980s, few centers successfully performed the Norwood operation. Publications demonstrated in-hospital survival varying between 42% and 66%, with the majority of deaths occurring within the first 24 hours after surgery as a result of low cardiac outflow. The long-term survival varied between 21% and 44% [13-15].

Data from the Congenital Heart Surgeons Society Data Center, in a prospective study involving 29 institutions, showed survival of 72%, 60% and 54%, respectively, at one month, one year and five years after staged correction. A total of 710 children with HSLS were analyzed between 1994 and 2000. The risk factors for death after the first stage included individual (low birth weight, small ascending aorta and older age in the Norwood operation), institutional and surgical aspects (shunt originating from the aorta, long circulatory arrest and involvement of the ascending aorta). Risk factors for death after the second stage included younger age at cavo-pulmonary shunt and the necessity of atrioventricular valvuloplasty [15].

Tweddell et al. [16] reported improvement in hospital survival from 53% in the operations performed between 1992 and the middle of 1996, to 93% in a series of 81 patients submitted to the Norwood operation using mBT grafts between July 1996 and October 2001. It must be mentioned, however, that this series was not made up of only infants with classical HSLS; 23% had anatomical variants that resulted in the left ventricle becoming the systemic ventricle.

Although more recent data reflect a trend of continued improvement in the results of the Norwood operation for HSLS in centers of excellence in pediatric cardiology throughout the world, mortality remains high, especially in institutions with less experience. Checchia et al. [17], using data from hospitals that are members of the Child Health Corporation of America, studied 1105 newborns with diagnoses of HSLS between 1998 and 2001. They observed that 801 were submitted to the Norwood procedure, 39 were submitted at heart transplantation and the remaining 265 (24%) were not submitted to surgical interventions. The infants submitted to the Norwood operation had a hospital survival rate of 68% (546/801). The four institutions with the greatest number of operations presented a 78% survival rate and the other 25 institutions had, on average, 59% of hospital survival.

Recent clinical trials express concern about the risk of cognitive neurological, neuromotor and psychosocial problems that sometimes occur after these operations [18]. Cyanosis, cardiac insufficiency and preexisting abnormities of the central nervous system that are associated with HSLS, as well as CPB and hypothermic circulatory arrest used in the staged correction, can cause neurological damage. Low flow selective cerebral perfusion has been used by some groups as an alternative to total circulatory arrest [16,19]. Data from a randomized pilot study did not suggest that selective cerebral perfusion improves the neuropsychomotor development of children submitted to the Norwood operation, when compared to hypothermic circulatory arrest [20]. Another study demonstrated that hypothermic circulatory arrest does not harm neurological development as long as its duration is short [21]. Multicentric clinical trials will be necessary to elucidate this question, but the utilization of techniques that decrease or eliminate circulatory arrest is probably safer.

The results obtained using the strategy described herein are similar to those in internationally published reports as only newborns with HSLS were included in this series. Additionally, the size of the ascending aorta and the presence of tricuspid insufficiency were not exclusion criteria, with the Norwood operation being contraindicated in less than 5% cases. Moreover, this technique allows short durations of hypothermic circulatory arrest, probably contributing to a greater survival that is accompanied by a lower risk of impairment to the nervous system.

Our group has used the ground-breaking technique with autologous pericardium treated in glutaraldehyde for aortic arch and descending aorta amplification in HSLS since 1999. This is a simple solution, which apart from the advantages of low cost, easy availability and immunologically compatibility, results in good neoaorta morphology and a low long-term incidence of coarctation [22].

One of the most commonly used techniques in aortic arch reconstruction includes the use of pulmonary homografts which are expensive, difficult to obtain and immunological sensitive. Moreover, these grafts may cause future problems if heart transplantation is necessary [16,23]. The direct anastomosis of the aorta is technically more complex and not always feasible, even though no significant differences have been reported between this technique and homologous grafts in respect to the necessity of re-interventions of the aortic arch [5,6,8].

The recent popularity of the RV-PA tube made the postoperative course more stable, however the impact on survival is still controversial. The experience of this group, showed better results in relation to hospital survival and to mortality between the stages, however statistical significance was only identified using the Fishers exact test in respect to the last variable. Sano et al. [9] reported a survival rate of 84% (61/73) for patients submitted to the Norwood operation using this technique in three centers in Japan between 1998 and 2002. They recognized the surgeon's experience and mechanical ventilation prior to the operation as risk factors. The RV-PA tube provides anterograde flow to the pulmonary arteries only during systole; it can have a larger diameter with less incidence of acute occlusion than the mBT technique, but it allows reverse flow during diastole leading to ventricular volume overload. Moreover, the necessity of right ventriculotomy, theoretically, increases the risk of cardiac arrhythmia and of reduced ventricular function in these patients with univentricular physiology. However, mBT provides anterograde flow through the tube during the entire cardiac cycle. It is controversial if this technique causes more growth of the pulmonary arteries [10,24], but the lower diastolic pressures in the aorta affect the coronary artery blood flow [10,11] and, possibly, the heart function by subendocardial hypoperfusion.

The conclusion of Sano et al. [9] that it will be possible to improve the results of many surgeons using the RV-PA connection were based on a report of a series of cases. Moreover, other reports showing improved survival rates using the RV-PA tube utilized the history of patients with mBT as the control series [10,11,24].

Studies that proved that there is no impact on the survival rate in respect to the type of tube used, apart from comparing non-concurrent groups, had very small populations of newborns [25,26]. Patients with the RV-PA tube presented greater mortality and indication for transplant after the second stage. The comparative study of Tabbut et al. [27], performed using concurrent patients, showed that there is no significant difference in the mortality rate between the RV-PA and mBT Groups. However the study was not randomized and presented some selection bias, such as the inclusion of patients with systemic left ventricles, which theoretically have better prognosis, only in the mBT group, and a preferential inclusion of patients with aortic valve atresia in the RV-PA group.

In spite of the good current results using the mBT technique in the first stage, there is still a risk of from 4% to 15% of deaths before the second stage [15,28,29], although the utilization of a home monitoring program may reduce this risk [30]. The use of a RV-PA tube decreases the interstage mortality among survivors of the Norwood operation, according to the study by Pizarro et al. [31], in which 40 of 46 patients in the mBT Group and 49 of 50 patients in the RV-PA Group tube completed the Hemi-Fontan stage with interim mortality rates of 13% and 2%, respectively. However, groups that present excellent results using the mBT technique showed similar results with the Sano modification. This may indicate that this modification is associated to an improvement in the results of the groups that had close to average mortality rates, maybe because the RV-PA tube facilitates the postoperative management. Randomized studies are necessary to identify possible advantages of this technique.

The main limitation of the current study in respect to analysis of comparative results between the two techniques is that the groups are not concurrent. Although, they have similar clinical characteristics and postoperative care, it is undeniable that the latter group benefited from the greater experience of the surgery team. Also the practice of keeping the majority of the patients in hospital until the second stage was gradually instituted, benefiting the most recent patients in respect to interstage survival.

CONCLUSIONS

The surgical strategy applied in the Norwood operation of newborns with HSLS, utilizing treated autologous pericardium for the amplification of the aortic arch, resulted in a short duration of circulatory arrest and mortality comparable to the best rates in international centers, obtaining good neoaorta morphology and low incidence of aorta coarctation.

The better immediate survival with the RV-PA tube was not significant when compared to the mBT procedure; however, the lower mortality in the interstage period and the greater medium-term survival in the RV-PA group were statistically significant advantages.

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