ORIGINAL ARTICLE

Braz J Cardiovasc Surg 2005; 20(1): 1-7

Bidirectional Glenn procedure in the staged treatment of hypoplastic left heart syndrome: early and late results

Operação de Glenn bidirecional no tratamento estagiado da síndrome de hipoplasia do coração esquerdo: resultados imediatos e tardios

Luciana da FONSECA, José Pedro da SILVA, Sônia Meiken FRANCHI, Rodrigo Moreira CASTRO, Daniel Orselli COMPARATO, José Francisco BAUMGRATZ

RBCCV 44205-722

Abstract

Objective: **To report early and long-term results of bidirectional Glenn procedure as the second stage for Hypoplastic Left Heart Syndrome (HLHS) treatment and describe a ministernotomy technique.**

Method: From March 1998 to February 2004, 15 patients who had been previously submitted to the Norwood procedure underwent elective partial cavopulmonary anastomosis. Ages ranged from 2 to 6 months (mean 3.46 ± 0.83 months). Six were male. Ministernotomy was performed in 11 (73.3%) patients. In order to obtain adequate initial blood oxygenation, an accessory 3-mm PTFE shunt was used in 9 patients and the right ventricle-pulmonary artery tube was maintained in one case. Clinical and echocardiographic follow-up was obtained in all patients.

Results: **Hospital survival was 86.6%, with one early death caused by surgical bleeding and another due to hypoxemia. Postoperative echocardiogram showed some early blood flow in the PTFE shunt and its closure in the long-term follow-up. There were two late deaths, one due to tracheostomy complications and another due to infectious meningitis. Seven patients, who are waiting for the third stage, are doing well clinically. Four patients have already undergone the third stage, successfully. The echocardiograms of 11 late survival patients showed good right ventricular function, no tricuspid regurgitation and good cavopulmonary blood flow over a 2.5-year mean follow-up time.**

Conclusions: **The Glenn procedure in HLHS resulted in a low mortality rate and satisfactory long-term outcomes, and can be performed using the ministernotomy technique. An accessory source of systemic-pulmonary flow in very small infants seems to improve the oxygen saturation.**

Descriptors: **Hypoplastic left heart syndrome, surgery. Heart defects, congenital, surgery. Heart bypass, right, surgery.**

Work performed in the Hospital Beneficência Portuguesa of São Paulo

Correspondence address: Luciana da Fonseca. Rua Maestro Cardim, 769, sala 202, bloco I – Paraíso São Paulo – SP. CEP 01323-001.
Tel: (11) 3285-4393. Fax: (11)3285-4429.
E-mail: lufonseca@uol.com.br

Article received in May, 2004
Article accepted in December, 2004

Resumo

Objetivo: **Relatar os resultados imediatos e tardios da operação de Glenn bidirecional como segundo estágio do tratamento da Síndrome de Hipoplasia do Coração Esquerdo (SHCE) e descrever a técnica de miniesternotomia utilizada.**

Método: **Entre março de 1998 e fevereiro de 2004, 15 pacientes com operação de Norwood prévia foram submetidos eletivamente à derivação cavopulmonar. As idades variaram de 2 a 6 meses (média 3,46 ± 0,83 meses), sendo seis pacientes do sexo masculino. Foram realizadas miniesternotomias em 11 (73,3%) casos. Para adequada oxigenação sangüínea inicial foi associado enxerto sistêmico-pulmonar de 3 mm em nove casos e manutenção do enxerto VD-TP em um caso. Acompanhamento clínico e ecocardiográfico foi realizado em todos os pacientes.**

Resultados: **A sobrevida hospitalar foi de 86,6%, ocorrendo um óbito por sangramento e outro por hipóxia. O ecocardiograma imediato mostrava fluxo pelo enxerto de PTFE nos dez pacientes em que foi utilizado, ocorrendo seu fechamento no controle tardio. Ocorreram dois (13,3%) óbitos tardios, um por complicação de traqueostomia e outro por meningite bacteriana. Sete pacientes aguardam o terceiro estágio, estando assintomáticos. Quatro foram submetidos ao terceiro estágio com sucesso. O ecocardiograma dos 11 pacientes sobreviventes tardios mostra boa função do ventrículo direito, sem insuficiência tricúspide e bom fluxo pela anastomose cavo-pulmonar, num seguimento médio de 2 anos e 5 meses.**

Conclusões**: A operação de Glenn na SHCE apresenta baixa mortalidade hospitalar, com resultados satisfatórios em longo prazo, podendo ser realizada através de miniesternotomia A associação de fluxo sistêmico-pulmonar acessório em crianças de baixa idade parece melhorar a saturação de oxigênio.**

Descritores: **Síndrome do coração esquerdo hipoplásico, cirurgia. Cardiopatias congênitas, cirurgia. Derivação cardíaca direita, cirurgia.**

INTRODUCTION

The term *Hypoplastic Left Heart Syndrome (HLHS)* was introduced by NOONAN and NADAS, in 1958, to describe the morphologic characteristics of combined mitral and aortic atresia [1].The incidence of HLHS is from 1 in 4000 to 6000 life births and the mortality without surgery exceeds 90% in the first year [2].

Several attempts of surgical repair were performed in the 1970s [3] but without success over the short term. The first long-term successful surgical repair was presented by NORWOOD et al. [4], in a series of children who underwent surgery between 1979 and 1981, allowing development of the univentricular Fontan-type repair in 1983 [5], utilizing the right ventricle as a systemic ventricle. The Norwood operation in the 1980s initially showed low survival (42% to 66%) and over the long term (21% to 44%), with the majority of the deaths occurring within the first 24 hours after surgery due to cardiovascular collapse [6,7]. Technical advances, a better understanding of the physiology of the Norwood surgery and refinement of the postoperative management have resulted in better results [6,8,9]. However, a risk of from 10% to 15% of late deaths remains before the second stage of the operation [10,11].

SANO et al. [12] introduced a modification to the original technique of Norwood, utilizing a PTFE graft anastomosed between the right ventricle (RV) and the pulmonary trunk (PT) making the management of the patient in the postoperative period easier by avoiding a drop during diastole in the coronary flow caused by a "shunt" to the lungs, which occurs with the systemic-pulmonary anastomosis [13].

The surgical treatment of HLHS has three phases:

l) **Norwood operation**

The Norwood procedure is performed within the first few days of life and consists of the anastomosis of the pulmonary trunk to the aorta with enlargement of aortic arch, generally using autologous pericardium treated with glutaraldehyde, associated to atrial septectomy and the placement of a systemic-pulmonary PTFE graft. Recently a RV-pulmonary graft (Sano modification) has been utilized.

2) **Second stage**

Partial cavopulmonary shunt (bidirectional Glenn or Hemi-Fontan procedure) where the flow of the superior vena cava is shunted to the pulmonary artery. It is performed when the patients is between 2 and 10 months of age [14-17].

3) **Third stage**

Fontan surgery or total cavopulmonary shunt, with the flow of the inferior vena cava directed to the lungs. It is performed at ages of from 18 and 24 months.

The alternative, heart transplantation, for the treatment of this syndrome has the disadvantages of limited availability of donors and the necessity of long-term immunosuppression [9,18].

The aim of this study is to report the ministernotomy surgical technique utilized in the second stage and the long-term evolution of patients with HLHS who electively underwent the bidirectional Glen procedure.

METHOD

From March 1998 to February 2004, 15 children with HLHS, who electively underwent the bidirectional Glenn procedure, were investigated. The mean age was 3.46 months

(range from 2 to 6 months) and the mean weight was 4.56 kg (range from 3.5 to 5.5 kg). Six (40%) of the patients were male. The first stage of Norwood procedure had been performed in these children in the neonatal period.

The children who had been submitted to the second stage in an emergency situation (severe hypoxia or low heart output syndrome after the Norwood procedure) were excluded from this group.

The preoperative echocardiogram demonstrated normal RV systolic function in 10 patients and moderately depressed in five. Slight tricuspid regurgitation was seen in five patients and moderate regurgitation in three.

The data of the patient report cards in respect to the preoperative, trans-operative and postoperative periods were retrospectively analyzed. Long-term clinical evaluation and echocardiograms were performed for all children, with magnetic angioresonance, to plan the second or third stages, performed in ten patients.

The vasoactive drugs used, which were necessary in most of the cases, were dopamine (5-15 μg/kg/min) and milrinone (0.25-0.75 μg/kg/min), with dobutamine (5-15 μg/kg/min), nitroprusside and noradrenaline being utilized in some cases.

To prevent thromboembolic events, the patients were medicated with acetylsalicylic acid (5 mg/kg/day), low molecular weight heparin (1 mg/kg/day) or dipiridamol (2 mg/kg/day), in the late postoperative period

Operative technique

The chest was opened by normal sternotomy in four (26.7%) patients and by inverted L hemisternotomy up to the third right intercostal space in 10 (66.6%) patients. Inverted T hemisternotomy was used in one (6.7%) patient who presented with the superior vena to the left. With the dissection of adherences and removal of the right lobe of the thymus, the anterior face of the aorta was exposed and cannulated after systemic heparinization (5 mg/kg). The right atrial appendage was exposed where a venous drainage catheter was placed. The superior cava vena was dissected and then selectively cannulated. The azygos vein was sectioned and sutured. Cardiopulmonary bypass was established and the patient was cooled to 30 ºC.

The PTFE graft between the right subclavian artery and pulmonary artery was sectioned and sutured immediately after the establishing of the cardiopulmonary bypass. Aortic clamping was not used. The superior cava vena was sutured proximal to its connection in the right atrium, and soon after it was sectioned above this suture. The right pulmonary artery was opened and end-to-side anastomosis to the superior cava vena was performed utilizing 6.0 PDS absorbable thread. The operative field is shown in Figure 1. Some technical modifications were necessary in some of the cases, which were as follows:

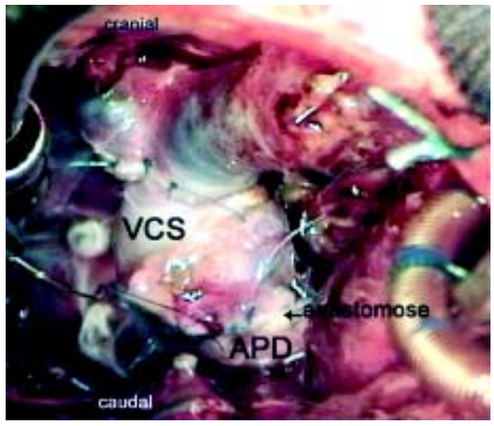


Fig. 1 - Operative field. Anastomosis of the superior vena cava in the right pulmonary artery

Aortic clamping and anterograde cardioplegia were performed in one patient, who also required enlargement of the left pulmonary artery.

In the third patient total circulation arrest was necessity to perform tricuspid valvuloplasty.

Association of a pulmonary systemic tube between the right subclavian artery and pulmonary artery was necessary in the sixth patient due to hypoxia (oxygen saturation less than 60%), in the immediate postoperative period. After this case, we opted to routinely use a thinner graft (3 mm) sutured to the old PTFE tube, clamped and sectioned during cardiopulmonary bypass, guaranteeing an adequate pulmonary flow in the postoperative period. This provided good oxygenation in all subsequent patients.

In the last patient, the only one in this series who was submitted to the Sano modification during the first stage, an inverted T hemisternotomy was performed in the second stage, due to the presence of a left cava vena and so the bilateral Glenn procedure was performed and the RV-PT graft was maintained.

RESULTS

Hospital survival was 86.6% with two deaths: The first, a patient operated on by complete sternotomy died in the immediate postoperative period due to excessive bleeding. The second patient presented with significant hypoxia when the cardiopulmonary bypass was removed and required an increase of the pulmonary flow using a PTFE graft. The echocardiogram showed an adequate anastomosis of the cava vena in the pulmonary artery without obstructions to the flow. The patient died five days after, due to right ventricle failure, probably initiated by the brief period of hypoxia to which the infant was submitted in the postoperative period.

The postoperative complications were: bronchopneumonia and ischemic stroke in one (6.7%) patient, phrenic paralysis in two (13.3%) patients with the necessity of surgical repair (plicature of the diaphragm), operative wound infection in two (13.3%) patients, mediastinal bleeding and slight subdurale hematoma in one (6.7%) patient and chylothorax to the left in the patient who presented with the left cava vena (6.7%). Due to these complications, the time of stay in the intensive care unit was prolonged, with a mean of 12.3 ± 9.01 days (n=12) varying from 3 to 39 days, even excluding the second patient, who remained for one year and six months under treatment of stenosis of the trachea with tracheostomy that had already been diagnosed before the second stage. All patients presented sinusal rhythm in the postoperative period.

The average peripheral blood saturation was 82% in the 13 patients, at hospital discharge.

The mean follow-up time of these patients was two years and five months. No patient presented with motor neurologic, intellectual or cognitive sequels in the long-term follow up.

There were two (13.3%) late deaths, one due to complications of a tracheostomy in an infant of two years and two months, who presented with sudden death three months after being released from hospital and the other due to infective meningitis at nine months of age.

An echocardiogram in the first week after surgery showed flow through the PTFE graft in all patients in whom it was utilized, with its closure evidenced in the long-term follow-up, but, without the occurrence of significant cyanosis at this phase.

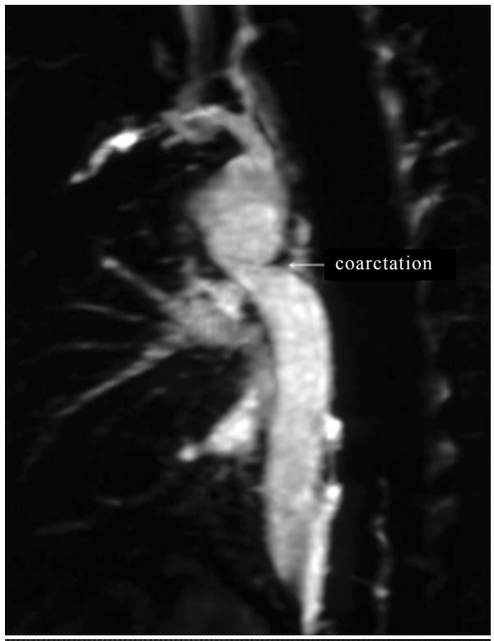
Three patients presented with late aortic coarctation, diagnosed by echocardiogram and angioresonance with surgical repair performed without complications in two patients after the second stage and the other patient after the third stage (Figure 2).

Four patients were successfully submitted to the Fontan procedure (third stage) at two years of age and were clinically well in the long-term follow up of from 1 to 3 years after the third stage. Protein-loosing enteropathy occurred in one patient after the Fontan procedure and re-hospitalization and multiple re-interventions (dilatation of the Fontan fenestration, dilatation of the aortic coarctation, embolization of arterial-venous fistula, AV valvuloplasty and surgical repair of aortic coarctation) were necessary to reduce the high venous blood pressure.

Seven patients are still waiting for the third stage without symptoms.

A patient, in the preoperative evaluation of the third stage, presented severe obstruction of the left pulmonary artery root with hypo-development, evidenced by angioresonance, associated with aortic coarctation. Apart from the repair of the aortic coarctation, anastomosis of a 5-mm PTFE graft between the descending aorta and left pulmonary artery was opted for in this case to improve the development of the left pulmonary artery in preparation for the third stage.

A



B

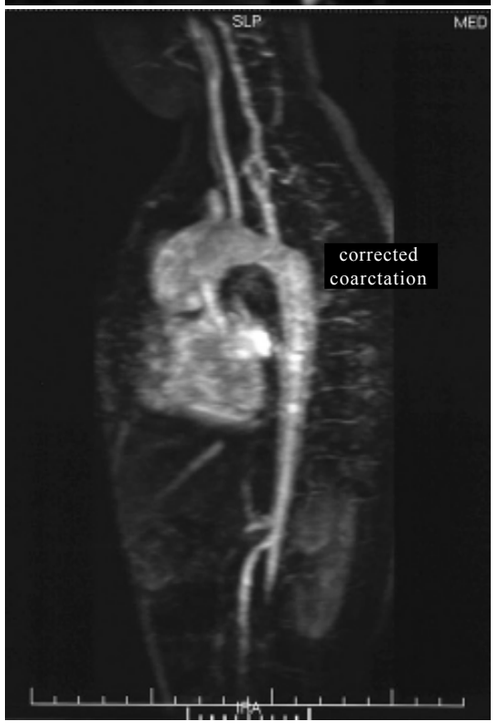


Fig. 2 - Chest angiorressonancia. A- Aortic coarction de aorta. B - Cavopulmonary Anastomosis

The long-term evolution of patients is illustrated in Figure 3.

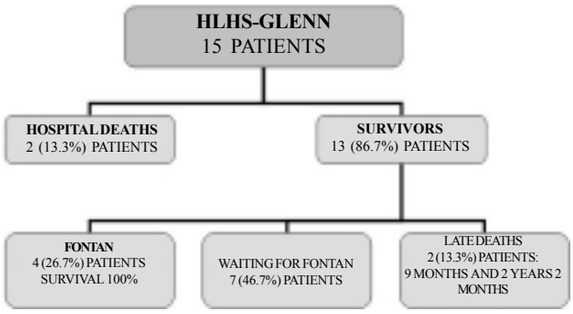


Fig. 3 - Diagram of the evolution after the second stage

Late thromboembolic complications did not occur as nine patients used acetylsalicylic acid, one used low molecular weight heparin and another used dipiridamol.

The echocardiograms of 11 long-term survivors showed the right ventricles without dysfunction, competent tricuspid valves and good flows through the cavopulmonary anastomoses over a mean time of 2 years and five months of follow up.

COMMENTS

The superior cava vena-pulmonary artery shunt (bidirectional Glenn procedure) has been utilized in the staged treatment of heart diseases with single functional ventricles. In HLHS, this second stage is necessary because the children develop progressive cyanosis, due to the lack of growth of the systemic-pulmonary graft (SPG), as well as the risk of its acute occlusion. The superior cava vena-pulmonary artery shunt increases the blood oxygenation and eliminates or decreases the two hemodynamic alterations caused by the SPG, which are volume overload of the RV and the decrease of the coronary flow due to a diastolic drop in the arterial pressure.

The procedure in three stages was utilized by Norwood from 1989. By 1992, 138 patients had been submitted to the hemi-Fontan surgery, with one death and 82 patients who completed the Fontan procedure, with seven early deaths and three late deaths [19].

The association of another source of pulmonary blood flow was reported in children with single ventricles submitted to the Glenn procedure before four months of age, to immediately increase the oxygenation after removal of the cardiopulmonary bypass [20].

The arguments to maintain an extra source of pulmonary flow, whether directly from the right ventricle or from a systemic-pulmonary graft, include the increase in the arterial oxygenation in the postoperative period, the promotion of the growth of the pulmonary arteries and possibly, prevention of the development over the long term of arteriovenous pulmonary and collateral systemic-pulmonary fistulae [21-23].

In the national literature, there was an early death in a series of five children submitted to the Norwood procedure from January to December of 2002, excluding those children with an aorta smaller than 4 mm or with severe infections. The Glenn procedure was performed in 1 ten-month-old infant so the mortality after the first stage was 25% (one case after 2 months) [24].

In this series, the indication of Glenn procedure was made at an early age, mean of 3.46 months, with the aim of decreasing the mortality in the period between the first and the second stages, which was also high. The hospital mortality was relatively high (13.3%) due to two deaths that occurred at the beginning of this series in 1998. One of them was due to postoperative hypoxia and the other surgical bleeding in the adherences between the heart and the sternum. The postoperative hypoxia may be related to the young age [20]. This was solved with the maintenance of a systemic-pulmonary graft, after reducing its caliber, which guaranteed better oxygenation in subsequent cases. There were no mortalities in the last nine patients of the series, in which a tube to increase the pulmonary blood flow in the immediate postoperative period was routinely associated. This extra flow, apart from increasing the blood oxygenation, facilitates the growth of the pulmonary arteries.

A study by JAQUISS et al. [25] compared children with ages of 94 ± 21 days (very young) and children with 164 ± 44 days (the usual age) suffering from HLHS submitted to the second stage of the Norwood procedure. The advantages of the cavopulmonary anastomosis before four months of life are the elimination of the volume overload imposed to the right ventricle and the shortening of the period of high risk of mortality between the first and second stages. The disadvantages include lower oxygenation, longer periods of mechanic ventilation, pleural effusions and longer ICU and hospital stays, however without increasing the risk of mortality. However, these authors did not utilize additional pulmonary flow and obtained saturation levels of 75 ± 7%, while in this series the observed mean saturation was 82%.

The performance of ministernotomy in 73% of the cases reduced the dissection area with a reduction of the bleeding.

The late mortality in two cases, due to the complications of tracheostomy in one and infective meningitis in the other, did not have any direct relationship to the surgical procedure.

The function of the sinusal node was maintained in all patients from the immediate postoperative period.

Early or late thromboembolic complications did not occur,

suggesting that the platelet anti-aggregating is enough for prophylaxis in the majority of the patients.

Angioresonance was useful in the evaluation of the anatomy of the pulmonary vessels and of the aorta, helping in the planning of the surgery. It was performed instead of heart catheterism with the advantages of being a noninvasive method, but, with the inconvenience of not enabling the measurement of the intracavitary pressures.

The later surgeries (aortic coarctation repair in three patients and tricuspid re-plasty in one patient) were not associated with the technique employed for the cavopulmonary shunt.

All late survivors are well and asymptomatic.

CONCLUSIONS

The Glenn procedure (second stage of Norwood procedure) in patients with hypoplastic left heart syndrome presents low mortality rates when selectively performed, with satisfactory results over the long term and can be achieved by ministernotomy. The association of an extra systemic-pulmonary flow in younger children seems to improve the oxygen saturation.

BIBLIOGRAPHIC REFERENCES

1. Noonan JA, Nadas AS. The hypoplastic left heart syndrome: an analysis of 101 cases. Pediatr Clin North Am. 1958;5(4):1029-56.

2. Fyler DC. Report of the New England Regional Infant Cardiac Program. Pediatrics. 1980;65(suppl):375-461.

3. Mohri H, Horiuchi T, Haneda K, Sato S, Kahata O, Ohmi M et al. Surgical treatment of hypoplastic left heart syndrome: case reports. J Thorac Cardiovasc Surg. 1979;78(2):223-8.

4. Norwood WI, Lang P, Castaneda AR, Campbell DN. Experience with operations for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 1981;82(4):511-9.

5. Norwood WI, Lang P, Hansen DD. Physiologic repair of aortic atresia-hypoplastic left heart syndrome. N Engl J Med. 1983;308(1):23-6.

6. Iannettoni MD, Bove EL, Mosca RS, Lupinetti FM, Dorostkar PC, Ludomirsky A et al. Improving results with first stage palliation for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 1994;107(3):934-40.

7. Meliones JN, Snider AR, Bove EL, Rosenthal A, Rosen DA. Longitudinal results after first-stage palliation for hypoplastic left heart syndrome. Circulation. 1990;82(5 suppl):IV151-6.

8. Pearl JM. Right ventricular-pulmonary artery connection in stage 1 palliation of hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 2003;126(5):1268-70.

9. Chang RK, Chen AY, Klitzner TS. Clinical management of infants with hypoplastic left heart syndrome in the United States, 1988-1997. Pediatrics. 2002;110(2 pt 1):292-8.

10. Pearl JM, Nelson DP, Schwartz SM, Manning PB. First stage palliation for hypoplastic left heart syndrome in the twenty-first century. Ann Thorac Surg. 2002;73(1):331-40.

11. Azakie A, Merklinger SL, McCrindle BW, van Arsdell GS, Lee KJ, Benson LN et al. Evolving strategies and improving outcomes of the modified Norwood procedure: a 10-year single institution experience. Ann Thorac Surg. 2001;72(4):1349-53.

12. Sano S, Ishino K, Kawada M, Arai S, Kasahara S, Asai T et al. Right ventricle-pulmonary artery shunt in first stage palliation of hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 2003;126(2):504-10.

13. Forbess JM. Pre-stage II mortality after the Norwood operation: addressing the next challenge. J Thorac Cardiovasc Surg. 2003;126(5):1257-8.

14. Douglas WI, Goldberg CS, Mosca RS, Law IH, Bove EL. Hemi-Fontan procedure for hypoplastic left heart syndrome: outcome and suitability for Fontan. Ann Thorac Surg. 1999;68(4):1361-8.

15. Bando K, Turrentine MW, Sun K, Sharp TG, Caldwell RL, Darragh RK et al. Surgical management of hypoplastic left heart syndrome. Ann Thorac Surg. 1996;62(1):70-7.

16. Norwood WI, Jacobs ML. Fontan's procedure in two stages. Am J Surg. 1993;166(5):548-51.

17. Bove EL, Lloyd TR. Staged reconstruction for hypoplastic left heart syndrome: contemporary results. Ann Surg. 1996;224(3):387-95.

18. Stuart AG, Wren C, Sharples PM, Hunter S, Hey EN. Hypoplastic left heart syndrome: more potential transplant recipients than suitable donors. Lancet. 1991;337(8747):957-9.

19. Norwood WI, Jacobs ML,Murphy JD. Fontan procedure for hypoplastic left heart syndrome. Ann Thorac Surg. 1992;54(6):1025-30.

20. Reddy VM, Liddicoat JR, Hanley FL. Primary bidirectional superior cavopulmonary shunt in infants between 1 and 4 months of age. Ann Thorac Surg. 1995;59(5):1120-6.

21. Caspi J, Pettitt TW, Ferguson TB, Stopa AR, Sandhu SK. Effects of controlled antegrade pulmonary blood flow on cardiac function after bidirectional cavopulmonary anastomosis. Ann Thorac Surg. 2003;76(6):1917-22.

22. Miyaji K, Shimada M, Sekiguchi A, Ishizawa A, Isoda T. Usefulness of pulsatile bidirectional cavopulmonary shunt in high-risk Fontan patients. Ann Thorac Surg. 1996;61(3):845-50.

23. Uemura H, Yagihara T, Kawashima Y, Okada K, Kamiya T, Anderson RH. Use of the bidirectional Glenn procedure in the presence of forward flow from the ventricles to the pulmonary arteries. Circulation. 1995;92(9 suppl):II228-32.

24. Fantini FA, Gontijo Filho B, Martins C, Lopes RM, Heiden E, Vrandecic E et al. A operação de Norwood modificada para tratamento da síndrome de hipoplasia do coração esquerdo. Rev Bras Cir Cardiovasc. 2004;19(1):42-6.

25. Jaquiss RD, Ghanayem NS, Hoffman GM, Fedderly RT, Cava JR, Mussatto KA et al. Early cavopulmonary anastomosis in very young infants after the Norwood procedure: impact on oxygenation, resource utilization and mortality. J Thorac Cardiovasc Surg. 2004;127(4):982-9.