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Abstract

Objective: To show current results of a modified Norwood procedure, where only autologous tissues are used in the reconstruction of the new aortic arch.

Method: Between January and December 2002, five neonates with hypoplastic left heart syndrome underwent the modified Norwood operation. Their ages ranged from two to nine days and their weight from 2.5 to 3.7 kg. The mean diameter of the ascending aorta was 6.2 mm. The surgical technique described by FRASER and MEE was employed using exclusively autologous tissue to achieve aortic arch reconstruction.

Results: The systemic-pulmonary shunt was performed using 3-mm PTFE grafts in three patients and 3.5-mm grafts in the other two. Deep hypothermic and circulatory arrest were used in all patients with cardiopulmonary bypass times ranging from 128 to 212 minutes (mean: 154) and circulatory arrest from 41-60 min (mean: 52). There were no intraoperative deaths and all patients underwent delayed sternal closure. There was one postoperative death (9th day) caused by sepsis. There was also one death two months after discharge caused by milk aspiration and pneumonia. One child underwent a 2-directional Glenn and the other two are still waiting for the second stage.

Conclusion: After this initial experience with this modified Norwood operation we think this is an excellent alternative to correct hypoplastic left heart syndrome in neonates.

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

Resumo

Objetivo: Apresentar os resultados do tratamento da síndrome de hipoplasia do coração esquerdo (SHCE) com técnica de Norwood modificada, na qual somente são usados tecidos autólogos para a reconstrução do arco aórtico.

Método: De janeiro a dezembro de 2002, cinco recém-nascidos com idade variando de dois a nove dias (média 5,0 dias) foram submetidos a operação de Norwood modificada.

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O diâmetro da aorta ascendente variou de 5 a 8 mm (média 6,2 mm). Foi empregada técnica na qual o arco aórtico e seus ramos foram amplamente dissecados, permitindo a reconstrução de um novo arco aórtico tendo a artéria pulmonar como via de saída, e com a utilização exclusiva de tecidos do próprio paciente. Anastomose sistêmico-pulmonar com politetrafluoretileno expandido de 3,0 mm foi utilizada em três pacientes e de 3,5 mm em dois pacientes. Todos foram operados com parada cardiocirculatória total.

Resultados: **O tempo de parada cardiocirculatória variou de 41 a 60 minutos (média 52,8 minutos). Todas as crianças sobreviveram ao ato operatório e foram encaminhadas para a unidade de terapia intensiva com o esterno aberto. Ocorreu um óbito no 9º dia de pós-operatório por sepse, o que resultou numa taxa de sobrevivência imediata de 80%. Ocorreu um óbito tardio por pneumonia de aspiração aos dois meses. Uma criança foi submetida ao segundo estágio da operação (cavo pulmonar) e encontra-se bem. Duas crianças estão em acompanhamento aguardando o próximo estágio. Em nenhuma delas existem evidências de obstrução do novo arco aórtico.**

Conclusão: **A técnica de Norwood modificada mostrou-se eficaz e com risco cirúrgico aceitável para o tratamento da SHCE, sem evidências de obstrução do arco aórtico reconstruído.**

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

INTRODUCTION

Hypoplastic left heart syndrome (HLHS) is a disease in which the left ventricle and the aorta present varying degrees of hypoplasia. It is always accompanied by stenosis or atresia of the mitral and/or aortic valves. It makes up around 2% of all congenital heart diseases being the fourth most commonly diagnosed anomaly in the first year of life. Without treatment it is fatal in 100% of the cases, with 95% of children dying within their first month of life. It is still the most frequent cause of death by heart disease in the first week of life [1].

Currently, there are two ways to treat: neonatal heart transplantation and staged palliative reconstruction as described by NORWOOD et al. [2]. Heart transplantation has important limitations, which are the lack of compatible donors and the adverse side effects of long-term immunosuppression [3]. Thus the Norwood operation is an attractive alternative for the treatment of HLHS. The first stage involves the creation of an ample connection between the systemic right ventricle through the pulmonary artery branch and the descending aorta and the reconstructed aortic arch and the establishing of a controlled source of pulmonary flow through a systemic-pulmonary anastomosis. In the second stage, the systemic-pulmonary shunt is replaced by a 2-directional cavopulmonary anastomosis (the modified Glenn procedure). The third step is using the Fontan operation or similar, thereby completing the treatment.

The Norwood operation, however, continues to be a challenge, with high immediate and late mortality rates [4]. The classical version, in which the ascending aorta and the aortic arch are enlarged by materials such as homografts or bovine pericardium, has a high incidence of aortic arch obstruction [5]. Several neo-aortic reconstruction techniques without using heterologous material have now been published [6,7]. The aim of this study is to present the results of the treatment of HLHS obtained in our department utilizing the Norwood operation modified by FRASER & MEE [8]. In this technique the aortic arch and the supra-aortic vessels are amply mobilized and anastomosed to the proximal pulmonary artery and to the descending aorta, only using the patient's tissues.

METHOD

From January to December 2002, five newborn babies (2 male and 3 male) suffering from HLHS were submitted to the modified Norwood operation. The ages ranged from 2 to 9 days (mean 5.0 ± 2.7 days) and the weights varied from 2.5 to 3.7 kg (mean 3.0 ± 0.4 kg).

The five patients presented with classical HLHS, defined as atresia or stenosis of the mitral and/or aortic valves, hypoplasia of the left ventricle, the great vessels concordant and an intact interventricular septum. One child also suffered from a type B interruption of the aortic arch and another presented with left atrial isomerism with the left superior vena cava patent. One patient had an interruption of the inferior vena cava, with venous drainage to the azygos system.

The diameter of the ascending aorta, measured by preoperative echocardiography, ranged from 5 to 8 mm (mean 6.2 ± 1.3 mm).

In the preoperative period, three patients presented with stable clinical conditions, with a good peripheral perfusion and diuresis. One child presented with hemodynamic instability and significant acidosis, after being electively intubated and receiving inotropic support the day previous to the operation and the other was transferred from another hospital already intubated. All were taking prostaglandin E1.

It is important to stress that the established protocol for this first stage excluded patients with aortas smaller than 4 mm and those with severe infectious complications.

In the first stage, the surgical technique employed was median sternotomy and total thymectomy, performed in all children, allowing the removal of a pericardial patch that, after being prepared in a 4% glutaraldehyde solution, was used for the reconstruction of the bifurcation of the pulmonary artery. The ascending aorta, aortic arch, descending aorta and the supra-aortic branches were extensively mobilized, as was the pulmonary artery branch and the right and left branches. The cardiopulmonary bypass (CPB) circuit was established with an arterial cannula placed in the ductus arteriosus and a single cannula in the right atrium.

With the patient with aortic arch interruption, another cannula was placed in the aorta near the brachiocephalic branch. After installation of the CPB, the ductus arteriosus was proximally connected to the arterial cannula to avoid volumetric overload of the ventricle. During the cooling phase, the pulmonary artery branch was obliquely cross-sectioned near to the root of the pulmonary branches and the distal neck was closed with an autologous pericardial patch. Under deep hypothermia of 16 °C and total circulatory arrest (TCA), sanguineous cardioplegia with lidocaine and magnesium was injected by the arterial cannula, after clamping the descending aorta and the supra-aortic branches. All the cannulae were removed and atrioseptostomy was performed through the atrial cannulation pouch. All the ductal tissue was resected and the lesser curve of the aortic arch was amply widened from the subclavian artery to the ascending artery. The arch was reconstructed by anastomosis of the descending aorta to the isthmus and the distal aortic arch. The pulmonary artery branch was then anastomosed to the anterior assemblage and to the ascending aorta. During the re-warming phase, systemic-pulmonary anastomosis (modified Blalock-Taussig) was constructed between the right innominate artery and the right branch of the pulmonary artery, using 3-mm polytetrafluoroethylene (PTFE) prostheses in three patients and 3.5-mm in two (Figure 1).

Before the second stage, the patients were submitted to echocardiographic and cinecardiographic studies. The 2-directional Glenn operation was performed with CPB but without cardiac arrest. The PTFE prosthesis implanted in the first operation was dried and the superior vena cava was anastomosed to the same pulmonary arteriotomy which was conveniently increased.

RESULTS

The CPB and TCA times varied from 128 to 212 minutes (mean: 154 minutes) and from 41 to 60 minutes (mean: 52.8 ± 9.0 minutes) respectively. With small recently introduced technical modifications, the TCA time was reduced to nearly 40 minutes in the latter patients.
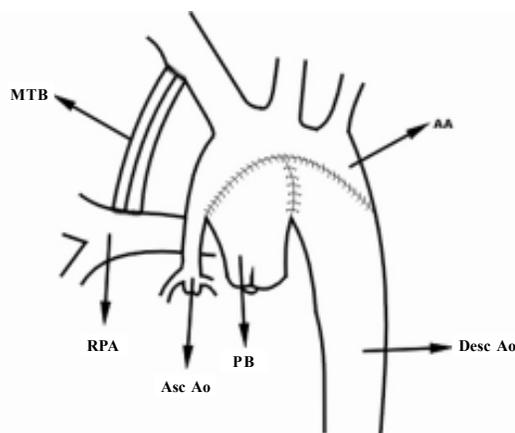
AA = Aortic arch; Asc Ao = Ascending Aorta; Desc Ao = Descending Aorta; PB = Pulmonary Branch; RPA = Right Pulmonary Artery; MBT = Modified Blalock-Taussig

Fig. 1: Diagram of the surgical technique employed.

All the children survived the operation and were sent to the intensive care unit (ICU) in stable clinical conditions. The sternum was closed 1 to 3 days after (mean: 2 ± 0.7 days) without evidence of complications related to the tactic. Intubation time varied from 2 to 9 days (mean: 4.0 ± 2.8 days) and stay in the ICU was from 6 to 9 days (mean: 6.8 ± 1.3 days).

No surgical re-interventions were necessary in the postoperative period. The clinical complications observed were that one child suffered pulmonary atelectasis and another sepsis, which was responsible for the only hospital mortality that occurred. No neurological or renal complications were observed.

The time of hospitalization was on average 10 days. Four children were released making an 80% immediate survival rate.

In the late postoperative follow up period, one death occurred two months after hospital release. The child was attended in another hospital and was diagnosed as having respiratory insufficiency due to milk aspiration. It was not possible to make an autopsy to confirm the cause of death.

The three survivors have already undergone a study by cardiac catheterism, with the modified Blalock-Taussigs continuing patent, without evidence of distortion or stenosis of the pulmonary artery and there is no evidence of obstruction of the neo-aortic arch of any of them. One child was submitted to the second stage with the preparation of the 2-directional Glenn-type cavopulmonary anastomosis at ten months of age. This patient had an excellent postoperative evolution. The other two patients are waiting to perform the second stage. However in the 12-months follow up a survival rate of 60% was recorded.

COMMENTS

The normally reported immediate mortality rate in the treatment for HLHS is 60% [9]. However, with modifications in the operative technique, the CPB technology and in the preoperative management by multidisciplinary teams, survival of up to 80 or 90% has been reported [4,10]. Among the risk factors for mortality after the Norwood operation are low birth weight [11], anatomic subtype diagnosis [12], associated cardiac anomalies [13], preoperative clinical conditions including the necessity of mechanical ventilation [4] and the diameter of the ascending aorta [10]. As this is a small group of patients and an initial experience, our series does not allow an analysis of these risk factors, however in general the patients were of a lesser surgical risk, which, in part, explains the 80% immediate and 60% late survival rates. It is important to state that the patients were related as the only inclusion criterion was an aorta equal to or greater than 4 mm.

One of the critical points in the original operation described by NORWOOD et al. [2] involved the extensive use of widening patches on the delicate and hypoplastic aorta of newborn babies. These patches, whether homografts or heterografts, certainly will not accompany the growth of the children and they also suffer degenerative alterations which evidently compromise the late results [6]. Among the alterations reported in the literature [14-16], we opted for the technique described by FRASER & MEE [8], which effectively only utilized autologous tissue in the reconstruction of the neo-aorta, even in the presence of coarctation or interruption of the aortic arch. In these five patients, it was not necessary to implant the ascending aorta in the pulmonary artery branch, as subsequently suggested by POIRIER et al. [10]. This was firstly because we opened the lesser curvature of the aortic arch extensively, to very close to the root of the left coronary artery and secondly because the degree of hypoplasia of the ascending aorta in our series was less than those described by POIRIER et al. [10].

Another important point in relation to the technique is the diameter of the PTFE prostheses employed in the construction of the systemic-pulmonary anastomoses. Analyzing the physiology of the circulation using a computer model of the Norwood operation, MIGLIAVACCA et al. [17] observed that the use of derivations of a greater diameter, shunts a greater proportion of the cardiac output to the lungs, diminishing with this the systemic perfusion. Thus, some authors have suggested the use of PTFE prostheses of a maximum of 3 mm, which facilitates the management of the pulmonary/systemic flow ratio (Qp/Qs) after correction [6]. We utilized prostheses of 3 mm in newborn babies of up to 3 kg and 3.5 mm in heavier children.

However the persistent incidence of death during the first year of life among the patients who survived the first stage is a concern. In some studies, the percentage can be as high as 12 to 15%, death is generally sudden, unexplainable and before performing the 2-directional cavopulmonary anastomosis [13]. One of the possible causes is an inadequate pulmonary flow, caused by occlusion of the modified Blalock-Taussig, that might have been the cause of the late death observed in one of our cases, as the patient died of frank respiratory insufficiency, initially attributed to a probable aspiration of milk. For this, the authors tried to shorten the interval between the first stage and performing the cavopulmonary shunt [4]. Another very attractive alternative is the modified technique proposed by SANO et al. [18], that consists of the preparation of right ventricle-pulmonary artery conduit using a 5-mm PTFE prosthesis. Although an incision in the anterior wall of the systemic right ventricle is required, the flow by the pulmonary artery is anterograde and also larger prostheses are occluded with greater difficulty.

One of the advantages of the present technique is the low incidence of obstruction of the new aortic arch. In a recent publication with the results of this technique, POIRIER et al. [10] observed that only 5% of the patients operated on required an operation to correct obstruction of the new aortic arch. Utilizing a similar surgical technique on a series of 120 patients, ISHINO et al. [19] found 16 patients (23%) had obstructions of the new aortic arch, of which 10 were successfully dilated using a balloon and six were corrected surgically. In our series, although small, presence of obstructions of the aortic neo-arch were not evidenced.

CONCLUSIONS

Although the sample was small and the follow-up time limited, the modified Norwood operation, when only autologous tissues were employed for the reconstruction of the systemic outflow tract of the heart and the aortic arch, was seen to be efficacious with a good surgical result and without evidence of obstruction of the reconstructed aortic arch.

BIBLIOGRAPHIC REFERENCES

1   Lloyd TR, Marvin Jr. WJ. Age at death in the hypoplastic left heart syndrome: multivariate analysis and importance of the coronary arteries. Am Heart J 1989;117:1337-43.

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

3   Razzouk AJ, Chinnock RE, Gundry SR, Johnston JK, Larsen RL, Baum MF et al. Transplantation as a primary treatment for hypoplastic left heart syndrome: intermediate-term results. Ann Thorac Surg 1996; 62:1-8.

4   Azakie T, 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;72:1349-53.

5   Bartram U, Grunenfelder J, Van Praagh R. Causes of death after the modified Norwood procedure: a study of 122 postmortem cases. Ann Thorac Surg 1997;64:1795-802.

6   Gutgesell HP, Gibson J. Management of hypoplastic left heart syndrome in the 1990s. Am J Cardiol 2002; 89:842-6.

7   Nagy ZL, Parsons JM, Watterson KG. Repair of aortic atresia and hypoplastic left heart syndrome without using graft material. Eur J Cardiothorac Surg 2000;17: 85-7.

8   Fraser Jr. CD, Mee RB. Modified Norwood procedure for hypoplastic left heart syndrome. Ann Thorac Surg 1995; 60 (6 suppl):S546-9.

9   Maruszewski B, Tobota Z. The European Congenital Heart Defects Surgery Data Base Experience: Pediatric. European Cardiothoracic Surgical Registry of the European Association for Cardio-Thoracic Surgery. Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery 2002:143-7.

10 Poirier NC, Drummond-Webb JJ, Hisamochi K, Imamura M, Harrison AM, Mee RB et al. Modified Norwood procedure with a high-flow cardiopulmonary bypass strategy results in low mortality without late arch obstruction. J Thorac Cardiovasc Surg 2000; 120: 875-84.

11 Malec E, Januszewska K, Kolz J, Pajak J. Factors influencing early outcome of Norwood procedure for hypoplastic left heart syndrome. Eur J Cardiothorac Surg 2000; 18: 202-6.

12 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; 62:70-7.

13 Gaynor JW, Mahle WT, Cohen MI, Ittenbach RF, DeCampli WM, Steven JM et al. Risk factors for mortality after Norwood procedure. Eur J Cardiothorac Surg 2002; 22:82-9.

14 Jacobs ML, Rychik J, Murphy JD, Nicolson SC, Steven JM, Norwood WI. Results of Norwood's operation for lesions other than hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 1995;110: 1555-62.

15 Mosca RS, Hennein HA, Kulik TJ, Crowley DC, Michelfelder EC, Ludomirsky A et al. Modified Norwood operation for single left ventricle and ventriculoarterial discordance: an improved surgical technique. Ann Thorac Surg 1997;64:1126-32.

16 Bu'Lock FA, Stumper O, Jagtap R, Silove ED, DeGiovanni JV, Wright JG et al. Surgery for infants with a hypoplastic systemic ventricle and severe outflow obstruction: early results with a modified Norwood procedure. Br Heart J 1995; 73:456-61.

17 Migliavacca F, Pennati G, Dubini G, Fumero R, Pietrabissa R, Urcelay G et al. Modeling of the Norwood circulation: effects of shunt size, vascular resistances, and heart rate. Am J Physiol Heart Circ Physiol 2001; 280: H2076-86.

18 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;126:504-10.

19 Ishino K, Stumper O, De Giovanni JJ, Silove ED, Wright JG, Sethia B et al. The modified Norwood procedure for hypoplastic left heart syndrome: early to intermediate results of 120 patients with particular reference to aortic arch repair. J Thorac Cardiovasc Surg. 1999; 117:920-30.