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The hypoplastic left heart syndrome is not a risk factor for Fontan operation

A síndrome do coração esquerdo hipoplásico não constitui fator de risco para operação de Fontan

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Abstract

Objective: **To demonstrate the hospital mortality of children undergoing the Fontan operation and determine whether the hypoplastic left heart syndrome (HLHS) is a risk factor for mortality.**

Methods: **From August 2001 to June 2008, 76 patients underwent Fontan operation, and were divided into two groups: group A with 54 patients, being 31 (40.7%) patients with tricuspid atresia and variants, six (7.8 %) patients of left ventricle double inlet tract, four (5.3%) patients of complete atrioventricular septal defect and 13 (17.1%) patients of other complex congenital heart disease and Group B consisting of patients with HLHS, a total of 22 (28.9%) patients.**

Results: **Patients in group A had a mean age of 6.47 years ± 4.83 and those in group B of 2.08 years ± 0.24 P <0.001, the average weight was 22.42 ± 11.04 against 12.99 ± 1.2 *P* = 0.016, the mean CPB time was 119.5 min against 113.3 min *P* = 0.0 with an average time of clamping of 74.8 min and 73.5 min *P* = 0.75. The mean ICU stay was 4.1 days for group A against 7.52 days for the group B *P* = 0.0003. In total (group A + B), three patients died, with a hospital mortality of 3.9%, being one patient with HLHS (1.3%) (*P* <0.001, 95% CI 0.001 to 0.228).**

Conclusion: **Our study shows that despite higher morbidity, the HLHS is not a risk factor for hospital mortality.**

Descriptors: **Congenital heart defects/surgery. Echocardiography. Hypothermia.**

Resumo

Objetivo: **Demonstrar a mortalidade hospitalar de crianças submetidas à operação de Fontan e determinar se a síndrome do coração esquerdo hipoplásico (SCEH) constitui fator de risco para mortalidade.**

Métodos: **De agosto 2001 a junho 2008, 76 pacientes foram submetidos à operação de Fontan, sendo divididos em dois grupos: grupo A com 54 pacientes, sendo 31 (40,7%) portadores de atresia tricúspide e variantes, seis (7,8%) de dupla via de entrada ventrículo esquerdo, quatro (5,3%) de defeito do septo atrioventricular total e 13 (17,1%) de outras cardiopatias congênitas complexas; e grupo B constituído por portadores de SCEH, num total de 22 (28,9%) pacientes.**

Resultados: **Os pacientes do grupo A tiveram média de idade de 6,47 anos ± 4,83 e do grupo B de 2,08 anos ± 0,24 *P*<0,001; a média de peso foi de 22,42 ± 11,04 contra 12,99 ± 1,2 *P*=0,016; o tempo médio de CEC foi de 119,5 min contra 113,3 min *P*=0,0, com tempo médio de pinçamento aórtico de 74,8 min e 73,5 min *P*= 0,75. O tempo médio de permanência em UTI foi 4,1 dias para o grupo A contra 7,52 dias para o grupo B *P*= 0,0003. No total (grupo A + B), três pacientes foram a óbito, com mortalidade hospitalar de 3,9%, sendo um paciente portador de SHCE (1,3%) (*P*<0,001; IC95% 0,001 - 0,228).**

Conclusão: **Nosso estudo evidencia que, apesar de maior morbidade, a SCEH não constitui um fator de risco para mortalidade hospitalar.**

Descritores: **Cardiopatias congênitas/cirurgia. Ecocardiografia. Hipotermia.**

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INTRODUCTION

After introducing the concept of total cavopulmonary anastomosis in 1971 by Fontan and Baudet [1] for the treatment of children with functional single ventricle, major changes have occurred in recent years, including the surgical techniques, indications and postoperative management. Some changes, such as the surgical staging of heart disease, fenestration of the conduit, lateral tunnel, extracardiac Fontan and the improvement of preoperative and postoperative, either by the evolution of diagnostic methods or by introducing new drugs, made the morbidity and mortality of the Fontan operation dramatically diminished over the years.

However, despite this great progress, some studies suggest that the hypoplastic left heart syndrome (HLHS) is a risk factor for mortality in children submitted to the Fontan operation [2] and others show no increased mortality [3]. Therefore, the proposal of this study is to evaluate the mortality rate of Fontan operation and determine if the HSLS is a risk factor.

METHODS

By means of surveying were evaluated the medical records of all cases of patients undergoing the Fontan procedure between August 2001 and June 2008, by the cardio-surgical team of Dr. José Pedro da Silva at Hospital Beneficência Portuguesa of São Paulo. The protocol used for this work was approved by the Ethics Research Committee of the institution (Number 0007036000008).

We included in group A all patients undergoing the Fontan operation, except those with HLHS, and in group B, all patients undergoing the Fontan procedure holding HLHS. Regarding the diagnosis, 22 (28.9%) patients had HLHS, 31 (40.7%) with tricuspid atresia and its variants, six (7.8%) of left ventricle double-inlet tract, four (5 3%) of complex complete atrioventricular septal defect and the 13 (17.1%) remaining from other complex congenital heart disease. We made comparisons between groups A and B, in addition to mortality, the following variables: weight, age, gender, duration of cardiopulmonary bypass (CPB), aortic clamping time and length of stay in ICU. In order to verify the association between the successful events and hospital deaths by HSLS, it was used the Fisher's exact test, $P < 0.05$ and the confidence interval of 95%.

Surgical technique

All patients underwent surgery using conventional cardiopulmonary bypass with membrane oxygenator, with insertion of an arterial cannula in the distal ascending aorta and bicaval cannulation for venous return, under moderate hypothermia (25 ° C) and performed myocardial protection with cold blood by anterograde tract. In all cases, ultrafiltration was performed during CPB. The surgical technique used to perform the Fontan operation was intracardiac lateral tunnel in 25 (32.8%) patients, extracardiac conduit in seven (9.2%) patients and extracardiac conduit made in 44 (57.8%) patients. The extracardiac conduit was constructed using composite tissue of PTFE (polytetrafluoroethylene) fenestrated for the construction of the medial wall and autologous pericardium *in situ* in its sidewall, which may promote growth of the tube, following the child's development.

RESULTS

Out of the 76 patients who underwent Fontan operation, three patients died, with a hospital mortality of 3.9%, being one (4.54%) patient with HLHS. The results are shown in Table 1.

Table 1. Immediate results of the Fontan operation.

Group	Group A	Group B	*P*
Number of patients	54	22	
Age (mean ± SD)	6.47 years ± 4.83	2.08 years ± 0,24	0.0001
Male (n / %)	34(60.7%)	12(54.5%)	0.389
Weight (mean ± SD)	22.42 ± 11.04	12.99 ± 1.2	0.016
CPB time (mean)	119.5 min	113.3 min	
Aortic clamping time (mean)	74.8 min	73.5 min	0.75
ICU stay (mean)	4.1 days	7.52	0.0003
Mortality	3.57%	4.54%	*P*<0.001 IC95% (0.001-0.228)

SD: standard deviation; CPB: cardiopulmonary bypass

There was no significant difference between the CPB and aortic clamping times between the groups.

Age and weight were significantly lower in group B, which may have contributed to longer stays in ICU in this group of patients.

DISCUSSION

The evolution of the management of neonates with single ventricle in recent years has increased the number of patients eligible for the Fontan operation. This increase is due in part to the great development of diagnostic methods, especially in echocardiography, which allowed the early diagnosis of various heart diseases and, consequently, their immediate intervention, combined obviously with the appropriate surgical techniques for maintaining adequate pulmonary blood flow. Furthermore, the introduction of prostaglandin and its fundamental use in cyanogenic heart diseases allowed newborns to be elective for surgery, largely avoiding the inconvenience of emergency surgeries.

Major advances have occurred, also with the Fontan operation, with a variety of technical modifications [1-4], including surgical staging between the partial bidirectional cavopulmonary anastomosis (Glenn) and total cavopulmonary anastomosis [5,6], the creation of a intra-atrial lateral tunnel [7] from right to left shunt through fenestration of a conduit or intra-atrial patch [8-10] and the creation of an extracardiac conduit [10]. More recently, some studies have shown that ultrafiltration would be another additive to improve performance. Thus, the morbidity and mortality of children undergoing the Fontan operation have declined dramatically in recent years, also influencing in this result the improvement of CPB, the anesthetic management and ICU care.

Gentles et al. [11], from the Children's Hospital in Boston, published a review article from 1973 to 1991, where 500 children underwent Fontan operation. In this study, it was found that mean high pulmonary arterial pressure, very young children, presence of heterotaxy syndrome and systemic atrioventricular valve as the tricuspid valve were identified as risk factors for the failure of the operation. HLHS occurred in less than 10% of cases and it was identified as a risk factor for failure.

Van Arsdell et al. [12], University of Toronto, published a study with 100 consecutive Fontan operations between 1991 and 1995. The mortality of the first 50 patients was 16% versus 0% in the last 50. They concluded that although the characteristics and risk factors of patients being equal, some innovations such as extracardiac Fontan and modified ultrafiltration after CPB significantly decreased the mortality of patients with the most recent surgeries.

Mosca et al. [13], the University of Michigan, published in 2000 a study with 100 patients who underwent Fontan operation between 1992 and 1998. All patients had HLHS, with two different surgical techniques employed in this group. Over the past five years of review, they have shown excellent results, with success rates of 98%, credited to the use of modified ultrafiltration and also the introduction of fenestration of the intra-atrial lateral tunnel.

Koutlas et al. [14], Children's Hospital of Philadelphia, reported good results in the Fontan operation after the use of modified ultrafiltration, concluding that this procedure contributes to the improvement of surgical outcomes. Several large referral centers have reported great hemodynamic improvement of the patients undergoing the Fontan operation after the introduction of right-left shunt [15,16], including improvement of preload, cardiac output, oxygen delivery, decreased central venous pressure and decreased of pleural effusions.

Tweddell et al. [17], Children's Hospital of Wisconsin, reported excellent results for the Fontan operation, with a mortality of about 3% for a total of 256 patients. The following aspects were considered surgery failure in this study: death, need for cardiac transplantation or need to undo the surgery.

Meyer et al. [18], Children's Hospital of Philadelphia, published a study of 160 children undergoing Fontan operation, from January 2000 to December 2004 using CPB and aortic clamping, with a success rate of 98%. Out of the patients operated, 71% had HLHS.

Clearly, in our group, the management of patients with single ventricle undergoing Fontan operation has evolved in recent years, many of these advances being incorporated in our patients, and crediting this success largely to the improvement of CPB, as well as the use ultrafiltration, causing it to decrease the inflammatory process of the CPB. Another determining factor is the uniformity of conduct in the postoperative period, with specific protocols for the management of such patients, being that in the last three years it has been also reduced the length of stay in ICU.

CONCLUSION

Our study shows a considerable number of patients undergoing the Fontan operation, patients with HLHS and it clearly demonstrates the reduction of hospital mortality in recent times, equating to the results of major international centers. Moreover, this shows that, despite the time of ICU admission to be higher in HLHS, it is not a risk factor for mortality in this stage of treatment, showing the great progress in dealing with this disease. The continuity of this work is becoming so vital for the monitoring and evolution of patients in the medium and long term, to determine not only morbid-mortality but also quality of life of these patients.

REFERENCES

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax. 1971;26(3):240-8.

2. Mahle WT, Spray TL, Wernovsky G, Gaynor JW, Clark BJ 3rd. Survival after reconstructive surgery for hypoplastic left heart syndrome: a 15-year experience from a single institution. Circulation. 2000;102(19 Suppl 3):III136-41.

3. Mosca RS, Kulik TJ, Goldberg CS, Vermilion RP, Charpie JR, Crowley DC, et al. Early results of the fontan procedure in one hundred consecutive patients with hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 2000;119(6):1110-8.

4. Fontan F, Deville C, Quaegebeur J, Ottenkamp J, Sourdille N, Choussat A, et al. Repair of tricuspid atresia in 100 pacients. J Thorac Cardiovasc Surg. 1983;85(5):647-60.

5. Hopkins RA, Armstrong BE, Serwer GA, Peterson RJ, Oldham HN Jr. Physiological rationale for a bidirectional cavopulmonary shunt. A versatile complement to the Fontan principle. J Thorac Cardiovasc Surg. 1985;90(3):391-8.

6. Mazzera E, Corno A, Picardo S, Di Donato R, Marino B, Costa D, et al. Bidirectional cavopulmonary shunts: clinical applications as staged or definitive palliation. Ann Thorac Surg. 1989;47(3):415-20.

7. de Leval MR, Kilner P, Gewillig M, Bull C. Total cavopulmonary connection: a logical alternative to atriopulmonary connection for complex Fontan operations. Experimental studies and early clinical experience. J Thorac Cardiovasc Surg. 1988;96(5):682-95.

8. Bridges ND, Lock JE, Castaneda AR. Baffle fenestration with subsequent transcatheter closure. Modification of the Fontan operation for patients at increased risk. Circulation. 1990;82(5):1681-9.

9. Bridges ND, Mayer JE Jr, Lock JE, Jonas RA, Hanley FL, Keane JF, et al. Effect of baffle fenestration on outcome of the modified Fontan operation. Circulation. 1992;86(6):1762-9.

10. Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit. A new form of right heart bypass. J Thorac Cardiovasc Surg. 1990;100(2):228-32.

11. Gentles TL, Mayer JE Jr, Gauvreau K, Newburger JW, Lock JE, Kupferschmid JP, et al. Fontan operation in five hundred consecutive patients: factors influencing early and late outcome. J Thorac Cardiovasc Surg. 1997;114(3):376-91.

12. Van Arsdell GS, McCrindle BW, Einarson KD, Lee KJ, Oag E, Caldarone CA, et al. Interventions associated with minimal Fontan mortality. Ann Thorac Surg. 2000;70(2):568-74.

13. Mosca RS, Kulik TJ, Goldberg CS, Vermilion RP, Charpie JR, Crowley DC, et al. Early results of the Fontan procedure in one hundred consecutive patients with hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 2000;119(6):1110-8.

14. Koutlas TC, Gaynor JW, Nicolson SC, Steven JM, Wernovsky G, Spray TL. Modified ultrafiltration reduces postoperative morbidity after cavopulmonary connection. Ann Thorac Surg. 1997;64(1):37-42.

15. Hijazi ZM, Fahey JT, Kleinman CS, Kopf GS, Hellenbrand WE. Hemodynamic evaluation before and after closure of fenestrated Fontan. An acute study of changes in oxygen delivery. Circulation. 1992;86(1):196-202.

16. Bridges ND, Lock JE, Mayer JE Jr, Burnett J, Castaneda AR. Cardiac catheterization and test occlusion of the interatrial communication after the fenestrated Fontan operation. J Am Coll Cardiol. 1995;25(7):1712-7.

17. Tweddell JS, Nersesian M, Mussatto KA, Nugent M, Simpson P, Mitchell ME, et al. Fontan palliation in the modern era: factors impacting mortality and morbidity. Ann Thorac Surg. 2009;88(4):1291-9.

18. Meyer DB, Zamora B, Spray TL. Outcomes of the Fontan procedure using cardiopulmonary bypass with aortic cross-clamping. Ann Thorac Surg. 2006;82(5):1611-8.