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Adults with congenital heart disease undergoing first surgery: prevalence and outcomes at a tertiary hospital

Adultos com cardiopatia congênita submetidos à primeira cirurgia: prevalência e resultados em um hospital terciário

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Abstract

Introduction: **Surgical treatment of congenital heart disease in adults showed a significant growth in recent years. But even so, the number of patients who reach adulthood without adequate surgical treatment remains high.**

Objective: **To demonstrate the results and hospital diagnoses of adult patients with congenital heart disease underwent the first surgery.**

Methods: **A retrospective analysis of records of patients operated for correction of congenital heart disease and age greater than or equal to 18 years. The exclusion criterium was surgery for reoperation. Period analyzed was from December 2007 to December 2010 with inclusion of 79 patients.**

Results: **The atrial septal defects were the most prevalent (53.1%), followed by VSD (15.2%), the coarctation (6.3%) and partial atrioventricular canal (6.3%). Thirteen (16.4%) patients had associated disease acquired and 14 (17.7%) congenital disease. Thirty-three (41.8%) patients had pulmonary hypertension. The average hospital stay in ICU and hospital were 3.9 and 14.5 days, respectively. Complications occurred in 18 (22.8%) patients, with infections being the most common. The hospital mortality was two (2.5%) patients.**

Conclusion: **The treatment of congenital heart disease in adults as first surgery has very favorable results. However, in our series, there was an increased length of stay in ICU and hospital.**

Descriptors: **Adult. Heart defects, congenital. Cardiovascular surgical procedures.**

Resumo

Introdução: **O tratamento cirúrgico da cardiopatia congênita em adultos apresentou importante crescimento nos últimos anos. Contudo, ainda assim, o número de pacientes que atingem a idade adulta sem tratamento cirúrgico adequado permanece elevado.**

Objetivo: **Avaliar os resultados hospitalares e diagnósticos dos pacientes adultos com cardiopatia congênita submetidos à primeira operação.**

Métodos: **Estudo retrospectivo, que analisou prontuários de pacientes operados para correção de cardiopatia congênita com idade maior ou igual a 18 anos. O critério de exclusão foi cirurgia para reoperação. Foi analisado o período entre dezembro de 2007 e dezembro de 2010, com inclusão de 79 pacientes.**

Resultados: **Os defeitos do septo atrial foram os mais prevalentes (53,1%), seguidos de comunicação interventricular (15,2%), coarctação da aorta (6,3%) e**

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Abbreviations, acronyms and symbols	
AF	Atrial fibrilation
ASD	Atrial septal defect
CAVB	Complete atrioventricular block
CHDA	Congenital heart diseases in adults
IAC	Interatrial communication
ICU	Intensive care unit
IVC	Interventricular communication
POD	Postoperative day
SUS	Unified Health System
VSD	Ventricular septal defect

canal atrioventricular parcial (6,3%). Treze (16,4%) pacientes apresentavam doença associada adquirida e 14 pacientes (17,7%), congênita. Trinta e três (41,8%) pacientes apresentavam hipertensão pulmonar. O tempo médio de internamento em UTI e hospitalar foi de 3,9 e 14,5 dias, respectivamente. Complicações ocorreram em 18 (22,8%) pacientes, sendo as infecciosas as mais comuns. A mortalidade hospitalar foi de dois (2,5%) pacientes.

Conclusão: **O tratamento da cardiopatia congênita em adultos como primeira cirurgia apresentou resultado bastante favorável. Contudo, em nossa série, houve maior tempo de internamento em UTI e hospitalar.**

Descritores: **Adulto. Cardiopatias congênitas. Procedimentos cirúrgicos cardiovasculares.**

INTRODUCTION

The surgical treatment of congenital heart disease in adults (CHDA) showed significant growth in recent decades [1,2], with predictions that in a few years the number of patients alive adults and children with congenital heart disease will be matched [3]. This is due to advances in treatment and diagnosis reached, however, still remains a multifactorial problem: the existence of a significant portion of patients surgically treated in adulthood that should have been corrected in childhood or adolescence.

In our environment, socioeconomic factors are identified in a significant portion of the problem. But nations have overcome the financial barrier continue to show their results with a significant proportion of patients treated as first surgery in adulthood [4].

Another factor that interferes with the proper treatment of this group of patients is currently recognizes the need to monitor patients with CCA in centers specializing in this type of disease, which differs from the adult acquired heart disease and children with congenital heart disease [3].

The objective of this study is to evaluate the results and hospital diagnoses of patients surgically treated adults with congenital heart disease as initial surgery in a tertiary hospital with structuring latest in cardiology and cardiovascular surgery.

METHODS

Retrospective study through analysis of medical records of patients who underwent surgery as the first surgery and congenital heart disease aged greater than or equal to 18 years. Patients who underwent reoperation in the same hospital performed the first surgery as well as patients with acquired diseases associated with congenital heart disease were not excluded.

The exclusion criterion was the patient admitted for reoperation.

There were analyzed the patients operated for correction of bicuspid aortic valve, because the intraoperative echocardiographic diagnosis and were often not documented, leading to a figure that does not represent reality.

The diagnosis of pulmonary hypertension was considered when there was a mean pressure greater than 25 mmHg or systolic blood pressure greater than 30 mmHg [5].

The charts in the period between December 2007 and December 2010, 79 patients with inclusion in this study.

RESULTS

There was a predominance of females with 49 (62%) patients. The mean age was 34 years (range 18-63 years), with 23 (29.1%) patients above 40 years. In Table 1, the diagnoses are being considered as the main congenital disease that prompted the surgery. As a secondary diagnosis was considered the associated disease (congenital or acquired), which was also treated at the same hospital.

Table 1. Diagnostics.

Main diagnosis	N (%)	Secondary diagnosis (N)
secundum ASD	40 (50.6)	MS (2) TR (5), PS (4), AoR (1) and CA (1)
Sinus venosus	2 (2.5)	ASD (1)
Partial atrioventricular canal	5 (6.3)	preoperatively CAVB (1)
Atrioventricular canal intermediate	1 (1.3)	-
Atrial septal aneurysm	1 (1.3)	TR
Interventricular communication	12 (15.2)	PS (5), AoR (2), MS (1), Endoc. (2)
Patent ductus arteriosus	1 (1.3)	
Aortic coarctatio	5 (6.3)	AoR (1) and CA (1)
Interrupted aortic arch I	1 (1.3)	AoS
Subaortic membrane	2 (2.5)	IVC (1)
Supra-aortic stenosis	1 (1.3)	Sd. Williams
Sinus of Valsalva aneurysm	2 (2.5)	IVC (1)
Cor triatriatum	1 (1.3)	PS, Situs inversus
Tetralogy of Fallot	4 (5.1)	-
Pulmonary stenosis	1 (1.3)	-

ASD = atrial septal defect, MS = mitral stenosis, TR = tricuspid regurgitation, PS = pulmonary stenosis, AoR = aortic regurgitation, CA = coronary atherosclerosis, CAVB = complete atrioventricular block, Endoc. = Endocarditis, AoS = aortic stenosis, IVC = interventricular communication, Sd. = syndrome

The atrial septal defects were the most prevalent with 53.1% (42 patients). Two cases presented with unique physiological atrium, yet both still existed a remnant of the septal region of the tricuspid annulus and are not therefore classified as single anatomical atrium.

The association of atrial septal defect (ASD) with mitral stenosis was present in two (2.5%) patients, which characterizes the syndrome Lutembacher.

The second most prevalent disease was congenital ventricular septal defect (VSD), and in all cases classified as perimembranous. Only one patient had association with other defect perimembranous VSD muscular type. In two patients with small defects and hemodynamic repercussion, surgery was performed for the presence of endocarditis. In one of them there was a significant involvement of the tricuspid valve, which required repair, persisting with moderate impairment postoperatively.

Tetralogy of Fallot was treated in four patients and transannular enlargement was employed in both cases. One of the patients had VSD doubly related, featuring the Eastern Fallot.

The presence of associated disease occurred in 27 (34.2%) patients, pulmonary stenosis associated with congenital disease most frequent (10 patients, 12.6%). The tricuspid regurgitation was the most common acquired disease, occurring in 5 (6.3%) patients.

The involvement of the aortic valve was present in 5 patients, but only one diagnosed with bicuspid. In another patient with VSD, the mechanism was the collapse of the non-coronary leaflet. The others had no definite etiology.

Table 2. Hospital complications.

complication	N
Respiratory infection	7
Reoperation*	6
Atrial fibrillation	5
Moderate-severe pericardial effusion	3
Pleural effusion	2
Postoperative bleeding	2
CAVB provisional	2
CAVB permanent	1
mediastinitis	1
Wound infection	1
Urinary tract infection	1
ICS	1
pneumomediastinum	1

(*) = included bleeding and mediastinitis, CAVB = complete atrioventricular block, ICS = ischemic cerebral stroke

The mean hospital stay was 14.3 days, ranging from 5 to 99 days. The average length of stay in the intensive care unit (ICU) ranged from 2 to 29 days, with an average of 3.9 days.

In Table 2, are related complications hospital. These occurred in 18 (22.8%) patients, however with more than one event per patient in some cases. The most common were infections (nine cases - 11.4%). Six patients underwent

reoperation, two for bleeding and one for mediastinitis. In two other patients, we needed a valve replacement plasties unsatisfactory (mitral and aortic) and one case of rupture of the patch VSD and hemodynamic instability on day 4 postoperatively (POD) in a patient corrected with Fallot, which until then, was quite favorable evolution. This was reoperated of urgency, showing excellent progress.

Complications related to cardiac rhythm occurred in 8 (10.1%) patients, atrial fibrillation (AF) the most common. In two cases of complete atrioventricular block (CAVB) provisional, the first was reversed on ICU admission, in a patient who had undergone VSD correction. The second CAVB began on the 1st postoperative day in a patient subjected to correction of ostium secundum IAC, being reversed in the 3rd POD.

In patients with pericardial effusion in two drainage was necessary. Among patients with pleural effusion, was necessary to perform a thoracentesis.

A patient who presented with neurological deficit ischemia diagnosis confirmed by CT. This patient had already submitted event preoperatively classified as transient ischemic attack, but postoperatively the new change persisted for more than 48 h. However, the patient was discharged with neurological symptoms resolved without deficit.

In 76 (96.2%) patients, the preoperative rhythm was sinus, two had AF and CAVB. At hospital discharge, the two patients with preoperative AF and one with postoperative AF remained arrhythmia. The CAVB patients with preoperative pacemaker implanted and one case of permanent CAVB died. Then, at hospital discharge, 94.9% of patients were in sinus rhythm (excluding the patient who died).

Cyanosis was present in only 5 (6.3%) patients in this series, presenting them with a significant and polycythemia hematocrit above 65%.

Pulmonary hypertension was diagnosed in 33 (41.8%) patients.

The catheter was used in 36 (45.6%) patients, in order to complement the diagnosis and / or coronary angiography. In five (6.3%) patients, angiography was used.

Mortality was two (2.5%) patients. The first was a young patient with secundum ASD and mild pulmonary hypertension, which had an uneventful surgery, but evolved with lung respiratory distress syndrome of acute on the 1st POD and progressive worsening. Postoperative tests did not identify cardiovascular causes. The patient died on POD 23 due to respiratory failure. The second case was a patient with partial atrioventricular canal and cavity very serious increase of the four heart chambers. The patient was operated and closure of cleft mitral wide. The valve was insufficient, then was re done and return. CAVB presented to the 1st POD, followed by FA with adequate response has not been indicated pacemaker. However, after a prolonged postoperative course, now in recovery, had severe bradycardia at 28 POD, not having access to a temporary pacemaker. This patient was the only case that would require a permanent pacemaker complication in this series (1.3%).

DISCUSSION

The surgical treatment of congenital heart disease in adults is increasing and with that there is the need for a multidisciplinary team to monitor these patients [3]. Our hospital is a tertiary unit with exclusive service to the Unified Health System (UHS), and started its activities in cardiology and cardiovascular surgery Cardiopediatrics in August 2006. As of December 2007, there was increased interest in the treatment of CCA. Then at the end of 2010, was created a specific clinic for these patients. Having an initial target of operating 4-8 patients per month since 2008, was achieved only an average of 2.4 cases / month in the last three years. Several factors are implicated in this deficit and this discussion is beyond the aim of this work. However, it is noteworthy that two large European series [4,6] were published recently, with an incidence of 69% and 75% of first surgeries in the treatment of CCA. Thus, it is clear that serious socioeconomic problems, such as occur in our environment, are not the only factors responsible for the large number of patients with CCA without initial surgical treatment.

In 2007, a European study was published with 2012 patients from 19 different centers [4]. Of these, in 1509 patients, treatment of the CCA was offered as the first surgery. However, this study analyzed the congenital aortic valve (10.7%), which did not occur in our series. Thus, we performed a comparison of diagnoses found in our work and in the European study, however excluding the percentage of patients treated aortic valve in the latter. There was a similarity in prevalence of diagnoses in general, and the ostium secundum IAC, septal defect and aortic coarctation partial atrioventricular were diseases most prevalent in both studies, and our share of them, respectively: 48.2% *versus* 50.6%, 7.2% *versus* 6.3% and 4% *versus* 6.3%.

The interventricular communications were also frequent, but differ in higher percentage (15.2% versus 7.3%), getting our group with a high percentage of this type of disease. Initially this difference is justified because it is a disease with early manifestation and therefore treated in childhood. But the reality in our country is still different, getting many patients without treatment. Another explanation lies in the associated diseases, which in our series have pulmonary stenosis diagnosed in almost half of patients with VSD. There were also two cases of small

VSDs and mild rebound, treated for endocarditis. That is, of the 12 patients treated with VSD in our study, 7 there was the evolution of communications unusual large, which may be related to the possibility of treatment in adulthood. The European study makes no reference to individualized presence of associated disease.

Work on the treatment of tetralogy of Fallot demonstrated, even in our country, the majority of patients are treated in adulthood presents good performance and favorable anatomy diseases [7,8]. However, in our series, the four patients operated on in two there was a need for transannular enlargement, increasing the proportion of patients with unfavorable anatomy, which may be due to the small number of patients in this analysis. We should then await future results with more patients to confirm whether or not this reality in our population. All recovered uneventfully.

The complication rate in our group was 22.8%, as being the most prevalent infections. Cardiac arrhythmias were also very frequent with 10.1% and 7.6% with reoperations. These data are in accordance with the findings of the study of Padalino et al. [6] in a series of 628 patients undergoing primary surgery and CHDA. Only the incidence of infection was not reported by the group. However, the group of Putman et al. [9] in a 17-year experience with 830 patients with CHDA, reported hospital morbidity of 33.1%, with cardiac arrhythmia being the most frequent. This result is higher than ours, but the group Putman evaluated the mechanical ventilation as a complication and it was not done by us. Another factor to explain this difference was the presence of complex cases in the study by Putman et al. [9], which certainly are related to increased morbidity, although this inference was not reported in the study. The infection rate was 2.8% of them being below the presented in this study.

The rate of ICU stay in this series was higher when compared to the results of Padalino et al. [6] (1.3 versus 3.9 days) and European study [4] (2.4 versus 3.9 days). The same happens with our rate of hospitalization (10.5 versus 14.4 days) [4]. The learning curve and the adequacy of an initial service are heavily involved in this difference, moreover, the incidence of infection in this series (11.4%) with increased need for antibiotics that may be associated with longer hospitalization.

A major concern in the treatment of patients with CHDA is the presence of pulmonary hypertension, since the increased pulmonary with possibility of developing hyper-fixed lung resistance occurs in these patients. Studies demonstrate that 10% to 15% of these patients develop pulmonary hypertension in various degrees [10,11]. In the present study, pulmonary hypertension was present in 33 (41.8%) patients. But even in these patients, there was no greater difficulty postoperatively. So, an interesting reference is the work of Sachweh et al. [12] demonstrated that in a number of adults with IAC and pulmonary hypertension, that even patients with severe hypertension preoperatively, had a good postoperative course, and found no relationship between pulmonary pressure preoperative biopsy pulmonary and surgical morbidity. This study brings to light a discussion of the methods and results of the evaluation of pulmonary hypertension, as well as the evolution of these patients in a specific type of disease. This is beyond the aim of our study, mainly by the heterogeneity of our group with pulmonary hypertension, but it is pertinent to note that our patients had good graft.

The preoperative diagnostic evaluation of congenital heart disease is made in large numbers of patients by echocardiography combined with Doppler mainly in the study of diseases of small to medium complexity. However, when this occurs in CHDA, other factors such as the need for coronary angiography or evaluation of pulmonary hypertension, necessitate the use of cardiac catheterization. In our group, while not presenting complexes cases, the use of catheterization was 45.6% (36 patients), which corresponds to a percentage higher than found in other studies [4,6]. Even with 29.1% of our patients having age above 40 years, still considered high indication for catheterization. One explanation may lie in the lack of uniformity in our conduct with patients who were prepared by different doctors and some of them were referred from another hospital with preoperative study performed. However, with the specialized ambulatory outpatient surgical center, we hope to reduce this number or even confirm an increased need this method in our population.

Patients with CHDA, to be operated later, when structural changes are more pronounced cardiopulmonary should have higher mortality. However, several authors demonstrate low mortality rate, even in our country [1,4,6,9,13-16], even when excluding patients treated by ostium secundum IAC, for these represent a simpler disease and low mortality. In our group there were also low mortality (2.5%), emphasizing that it is a relatively new service. However, complex congenital heart disease were not treated or palliative surgeries performed in this series, configuring these, especially the hearts of univentricular physiology as risk factors for hospital mortality [6].

Limitations of the study

The main limitation of this study is that it is retrospective, with all the biases of this type of research. The lack of outpatient data also hampers further analysis of the results, but in our country, with a large proportion of patients coming from the state, the loss to follow-up is still very common. But, with the implementation of CCA clinic in our hospital, we are confident that this monitoring can be done more systematically.

CONCLUSION

The treatment of CCA as first surgery presents a very favorable outcome. However, in this series, there was a longer hospital and ICU.

REFERENCES

1. Moons P, Engelfriet P, Kaemmerer H, Meijboom FJ, Oechslin E, Mulder BJ; Expert Committee of Euro Heart Survey on Adult Congenital Heart Disease. Delivery of care for adult patients with congenital heart disease in Europe: results from the Euro Heart Survey. Eur Heart J. 2006;27(11):1324-30.

2. Hoffman JI, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. Am Heart J. 2004;147(3):425-39.

3. Webb GD. Care of adults with congenital heart disease: a challenge for the new millennium. Thorac Cardiovasc Surg. 2001;49(1):30-4.

4. Vida VL, Berggren H, Brawn WJ, Daenen W, Di Carlo D, Di Donato R, et al. Risk of surgery for congenital heart disease in the adult: a multicentered European study. Ann Thorac Surg. 2007;83(1):161-8.

5. Carvalho ACC, Almeida RD, Lopes AA. Diagnóstico da hipertensão pulmonar. Diagnóstico, avaliação e terapêutica da hipertensão pulmonar. Diretrizes da Sociedade Brasileira de Cardiologia. 2005:20-8. Disponível em: http://publicacoes.cardiol.br/consenso/2005/039.pdf

6. Padalino MA, Speggiorin S, Rizzoli, Crupi G, Vida VL, Bernabei M, et al. Midterm results of surgical intervention for congenital heart disease in adults: an Italian multicenter study. J Thorac Cardiovasc Surg. 2007;134(1):106-13.

7. Moraes RC, Rodrigues JV, Gomes CA, Tenório EA, Neto FM, Santos CL, et al. Tratamento cirúrgico da Tetralogia de Fallot em Adultos. Rev Bras Cir Cardiovasc. 1991;6(2):80-4.

8. Beach PM Jr, Bowman FO Jr, Kaiser GA, Malm JR. Total correction of tetralogy of Fallot in adolescents and adults. Circulation. 1971;43(5 Suppl):I37-43.

9. Putman LM, van Gameren M, Meijboom FJ, de Jong PL, Roos-Hesselink JW, Witsenburg M, et al. Seventeen years of adult congenital heart surgery: a single centre experience. Eur J Cardiothorac Surg. 2009;36(1):96-104.

10. Diller GP, Gatzoulis MA. Pulmonary vascular disease in adults with congenital heart disease. Circulation. 2007;115(8):1039-50.

11. Steele PM, Fuster V, Cohen M, Ritter DG, McGoon DC. Isolated atrial septal defect with pulmonary vascular obstructive disease: long-term follow-up and prediction of outcome after surgical correction. Circulation. 1987;76(5):1037-42.

12. Sachweh JS, Daebritz SH, Hermanns B, Fausten B, Jockenhoevel S, Handt S, et al. Hypertensive pulmonary vascular disease in adults with secundum or sinus venosus atrial septal defect. Ann Thorac Surg. 2006;81(1):207-13.

13. Stellin G, Vida VL, Padalino MA, Rizzoli G; European Congenital Heart Surgeons Association. Surgical outcome for congenital heart malformations in the adult age: a multicentric European study. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:95-101.

14. Srinathan SK, Bonser RS, Sethia B, Thorne SA, Brawn WJ, Barron DJ. Changing practice of cardiac surgery in adult patients with congenital heart disease. Heart. 2004;91(2):207-12.

15. Jatene MB, Abuchaim DCS, Junior JLO, Riso A, Tanamati C, Miura N, et al. Resultados do tratamento cirúrgico da coarctação de aorta em adultos. Rev Bras Cir Cardiovasc. 2009;24(3):346-53.

16. Lisboa LAF, Abreu Filho CAC, Dallan LAO, Rochitte CE, Souza JM, Oliveira SA. Tratamento cirúrgico da coarctação do arco aórtico em adulto: avaliação clínica e angiográfica tardia da técnica extra-anatômica. Rev Bras Cir Cardiovasc. 2001;16(3):187-94.