Marcelo Felipe Kozak

Fatores Associados à Insuficiência Moderada ou Importante da Valva Atrioventricular Esquerda no Primeiro Mês após Correção de Defeito de Septo Atrioventricular

São José Do Rio Preto

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Orientador: Prof. Dr. Airton Camacho Moscardini

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SUMÁRIO

DEDICATÓRIA

Dedico esse trabalho à minha filha Beatriz, meu maior orgulho e razão de viver. Seu carinho, sua voz, sua inocência pueril e sua beleza sobrenatural, algo que apenas um pai enxerga no filho, são apaixonantes e estimulantes.

Devo, também, dedicar esse trabalho à minha esposa Ana Carolina, por seu amor, amizade, respeito e companheirismo.

Não posso deixar de lembrar de meus pais, Jorge e Indiaoara, responsáveis pela minha formação moral e educacional, da qual muito me orgulho. Mesmo com a perda de um filho, meu irmão Igor, continuam admirando e incentivando seus outros filhos a alçarem voos cada vez mais altos, sempre seguindo os preceitos ensinados a nós desde a infância: honestidade, perseverância e humildade. Além disso, têm a capacidade de me fazer sentir querido em qualquer momento.

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EPÍGRAFE

“Vous êtes belles, mais vous êtes vides. On ne peut pas mourir pour vous. Bien sûr, ma rose à moi, un passant ordinaire croirait qu'elle vous ressemble. Mais à elle seule elle est plus importante que vous toutes, puisque c'est elle que j'ai arrosée. Puisque c'est elle que j'ai mise sous globe. Puisque c'est elle que j'ai abritée par le paravent. Puisque c'est elle dont j'ai tué les chenilles (sauf les deux ou trois pour les papillons). Puisque c'est elle que j'ai écoutée se plaindre, ou se vanter, ou même quelquefois se taire. Puisque c'est ma rose.**”**

“Sois belas, mas vazias. Não se pode morrer por vós. Minha rosa, sem dúvida um transeunte qualquer pensaria que se parece convosco. Ela sozinha é, porém, mais importante que vós todas, pois foi ela que eu reguei. Foi nela que pus a redoma. Foi ela que abriguei com o para-vento. Foi dela que matei as larvas. Foi ela que escutei queixar-se ou gabar-se, ou mesmo calar-se algumas vezes. É a minha rosa.”

“O Pequeno Príncipe” (cap. 21)

Antoine de Saint-Exupéry

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LISTA DE ABREVIATURAS E SÍMBOLOS

AV: atrioventricular

DSAV: defeito do septo atrioventricular

CIA: comunicação interatrial

CIV: comunicação interventricular

IVAVE: insuficiência da valva AV esquerda

%: porcento

Artigo II

LAVVR: left atrioventricular valve regurgitation

AVSD: atrioventricular septal defect

AV: atrioventricular

ASD: atrial septal defect

VSD: ventricular septal defect

RESUMO

Introdução: Uma das complicações mais frequentes e importantes do tratamento cirúrgico do defeito de septo atrioventricular (DSAV) é a insuficiência residual da valva atrioventricular esquerda, tanto nas formas totais, como parciais e transicionais. Dessa forma, esse estudo foi conduzido para identificar fatores de risco associados à insuficiência da valva atrioventricular esquerda (IVAVE) de grau moderado ou importante nos primeiros 30 dias após correção de defeito de DSAV. *Métodos:* Dados de 104 pacientes com DSAV operados entre 2002 e 2010 foram avaliados retrospectivamente, sendo estudados os seguintes fatores de risco: idade e peso no momento da correção, ausência de síndrome de Down, grau de insuficiência da valva atrioventricular (AV) antes da correção, anormalidades na valva AV e uso de anuloplastia. Os pacientes foram separados em dois grupos de acordo com o tipo de DSAV: grupo I (total) e grupo II (parcial e transicional). Características dos 53 pacientes do grupo I: a mediana da idade foi de 6,7 meses e a do peso de 5,3 Kg; 86,8% tinham síndrome de Down; antes da operação, 26 pacientes apresentavam insuficiência pelo menos moderada da valva AV (49.1%); anuloplastia foi realizada em 34% dos pacientes; anormalidades na valva AV foram encontradas em 11.3% dos casos. Características dos 51 pacientes do grupo II: a mediana da idade foi de 4,1 anos e a do peso de 13,4 Kg; 37,2% tinham síndrome de Down; antes da operação, 23 pacientes apresentavam IVAVE pelo menos moderada (45,1%); anormalidades na valva AV foram encontradas em 17,6% dos casos; anuloplastia foi realizada em 21,6% dos pacientes. *Resultados*: Grupo I – Após a correção cirúrgica, 21 casos apresentaram IVAVE pelo menos moderada (39,6%). Pela análise multivariada, o único fator associado com IVAVE pelo menos moderada no pós-operatório foi ausência de

síndrome de Down (p = 0,03). Grupo II - Após a correção cirúrgica, 12 casos apresentaram IVAVE pelo menos moderada (23,5%). Pela análise univariada, apenas a ausência de síndrome de Down teve significância estatística (p = 0.02). Porém, após análise multivariada, nenhum dos fatores teve significância estatística. *Conclusão*: Ausência de síndrome de Down foi determinante de IVAVE moderada ou importante nos primeiros 30 dias após correção de DSAV total. Todavia, nenhum dos fatores estudados foi determinante para tais graus de IVAVE entre os pacientes com DSAV parcial e transicional.

ABSTRACT

Introduction: One of the most often and important complications after surgical treatment of atrioventricular septal defects is the left atrioventricular valve insufficiency. So, this study was conducted to identify risk factors for moderate or severe left atrioventricular valve regurgitation within 30 days of surgical repair of atrioventricular septal defects at our center. *Methods:* This was a retrospective study in which we evaluated the results of 104 consecutive patients that were operated on at our practice between 2002 and 2010. The following associated factors were considered: age, weight, Down syndrome, grade of preoperative atrioventricular valve regurgitation, abnormalities on the atrioventricular valve and the use of annuloplasty. Patients were separated in two groups according to type of AVSD: group I (complete) and group II (incomplete – partial and transitional). Characteristics of the 53 patients of the group I: the median patient age at the time of repair was 6.7 months; the median weight was 5.3 Kg; 86.8% had Down syndrome; at the time of preoperative evaluation, there were 26 cases with moderate or severe atrioventricular valve regurgitation (49.1%); annuloplasty was perfored in 34%; abnormalities on the valve were found in 11.3% of the cases. Characteristics of the 51 patients of the group II: The median patient age at the time of repair was 4.1 years; the median weight was 13.4 Kg; 37.2% had Down syndrome; at the time of preoperative evaluation, there were 23 cases with moderate or grater LAVVR (45.1%); abnormalities on the AV valve were found in 17.6% of the cases; annuloplasty was performed in 21.6% of the patients. *Results*: Group I - At the time of post-operative evaluation, there were 21 cases with moderate or severe left atrioventricular valve regurgitation (39.6%). After performing a multivariate analysis, the only significant factor associated with these grades of insufficiency within

30 days of surgical correction of complete atrioventricular septal defect was the absence of Down syndrome (p = 0.03). Group II - At the time of postoperative evaluation, there were 12 cases with moderate or greater LAVVR (23.5%). During univariate analysis, only absence of Down syndrome was statistically significant (p = 0.02). However, after a multivariate analysis, none of the factors reached significance. *Conclusion*: Absence of Down syndrome proved to be associated with moderate or severe post-operative left atrioventricular valve regurgitation in patients with complete AVSD. However, none of the factors studied was determinant of a moderate or greater LAVVR within the first 30 days of repair of incomplete AVSD at our center.

INTRODUÇÃO

A junção atrioventricular (AV) tem sua origem embriológica a partir do 25º dia de gestação, após a rotação para a direita do tubo cardíaco. Ao final da 5ª semana, o ventrículo esquerdo primitivo suporta grande parte da circunferência do canal AV, o qual tem sua luz ocupada por duas grandes massas mesenquimais, os coxins endocárdicos AV superior e inferior. A fusão desses coxins por volta da 6ª semana de gestação divide o canal AV no que serão os primórdios das junções AV direita e esquerda.(1)

A septação do canal AV é seguida, ao mesmo tempo, pela septação dos ventrículos e dos átrios. Um dos componentes da septação atrial é a chamada espinha vestibular, que se transformará na porção muscular inferior do septo atrial. Trata-se de uma massa mesenquimal originada da região dorsal, que cresce a partir da base atrial para se fundir, juntamente com a "capa" mesenquimal originada da fusão dos coxins endocárdicos, ao septum primum, originado do teto do átrio.(1)

A formação das valvas AV ocorre na sequência, com participação importante do miocárdio do canal AV para a formação da valva AV direita e do sistema de condução, e do coxim lateral esquerdo e da presença da via de saída do ventrículo esquerdo entre os anéis AV para formação adequada da valva AV esquerda.(1)

Anormalidades nesse processo dão origem ao defeito do septo atrioventricular (DSAV), também conhecido como defeito do canal AV. Por muitos anos, a falha completa ou incompleta na fusão dos coxins foi considerada como a única causa das diversas formas de DSAV, motivo pelo qual essa doença também ficou conhecida como defeito do coxim endocárdico. Hoje, a mesma importância tem sido dada à espinha

vestibular, motivo pelo qual o nome defeito do coxim endocárdico não tem sido mais utilizado.

A etiologia do DSAV é multifatorial, estando envolvidos fatores ambientais e genéticos. Entre os ambientais são citados: idade materna avançada, diabetes gestacional, uso de anti-tussígenos, história familiar de cardiopatia congênita e exposição paterna à radiação ionizante.[2] Com relação aos genéticos, acredita-se que algum gene do cromossomo 21 esteja envolvido na septação AV, dada a forte associação entre DSAV e síndrome de Down, mas até hoje esse gene não foi identificado. Por outro lado, em 2003, foi isolado o primeiro gene considerado fator de risco para DSAV em pacientes sem síndrome de Down, o gene CRELD1, localizado no cromossomo 3. Mutações nesse gene seriam responsáveis por parte dos casos.[3]

O DSAV acomete mais frequentemente filhos de mães acima de 34 anos e indivíduos com síndrome de Down, e tem uma incidência em torno de 0,35/1000 nascidos vivos/ano,[4] podendo chegando a 170/1000/ano em pacientes com síndrome de Down.[5]

Todos os casos de DSAV têm as seguintes características anatômicas: junção AV comum por meio de uma valva AV única com cinco folhetos (dois exclusivos do ventrículo direito, um do ventrículo esquerdo e outros dois, chamados folhetos-ponte, com porções para ambos os ventrículos, fazendo com que o componente esquerdo da valva tenha três folhetos); desproporção entre as vias de entrada e saída do ventrículo esquerdo, a última, mais alongada e estreita com a aorta anteriorizada e não "encaixada"; posição anômala dos músculos papilares do ventrículo esquerdo, que assumem uma relação mais ântero-posterior, ao contrário do usual, látero-medial.[6]

Sua classificação é feita de acordo com o número de orifícios valvares, a localização e o tamanho das comunicações intracardíacas. O DSAV total, ou forma completa, é o tipo mais comum e inclui um grande defeito septal com uma comunicação interatrial (CIA) ostium primum contígua a uma comunicação interventricular (CIV), geralmente de grande tamanho, e um orifício AV único para a valva AV comum, a qual "flutua" sobre o septo interventricular. Havendo fusão entre os folhetos-ponte, a valva fica dividida em dois orifícios, uma das características da forma parcial, que frequentemente inclui uma CIA ostium primum de grande tamanho e uma fenda na valva AV esquerda direcionada à porção mediana do septo interventricular.[7]

Há ainda a forma transicional, que se diferencia da parcial pela presença de uma CIV logo abaixo do plano AV, geralmente restritiva, e a forma intermediária, que difere da total pela presença de dois orifícios AV.[8] Casos mais raros sem comunicações intracardíacas já foram descritos,[9] demonstrando que a presença de tais comunicações não é obrigatória para o diagnóstico.

Com relação à valva AV, além da presença dos cinco folhetos, outras anormalidades podem ser encontradas: músculo papilar único, duplo orifício à esquerda, displasia e hipoplasia dos folhetos, fendas acessórias, anomalias de inserção das cordoalhas e valva AV esquerda em paraquedas.[10-14]

As manifestações clínicas e o prognóstico dependem das variações anatômicas citadas acima, do grau de insuficiência da valva AV e do grau de resistência vascular pulmonar.[8,15,16] Há também casos em que a relação do anel AV com os ventrículos é desproporcional, havendo dominância de um ou de outro ventrículo, as chamadas formas desbalanceadas, que podem se comportar como corações com fisiologia univentricular.[10] Assim, podemos encontrar desde pacientes adultos assintomáticos, até

crianças com menos de três meses em franca insuficiência cardíaca e, mais raramente, pacientes com algum grau de cianose.[8,12,16,17] De maneira geral, pacientes com a forma completa manifestam sintomas antes do que aqueles com as formas incompletas e normalmente são operados mais cedo.[8,18]

A finalidade do tratamento do DSAV é controlar a insuficiência cardíaca, manter o crescimento adequado da criança e prevenir o desenvolvimento de doença vasoclusiva pulmonar.[19] Isso implica quase sempre em fechar as comunicações intracardíacas e dividir a valva AV em dois componentes, um para cada ventrículo, preservando ou melhorando a competência da valva AV. Pacientes não operados e submetidos apenas a tratamento clínico apresentam menor sobrevida quando comparados à população geral, mesmo nas formas incompletas.[20]

Atualmente, com a possibilidade de diagnóstico fetal,[21] facilitando o manejo clínico desde o nascimento; com a indicação cirúrgica mais precoce,[15,17,19,22] evitando repetidas internações por insuficiência cardíaca descompensada e, também, a progressão para doença vascular pulmonar; e com as mudanças nos cuidados peri-operatórios,[23] esses pacientes têm uma maior sobrevida.

Embora os números atuais sejam cada vez mais aceitáveis no que se referem à mortalidade pós-operatória em 30 dias, abaixo dos 5% em alguns serviços, tanto para a forma total,[17,19,22,24,25] como para a parcial/transicional,[12,14,26,27] as taxas de reoperação por insuficiência da valva AV esquerda (IVAVE) continuam relativamente altas e sem grandes mudanças ao longo dos anos, variando de 4 a 22% para o DSAV total e de 7 a 22% para o DSAV parcial/transicional,[12-14,17,23,24,26,28-31] dependendo do serviço, da época de realização da correção cirúrgica e do tempo de acompanhamento.

Apesar de serem descritos casos de melhora da IVAVE após a alta hospitalar,[25] preocupa o fato de que aproximadamente entre 9% e 17% dos pacientes com DSAV total e entre 6% e 20% dos pacientes com DSAV parcial/transicional recebam alta hospitalar com graus significativos de IVAVE.[12,27,32,33] Além, é claro, dos casos que necessitam reoperação durante a mesma internação.[12,14,34]

A maioria dos trabalhos tem avaliado fatores predisponentes de reoperação tardia, sendo comumente observada forte relação entre reoperação e não fechamento da fenda da valva AV esquerda.[29,35,36] Outros fatores também são citados, mas com menos consistência: malformações na valva AV, ausência de síndrome de Down, baixo peso, grau de insuficiência da valva AV antes da operação e no pós-operatório imediato, idade inferior a 3 meses e ângulo mais agudo da valva AV.[11,12,17,26,30-32,35,37]

No entanto, poucos foram os trabalhos que estudaram fatores relacionados à IVAVE encontrada nos primeiros 30 dias após a correção,[18,27] apesar de se saber que a presença de lesão residual com repercussão hemodinâmica nesse período está associada com maior número de complicações, incluindo óbito e reoperação.[18,23,31,33,37] Portanto, entender melhor a estrutura e a fisiologia da valva AV, bem como determinar os fatores predisponentes de um pior resultado cirúrgico imediato são fundamentais.

Objetivo

O objetivo desse estudo foi identificar fatores associados à IVAVE moderada ou importante nos primeiros 30 dias após a correção cirúrgica de DSAV nos pacientes operados em nosso serviço.

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Factors Associated with Left Atrioventricular Valve Regurgitation within 30 days of Surgical Repair of Complete Atrioventricular Septal Defect

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Factors Associated with Left Atrioventricular Valve Regurgitation within 30 days of Surgical Repair of Complete Atrioventricular Septal Defect

Running head: Left Atrioventricular Valve Regurgitation

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ABSTRACT

Introduction: The aim of this study was to identify risk factors for moderate or severe left atrioventricular valve regurgitation within 30 days of surgical repair of complete atrioventricular septal defect at our center.

Methods: This was a retrospective study in which we evaluated the results of 53 consecutive patients that were operated on at our practice between 2002 and 2010. The following associated factors were considered: age, weight, Down syndrome, grade of preoperative atrioventricular valve regurgitation, abnormalities on the atrioventricular valve and the use of annuloplasty. The median patient age at the time of repair was 6.7 months; the median weight was 5.3 Kg; 86.8% had Down syndrome. At the time of preoperative evaluation, there were 26 cases with moderate or severe atrioventricular valve regurgitation (49.1%). Annuloplasty was done in 34%; abnormalities on the valve were found in 11.3% of the cases.

Results: At the time of post-operative evaluation, there were 21 cases with moderate or severe left atrioventricular valve regurgitation (39.6%). After performing a multivariate analysis, the only significant factor associated with these grades of insufficiency within 30 days of surgical correction of complete atrioventricular septal defect was the absence of Down syndrome (*p* equal to 0.03). Moderate or severe pre-operative insufficiency and the presence of atrioventricular valve abnormalities reached a marginal significance only under univariate analyses.

Conclusion: Absence of Down syndrome proved to be associated with moderate or severe post-operative left atrioventricular valve regurgitation in our patients.

INTRODUCTION

Although mortality rates are becoming more acceptable, between 9% and 17% of patients with complete atrioventricular septal defect are discharged from the hospital with significant residual left atrioventricular valve regurgitation. [1,2] Aside from the obvious risks of reoperation that it creates, the presence of left atrioventricular valve regurgitation with hemodynamic compromise in this period can increase time of hospital stay, morbidity and risk of death, and can also demand more long-term attention and care.[1-5]

To improve these statistics, a clear outline of the predisposing factors leading to a poorer immediate surgical outcome is essential. Factors associated with reoperation, like abnormalities on the atrioventricular valve, the non closure of the atrioventricular valve cleft, absence of Down syndrome, low weight, preoperative atrioventricular valve regurgitation, age less than 3 months and a more acute angle of the common atrioventricular valve have been cited.[1,4,6-9] However, few researchers studied factors associated with significant immediate post-operative left atrioventricular valve regurgitation.[5]

The purpose of this study was to identify factors associated with moderate or severe left atrioventricular valve regurgitation within 30 days of surgical repair at our center.

PATIENTS AND METHODS

This study was approved by the Ethics Committee of our institution, which waived the need for patient consent. The medical records of all patients who had undergone definitive repair of complete atrioventricular septal defect at our practice between March 2002 and April 2010 were retrospectively reviewed. Patients with complex cardiac anomalies such as tetralogy of

Fallot and double outlet right ventricle, as well as those who had a previous pulmonary banding were excluded.

As for the institution's characteristics, it is a general hospital in a large developing country in which there is a division of Pediatric Cardiology and Cardiovascular Surgery that started performing operations in 2002. Most patients are referred for surgical treatment from other parts of the country. After treatment, they return to their respective regions where they are clinically managed. However, most of these patients have no way to come back to receive follow-ups at our facilities, which hinders long-term follow-up.

The diagnosis was made with the use of transthoracic Echocardiography. Patients in whom there was a clinical suspicious of pulmonary vascular disease underwent heart catheterization. Echocardiograms were performed before and after operations by one of two physicians using commercially available machines, HDI 5000CV (ATL Ultrasound), Envisor-C and HD11 (Philips Ultrasound, Bothell, WA, USA), with 3 to 8 MHz probes. For further analysis, there were considered the last exam before surgery and the exam that was closest to the 30th post-operative day, while still being within 1 month of the repair.

The following factors were analyzed: age, gender, Down syndrome, grade of preoperative atrioventricular valve regurgitation, abnormalities on the atrioventricular valve, and the use of annuloplasty.

Atrioventricular valve regurgitation was subjectively divided into 4 grades based on the appearance of the color Doppler jets in relation to the surrounding chambers. 1: absent or trivial; 2: mild; 3: moderate; 4: severe. [10] The categorizations were based just on official written summaries of the exams. Images stored on tapes or in digital media were not accessed.

Abnormalities on the atrioventricular valve morphology were subjectively described by the surgeon. Transesophageal echocardiography was not available during the period in which the patients were operated on.

Statistical Analysis

Continuous variables were expressed as median, and comparisons were made using the two-sided Mann-Whitney test. Cathegorical variables were expressed using frequency distribution and percentages, and comparisons were made using the Fisher exact test.

Univariate odds ratios and their 95% confidence intervals (95% CI) were estimated for variables found to have a statistically significant (*p* lower than or equal to 0.05) relationship with post-operative left atrioventricular valve regurgitation grade moderate or severe. These variables were included in the multivariate analysis, which was completed using logistic regression. A *p*-value of 0.05 or less was considered significant.

All statistical analyses were conducted using the software StatsDirect, version 2.7.2. 2008 (Cheshire, UK).

Patient Population

We included 53 patients (37 females and 16 males), 46 with Down syndrome (86.8%). Age at the time of repair ranged from 2.7 months to 3 years (median 6.7 months); 37.7% of the patients were 6 months old or younger, and 83% were 1 year old or younger. Weight varied between 2.9 and 13 Kg (mean 5.3) (Table 1).

The majority presented minor associated heart defects, the most common of which were ostium secundum atrial septal defect (45.8%) and patent ductus arteriosus (30.2%) (Table 2).

Preoperative cardiac catheterization was used in 9 of the 53 patients (17%) in order to evaluate pulmonary vascular resistance. The age of these 9

patients ranged from 6.7 to 31.4 months (mean 13.3 months, which was almost double the mean age of the entire group). Their characteristics can be found in Table 3.

At the time of preoperative evaluation, there were 4 cases with grade 1 atrioventricular valve regurgitation (7.5%), 23 with grade 2 (43.4%), 16 with grade 3 (30.2%), and 10 with grade 4 (18.9%). Considering both the presence of Down syndrome and the grade of atrioventricular valve regurgitation, patients with Down syndrome had lower grades of pre-operative atrioventricular valve regurgitation than those without Down syndrome, but not statistically significant (*p* equal to 0.2).

Six cases (11.3%) of abnormalities on the atrioventricular valve morphology were found: small left atrioventricular valve orifice (3), hypoplastic left mural leaflet (1), accessory cleft (1), and grossly malformed valve (1). Among the 46 patients with Down syndrome, 5 (10.9%) had abnormalities on the atrioventricular valve, while 1 of the 7 patients without Down syndrome (14.3%) had an abnormality (*p* superior to 0.99).

Operative and post-operative management

Surgery using a median sternotomy was performed by the same surgeon on all patients, using continuous extracorporeal circulation by ascending aortic and bicaval cannulation with deep hypothermia (rectal temperature 22°C) in 2 patients, while moderate hypothermia (rectal temperature 25-28°C) was used on the other 51. The cardiopulmonary bypass time varied between 66 and 200 minutes (median 105), and the aortic cross-clamp time between 42 and 180 minutes (median 78.5). When present, a patent ductus arteriosus was ligated as cardiopulmonary bypass began. Antegrade cold crystalloid cardioplegia was used at 20-minute intervals for myocardial preservation.

The two-patch technique with preserved bovine pericardium was used in 50 patients (94.3%); the other 3 were repaired using the single-patch modified

technique, because the surgeon considered their ventricular septal defects to be too small.

The zone of apposition or cleft was completely closed in 49 patients (92.4%), was partially closed in 1 patient, and was left open in 3 patients (in two patients the annulus was too small, and in one patient, the posterior annuloplasty had reduced the diameter of the valvar orifice, which made it impossible to perform this maneuver).

Both cleft closure and posterior annuloplasty were used in 18 patients (34%) who presented annular dilation based on the surgeon's judgment. There was no difference between the pre-operative grade of atrioventricular valve regurgitation of these 18 patients comparing to the other 35 patients (*p* equal to 0.97).

All patients left the operating room intubated and with the use of inotropic and/or vasodilator drugs according to their clinical status and operative findings. These procedures were removed based on the judgment of the critical care physician. Information regarding the post-bypass grade of left atrioventricular valve regurgitation was not available for all patients, neither by transesophageal Echocardiography, nor by saline infusion into the left ventricle to test the valve competence.

The post-operative time on a mechanical ventilator ranged from 4 to 1699.3 hours (median 19.9) and the time of inotropic support varied between 24 and 1146 hours (median 81).

RESULTS

The post-operative length of hospital stay ranged from 1 to 149 days (mean 11.5). There were 4 deaths (7.1%), 1 within the first 24 hours due to cardiogenic shock caused by left ventricular dysfunction; another from a 3rd

degree atrioventricular block on the 12nd post-operative day and before pacemaker implantation; 1 due to sepsis on the 24th post-operative day and another on the 25th post-operative day from multi-system organ failure. The post-operative left atrioventricular valve regurgitation grade on these patients was 2, 1, 3 and 4 respectively (their pre-operative atrioventricular valve regurgitation grades were respectively 3, 3, 3 and 2).

The echocardiograms considered for analyses were performed between 1 and 29 days (mean 12.1 plus or minus 8.5) after repair. At the time of post-operative evaluation, there were 5 cases with grade 1 left atrioventricular valve regurgitation (9.4%), 27 with grade 2 (50.9%), 18 with grade 3 (34%), and 3 with grade 4 (5.7%). The difference between the pre- and post-operative grades of valve regurgitation was marginally significant (*p* equal to 0.06). There was a partial or complete improvement of atrioventricular valve regurgitation or maintenance of a trivial or mild atrioventricular valve regurgitation in 35 patients (66%). A one-grade worsening was found in 12 patients (22.6%).

Of the 4 cases in which the cleft was left partially or completely open, there was no significant difference between pre- and post-operative atrioventricular valve regurgitation (*p* superior to 0.99). However, among those in which the cleft was closed, this difference was significant (*p* equal to 0.05). Among those who had undergone annuloplasty, this difference was not significant (*p* equal to 0.4), and among those who had not undergone annuloplasty, this difference was only marginally significant (*p* equal to 0.08).

Other findings on post-operative echocardiograms included the following: 10 cases of right atrioventricular valve regurgitation (18.9%), 7 cases of small ventricular septal defects (13.2%), 3 cases of pericardial effusion (5.7%), 3 cases of pulmonary hypertension (5.7%), 2 cases of small atrial septal defect (3.8%) and 1 case of left atrioventricular valve stenosis (1.9%).

Table 4 e 5 shows the comparisons of data between patients with a post-operative left atrioventricular valve regurgitation grade trivial or mild and those with a left atrioventricular valve regurgitation grade moderate or severe. According to a univariate analysis, absence of Down syndrome was proved to be the only factor associated with moderate or severe left atrioventricular valve regurgitation after correction (*p* equal to 0.01). Presence of at least mild pre-operative atrioventricular valve regurgitation and presence of atrioventricular valve abnormality were only marginally significant. According to a multivariate analysis, only absence of Down syndrome continued to be statistically significant (*p* equal to 0.03).

DISCUSSION

In this study, the only risk factor that was considered significant after multivariate analysis for post-operative left atrioventricular valve regurgitation grade moderate or severe within 30 days of surgical repair was the absence of Down syndrome (*p* equal to 0.03).

Some papers have reported the absence of Down syndrome as a risk factor for reoperation.[8,11] Dodge-Khatami et al. stated that the atrioventricular valve of the patient with Down syndrome has more tissue, which may make the surgical repair easier.[1] If we take into consideration that there were no statistically significant differences in the pre-operative grade of atrioventricular valve regurgitation among Down and non-Down patients, it seems reasonable to believe in statements like that. However, we didn't find any published information correlating the presence of Down syndrome with the amount of tissue or histological aspects of the atrioventricular valve.

In the study by Kanani et al, in which the anatomy of the subvalvar apparatus of normal hearts was compared to those of hearts with atrioventricular septal defect, the structural and geometric disarray of the

tendinous cords of those hearts with atrioventricular septal defect was clearly visible, along with its possible rule on the mechanisms of valve regurgitation.[12] However, there was no mention if patients with Down syndrome were included in the study. Studies addressed to explain this very common finding, like histopathological comparisons of the valve and subvalve apparatus of patients with and without Down syndrome, must be done or, maybe, the surgical approach to patients without Down syndrome should be revisited.

Mild or larger atrioventricular valve regurgitation as a risk factor for post-operative left atrioventricular valve regurgitation moderate or severe was marginally significant only under univariate analyses (p equal to 0.14). Significant preoperative left atrioventricular valve regurgitation eventually leads to left ventricle dilation, changing the cordal axis and influencing the mechanism of the valve closure, what may explain these results.[12] However, in a more recent work by Bharucha et al, the grade of preoperative atrioventricular valve regurgitation didn't influence the results.[9] With the use of three-dimensional echocardiography, they found that a more acute angle of the components of the common atrioventricular valve against the plane of the common atrioventricular junction would be a predictor of postoperative valve function. So, it is clear that with the use of new diagnostic tools, a better understanding of both the atrioventricular valve structure and function will add new perspectives on the management of this disease.

The results of this study could not prove any relationship between the presence of malformation and moderate or severe post-operative left atrioventricular valve regurgitation. Some studies found the atrioventricular valve malformation to be associated with reoperation or valve replacement.[6,8] However, some cases of atrioventricular valve malformation have been subjectively diagnosed.[6] Ando et al. found a weak correlation

between preoperative echocardiographic findings and the surgeon's judgment in regard to the diagnosis of these malformations.[13] Furthermore, there is little consistency between the findings of two- and three-dimensional Echocardiography in respect to the analyses of atrioventricular valve abnormalities. In the study by Takahashi et al, for instance, the correlation between the findings of both methods was lower than 46% in the evaluation of the mural leaflet and in the evaluation of the commissural abnormalities of the left atrioventricular valve leaflets.[14] The three-dimensional echocardiogram was more accurate and more reliable.

With respect to the approach of the zone of apposition, some studies have reported that not closing it would be a risk factor for reoperation, and it has therefore been adopted at our institution.[15,16] However, in 4 cases in this study (7.5%) the cleft was not completely closed due to the risk of stenosis. In these cases, there was no alteration in the functional status of the valve, differently of those who had this procedure performed, who clearly had some benefit from this approach.

The rate of annuloplasty in this study (34%) was similar to that reported by Suzuki et al. (30%), and much higher than those reported by Dragulescu et al. (4.7%) and by Stellin et al. (2.5%).[6,17,2] These different rates show that there is no well established pattern on this item. Padala et al. showed, in vitro, the importance of the rule of the annular dilation for the atrioventricular valve regurgitation grade: they concluded that performing only the cleft closure is not enough to avoid the atrioventricular valve regurgitation, and that would have a real benefit in shortening the annulus size.[18] Such benefit was not found in the present study; however we cannot confirm this lack of influence, because individual variances were not analyzed. For example, it would be necessary to study the grade of left atrioventricular valve regurgitation on a specific patient had the annuloplasty not been done.

Comments

Some might say that that extremely high number of patients with Down syndrome in this study (86.8%) could suggest some biased referral pattern. In the literature this rate varies between 65% and 75%.[20,21] Only Backer et al. found such a high number (92.4%).[22] Actually, it can be explained by some reasons: termination of pregnancy is not allowed in our country as it is in some developed countries;[23] and there is a center of Fetal Medicine at our facilities which refers all patients with genetic syndromes for our diagnostic department.

Study Limitations

It was a retrospective study, and was therefore subject to limitations in terms of how correctly the information in the medical records was well filled.

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Table 1. Characteristics of the 53 patients enrolled in the study.

Characteristic	N (%)
Age at the time of repair in months (median)	6.7
Female	37 (69.8%)
Weight in Kg (median)	5.3
Down syndrome	46 (86.8%)
Grade 1 AVVR	4 (7.5%)
Grade 2 AVVR	23 (43.4%)
Grade 3 AVVR	16 (30.2%)
Grade 4 AVVR	10 (18.9%)
AV valve abnormalities	6 (11.3%)

AVVR: atrioventricular valve regurgitation; AV: atrioventricular.

Table 2. Associated minor heart malformations.

Defect	N (%)
ASD	24 (45.3%)
PDA	16 (30.2%)
Left SCV	2 (3.8%)
RPA stenosis	1 (1.9%)
VSD	1 (1.9%)

ASD: atrial septal defect; PDA: persistent ductus arteriosus; SCV: superior caval vein; RPA: right pulmonary artery; VSD: ventricular septal defect.

Table 3. Clinical data of the 9 patients who had undergone right heart catheterization.

Patient	Age (mo)	DS	AVVR grade	MPAP	PVR/SVR	PO LAVVR grade
1	24.2	Y	4	30	NA	3
2	7.4	N	4	32	0.1	3
3	31.4	Y	4	33	0.37	3
4	6.7	Y	4	33	0.78	2
5	9.2	Y	4	40	0.3	2
6	13.3	Y	3	40	0.33	2
7	11.7	Y	2	44	0.38	3
8	16.5	Y	2	45	0.22	2
9	15.6	Y	4	39	NA	2

mo: months; DS: Down syndrome; AVVR: atrioventricular valve regurgitation; MPAP: mean pulmonary artery pressure; PVR: pulmonary vascular resistance; SVR: systemic vascular resistance; PO LAVVR: post-operative left atrioventricular valve regurgitation; Y: yes; N: no; NA: non available.

Table 4. Univariate analysis of preoperative and intra-operative factors related to postoperative left atrioventricular valve regurgitation grade moderate or severe.

FACTOR	Trivial or Mild LAVVR	Moderate or Severe LAVVR	UNIVARIATE		
	(n = 32)	(n = 21)	OR	CI (95%)	*p*
Age in months (median)	7.7	6.1			0.63
Weight in Kg (median)	5.4	5			0.93
Down syndrome	31 (96.9%)	15 (71.4%)	0.08	0.002-0.791	0.01
Preoperative AVVR ≥ 2	28 (87.5%)	21 (100%)			0.14
Annuloplasty	11 (34.4%)	7 (33.3%)			>0.99
AV valve abnormality	2 (6.2%)	4 (19%)			0.20

LAVVR: left atrioventricular valve regurgitation; AVVR: atrioventricular valve regurgitation; AV: atrioventricular; n: number; =: equal to; >: superior to; ≥: superior or equal to; OR = odds ratio; CI = confidence interval.

Table 5. Multivariate analysis of preoperative and intra-operative factors related to postoperative left atrioventricular valve regurgitation grade moderate or severe.

FACTOR	Trivial or Mild LAVVR	Moderate or Severe LAVVR	UNIVARIATE		
	n =32	n = 21	OR	CI 95%	*p*
Down syndrome	31 (96.9%)	15 (71.4%)	0.08	0.009-0.774	0.03
Pre-operative AVVR ≥ 2	28 (87.5%)	21 (100%)			0.99
AV valve abnormality	2 (6.2%)	4 (19%)			0.21

LAVVR: left atrioventricular valve regurgitation; AVVR: atrioventricular valve regurgitation; AV: atrioventricular; n: number; ≥: superior or equal to; =: equal to; OR = odds ratio; CI: confidence interval.

Factors Associated with Moderate or Severe Left Atrioventricular Valve Regurgitation Within 30 days of Repair of Incomplete Atrioventricular Septal Defects

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Keywords:	congenital heart disease, valve, septal defect

Factors Associated with Moderate or Severe Left Atrioventricular Valve Regurgitation Within 30 days of Repair of Incomplete Atrioventricular Septal Defects

Running head: Left Atrioventricular Valve Regurgitation

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ABSTRACT

Objective. This study was conducted to determine factors associated with moderate or greater left atrioventricular valve regurgitation (LAVVR) within 30 days of surgical repair of incomplete atrioventricular septal defect (AVSD).

Methods. We evaluated the results of 51 consecutive patients 14 years old and younger presenting with incomplete AVSD that were operated on at our practice between 2002 and 2010. The following associated factors were considered: age, weight, absence of Down syndrome, grade of preoperative LAVVR, abnormalities on the left atrioventricular valve and the use of annuloplasty. The median patient age at the time of repair was 4.1 years; the median weight was 13.4 Kg; 37.2% had Down syndrome. At the time of preoperative evaluation, there were 23 cases with moderate or grater LAVVR (45.1%). Abnormalities on the AV valve were found in 17.6% of the cases; annuloplasty was performed in 21.6% of the patients.

Results. At the time of postoperative evaluation, there were 12 cases with moderate or greater LAVVR (23.5%). During univariate analysis, only absence of Down syndrome was statistically significant (p = 0.02). However, after a multivariate analysis, none of the factors reached significance.

Conclusion. None of the factors studied was determinant of a moderate or greater LAVVR within the first 30 days of repair of incomplete AVSD at our center.

Keywords: congenital heart disease; valve; septal defects

INTRODUCTION

The classification of atrioventricular septal defect (AVSD) depends on the number of valve orifices and on the location and size of the septal defect in question [18]. Partial AVSD presents two distinct atrioventricular (AV) valve orifices, an often large ostium primum atrial septal defect (ASD), and a cleft on the left AV valve. Transitional AVSD also presents two distinct AV valve orifices, an ostium primum ASD, the cleft, and usually a small ventricular septal defect (VSD), immediately beneath the AV valve plane [8]. Both are considered incomplete forms of AVSD and since the anatomical differences and hemodynamic compromise are quite unremarkable, data from children with either defect can be combined for analysis.

Considering the natural history [17], and the fact that mortality rates are lower than 5% in many centers [1,3,10,14,15,19,23,24], surgical treatment must be indicated. However, reoperation rates for left atrioventricular valve regurgitation (LAVVR) are still relatively high, and have shown no alterations over time, varying between 7% and 22% [1,3, 9,14, 24,32].

Many factors have been proposed as being involved in reoperation, including post-operative LAVVR [3,23,24,32]. Some studies have shown that between 6% and 20% of patients with incomplete AVSD are discharged from hospital with significant residual LAVVR [3,23], but there is a lack of data reporting factors associated with immediate post-operative LAVVR, although there are reports of valve replacement within 30 days of operation [3,9,28] and in-hospital deaths [9] due to significant immediate post-operative LAVVR.

To improve these statistics, a clear outline of the predisposing factors leading to a poorer immediate surgical outcome is essential. The goal of this study was to test some of the previous risk factors published in the literature on an at least moderate LAVVR within 30 days of surgical repair at our center.

PATIENTS AND METHODS

This study was approved by the Ethics Committee of our institution, which waived the need for patient consent. As for the institution's characteristics, it is a general hospital in a large developing

country in which there is a division of Pediatric Cardiology and Cardiovascular Surgery that started performing operations in 2002. Most patients are referred for surgical treatment from other parts of the country. After treatment, they return to their respective regions where they are clinically managed. However, most of these patients do not to come back to receive follow-ups at our facilities, which hinders long-term follow-up.

The medical records of all patients 14 years old and younger who had undergone definitive repair of partial and transitional AVSD at our practice between March 2002 and April 2010 were retrospectively reviewed. Patients with any right ventricle obstruction, and those who had a previous pulmonary banding were excluded.

The diagnosis was made with the use of transthoracic Echocardiography, according to the current nomenclature [18]. Echocardiograms were performed before and after operations by one of two physicians using commercially available machines, HDI 5000CV (ATL Ultrasound), Envisor-C and HD11 (Philips Ultrasound, Bothell, WA, USA), with 3 to 8 MHz probes. For further analysis, there were considered the last exam before surgery and the exam that was closest to the 30th post-operative day, while still being within 1 month of the repair.

The following pre-operative factors were analyzed: age, weight, absence of Down syndrome and LAVVR grade. The following intraoperative factors were analyzed: abnormalities on the AV valve morphology and the use of annuloplasty.

LAVVR was subjectively divided into 4 grades based on the appearance of the color Doppler jets in relation to the left atrium [34]: I = absent or trivial; II = mild; III = moderate; IV = severe. The categorizations were based just on official written summaries of the exams. Images stored on tapes or in digital media were not accessed.

Abnormalities on the AV valve morphology were subjectively described by the surgeon. Transesophageal echocardiography was not available during the period in which the patients were operated on.

Statistical Analysis

Continuous variables were expressed as mean or median, and comparisons were made using the two-sided Mann-Whitney test. Categorical variables were expressed using frequency distribution and percentages, and comparisons were made using the Fisher exact test. For the analyses of variances, the Kruskal-Wallis test was used. Univariate odds ratios and their 95% confidence intervals (95% CI) were estimated for variables found to have a statistically significant ($p \leq 0.05$) or borderline significant ($p \leq 0.2$) relationship with moderate or greater post-operative LAVVR. These variables were included in the multivariate analysis, which was completed using logistic regression. A p-value of 0.05 or less was considered significant. All statistical analyses were conducted using the software StatsDirect, version 2.7.2. 2008 (Cheshire, UK).

Patient Population

We included 51 patients (27 females and 24 males), 35 with partial AVSD and 16 with transitional AVSD, 32 without Down syndrome (62.7%). Age at the time of repair ranged from 4.2 months to 12.8 years (median = 4.1 years). Weight varied between 3.8 and 44.3 Kg (median = 13.4 Kg). At the time of preoperative evaluation, there were 4 cases with grade I LAVVR (7.9%), 24 with grade II (47%), 15 with grade III (29.4%), and 8 with grade IV (15.7%) (Table 1). Less than 50% of the patients presented minor associated heart defects (Table 2).

Considering both the presence of Down syndrome and the grade of atrioventricular valve regurgitation, patients with Down syndrome had lower grades of pre-operative atrioventricular valve regurgitation than those without Down syndrome, but not statistically significant ($p = 0.17$).

Nine cases (17.6%) of abnormalities on the AV valve morphology were found and they were described according to the surgeon's report: dysplastic valve leaflets (5); accessory cleft (2), tendinous cordae rupture (1), and small left AV valve orifice (1). Among the 19 patients with Down syndrome, 2 (10.5%) presented one of these abnormalities; while among the 32 without Down syndrome, 7 (21.9%) presented it ($p = 0.7$).

Operative and postoperative management

Surgery using a median sternotomy was performed by the same surgeon on all patients, using continuous extracorporeal circulation by ascending aortic and bicaval cannulation. When present, a patent ductus arteriosus was ligated as cardiopulmonary bypass began. Deep hypothermia (rectal temperature 22°C) and total circulatory arrest was used in 1 patient who presented coarctation of the aorta. Moderate hypothermia (rectal temperature 25-30°C) was used in 29 patients, while mild hypothermia (rectal temperature 30-34°C) was used on the other 21. The cardiopulmonary bypass time varied between 34 and 114 minutes (median 72), and the aortic cross-clamp time between 19 and 89 minutes (median 48). Antegrade cold crystalloid cardioplegia was used at 20-minute intervals for myocardial preservation.

The ASD was closed with a preserved bovine pericardium patch in 50 of the 51 cases, and with direct suture in 1 case. The surgeon didn't find any evidence of VSD in 8 of the 16 patients with transitional AVSD. Of these same 16 patients, the VSD was closed with direct suture in 3, it was closed with a preserved bovine pericardium patch in 2, it was closed with the same patch used for the closure of the ASD in 2, and in the 1 patient remaining, it was closed using implantation of the superior bridging left on the superior rim of the defect.

As for the closure of the cleft, our routine has been to close it whenever possible, even in cases with absent or minimal LAVVR. Thus, the cleft was completely closed with interrupted 6-0 polypropilene sutures in 49 patients (96.1%); in the other 2 patients, the reason why it was left open was not found in the reports. For those 11 patients (21.6%) who presented annular dilation based on the surgeon's judgment, both cleft closure and posterior annuloplasty were used. There were no statistically significant differences in the pre-operative grade of LAVVR of these 11 patients when comparing to those who had not undergone annuloplasty ($p = 0.94$).

All patients left the operating room intubated and with the use of inotropic and/or vasodilator drugs according to their clinical status and operative findings. These procedures were removed based on the judgment of the critical care physician.

RESULTS

The postoperative time on a mechanical ventilator ranged from 2.6 to 44.7 hours (median 7.2) and the time of inotropic support varied between 10 and 157 hours (median 46). The postoperative length of hospital stay ranged from 1 to 22 days (mean 7). There were two deaths (3.9%), one within the first 24 hours due to a 3rd degree AV block (temporary pacing wires didn't work well) and another from shock on the 2nd postoperative day. The post-operative LAVVR grades on these patients were II and I, respectively. Before treatment, both of these patients had a grade I LAVVR.

Postoperative echocardiographic analyses occurred between zero and 30 days (mean 12.6 ± 9.4). At the time of postoperative evaluation, there were 8 cases with grade I LAVVR (15.7%), 31 with grade II (60.8%), 11 with grade III (21.5%), and 1 with grade IV (2%) (Figure 1). The variance between pre- and post-operative grades of valve regurgitation was significant ($p = 0.018$).

In 17 patients, the trivial or mild LAVVR found before operation was maintained after the procedure. In 15 of the 23 patients with moderate or severe pre-operative LAVVR (65.2%), the post-operative LAVVR was found to be trivial or mild. A one-grade worsening was found in 8 patients (15.7%). Of the 2 cases in which the cleft was left open, the grade of LAVVR didn't change. Among those who had undergone annuloplasty, the amount of improvement was lower than among those who had not undergone this procedure, but without statistic significance ($p = 0.84$).

Other findings on postoperative echocardiograms were seen in 26 patients and included the following: 12 cases of insignificant residual VSD, which is defined as a VSD < 3 mm (23.5%; 8 of them were not seen during surgery, confirming that they were too small), 8 cases of pericardial effusion (15.7%), 4 cases of insignificant residual ASD, which is defined as an ASD < 3 mm (7.8%), 4 cases of right ventricular dysfunction (7.8%), 2 cases of mild left AV valve stenosis (3.9%), 1 case of left ventricular dysfunction (2%), 1 case of pulmonary hypertension (2%), and 1 case of a fistula from the left ventricle to the right atrium (2%).

As shown in table 3, univariate analysis revealed that absence of Down syndrome was determinant for post-operative LAVVR grade ≥ III ($p = 0.02$). Severe pre-operative LAVVR and AV

valve abnormalities were only marginally significant (p = 0.07 and 0.19, respectively). However, as shown in table 4, none of the factors were found to be determinant for post-operative LAVVR grade ≥ III. Absence of Down syndrome reached only a borderline significance (p = 0.06).

DISCUSSION

It is important to note that, despite different ages, different weight at repair, and different physiology [8], patients with both complete and incomplete AVSD present a similar risk of reoperation for LAVVR [1-4,9,11-14, 20,22,24-27,29,32,33]. What both variations of the same disease have in common are the typical anatomical landmarks of AVSD (a common 5-leaflet AV valve, distinct papillary muscle displacement and a narrow and elongated left ventricle outflow tract) [16,21], as well as a high prevalence of individuals with Down syndrome [31]. Therefore, the clue to understanding this frequent complication may be related more to these two aspects than to another factor such as age, weight, or AV valve malformation.

In the study by Kanani et al [19], in which the anatomy of the subvalvar apparatus of normal hearts was compared to those of hearts with complete and incomplete AVSD with an intact left AV valve, the structural and geometric disarray of the tendinous cords of those hearts with AVSD was clearly visible, along with its possible rule on the mechanisms of valve regurgitation. Moreover, in the work by Bharucha et al [6], with the use of three-dimensional echocardiography, it was found that a more acute angle of the components of the common atrioventricular valve against the plane of the common atrioventricular junction would be a predictor of postoperative valve function. Weight at repair, Down syndrome and pre-operative grade of LAVVR did not influence the results. This factor was not tested in our study, nor in other studies in the literature.

In our study, which was performed in a developing country without the use of intra-operative transesophageal echocardiography, a decrease of the number of patients with moderate or grater LAVVR from 45.1% to 23.5% was very similar to that recently reported by Minich et al [23] in a study evaluating data from seven centers in North America. These results show that all techniques that

were used, including cleft closure and annuloplasty, had an impact, but not enough of an impact to completely avoid early post-operative LAVVR.

As for the Down syndrome factor, AVSD is a real model to study the impact of Down syndrome on outcomes, since there are no other situations in congenital heart diseases in which there is such a high prevalence of a chromosomal disorder. Statements such as “In Down patients valve tissue is more abundant and allows for an easier reconstruction” [12] are often seen. It suggests that patients without Down syndrome would have a higher risk of worse surgical outcomes, as found by Welke et al [32] and by Lange et al [20]. However, not all studies confirm this relationship. Murashita et al [24] and Al-Hay et al [1], for instance, did not find any significant differences in the results. In the aforementioned study by Kanani et al [19], there was no mention if hearts of patients with Down syndrome were included. Perhaps, histopathological comparisons of the valve and subvalve apparatus of patients with and without Down syndrome might answer this question.

Considering the presence of AV valve malformation, there is little consistency between the findings of two- and three-dimensional Echocardiography. In the study by Takahashi et al [30], for instance, the correlation between the findings of both methods was lower than 46% in the evaluation of the mural leaflet and in the evaluation of the commissural abnormalities of the left AV valve leaflets. The three-dimensional echocardiogram was more accurate and more reliable. Therefore, it is clear that the way in which AV valve malformations or abnormalities are diagnosed may influence the results.

In conclusion, we were not able to identify any risk factor for post-operative LAVVR at our center. It may be due to the limited number of patients enrolled in the study, or more probably, to the fact that we did not research the correct factor, if it actually exists. We think that in the era of three-dimensional echocardiography, new points of view will appear and will help to better understand some aspects of this disease that still need to be explained [5].

Study Limitations

It was a retrospective study, and was therefore subject to limitations in terms of how correctly the information in the medical records was filed. The small number of patients may, in some ways, account for the lack of statistical significance among the factors studied.

Table 1. Pre- and intra-operative characteristics of the patients enrolled in the study.

Characteristic	n (%)
Age at the time of repair in years (median)	4.1
Female	27 (52.9%)
Weight in Kg (median)	13.4
Down syndrome	19 (37.2%)
Grade I LAVVR	4 (7.9%)
Grade II LAVVR	24 (47%)
Grade III LAVVR	15 (29.4%)
Grade IV LAVVR	8 (15.7%)
Atrioventricular valve abnormalities	9 (17.6%)
Annuloplasty	11 (21.6%)

LAVVR = left atrioventricular valve regurgitation

Table 2. Associated Minor Heart Defects* Noted before Surgery.

Defect	n	%
Ostium secundum atrial septal defect	15	29.4
Common atrium	3	5.8
Additional ventricular septal defect	2	3.9
Patent ductus arteriosus	1	2
Left superior caval vein	1	2
Left atrium isomerism	1	2
Coarctation of the aorta	1	2
Subaortic stenosis	1	2

*Defects not mutually exclusive.

Table 3. Univariate Relations between Variables and Grade ≥ III Postoperative Left Atrioventricular Valve Regurgitation (LAVVR).

	LAVVR ≤ II	LAVVR ≥ III	Univariate		
	n = 39	n = 12	OR	95% CI	*P*
Age in months (median)	53.3	45.3			0.97
Weight in Kg (median)	13.1	15.4			0.87
Abscence of Down syndrome	21 (53.8%)	11 (91.7%)	9.43	1.12-427.9	0.02
Grade IV Preoperative LAVVR	4 (10.2%)	4 (33.3%)	4.4	0.64-28.31	0.07
AV valve abnormality	5 (12.8%)	4 (33.3%)	3.4	0.53-19.7	0.19
Annuloplasty	8 (20.5%)	3 (25%)			0.7

AV = atrioventricular; CI = confidence interval; OR = odds ratio.

Table 4. Multivariate Relations between Variables and Grade ≥ III Postoperative Left Atrioventricular Valve Regurgitation (LAVVR).

	LAVVR ≤ II	LAVVR ≥ III	Multivariate		
	n = 39	n = 12	OR	95% CI	*p*
Abscence of Down syndrome	21 (53.8%)	11 (91.7%)	8.4	0.9-79.5	0.06
Grade IV Preoperative LAVVR	4 (10.2%)	4 (33.3%)			0.25
AV valve abnormality	5 (12.8%)	4 (33.3%)			0.32

AV = atrioventricular; CI = confidence interval; OR = odds ratio.

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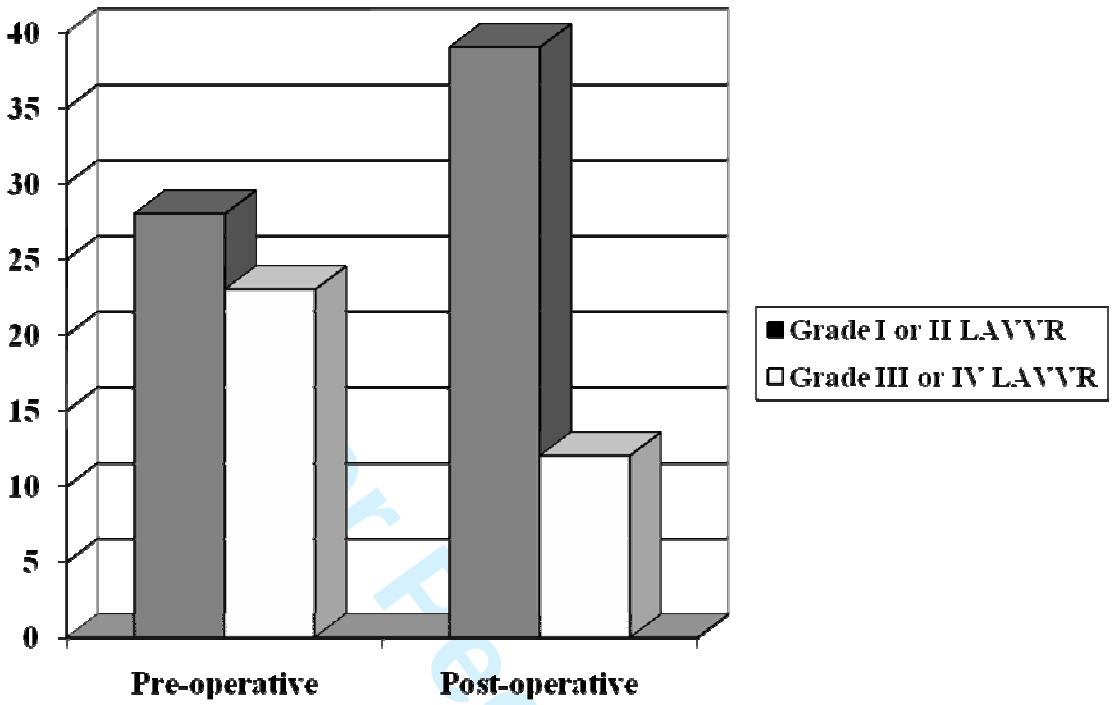

CONCLUSÃO

Entre os pacientes com defeito de septo atrioventricular total, o único fator de risco determinante de insuficiência moderada ou importante da valva atrioventricular esquerda nos primeiros 30 dias após correção cirúrgica foi a ausência de síndrome de Down. Entre os pacientes com defeito de septo atrioventricular parcial e transicional, não foi possível identificar os fatores de risco determinantes desse mesmo grau de insuficiência valvar.

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