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Results and Evolution of Surgical Repair of Auriculoventricular Defect in Patients of Late Presentation

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Abstract

The atrioventricular septal defects progressively develop an irreversible lung damage, leaving these patients out of surgical treatment. These patients should be treated between 3 and 6 months of age before the pulmonary vascular lesion begins, but in our institution we see the task of operating them later, which is the reason why we carried out the present study.

Methods: It is a retrospective, cross-sectional and descriptive study, where we studied a total of 67 patients who underwent correction surgery of atrioventricular septal defect older than 1 year in our institution between January 2000 and December 2015. Of these, 64 patients survived hospital discharge, of whom 55 had available follow-up data at a median postoperative time point of 4.7 (1-14) years.

Results: The pulmonary systolic pressure in the preoperative period was classified as severe pulmonary hypertension in 48 (71.6%) patients, giving a turn in the postoperative period where the highest percentage 74% (50 patients) presented normal pulmonary pressures, observing the same pattern during follow-up 70% (38 patients). The mortality in the immediate postoperative period was 4.4%. As a risk factor for mortality we observed high pulmonary vascular resistance with a P = 0.009. The percentage of reoperation is 7.2% and these do not have any risk factor that conditions this complication.

Conclusion: Treating patients older than one year with an atrioventricular septal defect is a safe procedure with a low rate of mortality and morbidity, with a significant decrease in systolic pressure of the pulmonary artery both in the immediate postoperative period and in long-term follow-up.

Keywords: Atrioventricular septal defect; pulmonary hypertension

Introduction

Atrioventricular septal defects are cardiac malformations characterized by impaired development of the embryonic endocardial bearings and the primitive atrioventricular canal. It has an incidence of 4-5.3 per 10,000 live births, encompasses 7% of all congenital heart defects, and is often associated with Down syndrome [1].

The clinical and pathophysiological consequences of atrioventricular septal defects will depend on the magnitude of the short circuit and the anatomical site where it is located and, as time passes, the pulmonary vascular resistance will be a regulator of the short circuit at the ventricular level [2].

In general, heart disease with a left-to-right shunt is symptomatic when 50% or more of the flow that reaches the left side of the heart is carried to the right side, that is, the pulmonary flow is 2 or more times the systemic flow (Qp / Qs greater than or equal to 2). Pulmonary hyperflow when associated with pulmonary systolic hypertension leads to a thickening of the tunica media and the arteriolar intima causing increased pulmonary vascular resistance, affecting pulmonary diastolic pressure. This intimal process can progress to hyalinization and fibrosis, and cause arteriolar thrombosis, becoming irreversible and progressive damage called obstructive pulmonary vascular disease, and pulmonary vascular resistance can reach such high values that it minimizes the short circuit from left to right and then reverses it from the right to the left causing cyanosis (Einsenmeyer syndrome) [2].

Due to the aforementioned, the appropriate age to intervene surgically is younger and younger, due to the need to avoid damage to the pulmonary vasculature suffered by these patients with late diagnoses or poorly performed follow-ups. Given that these children arrive at our center without prior medical attention, we are tasked with evaluating them at an older age in order to offer them surgical resolution. For this reason, it is important to describe the evolution of these patients who underwent correction of the late-presenting atrioventricular septation defect.

Materials and Methods

It is a retrospective, cross-sectional and descriptive study in which a total of 110 cases of surgical correction of the AV canal were observed in our institution between January 2000 and December 2015. 67 were surgically corrected who were older than 1 year, average age 3.23 (1-17 years). All patients who had other associated congenital heart defects, who have been lost to follow-up and do not have complete records, were excluded Figure 1.

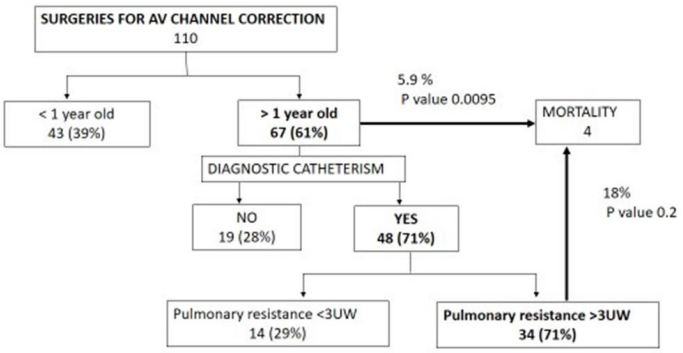

Figure 1: Diagnostic catheterization

All patients were diagnosed by echocardiography. According to Rastelli's classification, we have 60 type A (89.5%), 5 with type B (7.4%) and 2 with type C (2.9%). Table 1. Pulmonary systolic pressure measurement was also performed, classifying them as normotensive pressures less than 40 mm Hg, mild hypertession of 40 - 54 mmHg, moderate 55 to 64 mmHg, and severe greater than 65 mmHg.

Diagnostic catheterization was performed in all patients who were classified as severe pulmonary hypertensive to measure pulmonary pressure and pulmonary vascular resistance. A total of 14 (29%) patients presented low vascular resistance (<6 UW / m2) (9) and the rest underwent a pharmacological challenge, observing a decrease in resistance, supporting the operability of the patient. Figure 1.

Surgical Technique: The repair of complete atrioventricular canal defects has been described using single patch, two patch and modified single patch techniques. The technique used in our center depends on the experience and criteria of each surgeon. The cardiopulmonary bypass method is similar for the different techniques. Continuous cardiopulmonary bypass with moderate hypothermia (28-32° C) and antegrade cardioplegia have been used routinely for myocardial protection.

The single patch technique uses a bovine pericardium patch treated in our center, it begins by floating the AV valve leaflets in the closed position. The line of apposition of the anterior and posterior common leaflets is noted and an imaginary line is created that delimits the components of the right and left valves, marking the coaptation of the anterior and posterior common leaflets with a horizontal mattress suture. The ventricular septum ridge is sutured to the bottom of the patch with multiple interrupted horizontal mattress sutures. The components to the left of the common AV valve tissue are secured to the patch in

the appropriate plane with multiple interrupted mattress sutures. Whenever possible, these same sutures can be used to anchor the components of the right AV valve to the patch. The atrial portion of the defect is closed by suturing the upper edge of the patch to the lower edge of the atrial septal defect.

The repair of two patches in the same way is used a bovine pericardium patch treated in our center to close the ventricular septum defect. The valve leaflets are sutured to the top of the patch of the ventricular septal defect. The area of apposition of the left AV valve is closed with a suture and a separate pericardial patch is used to close the atrial septal defect.

With the modified single-patch technique, the ventricular septum defect is closed by placing a series of 5-0 polypropylene sutures supported on the right side of the interventricular septum ridge. These sutures are then sequentially passed through the anterior and posterior common leaflets at a predetermined site to separate the right and left AV valves. The sutures are then placed through the edge of a bovine pericardial patch treated at the institute. These sutures are then tied. The atrial component of the AV canal defect is closed with the pericardial patch.

Once the procedure was completed, a transesophageal echocardiographic evaluation was performed for all AV canal cases.

Statistic analysis

Continuous variables are reported as mean, or median with range, and categorical variables are presented as frequencies and percentage of the total.

Groups were compared using Fisher's exact test for categorical variables.

Results

Table 1: Pre-Surgical Characteristics

N = 67	
Age	3.23 (1-17) años
Sex (female)	40 (59%)
Down syndrome	41 (61%)
Rastelli classification Tipe A Tipe B Tipe C	 60 (89%) 5 (7.4%) 2 (2.9%)
NYH functional class II III	 85% 15%
LVEF %	66 (33-77)
Pulmonary artery banding	7 (10.4%)

41 (61%) patients present as Down Syndrome comorbidity. The previous preoperative functional class in which these patients were found were class II - III, 85-15% respectively. Table 1.

The mean pulmonary systolic pressure measured by catheterization or by echocardiography was 68.9 (28 - 105) mmHg. 3 (4.4%) patients presented pulmonary pressures within normal ranges, 2 (2.9%) presented mild pulmonary hypertension, 8 (11.9%) presented moderate pulmonary hypertension and 48 (71.6%) presented severe pulmonary hypertension. Those with severe hypertension, 14 was equaled systemic pulmonary pressure. Table 4. 48 (71%) patients underwent preoperative catheterization. 14 (29%) patients presented low pulmonary resistance and the rest were observed to be elevated but mobile. Figure 1. Statistical analysis was performed where it was observed that high lung resistances do not have significant statistical value comparing the variables of Down syndrome, central valve insufficiency, reoperations and mortality. Table 5.

Table 2: Surgical Characteristics

N = 67	
Surgical technique Unipatch Modified unipatch 2 patches	 57 (85 %) 7 (10.4%) 3 (4.4%)
Aortic clamping time	81 (30-133)
Extracorporeal circulation time	121 (58 -296)
Morbidity < 30 days	9 (13.4%)
Mortality	3 (4.4%)

Table 3: Follow-up

N = 55	
Follow-up (years)	4.7 (1 – 14)
Morbidity > 30 days	5 (7.4%)
AV valve reoperations	4 (7.2%)
Pacemaker implants	9 (16.3%)
Current treatment None Sildenafil Diuretic	 14 (25.4%) 3 (5.4%) 41 (74.5%)
Antihypertensive Digoxin	21 (38%) 3 (5.4%)

Table 4: Measurement of Pulmonary Artery Systolic Pressure (mmHg)

N° 67	Preop	Postop	Follow-up N° 54	P value
PASP	68.9 (28-105)	31.9 (16 – 75)	34 (13-76)	
- Normal	3 (4.4%)	50 (74%)	38 (70%)	0.0001
- Mild	2 (2.9%)	9 (13.4%)	9 (16.6%)	0.054
- Moderate	8 (11.9%)	5 (7.4%)	4 (7.4%)	0.29
- Severe	48 (71.6%)	4 (5.9%)	3 (5.5%)	0.0001

Table 5: Patients with Low and High but Mobile Pulmonary Resistances

	Pulmonary resistance < 6 UW/m2 14 (29%)	Pulmonary resistance > 6 UW/m2 34 (71%)	P Value
Down syndrome	6 (42%)	15 (44%)	6 (42%)
Central Valve regurgitation	6 (42%)	14(41%)	6 (42%)
Reinterventions	1 (7%)	-	1 (7%)
Mortality	-	4 (11%)	-

7 (10.4%) patients previously underwent a pulmonary artery banding and one had pulmonary stenosis as an associated pathology. Table 1.

3 techniques were used for surgical correction, the unipatch technique in 57 patients (85%), the unipatch modified technique in 7 (10.4%) and two patches in 3 (4.4%). The patch was fenestrated in 3. In 44 the plasty was performed at the level of the left AV valve, in 3 the rings were placed and in 20 no procedure was performed on the valve. Plasty was performed on the right AV valve in 10. The average aortic clamp time was 81 minutes (30-133), the extracorporeal circulation time 121 minutes (58 - 296). Table 2.

Intracavitary catheters were installed in 33 patients. The measurement of systolic pressure at admission to post-surgical therapy was 31 mmHg (11-75). The blood pressure measured during the hospital stay by average echocardiography was 31.9 (16 - 75) mmHg. Pulmonary pressure was normal in 50 (74%) patients, pulmonary hypertension was classified as mild in 9 (13.4%), moderate in 4 (7.4%) and severe in 4 (5.9%). Performing the statistical analysis comparing the pulmonary pressures in the pre and post-surgery, the pulmonary pressure is normalized significantly, representing a P value of 0.0001. Table 4.

Early mortality in 3 patients (4.4%), the cause of death in 2 patients was cardiogenic shock and in 1 septic shock. One patient presented late mortality and the cause was septic shock. Graph 2. Average length of hospital stay was 12 days (5-68). Table 3.

Follow-up was lost in 9 patients. In the rest, the average follow-up time was 4.7 years (1-14). 9 patients presented early post-surgical complications, 1 mediastinits, 1 endocarditis, 1 left AV valve insufficiency that required surgical intervention, 6 patients presented complete AV block that required pacemaker implantation. Table 3. 5 patients presented late post-surgical complications, 1 endocarditis, 1 severe left AV valve insufficiency that required reoperation and 3 complete AV block that required pacemaker implantation.

The most recently measured pulmonary systolic pressure in the mean follow-up was 34 mmHg (13-76). Pulmonary pressure was normal in 38 (70%), pulmonary hypertension was classified as mild in 9 (16.6%), moderate in 4 (7.4%) and severe in 3 (5.5%). Table 4. All are in functional class I.

At follow-up, 21 patients do not require medical treatment. Only 3 receive Sildenafil, the 3 patients who have severe pulmonary hypertension. 3 of them digoxin. All those requiring treatment have diuretics and 21 patients also receive oral antihypertensive drugs. Table 3.

Discussion

In this 15-year study, we observed that 110 patients underwent surgery to correct the ventricular atrium septum defect, of which 61% of the total presented ages older than one year of life and this is due to a poor early diagnosis that includes the lack of gathering the population to detect cardiac pathologies in our environment. There are several studies in the literature that state that the best time for surgical intervention is between the first 3 to 6 months of life, which is when the natural course of pulmonary vascular disease begins [1-3, 5], and even demonstrated in the study by G. Stellin [8] and collaborators, which further reduces reoperation rates in operated patients younger than 3 months. With these references and with the clear indications, they led us to the question that promote to this study to demonstrate that despite operating patients with established pulmonary vascular disease, our numbers are comparable.

In the study by G. Salil *et al.* [1] they compare two eras of early surgical treatment, before the 90's and a late one after the 90's where they present a mortality rate of 10% and a reoperation rate of 13% of their total sample, it is clear that the age in the patients sometimes corrected in the early age were children older than 1 year and in the late age they were younger than one year, the statistical analysis where they are old is not a risk factor for mortality instead it is a risk factor for reoperation. If we analyze our data, the mortality they had was 4.4%, very similar to the study carried out in 7 different hospitals in North America [4], which was 4%.

Delayed presentation in patients with trisomy 21 is also unusual, but one of the factors that allows these patients to progress relatively without symptoms until an older age may be the protective vasoconstrictive effect of hypoxia secondary to sleep apnea that these patients present. [7]. In our study these patients represent 61% of our population similar to the study carried out by O. Xie *et al.* [6] where the operated patients older than 6 months of age with trisomy 21 were 60% of their population.

We observe the pulmonary pressures in the preoperative period where more than 90% of the patients present pulmonary hipertension and of these 71% are severe, but once the defect is corrected, the pulmonary pressures reach normal figures during the immediate postoperative period, even maintaining normal in the followup over the years. The study carried out by P. Bambul Heck *et al.* [2] where they study the reversibility of preoperative hypertension in patients with ventricular and atrioventricular septation defects, found that in the postoperative period 1.3% of their patients maintained high pulmonary pressures requiring treatment. with pulmonary antihypertensive drugs. In our study, they represent 5.5% and these patients are the only ones receiving pulmonary antihypertensive treatment [9].

Not all patients underwent preoperative diagnostic catheterization, but of the total number of patients undergoing this study, we observed that 71% of these had high but mobile lung resistance to oxygen challenge. A striking fact is that the patients who died both in the immediate and late post-operative periods did undergo this diagnostic study and had high lung resistance.

Conclusion

Treating patients older than one year with an atrioventricular septal defect is a safe procedure with a low mortality and morbidity rate, with a significant decrease in pulmonary artery systolic pressure both in the immediate postoperative period and in long-term follow-up. As a risk factor, mortality would be related to high pulmonary resistance, however we find ourselves in the limitation that not all our patients underwent preoperative diagnostic catheterization.

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