ARTIGO ORIGINAL ORIGINAL ARTICLE

TWELVE YEARS OF COMPLETE ATRIOVENTRICULAR SEPTAL DEFECT REPAIR

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Abstract

Background: Surgical repair is the standard treatment for complete atrioventricular septal defect. At our institution, this repair is performed by single patch, modified single patch or two patch techniques, according to the surgeon preferences and the surgical anatomy of the defect. The goal of this study was to evaluate our results from the last twelve years.

Methods: From June 2006 to June 2018, 81 children with complete atrioventricular septal defect (without tetralogy of Fallot or unbalanced ventricles) were submitted to surgical repair at our institution. Data from all patients was retrospectively collected and evaluated.

Results: The average age was 6.9 ± 13.7 months and 84% had Down syndrome. Eighty percent were symptomatic and 6 patients were previously submitted to pulmonary artery banding. No more that mild left atrioventricular valve insufficiency was found in 84% and 89% of the patients, at discharge and follow-up, respectively. Small residual septal defects were present in 27% at discharge; during follow-up, 41% of these closed spontaneously. Pulmonary hypertension at discharge and follow-up appeared in 3.7% and 1.3%, respectively. Permanente pacemaker was implanted in 3 patients. Left ventricle outflow tract obstruction was found in 3 patients and 2 needed surgical correction. At follow-up (40 ± 38 months), 90% of the patients presented NYHA functional class I. No significant differences in the main repair outcomes were found between techniques, with the exception of small residual septal defects, although the groups were unmatched.

Conclusions: Overall and regardless of the technique used for the repair of complete AVSD, good early and midterm outcomes were achieved.

INTRODUCTION

Atrioventricular septal defects (AVSD) are a group of common congenital heart defects with an *ostium primum* defect and an (separated or common) atrioventricular (AV) orifice; an inlet ventricular septal defect may or not be present.[1] According to the ventricular defect size and the status of the AV valves, it can be classified as complete, intermediate or partial. The complete type presents with two components, one ventricular (ventricular septal defect - VSD) and another atrial (atrial septal defect - ASD), and a single common AV valve. In addition, other heart anomalies can coexist, sometimes as part of syndromes, most frequently, Down syndrome.[2]

If untreated, 50% of the patients with complete AVSD die in the first year of life due to heart failure and pulmonary infections. Untreated patients develop pulmonary hypertension (PHT) and Eisenmenger syndrome, therefore, an early surgical intervention is necessary.[3] However, the surgical treatment is challenging, requiring closure of the septal defects and repair of any AV valves dysfunction. Currently, 3 different techniques are commonly used: single patch technique (that corrects the septal defect with one single patch, dividing the bridging leaflets and then reattaching them to the patch)[4]; two-patch technique (that uses separated patches to correct de ASD and VSD, fixing the valve leaflets between them)[5]; and modified single patch technique or "Australian" technique (where the VSD is directly attached to the valve leaflets and the ASD closed with a patch).[6] So far, none have shown superiority over the others.[7,8]

The classic single patch technique was the method of first choice in our initial experience, with the modified single patch technique used by favorable anatomy. In 2013 the two-patch technique was added to the routine.

The aim of this study was to review the results of our experience using these 3 techniques to repair complete AVSD over the last 12 years.

PATIENTS AND METHODS

Population and Methods

Between June 2006 and June 2018, 81 children with complete AVSD, without tetralogy of Fallot or unbalanced ventricles, were submitted to surgical repair at our center. Single-patch technique was used in 41 patients; modified single patch technique and two-patch techniques were used in 20 patients each. The clinical data of all patients was retrospectively reviewed, with respect to patients' preoperative characteristics, operative parameters, perioperative outcomes and follow up.

Surgical aspects

Operations were performed after median sternotomy, on cardio-pulmonary bypass with bicaval cannulation (in mild to moderate hypothermia). Cardioplegic arrest was achieved with crystalloid cardioplegia. Intraoperative echocardiography was routinely performed in each patient; suboptimal results - severe AV regurgitation; moderate/severe AV or left ventricle outflow tract [LVOT] obstruction stenosis; residual ASD/VSD> 2 mm - were revised.

Statistical analysis

Statistical analysis was carried out using SPSS v24. Continuous variables were treated as mean and standard deviation and compared with t-student and ANOVA tests. Categorical variables were summarized as the number and/or percentage of subjects in each category and compared with Chi square/Fisher's exact testes.

RESULTS

Baseline

Overall, the patients were 6.9 ± 13.7 months old, 52% were female, the mean weight was 5.2 ± 2.9 kg, 84% had Down syndrome, 69% had another atrial septum defect, and 7.4% were previously submitted to PA banding. Eighty percent were symptomatic.

Baseline characteristics for each technique are summarized in table 1. The group of patients treated with the modified single patch technique (mSPG) were older on average, had more body weight, more Rastelli type A defects, less cases of Down syndrome, lower frequency of PHT and no previous PA banding. The groups of patients treated with the single patch (SPG) and two patch (TPG) techniques were more similar in the baseline characteristics.

Surgical Procedure

Surgical times varied with concomitant procedures and reoperation. The mean surgical times of the patients treated with a simple combination of procedures (AVSD and left AV valve valve repair, with/without persistent ductus arteriosus ligation) is described in the Table 2.

Closure of preexisting PFO/ASD II was the most common additional procedure in all groups (Table 3).

Intraoperative revision occurred less in the SPG (4.8%); TPG and mSPG had similar number of revisions (25% and 20%, respectively). Left or right AV-valve repair was the most frequent reason for revision.

Table 1 **Base characteristics of the patients by technique**

	Two Patch n=20	Single Patch n=41	Modified Single Patch n=20	*p*
Sex (female/male)	50/50 %	49/51 %	50/45 %	0.945
Age (months)	5.4 ± 3.7	6.4 ± 16.0	9.3 ± 15.2	0.636
Weight (kg)	5.2 ± 1.2	4.6 ± 3.2	6.3 ± 3.1	0.083
Height (cm)	60.4 ± 6.6	56.0 ± 12.9	54.5 ± 13.1	0.032
Rastelli type (A, B, C)	45 | 5 | 50 %	19 | 5 | 76 %	60 | 0 | 35 %	0.011
NYHA class (I, II, III, IV)	30 | 25 | 25 | 20 %	12 | 42 | 44 | 2 %	25 | 55 | 20 | 0 %	0.014
Comorbidity				
Down syndrome	95 %	85 %	70 %	0.028
PFO or ASD type II	85 %	59 %	75 %	0.089
Hypothyroidism	30 %	24 %	25 %	0.890
Former premature baby	15 %	7 %	5 %	0.484
Previous PA banding	20 %	5 %	0 %	0.037
Patent ductus arteriosus	10 %	5 %	20 %	0.178
Left superior vena cava	10 %	7 %	5 %	0.833
Pulmonary hypertension	70 %	78 %	45 %	0.034

ASD - Atrial septal defect; NYHA - New York Heart Association; PA - Pulmonary artery; PFO - patent foramen ovale; RBBB - Right bundle branch block
Continuous variables are presented in the format mean ± standard deviation (minimum - maximum values).

Table 2 **Surgical times**

	Single Patch n=29	Modified Single Patch n=15	*p*
Aorta Clamping (minutes)	119 ± 21.1 (79)	85 ± 18.4 (60)	101 ± 14.8 (77)
Cardiopulmonary bypass (minutes)	179 ± 31.1 (108)	128 ± 30.1 (87)	150 ± 32.1 (113)
Operative time (minutes)	261 ± 34.8 (192)	213 ± 30.4 (160)	229 ± 32.3 (180)

Only cases were atrioventricular septal defect repair plus left atrioventricular valve repair plus interatrial communication closure with or without ductus arteriosus ligation were performed are included in this table. The groups were unmatched.
Continuous variables are presented in the format mean ± standard deviation (minimum value).

Table 3 **Surgical procedures**

	Single Patch n=41	Modified Single Patch n=20	Two Patch n=20	*p**
Additional procedures	-	-	-	-
Right AV valve repair	6 (15%)	1 (5%)	0	0.129
PFO/ASD II correction	22 (54%)	15 (75%)	17 (85%)	0.034
by direct suture	14 (34%)	13 (65%)	11 (55%)	-
with pericardium patch	8 (20%)	2 (10%)	6 (30%)	-
Ductus arteriosus ligation	2 (4.8%)	5 (25%)	2 (10%)	0.063
PA de-banding (± augment plasty)	2 (4.8%)	0	4 (20%)	0.037
Intra operative revision	2 (4.8%)	4 (20%)	5 (25%)	0.318
Left AV valve insufficiency	1 (2.4%)	2 (10%)	4 (20%)	-
Right AV valve insufficiency	0	2 (10%)	0	-
VSD	1 (2.4%)	0	(1)	-
PA stenosis	0	0	1 (5%)	-

ASD - Atrial septal defect; AV - Atrioventricular; AVSD - Atrioventricular septal defect; PA - Pulmonary artery; PFO - Patent foramen ovale; VSD - Ventricular septal defect.
*The groups were unmatched.

ICU course

The mSPG had a shorter ventilation (4.1 ± 3.0 days), needed less catecolamines (4.6 ± 3.0 days) and phosphodiasterase inhibitors (2.3 ± 2.4 days) support and a shorter ICU (8.7 ± 5.4 days) and in-hospital (17.4 ± 7.5 days) stay (Table 4), whereas the amount of catecholamines and PDI didn't reach statistical significance.

Despite considerable numeric differences in the studied complications, none reached statistical significance, with urinary infection with identified agent being the exception (see Table 4 for details). There was no wound infection in the postoperative period.

Atrioventricular valves

Overall, 77% of the patients presented with a mild or trivial left AV valve insufficiency preoperatively. No more than a mild regurgitation was found in 84% at discharge and in 89% at follow-up; four patients (3 in the SPG and 1 in the TPG) needed to be reoperated in the early postoperative period due to severe left AV valve insufficiency. Freedom from moderate to severe regurgitation preoperative, postoperative and at follow-up were, 76%, 85% and 90% in the TPG, 78%, 88% and 85% in the SPG, and 75%, 75% and 90% in the mSPG, respectively (Figure 1A).

For the right AV valve, not more than a mild insufficiency was present in 95% of the patients at discharge and 96% at follow-up. Groups details are represented in Figure 1B.

There was no relevant (more than mild) stenosis of either the right or left AV valves, both early postoperative or during follow up.

None of the groups reached statistical significance, regarding the left and right AV valves insufficiency, in the in-hospital and follow-up phases.

Residual septal defects

During in-hospital stay, 1 patient in the SPG (2.4%) needed reoperation due to a large residual VSD; in the TPG, 1 patient was reoperated because of severe left AV valve insufficiency (as mentioned above) and, concomitantly, the VSD was also revised because of residual large VSD. In both cases, the VSD could be almost completely closed. At discharge, 45% of the patients in the TPG presented with small residual septal defects (ASD or VSD) (<2mm); the incidence was 35% in the mSPG and 17% in

Table 4 **Early post-operative results**

	Single Patch n=41	Modified Single Patch n=20	Two Patch n=20	*p**
Ventilation (days)	7.1 ± 3.8	4.1 ± 3.0	6.9 ± 4.0	0.011
Catecolamines (days)	5.9 ± 3.1	4.6 ± 3.0	5.9 ± 3.7	0.427
Phosphodiasterase inhibitors (days)	6.4 ± 8.1	2.3 ± 2.4	5.3 ± 4.1	0.113
ICU stay (days)	14.5 ± 11.5	8.7 ± 5.4	17.8 ± 11.8	0.026
Hospital stay (days)	26.5 ± 15.4	17.4 ± 7.5	31.9 ± 16.3	0.006
Post-operative complications Suspected infection (at least one) [confirmed with identified agent]	71 % [27 %]	45 % [10 %]	70 % [60 %]	0.119 [0.002]
Suspected infection (at least one) [confirmed with identified agent]	12 % [10 %]	10 % [10 %]	35 % [35 %]	0.053 [0.029]
Suspected urinary infection [confirmed with identified agent]	34 % [17 %]	30 % [5 %]	40 % [20 %]	0.799 [0.347]
Suspected respiratory infection [confirmed with identified agent]	2 % [2 %]	0% [0 %]	5 % [5 %]	0.595
Pulmonary hypertension crisis	17 %	20 %	45 %	0.051
Arrhythmia**	32 %	15 %	30 %	0.367
Permanent pacemaker implantation	2.4 %	5 %	5 %	0.830
Chylothorax	17 %	15 %	30 %	0.407
Atelectasis or Pneumothorax (significant)	20 %	20 %	30 %	0.628
Death	0 %	0 %	0 %	-

ASD - Atrial septal defect; AV - Atrioventricular; AVSD - Atrioventricular septal defect; PA - Pulmonary artery; PFO - Patent foramen ovale; VSD - Ventricular septal defect.
*The groups were unmatched.

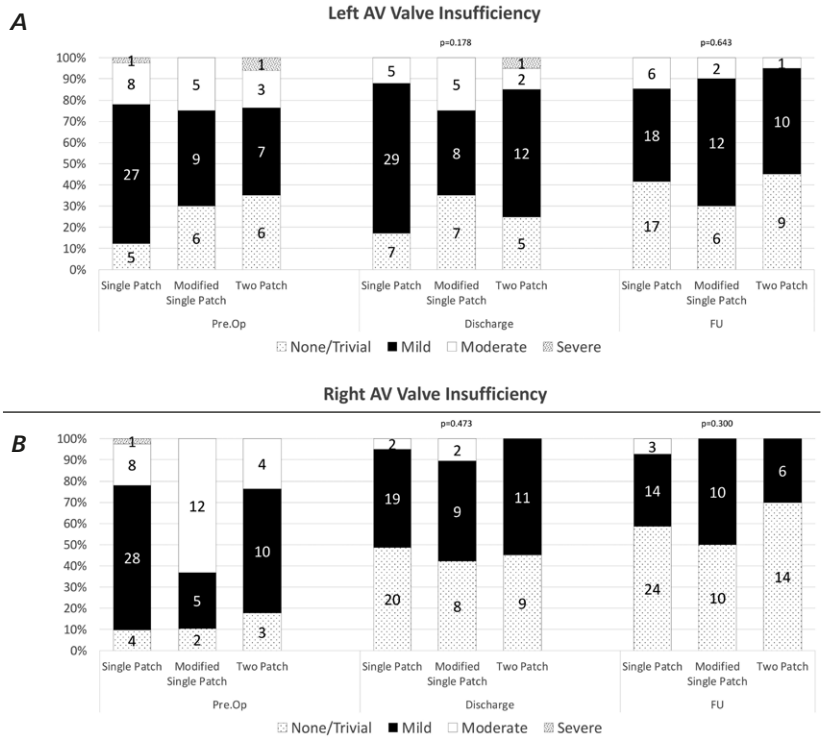

Figure 1

*Bar charts representing progression of left (**A**) and right (**B**) atrioventricular valve insufficiency for each group. The contemplated stages are: status previous to surgery (Pre.Op); status in the last in-hospital evaluation (Discharge); and status at the last follow-up (FU). The numbers inside the bars are the number of patients with the respective level of insufficiency. On top of each stage is the p value for the comparison between groups on the respective stage. The groups were unmatched.*

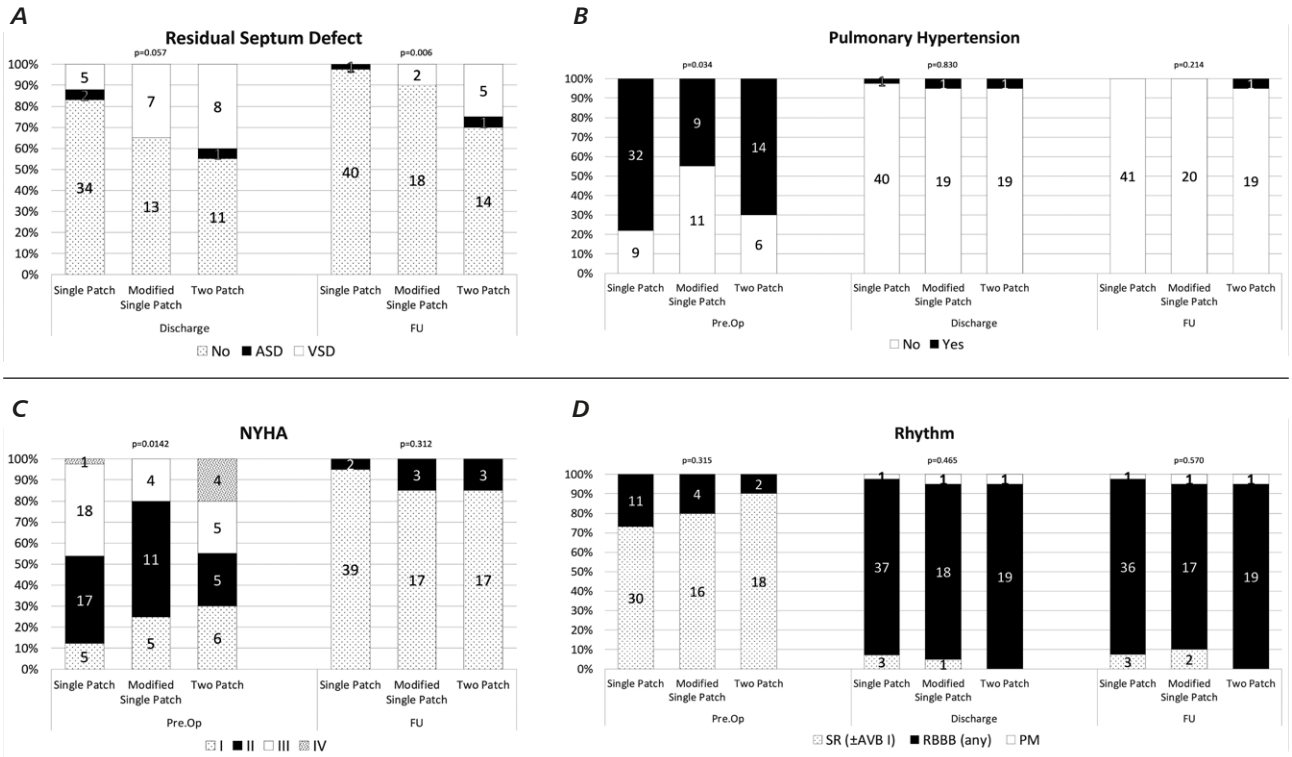

Figure 2

*Bar charts representing progression of residual septal defects (**A**), pulmonary hypertension (**B**), heart rhythm (**C**) and New York Heart Association (NYHA) class. The contemplated stages are: pre-operative status (Pre.Op); status in the last in-hospital evaluation (Discharge); and status at the last follow-up (FU). The numbers inside the bars are the number of patients with the respective level of insufficiency. On top of each stage is the p value for the comparison between groups on the respective stage. The groups were unmatched.*
ASD - Atrial septal defect; AVB I – First-degree atrioventricular block; PM - Permanent pacemaker; SR - Sinus rhythm; RBBB - Right bundle branch block; VSD - Ventricular septal defect.

the SPG (Figure 2A). During follow-up, the incidence, due to spontaneous closure, reduced to 30%, 10% and 2.4% (p=0.006), respectively. None of the residual defects was hemodynamically relevant.

Pulmonary hypertension

Prior to surgery, PHT was present in 68% of the patients, with the incidence varying between groups (see Table 1 for details). After surgery, PHT crisis occurred in 45% of the TPG, 20% of the mSPG and 17% of the SPG. At discharge, 1 patient in each group had PHT (Figure 2B). At follow-up, only 1 patient (from the TPG) remained with high pressure levels.

Statically significant differences between groups were only found in the preoperative frequency of PHT (p=0.034).

Rhythm

In the early postoperative period, temporary, but clinically relevant, arrhythmic episodes (such as heart block and junctional ectopic tachycardia) occurred in 30% of the TPG and SPG; in the mSPG the frequency was 15% (Figure 1D).

Permanent pacemaker (PM) implantation after surgery was 2.4% in the SPG and 5% in both mSPG and TPG.

Right bundle branch block (RBBB) was the most common permanent rhythm anomaly in all groups; overall it was 91%. All the 17 patients that previously to surgery had an AV block (AV block I or RBBB) sustained RBBB after surgery. New RBBB after procedure was 89%. This outcome was kept during follow-up, similarly, in all groups.

No statically significant differences were found between groups, regarding rhythm anomalies.

Survival, functional status and other follow--up data

Mean follow-up time was different between groups: 17.1 ± 15.8 months in TPG, 30.6 ± 34.7 months in mSPG and 57.2 ± 39.7 months in SPG (p<0.001).

No patient died in the perioperative period or during the follow-up.

More than 85% of the patients presented NYHA functional class I at follow-up, without meaningful differences between the groups; the remaining were at class II (Figure 2C).

LVOT obstruction was found in 3 patients: 2 in the SPG and 1 in the TPG. In the 2 patients from the SPG the obstruction needed surgical correction; these were the only AVSD-related reoperations that occurred in all groups during follow-up.

During follow-up, 1 case of endocarditis was found in the SPG, 28 months after surgery, and 1 case of

sepsis occurred in the mSPG, 16 months post-surgery. Four patients in SPG, 4 in the TPG and 1 in the mSPG were hospitalized due to respiratory infection, at least once; all but one, had Down syndrome.

COMMENT

The definitive treatment for complete AVSD is the surgical repair. Several techniques have emerged to resolve this challenging congenital heart defect, with three of them being commonly used: single patch technique, two-patch, and modified single patch technique. None have shown superiority over the others and, regardless of the technique, several post-operative complications (left AV valve dysfunction, conduction system disturbance, postoperative pulmonary hypertensive crisis etc.) are known to occur.

All three techniques are being currently used at our institution. The technique of repair is generally decided intraoperative according to the anatomy and the surgeon's preferences, which outlines the reason for the different population baseline characteristics between the groups. A small VSD usually leads to smaller left to right shunt and, consequently, less PHT and symptoms. This allows the patients to mature more before needing surgery (either PA banding or defect repair), which leads to a faster and less complicated recovery. The modified single patch technique was mainly used when the VSD was small and this very important anatomical advantage was the reason why mSPG presented overall better results. Despite this, in general, all three techniques resulted in satisfactory surgical and clinical results, which improved during follow-up. Some patients needed reoperation in the early postoperative period due to severe left AV valve regurgitation. In particular, the number of intraoperative revisions in the TPG and mSPG was high. This is justified by the small number of patients and the initial learning curve. Although one of the patients was discharged with unsatisfactory results regarding the left AV valve, in the follow up the overall patient population showed spontaneous improvement with no cases of severe left AV valve insufficiency.

Right AV valve regurgitation was greatly reduced with surgery, which was also favored from the change in pressure after closing the intracardiac shunts, positioning the right AV-valve in the low-pressure circulation. Small and hemodynamically not relevant residual septal defects were not uncommon in the early postoperative period, being most frequent in the TPG; this may be related to the additional location for possible residual AVSD that the two-patch technique presents. With time, the residual defects tended to close spontaneously, and the differences found between groups (favoring the single patch technique) may result from the different follow-up times (longer in the SPG). PHT improved, as anticipated, greatly during recovery. Despite high frequencies of PHT before surgery and PHT crisis in the first days after operation, only a minority of patients showed signs of elevated pulmonary pressure at discharge, that persisted in only one patient at follow-up. All structural and physiologic improvements reflected in the functional status, with at least 90% in NYHA class I, and the remaining in class II, and in the survival, with no mortality.

Permanent PM implantation and LVOT obstruction are serious complications associated with AVSD repair. Global PM implantation was 3.7% (with 1 case in each group). Regardless of the technique, RBBB was much more frequent. This conduction anomaly is underreported, since it is considered less problematic than complete AV Block. However, an increased risk of mortality has been found in adults with RBBB (both in the general population and in patients with heart disease) and it can be expected that this may also have impact on the child's long-term prognosis.[9,10] LVOT obstruction also occurred in 3.7% of the patients (2 in the SPG; 1 in the TPG), and 2 of those patients required surgical treatment.

After literature research, we found the incidence of the major complications after surgical correction for complete AVSD in our study (PM implantation, AV-Valve regurgitation, LVOT obstruction, etc) comparable with the ones in early reports.[5,7,8,11]

Infections during the recovery of a surgery is a well-known risk factor for mortality. For this reason, antibiotics are in many cases initiated in presence of suggestive symptoms/signs or elevated inflammatory markers, without a previous clinical diagnosis of infection. Detailed information about infections after complete AVSD repair is not always present in the literature. Reports of sepsis ranging from 5.3 to 7.5% and respiratory infection from 8.7 to 59% were found[12,13,14,15]; these also report a few cases of infection-related mortality. By direct comparison we obtained less cases of sepsis. Long ventilation demand, long ICU and hospital stay on a very young population with 84% Down syndrome patients is probably the reason for the number of infections. Supporting this idea is the fact that both TPG and SPG, with a more fragile population and longer ventilation length and ICU stay, acquired more infections, comparing to the mSPD. On the other hand, these differences may exist due to a low number of patients. All cases were treated and no associated mortality occurred. Concerning infections during follow-up, respiratory infections were the most frequent. Endocarditis/sepsis occurred in 2 patients of the SPG (2.5% globally, 4.9% within group), more than 1 year from surgery. The patents were hospitalized, properly treated and recovered good functional status without sequels.

Many surgical groups have compared the different techniques of repair for CAVSD, with none showing definitive superiority over the other. We added the two patch technique to our routine in order to expand our ability to better treat the variety of surgical anatomies of the AV canal, with the surgeon deciding, intraoperative, which technique would best fit the patient. The mandatory echocardiography in the operating room helped us revise unsatisfactory results immediately, reducing the need for late re-operations and, probably, also the overall length of

hospital stay. Furthermore, residual AV-Valve insufficiency tended to reduce during follow up and small septal defects to close spontaneously, allowing the patients to achieve a good functional status with excellent survival and high freedom from re-intervention over time.

Study limitations

This report shows several limitations like the low number of patients and the relative short follow-up times of the mSPG and the TPG. Furthermore, the groups were not matched.

Disclosures and Freedom of Investigation

The authors report no conflicts of interest regarding the content of this study.

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