

Cone Reconstruction in Ebstein's Anomaly Repair: Early and Long-term Results

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Abstract

Background: The main Ebstein anomaly (EA) repairs are based on the monocusp reconstruction of the tricuspid valve and are limited by the frequent need for replacement or the high recurrence of valve regurgitation.

Objectives: To evaluate the feasibility and effects of anatomical repair of Ebstein's anomaly using the cone reconstruction technique on patients' clinical evaluation, tricuspid valve function and right ventricular morphology.

Methods: We compared the clinical, echocardiographic and radiological data of 52 consecutive patients, with a mean age of 18.5 ± 13.8 years, submitted to the cone reconstruction technique, obtained in the preoperative, early postoperative (EPO) and long-term (LPO) periods.

Results: There were two in-hospital deaths (3.8%) and two more during the follow-up. Mean functional class of pre-operative heart failure improved from 2.2 to 1.2 after 57 months of mean follow up of 97% of patients ($p < 0.001$). The mean degree of preoperative tricuspid regurgitation decreased from 3.6 to 1.6 in the EPO ($p < 0.001$), remaining at 1.9 in LPO period ($p > 0.05$). The indexed RV functional area increased from 8.53 ± 7.02 cm²/m² preoperatively to 21.01 ± 6.87 cm²/m² in the EPO ($p < 0.001$) and remained unchanged at 20.28 ± 5.26 cm²/m² in LPO period ($p > 0.05$). The mean cardiothoracic ratio was decreased from 0.66 ± 0.09 to 0.54 ± 0.06 ($p < 0.001$) in the long term.

Conclusions: The cone technique showed low in-hospital mortality, resulting in an effective and long-lasting repair of tricuspid regurgitation, restoring the functional area of the right ventricle and allowing reverse remodeling of the heart and clinical improvement in most patients in the long term. (Arq Bras Cardiol 2011; 97(3) : 199-208)

Keywords: Ebstein anomaly/physiopathology; ebstein anomaly/surgery; tricuspid valve/surgery; perioperative period.

Introduction

Ebstein's anomaly, due to the complexity of the anatomical and functional alterations that involve the tricuspid valve and right ventricle, has resulted in the development of different surgical techniques for its correction.

In 1972, Danielson et al¹ developed a technique for tricuspid valve repair, modified from Hardy's operation², which became one of the most frequently used in the treatment of Ebstein's anomaly. This technique, which includes transversal plication of the atrialized portion of the RV, results in a tricuspid valve formed by a single leaflet (monocusp) coapted to the interventricular septum. However, the need to have a large and mobile anterior valve to achieve the objective of correcting right AV regurgitation

limits this operation to a restrict group of anatomical variations, which require tricuspid valve replacement in 36% to 65% of cases³⁻⁵.

Carpentier et al⁶ reported a new technique in 1988, with longitudinal plication of the right ventricle and repositioning of the right AV valve to the anatomically correct level, with remodeling and strengthening of the valve annulus with a prosthetic annulus, resulting in good right ventricular morphology, and showing its capacity of being applied to most anatomical presentations of the disease, but mortality was high (14%) in its initial series, with frequent long-term complications. The subsequent publication of a similar technique showed high incidence of moderate and important tricuspid regurgitation⁷.

In 1989, we developed a new surgical technique, called cone reconstruction of the tricuspid valve, aiming at the reconstruction of the valve similar to a normal one⁸. After 1993, the surgery's technical concepts were standardized and we started to use it routinely for the treatment of patients with Ebstein's anomaly. This surgery was applied to the first 40 consecutive patients with a mortality of

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2.5% and no valve replacement, with excellent immediate echocardiographic results, clinical improvement and low incidence of reoperation after a medium-term follow-up⁹.

Objectives

To assess the technical viability of the cone reconstruction for Ebstein's anomaly, evaluating its effects on the clinical evolution of patients and the morphology and function of the tricuspid valve and right ventricle in the early postoperative and long-term periods.

Methods

The operations were carried out at the Hospital São Joaquim da Real e Benemérita Associação Portuguesa de Beneficência de São Paulo by two surgeons from the same team.

The long-term echocardiographic assessment was performed at Instituto do Coração do Hospital das Clínicas da Universidade de São Paulo (INCOR), after obtaining patients' informed consent and project approval by the Research Ethics Committee of Faculdade de Medicina da Universidade de São Paulo, protocol number 055/05.

Study population

The population of the present study consists of 52 patients with Ebstein's anomaly consecutively submitted to surgical repair with the cone reconstruction technique, between November 1993 and December 2006, whose clinical data are shown in Table 1. Surgical treatment was indicated according to previously described criteria⁹ and the same technique was used in all patients with surgical indication, regardless of the severity and anatomical type. Two neonates submitted to another type of surgical procedure were not included in this series.

Surgical Technique

Using cardiopulmonary bypass (CPB) with moderate systemic hypothermia and myocardial protection with blood cardioplegia, a right oblique atriotomy was performed, proximally detaching the anterior and posterior tricuspid megaleaflets. Then, the abnormal papillary muscles and other tissues between the valves and right ventricular wall were severed, taking special care to preserve the attachment to the right ventricular apex. The free border of the posterior leaflet is rotated clockwise and sutured in the septal border of the anterior leaflet, transforming the new tricuspid valve in a cone with a fixed vertex at the apex of the right ventricle. Then, the longitudinal plication of the atrialized RV is performed, excluding its thin portion.

The new valve annulus is fitted at the anatomically correct level by means of plication of the true annulus, adapting it to the base of the previously constructed cone. The valve is attached to true annulus by interrupted sutures. Subsequently, a continuous suture is performed with reinforcement of the new junction of the tricuspid valve to the annulus, using very superficial stitches close to the area of the atrioventricular node. This suture uses polypropylene thread in adult patients and polydioxanone (PDS) in children, when there is interest in AV junction growth.

Finally, regarding the closing of the atrial septal defect, the capacity to allow the right-to-left flow is preserved by partially closing of the foramen ovale. The *ostium secundum* type atrial septal defect, if present, is closed with the valve technique. More technical details can be seen in other publications^{9,10}.

Additional surgical procedures added to the repair consisted of closing the ventricular septal defect, mitral valve repair, correction of partial anomalous pulmonary vein drainage, three extensions of the RV outflow tract and 11 cases of anomalous conduction over accessory pathways.

Table 1 – Preoperative clinical characteristics of the 52 patients

Variable	Values
Age: range (mean ± SD)	0.25 - 49 years (18,5 ± 13,8)
Sex	
Male	28 (53.8%)
Female	24 (46.2%)
Cyanosis	30 (58%)
Edema	3 (5.8%)
LV cardiomyopathy	2 (3.8%)
Associated cardiac defects	
ASD – Foramen ovale	46 (88.5%)
WPW	12 (23.1%)
VSD	1 (1.9%)
Pulmonary stenosis	3 (5.8%)
Partial anomalous pulmonary venous drainage	1 (1.9%)

LV – left ventricle; SD – standard-deviation; ASD – atrial septal defect; VSD – ventricular septal defect; WPW – Wolff-Parkinson-White syndrome.

Clinical outcome

The following were considered relevant: presence of cyanosis, cardiac arrhythmias especially WPW, data related to cardiopulmonary bypass time, time of aortic clamping, length of intensive care unit (ICU) stay, length of hospital stay and in-hospital mortality, defined as death in the first month of postoperative period and functional class according to the NYHA.

The long-term clinical follow-up was achieved through physical examination or telephone interview, following a specific protocol form, analyzing the following outcomes: death, reoperation, arrhythmias and need for pacemakers.

Chest X-ray

Chest x-rays of patients in frontal view, obtained in the preoperative period, were used to calculate the cardiothoracic ratios, which were compared with the ratios during the long-term evolution. The cardiothoracic ratio was calculated using the method described by Danzer¹¹.

Echocardiogram

The echocardiographic studies obtained before surgery (preoperative) and at hospital discharge (early postoperative) were recorded for later evaluation by the same echocardiographer responsible for long-term echocardiographic studies.

The anteroposterior diameter of the true tricuspid annulus and heart cavity dimensions were measured in the four-chamber view. We evaluated tricuspid valve function regarding stenosis, regurgitation or both. Valve regurgitation was assessed by the modified method of Suzuki et al¹². Heart cavity areas were measured and indexed by body surface area. The cavity areas were estimated by planimetry using a modified two-dimensional mode.

Statistical analysis

Data are expressed as mean and standard deviation (SD). Qualitative variables were expressed as a percentage. We used the GraphPad Prism software (version 4.0, GraphPad Software, Inc., San Diego, Calif.). The descriptive level was 0.05. For continuous variables, we used to compare means using the unpaired t-test, two-tailed; for categorical variables functional class pre-and post-operative, the Wilcoxon test, and for the grades of tricuspid insufficiency, the Friedman test. Changes in size of the tricuspid valve annulus and RV area were analyzed by repeated measures of variance (ANOVA), with completion by the Bonferroni test. The actuarial survival was determined by the Kaplan-Meier method, and data presented in the standard error.

Numerical data are expressed as mean and standard deviation (SD). Qualitative variables were expressed as percentage. We used the GraphPad Prism software (version 4.0, GraphPad Software, Inc., San Diego, Calif.). Significance was set at 0.05. For continuous variables, means were compared using the two-tailed unpaired t-test; for the categorical variables pre-and postoperative functional class, the Wilcoxon test was used and for the tricuspid

regurgitation grades, Friedman's test was used. Changes in size of the tricuspid valve annulus and RV area were analyzed by repeated measures of analysis of variance (ANOVA), complemented by Bonferroni test. The actuarial survival curve was determined by the Kaplan-Meier method and values are presented by standard error.

Results

Perioperative and early postoperative periods

The CPB time ranged from 45 to 185 minutes (mean 112.3 ± 33.23 minutes) and aortic clamping, 25 to 115 minutes (mean 74.35 ± 21.93 minutes). All patients underwent tricuspid valve repair with cone reconstruction technique and tricuspid valve replacement was not required in any patient.

The mean length of ICU stay was 4 ± 4.7 days, ranging from 1 to 31, and length of hospital stay ranged from 7 to 45 days, with a mean of 13.7 ± 8.6 . In-hospital mortality was 3.8% (n = 2); the first patient died due to biventricular cardiomyopathy by prolonged hypoxia in the preoperative period and the low cardiac output was the cause of death on the fourth postoperative day. The second, in a patient aged 40, died on the seventh postoperative day due to right heart failure. In both cases, tricuspid valvuloplasty was effective in the immediate postoperative echocardiographic assessment.

Long-term follow-up

These data were obtained with the follow-up of 49 patients, 97.9% of the 50 who survived the hospital phase. There were two deaths: one due to tricuspid valve endocarditis after dental infection, three years after the surgery, and the second seven years after surgery due to right ventricular dysfunction and ventricular arrhythmias.

Data on heart failure functional class (NYHA) with a follow-up of 9.6 to 162 months (mean $57.44 \text{ m} \pm 45.14$ months), collected in the last evaluation, compared with those obtained preoperatively in 47 patients (97.9%) (Table 2) showed significant functional class improvement in the long-term postoperative period ($p < 0.001$). The survival curve estimated by the Kaplan-Meier method (Figure 1), which includes hospital mortality, showed survival values of $94.1 \pm 3.2\%$ from one month to six years of follow-up and of $86.2 \pm 8\%$ after seven years of follow-up. Reoperations were necessary in four patients and tricuspid re-plasty was performed in all of them in the third, fourth, fifth and tenth years after surgery, as described: significant tricuspid regurgitation in two cases, suture dehiscence of septal portion and presence of previously undiagnosed WPW associated with a small wound dehiscence in the third patient. These patients had good subsequent outcome. The other patient, after several attempts at catheter ablation of arrhythmias, was reoperated by another team and the cause of valve failure remained unclear; she died two years later due to right ventricular dysfunction and ventricular arrhythmia.

Three patients had atrial arrhythmias, and one underwent successful ablation and the others were clinically controlled. Of the 11 patients with Wolff-Parkinson-White (WPW) syndrome

undergoing surgical section of the anomalous bundle in the same surgical procedure, success was obtained in 10. The other patient underwent repeated ablation attempts, developing tricuspid regurgitation, which required reoperation in the fifth postoperative year.

Pacemaker implantation was necessary due to a first-degree AV block, which after the use of amiodarone, showed intermittent episodes of total AV block one year after surgery. A patient with severe biventricular cardiomyopathy had a stroke on the sixth postoperative year and is currently in the ninth postoperative year. Although he receives appropriate therapy for heart failure, he remains in functional class III. The remaining patients have remained event-free; one patient even had an uneventful pregnancy to term, two years after surgery. There was no need for tricuspid valve replacement by prosthesis in any patient in this series.

Chest X-ray – Cardiothoracic ratio

There was a significant reduction in the cardiothoracic ratio of patients from 0.66 +/- 0.09, CI: 0.56 to 0.97 preoperatively, to 0.54 +/- 0.06, CI: 0.44 to 0.66 (p

<0.0001) in studies carried out in the same patients with a mean follow-up of 37.2 months.

Echocardiographic results

Full echocardiographic studies were obtained (preoperative, early postoperative and long-term postoperative) in 40 patients. There was a decrease in the degree of right AV valve regurgitation in the early postoperative (EPO) when compared with the preoperative period, and this decrease was maintained in the long term (Table 3, Figure 2). The tricuspid valve annulus was smaller in the EPO (42.8 +/- 12mm to 19 mm +/- 6, p <0.01), but grew up in the long-term evaluation (22 + 6 mm), although it was not statistically significant. This initial decrease caused acceleration in the flow of AV valve in five patients - one of them even had mild stenosis (mean gradient of 7 mmHg) in the long term.

Regarding the dimension of the RV, the measured areas (cm²/m²) indexed by body surface areas of the patients showed an increase in functional RV when comparing the preoperative (8.53 ± 7, 95%CI: 2.13 - 32) and the early postoperative period (21 ± 7, 95%CI: 8.59 - 38.8), p < 0.01, remaining unchanged in the long term (21 ± 5, 95%CI: 12 - 32.4), p > ± 0.05, when

Table 2 – Comparison of paired heart failure functional classes between the preoperative and long-term postoperative periods

Functional class	Preoperative (n)	LPO	Valor de p
Class I	5	44	
Class II	11	2	p < 0.0001
Class III	27	1	
Class IV	4	0	

LPO- long-term postoperative period.

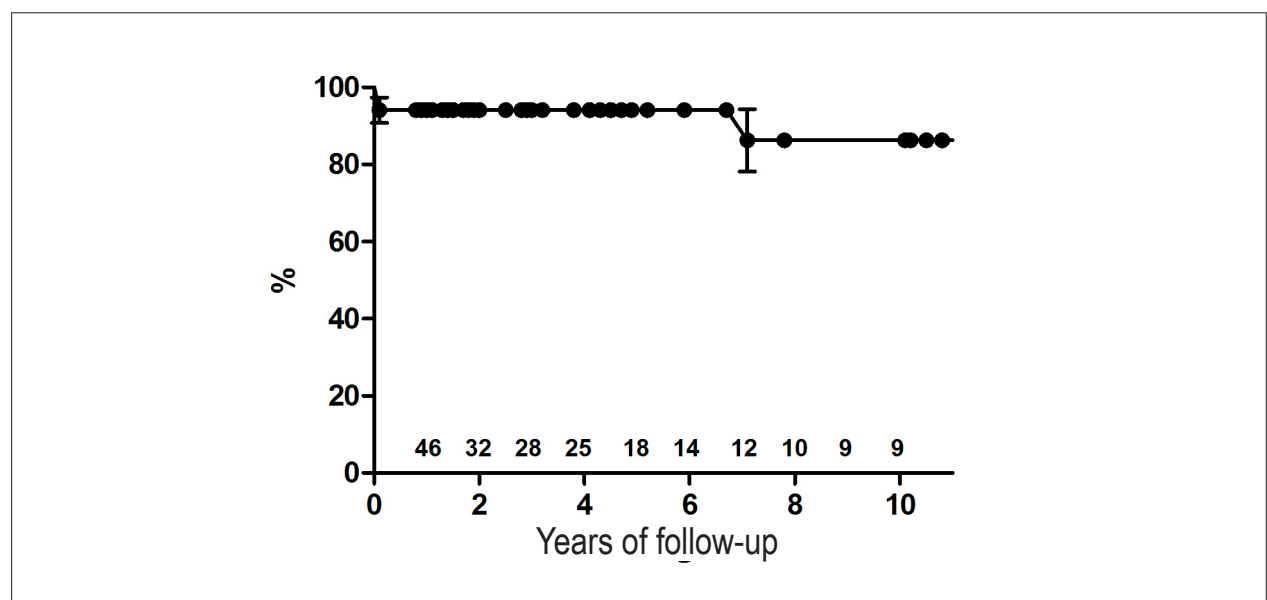


Figure 1 – Actuarial survival curve estimated by Kaplan-Meier shows survival of 94.1 ± 3.2% from 1 month to 6 years of follow-up and 86.2 ± 8% after 7 years. Data are presented with standard error and the numbers indicate patients at risk in each period.

compared with those obtained in the EPO (Figure 3).

There was an indexed decrease in the RA when comparing the preoperative ($29 \pm 12 \text{ cm}^2/\text{m}^2$, 95%CI: -7.78-16.3) and the early postoperative period ($17 \pm 6.6 \text{ cm}^2/\text{m}^2$, 95%CI: -10.8 - 19.35), $p < 0.05$. In the long term, there was a slight decrease of the right atrium ($14 \pm 5.4 \text{ cm}^2/\text{m}^2$, 95%CI: 1.2 - 7.3) compared to the EPO, which was not statistically significant (Figure 4).

Figure 5 illustrates the echocardiographic evolution of a patient, showing the functional right ventricular restoration and the postoperative tricuspid valve anatomy.

Discussion

The cone technique was designed according to some concepts of Carpentier et al⁶, in order to fit the leaflets of the tricuspid valve to the true annulus position and the longitudinal plication of the atrialized right ventricle, restoring ventricular morphology and volume, but it results in anatomically much different valve. Carpentier et al⁶ moved the tricuspid valve, displaced into the right ventricle interior, bringing it to the normal level of the right AV annulus and turning it partially only to reach the septal region. Therefore, reimplantation occurs in the monocusp format, with diastolic flow out of the valve center. In the cone reconstruction, the full clockwise rotation is made, from the lateral margin of all the anterior and posterior leaflets of the tricuspid valve, thus preserving of fixations in the right ventricular apex. Furthermore, the full circumference of the base of this cone is stitched on the normal level of the tricuspid annulus, including the septal region, resulting in central blood flow and restoring the septal leaflet function.

Hetzer et al¹³ published a new technique for tricuspid valve plasty, which places the valve mechanism at the level of the true AV annulus with the use of leaflets with greater mobility without plication of the atrialized RV chamber, thus excluding the leaflets with anomalous insertion, creating one or two valve orifices. This technique, which must be adapted to each case, requires a significant amount of mobile anterior leaflet, which limits its use to cases of more favorable anatomy.

Moreover, as the stitches are relatively deep, there was a higher frequency of complete atrioventricular block, requiring implantation of a pacemaker in 10.5% of cases in that series.

In the present series, there was no occurrence of atrioventricular block in the immediate postoperative period, similar to the work of Wu and Huang^{14,15}, who sutured a fresh autologous pericardial patch just below the right AV annulus to reconstruct the septal leaflet, emphasizing the importance of a superficial suture in this area.

Surgical times were quite variable, reflecting the broad spectrum of anatomical variations of Ebstein's anomaly and hence variations in the complexity of repair operations. The mean time of myocardial ischemia, 74 minutes, was higher than the average time of 38 minutes, from the initial series by Carpentier et al⁶ with 14 patients. However, using the current methods of myocardial preservation, this time is compatible with good cardiac performance after surgery.

Postoperative clinical outcome

The immediate clinical results, with an in-hospital mortality rate of 3.8%, short hospitalization duration and absence of valve replacement can be considered very good. The Mayo Clinic group¹⁶, which uses Danielson's technique, had a mortality rate of 5.8% in valve repair in children younger than 12 years, but the valve repair was only possible in 27% of 186 children, with the tricuspid valve being replaced by prosthesis in 62% of cases. The experience of the Carpentier group, representing the second largest series among the ones published, had a mortality rate of 9%¹⁷. The publication by Sarris et al¹⁸, reporting the collective results of 13 institutions associated with the European Congenital Heart Surgeons Association, showed an in-hospital mortality of 13.3%, although it included newborns, which are a higher-risk group.

Regarding the tricuspid valve replacement in Ebstein's anomaly, Kiziltan et al¹⁹ reviewed their series of 323 patients with tricuspid valve replacement performed in 48.9% of patients. In the long-term results, 97.5% of patients were free of bioprosthetic valve replacement after five years, and

Table 3 – Grade of tricuspid valve regurgitation: comparison of repeated measurements in 40 patients

Tricuspid regurgitation	Pre (n)	EPO (n)	LPO (n)
Grade 1	0	19	11
Grade 2	1	17	22
Grade 3	15	4	7
Grade 4	24	0	0

Comparison	Friedman's test p value
EPO - Pre	< 0.001
LPO - Pre	< 0.001
LPO - EPO	>0.05

Pre - preoperative, EPO – early postoperative, LPO- long-term postoperative.

80.6% after 10 years. There was no statistical difference for reoperation at 10 and 12 years between valve replacement and tricuspid valvuloplasty results, perhaps related to the large

size of implanted bioprosthesis in relation to the size of the patient, and right ventricular systolic pressure, generally low in patients after Ebstein's anomaly repair. However, it is a study

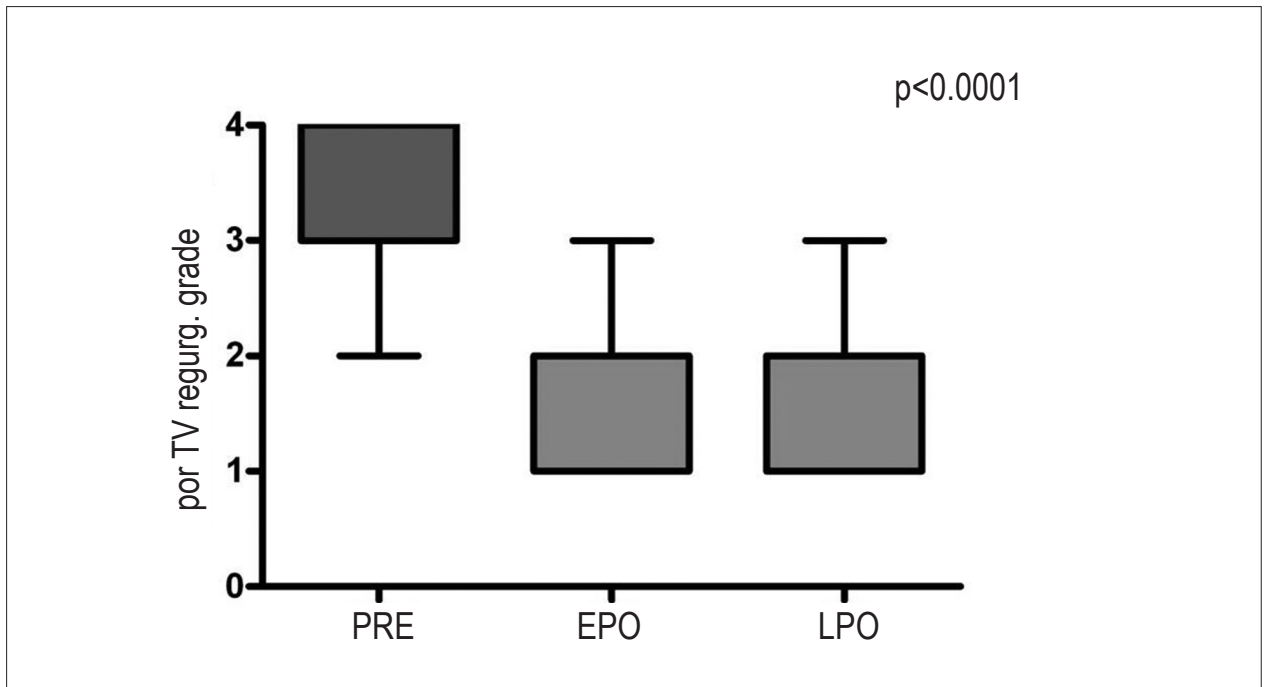


Figure 2 – Tricuspid valve regurgitation: Comparison between the pre-operative (PRE), early postoperative (EPO) and long-term postoperative (LPO) periods. Mean \pm 95% confidence interval. EPO – early postoperative, LPO – long-term postoperative, TV – tricuspid valve.

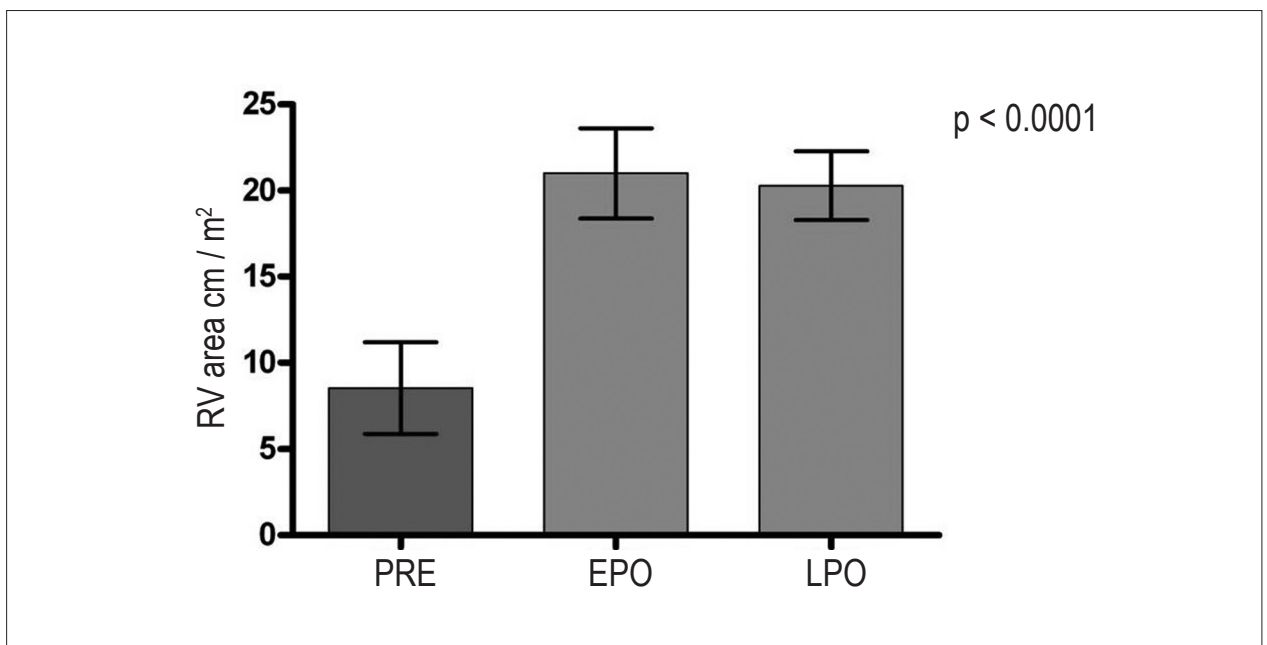


Figure 3 – Functional area of the right ventricle, indexed by body surface area in cm^2/m^2 : Comparison between measurements in the preoperative, early postoperative and long-term post-operative periods. Mean \pm 95% confidence interval. RV – right ventricle, EPO – early postoperative, LPO – long-term postoperative.

that compares the replacement with the monocusp technique of tricuspid valvuloplasty, for a limited period, disclosing a tendency toward the increase in replacements after 10 years, which does not reduce the importance of attempting efficient and durable tricuspid valvuloplasty, especially in children.

In general, the surgical technique for handling ASD in Ebstein's anomaly has not been considered by the authors and all of them performed only routine closing. In this series, valved ASD was always maintained to allow blood flow only from right to left. This proved to be useful in the immediate postoperative course of six patients with significant right ventricular dysfunction, who maintained an adequate cardiac output at the expense of a moderate drop in oxygen saturation, which increased as soon as right ventricular function improved. This strategy of right ventricular decompression made the cavopulmonary shunt unnecessary in our patients. In this sense, Chauvaud et al²⁰ reported better results with the use of bidirectional cavopulmonary anastomosis, together with Carpentier's operation, in patients with severe right ventricular dysfunction. These authors used this technique in 36% of patients in order to reduce right ventricular preload in cases of severe right ventricular dysfunction, thus obtaining a significant reduction in mortality caused by right ventricular failure. In our series, we had no need to employ cavopulmonary anastomosis.

The two series, however, had different characteristics as our patients were younger and had a lower cardiothoracic index than those in the series by Chauvaud et al²⁰.

The long-term clinical results showed a significant reduction in heart failure functional class, low incidence of arrhythmias and mortality. Few reoperations were necessary; the main cause was suture dehiscence in the septal region of the tricuspid valve.

Cardiothoracic ratio

Preoperatively, the mean cardiothoracic index of 0.66 was reduced to 0.54 in the late postoperative period, approaching the normal range. This index is important in the evaluation of Ebstein's anomaly, in which the left ventricle and atrium have normal or even below normal dimensions, with the increased cardiac silhouette determined by the dimensions of the right atrium and ventricle. Exceptions to this rule are cases that develop left ventricular cardiomyopathy. As in this series there was just one case of left ventricular dysfunction among the survivors, the reverse remodeling of the heart is related to the reverse remodeling of the right heart chambers.

Echocardiographic results related to valve function

The results related to the right AV valve anatomy and function were good and long-lasting. The operations described by Carpentier et al⁶ and Quaegebeur et al²¹ showed a considerable incidence of tricuspid regurgitation in the long term. In the study by Chen et al⁷ (Quaegebeur group), which evaluated 25 patients, 16 echocardiograms were obtained in the mid-term, disclosing moderate (18%) and significant (12%) regurgitation and two patients needed valve replacement. With the cone reconstruction, there was a small recurrence of regurgitation and a significant decrease in tricuspid valve regurgitation was maintained in the long term, with no need for valve replacement in any patient.

Tricuspid valve stenosis was observed in only one patient in the long term, which was discreet at the echocardiography and with no clinical consequences. The inclusion of the septal valve to increase the valve appears to be important in the prevention of stenosis, particularly

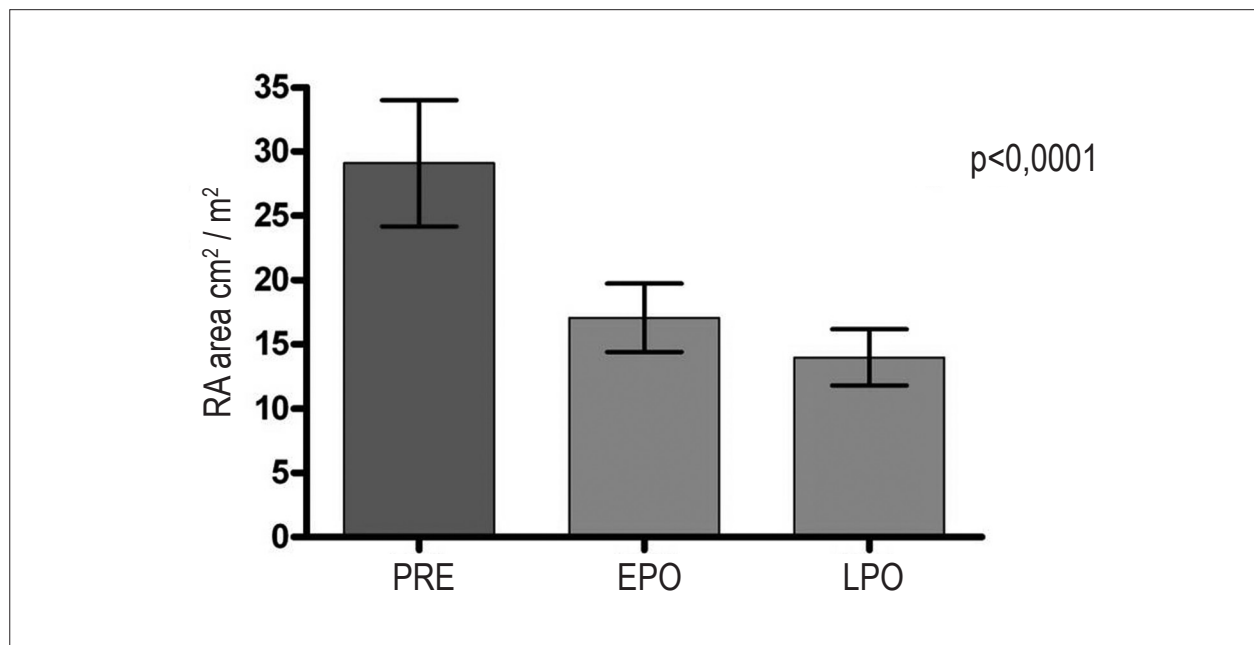


Figure 4 – Right atrium area indexed by body surface area in cm²/m²: Comparison of measurements in the preoperative, early postoperative and long-term postoperative periods. Mean \pm 95% confidence interval. RA - right atrium, EPO - early postoperative, LPO - long-term postoperative.

in adult patients. The rotation of the posterior leaflet tissue to be fitted to the hypoplastic septal leaflet, completing its proximal aspect, is a helpful maneuver to obtain a larger tricuspid valve. There was significant reduction in the annulus, as a result of the surgical intervention and long-term growth, which appears to be beneficial in children.

Not using the prosthetic annulus in the cone reconstruction technique of the tricuspid valve appears to be important in

children, in whom the permanent size limitation of the tricuspid annulus, already decreased, could result in stenosis in the long term. Moreover, the annulus flexibility allows the reduction in the tricuspid annulus during systole, contributing to the valve closing mechanism. The possibility of this annulus growth and flexibility, good long-term clinical outcome and few reoperations in this series of patients are arguments against the routine use of a prosthetic annulus.

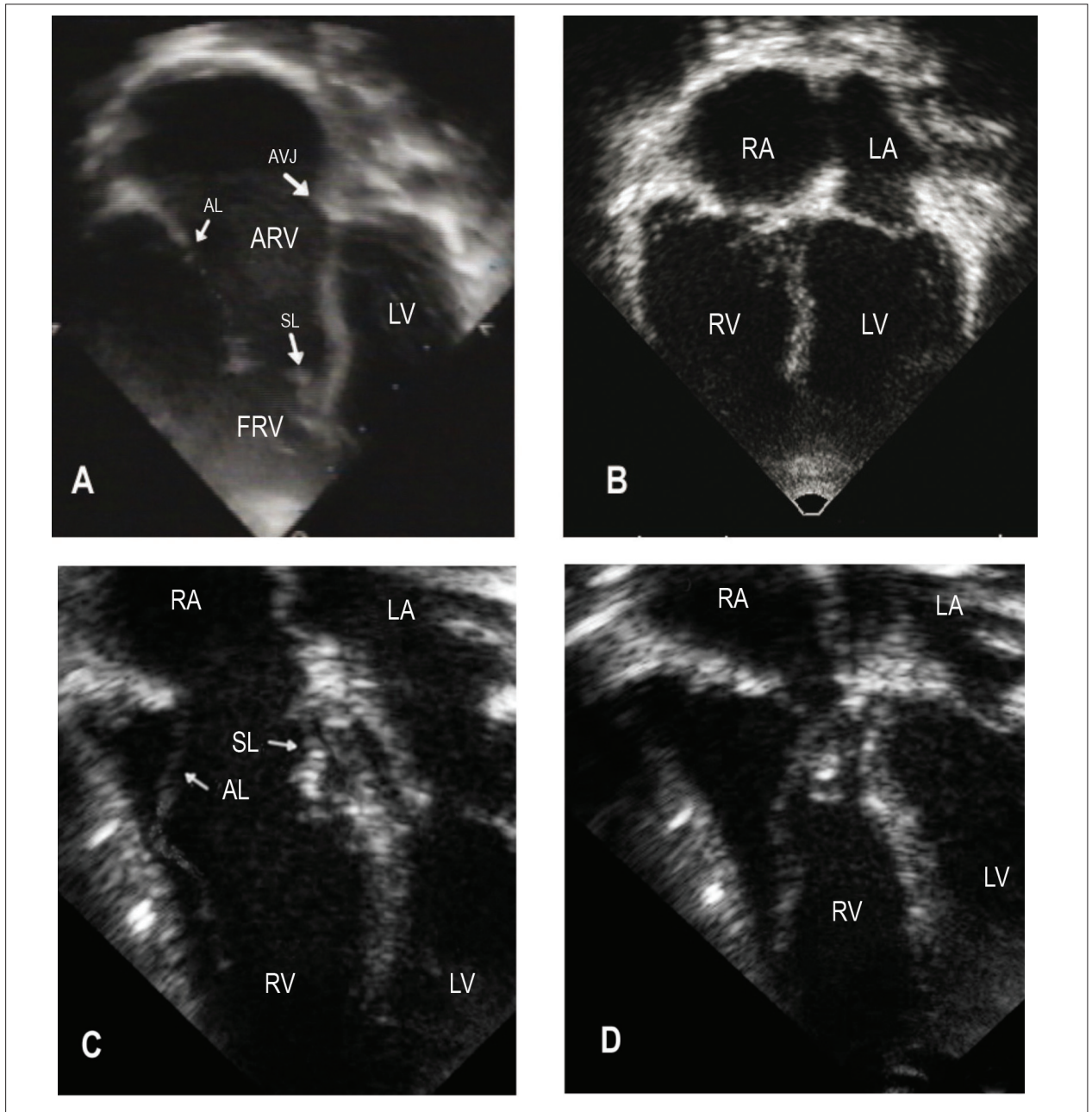


Figure 5 – Two-dimensional echocardiography showing morphology of small functional right ventricle preoperatively (A) was restored postoperatively with the repositioning of the tricuspid valve in the atrioventricular junction (B), the good opening of the tricuspid valve in diastole (C) and its closing and adequate leaflet coaptation during systole (D); RA - right atrium; LA - left atrium; AVJ - atrioventricular junction; FRV - functional right ventricle; ARV - atrialized right ventricle; RV - right ventricle; LV - left ventricle; AL - anterior leaflet of the tricuspid valve and SL - septal leaflet of the tricuspid valve.

Right ventricular restoration due to the longitudinal plication of the RV and the fitting of the tricuspid valve in its actual annulus was demonstrated by the considerable increase in the functional area of the RV, immediately after the surgery, after using the cone reconstruction technique. In this respect, this technique differs from that of Danielson¹, in which the transversal plication of the atrialized portion of the RV, leaving the AV valve in the same place where it exists in the RV, does not increase the size of the functional RV.

The two-dimensional echocardiography has limitations when assessing the right ventricular morphology and function. While the proximity of the mitral and aortic valves gives the LV its pyriform shape, the geometry of the RV is more complex, as the tricuspid valve is distant from the pulmonary valve. These morphological characteristics make it difficult to perform a geometric analysis of volumes and additionally, they do not allow the full view of the ventricle in a single echocardiographic projection²². In addition to the geometric complexity, there is greater difficulty in perfectly defining the endocardium along its entire length. Measurements of ejection fraction in the one-dimensional mode were not applicable due to the abnormal septal wall motion and geometry distortion of the ventricles. Eidem et al²³ suggest that the Tei index, or myocardial performance index, is useful as a quantitative measure of ventricular function in these patients. The three-dimensional echocardiography and tissue Doppler could help define the boundaries of the RV, but these techniques are not available in the initial studies of this series.

We chose areas, not volumes, due to previous studies that used these measurements to assess the severity of

Ebstein's anomaly, such as the Celermajor index²⁴, or the sum of the areas of the right atrium and atrialized right ventricle divided by the sum of the functional RV areas, LA and LV.

It is necessary to conduct further prospective studies with resonance imaging or computed tomography, which are more accurate methods to evaluate the morphology and size of the RV, in order to elucidate the function and reverse remodeling of this cavity.

Conclusions

The cone reconstruction technique to correct Ebstein's anomaly is feasible, with low in-hospital mortality and corrects tricuspid valve regurgitation without the need for valve replacement, which results in clinical improvement, right ventricular functional area restoration and reverse remodeling of the heart in most patients.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

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Study Association

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