Mid-term Outcome Of Surgical Treatment In Pediatric Patients With Ebstein's Anomaly: A Single-center Cohort Study.

Jia-chen Li
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Xian-chao Jiang
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Si-meng Zhang
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Jin-yang Liu
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Ya-juan Zhang
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Jun Yan
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Shou-jun Li
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital
Qiang Wang (wqcory@126.com)
Chinese Academy of Medical Sciences & Peking Union Medical College Fuwai Hospital

Research article

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Abstract

Background: Ebstein's anomaly is a malformation of the tricuspid valve and myopathy of the right ventricle. Surgery is now the main treatment for the defect. To summarize our surgical results and experience based on patients with Ebstein's anomaly who were under 7 years of age and treated with different surgical treatments.

Materials and Methods: From January 2010 to December 2019, 80 patients under 7 years old who were diagnosed of Ebstein's anomaly and underwent different surgical treatments were consecutively enrolled and followed up in detail.

Results: The median age of the 80 patients at the time of surgery was 3.63 years. Sixty-four (80.00%) patients underwent biventricular repair while 13 (16.25%) underwent 1.5-ventricle repair. With the median follow-up 27.50 months, the long-term survival of the total cohort, 1.5-ventricular repair and biventricular repair was 82.35%, 91.67% and 100%, respectively. The long-term freedom from reoperation rate was 97.50%, 92.31% and 98.44%, respectively. Mild, moderate and severe TR before surgery occurred in 6 (7.50%), 18 (22.50%) and 56 (70.00%), respectively. The early outcomes of 78 patients were 65 (83.33%), 11 (14.11%) and 2 (2.56%); the mid-term outcomes of 72 patients were 49 (68.06%), 19 (26.38%) and 4 (5.56%). Both early and long-term valve regurgitation were significantly decreased ($p< 0.001$) compared with preoperative condition. No more severe regurgitation occurred ($p=0.404$), though some early mild regurgitation became acceptable moderate regurgitation during long-term follow-up ($p=0.036$). Compared with Carpentier procedure, cone procedure had better long-term effect, while the effect of whole-valve technique needed more operation and long-term follow-up.

Conclusion: The reoperation rate and mid-term mortality of surgical treatment for Ebstein's anomaly were both low, tricuspid regurgitation was significantly improved during mid-term follow up. Cone procedure had the best mid-term effect among anatomic repair.

Introduction

Ebstein's anomaly (EA) is a malformation of the tricuspid valve (TV) and myopathy of the right ventricle (RV) [1]. The incidence is 1 per 200,000 live births and it accounts for 1% of congenital heart disease [2]. The core and critical distinguishing characteristic of EA from other congenital regurgitant lesions is the degree of apical displacement of the septal leaflet ($\geq$ 8 mm/m^2 body surface area) [3]. The pathological characteristics of each EA patient is different, which makes the surgical management complex and individualized, especially when it is treated with tricuspid valvuloplasty. A number of univentricular palliation and biventricular repair methods, including various modifications of techniques, have been described [4–5]. Studies on surgical outcomes in patients with EA often include a majority of adults; consequently, long-term outcomes of biventricular repair are generally good. However, there are few studies on neonates and young children with large sample sizes. We therefore conducted a retrospective study on patients with EA who were under 7 years of age and treated with different surgical treatments based on the recent 10-year data of our institution and summarized our surgical experience.

Materials And Methods

Patient selection and data collection

Eighty patients under 7 years old who were diagnosed with EA and underwent different surgical treatments were consecutively enrolled in our research from January 2010 to December 2019. The diagnosis mainly relied on echocardiography and transesophageal ultrasound and was confirmed during the intraoperative probe. The diagnostic criterion was apical displacement of the TV from the atrioventricular ring >0.8 cm/m^2 body surface area [6]. Children with other complex congenital heart diseases, including pulmonary atresia, tetralogy of Fallot and corrected transposition of the great arteries were excluded. Demographic data and clinical characteristics included sex, age, weight, body length, cardiac functional class, presence of cyanosis, Carpentier type, associated cardiac anomalies, the presence of preoperative arrhythmias, prior cardiac operations and preoperative degree of tricuspid regurgitation (TR). Surgical procedures and postoperative parameters, including cardiopulmonary bypass (CPB) time, aortic cross clamp time, mechanical ventilation time, intensive care unit (ICU) stay, complications, mortality, cause of death, and TV and RV function, when available, were also comprehensively evaluated. Patients' records and postoperative follow-up were conducted through our outpatient service and over the telephone.

Surgical methods standard

In general, surgical procedures can be divided into three categories, biventricular repair, 1.5-ventricle repair or univentricular palliation. The decision for the type of repair was based on the ratio of the atrialized right ventricle (aRV) to the functional right ventricle (fRV), the size of the TV annulus and the size of the pulmonary artery in addition to the severity of TR. Biventricular repair was performed in patients with a retained RV function and an acceptable anatomy and fRV in the absence of severe pulmonary valve stenosis or atresia. Due to the lack of preoperative quantitative evaluation, most of the decisions for 1.5-ventricle repair were made by the surgeon during the operation. In patients with extensive RV failure, severe TR and/or the presence of other relevant cardiac comorbidities, e.g., PA, a univentricular palliation was the intended surgical strategy. All operations were analyzed as separate events.

Evaluation standard

There are some evaluation criteria that can help us understand the preoperative condition and postoperative recovery of patients. Cyanosis was determined when the increasing level of deoxyhemoglobin in the blood resulted in a purplish appearance of the skin mucous membrane. The severity of EA was evaluated by echocardiography and transesophageal echocardiography and determined by the internationally recognized Carpentier type. Early outcome mainly referred to death or reoperation within 30 days after surgery or before hospital discharge. Mid-term outcome mainly referred to death or reoperation within one to five years after surgery.

Statistical analysis

Descriptive statistics for the 80 patients are reported. Data are presented as means ± standard deviations (SDs) for continuous data with a normal distribution, as medians (25th percentile, 75th percentile) for continuous data with a nonnormal distribution, or as numbers and percentages for categorical variables, as appropriate. factors for early mortality. Freedom from reoperation and cumulative survival rates were analyzed according to the Kaplan-Meier test. There was insufficient incidence of deaths and reoperations to perform an analysis to identify risk. Differences in the frequency of TR at preoperative, early and mid-term follow-up examinations were performed using the chi-square test and Fisher test. Statistical significance was defined as $P<0.05$ with two-tailed distributions. All statistical analyses were performed with SPSS version 22.0 software (SPSS, Inc., Chicago, IL, USA) and GraphPad Prism version 5.0 software (GraphPad Software, Inc., La Jolla, CA, USA).

Results

Patient profiles

The demographic data and clinical characteristics of the patients are summarized in **Table 1**.

Surgical procedures and postoperative data

The surgical procedures performed and postoperative data are presented in **Table 2**. Of the 80 patients, two (2.50%) patients underwent the Glenn procedure because of tricuspid orifice stenosis and right ventricle dysplasia. Tricuspid valve replacement (TVR, $29^{\#}$, Medtronic, Bio-valve) was performed for one (1.25%) patient because of severe tricuspid valve dysplasia. Two (2.50%) patients underwent unexpected reoperation, and 1 patient underwent rerepair because of recurrent TR on the 6^{th} day postoperatively; another patient received the Glenn procedure because of acute right heart failure on the 4^{th} day postoperatively. The early mortality was 1.25%, and 1 patient's venous pressure increased and blood pressure decreased after biventricular repair. The Glenn procedure and extracorporeal membrane oxygenation were then performed, but the patient consistently could not maintain spontaneous breathing after surgery because of poor right heart function and passed away on the 38th day after the operation.

Surgical technique

The detailed surgical strategies are shown in **Table 3**. Regular methods of EA surgical correction included the Danielson procedure, Devage procedure, Carpentier procedure, cone procedure, and whole-valve technique. The main procedures applied in our center were as follows. 1) Danielson procedure: lateral aRV plication, and the lobes were raised to the level of the normal valve ring. 2) Devage procedure: plication with 1 or 2 mattress sutures from the junction of the anterior and posterior lobe to the junction of the septal and posterior lobe, whereby the apical aspects of the septal and anterior leaflets would effectively create a bicuspid valve. 3) Carpentier procedure: most of the anterior valve and adjacent part of posterior valve were detached and sutured at the level of the normal tricuspid valve ring, longitudinal aRV plication, such that the TV became a single valve structure dominated by the anterior valve. 4) Cone procedure: all three lobes were cut off at the root of the lobe, the adhesion of the subvalve tissue was released, and the three lobes were then structured to be a cone-like body, clockwise rotated and sutured into the normal valve ring. 5) Whole-valve technique: an optimized and improved technique based on the cone procedure in our center. In the case of tricuspid dysplasia, the free margin of each lobe is actively widened with fresh autologous pericardium to ensure sufficient alignment between the lobes. Among the 13 patients, 4 received the intended bidirectional cavopulmonary shunt (BCPS) as a preoperative plan, the other 9 received unplanned BCPS because of poor RV function during the operation. Other modified techniques based on regular methods, including right ventricle plication, TV annuloplasty plication and autologous leaflet augmentation, prosthetic ring or artificial chordae, were not used in the enrolled patients. ASD subtotal closure was also considered in the research.

Follow-up and mid-term outcome

The median follow-up of the 80 patients was 27.50 (10.25, 56.75) months, with 3 patients loss to follow-up after discharge, and the effective follow-up rate was 96.25%. One early death occurred after 1.5 ventricular repair on the 38th day after the operation, as mentioned above. One late death occurred in the 6th year after the Glenn procedure because of RV dysplasia. As shown by the survival curve in **Figure 1a**, the overall survival was 98.71% at 4 years (50 months) and 82.35% at 8 years (100 months). The survival of patients with 1.5-ventricular repair was 91.67% at 4 years and at 8 years, and no patients who received biventricular repair have died to date. The freedom from reoperation curve is shown in **Figure 1b**. The long-term freedom from reoperation rate of overall, 1.5-ventricular repair and biventricular repair was 97.50%, 92.31% and 98.44%, respectively. Except for 2 patients who received tricuspid valve rerepair and the Glenn procedure on the 6th day and the 4th day after surgery, no patients received reoperation to date. Of the 6 cases with arrhythmia in the early postoperative period, 1 patient had atrioventricular block of the second degree, 3 had untreated transient ventricular tachycardia, and 2 improved spontaneously after discharge. Other clinically important complications, including right ventricular outflow tract obstruction, left ventricular outflow tract obstruction or TV stenosis, were not observed either before or after surgical repair.

Preoperative, predismissal, and mid-term findings observed for TR are summarized in **Figure 2a**. Of the 80 patients, mild, moderate and severe TR before surgery occurred in 6 (7.50%), 18 (22.50%) and 56 (70.00%), respectively. Of these, the early outcomes of 78 patients (postoperative echocardiography for 2 patients' could not be found because of systematic failure) were 65 (83.33%), 11 (14.11%) and 2 (2.56%), and the mid-term outcomes of 72 patients (3 patients were lost to follow-up after discharge; 3 patients received no reexamination after discharge) were 49 (68.06%), 19 (26.38%) and 4 (5.56%), respectively. When comparing preoperative (pre-op) with dismissal, preoperative with mid-term, and dismissal with mid-term echocardiograms, we divided the cases into two groups (mild group vs moderate-severe group) and found that both early and long-term degrees of TR were significantly decreased (**Table 4**, p< 0.001). Compared with the early outcome, the long-term TR degree increased (**Table 4**, p=0.036) between the two groups. However, when separately comparing the mild group with the severe group, we found that no severe TR occurred (**Table 5**, p=0.404). It could be speculated that some early mild TR cases had become cases of acceptable moderate TR. In the 63 patients with anatomic repair, 8 cases underwent whole-valve technique, 24 cases underwent cone procedure and 31 cases underwent Carpentier procedure. Preoperative, predismissal, and mid-term findings observed for TR with whole-valve technique are summarized in **Figure 2b.** The preoperative number (%) of mild, moderate and severe tricuspid regurgitation was 0, 2 (25.00%) and 6 (75.00%), respectively. The early outcome was 7 (87.50%), 1 (12.50%) and 0. The mid-term outcome was 7 (87.50%), 0 and 1 (12.50%). As the modified technique was carried out in recent few years with insufficient cases and follow-up time, the effect of whole-valve surgery needed further follow-up and observation. Preoperative, predismissal, and mid-term findings observed for TR with cone and Carpentier procedure are summarized in **Figure 2c** and **Figure 2d**, respectively. The preoperative number (%) of mild, moderate and severe tricuspid regurgitation in cone procedure was 4 (16.67%), 3 (12.50%) and 17 (70.83%), respectively. The early outcome was 23 (95.83%), 1 (4.17%) and 0. The mid-term outcome was 22 (95.65%), 1 (4.35%) and 0. The preoperative number (%) of mild, moderate and severe tricuspid regurgitation in Carpentier procedure was 4 (12.90%), 7 (22.58%) and 20 (64.52%), respectively. The early outcome was 30 (96.77%), 1 (3.23%) and 0. The mid-term outcome was 21 (80.77%), 2 (7.69%) and 3 (11.54%). By evaluating long-term survival (100% vs 97.06%, p=0.58), freedom from reoperation (100% vs 97.06%, p=0.58) and TR, the long-term effect of cone surgery was significantly better than that of Carpentier surgery.

Discussion

EA's anatomical features [2] include 1) displaced and dysplastic septal and posterior leaflets; 2) an elongated, fenestrated, or dysplastic anterior leaflet; 3) a markedly dilated atrialized right ventricle that becomes thin and dyskinetic; and 4) a displaced functional annulus and a markedly expanded anatomic annulus. Many procedures have been reported based on this anatomical theory. In general, surgical methods can be divided into three categories: biventricular repair, 1.5 ventricle repair and univentricular palliation. In this research, different surgical methods and relative long-term prognosis of children with EA in our institution were systematically summarized for the first time. It was a large-sample sized cohort study for Chinese EA children, and it was of great significance to understand the status quo of diagnosis and treatment for such disease in China.

Outcome summary

Among the patients diagnosed with EA under 7 years old in our study, 6 (7.50%) were younger than 1 year old, and 19 (23.75%) were between 1 and 2 years old. Twelve (15.00%) cases were serious Carpentier type III-IV. There were 64 biventricular repairs, 13 1.5-ventricle repairs, 2 univentricular palliations and 1 TVR. Compared to previously reported studies, the proportion of biventricular repair was high but the proportion of 1.5-ventricle repair and univentricular palliation rather low in our center. To date, we have not performed the Starnes procedure for neonates who must undergo emergency surgery. During the last 10 years, our concept of biventricular repair has constantly been updated. The Carpentier procedure was the conventional method in the early stage, and the cone and its modified methods gradually dominated in the late stage. The overall outcome of the operations was worthy of recognition. The rates of long-term mortality and reoperation were both very low. TR was significantly improved, and the proportion of severe regurgitation decreased from 70% before surgery to 5.56% during long-term follow-up. Serious long-term complications did not occur except for 6 cases of untreated arrhythmia.

Indications for surgery

Traditional indications for surgery included symptoms of cyanosis, fatigue, paradoxical embolism, decreasing exercise performance (preferably documented by exercise testing), progressive RV enlargement, progressive RV dilation or reduction of systolic function by echocardiography and/or appearance of atrial or ventricular arrhythmias [7]. The timing of surgery for EA continued to be controversial, particularly when confronted with an asymptomatic patient with normal exercise tolerance. In fact, the vast majority of patients with EA require surgery in their lifetime. Dearani indicated [7] that the operation should occur in early childhood as long as the surgical team can provide a reproducible and predictable satisfactory tricuspid repair. Their repair rate in children approached 100%, and the risk of operation was <1%, as reported in a large sample-size retrospective study [8]. Similar results could be drawn in our research. The overall survival was 98.71% at 4 years and 82.35% at 8 years, and none of the patients who received biventricular repair have died to date. Among these patients, only 22.50% presented with symptoms of cyanosis. Therefore, we may delay the timing of the operation based only on symptoms.

Experience of surgical intervention

The original method from the 1960s was to uplift the descending valve, including the Hardy technique and Danielson technique [9]. In the 1990s, the Carpentier technique [10] and subsequent Quaegebeur technique [11] incorporated surgical delamination of the anterior leaflet with annular reattachment to optimize monocusp mobility for ventricular septal coaptation. Since 2007, the cone reconstruction [12] technique has become one of the most widely recognized surgical procedures for EA. It is an extension of the previous technique in that septal and inferior leaflet tissues were mobilized, after which the sides of all mobilized leaflets were connected in a manner to create 360 degrees of leaflet tissue. Near-anatomic repair could be achieved when the newly constructed “cone” was reattached to the true annulus. After that, many other studies have reported their modified cone technique and achieved good results, including ringed annuloplasty, closure of leaflet fenestration, Sebening stitch, and artificial chordae among others [8]. According to our research, the long-term effect of cone surgery was significantly better than that of Carpentier surgery. However, during our clinical treatment and subsequent re-examination, we found that with the cone technique, some patients still showed different degrees of TR and RV enlargement at the later stage, even though the initial postoperative effect was good, especially for those with severe tricuspid dysplasia. From January 2018, we began our modified method named the “whole-valve technique”. Our experiences are as follows: 1) all the clingy leaflets of the tricuspid valve sufficiently dissociated from the right ventricular wall to guarantee effective closure of the detached part of the anterior and posterior leaflets; 2) in the cases of tricuspid valve dysplasia, fresh pericardial tablets were actively used for splicing the detached part of each leaflet; and 3) new leaflets were sutured to the plication of the true tricuspid annulus. To date, a total of 8 cases of this technique have been performed, and the 2-year follow-up was satisfactory, including 2 Carpentier type IV cases. No additional Glenn procedure has been performed, and more cases and longer follow-up times will receive further attention. In addition, the modified Devage procedure, also named the “play it where it lies” technique [13], was performed in 2 patients, both of whom underwent the intended Glenn procedure and had moderate TR at the latest reexamination. In our opinion, with regard to the choice of surgical procedure, the “play it where it lies” technique should be chosen with caution. Due to the small number of cases, we were unable to summarize the effective TVR experience and indications for young children.

When should we perform BCPS?

BCPS is applied selectively in patients to unload RV volume in the setting of severe RV dysfunction. Studies have shown that BDPS decreases the volume of the enlarged, dysfunctional right ventricle by 35–45% (depending on the patient’s age), providing substantial preload to the underfilled left ventricle and reducing the hemodynamic stress on a complex TV repair [7]. A multicenter retrospective study [13] from the Netherlands reported that between 1980 and 2013, 63 children (0-112 months) underwent 109 operations. Two of 37 biventricular repair patients received 1.5-ventricle repair at 11 and 105 months after surgery. Five 1.5-ventricle repair patients received BCPS in the first surgery. The other 21 patients underwent univentricular palliation surgery due to extensive RV failure, severe TR or the presence of significant other cardiac abnormalities. In terms of this study, 7 1.5-ventricle repair cases were performed in 109 operations (6.4%), and the rate of 1.5-ventricle repair was rather low compared with our research. In our center, the rate of 1.5-ventricular repair was approximately 16.9%, which was higher than that reported in the Netherlands, yet it was significantly lower compared with the 30% at the Mayo Clinic, USA [8] and the more than 50% at Shanghai Children’s Medical Center, China [15]. The study from Shanghai Children’s Medical Center also suggested that the BCPS procedure should be presented when a preoperatively severely dilated RV (aRV>50%) with the anterior leaflet causes significant RV outflow tract obstruction, outstanding TV morphologic abnormity, and hemodynamic instability after separation from cardiopulmonary bypass, regardless of the presence of RV failure or severe cyanosis. According to Dr Dearani’s latest study [1], BCPS is generally feasible, as pulmonary hypertension is rare in EA, but should be used with caution because it may increase left ventricular (LV) end-diastolic pressure in the setting of moderate to severe LV dysfunction. Indications for BCPS in adults include both heart failure indications (RV end-diastolic volume > 250 mL/m, RV ejection fraction < 25%, D-shaped left ventricle, postrepair right atrial pressure to left atrial pressure ratio > 1.5:1, and postrepair cardiac output depression) and no indications of heart failure (complex TV repair tension reduction and postrepair TV stenosis with a mean gradient > 8–10 mmHg). BCPS contraindications related to pulmonary hypertension include a mean pulmonary artery pressure > 20 mmHg, pulmonary arteriolar resistance > 4 Woods units, and elevated left heart pressure (LV end diastolic pressure or left atrial pressure > 12 mmHg). In our opinion, BCPS should be actively prepared when the tricuspid valve orifice is smaller than normal or the proportion of aRV is larger than that of fRV. In addition, ASD subtotal closure is a good method to unload RV. Although saturation was sacrificed to some extent, the long-term prognosis of the TV and RV was not affected according to our follow-up.

Study limitations

This study is limited by its retrospective nature. The comparison between cone procedure and Carpentier procedure could be biased because of the age of operations. More arguments of RV function should be provided to assess the patient's preoperative condition and postoperative recovery. Due to the low number of deaths and reoperations, reliable analysis to identify risk factors could not be performed.

Conclusion

The reoperation rate and mid-term mortality of surgical treatment for Ebstein's anomaly were both low, tricuspid regurgitation was significantly improved during mid-term follow up. Cone procedure had the best mid-term effect among anatomic repair.

Abbreviations

EA = Ebstein's anomaly, TV = tricuspid valve, RV = right ventricle, TR = tricuspid regurgitation, CPB = cardiopulmonary bypass, ICU = intensive care unit, aRV = atrialized right ventricle, fRV = functional right ventricle, SDs = standard deviations, BCPS = bidirectional cavopulmonary shunt.

Declarations

Ethics approval and consent to participate

The study protocol was approved by the Institutional Review Board of Fuwai Hospital. The requirement to obtain informed consent was waived because of the retrospective nature of the study.

Consent for publication

Consent was obtained from the patients or their relatives.

Availability of data and materials

The datasets used or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors have declared that no interest.

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Authors' contributions

(I) Conception and design: JC Li, Qiang Wang; (II) Administrative support: J Yan, SJ Li; (III) Provision of study materials or patients: J Yan, SJ Li, Q Wang; (IV) Collection and assembly of data: JC Li, XC Jiang, SM Zhang; (V) Data analysis and interpretation: JY Liu, YJ Zhang; (VI) Manuscript writing: All authors; (VII) Final approval of manuscript: All authors.

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Tables

Table 1
Demographic data and clinical characteristics.

Variables	Total (N = 80)
[a] Male	44 (55.0%)
[b] Median age in year (min; max)	3.63 (1.80, 6.00)
[a] Age1 year	4 (5.00%)
[c] Wight (kg)	16.09 ± 6.33
[c] Body length (cm)	99.14 ± 18.34
[a] Cyanosis	18 (22.50%)
[a] Carpentier type III-IV	12 (15.0%)
[a] Associated anomaly	
Atrial septal defect / Patent foramen ovale	63 (78.75%)
Ventricular septal defect	9 (11.25%)
Pulmonary stenosis	4 (5.00%)
Pulmonary arterial hypertension	2 (2.50%)
Patent ductus arteriosus	4 (5.00%)
*Others	5 (6.25%)
[a] Arrhythmia	4 (5.00%)
[a] Previous ablation	1 (1.25%)
[a] Prior cardiac operation	0 (0)
[a] Severity of tricuspid regurgitation	
Mild	6 (7.50%)
Moderate	18 (22.50%)
Severe	56 (70.00%)

Values are n (%) a or median (25%, 75%) b or mean ± SD c. *Others included 1 coronary left ventricular fistula, 1 persistent left superior vena cava, 1 right ventricular dysplasia syndrome, 1 aortopulmonary collateral vessels and 1 moderate mitral regurgitation. (SDs = standard deviations)

Table 2
Surgical procedures and postoperative data.

Variables	Total (N = 80)
[c] Cardiopulmonary bypass time, min	114.96 ± 42.41
[c] Aortic cross clamp time, min	75.34 ± 25.41
[a] Surgical strategy	
Biventricular repair	64 (83.75%)
1.5-ventricle repair	13 (16.25%)
Univentricular palliation	2 (2.50%)
Tricuspid valve replacement	1 (1.25%)
[a] Combined surgery	
Right ventricle plication	68 (85.00%)
Atrial septal defect / Patent foramen ovale closure	44 (55.00%)
Antiarrhythmic procedure	3 (3.75%)
*Others	20 (25.00%)
[b] Mechanical ventilation time, hour	11.00 (6.26, 20.75)
[b] ICU stay, day	2.00 (1.00, 3.00)
[b] Postoperative length of stay, day	8.00 (7.00, 11.00)
[a] Postoperative complication	
Postoperative arrhythmia	6 (7.5%)
Unexpected reoperation	2 (2.50%)
Recurrent tricuspid regurgitation	1 (1.25%)
Acute right heart failure	1 (1.25%)
Delayed sternal closure	1 (1.25%)
Extracorporeal membrane oxygenation	1 (1.25%)
Peritoneal dialysis	0 (0)

Values are n (%) a or median (25%, 75%) b or mean ± SD c. *Others included 9 VSD repair, 2 pulmonary valvuloplasty, 1 pulmonary angioplasty, 4 PDA ligation, 1 coronary left ventricular fistula repair, 1 right ventricular outflow tract dredging, 1 aortopulmonary collateral vessels embolization and 1 mitral valvuloplasty. (SDs = standard deviations)

Variables	Total (N = 80)
Massive pleural effusion	1 (1.25%)
[a] Early mortality	1 (1.25%)
Values are n (%) a or median (25%, 75%) b or mean ± SD c. *Others included 9 VSD repair, 2 pulmonary valvuloplasty, 1 pulmonary angioplasty, 4 PDA ligation, 1 coronary left ventricular fistula repair, 1 right ventricular outflow tract dredging, 1 aortopulmonary collateral vessels embolization and 1 mitral valvuloplasty. (SDs = standard deviations)	

Table 3

Detailed surgical strategy.

Added / Modified procedures Repair Strategy (N = 77)	Right ventricle plication (N = 71)	ASD subtotal closure (N = 18)	TV annuloplasty plication (N = 59)	Autologous leaflet augmentation (N = 40)
Biventricular repair (N = 64)				
Danielson (N = 1)	1	0	1	0
Carpentier (N = 31)	28	5	26	13
Cone (N = 24)	22	5	18	13
Whole valve technique (N = 8)	8	2	4	8
1.5-ventricle repair (N = 13)				
Danielson (N = 1)	1	0	1	0
Devage (N = 2)	2	1	1	0
Carpentier (N = 5)	5	2	5	4
Cone (N = 5)	4	3	3	2
Detailed items included biventricular repair and 1.5-ventricle repair, right ventricle plication, BCPS, ASD subtotal closure, TV annuloplasty plication and autologous leaflet augmentation. (BCPS = Bidirectional cavo-pulmonary shunt, ASD = atrial septal defect, TV = tricuspid valve)				

Table 4

Comparison of mild and moderate-severe tricuspid regurgitation between preoperative, early and mid-term echocardiograms.

Variables		Preoperative (N = 80)	Early outcome (N = 78)	Mid-term outcome (N = 72)
Tricuspid regurgitation	**Mild**	6	65	49
	Moderate-severe	74	13	23
P	**Compared with preoperative**	-	<0.001	<0.001
	Compared with early outcome	-	-	0.036

Table 5

Comparison of mild and severe tricuspid regurgitation between early and mid-term echocardiograms.

Variables		Early outcome (N = 67)	Mid-term outcome (N = 53)	Total
Tricuspid regurgitation	**Mild**	65	49	114
	Severe	2	4	6
p				0.404

Figures

Figure 1a

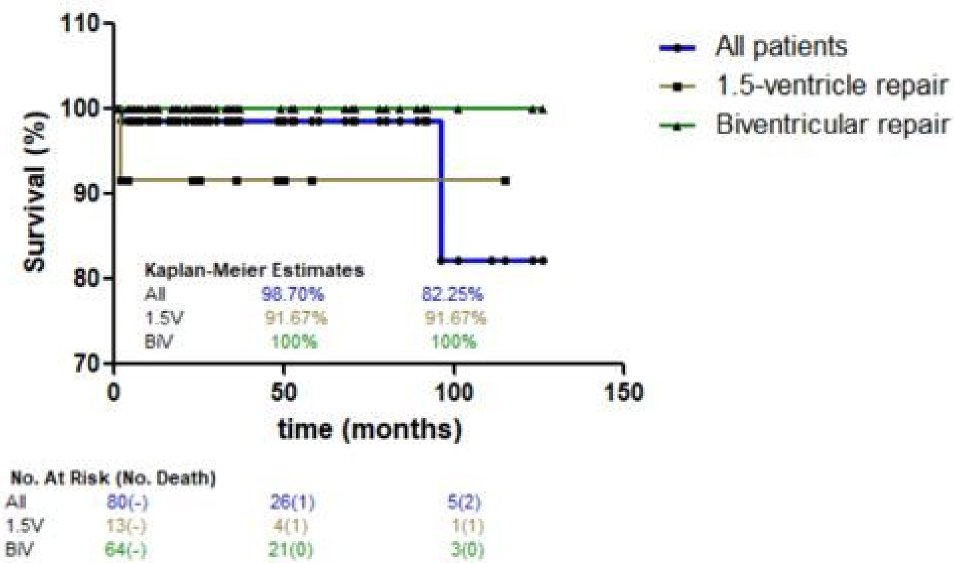

Figure 1b

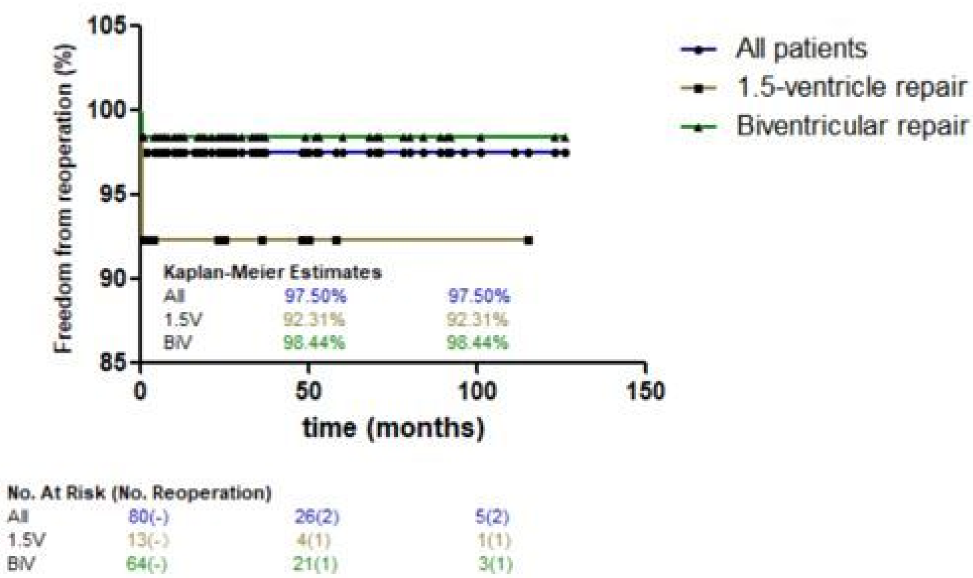

Figure 1

Mid-term survival and freedom from reoperation of all patients (blue line), patients underwent 1.5 ventricular repair (yellow line) and patients underwent biventricular repair (green line).

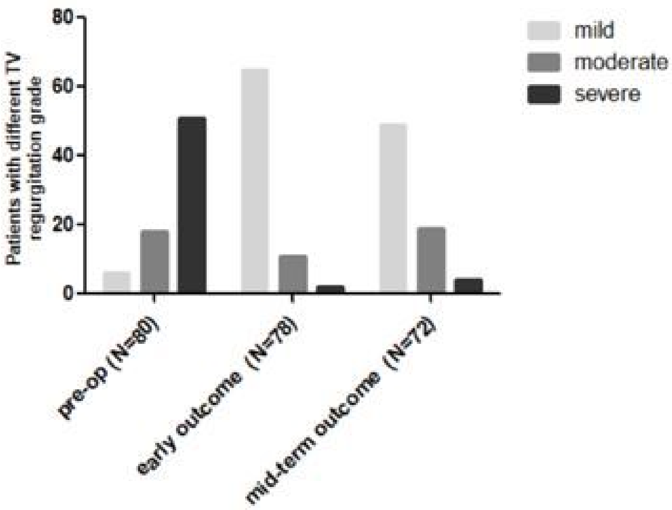

Figure 2a

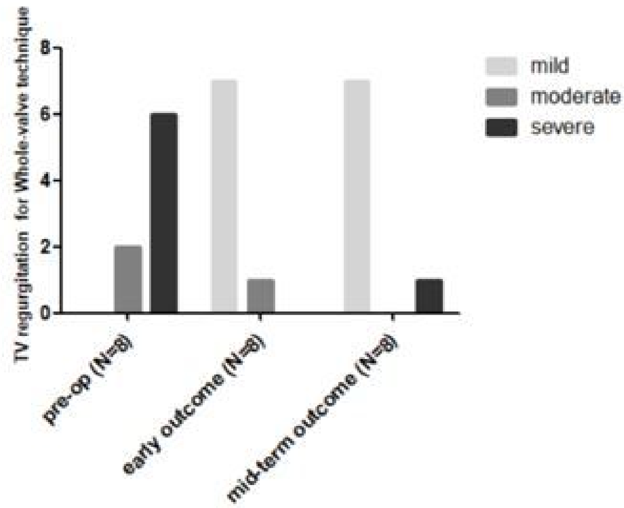

Figure 2b

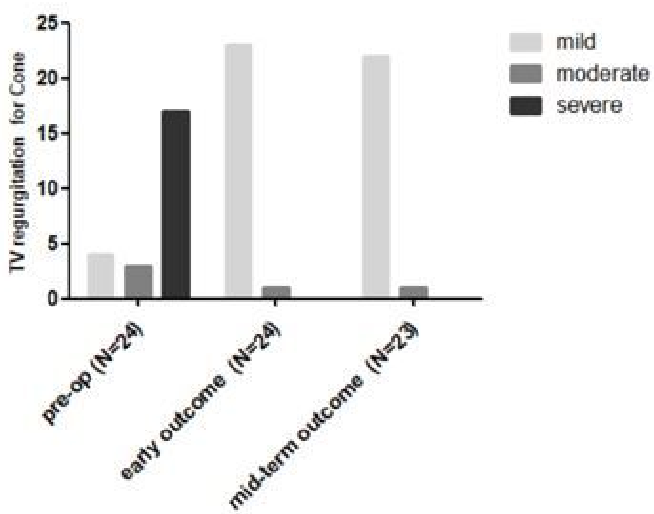

Figure 2c

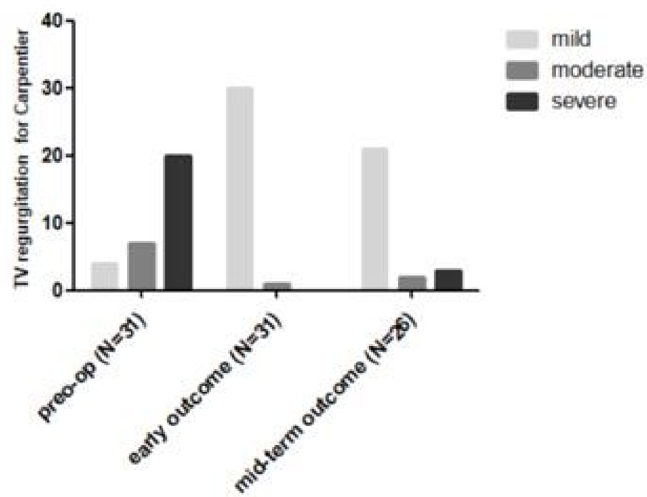

Figure 2d

Figure 2

Paired comparisons of echocardiographic changes after surgical repair. 2a. Paired comparisons after surgical repair. 2b. Paired comparisons after Whole-valve technique repair. 2c. Paired comparisons after Cone repair. 2d. Paired comparisons after Carpentier repair. (pre-op=preoperative result, TV=tricuspid valve)