



Mid-term outcomes of individualized surgeries in patients with Ebstein's anomaly

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Abstract

The variable anatomy of Ebstein's anomaly leads to its various surgical procedures. The long-term outcomes of different operations were not well established. Thirty-five patients with Ebstein's anomaly who underwent operations from 2006 to 2018 in our department were retrospectively reviewed. Individualized surgical plans were performed according to the preoperative echocardiography and surgeons' preference. Tricuspid repair, either Danielson's or Carpentier's technique, was the primary choice in patients who had sufficient tricuspid leaflets and adequate right ventricle, while tricuspid replacement was used when a reliable repair is not achievable. Additional bidirectional cavopulmonary shunt was performed in those who had unstable hemodynamics despite of high central venous pressure after separation from cardiopulmonary bypass. The perioperative and follow-up data were collected. The age was 26.9 (0.6–54) years [16 children (age < 14, and 19 adults (age ≥ 14)]. Preoperative tricuspid regurgitation was severe in 30, moderate in 4, and mild in the remaining 1 patient. Preoperative cardiac-associated malformations include 20 atrial septal defects, 2 ventricular septal defects, 2 pulmonary stenosis, and 1 sub aortic ridge, and these were operated simultaneously. Among all the surgical patients, 2 needed additional reoperation during the same admission, and ultimately, 29 patients had biventricular repair, including 21 tricuspid repair and 8 replacements. The other 6 patients had cavopulmonary connection and achieved 1.5 ventricular repair (3 tricuspid repair and 3 replacements). In all the 24 tricuspid repair patients, Danielson's procedure was used in 17, while Carpentier's technique was used in the other 7 patients. The average cardiopulmonary bypass time was 90 ± 28 min and cross-clamp time was 48 ± 24 min. There were 2 perioperative deaths (5.7%) and no third-degree atrioventricular block. The postoperative in hospital stay was 13.7 ± 9.6 days. In the 33 survivors who were followed up at a median of 29.2 months, 6 patients had severe tricuspid regurgitation, and 2 of them underwent tricuspid replacement. The 5-year freedom from severe tricuspid dysfunction or reoperation was 78.5%, and no difference was found between children and adults, neither between different surgical choices. The surgeries of Ebstein's anomaly were variable, and individualized operation achieved reasonable short- and mid-term results. However, severe tricuspid regurgitation during the follow-up was not neglectable, and reoperation in such cases also achieved good outcomes. New repair strategy such as cone repair may be considered.

Keywords Ebstein's anomaly · Surgery · Congenital heart disease

Background

Ebstein's anomaly (EA) is a rare form of congenital heart disease, characterized with deformation of tricuspid valve, varied degrees of right-ventricular hypotrophy and potentially associated anomaly such as atrial septal defect et al.

Depending on the different morphologies of tricuspid valve and right ventricle, patients with Ebstein's anomaly could be lethal with severe symptoms in neonates or asymptomatic in 70 years old. Surgery is offered to those who had decreased excise tolerance with moderate-to-severe tricuspid regurgitation or cyanosis. Various operations were developed to treat this complex cardiac defect.

Pioneer surgeons from all over the world tried to repair this anomaly with different techniques. Typical repair included three techniques. First, Danielson et al. at Mayo Clinic plicated atrialized right ventricle horizontally, repaired of tricuspid valve in the presence of a mobile and

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well-formed anterior leaflet by posterior annuloplasty, and fixed the other associated defects [1–3]. Second, Carpentier et al. from France used a different procedure. The anterior and posterior leaflets are detached and mobilized, and the atrialized right ventricle is plicated vertically, then the detached leaflets are sewed to the anatomic tricuspid annulus, and this technique greatly extends the possibility of repair [4, 5]. The third and most recent technique was developed by Brazilian surgeon Da Silva [6–8]. He mobilized almost all the tricuspid leaflets, and rotated the leaflets clockwise, and then reattached them to the anatomic annulus, plus minus reinforcement with a ring as necessary, so the hinge point of the new valve was at the anatomic tricuspid annulus. When acceptable repair results could not be achieved, tricuspid replacement should be performed. Since Ebstein's anomaly is a very rare cardiac defect, any data about this disease may add new information and help the decision-making of surgical choice in the future. Here, we summarized our mid-term surgical outcomes of this disease.

Patients and methods

This retrospective study was approved by our institutional research ethics board, and informed consent was received. Patients who admitted into cardiothoracic surgery department of our hospital with diagnosis of EA from 2006 to 2018 were retrospectively reviewed. In total, there are 41 patients, and 6 of them did not receive surgery (contra-indications and other reasons), while the other 35 patients underwent operations with various techniques and were included in this study.

Individualized surgical plans were performed according to the preoperative echocardiography and surgeons' preference. The Great Ormond Street Echocardiogram (GOSE) Score was calculated according to the preoperative echocardiograms. This score is defined as the ratio of area of the right atrium and atrialized right ventricle to the combined area of the functional right ventricle, left atrium, and left ventricle [9]. Usually, if there were sufficient tricuspid leaflets and not severely hypoplastic right ventricle, a biventricular tricuspid valve repair was preferred. If the mobilized tricuspid tissue is enough to generate a competent valve with anticipated tricuspid annulus Z score more than -2 and the expected new right-ventricular volume is reasonable, a biventricular repair was preferred. If the tricuspid leaflets were not sufficient and right ventricle was small, tricuspid replacement was the choice. Whether an additional bidirectional Glenn procedure was performed depends on the hemodynamic stability after weaning off the cardiopulmonary bypass. Central venous pressure higher than 20 mmHg when all the other settings were optimized strongly suggested the need of additional Glenn procedure.

In those who required tricuspid replacement, both mechanical and biological valves were used, and mechanical valve was preferred in our department due to surgeon's preference and young ages of this cohort. The perioperative and follow-up data were collected.

Operations

The majority of our patients underwent a sternotomy to complete the operation. The cardiopulmonary bypass was established by aortic bicaval cannulation. Usually, the SVC cannula was placed high at the junction of innominate vein in case of potential cavopulmonary connection during the operation. After the heart was arrested, the tricuspid valve and right ventricle were inspected, and repair or replacement strategy was finally decided at this moment, in combination with the preoperative echocardiographic findings.

In term of tricuspid repair, Danielson's technique was used in the earlier era, while Carpentier's repair technique was used in the recent 5 years. Briefly, Danielson's technique includes a unicuspid repair of deformed tricuspid valve and horizontal plication of atrialized right ventricle. An annuloplasty ring was placed according to the patient's age and surgeon's judgement. Carpentier's repair contained the detachment of anterior and posterior leaflets, resection of secondary paravalvular apparatus, and vertical plication of the atrialized right ventricle, and then sewed the mobilized leaflets to the anatomic tricuspid annulus. Intraoperative transesophageal echocardiography was performed to evaluate the result of repair, and subsequent re-repair or replacement was undertaken if necessary.

If tricuspid valve replacement was the choice, we routinely preserved the native valve tissue and subvalvular apparatus as much as possible. Interrupted pledgeted stitches was placed. Usually, the stitches were placed at the annulus at the anterior part; however, the stitches around posterior and septal annulus area were placed above the anatomic annulus and towards to the right atrium. The coronary sinus was isolated in the right atrium. A biological or mechanical bi-leaflet valve was placed.

Perioperative management and follow-up

If a patient underwent simultaneous bidirectional cavopulmonary connection, the positive end-expiratory pressure of ventilation was set very low or zero. The patients were extubated as early as possible if hemodynamics was stable and there were no major complications. Anticoagulation plan was started at the first or second postoperative day. Warfarin was given in the first half-year post operation in those who underwent a repair with annuloplasty ring or biological valve replacement, while, in those who had mechanical valve replacement, warfarin would be taken in the whole life. Our

target INR is 2.0–3.0. Pre-discharge echocardiography was performed routinely, and the patients were required to come back to our hospital for postoperative follow-up at 6 months, and then followed up at their regional hospitals.

Statistical analysis

Data were presented as frequency (percentage), median (interquartile range), or mean \pm standard deviation. Differences between groups were tested with the Chi-square test or Fisher's exact test for discrete variables. The Mann–Whitney *U* test or independent *t* test was used for continuous variables between groups. Freedom from reintervention and severe tricuspid dysfunction were analyzed by the Kaplan–Meier method and the log-rank test. Data analysis was performed with the STATA 14.0 software (STATA Corporation, College Station, TX).

Results

Baseline data

Demographic data of the 35 surgical patients are summarized in Table 1. There are 20 males and 15 females. The age at operation ranged from 7 months to 62 years, with median age of 14. With a cut-off age of 14 years, the patients were categorized into pediatric group ($N=16$) and adult group ($N=19$). Preoperative cardiac-associated malformations include 20 atrial septal defects, 2 ventricular septal defects, 2 pulmonary stenosis, and 1 sub aortic ridge. All these anomalies were operated simultaneously. The median GOSE score was 0.8 (range 0.4–1.6) and the mean was 0.78 ± 0.31 . The ratio was used to define four grades of increasing severity. Eight patients had grade 1 (< 0.5), 18 patients had grade 2 (0.5–0.99), 8 patients had grade 3 (1.0–1.49), and 1 patient had grade 4 (≥ 1.5). Preoperative tricuspid regurgitation was severe in 30 (85.7%) patients, moderate in 4 (11.4%) patients, and mild in 1 (2.9%) patient. Preoperative electrocardiogram indicated 4 first-degree atrioventricular blocks (11.4%), 11 right bundle branch blocks (8 complete and 3 incomplete), 1 atrial fibrillation, and 1 atrial flutter. No other severe arrhythmia was found. The NYHA heart function was grade I in 3 patients, grade II in 14 patients, and grade III in the remaining 18 patients.

Operative results

Figure 1 demonstrates the flowchart of all the surgical EA patients in this study. In the 35 patients, the majority had sternotomy (34 patients), and the remaining one had right anterior thoracotomy to complete the operation. Two patients required reoperations (1 dehiscence of repair stitch

Table 1 Baseline demographic data

Demographics	Results
Male	20 (57.1%)
Age (yrs, IQR)	14 (3–40)
Adults/Children	19/16 (54% vs 46%)
Associated cardiac lesions	
ASD or PFO	20 (57.1%)
VSD	2 (5.7%)
PS	2 (5.7%)
Subaortic ridge with LVOTO	1 (2.9%)
GOSE score	0.78 ± 0.31
Grade 1 (< 0.5)	8 (22.9%)
Grade 2 (0.5–0.99)	18 (51.4%)
Grade 3 (1.0–1.49)	8 (22.9%)
Grade 4 (≥ 1.5)	1 (2.9%)
Degree of tricuspid regurgitation	
Severe	30 (85.7%)
Moderate	4 (11.4%)
Mild	1 (2.9%)
Electrocardiogram	
I degree AVB	4 (11.4%)
RBBB	8 (22.9%)
IRBBB	3 (8.6%)
Atrial fibrillation	1 (2.9%)
Atrial flutter	1 (2.9%)

ASD atrial septal defect, PFO patent foramen ovale, VSD ventricular septal defect, LVOTO left ventricular outflow tract obstruction, GOSE score Great Ormond Street Echocardiogram Score, AVB atrioventricular block, RBBB right bundle branch block, IRBBB incomplete right bundle branch block

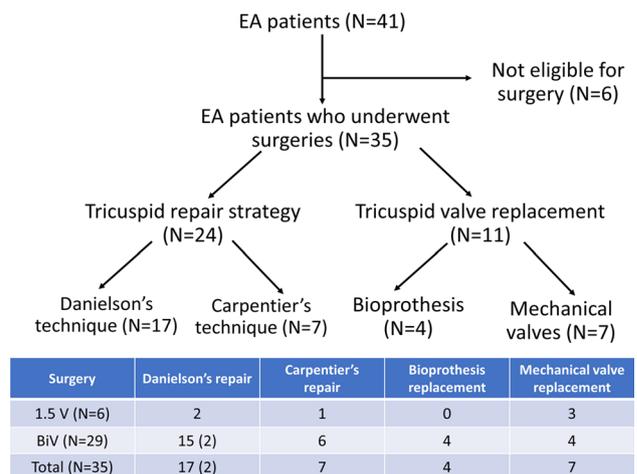


Fig. 1 Flowchart of the surgical patients included in this study. The number in the bracket in the lower table represented patients who had reoperation of the tricuspid valve during the follow-up

and 1 right-ventricular failure) during the same admission, and ultimately tricuspid repair was successfully performed in 24 patients (17 Danielson's repair and 7 Carpentier's repair). Among these 24 patients, an annuloplasty ring was placed in 6 adults and 1 adolescent. In the remaining 11 EA patients, tricuspid valve replacement was applied (4 biological and 7 mechanical valves). The age in replacement patients was older than those who had repair surgery (37 ± 13.7 vs 17.4 ± 19 , $P < 0.01$). In total, 29 patients achieved biventricular repair, while an additional bidirectional Glenn procedure was required in 6 patients (2 Danielson's repair, 1 Carpentier's repair, and 3 mechanical valve replacement). The atrial septal defect was left open intentionally in 6 patients. One patient needed delayed chest closure.

Table 2 summarizes the perioperative data. The average cardiopulmonary bypass time was 90 ± 28 min and cross-clamp time was 48 ± 24 min in all the patients. Carpentier's repair technique required longer cross-clamp time and bypass time compared with the other two techniques (both $P < 0.01$). In total, there were two deaths (5.7%). The first patient had preoperative chronic kidney failure on dialysis, and he had a biological valve replacement, but he developed

low cardiac output syndrome after operation and died 1 week later. The second patient had severe right-ventricular failure preoperatively (GOSE score = 1.6), and he underwent a tricuspid valve replacement and partial closure of atrial septal defect; however, he had refractory arrhythmia postoperation and died 10 days after operation. There was no third-degree atrioventricular block and pacemaker implantation. Postoperative complications included one bleeding, two low cardiac output syndromes, and one wound infection. The average ICU stay was 5.4 ± 5.0 days, and the postoperative in-hospital stay was 13.7 ± 9.6 days. Predischarge echocardiography revealed normal functional prosthetic valve in all the 11 replacement patients. In the other 24 repaired patients, 4 patients had nil or trivial, 17 patients had mild, 2 patients had moderate, and 1 patient had severe tricuspid regurgitation. No significant tricuspid stenosis was found. There was no difference in term of tricuspid function between the two repair techniques.

Follow-up results

The 33 hospital survivors were followed 29.2 (3.1–62.8) months. There was no late death. Six patients had severe tricuspid regurgitation (predischarge regurgitation: 1 severe, 2 moderate, 3 mild; repair strategy: 5 biventricular and one 1.5 ventricular; repair techniques: 5 Danielson's and 1 Carpentier's; annuloplasty ring: 2), and two of them underwent tricuspid replacement 5- and 10-year after initial operation, respectively. The 6-month and 3-year freedom from severe tricuspid dysfunction (stenosis or regurgitation) or reoperation was 94.4% and 78.5%, and no difference was found between different initial surgical choices ($P = 0.22$) (Fig. 2), GOSE score grade ($P = 0.72$), not either between children and adults ($P = 0.05$) or whether using biventricular repair ($P = 0.87$). NYHA heart function was grade 1 in 26 patients, and was grade 2 in the other 7 survivors.

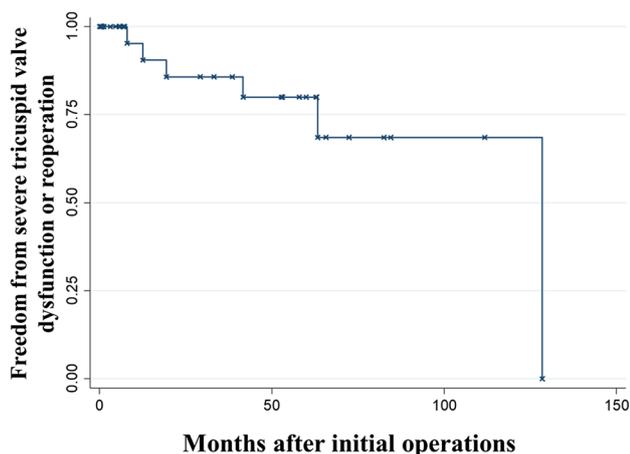
Discussion

This retrospective study indicated either biventricular or one and a half ventricular strategy with repair or replacement of the displaced tricuspid valve in EA patients achieved reasonable short- and mid-term outcomes. Early and late risk of death was not high, and most of the patients had increased NYHA heart function after the surgery; however, a number of patients had more than moderate tricuspid regurgitation at the latest follow-up, and a few of them need reoperation of the failed valve. These patients need close surveillance during the follow-up.

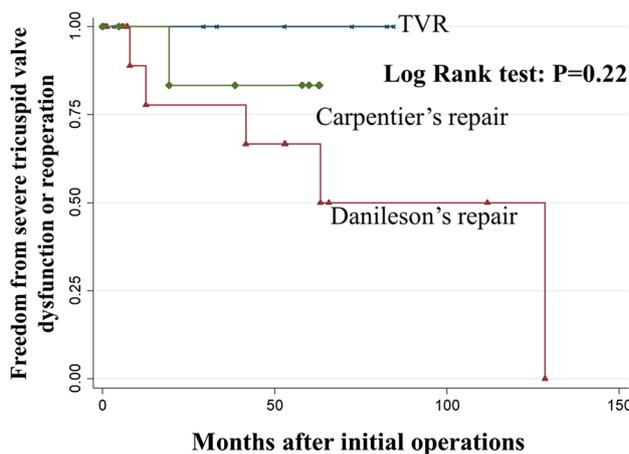
EA has very varied spectrum of the tricuspid valve, development of right ventricle, and associated cardiac lesion, and no two hearts of EA patients are the same. The clinical

Table 2 Perioperative data

Perioperative data	Results
CPBT (min)	90 ± 28
Danielson's repair	71 ± 22
Carpentier's repair	123 ± 15
Tricuspid replacement	100 ± 21
ACCT (min)	48 ± 24
Danielson's repair	36 ± 11
Carpentier's repair	86 ± 14
Tricuspid replacement	44 ± 21
Hospital death	2 (5.7%)
Intubation hours	51.1 ± 69.5 (20, IQR 9–66)
Complications	
Bleeding	1 (2.9%)
Low cardiac output syndrome	2 (5.7%)
Prolonged intubation (> 24 h)	11 (31.4%)
Wound infection	1 (2.9%)
Continuous renal replacement therapy	1 (2.9%)
Delayed chest closure	1 (2.9%)
ICU days	5.4 ± 5.0 (4, IQR 3–7)
Postop stay	13.7 ± 9.6 (1, IQR 7–20)
Predischarge tricuspid valve function	
None or trivial regurgitation	4 (11.4%)
Mild regurgitation	17 (48.6%)
Moderate regurgitation	2 (5.7%)
Severe regurgitation	1 (2.9%)
Well-functioned prosthesis	11 (31.4%)



Months	0	6	12	24	36	48	60
Number at risk	33	23	20	18	16	14	9



Months	0	6	12	24	36	48	60
Danielson's	17	11	8	7	7	6	4
Carpentier's	7	6	6	5	5	4	2
Replacement	9	6	6	6	4	4	3

Fig. 2 Mid-term function of repaired and replaced tricuspid valve. The left column includes all the hospital survivors ($N=33$), and the right column was grouped by different surgical techniques. Tricuspid

dysfunction was defined as any severe regurgitation or tricuspid reoperation after repair, and malfunction of the prosthesis after replacement

manifestations of this diseases also varied a lot, from completely asymptomatic in adulthood to very severe in neonates. The earlier it has been diagnosed, the worse outcomes the patient has. The natural history of EA is not benign [10, 11], the EA neonates who need earlier surgery had a very high risk of mortality up to 50% [12, 13], and those less severe patients gradually developed decreased exercise capacity, cyanosis if an atrial septal defect exists, supraventricular arrhythmia due to the distend right atrium, and even a significant heart failure compromised to both right and left ventricles. When EA patients had the above-mentioned symptoms along with moderate or severe regurgitation on echocardiography, surgery should be considered.

In the past half a century, pioneer surgeons tried to correct this complex cardiac defect. The main principle of the surgery contains the elimination of the tricuspid regurgitation, repair of the associated cardiac defect such as PFO, plication of the atrialized right ventricle, and, sometimes, right atrial reduction. Ablation with different kinds of energy source has been offered to those who have evidence of arrhythmia preoperatively in some centers. The early concept to achieve a competent tricuspid valve is to make the valve unicuspid, and this valve leaflet coaptates to the ventricular septum. Danielson et al. from Mayo Clinic put a pledged stitch in the base of papillary muscle of right-ventricular free wall, and approximate the valve to the ventricular septum; in addition, the tricuspid annulus was reduced with annuloplasty stitches or a ring [3]. Alternatively, Carpentier and Quaegebeur [4, 14] detached the anterior and posterior leaflets, and resected the subvalvular connections between the leaflets

and ventricular wall, only left the main chords on the free edge of the leaflets. This step made the leaflets much more mobile, and the leaflets then re-attached to the anatomic tricuspid annulus after the longitudinal plication of the atrialized ventricle. Carpentier et al. strongly suggest using an annuloplasty ring to reinforce the repair, which yielded the excellent outcomes [4, 5]. However, the durability of this unicuspid repair strategy was not sustained very well, and a number of patients suffered severe tricuspid regurgitation and reoperation in the long term [15, 16]. In the early era of 2000s, anatomic repair with tri-leaflet repositioned to the true annulus was tried by Wu et al. based on the Carpentier's techniques [17, 18]. He detached all the leaflets, and patch the septal leaflets with autologous pericardium, and then re-attached the leaflets to the true annulus; during the procedure, the atrialized ventricle was resected. The reported short- and mid-term outcomes was excellent [19]; however, most surgeons are concerned with the long-term outcomes of the pericardium patch and the aggressive management of the atrialized ventricle, and thus, this procedure was not reproduced widely by the other surgeons. On the contrast, Da Silva et al. freed the septal leaflets, as well, rotated the valve clockwise and made a new conic valve, and then re-attached to the downsized true annulus [6]. The outcomes from his own center were excellent [7, 8], and were proved to be very effective and reproducible in a few large centers [20–25]. Despite the huge success, Dearani also emphasized a steep learning curve of this encouraging cone repair technique, and more recently, he made a few modifications based on cone repair, and the most important one is to use

glutaraldehyde autologous pericardium or Cormatrix to enlarge or reconstruct the hypoplastic tricuspid leaflets, but the long-term outcome of these modifications needs close follow-up [25]. New ideal graft for leaflet enlargement/replacement still needs to be refined.

In our study, the outcomes included mortality, late reoperation, late tricuspid function, and heart function. The in-hospital mortality is 5.7%, which is in the range from the previous studies [5, 12, 16, 25–27]. These two patients both had severe preoperative morbidities, one had chronic renal failure on dialysis, and the other had severe right-ventricular failure, indicating surgery should be recommended before a significant organ failure occurs. In total, 2 patients needed reoperation and both of them had a previous Danielson's repair in childhood. Their residual tricuspid regurgitation gradually increased and, finally, had the valve replaced at 5 and 10 years later. Reoperation of the tricuspid valve was needed in a few patients almost in all the previous reports, and majorly because of recurrence of severe regurgitation. Brown et al. reported 36 reoperation in 182 Danielson's repair patients [16], and Chauvaud et al. reported 16/191 Carpentier's repair in the next 20 years post the initial operations [5]. In terms of cone repair technique, need for late reoperation was not completely eliminated, and the incidence ranged from 1.2 to 13% [23–25, 28]. Dehiscence of the previous repair stitches is the main mechanism of recurrent regurgitation founded during the reoperation, and most patients benefit from re-repair [24, 25, 29]. Late pacemaker implantation was another important reason for reoperation [22, 24]. Tricuspid stenosis is rarely seen.

Reoperation for tricuspid regurgitation may underestimate the long-term function of the repaired tricuspid valve, since some patients may have more than moderate regurgitation, but not receive reoperation yet. Thus, we used the combination of reoperation and more than moderate tricuspid regurgitation as a composite outcome of the tricuspid valve function. In our study, the 5-year freedom from this composite outcome was 78.5%. Several literatures reported long-term tricuspid valve regurgitation after cone repair, and found that 2–13.5% patients had more than moderate regurgitation in the follow-up [20, 23, 27, 30], despite a significant tricuspid function improvement was achieved before dismissal [22].

When a good repair is not achievable, replacement of the tricuspid valve is the choice. In children and young adults, timing of the replacement procedure should be deferred as much as possible for complication of anticoagulation and the prosthetic failure, and this is another reason of high incidence of repair in children in addition to the application of cone repair technique. However, in some adults, especially when age is over 50 years, replacement of the valve should be given with a low threshold, since a replacement is better than an imperfect repair [16, 31, 32] in this cohort. Usually, biological valve is preferred for old patients, and mechanical

valve is the choice for young patients. In this study, we used more mechanical valve than biological valve, because the ages of our patients were relatively young. Another reason is that, based on our clinical experience, the incidence of thrombosis after mechanical valve replacement is less frequent in Asian population compared with western population, and we did not see much thrombosis events in Chinese patients [33, 34].

Determinants of a success operation in EA patients include not only the anatomic feature of the tricuspid valve, but also the development of the right ventricle. The atrialized right ventricle was plicated or resected in most studies and was incorporated into the new right ventricle in both Carpentier's technique and cone repair. However, some patients still had small right ventricle or a significant decreased right-ventricular function, and a bidirectional cavopulmonary connection should be given, because it reduces the preload of right ventricle and fills the left heart better, and, therefore, finally improves the postoperative recovery and late outcome. We used an additional cavopulmonary connection in 6 patients (3 repair and 3 replacement), and all the 6 patients were well in recent follow-up. The concern of SVC syndrome and decreased exercise capacity was not seen in our patients. The use of cavopulmonary connection depends on different patient's age, surgeons' preference, and repair techniques. The incidence ranges from 2 to 66.7%, and most of the patients were in good status [20, 25, 28, 32].

Arrhythmia is frequently seen in EA patients perioperatively, even in those who did not receive any operation, and this is a long-term issue which we need to face [11]. In general, if a patient had severe arrhythmia before operation, then an ablation procedure is recommended during the EA operation; however, it is controversial whether to perform a prophylactic ablation of atrial arrhythmia in those who did not have any onset of arrhythmia. Dearani et al. did the concomitant ablation procedure in up to 40% patients [23], and Anderson et al. reported a 33% rate of anti-arrhythmia procedure [28], while Wu et al. did not perform any anti-arrhythmia operation [19]. In our study, one patient had the previous atrial fibrillation, and he had a concomitant Cox maze IV operation with radiofrequency ablation, and sinus rhythm was sustained at the latest follow-up. We did not do any prophylactic ablation procedure. Although Al-Najashi et al. reported up to 41% occurrence of atrial arrhythmia [35], we did not catch new significant arrhythmia during the follow-up.

Currently, most of the studies of EA patients not only focus on the anatomic results, but also focus on the functional outcomes of the patient and right ventricle. The majority patients in our studies reported increased NYHA heart function after the operation, and most of them are in grade I and grade II at the latest follow-up. The traditional echocardiography is not an ideal modality to evaluate the

right-ventricular function, and MRI is preferred [36]. Lange et al. reported the remodel right ventricle functioned better after cone repair [37]. We did not perform the MRI study of the right ventricle; however, according to a latest MRI study, Beroukhim et al. found reduced tricuspid regurgitation, and stroke volume of right ventricle, in contrast to the increased stroke volume of left ventricle, and improved left-ventricular synchrony after cone repair in 20 patients [38].

The youngest patient in our study was 7 months old at the EA surgery, and we did not have neonates and very small infants in this study, so our results need to be inferred with cautions. EA surgery in neonates and very small infants is much more complicated than those who had operation in later age. Small age at operation was a significant risk factor for EA surgery [12, 13, 39]. Usually, a Starnes operation is preferred in symptomatic neonates, and most of these patients go to single ventricular palliations. Alternatively, Knott-Craig et al. reported success biventricular repair in this difficult subgroup, but the patients' number is limited and longer follow-up is needed [40, 41].

This retrospective study had several limitations. First, despite this study contained patients in the last 12 years, the patient number is small due to low incidence of this disease, and only about two-thirds of them had follow-up data more than 6 months, because the original contact number was not valid now in our early patients. Second, most of our patients had nearly normal preoperative ECG, so we did not perform electrophysiologic study during the initial surgery, and did not focus on the rhythm issue after the surgery, which had been reported in the other literatures. Finally, only Danielson's and Carpentier's repair techniques were used in our department because of surgeons' preference, and we were not able to summarize our own results of the most recent cone repair technique, which we will try in the near future.

Conclusions

The surgeries of EA were variable, and individualized operation achieved good short-term results and reasonable mid-term outcomes. However, severe tricuspid regurgitation during the follow-up was not neglectable in either Danielson's or Carpentier's repair patients, and reoperation in such cases may achieve good outcomes. Cone repair may be considered in our future for its anatomic advantages.

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Compliance with ethical standards

Conflict of interest None for all authors.

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