Cardiovascular Topics

Surgical experience in adults with Ebstein’s anomaly: long-term results
Ozge Altas, Sabit Sarikaya

Objective: The aim of this study was to review late results of the surgical treatment of Ebstein’s anomaly with reconstruction and replacement in adults.

Methods: Medical records of 28 consecutive patients operated on between 1991 and 2014 were reviewed retrospectively. Surgical repair was performed in 19 (67.9%) patients (Hardy: two, Danielson: three, modified Danielson: six, Carpentier: three, Kay annuloplasty reinforced with ring: two), whereas tricuspid valve replacement was performed in nine patients (32.1%). Primary long-term outcomes consisted of right ventricular function, survival and freedom from re-operation. We evaluated the additional impacts of residual tricuspid insufficiency and type of surgery on survival.

Results: In-hospital mortality rate was 7.1% (n = 2) due to low cardiac output status and sepsis. Patients showed a significant postoperative decrease in tricuspid regurgitation (p < 0.001), right atrial size (p < 0.001) and pulmonary hypertension (p = 0.002). The mean follow-up time was 140 ± 71.4 months, with a median of 126 months (105–192). Late mortality occurred in two patients and there was no significant difference in terms of survival based on residual tricuspid insufficiency (p = 0.57) and type of surgery (p = 0.094). Overall survival rates were 89.3, 85.4, 85.4 and 68.3% at five, 10, 15 and 20 years, respectively.

Conclusion: Although complex leaflet reconstruction techniques have evolved to achieve a more physiological and durable repair, both approaches can be performed safely on specific patients and can be alternated, with acceptable rates of survival and re-operation.

Keywords: tricuspid valve, Ebstein anomaly, congenital heart disease

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Having accounted for 1% of congenital heart disease, Ebstein’s anomaly (EA) is associated with abnormalities of the tricuspid valve (TV) leaflets and right ventricle (RV), including atrialisation due to apical displacement of the tricuspid annulus. The surgical strategy for patients with EA is controversial and varies according to anatomical severity, however significant progress has been made since the early 1950s in understanding and managing the disease.

Treatment was formerly directed towards valve repair rather than replacement due to unsatisfactory late results of prosthetic valves.1-3 Lately, leaflet reconstruction techniques, such as Carpentier and cone repairs, have become the first-choice treatment when available, providing good anatomical and physiological results.4,5

Classic approaches emphasise monocuspid coaptation of the TV and cause off-centre diastolic flow, leading to late valvular regurgitation. On the other hand, the modern approach creates near-anatomical repair by re-attaching leaflets clockwise to the true annulus and induces central blood flow.1 However, these types of interventions cannot be offered to all patients. It is not feasible to perform in the presence of pulmonary hypertension (PHT), a severely enlarged RV or when there is lack of septal leaflet tissue.

This report summarises our experience with the surgical management of EA using various techniques and assessing postoperative complications, long-term survival and freedom from re-operation.

Methods
Ethical approval was obtained from our institutional ethics committee. Informed patient consent was waived due to the retrospective nature of the study.

From March 1991 to December 2014, the medical records of 28 patients undergoing surgery for EA were examined and included in a retrospective study evaluating the long-term follow up of at least five years. Patients having significant tricuspid insufficiency (TI) with symptoms of dyspnoea or right-sided heart failure defined as New York Heart Association (NYHA) class III or IV, progressive cardiomegaly, rhythm disturbances and paradoxical embolism were indicated for surgery.1 Patients were excluded if the surgery was performed by the congenital heart surgery team or if they had congenitally corrected transposition of the great arteries or pulmonary atresia with intact ventricular septum and complex conotruncal abnormalities.

Data of the demographic variables, intra-operative process and postoperative outcomes were collected retrospectively. Demographic and clinical data are presented in Table 1.
Pulmonary embolism due to a right atrial thrombus was the first finding in one patient. Other patients mostly suffered from decreased exercise capacity, along with dyspnoea (82.1%) and oedema (39.2%). Cardiovascular risk factors were diabetes mellitus in one patient (3.6%), hyperlipidaemia in seven (25%), arterial hypertension in six (21.4%), and smoking was present in three patients (10.7%).

Pre-operative cardiac assessment was done with transthoracic echocardiography to diagnose associated pathologies and the data are summarised in Table 2. Previous cardiac operations included partial biventricular repair for pulmonary atresia (n = 1), mitral valve replacement (MVR) and aortic valve replacement (AVR) (n = 1), and open mitral commissurotomy (n = 1). The last follow up was performed by review of the results of any recent examinations at our institution in 2020.

Early operative management in our centre typically includes the simpler and more familiar method, which is repair or replacement of the TV and plication of the atrialised RV, concomitant with correction of the associated anomaly. Tricuspid repair, which is the main goal of surgery, evolved through various modifications. The integrity of the anterior leaflet and the attachment location are the first steps to be checked for successful repair.

Initially, correct physiological anatomy of the valve was achieved by bringing the hinge line of the septal and posterior leaflet parallel to the annulus, by transverse plication of the atrialised RV (Hunter–Lillehei–Hardy technique, n = 2). Subsequently, we carried out posterior free-wall plication of the RV and reduction of the right atrium (RA), causing a form of monocuspid valve with/without anterior papillary muscle approximation to the septum, which was centred (classical/modified Danielson technique, n = 3/9). Later, the techniques targeted the leaflets instead of the ventricle for reconstruction of the valve.

The plan was switched to re-attach the anterior leaflet to the true annulus by enforcing it with a ring. The posterior wall was longitudinally plicated to obtain sufficient coaptation (Carpentier technique, n = 3). Additionally, Kay annuloplasty, reinforcing with a flexible ring, was utilised in two patients. Valve repair without a ring was performed in patients with minimal annular dilation and less severe pulmonary hypertension. In cases of severe RV dysfunction and more severe PHT, ring annuloplasty was chosen to form annular stabilisation, avoiding inevitable redilatation with deterioration of valve repair. The repair was checked with a saline test, as well as intra-operative echocardiography.

Unfulfilled leaflet coaptation can be repaired either by bicuspidisation or reshaping of the valve with a pericardial patch. Consequently, valve replacement (n = 9) with a tissue prosthesis at the level of the true tricuspid annulus was preferred when repair was not feasible. The valve tissue adjacent to the RV outflow tract was excised to avoid obstruction of the tract. A flow chart of interventions can be seen in Fig. 1. Surgical procedures are documented in Table 3.

Since postoperative ventricular arrhythmias were common, we preferred to plicate or resect the thin, atrialised ventricle to maintain contractility. Precautions were typically taken on suture lines to avoid injury to the conduction system or right coronary artery. One patient underwent MVR due to severe regurgitation. Radiofrequency ablation was utilised in one patient (3.6%) with chronic atrial fibrillation (AF). Mean cardiopulmonary bypass time was 106.3 ± 30.7 minutes with a mean aortic cross-clamp time of 65.2 ± 19.5 minutes.

### Statistical analysis

Descriptive statistics for categorical variables are given as frequency and percentage, and continuous variables as mean ± SD or median (min–max). Variables of pre- and post-operative echocardiographic evaluations were compared with either the

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**Table 1. Clinical characteristics of patients (n = 28)**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Mean ± SD or number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>33.7 ± 17.7</td>
</tr>
<tr>
<td>Gender (female)</td>
<td>15 (53.6)</td>
</tr>
<tr>
<td>NYHA class: III</td>
<td>9 (32.1)</td>
</tr>
<tr>
<td>NYHA class: IV</td>
<td>17 (60.7)</td>
</tr>
<tr>
<td>Dyspnoea</td>
<td>23 (82.1)</td>
</tr>
<tr>
<td>Oedema</td>
<td>11 (39.2)</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>5 (17.8)</td>
</tr>
<tr>
<td>Palpitations</td>
<td>21 (75)</td>
</tr>
<tr>
<td>NYHA: New York Heart Association</td>
<td></td>
</tr>
</tbody>
</table>

**Table 2. Pre-operative echocardiographic evaluation of patients**

<table>
<thead>
<tr>
<th>Echocardiographic data</th>
<th>Number (%) or mean ± SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>TR ≥ 2</td>
<td>23 (82.1)</td>
</tr>
<tr>
<td>PHT (mmHg)</td>
<td>52.7 ± 12.6</td>
</tr>
<tr>
<td>RAD (cm)</td>
<td>6.1 ± 0.6</td>
</tr>
<tr>
<td>RVEDD (mm)</td>
<td>37.7 ± 11</td>
</tr>
<tr>
<td>RVESD (mm)</td>
<td>52.4 ± 14.1</td>
</tr>
<tr>
<td>RVEF (%)</td>
<td>60.3 ± 5.8</td>
</tr>
<tr>
<td>Low LVEF (%)</td>
<td>2 (7.1)</td>
</tr>
</tbody>
</table>

**Carpentier classification**

A-B 23 (82.1)
C-D 5 (17.8)

**Associated cardiac anomalies**

ASD 10 (35.7)
PFO 1 (3.6)
VSD 2 (7.1)
AVR 4 (14.3)
AR 3 (10.7)

**Arrhythmias**

Atrial fibrillation 1 (3.6)
Supraventricular tachycardia 10 (35.7)

**Prior cardiac surgery**

MVR + TVR 3 (10.7)
AVR + MVR 1
AMV + tricuspid reconstruction 1

**ASD: atrial septal defect; AR: aortic regurgitation; LVEF: left ventricular ejection fraction; MR: mitral regurgitation; PFO: patent foramen ovale; PHT: pulmonary hypertension; RAD: right atrial diameter; RV: right ventricle; RVEDD: right ventricular end-diastolic diameter; RVESD: right ventricular end-systolic diameter; TR: tricuspid regurgitation; VSD: ventricular septal defect.**

**Table 3. Operative data**

<table>
<thead>
<tr>
<th>Operative data</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tricuspid valve repair</td>
<td>19 (67.9)</td>
</tr>
<tr>
<td>Hardy</td>
<td>2 (10.5)</td>
</tr>
<tr>
<td>Danielson/modified Danielson</td>
<td>3/9 (63.2)</td>
</tr>
<tr>
<td>Carpenter</td>
<td>3 (15.8)</td>
</tr>
<tr>
<td>Kay annuloplasty</td>
<td>2 (10.5)</td>
</tr>
<tr>
<td>Tricuspid valve replacement</td>
<td>9 (32.1)</td>
</tr>
<tr>
<td>ASD closure</td>
<td>11 (39.3)</td>
</tr>
<tr>
<td>VSD closure</td>
<td>2 (10.5)</td>
</tr>
<tr>
<td>MVR</td>
<td>1 (3.6)</td>
</tr>
</tbody>
</table>

ASD: atrial septal defect; MVR: mitral valve replacement; VSD: ventricular septal defect.
paired samples t-test or Wilcoxon–rank sum test, as appropriate. Survival was estimated using the Kaplan–Meier method and age- and gender-matched groups were compared using the log-rank test. Analyses were performed with SPSS 15.0.1 for Windows (SPSS Inc, Chicago, IL, USA) and \( p < 0.05 \) was considered statistically significant.

**Results**

Follow up was available for 21 of the 23 survivors (91.3%) and confirmed by clinical evaluation, outside physician report or death notice. The mean age of patients was 33.7 ± 17.7 years (range: 13–54), of whom 15 (53.6%) were female. Most patients were in sinus rhythm pre-operatively. One patient was in chronic AF, whereas a tachycardia-dependent right bundle branch block was present in five patients at the time of the operation. Three patients had had previous cardiac surgery and 14 (50%) needed a concomitant procedure. Carpentier type B anomalies were the most frequent type (\( n = 12 \)), followed by type A (\( n = 11 \)) and C anomalies (\( n = 5 \)). Mean intensive care unit and hospital stays were 6.3 ± 7.9 and 13.2 ± 10.2 days, respectively.

Early mortality was observed in three patients [tricuspid valve replacement (TVR): two, Danielson: one] (10.7%) due to low cardiac output syndrome, where one patient underwent a re-do TVR. The other patients already had pre-operative biventricular dysfunction and developed renal failure. Intra-aortic balloon pump support was provided in all three patients. Two patients (TVR: one, modified Danielson: one) (7.1%) died during the late postoperative period. Causes of mortality were cerebrovascular accident and multi-organ failure (the patient who underwent Danielson repair). Postoperative infection involving the lungs was observed in both patients.

Median follow up was 126 months [interquartile range 105–192]. Overall survival rate was 89.3, 85.4, 85.4 and 68.3% at five, 10, 15 and 20 years, respectively (Fig. 1). At long-term follow up, the NYHA class was II in five patients and III in two patients.

Further analyses were reviewed to see how subgroup movement affected the results, even if the number per group was small, and the patients were divided into two groups (group I: tricuspid reconstruction, group II: TVR) to assess the impact of procedural intervention and residual TI on survival. We concluded that there was no statistically significant difference at survival according to the type of surgery (group I: 94.7% at five years, 89.2% at 10, 15 and 20 years; group II: 77.8% at five, 10 and 15 years) (\( p = 0.094 \), Fig. 2) and the presence of residual TI (\( \geq 2 \)) (\( p = 0.57 \), Fig. 3).

None of the patients required early re-operation for recurrent TI. Freedom from late re-operation was 71.1% at 20 years (Fig. 4). Reasons for re-operation were prosthetic dysfunction.
Survival curves for surgery type
Based on Kaplan–Meier estimates

Fig. 3. Survival according to the type of surgery (group I: tricuspid reconstruction vs group II: TVR).

Survival curves for surgery type
Based on Kaplan–Meier estimates

Log-rank
$p = 0.094$

Number at risk

<table>
<thead>
<tr>
<th>Surgery type = 1</th>
<th>19</th>
<th>19</th>
<th>19</th>
<th>18</th>
<th>17</th>
<th>14</th>
<th>10</th>
<th>9</th>
<th>8</th>
<th>4</th>
<th>2</th>
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</thead>
<tbody>
<tr>
<td>Surgery type = 2</td>
<td>9</td>
<td>7</td>
<td>7</td>
<td>7</td>
<td>6</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Log-rank
$p = 0.57$

Number at risk

<table>
<thead>
<tr>
<th>Residual TI &lt; 2 (1)</th>
<th>14</th>
<th>13</th>
<th>8</th>
<th>4</th>
<th>2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Residual TI ≥ 2 (1)</td>
<td>14</td>
<td>12</td>
<td>10</td>
<td>5</td>
<td>0</td>
</tr>
</tbody>
</table>

Fig. 4. Survival according to residual TI (0 = residual TI < 2, 1 = residual TI ≥ 2)
in one patient at 17.1 years and recurrent TI in two patients who underwent valve replacement at 7.3 and 16 years. One patient was found to have immobile bioprosthesis cusps three years after surgery, and was treated with recombinitate tissue plasminogen activator and discharged with a normo-functional bioprosthesis. There was no known early or late bioprosthetic valve endocarditis reported from our institution.

Consecutive echocardiographic assessment showed a reduction in the RA size (6.1 ± 0.6 to 4.4 ± 0.5 cm, \( p < 0.001 \)) with good functioning of the reconstructed TV. Likewise, improvement in NYHA functional class was observed (\( p < 0.001 \)). Pre- and postoperative echocardiographic evaluation showed that there were significant declines in TI, PHT and right ventricular ejection function as well (\( p < 0.05 \)). At a recent follow up, only one patient was found with moderate TI, whereas the others had no or mild regurgitation. Comparative echocardiographic evaluation of the patients can be seen in Table 4.

Anti-arrhythmic therapy was required in 10 patients due to paroxysmal supraventricular tachycardia. Even though Wolf–Parkinson–White syndrome is commonly associated with EA, none of our patients showed its electrocardiographic characteristic. One patient underwent concomitant radiofrequency ablation due to AF. Atrioventricular block was observed in six patients (21.4%) following surgery; one patient who underwent Danielson repair eventually needed a permanent pacemaker, and five more patients recovered to sinus rhythm before discharge. Two patients experienced AF and managed medically during follow up. Renal failure was observed in three patients, and one needed haemodialysis and died of multi-organ failure. Two of three patients with bleeding needed re-exploration for pericardial tamponade. Postoperative complications are shown in Table 5.

**Discussion**

Numerous variants of EA have been challenging for the surgeon since the earliest repair techniques. Advances in techniques have improved the early and late survival of patients. It is crucial to determine an appropriate time for surgical intervention. The pathological process, deformity and displacement of the TV cause severe TI and RA dilatation, in addition to conduction abnormalities or atrioventricular accessory conduction pathways. Patients may present with different forms of arrhythmia. Studies show 17 to 30% of patients presented with tachycardia or accessory pathways. Most of the patients, except five with supraventricular tachycardia, had sinus rhythm in our study. Of the five, one patient who underwent TVR, died showing ventricular pre-excitations. Ventricular dysrhythmias appear to be lethal in postoperative patients who also have massive cardiomegaly, and are associated with poorer long-term results. Right-sided or bi-atrial maze is the best option for AF, but it also can be managed with radiofrequency ablation, which we preferred in one patient.

Various TV repair techniques have been described and commonly include the cone technique, Carpenter repair, and modifications of the Danielson technique. Although valve replacement can be done safely, repair of the deformed TV is the goal of surgical therapy due to the risk of thromboembolism and valve failure. Even though the modern approach of cone reconstruction is safely applicable and showed favourable impact on right ventricular remodelling, it is not feasible to perform in all patients because of significant valve abnormalities such as inadequate size or attachment of the free edge of the leaflet to the ventricular wall, massive RV and PHT, and relatively older age.

Our approach mainly focused on the monocusp repair technique in severely displaced anterior leaflets and plication of the atrialised RV with satisfactory survival and freedom from re-operation rates. We did not perform cone repair because of anatomical variability [left ventricular ejection fraction in two patients, presence of severe right atrioventricular dilatation in three patients, lack of septal leaflet tissue in five patients] and an inadequate learning curve due to the rarity of the procedure. Initial simplified methods did not improve the anatomical anomaly, however, we achieved satisfactory results by eliminating TI. We prefer these methods to extirpate the progressive RV failure by avoiding surgical/myocardial injury, and this approach may be preferred, especially in patients with advanced age. In the presence of severe TV annulus or RV dilatation and in the absence of a complete septal leaflet, TV replacement should be considered to reduce cross-clamp time.

Both mechanical and biological valves were used with similar re-operation rates, but with higher survival rates in favour of a biological prosthesis. It is crucial not to underestimate the increased mortality and complication rate with valve replacement as a risk factor. As TVR is a general risk factor, our study showed no statistically significant difference in the impact of procedural intervention on survival (\( p > 0.094 \)). Due to the increased interest in valve-in-valve procedures and avoidance of chronic anticoagulation, bioprosthetic valves generally were preferred, with good durability.

Currently, early mortality rates range from 2.4 to 9%, whereas late mortality rates are reported up to 31%. In our study, early and late mortality rates were 10.7 and 7.1%, respectively.
These survival rates are acceptable. Even the largest series reported 94, 90, 86 and 76% late survival rates at five, 10, 15 and 20 years, respectively, in patients undergoing modern cone repair.\textsuperscript{22} It would not be right to analyse the risk factors related to peri-operative mortality in our study due to limited functional assessment. However, in other studies, pre-operative RV and LV function have been accepted risk factors for mortality.\textsuperscript{23} Mitral regurgitation, RV outflow tract obstruction and more than moderate ventricular function were reported to be the predictors of late mortality.\textsuperscript{24} A slightly increased early mortality rate was observed in three patients in our study because two of the patients had pre-operative biventricular failure and the third underwent redo operation.

In all cases undergoing TV repair, we had low incidence of recurrent TI, as well as good RV function in the long term. In our institutional experience, freedom from re-operation was 71.1\% at 20 years (Fig. 5) and no patients required early re-operation for recurrent TI. Limitations with the design of this study are the variability of procedures performed in the repair group, and the lack of variables in echocardiographic parameters and recent modern examinations [such as magnetic resonance imaging (MRI) or exercise testing]. Despite these limitations, the improvement in clinical function and survival showed satisfactory results.

Late outcomes regarding freedom from re-operation following cone reconstruction are limited, however Holst \textit{et al}. reported 98.8\% at six years in adult patients.\textsuperscript{17} Moreover, in non-cone repairs, freedom from re-operation at 10 years was reported between 77 and 82\%.\textsuperscript{22} Some other small group studies reported the rates between 88.7 and 92.9\% at 10 to 20 years of follow up.\textsuperscript{23,24} Pre-operative echocardiographic assessment is essential to determine the fastening sites of the anterior leaflet. Positioning of the papillary muscles is important for assessment of leaflet mobility. Consequently, it might result in important residual TV regurgitation following surgical repair.

McLellan-Tobert \textit{et al}. reported improved exercise tolerance in adults following TV surgery.\textsuperscript{13} Although late functional assessments are limited, it is certainly clear that patients showed improvement in NYHA class compared to their pre-operative status (\(p < 0.001\)). Likewise, many groups have reported excellent results with non-cone repair.\textsuperscript{22,23} The majority of our patients showed significant decreases in RA size and PHT (\(p < 0.001\) and \(p = 0.002\)). We preferred to use ring annuloplasty in two patients with severe RV dysfunction and PHT, in contrast to modern approaches (Carpentier repair, cone reconstruction).

Some studies reviewed the use of ring annuloplasty over De Vega’s suture annuloplasty in non-EA patients, and found no significant difference in outcome between the two techniques.\textsuperscript{23} Our results demonstrate that most patients were able to undergo a satisfactory repair with marked reduction in the degree of TI. Furthermore, a bidirectional cavopulmonary shunt or atrial septal fenestration can be pursued to off-load RV volume in the presence of RV failure and dilatation. PHT and valvular disease of the left side should be evaluated prior to shunting to avoid left ventricular end-diastolic pressure.\textsuperscript{1}

It is stated with cone repair that anatomical restoration of the entire RV induces larger effective stroke volume, causing preferable RV remodelling. Holst and colleagues showed reduction in TI and RV size, as well as an increase in RV fractional area (\(p < 0.0001\)).\textsuperscript{17} However, the mean follow up was much less (3.5 \pm 2.5 years) than the two decades of our patients’ follow up. Li \textit{et al}. found a more synchronised RV movement pattern and decreased functional RV volume by evaluating tricuspid annular movement synchronicity index.\textsuperscript{24} Recently, MRI has been used to show increased pulmonary flow and decreased functional right ventricular end-diastolic volume, and these studies concluded that there was significant improvement in clinical status.\textsuperscript{25}

The primary limitation of this study was the difficulty in collecting a large number of cases with each procedure, therefore it is hard to draw conclusions. The study consisted of a small group of patients in the period before the emergence of cone repair, therefore we are not able to generalise our study against the clinical outcomes of more recent studies. However, there have been some contra-indicated patients, complicating the results of cone repair during the last decade. Hence, we preferred to use vertical plication of the atrialised RV portion in order to avoid paradoxical motion of the RV. We believe this approach provides clinical benefit in selected patients. Another partial limitation is that MRI has been used recently, allowing for wider analysis of RV function over echocardiography. It is not possible to use MRI for all patients due to lack of availability or patients being followed up by local physicians.

**Conclusion**

Although TV function and RV remodelling has been improved with the evolution of many different techniques, the interventions should be focused on more physiological reconstruction for the durability of surgical valve repair. Recent surgical techniques are in contrast to our study, however we have shown satisfactory results obtained with simple approaches. It is questionable to standardise operative techniques against the wide spectrum of anatomical disorders. Further long-term research is required on contemporary interventions for additional assessment.
The authors thank Dr Ali Karagoz for his assistance with the statistics used in this report.

References