

Original
Article

Cone Type Repair Has Become Our First Option in the Treatment of Adult Ebstein Anomaly

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Purpose: Different surgical procedures were defined due to degree of Ebstein anomaly. In this study, we are reporting our surgical experience of adult patients with Ebstein anomaly.

Methods: We analyzed the patients, who had operated with the diagnosis of Ebstein anomaly from March 2011 through February 2018, retrospectively. We evaluated patients in two groups: patients with cone type surgical repair were in Group 1 and patients with other surgical (Danielson, Carpentier) procedures were in Group 2.

Results: There were 23 patients in the study; 9 (39%) were in Group 1 and 14 (61%) were in Group 2. In the comparison of preoperative data, groups were statistically similar. Aortic cross clamp and cardiopulmonary bypass (CPB) times, need for inotropic agents, intubation time, postoperative >2 tricuspid regurgitation incidence, intensive care unit, and hospital staying times were statistically significantly higher in Group 2. There was one mortality (4.3%) in Group 2 due to sepsis.

Conclusion: Cone type repair can safely be performed in the repair of Ebstein anomaly with acceptable results. Our results are encouraging to prefer this technique as one of the first-line treatment of Ebstein anomaly. However, further randomized controlled studies are recommended to evaluate the efficacy of this surgical procedure.

Keywords: cone type repair, Ebstein anomaly, tricuspid valve

Introduction

Ebstein anomaly was first defined by Wilhelm Ebstein in 1866. Its prevalence is 0.5% among all congenital cardiac diseases.^{1,2} It is a rare malformation mainly involving

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tricuspid valve and right ventricle.³) Posterior and septal leaflets of tricuspid valves are placed down from the tricuspid annulus to the right ventricular apex. It may have different characteristics in a wide spectrum from the slightly downward location of septal leaflet to completely non-functional right ventricle. Characteristically, there is an atrialized right ventricle and a small chambered right ventricle with thin wall in the Ebstein anomaly.

There are different degrees of clinical presentations according to the leaflet hypoplasia and placement. Cyanosis is detected in nearly half of the patients.⁴) Dyspnea, tachycardia, syncope, and decreased exercise tolerance are the most common symptoms. Ebstein anomaly can easily be diagnosed with two-dimensional transthoracic echocardiography. Paradoxical embolism, increased cyanosis, extremely damaged tricuspid valve structure, persistent arrhythmia, and decreased functional capacity are the indications for the surgery.²)

Different surgical treatment alternatives were defined due to degree of cardiac malformation and leaflet conditions. Tricuspid valve repair, tricuspid valve replacement, atrialized right ventricle plication, one and half ventricle repair, Starnes procedure, heart transplantation, and surgical treatment of arrhythmias are most preferred surgical procedures. In this study, we are reporting our surgical experiences of cone type repair and comparison of its results with other surgical techniques.

Materials and Methods

We retrospectively analyzed the patients, who had been operated due to the diagnosis of Ebstein anomaly in our department from March 2011 through February 2018. Demographic details and clinical information of the patients were obtained from the medical records. Patients older than 18 years old with ≥ 3 tricuspid valve deficiency and New York Heart Association (NYHA) class 3–4 symptoms were included in the study. Patients with right ventricle outlet tract stenosis that necessitated intervention were excluded. We reviewed surgical reports and created two groups according to the surgical techniques. Patients with cone type surgical repair were in Group 1 and patients with other surgical (Danielson, Carpentier) procedures were in Group 2.

All patients were evaluated with transthoracic echocardiography in the preoperative and postoperative examinations. Patients over 40 years of age were evaluated with coronary angiography before the operation.

Surgical procedure

After median sternotomy, pericardium was carefully dissected and kept in glutaraldehyde containing solution. Cardiopulmonary bypass (CPB) was established with aortic and selective cannulation of both vena cava. After the right atriotomy, atrial septum, pulmonary venous return, tricuspid valve leaflets, chordal apparatus, papillary muscles, atrialized right ventricle, right ventricular volume, coronary sinus, possible conduction system, and right coronary artery were evaluated in detail.

In Group 1, tricuspid leaflets were mobilized and interior leaflet was made closer to septal leaflet by rotating clockwise. In patients with extremely poor leaflet structure, leaflet augmentation was applied with autologous pericardial patch. We used glutaraldehyde fixed pericardial patch in three patients. To maintain the soft consistency of the patch, we treated patch with glutaraldehyde for 3 minutes and washed it saline solution. Atrialized

right ventricle and inferior annulus were longitudinally plicated from the inside of the right ventricle. Attention was paid to avoid stenosis, and no prosthetic materials such as rings were used.

In Group 2, atrialized right ventricle was transversely plicated and tricuspid ring annuloplasty performed in Danielson procedure. During the Carpentier technique, atrialized right ventricle was longitudinally plicated and tricuspid reimplantation was done.

Atrial septal defect or patent foramen ovale was present in all patients and these defects were repaired with primary or glutaraldehyde fixed pericardial patch plasty. During the surgery, attention was paid to avoid right coronary artery distortion and conduction system damage. In patients with restricted right ventricular capacity and failure of weaning from the CPB despite of the inotropic support, one and half ventricle repair was added to the operation. Vena cava was separated from superior right atrium with protecting sinoatrial node and anastomosed to the right pulmonary artery.

Statistical analyses were generated using IBM Statistical Package for the Social Sciences 24.0 (SPSS 24.0, SPSS Inc., Chicago, IL, USA) software. Categorical variables are summarized as number and percentage, and continuous variables are summarized as mean \pm standard deviation. For the comparison of Group 1 and Group 2, t-test and Wilcoxon tests were used. $p < 0.05$ was regarded as statistically significant in the comparisons.

Results

A total of 23 patients between the ages of 21 and 64 years underwent operation with the diagnosis of Ebstein anomaly in the study period. There were 10 males (43.5%) and 13 females (56.5%). The most common symptom of the patients was dyspnea. In all, 14 patients described paroxysmal palpitation in the preoperative anamnesis. However, sinus rhythm was observed in all patients. There were nine patients in Group 1 and 14 patients (10 with Danielson procedure, 4 with Carpentier technique) in Group 2. No additional procedure for arrhythmia was performed during the surgery. There was no statistically significant difference among the groups for preoperative variables (**Table 1**).

Aortic cross clamping and CPB times were statistically significantly high in Group 2. There was no tricuspid valve replacement in any of the cases. Requirement of concomitant one and half ventricle repair was 1 (11.1%) and 2 (14.2%) in Group 1 and

Table 1 Comparison of preoperative data

| Variable | Group 1 (n = 9) | Group 2 (n = 14) | p value |
|-----------------------|------------------|------------------|---------|
| Age (year) | 42.3 ± 13.2 | 44.1 ± 11.6 | 0.925 |
| Gender (male/female) | 4 M/5 F | 6 M/8 F | 1.000 |
| Weight (kg) | 68.4 ± 10.1 | 67.1 ± 11.4 | 0.529 |
| NYHA Class | | | |
| Class 3 (n) | 6 (66.7%) | 9 (64.3%) | 0.803 |
| Class 4 (n) | 3 (33.3%) | 5 (35.7%) | 0.270 |
| Oxygen saturation (%) | 88.5 ± 5.2 | 89.3 ± 4.1 | 1.000 |
| Ejection fraction | 54 ± 3 | 52 ± 5 | 0.701 |
| GOSE score >1 | 1 (11.1%) | 2 (14.2%) | 0.107 |
| Albumin (g/dL) | 3.7 ± 0.6 | 3.9 ± 0.4 | 0.590 |
| Creatinine (mg/dL) | 0.9 ± 0.23 | 0.8 ± 0.27 | 0.353 |
| Hemoglobin (gr/dL) | 13.6 ± 2.1 | 14.4 ± 1.5 | 0.215 |
| INR | 1.1 ± 0.3 | 1.21 ± 0.24 | 0.409 |
| Platelet count | 161.000 ± 34.000 | 180.000 ± 27.000 | 0.095 |

INR: international normalized ratio; NYHA: New York Heart Association; GOSE: great ormond street echocardiogram

Table 2 Comparison of operative data

| Variable | Group 1 (n = 9) | Group 2 (n = 14) | p value |
|---|-----------------|------------------|---------|
| ACC time (minutes) | 76.3 ± 11.2 | 82.6 ± 13.5 | 0.007 |
| CPB time (minutes) | 91.5 ± 14.7 | 98.5 ± 16.4 | 0.032 |
| Temperature (°C) | 32.5 ± 1.3 | 32.3 ± 1.6 | 0.603 |
| Concomitant one and half ventricle repair (n) | 1 (11.1%) | 2 (14.2%) | 0.107 |
| Inotropic support (n) | 5 (55.5%) | 11 (78.5%) | 0.014 |

ACC: aortic cross clamp; CPB: cardiopulmonary bypass

Table 3 Comparison of postoperative data

| Variable | Group 1 (n = 9) | Group 2 (n = 14) | p value |
|--|-----------------|------------------|---------|
| Intubation time (hours) | 6.7 ± 2.4 | 8.2 ± 3.1 | 0.012 |
| Drainage (mL) | 610 ± 70 | 590 ± 80 | 0.805 |
| Hemoglobin at PO 1 st day (mg/dL) | 10.3 ± 1.5 | 10.8 ± 1.1 | 1.000 |
| Tricuspid insufficiency >2 | 1 (11.1%) | 3 (21.4%) | 0.009 |
| SVT (n) | 3 (33.3%) | 5 (35.7%) | 0.270 |
| ICU staying time (days) | 3.1 ± 1.2 | 4.3 ± 1.4 | 0.038 |
| Hospital staying time (days) | 7.4 ± 1.3 | 9.2 ± 1.7 | 0.024 |

ICU: intensive care unit; SVT: supraventricular tachycardia

Group 2, respectively. Patients, who had undergone concomitant one and half ventricle repair, were discharged from the hospital at postoperative 8th day with NYHA class 1–2 symptoms (**Table 2**).

Intubation time, intensive care, and hospital staying times were statistically significantly high in Group 2. Supraventricular tachycardia attacks were detected in eight patients. Of six were converted to normal sinus rhythm with medical treatment and remaining two were with electrical cardioversion (**Table 3**).

In the routine postoperative control transthoracic echocardiography, >2 tricuspid valve regurgitation was revealed in one patient in Group 1 (11.1%) and three patients (21.4%) in Group 2. In these patients, tricuspid

valve insufficiency was decreased from 4 degrees to 3 degrees and NYHA symptom scores was ≤2 postoperatively. Mean gradient of tricuspid valve was under 4 mmHg in all patients. There was no reoperation due to bleeding and no complete atrioventricular block that necessitated permanent pacemaker implantation. There was one mortality in Group 2 (4.3%) because of the sepsis.

Discussion

In the surgical treatment of Ebstein anomaly, various alternatives, from tricuspid valve repairs to heart transplantation were defined based on cardiac malformation. In this study, we evaluated our surgical approaches in the

patients with Ebstein anomaly over 20 years of age. We performed plication of atrialized right ventricle and tricuspid valve repair in all patients. According to our results, cone type repair had better operative and postoperative outcomes.

Cone type repair has been declared with the advantages of decreased reoperation, morbidity, and mortality rates.⁵⁾ Gradually increasing application prevalence of cone procedure was preferred especially since it is better tolerated by patients, provision of anatomic and physiological repair and low mortality rates.⁶⁾ During the cone procedure, different type of modifications including leaflet augmentation and ring annuloplasty might be applied successfully.⁷⁾ In our department, since our postoperative tricuspid valve repair success could not reach the desired level with the other techniques, we started to apply cone type repair in our last nine patients. We performed leaflet augmentation in patients with very inadequate leaflet tissue, but we did not prefer ring annuloplasty to avoid intracardiac prosthetic material. Aortic cross clamp and CPB times, inotropic agent usage, intubation time, postoperative >2 tricuspid insufficiency, intensive care unit, and hospital staying times were lower with cone procedure. Postoperative outcomes of these patients were encouraged us to keep in mind this technique as a first option for the treatment of Ebstein anomaly.

Ebstein anomaly was anatomically and functionally classified in four types considering the anterior leaflet mobility and atrialized right ventricular volume. While anterior leaflet mobility decreases from Type A to Type D, atrialized right ventricle volume increases. Repair chance of tricuspid valve is decreased after Type C.⁸⁾ Also it is reported that values over 1 in Great Ormond Street Echocardiogram (GOSE) score in the younger than 1-year old patients might be related with poor prognosis.⁹⁾ In our patients, there were one patient in Group 1 and 2 patients in Group 2 with >1 GOSE score. There was one mortality in Group 2 and this patient's GOSE score was 1.5.

After the surgery of Ebstein anomaly, residual tricuspid regurgitation might trigger right ventricular volume overloading, decline in right ventricular function, and significant intractable arrhythmias.¹⁰⁾ This clinical condition influence mid-term and long-term outcomes of the surgical procedure. In our first patients, more than moderate tricuspid valve regurgitation was 20%. Then, we started to apply cone type repair and the rate of more than moderate tricuspid regurgitation was decreased to 10%. Due to this encouraging results,

cone type repair has become our first option in the surgery of Ebstein disease.

Cyanosis, dyspnea, decreased exercise tolerance, tachycardia, chest pain, and syncope are the most common symptoms in Ebstein anomaly. Preoperative functional capacity of the patients (NYHA) was especially reported as the most important parameter on survival.¹¹⁾ One-third of our patients had NYHA Class 4 symptoms while remaining had Class 3 symptoms. We could not reveal any correlation with functional capacity and mortality due to relatively small groups in our study.

In Ebstein anomaly, postoperative mortality rates differ due to complexity and its specific surgical procedure. Jost et al.¹²⁾ reported that they performed biological tricuspid valve replacement for 73% of patients with Ebstein anomaly and plication was the treatment of choice for those with thinned and transparent right ventricle. In the Mayo Clinic series containing 539 patients, mortality in the first 30 days was reported as 4.9% in patients with tricuspid valve repair and as 6.8% in patients with tricuspid valve replacement.¹³⁾ While Chauvaud et al.¹⁴⁾ reported decreasing of mortality from 12% to 7% in their recent patients; Silva et al.¹⁵⁾ declared 3.8% mortality with cone type repair. Recently in another study from Mayo Clinic, it was stated that the mortality rates decreased to 1% with cone type repair.⁶⁾ We had only one mortality in our 23 patients that was in Group 2. There was no mortality in the last nine patients that we performed cone technique.

Postoperative myocardial infarction caused by right coronary artery distortion during the plication of atrialized right ventricle might develop as a complication. Adachi et al.¹⁶⁾ stated the risk of myocardial infarction as 0.4% after plication of atrialized right ventricle. We avoided possible right ventricle distortion in our cases and there was no postoperative myocardial infarction.

There are studies reporting that right ventricle deficiency risk would decrease with the addition of one and half ventricle repair to the operation in patients with Ebstein anomaly and very limited right ventricle capacity.¹⁷⁾ Right ventricular workload is decreased with bidirectional cavapulmonary anastomosis in patients with restricted right ventricular volume in surgical inspection and pulmonary artery pressure less than 18 mm Hg. Thus, postoperative patient toleration is increased and surgical outcome is improved. We performed concurrent one and half ventricle repair in three patients with restricted right ventricular capacity (13%). These patients well tolerated weaning from CPB and benefit from surgery.

There are studies reported that del Nido cardioplegia solution provides safer myocardial protection especially in congenital cardiac diseases.¹⁸⁾ In our department, we preferred mild hypothermia (32°C) and isothermal blood cardioplegia for myocardial protection. Thus, we avoid from deep hypothermia in our cases due to its potential side effects such as respiratory dysfunction and longer intensive care unit and hospital staying times.

The present study has the nature limitations of retrospective studies, and information are depended on the availability of the medical records. Magnetic resonance imaging (MRI) is helpful to evaluate ventricular diameter and pulmonary vascular structures. However, we gained only a few patients MRI data that did not enough to compare the groups. In this study, we compared the results of our last nine patients who had undergone cone type repair with other patients. Randomized-controlled studies about this subject will be more accurate. We could not reach detailed reports of echocardiographic examinations. However, data that we gained are sufficient to carry out the idea and is encouraging for future studies.

Conclusion

In the surgical treatment of Ebstein anomaly, cone type repair is more secure compared to other surgical procedures. According to our results, we prefer cone procedure as treatment of choice for Ebstein anomaly. However, more comprehensive studies comparing long-term results of different surgical treatment alternatives are required. At least, this technique should be kept in mind as a surgical alternative for the repair of Ebstein anomaly.

Disclosure Statement

The authors certify that they have no conflicts of interest in the manuscript.

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