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Successful surgery of Ebstein's anomaly of 11-year-old patient after having had two cardiac arrests

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Abstract

Successful surgery of Ebstein's anomaly of 11-year-old patient after having had two cardiac arrests

Ebstein's anomaly is a rarely seen congenital heart disease with high morbidity and mortality rates. Impaired function of tricuspid valve and partially atrialized right ventricle are the main characteristics of the disease. Patients may have symptom-free survival for long terms whereas intra uterine death may also be encountered due to wide range of the symptoms. Regurgitation of the tricuspid valve and right-sided heart failure are the foremost symptoms. Surgery must be performed in symptomatic patients without delay. Carpentier's approach to the surgical treatment of Ebstein's anomaly is one of the methods that can be applicable. In this study it is reported that successful surgical treatment of 11-year-old child with Ebstein's anomaly who had two cardiac arrests before operation, by using Carpentier's technique.

Keywords: Carpentier's Technique, Ebstein's Anomaly, Surgery

Öz

Ebstein anomalisi olan ve iki kez kardiyak arrest geçiren 11 yaşındaki hastanın başarılı cerrahisi

Ebstein anomalisi, yüksek morbidite ve mortalite oranlarına sahip, nadir görülen bir doğuştan kalp hastalığıdır. Triküspit kapağın işlev bozukluğu ve kısmen atriyalize olmuş sağ ventrikül, hastalığın temel özellikleridir. Hastalar uzun süreli semptomsuz sağkalıma sahip olabilirken, semptomların geniş bir yelpazesi nedeniyle rahim içi ölümle de karşılaşılabilir. Triküspit kapağın yetersizliği ve sağ kalp yetmezliği en başta gelen semptomlardır. Semptomatik hastalarda ameliyat gecikmeden yapılmalıdır. Carpentier'in Ebstein anomalisinin cerrahi tedavisine yaklaşımı, uygulanabilecek yöntemlerden biridir. Bu çalışmada, ameliyattan önce iki kez kardiyak arrest geçiren Ebstein anomalili 11 yaşındaki bir çocuğun Carpentier tekniği ile başarılı cerrahi tedavisi bildirildi.
Anahtar Kelimeler: Carpentier Tekniği, Ebstein Anomalisi, Cerrahi

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INTRODUCTION

Ebstein's anomaly is an uncommon but complex congenital defect involving the right ventricle (RV) and tricuspid valve (TV), accounts for 0.5 % of all congenital heart diseases (1). Main clinical features are apically localized septal leaflet of the TV and dilation of atrialized part of the RV, those may lead to several disorders including tricuspid regurgitation, low cardiac output and depressed RV function (2). Although congenital malformations are present at birth signs and symptoms may occur at any age and differs from arrhythmias, right-sided heart failure, cyanosis and sudden cardiac death to asymptomatic period due to anatomic severity and degree of right-to-left interatrial shunting (3). Surgical repair is indicated for patients with right heart dilation and progressive ventricular dysfunction. Various surgical methods are introduced in the literature and one of them is Carpentier's technique (4).

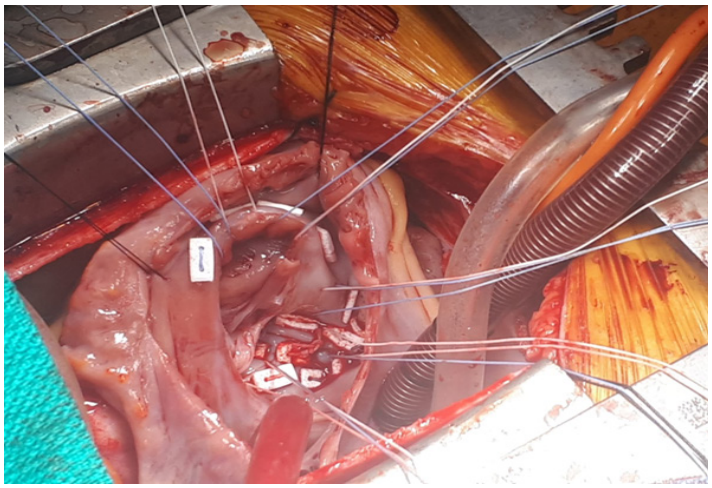
In this study we aimed to present the surgical treatment of 11-year-old patient with Ebstein's anomaly by using Carpentier's technique after having had two cardiac arrests.

CASE REPORT

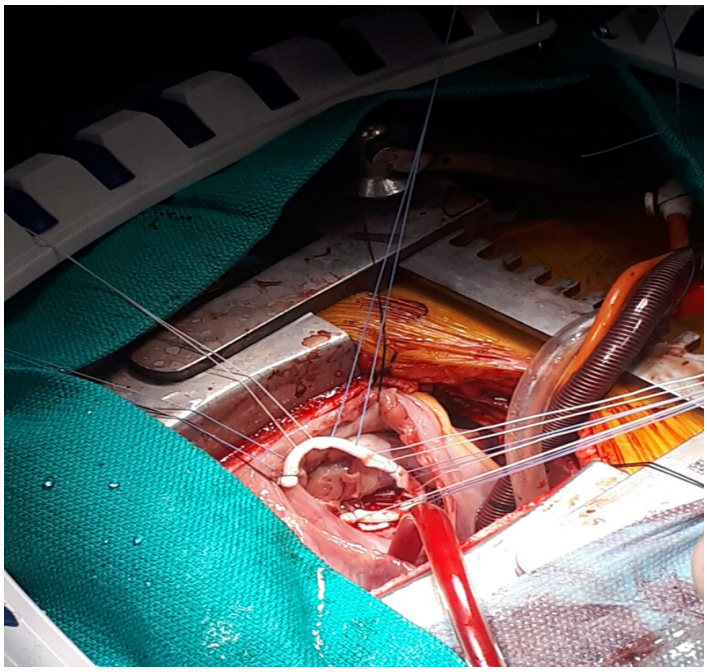
A 11-year-old Syrian immigrant girl was admitted to emergency department via ambulance with the complaint of general condition disorder. Patient was unconscious and initial physical examination revealed bradycardia (heart rate; 40/min), hypotension (arterial blood pressure; 60/30 mmHg) and shallow breathing. At the third minute after admission to the hospital, patient had cardiopulmonary arrest. Cardiopulmonary resuscitation (CPR) was started and she was intubated and stabilized with low-dose inotropic support (0.05 mcg/kg noradrenalin) within five minutes.

From the patient's history and medical data it was revealed that patient was diagnosed with Ebstein's anomaly two years ago and although surgery was recommended she was followed with medical treatment due to poor health system conditions in her country.

She had gradually increased exercise intolerance and fatigue for two years. There was no abnormality in laboratory findings and bed-sided detailed echocardiography was performed and Ebstein's anomaly was confirmed with the evidence of the apical displacement of the septal and posterior leaflets of the tricuspid valve with grade 3tricuspid regurgitation and right atrial enlargement with small right ventricle. There was a right ventricle outflow tract obstruction caused by anterior valve. Inlet portion of the right ventricle was atrialized. The left atrium and left ventricle were within normal limits and left ventricular ejection fraction was 50%. There was no additional abnormality including atrial septal defect or patent foramen ovale detected. After written informed consent taken from her parents, operation was decided and the patient was transferred to cardiovascular surgery intensive care unit. Polymorphic ventricular tachycardia was detected in ECG while follow up period. While preparing for the surgery the patient had second cardiac arrest attack. After CPR, continued three minutes, heart rhythm was achieved but operation was delayed because of hypotension. Operation was performed on the next day because of the stabilized clinical conditions including the arterial pressure values of 110/60 mmHg without inotropic support that permitted general anesthesia and surgical trauma. Carpentier's method was applied in the operation with annuloplasty to the TV by using 29 no Duran AnCoreand plication of the atrialized segment of the right ventricle under cardiopulmonary pump (Fig. 1,2).



Picture 1. Intraoperative sight of plicated atrialized segment of the right atrium and preparation of the ring replacement to the tricuspid valve



Picture 2. Intraoperative sight of the ring replacement to the tricuspid valve

Duration of cross clamp and operation was 45 min and 2 hours, respectively and duration of intensive care stay was 2 days. There was no need to inotropic support after operation and patient was extubated at 6th hours of postoperative period. Patient was discharged 7 days after operation with a mild tricuspid regurgitation. There were no clinical abnormalities detected during the postoperative first week and first month controls.

DISCUSSION

Ebstein's anomaly is the malformation of the TV and the myopathy of the right ventricle. Symptoms and the signs of the Ebstein's anomaly are variable. Thus, treatment modalities differ from the clinical situations of the patients. Although technological improvements in the area of cardiovascular surgery, survival of the Ebstein's anomaly is still under 50% of the patients older than 10years (5). Asymptomatic patients can be conservatively treated and kept under close follow-up whereas surgical procedure is required in the presence of right heart dilation and progressive impairment of ventricular systolic function (6). Surgical approach to TV regurgitation improves long-term outcome. Long-term survival over 20 years after the operation was reported up to be 90% in several studies and surgery should not be delayed if it is necessary because of the surgical risks and long-term mortality of the advanced disease (7).

Various surgical methods were introduced in treatment of Ebstein's anomaly and tricuspid valve repair is the main aim of the surgical intervention. It also includes right atrial reduction, RV plication and atrial septal closure. Although tricuspid repair is considerable it should be kept in mind that outcomes of tricuspid replacement in adult patients indicate that it is effective and safe (4).

Surgical treatment of Ebstein's anomaly remains a challenge. Hunter and Lillehei (8), Carpentier et al. (9), Danielson et al. (10), and Da Silva et al. (11) introduced a variety of surgical modalities in treatment of Ebstein's anomaly, to make the function of the TV better. Hunter's technique includes the transposition of the tricuspid valve to its normal location. According to the method described by Carpentieret al. (9) suggested the longitudinal plication of the RV and return of the TV to the correct level, reinforced with a prosthetic ring. Danielson et al. (10) applied a technique consists of a transverse plication of the atrialized RV, posterior tricuspid annuloplasty, and right reduction atrioplasty. Da Silva et al. (11) approached the surgical treatment of Ebstein's anomaly by using some principles of the Carpentier's concepts except prosthetic ring to the TV. Instead of that cone reconstruction was performed to the patients (12). In this procedure, a cone shape is formed from the anterior tricuspid valve by rotating clockwise and the base of this cone is attached to the true tricuspid valve ring. Atrialized part of the right ventricle is plicated or resected if necessary. It is accepted as the best anatomic repair because it allows compliant tissue coaptation at the atrioventricular junction (13).

The wide range of pathophysiologic and anatomic presentations of Ebstein's anomaly has made it difficult to reach uniform results with the surgical repair of this complex congenital heart disease and even today there seems to be no 'single' surgical approach that offers optimal results (14). There are four types of surgical anatomy described according to the surgical anatomy in Ebstein's anomaly. According to the type C, there is a displacement of septal and posterior leaflets, enlargement of right atrium, small right ventricle and restricted anterior leaflet motion and it was emphasized in the literature that reconstruction is not suitable if there is a right ventricle outflow tract obstruction caused by anterior valve which is stricked to the right ventricle (3, 15). As the mentioned case was involved in type C, Carpentier's technique was performed instead of Cone procedure in our case.

Polymorphic ventricular tachycardia (PMVT) is an arrhythmia faster than 100 beats per minute and characterized by rapid and frequent changes in QRS with axis, morphology or both. Its clinical symptoms are in association with Q-T interval. In a study conducted by Werf et al, efficacy of beta blocker therapy was shown in patients with PMVT (16). In our case as the Q-T interval was normal and absence of intraoperative apparent rhythm disorder, additional intervention was not performed and follow up with beta blocker therapy was decided.

CONCLUSION

In conclusion, although it is difficult to deal with Ebstein's anomaly, appropriate surgical treatment to the symptomatic patients without delay can save lives.

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Peer-Review

Both externally and internally peer reviewed.

Conflict of Interest

The authors declare that they have no conflict of interests regarding content of this article.

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The authors report no financial support regarding content of this article.

Ethical Declaration

Informed consent was obtained from the parents of the participant and Helsinki Declaration rules were followed to conduct this study.

Authorship Contributions

Concept: MAC, Design: OA, ATD, Supervising: MAC, ATD, Financing and equipment: MAC, OA, Data collection and entry: OA, Analysis and interpretation: ATD, OA, Literature search: ATD, MAC, OA, Writing: OA

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