Cone Reconstruction with Successful One and A Half Ventricle Repair for Ebstein Anomaly

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ABSTRACT

Ebstein anomaly is a rare pathology among congenital cardiac diseases with a prevalance of 0.5%. Ebstein anomaly patients suffer from arrhytmia, severe right ventricular dysfunction and left ventricular dysfunction due to cyanosis and right ventricular involvement. Most of the patients are presented at infancy, childhood and rarely after adolescence ages. Biventricular repair has a high mortality and morbidity and unacceptable functional results in Ebstein anomaly patients with severe right ventricular dysfunction, one and a half ventricle repair yields better results. In our case, we presented a one and a half ventricle repair with a successful Cone type repair in a 34-year-old male patient who referred to our clinic with cardiac arrest presentation and was detected to have functional type C Ebstein anomaly.

Key Words: Cone reconstruction; Ebstein anomaly; one and a half ventricle repair.

Cone Tipi Onarım Yapılan Ebstein Anomalili Hastada Eş Zamanlı Başarılı Bir Buçuk Ventrikül Tamiri

ÖZ

Ebstein anomalisi, doğumsal kalp hastalıkları arasında %0.5 oranında nadir görülen bir patolojidir. Ebstein anomalisi olan hastalarda siyanoz ve sağ ventrikül etkilenmesine bağlı aritmi, ciddi sağ ventrikül disfonksiyonu ve sıklıkla sol ventrikül disfonksiyonu görülebilmektedir. Bu hastalar bebeklik, çocukluk ya da daha nadir olarak ergenlik sonrası karşımıza çıkmaktadır. Ciddi sağ ventrikül disfonksiyonuna sahip Ebstein anomalili hastalarda biventriküler onarım yüksek mortalite ve morbidite ve kötü fonksiyonel sonuçlar ortaya çıkarabilmekte ve bir buçuk ventrikül onarımı ile daha iyi sonuçlar alınabilmektedir. Bu olgu sunumunda, kardiyak arrest öyküsü ile başvuran, fonksiyonel tip C Ebstein anomalisi tespit edilen 34 yaşında erkek hastada yapılan başarılı Cone tipi onarım ile birlikte bir buçuk ventrikül tamiri sunulmuştur.

Anahtar Kelimeler: Bir buçuk ventrikül tamiri; cone tipi onarım; Ebstein anomalisi.

INTRODUCTION

Ebstein anomaly (EA) is a rare malformation which has a prevalence of 0.5% among congenital cardiac diseases and it mainly involves the tricuspid valve and right ventricle[1,2]. It was separated into four types by Carpentier with an anatomic and functional classification. While atrialized right ventricle volume is quite small in Type A (5%), atrialized right ventricle volume increases in Type B (35%), atrialized ventricle is quite wide and has limited movement in Type C (51%) and the anterior leaflet movement is quite limited by short chordas. Tricuspid leaflet tissue decreased (8%), right ventricle wall became severely thinner and its contractility decreased in Type D[3].

The most common additional cardiac anomaly is atrial septal defect. Atrial tachyarrhythmias are the second common ones compared to atrial dilatation. One or more accessory transfer pathways of Wolff-Parkinson White syndrome are present in nearly 15% and atrioventricular nodal reentrant tachycardia is present in 2% of the patients[4]. Cyanosis is present in nearly half of the patients with EA.

The most common parameter on survival is the functional capacity of the preoperative patient (New York Heart Association)[5]. Decrease in functional capacity, increase in cyanosis,

Cite this article as: *Özbek B, Öztürker K, Tomrukçu F, Kara KA, Şavluk ÖF, Çevirme D, et al. Cone reconstruction with successful one and a half ventricle repair for Ebstein anomaly. Koşuyolu Heart J 2021;24(Suppl 1):S65-S67.*

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Submitted: 08.04.2021
Accepted: 12.05.2021
Available Online Date: 16.05.2021

persistent arythmia or paradoxal emboli primarily makes us consider the necessity of surgical treatment. Surgical treatment alternatives are tricuspid valve repairment, tricuspid valve replacement, one and a half ventricle repairment, atrialized right ventricle plication, right reduction atrioplasty, surgical treatment of arhythmias, concurrent treatment strategies or cardiac transplant. A 34-year-old male type C EA patient with a history of cardiac arrest was presented in this article.

CASE REPORT

The functional capacity of the patient who admitted to our clinic with a history of 10 minute lasting cardiac arrest (terminated after an effective cardiopulmonary resuscitation) was determined as NYHA (New York Heart Association) class 3. According to the blood gas analysis of the conscious and cooperative patient, oxygen saturation was 91%, arterial blood pressure was 91/58 mmHg, pulse rate was 65/min, in sinus rhythm and respiratory rate was 14/minute. In the transthoracic echocardiography, medium-advanced (degree 3-4) tricuspid insufficiency, tricuspid valve was located in 7.1 cm distal in anteroseptal zone (Figure 1), remaining right ventricle cavity was very low, secundum atrial septal defect was wide, GOSE index was 1 and the condition was evaluated as severe EA.

Cone type repair aiming to decrease postoperative right ventricle insufficiency risk and one and a half ventricle repairment were planned and made in the same session for the patient with limited right ventricle capacity. Intraoperative pulmonary artery pressure was 14 mmHg. Secundum atrial septal defect was completely covered with pericardial patch. Cross clamp time was 131 minutes and total perfusion time was 161 minutes. The patient left cardiopulmonary bypass without any problems and was extubated in the fifth hour after being hospitalized in intensive care unit and annular level of neo tricuspid valve was observed in control echocardiography, the coaptation was good, a weak deficiency was present, there was no narrowness, atrial septum was intact and the bidirectional cavopulmonary anastomosis was smooth (Figure 2).

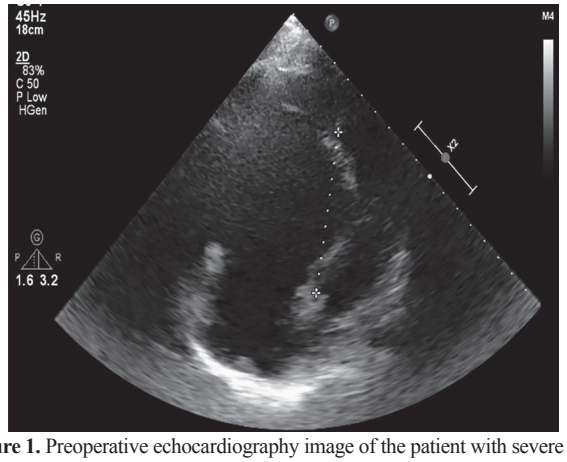

Figure 1. Preoperative echocardiography image of the patient with severe Ebstein anomaly and tricuspid valve located 7.1 cm distal in anteroseptal zone.

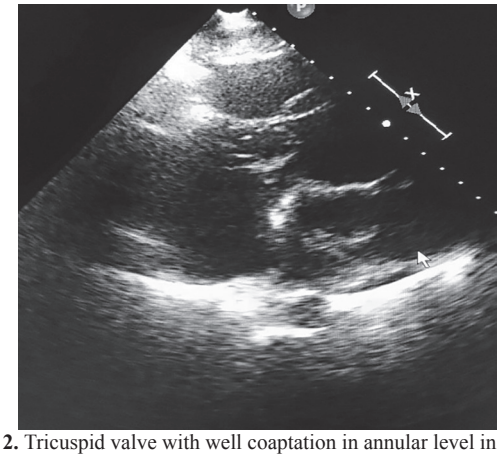

Figure 2. Tricuspid valve with well coaptation in annular level in echocardiography on postoperative fisrt day.

A 2.5 cm pericardial effusion was detected in the control echo of the patient taken in intensive care unit due to supraventricular tachycardia attack on postoperative third day. Subxiphoid pericardial tube drainage was made and pericardial effusion did not recur in the patient who returned to sinus rhythm. The patient was discharged on postoperative seventh day. Postoperative functional capacity was observed as NYHA class 1.

DISCUSSION

In EA patients, anomaly type, tricuspid valve insufficiency amount, right ventricle dysfunction degree, functional capacity and presence of other accompanying cardiac anomalies (pulmonary stenosis, atresia) are among the most important factors effecting operation mortality[1]. In addition to tricuspid and right ventricle reconstruction, one and a half ventricle repairment is suggested to be added to the treatment of patients referring with childhood and adolescence right ventricle dysfunction[6,7]. We applied this in our case.

Cone type reconstruction we applied in our patient is also used commonly in tricuspid and right ventricle repairment methods in addition to Carpentier type (longitudinal plication and tricuspid reimplantation) and Danielson type (transverse plication and tricuspid ring annuloplasty). Mobilisation of tricuspid leaflets is provided in cone type reconstruction and interior leaflet is made closer to septal leaflet side by rotating clockwise. It is completed with the inner longitudinal plication of atrialised right ventricle and inferior annulus[8-10].

In patients in whom one and a half ventricle repairment was also added to these methods, it was shown that right ventricle front load decreased, pulmonary artery flow increased, left ventricle filling pressure was increased and the left ventricle function recovered.

In patients with severe Ebstein anomaly with a primary right ventricle dysfunction as in our patient, we consider that the addition of one and a half ventricle repairment to surgical treatment would decrease mortality and reoperation need.

Informed Consent: Informed consent form was obtained from patient.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept/Design - BÖ, KÖ; Analysis/Interpretation - BÖ, ET; Data Collection - FT, KK; Writing - BÖ, DÇ; Critical Revision - ÖŞ, ET; Statistical Analysis - KÖ, FT; Obtained Funding - DÇ, KÖ; Overall Responsibility - BÖ, HC; Final Approval - All of authors.

Conflict of Interest: The authors have no conflicts of interest to declare

Financial Disclosure: The authors declared that this study has received no financial support.

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