Surgical Treatment of Ebstein's Anomaly in Adult with One and a Half Ventricle Repair: Case Report

Erişkin Ebstein Anomalisinde Bir Buçuk Ventrikül Cerrahi Onarım

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ABSTRACT Ebstein's anomaly is a rare congenital heart defect. Depends on the severity of malformations, different symptoms can be seen. Goals of surgical intervention are to increase pulmonary blood flow, to minimize tricuspid insufficiency, to reduce or eliminate right-to-left shunt, to optimize right ventricular function, and to reduce or to eliminate arrhythmias. In this article we reported surgical treatment of Ebstein's anomaly with the "one and a half ventricle repair" in two adult patients. With the use of one and a half ventricle repair; closure of the atrial septal defect resolves cyanosis, repair of tricuspid valve decreases the regurgitation from the valve and heart failure and systemic and pulmonary circulations are separated. One and a half ventricle repair can be utilized in patients with severe Ebstein's anomaly and impaired right ventricular function who are at high risk for biventricular repair.

Key Words: Ebstein anomaly; tricuspid valve insufficiency; heart defects, congenital

ÖZET Ebstein anomalisi nadir rastlanan doğumsal bir kalp defektidir. Malformasyonun ciddiyetine paralel olarak değişik semptomlar görülebilir. Cerrahi tedavinin amacı pulmoner kan akımını arttırmak, triküspit kapak yetersizliğini azaltmak, sağ sol şantı ortadan kaldırmak veya azaltmak, sağ ventrikül fonksiyonlarını düzeltmek ve aritmileri önlemek veya azaltmaktır. Bu bildiride "bir buçuk ventrikül tamir" ile cerrahi tedavi uygulanmış olan iki erişkin olgu sunulmaktadır. Bir buçuk ventrikül onarımı yöntemi sonunda atriyal septal defektin kapatılması siyanozu ortadan kaldırır, triküspit kapağın tamiri kapak ve kalp yetmezliğini azaltır, sistemik ve pulmoner dolaşım birbirinden ayrılmış olur. Düşük sağ ventrikül fonksiyonuna sahip olan ve biventriküler onarımın riskli olduğu hastalarda birbuçuk ventrikül tamir prosedürü uygulanabilir bir cerrahi tedavi seçeneğidir.

Anahtar Kelimeler: Ebstein anomalisi; triküspid kapak yetersizliği; kalp kusurları, doğumsal

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Ebstein's malformation is a rare cardiac defect accounting for less than 1% of all congenital heart diseases.[1] Ebstein's anomaly (E.A) is defined by a downward displacement of the annular attachments of the septal and posterior leaflets of the tricuspid valve into the inlet portion of the right ventricle. This defect is characterized by remarkable morphologic variability thus a broad spectrum of clinical presentation. Consequently, diagnosis may be made in symptomatic newborn infants, in children or in adults. The choice of treatment depends on the patients clinical presenta-

tion, morphology of the tricuspid valve, presence of right to left shunt via atrial septal defect, severity of right heart dilatation and dysfunction, or presence of atrial or ventricular arrhythmias.[2-4] Biventricular repair is usually possible for majority of patients. One and a half (1.5) ventricle repair can be used in patients with poor right ventricle function.[2,4,6,7]

CASE REPORTS

First case is a 48-year-old man, referred to our hospital with dyspne and fatigue symptoms, which onset 3 years ago. The heart rate was 42 per minute and the blood presure was 95/55 mmHg. Electrocardiogram showed sinus bradycardy and p pulmonale. In echocardiographic assessment E.A, secundum atrial septal defect , mild-moderate tricuspid regurgitation and global systolic dysfunction were detected. Left ventricle ejection fraction was 35%. In coronary angiograms coronary arteries are in normal pattern. He was in New York Heart Association (NYHA) functional class III.

Patient 2 is a 18-year-old woman, referred to our hospital with palpitation. The heart rate was 80 per minute and the blood presure was 105/60 mm Hg. Electrocardiogram showed; right axis deviation, p pulmonale and intermittent atrial arrhythmias. In echocardiographic assessment E.A, mild tricuspid regurgitation were seen. Left ventricle ejection fraction was 40%. She was in NYHA functional class III.

Aortic and bicaval cannulation were used for the cardiopulmonary bypass. The superior caval cannula was placed at the level of the innominate vein to facilitate bidirectional cavopulmonary anastomosis. At the beginning of the procedure the mean right pulmonary pressure was 16 mmHg in Patient 1 and 15 mmHg in Patient 2. Systemic mild hypothermia and intermittant normothermic blood cardioplegia were used during the cross clamp period. With the use of Carpentier technique tricuspid valve repair was performed, the redundant right atrial wall and appendage were excised, secundum atrial septal defect was closed with a patch of autologous pericardium (Figure 1-3). During rewarming period a bi-directional

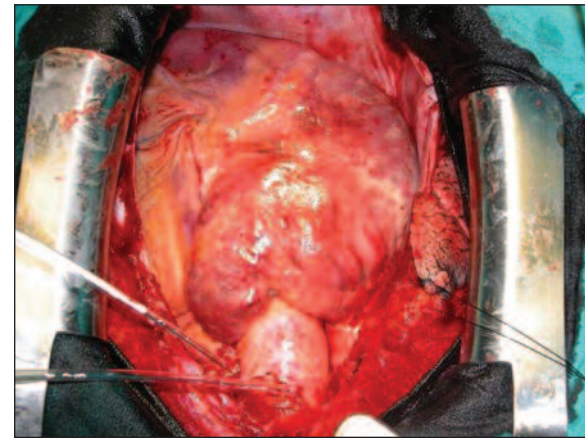

FIGURE 1: Operative view of atrialized right ventricle and dilated right atrium.

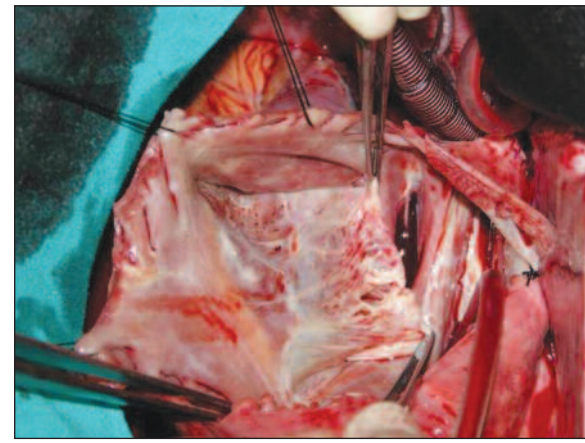

FIGURE 2: Operative view of tricuspid valve anatomy.

cavopulmonary shunt (BDCPS) was performed (Figure 4). The decision to proceed with a BDCPS was made preoperatively because of dilated and thin walled poorly right ventricular function, and a long history of tricuspid valve insufficiency

Operative and postoperative courses were uneventfull for both cases. Control echocardiograms showed a mild tricuspid regurgitation. Neither tricuspid stenosis nor residual intracardiac shunt was observed. Patient 1 was discharged on the 11th postoperative day with a 95% saturation and NYHA class I symptoms. Patient 2 was discharged on the 9th postoperative day with a 97% saturation and NYHA class I symptoms. Follow-up time was 8 months for patient 1 and 6 months for patient 2. Both cases did not have required reintervention or hospitalization for any reason. They have demon-

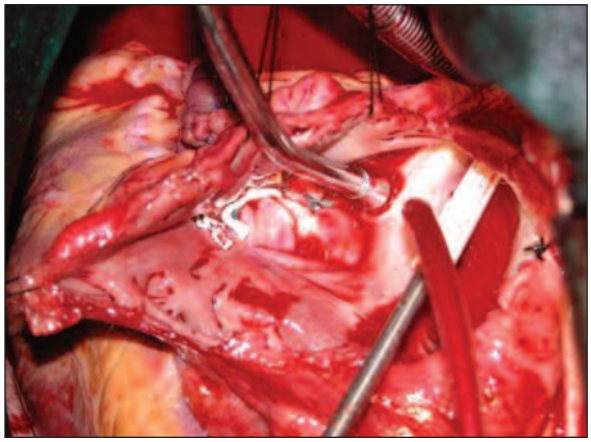

FIGURE 3: Operative view after the Carpentier repair technique.

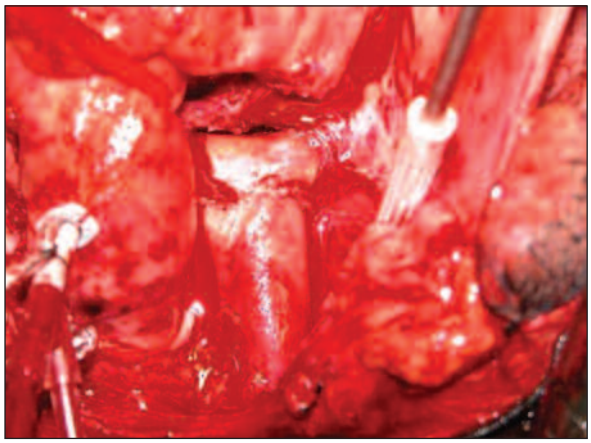

FIGURE 4: Bi-directional cavopulmonary shunt.

strated improved exercise tolerance. They didn't have evidence of heart insufficiency. Their oxygen saturation by pulse oximetry were over 93%.

DISCUSSION

E.A is a complex form of congenital heart diseases.[1,2] There is an inverse relationship between severity of symptoms and the age of the diagnosis. Patients with E.A have a reduced life expectancy.[1-3] The surgical management of E.A in adult patients is repair or replacement of the tricuspid valve in conjuction with closure of the atrial septal defect.[2,3] With the use of 1.5 ventricle repair; closure of the atrial septal defect resolves cyanosis, repair of tricuspid valve decreases the regurgitation from the valve and heart failure and systemic and pulmonary circulations are separated. The BDCPS diverts 40% of the systemic venous return from the right ventricle. The benefit of the BDCPS could be related to decreasing the work index of right ventricle and subsequently prevent postoperative ventricular dilatation.[6] The present cases showed no cyanosis and no futher episodes of right ventricle failure during follow up period. The indication of 1.5 ventricle repair in Ebstein's malformation has not been confirmed, although it decreases operative mortality and it is effective in patients with low cardiac output and acute right ventricle failure.[4,6,7] The need for a BDCPS should be anticipated with the evaluation of the functional right ventricle preoperatively.[4,5] Eventually, the decision is made after weaning from the cardiopulmonary bypass. High right and left atrial pressure measurements and low mixed-venous saturation measurements are good indicators for BDCPS.[4] In our patients, the decision to proceed with a BDCPS was made preoperatively because of dilated and thin walled poorly right ventricular function and a long history of tricuspid valve insufficiency.

There are several concerns with the use of BDCPS. The disadvantages include a longer operating time, loss of the catheter access to the right heart and superior vena cava syndrome.[3,4] In our cases, no undesirable effect of the BDCPS were noted.

In summary, 1.5 ventricul repair of Ebstein anomaly can be utilized in patients with severe Ebstein anomaly and impaired right ventricular function. It eliminates right to left intracardiac shunt, improves exercise tolerance and functional class who are at high risk for biventricular repair. In addition, early mortality can be decreased and the quality of life can be improved.

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