Ege Tıp Dergisi / *Ege Journal of Medicine* 2019;58(2):161-165

Surgical treatment of Ebstein anomaly: A unicenter experience

Ebstein anomalisinin cerrahi tedavisi: Tek merkez deneyimi

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Abstract

Aim: Ebstein anomaly is a rare congenital cardiac disorder that causing by failure of delamination of the tricuspid valve. Various surgical methods have been developed so far as treatment options. Our initial experience and short term follow up reported herein.

Materials and Methods: Ten patients underwent surgical treatment with a diagnosis of Ebstein anomaly in our center between 2007 and 2017. Two patients underwent one and a half ventricle repair and Danielson method applied to eight patients. The X-clamping and cardiopulmonary by-pass (CPB) timing, duration in intensive care unit (ICU), extubation time and need for blood product were compared between Danielson and one and a half ventricle repair applied patients.

Results: There was no mortality among patients. In the one and a half repair applied group both X-clamping (p=0.044) and CPB time (p=0.044) was longer compared to Danielson group. On the other hand there is no significantly difference between groups in ICU duration (p=0.400) or extubation timing (p=0.889). Furthermore, need for blood product and amount of drainage were not different between groups when two infant age cases were excepted while calculating.

Conclusion: Danielson method is safe and effective in the repair of Ebstein anomaly and can be applied at infant age with similar clinical outcomes. Nevertheless, one and a half repair option is also beneficial in patients with a large size atrialized right ventricular volume. The last decision should be confirmed by heart team considering patient characteristics and surgeons' past experience.

Keywords: Ebstein anomaly, Danielson method, one and a half ventricle repair.

Öz

Amaç: *Ebstein anomalisi triküspid kapak delaminasyonundaki yetersizlik sonucu oluşan nadir bir konjenital kalp hastalığıdır. Hastalığın tedavisinde farklı cerrahi yöntemler bugüne dek tanımlanmıştır. Bu çalışmada cerrahi deneyim ve kısa dönem cerrahi sonuçlarımızı belirtmekteyiz.*

Gereç ve Yöntem: *Çalışmaya 2007-2017 yılları arasında merkezimizde Ebstein anomalisi tanısı ile opere edilen on hasta dahil edildi. İki hastaya bir buçuk ventrikül tamiri uygulanırken diğer sekiz hasta Danielson metodu ile opere edildi. Bu hasta grupları arasında kros klemp ve kardiyopulmoner by-pass zamanları ile yoğun bakım ve entübasyon süreleri ve kan ürünü replasman gereksinimleri karşılaştırıldı.*

Bulgular: *Hastalarda mortalite gözlenmedi. Bir buçuk ventrikül tamiri uygulanan hastalarda Danielson uygulanan hasta grubuna göre hem kros klemp (p=0,044) hem de kardiyopulmoner by-pass süresi (p=0,044) daha az izlendi. Diğer yönden iki grup arasında yoğun bakım yatış süreleri (p=0,400) veya entübasyon süreleri (p=0,889) arasında anlamlı fark izlenmedi. Danielson ameliyatı uygulanan iki infant hasta hesaplamaya dahil edilmediği durumda yapılan karşılaştırmada, kan ürünü gereksinimi ve drenaj miktarlarında gruplar arasında belirgin fark izlenmedi.*

Sonuç: *Danielson metodu, infant yaş grubunda Ebstein anomalisi tedavisinde diğer tanımlanmış tamir yöntemlerine benzer klinik sonuçlarla, güvenli ve etkin bir biçimde uygulanabilir. Yine de bir buçuk ventrikül tamir seçeneği geniş atriyalize ventrikülü olan hastalarda faydalı bir yöntemdir. Nihai karar hastanın karakteri ve cerrahi ekibin geçmiş tecrübeleri göz önüne alınarak kalp ekibi tarafından verilmelidir.*

Anahtar Sözcükler: *Ebstein anomalisi, Danielson metodu, bir buçuk ventrikül onarımı.*

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Received: 01.03.2018 Accepted: 01.06.2018

Introduction

Ebstein anomaly was first described on a post-mortem 19 years old male, cyanotic case with dyspnea, palpitations, jugular venous distension, and cardiomegaly in 1866 by Wilhelm Ebstein at autopsy, afterwards in 1949 first ante-mortem case was diagnosed (1,2). The prevalence of this disorder is determined in various studies with a range between 0.03% and 0.06% among all congenital heart diseases (3-5). The most concomitant congenital heart disease is atrial septal defect (ASD) and Wolf-Parkinson-White syndrome may coexist in these patients up to 25% (6).

Ebstein anomaly is a rare congenital cardiac disorder that causing by failure of delamination of the tricuspid valve. This pathology leads to apical displacement of tricuspid valve and partial atrialization of right ventricle. Thus, manifest as from mild to severe right ventricular failure and tricuspid valve (TV) regurgitation. Generally, the anterior valve locates in usual line, besides septal and posterior valves stretched downward displacement into right ventricle. The Caprentier classification which is based on these two abnormalities is very beneficial to assess the severity of disease. The occurrence and timing of clinical presentation change due to widely varied hemodynamic results of the pathology. Therefore, careful medical management and a close echocardiographic follow up are advised to all diagnosed asymptomatic patients.

On the other hand, in presence of significant desaturation, reduced right ventricular function, paradoxical embolization, increased cardiothoracic index or limited functional capacity and occurrence of ventricular arrhythmias, surgical treatment is strongly recommended. Nowadays, by many surgeons in most centers, if two ventricle corrections are possible, developed repairs techniques are being frequently using safely with a satisfied late term results in the light of past experiences of Danielson, Carpentier and da Silva (7).

In clinic, we perform Danielson modification as first option in patients with favorable anatomy. Otherwise single or one and a half ventricular correction methods are considering temper to right ventricular hypoplasia. In this report, we describe our surgical experience and our short-term surgical results.

Material and Methods

Patient characteristics

Ten patients underwent surgical treatment with a diagnosis of Ebstein anomaly in our center between 2007 and 2017. The patients' hospital records were reviewed retrospectively. There were seven female and three male patients. As well as two of them were infant patients, the mean age was 17.4±3.6 years (8 months to 42 years) and mean weight was 34.8±9.6kg (4.9 - 80kg).

All patients were diagnosed via transthoracic echocardiography (TTE). Detailed physical examination, chest radiography and electrocardiography analysis results were recorded. Cardiothoracic index (CTI) results were measured and all patients had ≥0.5 rate. Eight patients were in sinus rhythm and other two were with right bundle branch block. Functional classes according to New York Heart Association (NYHA) were recorded. All patients had severe tricuspid valve regurgitation ('3-'4) and eight of them had concomitant ASD. On the other hand in one case, patent ductus arteriosus (PDA) was diagnosed preoperatively. Hence, over 70% atrialized right ventricle volume, two patients had significantly high The Great Ormond Street Echocardiography (GOSE) score.

The X-clamping and cardiopulmonary by-pass (CPB) timing, duration in intensive care unit (ICU), extubation time and need for blood product were compared in these two patients compared to other eight Danielson method applied patients.

Postoperative TTE were evaluated after seventh day of operation and amount of tricuspid valve regurgitation and left ventricle ejection fraction (LVEF) were examined.

Indications for operation

Indications for operation included fatigue or decreased exercise tolerance, cyanosis, and progressive right ventricular dilation. The goal of surgical intervention was to restore tricuspid valve competence before the development of significant right ventricular overload and dysfunction and prior to left ventricular (LV) dysfunction. Earlier operation was pursued if successful tricuspid valve repairs, rather than replacement (Table-1).

Statistical analysis

Statistical package for the Social Sciences for windows version 15.0 for windows (SPSS, Chicago, IL, USA) was used for acquired data analysis. Measured data were presented as mean±standard deviation. Due to the data does not set a normal distribution and limited sample size, whereas groups are including only eight and two cases respectively, a non-parametric statistical test (Mann-Whitney U) was used for comparison between the two groups instead of a parametric test. A p value of <0.05 was considered statistically significant.

Table-1. Preoprative Characterictics of Patients.

	Age/sex	Symptoms	NYHA class	CTI	Preoperative TR	ECG	Concomitant anomaly
1	10m/F	Dyspnea, cyanosis	II	0.65	'3	NSR	ASD
2	13y/F	Dyspnea, cyanosis	III	0.6	'3-4	NSR	ASD
3	18y/M	Exercise intolerance, dyspnea	IV	0.65	'4	NSR	ASD
4	24y/F	Exercise intolerance	II	0.6	'3	NSR	ASD
5	8m/F	Dyspnea, cyanosis	III	055	'3-4	NSR	ASD
6	42y/F	Exercise intolerance, palpitation	III	0.7	'3-4	RBBB	ASD+PDA
7	26y/F	Exercise intolerance, palpitation	III	0.6	'3	NSR	-
8	15y/F	Cyanosis, dyspnea	II	0.55	'3-4	NSR	ASD
9	35y/M	Exercise intolerance palpitation, chest pain	III	0.6	'3	RBBB	-
10	16y/M	Exercise intolerance, dyspnea	III	0.65	'3-4	NSR	ASD

NYHA: New York Heart Association, CTI: Cardiothoracic index, TR: Tricuspid regurgitation, ECG: Electrocardiography, NSR: Normal sinus rhythm, RBBB: Right bundle branch block, ASD: Atrial septal defect, PDA: Patent ductus arteriosus, M: Male, F: Female.

Table-2. Comparison and the Descriptive Data of the Groups.

		Mean	Median	Variance	Std. deviation	Std. Error	p value
X-clamp time (min)	1 ½ repair	91.50	91.50	60.500	7.77817	5.50000	0.044
	Danielson	57.75	60.00	100.214	10.01071	3.53932	
CPB time (min)	1 ½ repair	108.50	108.50	84.500	9.19239	6.50000	0.044
	Danielson	78.50	81.00	116.286	10.78359	3.81257	
ICU duration (h)	1 ½ repair	108.50	108.50	84.5000	9.19239	6.50000	0.400
	Danielson	78.50	81.00	116.268	10.78359	3.81257	
Extubation time (min)	1 ½ repair	165.00	165.00	450.000	21.21320	15.00000	0.889
	Danielson	161.25	165.00	1012.50	31.81981	11.25000	

ICU: Intensive care unit, CPB: Cardiopulmonary by-pass.

Table-3. Operations and the Results

	Operation	ICU (h)	Extubation time (min)	X-clamp time (min)	CPB time (min)	LVEF alteration	Postop TR
1	Danielson	20	120	42	56	+5%	-
2	Carpentier+Cone+BDGS+ring implantation+pacemaker implantation (1.5 ventricle)	26	180	97	115	-	'1-2
3	Valve replacement+BDGS (1.5 ventricle)	21	150	86	102	-	-
4	Danielson	22	150	62	80	+5-10%	'1
5	Danielson	24	120	67	90	-	-
6	Danielson+cryoablation+ ring implantation	20	180	65	80	-	'1-2
7	Danielson	22	210	68	86	+5-10%	-
8	Danielson	18	180	56	82	-	-
9	Danielson+ring implantation	24	150	44	70	+5%	'1-2
10	Danielson	21	180	58	84	-	-

ICU: Intensive care unit, CPB: Cardiopulmonary by-pass, LVEF: Left ventricle ejection fraction, TR: Tricuspid regurgitation, BDGS: Bidirectional Glenn shunt

Results

There was no mortality among patients. Danielson correction was performed to eight cases while other two patients underwent one and a half ventricle repair owing to insufficient effective right ventricular volume. In two patients, concomitant ASD were repaired with autologous pericardial graft and six of them were closed via continuous suturation. Detected PDA was ligated in one case. Aiming to correcting the tricuspid regurgitation in two cases 32 and 36 sized tricuspid rings were implanted while Danielson correction.

On the other hand in patients to whom underwent one and a half repair correction (age 13 and 18), additional surgical procedures were essential as usual. In one case Cone resection and Carpentier repair were performed and 32 mm sized tricuspid ring was implanted, the correction was completed with bidirectional Glenn shunt.

Addition to these, permanent epicardial pacemaker implantation was needed in that patient. In other case tricuspid repair was non-applicable, thus 33 mm sized mitral bioprothesis valve was implanted to tricuspid position for that purpose. As a result of it in one and a half ventricle repair applied group both X-clamping (p=0.044) and CPB time (p=0.044) was longer compared to Danielson group. On the other hand there is no significant difference between groups in ICU duration (p=0.400) or extubation timing (p=0.889), (Table-2). Furthermore, need for blood product and amount of drainage were not different between groups when two infant age cases were excepted while calculating.

In the ICU, only one patient needed to inhaler nitric oxide therapy for one day. Postoperative all patients had sinus rhythm except for pacemaker implanted one. Moreover, cryoablation was utilized in one patient during Danielson repair. There was no additional complication that requiring reoperation in the hospitalization or follow-up period.

Postoperatively, in only four cases mild tricuspid regurgitation ('1-'2) was measured via TTE. LVEF increment was measured at 4 cases up to 10%. Functional class at discharge was also better in all patients when compared to preoperative results (Table-3).

Discussion

In the present study we present our surgical results of patients with Ebstein anomaly. Mostly we prefer Danielson technique if anatomical defects are feasible for repair. This method involves, plication of atrialized right ventricular part towards basis of heart from apex and anterior tricuspid annuloplasty. Hereby, tricuspid leaflets are located more physiological line and closer to annulus. Otherwise, various options should be considered.

Mayo Clinic has the largest database about this disease worldwide so far, in their cohort, they've presented 539 cases and their long term results in 2006 (8). Similarly, ASD closure rate was 83% in this study as in response to our 80% result. Their mortality ratio was 5.9%. Thereby limited patient population as previously mentioned in text, we've seen no mortality. In addition, same clinic reported their results with Cone reconstruction in another study including 84 patients (9). Cone repair mostly favored in young patients which define complete delamination septal leaflet then rotation and reattaching to the true annular zone. Subsequently, plication of atrialized right ventricular part in the cone repair technique allows a leaflet-to-leaflet coaptation in contrast to other methods. Thus, surgeons have a chance to avoid valve replacement. However, additional annuloplasty ring implanting has still controversy. Some researchers associate it with a potential tricuspid stenosis in long-term follow up (10). But Mayo clinic study concludes that ring annuloplasty is safe and decrease regurgitation. Moreover, in our study, no need of reoperation was seen due to valve stenosis. Mayo clinic results indicate a steep learning curve and pose a great success with a 98% uneventful hospital discharge. As verifying these comments, despite we are not experienced in the Cone technique, we were able to perform a smooth procedure in an adolescent case and obtained a decrement in regurgitation with annuloplasty ring utilization, moreover NYHA functional class improved.

In younger infants, biventricular repair is feasible with good early results and provides satisfying functional status. Boston and colleagues (11) have a trial including 32 patients. 29 of them underwent valve repair and ten of them were treated with Danielson method. In all patients with preoperative $^{4}/_{4}$ regurgitation ends up with ¼ and NYHA class I or II. We achieved in two infant cases similar functional status to whom applied Danielson repair.

Allen and colleagues (12) have reported 15 (3.7%) patients who required permanent pace in their study. Also in one case we had to implant permanent pace. Despite, they indicate that basically, permanent pace requirement is more often in patients who need to valve replacement, we were able to maintain a non-regurgitan valve via tricuspid ring annuloplasty.

From a different view of point, although Ebstein anomaly is naming as a right side problem, also effects left part of heart. Particularly, left ventricular dysfunction reported in several studies (13-15). The left ventricular failure is multifactorial among Ebstein patients. RV volume overload easily depress LV function by causing compression and abnormal septal motion. Correcting the RV anomaly provides an ordinary pulmonary flow and enhances and regulates LV preload. Morgan et al. (13) remarked this recovery in their study. They reported 36 TV repaired and 12 replaced and one 1.5 ventricle repaired patients and determined no worsening LV ejectional function, contrarily measured improvement in all patients. Consequently, they claim that decreasing LV function thought to be an indication to promptly restore TV competence rather than a contraindication to TV operation. We infer from our results the same view. Nevertheless, impaired ventricle function and complicated Ebstein morphology cause to consider different approaches. Quinonez et al. (16) reported bidirectional Glenn shunt constructed patients' results. They determined LV function improvement postoperatively, also suggest utilizing bidirectional Glenn shunt with a reduced LV function and severe Ebstein anomaly who are at a high risk for biventricular repair. Two cases of us with 1.5 ventricle repair showed similar results as aforementioned study.

Conclusion

To summarize, different surgical treatment options should be considered regarding to a broad range variation of Ebstein anatomy. Particularly, valve repair methods should be thought as far as possible. Danielson method is safe and effective in the repair of Ebstein anomaly and can be applied at infant age with similar clinical outcomes. Nevertheless, one and a half repair option is also beneficial in patients with a large size atrialized right ventricular volume. The last decision should be confirmed by heart team considering patient characteristics and surgeons' past experience.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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