Arch Iranian Med 2006; 9 (4): 354 – 358

Original Article

Repair of Ebstein Anomaly: Early and Mid-Term Results

Hassan Radmehr MD*, Mehrdad Salehi MD*, Seyed-Khalil Forouzan-Nia MD**, Seyed-Ali Emami MD*, Seyed-Jalil Mirhoseini MD*, Mehdi Sanatkarfar MD•*

Background: **Ebstein anomaly is the downward displacement and adherence of dysplastic septal and posterior tricuspid leaflets into the right ventricle, thereby dividing the ventricle into a so-called atrialized chamber and a functionally reduced right ventricle. We evaluated the early and mid-term results of primary repair of Ebstein anomaly in adult patients.**

Methods: **Eight consecutive patients undergoing repair of Ebstein anomaly with Danielson technique at Imam Khomeini Medical Center, Tehran, Iran from January 1997 through July 2004 were evaluated. Functional and echocardiographic parameters were studied both preoperatively and postoperatively, as well as demographic status and adverse events.**

Results: **Hospital mortality occurred in one patient because of right ventricular failure. The average follow-up period was 5.3 ± 3.4 years (median: 3.8 years). The actuarial survival rate was 85.7 ± 4.8% at 7 years. During the follow-up, six patients were in New York Heart Association functional class I, and one patient was in class II. None of the patients required reoperation related to their Ebstein repair. One patient demonstrated atrioventricular dissociation perioperatively; however, only one patient required permanent pacemaker insertion later. One patient had minimal (1^+) regurgitation, with the jet localized along the anterior part of the ventricular septum. Two patients had residual tricuspid valve insufficiency (2^+) on echocardiography.**

Conclusion: **Ebstein repair has a good functional and hemodynamic outcome in adult patients.**

Archives of Iranian Medicine, Volume 9, Number 4, 2006: 354 – 358.

Keywords: Ebstein anomaly • functional class • tricuspid valve • ventricular failure

Introduction

Ebstein anomaly is the downward displacement and adherence of dysplastic septal and posterior tricuspid leaflets into the right ventricle, thereby dividing the ventricle into a so-called atrialized chamber and a functionally reduced right ventricle. Ebstein anomaly may manifest at any age. Its clinical presentation encompasses a spectrum of severity from death in untreated symptomatic neonates to long-term survival in some adults with or without repair.[1 – 4]

In adult patients, symptoms typically represent the effects of ongoing tricuspid regurgitation (TR) and right ventricular (RV) dysfunction. At any age, rhythm abnormalities are the cause of significant morbidity and sudden death.[5 – 7] During the past 2 decades, several innovative techniques have been promoted as alternatives to valve replacement for repair of Ebstein anomaly. Most of these techniques involve tricuspid valve restoration and some degree of plication of the atrialized ventricle.[8 – 17]

The aim of this study was to evaluate the outcomes of the patients who underwent repair of Ebstein anomaly in our center during a 7-year period.

Materials and Methods

From January 1997 through July 2004, eight patients who underwent repair of Ebstein anomaly in our center were included in this study. The mean age of the patients was 23.8 ± 10 years

Authors' affiliations: Department of Cardiovascular Surgery, *Tehran University of Medical Sciences, Tehran, **Yazd University of Medical Sciences, Yazd, Iran.
•**Corresponding author and reprints:** Mehdi Sanatkarfar MD, Department of Cardiovascular Surgery, Imam Khomeini Medical Center, Tehran University of Medical Sciences, Tehran, Iran. Fax: +98-21-66929977, E-mail: mehdi_sanatkar@yahoo.com.
Accepted for publication: 16 November 2005

(range: 13 – 32).

We evaluated the patients' characteristics such as age, gender, indication for surgical correction, associated lesions, preoperative radiographic cardiothoracic ratio, exercise testing (when available), presence of preoperative arrhythmia, and previous procedures.

Preoperative and postoperative echocardiograms were analyzed by one blinded cardiologist according to a modified grading scale ("Ebstein Severity Scale") , which was described by Carpentier and colleagues and Quaegebeur and colleagues.[9, 17] The echocardiographic analysis included parameters of right and left ventricular function, severity of TR, and degree of anterior tricuspid leaflet mobility. In addition, in postoperative studies, the level of displacement of the tricuspid apparatus was assessed.

Based on New York Heart Association (NYHA) functional class, before the operation two patients categorized in class II (25 %), five patients (62.5%) in class III, and one patient (12.5%) in class IV.

The mean cardiothoracic ratio was 0.65 ± 0.06 (0.5 – 0.8). Tricuspid valve insufficiency (TVI) was assessed by echocardiography and/or angiocardiography and graded from 1^+ to 4^+. TVI was 2^+ in one patient (12.5%), 3^+ in four patients (50%), and 4^+ in two patients (25%). Tricuspid valve stenosis was not present. The most frequent associated anomaly was atrial septal defect (ASD). None of the patients had undergone any previous operation.

Indications for surgery were functional disability in 6 cases (75%), cyanosis in one case (12.5%), and rhythm disturbances not improved by medical treatment in one case (12.5%).

All patients were evaluated clinically before the operation and followed up for a maximum of 7 years postoperatively. Routine transthoracic echocardiography was obtained before hospital discharge, after 6 months, and annually. Additional transthoracic echocardiography and exercise tests were obtained later postoperatively at the discretion of the referring cardiologist.

The goals of the surgery were to restore normal tricuspid function and to preserve RV contractility.

Valve repair technique

The operation was performed by median sternotomy. Total cardiopulmonary bypass was established by aortic and bicaval cannulation and tourniquets around the inferior and superior vena cava (IVC & SVC). Moderate hypothermia (25 – 28ºC) and cardioplegic arrest by cold crystalloid solution were accomplished. The right atrium was opened by an oblique incision from the right atrium towards IVC.

In six patients, repair by plication of the atrialized portion of the RV and posterior annuloplasty was performed (Danielson method) (Figure 1). The repair was successful in five patients, but failed in one patient (28-year-old female); therefore, tricuspid valve replacement (TVR) was done for her.

In two other patients, because of the deformity of the anterior leaflet, TVR was planned and performed by mechanical prosthetic valves.

The ASDs were also closed. There was not any ventricular septal defect, mitral valve insufficiency, pulmonary stenosis, and subaortic stenosis in our patients.

Two patients underwent TVR by mechanical prosthetic valves. Indications for replacement were: total adherence of the leaflet tissue to the ventricular wall (type D) in one of the patients, and partial atrioventricular (AV) defect with hypoplasia of the leaflet tissue in the other one (Figure 1).

Intraoperative radiofrequency ablation

In one patient with documented supraventricular arrhythmias preoperatively, we performed intraoperative radiofrequency ablation procedure (modified right-sided Maze) using a RF Cobra probe. In this case, we paid particular attention to avoid the fibers of the conduction

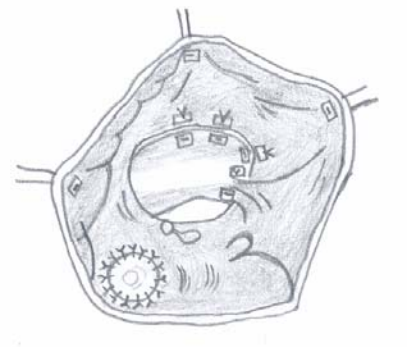

Figure 1. Operative view (final result).

system, especially during the creation of the so-called isthmus block (IVC-tricuspid annulus). We started intravenous amiodarone for this patient and switched it to oral amiodarone later. This regimen was continued for the first 3 postoperative months. Those patients on antiarrhythmic regimens preoperatively continued their medications postoperatively.

Statistical analysis

Hospital mortality was defined as death during the surgery or within 30 days after the surgery. The Kaplan-Meier method was used to calculate the actual survival and the probability of remaining free from reoperation. Continuous variables were evaluated using Student's t test and $P < 0.05$ was considered significant. The association between qualitative risk factors and perioperative death was evaluated by χ^2 test.

Results

Eight patients underwent primary repair. The average follow-up period was 4.3 ± 2.7 years. Patients' demographics are summarized in Table 1.

Hospital mortality occurred in one patient, a 32-year-old female patient with NYHA class IV who died in operating room. In this patient, the RV wall was very thin.

One patient (12.5%) demonstrated AV dissociation prior to the operation; however, only one patient (12.5%) required permanent pacemaker insertion. The remaining patients maintained normal sinus rhythm throughout the perioperative and postoperative periods. Nonlethal perioperative complications were arrhythmia in two cases (atrial fibrillation in one and paroxysmal atrial contraction (PAC) in the other one), who needed inotropic support in one case after operation, and external pacemaker in another case (Table 2). None of our patients died during the follow-up period. The actuarial survival rate was 85.7 ± 4.8% during follow-up.

Table 2. Postoperative complications.

Postoperative complications	Number
Infection	0
Bleeding	0
Renal failure	0
GI	0
Neurologic	0
Respiratory	0
Need for inotropic medications	1
Temporary pacemaker	1
Arrhythmia	2
Reoperation	0

Among the seven patients who were followed up for more than one year, six patients were in functional class (NYHA) I, and one patient was in class II. None of the patients required reoperation for the reasons related to their Ebstein repair. The tricuspid valve function was assessed by echocardiography at the last follow-up. One patient had minimal (1^+) regurgitation, with the jet localized along the anterior part of the ventricular septum. Two patients had residual tricuspid valve insufficiency (2^+) on echocardiography.

Tricuspid valve stenosis resolved in all patients. At 6-month and 1-year follow-ups, none of these patients had arrhythmia or were maintained on oral antiarrhythmic medications. Ejection fraction of the effective RV decreased after surgery from 55 ± 15% to 50% ± 10 ($P < 0.01$). Ejection fraction of the left ventricle increased after surgery from 50 ± 12% to 55 ± 15% ($P < 0.01$).

Discussion

It is well known that Ebstein anomaly has a wide clinical and anatomic spectrum.[18 – 20] Ebstein anomaly represents an unusual congenital cardiac lesion with variable clinical presentation. The characteristic anatomic feature of the disease is the downward displacement and adherence of dysplastic septal and posterior tricuspid leaflets

Table 1. Demographic characteristics of the patients.

Patients	Sex	Age	Indication for operation	Associated anomalies	NYHA class	Follow-up (yr)
1	F	13	Cyanosis, severe TR	ASD	II	3.4
2	F	15	Severe TR	ASD	III	4.5
3	F	32	CHF, severe TR	ASD	IV	Died
4	F	28	Severe TR	ASD	III	7
5	M	18	Severe TR	ASD	III	4
6	M	25	Severe TR	ASD	II	6.2
7	M	28	Severe TR	ASD	III	5
8	M	32	Severe TR, arrhythmia	ASD	III	7

CHF = congestive heart failure.

into the RV, thereby dividing the ventricle into a so-called atrialized chamber and a functionally reduced RV.[1 – 4]

Factors involved in tricuspid valve insufficiency are the enlargement of the tricuspid annulus, the restricted and downward septal leaflet, and the absence or restricted posterior leaflet.[21] In addition, the anterior leaflet is rarely normal and tethering is frequent.[18] The large anterior leaflet of the tricuspid valve may demonstrate restricted mobility because of muscular trabeculations to the RV free wall. Whereas early repairs sought primarily to address the essential problem of TR, recent techniques have each concentrated on reconstruction of these essential deformities: liberation of the anterior leaflet from its adhesions, exclusion or reduction of the atrialized ventricle, reinstitution of valvular integrity and competency by reattaching the anterior leaflet to the new annulus, and remodeling of the tricuspid valve annulus and inflow portion of the RV.[8 – 17]

In neonates with Ebstein anomaly, recent reports of successful outcomes with complete repair has challenged the notion that functional univentricular palliation is the only therapeutic option for these patients.[22 – 24]

Improved outcomes with repair during the past 2 decades have decreased the deterioration of right-sided heart function, TR, and potential development of arrhythmogenic foci.[1, 25, 26]

The goal of reconstructive valvular surgery is to restore a normal surface of cooptation of the leaflet tissue. In Ebstein anomaly, the anterior leaflet is the only effective leaflet capable of reaching the ventricular septum. Subsequently, the surface of cooptation is between the anterior leaflet and the ventricular septum. Techniques of valve enlargement have been developed with some success, but unknown long-term results.[27] Repair of the septal leaflet is technically demanding.[28] Mobilization of the anterior leaflet is based on the concept of a restricted anterior leaflet.[23]

The surgical limitation of valve repair is Carpentier type D disease, in which the individualization of the anterior leaflet from the RV wall is not complete. Valvular replacement in such cases appears prudent and effective.[18, 29] Ebstein anomaly is also a RV disease,[30] and most of the operative mortality is related to underestimation of RV contractility. Decreasing the RV's preload is an important part in the management of depressed RV contractility.[31 – 33]

Approximately 79% of patients with Ebstein anomaly demonstrate supraventricular or ventricular arrhythmias. Sudden death has been reported to occur in up to 20% of these patients.[34, 35] Although the pathologic anatomy in Ebstein anomaly is thought to provide an ideal substrate for aberrant conductive pathways, the appropriate treatment for their ablation remains unclear.

Another procedure for repair of Ebstein anomaly is Carpentier technique. In this technique, after detaching the ¾ of anterior and most of the posterior leaflet from the annulus, plication of the atrialized portion of the RV will be done vertically and the detached leaflet will be reattached to the anomaly by clockwise rotation. Finally, Carpentier ring will be placed to prevent recurrent TR. The result of this technique is as good as Danielson method. When the repair cannot be done successfully, TVR by bioprosthesis or mechanical valve is necessary.

We undertook the current study to evaluate our medium-term outcomes of repair of Ebstein anomaly at a single institution. Our technique for repair was not varied substantially during the 7-year period but was modified specifically with regard to patient age.

We concluded that repair of Ebstein anomaly by technique of Danielson is effective and the complications after operation and during follow-up were low and acceptable. This procedure can be very helpful for adult patient who suffer from Ebstein anomaly.

References

1 Mair DD. Ebstein's anomaly: natural history and management. *J Am Coll Cardiol*. 1992; 19: 1047 – 1048.

2 Attie F, Rosas M, Rijlaarsdam M, Alfonson B, Zabal C, Kuri J, et al. The adult patient with Ebstein anomaly: outcome in 72 unoperated patients. *Medicine (Baltimore)*. 2000; 79: 27 – 36.

3 Celermajer DS, Bull C, Till JA, Cullen S, Vassillikos V, Sullivan ID, et al. Ebstein's anomaly: presentation and outcome from fetus to adult. *J Am Coll Cardiol*. 1994; **23:** 170 – 176.

4 Spitaels SEC. Ebstein's anomaly of the tricuspid valve complexities and strategies. *Cardiol Clin*. 2002; **20:** 431 – 439.

5 Lo H, Lin F, Jong Y, Tseng Y, Wu T. Ebstein's anomaly with ventricular tachycardia: evidence for the arrhythmogenic role of the atrialized ventricle. *Am Heart J*. 1989; 117: 959 – 962.

6 Cappato R, Schlüter M, Weiss C, Antz M, Kopschyk DH, Hofman T, et al. Radiofrequency current catheter ablation of accessory atrioventricular pathways in Ebstein's anomaly. *Circulation*. 1996; **94:** 376 – 383.

7 Oh J, Holmes DR, Hayes DL, Porter CJ, Danielson GK.

Cardiac arrhythmias in patients with surgical repair of Ebstein's anomaly. *J Am Coll Cardiol*. 1985; **6:** 1351 – 1357.

8 DiRusso GB, Gaynor JW. Ebstein's anomaly: indications for repair and surgical technique. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu.* 1999; **2:** 35 – 50.

9 Carpentier A, Chauvaud S, Macé L, Relland J, Mihaileanu S, Marino JP, et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg.* 1988; **96:** 92 – 101.

10 Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC. Operative treatment of Ebstein's anomaly. *J Thorac Cardiovasc Surg*. 1992; **104:** 1195 – 1202.

11 Kupilik N, Simon P, Moidl R, Wollenek G, Marx M, Wolner E, et al. Valve-preserving treatment of Ebstein's anomaly: perioperative and follow-up results. *Thorac Cardiovasc Surg*. 1999; **47:** 229 – 234.

12 Mair DD, Seward JB, Driscoll DJ, Danielson GK. Surgical repair of Ebstein's anomaly: selection of patients and early and late operative results. *Circulation.* 1985; **72 suppl II:** II70 – II76.

13 Marianeschi SM, McElhinney DB, Reddy VM, Silverman NH, Hanley FL. Alternative approach to the repair of Ebstein's malformation: intracardiac repair with ventricular unloading. *Ann Thorac Surg.* 1998; **66:** 1546 – 1550.

14 Wu Q, Huang Z. Anatomic correction of Ebstein anomaly. *J Thorac Cardiovasc Surg*. 2001; **122:** 1237 –1238.

15 Augustin N, Schmidt-Habelmann P, Wottke M., Meisner H, Sebening F. Results after surgical repair of Ebstein's anomaly. *Ann Thorac Surg*. 1997; **63:** 1650 – 1656.

16 Hetzer R, Nagdyman N, Ewert P, Weng YG, Alexi-Meskhisvilli V, Berger F, et al. A modified repair technique for tricuspid incompetence in Ebstein's anomaly. *J Thorac Cardiovasc Surg*. 1998; **115:** 857 – 868.

17 Quaegebeur JM, Sreeram N, Fraser AG, Bogers AJJC, Stümper OFW, Hess J, et al. Surgery for Ebstein's anomaly: the clinical and echocardiographic evaluation of a new technique. *J Am Coll Cardiol*. 1991, **17:** 722 – 728.

18 Anderson RH, Vogel M, Ho SY. The functional anatomy of Ebstein's malformation of the tricuspid valve. In: Yacoub M, ed. *Annual of Cardiac Surgery*. London, Philadelphia: Current Science; 1995: 157 – 163.

19 Zuberbuhler JR, Allwork SP, Anderson RH. The spectrum of Ebstein's anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg*. 1979; **77:** 202 – 211.

20 Schreiber C, Cook A, Ho SY, Augustin N, Anderson RH. Morphologic spectrum of Ebstein's malformation: revisitation relative to surgical repair. *J Thorac Cardiovasc Surg*. 1999; **117:** 148 – 155.

21 Oechslin E, Buchholz S, Jenni R. Ebstein's anomaly in adults: Doppler-echocardiographic evaluation. *J Thorac Cardiovasc Surg*. 2000; **48:** 209 – 213.

22 Knott-Craig CJ, Overholt ED, Ward KE, Razook JD. Neonatal repair of Ebstein's anomaly: indications, surgical technique, and medium-term follow-up. *Ann Thorac Surg*. 2000; **69:** 1505 – 1510.

23 Knott-Craig CJ, Overhotld ED, Ward KE, Ringewald JM, Baker SS, Razook JD. Repair of Ebstein's anomaly in the symptomatic neonate: an evolution of technique with 7-year follow-up. *Ann Thorac Surg*. 2002; **73:** 1786 – 1793.

24 Starnes VA, Pitlick PT, Berstein D, Griffin ML, Choy M, Shumway NE. Ebstein's anomaly appearing in the neonate. *J Thorac Cardiovasc Surg*. 1991; **101:** 1082 – 1087.

25 Barber G, Danielson GK, Heise CT, Driscoll DJ. Cardiorespiratory response to exercise in Ebstein's anomaly. *Am J Cardiol*. 1985; **56:** 509 – 514.

26 Driscoll DJ, Mottram CD, Danielson GK. Spectrum of exercise intolerance in 45 patients with Ebstein's anomaly and observation on exercise tolerance in 11 patients after surgical repair. *J Am Coll Cardiol*. 1988; **11:** 831 – 836.

27 van Son J, Kinzel P, Mohr F. Pericardial patch augmentation of anterior tricuspid leaflet in Ebstein's anomaly. *Ann Thorac Surg.* 1998; **66:** 1831 – 1832.

28 Kaneko Y, Okabe H, Nagata N, Yasui S, Yamada S, Kobayashi J, et al. Repair of septal and posterior tricuspid leaflets in Ebstein's anomaly. *J Cardiol Surg*. 1998; **13:** 229 – 235.

29 Shauvaud S, Berrebi A, d'Attellis N, Mousseaux E, Hernigou A, Carpentier A. Ebstein's anomaly: repair based on functional analysis. *Eur J Crdiothorac Surg*. 2003; **23:** 525 – 531.

30 Celermajer DS, Bull C, Till JA, Cullen S, Vassillikos VP, Sullivan ID, et al. Ebstein's anomaly: presentation and outcome from fetus to adult. *J Am Coll Cardiol*. 1994; **23:** 170 – 176.

31 Chauvaud S, Fuzellier JF, Berrebi A. Bidirectional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high-risk patients. *Eur J Cardiothorac Surg*. 1998; **13:** 514 – 519.

32 Marianeschi SM, McElhinney DB, Reddy VM, Silverman NH, Hanley FL. Alternative approach to the repair of Ebstein's malformation: intracardiac repair with ventricular unloading. *Ann Thorac Surg*. 1998; **66:** 1546 – 1550.

33 Kreutzer C, Mayorquim RC, Kreutzer GO, Conejeros W, Roman MI, Vazquez H, et al. Experience with one and a half ventricle repair. *J Thorac Cardiovasc Surg.*1999; **117:** 662 – 668.

34 Chauvaud SM, Brancaccio G, Carpentier AF. Cardiac arrhythmia in patients undergoing surgical repair of Ebstein's anomaly. *Ann Thorac Surg*. 2001; **71:** 1547 –1552.

35 Misaki T, Watanabe G, Iwa T, Watanabe Y, Mukai K, Takahashi M, et al. Surgical treatment of patients with Wolf-Parkinson-White syndrome and associated Ebstein's anomaly. *J Thorac Cardiovasc Surg*. 1995; **110:** 1702 – 1707.