Journal of the American College of Cardiology
© 2012 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Vol. 59, No. 23, 2012
ISSN 0735-1097/$36.00
doi:10.1016/j.jacc.2012.03.020

Outcome of Cardiac Surgery in Patients 50 Years of Age or Older With Ebstein Anomaly

Survival and Functional Improvement

Christine H. Attenhofer Jost, MD,* Heidi M. Connolly, MD,† Christopher G. Scott, MS,‡ Harold M. Burkhart, MD,§ Carole A. Warnes, MD,† Joseph A. Dearani, MD§

Zurich, Switzerland; and Rochester, Minnesota

Objectives This study sought to analyze the presentation, surgical procedures, and outcomes in patients ≥50 years of age with Ebstein anomaly (EA).

Background Data on management and surgical outcomes in older patients with EA are limited.

Methods Operative and clinical data from patients with EA ≥50 years of age undergoing cardiac surgery at our center between October 1980 and January 2010 were analyzed.

Results During the study period, 89 procedures were performed in 81 patients with EA (63% women; mean [range] age 59 [50 to 79] years). Pre-operative symptoms included palpitations (n = 69), edema (n = 30), and previous stroke/transient ischemic attack (n = 21). Seventy-six patients (85%) had functional class III or IV symptoms, and 13 (16%) had previous cardiac surgery. Tricuspid valve surgery was necessary in 87 of the 89 procedures (98%): replacement in 65 (73%) and repair in 22 (25%). Three early deaths occurred (4%). On long-term follow-up (available in 73 of 78 early survivors), 63 patients (89%) had improved functional class and 13 patients died (19%). The 20-year survival was 65% versus 74% for age- and sex-matched controls (p = 0.001). The best predictors of late death were lack of post-operative improvement and older age at surgery.

Conclusions Although cardiac surgery in patients with EA ≥50 years of age was often complex, early mortality was low (4%) when surgery was performed at an experienced center. Long-term survival was good, although less than expected. These data suggested that surgery in older patients with EA may have to be performed earlier. (J Am Coll Cardiol 2012;59:2101–6) © 2012 by the American College of Cardiology Foundation

Ebstein anomaly (EA), a rare congenital heart malformation, occurs in approximately 1 to 5 per 200,000 live births (1). EA encompasses a wide anatomic spectrum of abnormalities of the tricuspid valve (TV) leaflets and right ventricle (RV), including atrialization due to apical displacement of the tricuspid annulus. Common associated anomalies include atrial septal defect (ASD) and patent foramen ovale, occurring in ≤90% of patients, and ventricular pre-excitation in approximately 15% of cases (2). Many patients with EA survive and require repair in adulthood (3). Age at clinical presentation varies considerably by anatomic severity and by other associated heart diseases. Clinical symptoms are age dependent and include cyanosis, right-sided heart failure, arrhythmias, decreased exercise tolerance, fatigue, sudden cardiac death, and paradoxical emboli (4). Optimal timing of intervention is often difficult and must be individualized (3,5–11).

Few data have been published on the operative outcomes of patients with EA who are ≥50 years of age at the time of surgery (11,12). Therefore, we sought to analyze clinical presentation, surgical interventions, and outcomes of all adults with EA at our institution who underwent operative repair at ≥50 years of age.

Methods

We retrospectively searched our database for patients ≥50 years of age who underwent operations for EA at Mayo Clinic (Rochester, Minnesota) between October 1980 and January 2010. Before 1980, no patients with EA in this age group had TV operations. The diagnosis of EA was based on echocardiography in all patients. Inclusion criteria were atrioventricular and ventriculoarterial concordance and 2 ventricles; we excluded patients with pulmonary atresia and

From the *Cardiovascular Center Zurich, Zurich, Switzerland; †Division of Cardiovascular Diseases, Mayo Clinic, Rochester, Minnesota; ‡Division of Biomedical Statistics and Informatics, Mayo Clinic, Rochester, Minnesota; and the §Division of Cardiovascular Surgery, Mayo Clinic, Rochester, Minnesota. All authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Manuscript received September 1, 2011; revised manuscript received February 23, 2012, accepted March 6, 2012.

Abbreviations and Acronyms

ASD = atrial septal defect
EA = Ebstein anomaly
LV = left ventricular
LVEF = left ventricular ejection fraction
RV = right ventricle/ventricular
TV = tricuspid valve

complex conotruncal abnormalities. The indications for surgery in patients with EA included dyspnea, heart failure, tachyarrhythmias not amenable to another therapy, ASD, progressive cardiomegaly, and other associated lesions. The Mayo Clinic Institutional Review Board approved this study. All patients gave informed consent to participate according to the guidelines of our ethical committee.

Operative technique. Operative management of EA included: 1) pre-operative electrophysiological mapping for localization and ablation of accessory conduction pathways in patients with ventricular pre-excitation; 2) closure of an interatrial communication, if present; 3) elimination of previously placed shunts and repair of associated anomalies; 4) performance of any indicated antiarrhythmia procedures; 5) selective plication of the atrialized RV; 6) reconstruction of the TV or TV replacement; and 7) excision of redundant right atrial wall (2,3,13). Our TV repair techniques for EA have evolved during the past 30 years (14).

Tricuspid valve surgery. Various modifications of the TV repair procedure were used to address the numerous variants of EA (3,13); in general, the procedures consisted of a monocusp repair at the level of the functional annulus. Plication was reserved for atrialized RV that was thinned and dyskinetic.

When TV replacement was performed, anterior leaflet tissue toward the RV outflow tract was excised to avoid potential RV outflow tract obstruction after bioprosthetic valve replacement (15). The position of the os of the coronary sinus relative to the prosthesis (draining into the right atrium or RV) was determined by the proximity of the coronary sinus to the conduction tissue. When there was sufficient distance between the 2 structures, the bioprosthesis was positioned so that the coronary sinus drained into the right atrium (i.e., the prosthesis went between the conduction tissue and the coronary sinus). When the 2 structures were close together, the prosthesis was positioned to the atrial side of both to avoid heart block. In this situation, the coronary sinus drained into the RV. Injury to the right coronary artery was avoided by deviating the suture line cephalad to the true TV annulus anteriorly and posterolaterally. The struts of the bioprosthetic valve were oriented so that they straddled the membranous septum and conduction tissue.

Antiarrhythmia surgery. In the earlier years of surgery, intraoperative electrophysiological mapping for localization and surgical division or cryoablation of the pathways were performed in patients with accessory conduction pathways. In the current era, pre-operative electrophysiological mapping and percutaneous ablation of accessory conduction pathways is performed. Paroxysmal and persistent atrial fibrillation are treated with a modified cut-and-sew Cox maze III procedure. In the presence of pre-operative atrial flutter, the right atrial isthmus is cryoablated as well. In the latter part of this series, cryoablation was applied from the amputated right atrial appendage to the TV annulus to decrease the possibility of damaging the arterial blood supply to the sinoatrial node. We currently prefer cryoablation to electrocautery at our center.

Statistical analysis. Demographic and other patient-related data were obtained from medical records. Follow-up information was obtained from clinic visits, correspondence from local physicians, mailed questionnaires, and Social Security Death Index (in all patients). Data are expressed as mean ± SD or number of patients (percentage). $p < 0.05$ was considered statistically significant. Early operative mortality was defined as death at any time during the index hospitalization or within 30 days of operation. Survival was estimated by using the Kaplan-Meier method and was compared with an age- and sex-matched population using the 1-sample log-rank test. Within the population of operative survivors, variables associated with survival were evaluated using Cox proportional hazards regression methods, and results are given as hazard ratios with 95% confidence intervals.

Results

We identified 81 patients (10 patients before 1990) who underwent 89 procedures for EA during the study period (1 patient had 3 operations, and 6 patients had 2 operations); the average age at operation was 59 years (range 50 to 79 years). The clinical characteristics of the patients are shown in Table 1. Average age at EA diagnosis was 43 years; 11 patients were >60 years of age at the time of diagnosis. Most patients were markedly symptomatic: 85% were in functional class III or IV at surgery. The most common symptoms included dyspnea, palpitations, cyanosis, and

Table 1 Clinical Characteristics

Patient characteristics (n = 81)	
Age at diagnosis, yrs	43 ± 19
Women	51 (63)
Pre-procedural characteristics (n = 89)	
Patient age at surgery, yrs	59 ± 8
New York Heart Association functional class	
I/II	13 (15)
III/IV	76 (85)
Dyspnea	74 (83)
Palpitations	69 (78)
Edema	30 (34)
Cyanosis	21 (24)
Stroke/TIA	21 (24)
Dizziness	16 (18)
Right-sided heart failure	13 (15)
Syncope	7 (8)
Clubbing	6 (7)

Values are mean ± SD or n (%).
TIA = transient ischemic attack.

peripheral edema (Table 1). The 46 women with prior pregnancies had of 1.7 ± 1.2 children. The hemoglobin value was 14.9 ± 2.0 g/dl (range 10.8 to 19.7 g/dl). Pre-operative left ventricular ejection fraction (LVEF) was 57% (11%) and was <50% in 23 patients (26%). A previous stroke or transient ischemic attack had occurred in 21 patients (24%).

Prior cardiac surgery had been performed in 13 patients (16%) 7 to 40 years previously. Prior operations included ASD closure in 10 patients, TV repair in 2 patients, and TV replacement in 2 patients.

Most patients were in sinus rhythm pre-operatively. Paroxysmal or persistent atrial fibrillation was present in 19 patients (21%) and pre-excitation in 7 patients (8%). Four patients had pacemakers, and 1 patient had an automatic internal cardiac defibrillator. Moderate or severe mitral regurgitation was present in 9 patients (10%). Cardiovascular risk factors included arterial hypertension in 29 patients (33%), hyperlipidemia in 29 patients (33%), smoking history in 18 patients (20%), and diabetes mellitus in 4 patients (4%). Of 75 patients (84%) who underwent coronary angiography, coronary artery disease was found in 15 patients (20%).

Surgical procedures performed are shown in Table 2, with prosthetic TV types shown in Table 3. Additionally, pacemaker-lead thrombus was removed from 3 patients at operation. One patient each had left atrial appendage ligation, excision of a blood cyst from the TV, and resection of a left ventricular (LV) aneurysm due to myocardial infarction. The cardiopulmonary bypass time was 112 ± 49 min (range 39 to 242 min), and mean cross-clamp time was 50 (29) min.

Two patients had a bidirectional cavopulmonary shunt as an adjunct to TV replacement. A 57-year-old woman with severely reduced RV function and mildly reduced LVEF (43%) remained severely symptomatic post-operatively during a 33-month follow-up. The other patient was a 56-year-old man with severely reduced RV function and mildly reduced LVEF (48%). His symptoms improved during a 38-month follow-up. Neither patient had symptoms attributable to increased superior vena cava pressure.

Table 2 Surgical Procedures Performed (N = 89)

Procedure Type	No. of Procedures (%)
Tricuspid valve operation	
Repair	22 (25)
Replacement	65 (73)
None	2 (2)
ASD/PFO closure	58 (65)
Right reduction atrioplasty	41 (46)
Anterior right pericardectomy	18 (20)
Plication of atrialized RV	12 (13)
Right-sided maze procedure	18 (20)
Ablation of accessory pathway	9 (10)
CABG	9 (10)
Mitral valve surgery	6 (7)
Repair	4 (5)
Replacement	2 (2)
Bidirectional Glenn operation	2 (2)
Aortic root surgery	2 (2)
Repair of PS	1 (1)
Permanent pacing	1 (1)

ASD = atrial septal defect; CABG = coronary artery bypass grafting; PFO = patent foramen ovale; PS = pulmonary stenosis; RV = right ventricle.

Table 3 Types of TV Prostheses Used for TV Replacement (n = 65)

Mechanical Starr-Edwards ball-cage prosthesis, model*	7 (11)
4M	1
5M	6
Mechanical bileaflet prosthesis, size (mm)	13 (20)
31	5
33	8
Porcine BPV, size (mm)	45 (69)
25	1
29	1
31	2
33	8
35	33

Values are n (%). *Used before 1993.
BPV = bioprosthetic TV prosthesis; TV = tricuspid valve.

Three patients (4%) died early, all with operations before 1995. A 72-year-old man died during TV repair, suture closure of an ASD, coronary artery bypass surgery, and resection of an LV aneurysm. A 71-year-old man died early post-operatively of ventricular arrhythmias after undergoing TV replacement with a 33-mm mechanical St. Jude prosthesis. A 65-year-old man with diabetes mellitus died on post-operative day 4 of a myocardial infarction and pulmonary embolism.

Long-term follow-up and survival. Follow-up was available for 73 of 78 survivors (94%); median follow-up was 84 months (range 2 to 404 months). Follow-up was confirmed by clinical evaluation in 33 patients, death notice in 12 patients, research questionnaire in 9 patients, outside physician report in 11 patients, or another contact in 8 patients.

For the 71 patients with clinical follow-up, improvement in functional class occurred in 63 patients (89%). Post-operatively, only 8 patients remained in functional class III or IV (11%). Reoperation was necessary in 8 patients (7 performed at our center). Reasons for reoperation (>1 reason possible) included TV replacement for recurrent tricuspid regurgitation (n = 3), TV replacement for prosthetic dysfunction (n = 4), TV re-repair for recurrent tricuspid regurgitation (n = 1), aortic valve replacement for aortic stenosis (n = 1), and mitral valve replacement for mitral regurgitation (n = 1). Freedom from reoperation is shown in Figure 1.

Of the 78 operative survivors, 13 died during follow-up (17%). Significant predictors of death by univariate analysis were lack of post-operative improvement ($p < 0.001$), older age at surgery ($p = 0.007$), pre-operative LVEF <50% ($p = 0.02$), diabetes mellitus ($p = 0.02$), and pre-operative history of heart failure ($p = 0.01$) (Table 4). Each of these

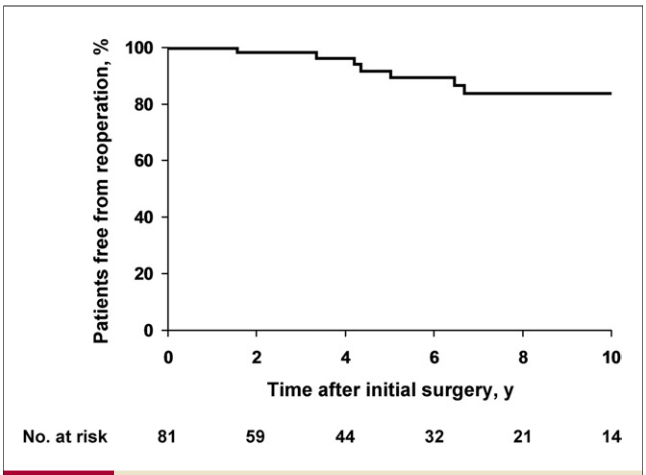

Figure 1 Kaplan-Meier Curve Showing Freedom From Reoperation

Freedom from tricuspid valve repair or replacement in the 81 study patients.

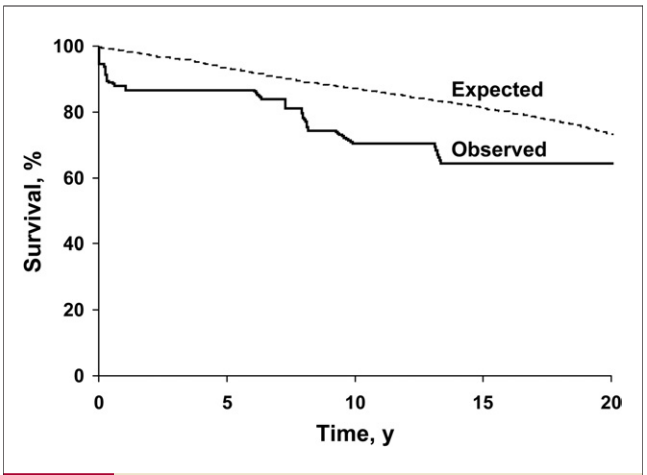

Figure 2 Kaplan-Meier Survival Curve

Overall survival of study patients (n = 81) versus expected survival ($p < 0.001$).

predictors remained significant after adjustment for age; however, no other predictors were significant after adjustment for lack of post-operative improvement. Observed long-term survival was lower than expected ($p = 0.001$) (Fig. 2). Actuarial 10-year survival was 71% vs. 88% expected, and actuarial 20-year survival was 65% vs 74% expected for an age- and sex-matched population.

Discussion

In our patients with EA undergoing surgery after age 50 years, early mortality was 4%; post-operative improvement was noted in most patients. Long-term survival was acceptable, although less than expected in an age- and sex-matched population. This study suggested that cardiac repair should be considered in patients with EA who are older and have symptoms or advanced heart disease.

Clinical presentation. In this age group, a high percentage of patients underwent operation because they were symptomatic—89% were in New York Heart Association functional class III or IV. Palpitations, edema, cyanosis, and a history suggestive of paradoxical emboli were common and most likely caused by advanced right-sided heart disease and high right atrial pressures.

Table 4 Univariate Predictors of Death During Follow-up

Parameter	HR (95% CI)	p Value
No post-operative improvement	10.0 (3.01–33.4)	<0.001
Pre-operative history of heart failure	4.42 (1.42–13.7)	0.01
Pre-operative LVEF <50%	3.59 (1.20–10.7)	0.02
Diabetes mellitus	6.76 (1.43–31.9)	0.02
Pulmonary hypertension	1.21 (0.36–4.08)	0.75
History of atrial fibrillation	0.95 (0.26–3.46)	0.94
Tricuspid valve replacement	1.63 (0.44–5.99)	0.46
Male	1.96 (0.65–5.89)	0.23
Age at surgery, per 10 yrs	2.53 (1.28–5.00)	0.007

CI = confidence interval; HR = hazard ratio; LVEF = left ventricular ejection fraction.

Tricuspid valve replacement. Because the myocardium in EA is myopathic, it is important to eliminate most of the tricuspid regurgitation so that RV remodeling can be optimized. Although we generally prefer TV repair for patients with EA, we have a lower threshold to proceed with TV replacement in older patients, particularly if severe RV or tricuspid annular enlargement or pulmonary hypertension is present because these findings have been correlated with poor TV repair durability. In our experience, TV replacement provides the best outcome in older patients with EA and has been performed with low early mortality and excellent durability, comparable to our results in 539 patients with EA undergoing surgery at a median age of 24 years (3). TV replacement was performed in 65 of 89 operations (73%) in this series.

Our reported experience notes that valve repair and replacement have similar freedoms from reoperation for recurrent TV problems (15). In the current series, 45 of 65 patients (69%) who had TV replacement received a bioprosthesis. Porcine bioprosthetic TVs are preferred to bovine pericardial prostheses because the latter are stiffer, which results in an increased transvalvular gradient, reduced cusp mobility, and possible late thrombus formation (15). The use of mechanical valves has decreased substantially in patients with EA at our institution because of the increased risk of thrombosis due to decreased RV function and low right atrial and RV pressures, which results in abnormal prosthetic disc motion, even in the presence of adequate warfarin anticoagulation (15). Therefore, even in patients with EA with atrial fibrillation, we have preferred bioprosthetic valve replacement. We have not identified any late problems with the coronary sinus draining into the RV. Questions may arise about the potential detrimental effect on LV function because of increased coronary sinus pressure when it drains into the RV. Analysis of matched patients with EA in our practice, comparing coronary sinus drainage into the right atrium versus the

RV, has shown no difference in LV function at late follow-up (Connolly HM, unpublished data).

Plication or resection of an atrialized RV is performed only if it is thinned out and transparent. We do not believe that this type of ventricular wall contributes to RV function (3). In this series of older patients with EA, plication of the atrialized RV was performed in only 13% of patients.

TV repair. Numerous TV repair techniques (14,16–18) have been described; the most common types include cone modification, Carpentier technique, and modifications of the Danielson technique (17,19,20). Thirteen patients in the current series had operations after the cone TV repair modification was introduced at our center (January 2007); TV replacement was performed in 11 of these patients, and 2 patients had TV repair, but the cone modification was not used. Thus, we do not have experience with TV repair using the cone technique in patients ≥50 years of age. We have used the cone repair in approximately 85% of our younger patients with EA, with a satisfactory repair in ≤90%.

Bidirectional cavopulmonary shunts have been described as being helpful in patients with severe EA who are at risk for right-sided heart failure (6). A bidirectional cavopulmonary shunt was performed on only 2 patients in our series, and neither patient had a single-ventricle repair (RV exclusion) or needed cardiac transplant during long-term follow-up.

Outcome in our patients was comparable to that in a large series of 539 patients with EA from our center, in which 20-year survival was 71% (mean [range] age at surgery 24 years [8 days to 79 years]) and early mortality was 4.9% (3). Thus, except in neonates with EA for whom early mortality is increased (21,22), the outcome of EA surgery might be age independent (23). Other centers have also reported excellent outcomes in their patients with EA; however, these series primarily involved younger patients (18).

Closure of any atrial septal communication. In most patients with EA, an interatrial communication is present, which is routinely closed during surgery. Some researchers have suggested that isolated closure of an interatrial communication in EA might cause right-sided heart failure. ASD or patent foramen ovale closure was also performed in 10 of our patients before TV surgery, and right-sided heart failure or sudden increase in TV regurgitation did not occur in any of our patients.

Right-sided maze procedure. The incidence of atrial arrhythmias increases with age in any patient, including those with EA; thus, we believe that the maze procedure should be applied liberally in patients with EA who require operation. Although the presence of pre-excitation might not affect outcome (3,4), the most current recommendation is to proceed with arrhythmia intervention before or during reparative surgery for EA (24). In the current era, we generally perform a biatrial maze procedure when atrial fibrillation is persistent. Overall, our approach is to address all abnormalities at the time of operation (25). In this series, we have no data on long-term success of arrhythmia procedures; however, success of these procedures in patients with EA has been shown (24,26).

Study limitations. The excellent outcomes in this group of patients with EA who are ≥50 years of age may be related to a less severe anatomic variant. This certainly is true compared with the defects in neonates; however, the same selection criteria for surgery were applied in these patients as in other adolescents or young adults with EA. There are no data comparing surgical treatment with nonsurgical treatment in these patients.

Cardiac magnetic resonance imaging was not used in this group of patients but may provide additional future data about the RV to risk-stratify patients for repair versus replacement.

Surgical techniques have evolved over the past 30 years in an effort to decrease the number of valve replacements. This has been largely successful in the pediatric and young adult age groups, but further improvements in repair techniques are necessary to ultimately decrease the number of TV replacements in patients ≥50 years of age.

In this study, the number of patients with events was small. Although some risk factors for mortality were identified, the low number of events resulted in limited study power. This is reflected in wide confidence intervals on estimated hazard ratios.

Conclusions

Cardiac surgery in patients with EA who are ≥50 years of age may be complex; however, if the operations are performed at a center with experience in congenital heart disease, early mortality is low (4%) and long-term outcome is excellent. Better survival is seen in patients with postoperative improvement in functional class and in those who are younger at the time of surgery. Cardiac surgery in EA at an older age is feasible but palliative, and long-term follow-up in these patients is required despite excellent outcome. Our data showed that surgery should be performed before EA disease is too advanced and patients are too symptomatic.

Reprint requests and correspondence: Dr. Heidi M. Connolly, Division of Cardiovascular Diseases, Mayo Clinic, 200 First Street SW, Rochester, Minnesota 55905. E-mail: connolly.heidi@mayo.edu.

REFERENCES

1. Keith JD, Rowe RD, Vlad P. Heart Disease in Infancy and Childhood. New York, NY: MacMillan, 1958:58.
2. Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC Jr. Operative treatment of Ebstein's anomaly. J Thorac Cardiovasc Surg 1992;104:1195–202.
3. Brown ML, Dearani JA, Danielson GK, et al. The outcomes of operations for 539 patients with Ebstein anomaly. J Thorac Cardiovasc Surg 2008;135:1120–36, e1–7.
4. Celermajer DS, Bull C, Till JA, et al. Ebstein's anomaly: presentation and outcome from fetus to adult. J Am Coll Cardiol 1994;23:170–6.
5. Chauvaud S. Ebstein's malformation: surgical treatment and results. Thorac Cardiovasc Surg 2000;48:220–3.

6. Chauvaud S, Fuzellier JF, Berrebi A, et al. Bi-directional cavopulmonary shunt associated with ventriculo and valvuloplasty in Ebstein's anomaly: benefits in high risk patients. Eur J Cardiothorac Surg 1998;13:514–9.
7. Dearani JA, Danielson GK. Surgical management of Ebstein's anomaly in the adult. Semin Thorac Cardiovasc Surg 2005;17:148–54.
8. Greason KL, Dearani JA, Theodoro DA, Porter CB, Warnes CA, Danielson GK. Surgical management of atrial tachyarrhythmias associated with congenital cardiac anomalies: Mayo Clinic experience. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2003;6:59–71.
9. Quinonez LG, Dearani JA, Puga FJ, et al. Results of the 1.5-ventricle repair for Ebstein anomaly and the failing right ventricle. J Thorac Cardiovasc Surg 2007;133:1303–10.
10. Theodoro DA, Danielson GK, Porter CJ, Warnes CA. Right-sided maze procedure for right atrial arrhythmias in congenital heart disease. Ann Thorac Surg 1998;65:149–53.
11. Joffe D, Gurvitz M, Stout K. Considerations of a one-and-one-half ventricle repair in a 47-year-old patient. Congenit Heart Dis 2008;3:69–72.
12. Nakayama T, Asano M, Matsumoto K, et al. Reconstruction of the right heart in an elderly woman with Ebstein's anomaly and severe heart failure. Ann Thorac Surg 2007;84:1745–6.
13. Dearani JA, O'Leary PW, Danielson GK. Surgical treatment of Ebstein's malformation: state of the art in 2006. Cardiol Young 2006;16 Suppl 3:12–20.
14. Danielson GK, Maloney JD, Devloo RA. Surgical repair of Ebstein's anomaly. Mayo Clin Proc 1979;54:185–92.
15. Brown ML, Dearani JA, Danielson GK, et al. Comparison of the outcome of porcine bioprosthetic versus mechanical prosthetic replacement of the tricuspid valve in the Ebstein anomaly. Am J Cardiol 2009;103:555–61.
16. Hetzer R, Nagdyman N, Ewert P, et al. A modified repair technique for tricuspid incompetence in Ebstein's anomaly. J Thorac Cardiovasc Surg 1998;115:857–68.
17. Chauvaud S, Berrebi A, d'Attellis N, Mousseaux E, Hernigou A, Carpentier A. Ebstein's anomaly: repair based on functional analysis. Eur J Cardiothorac Surg 2003;23:525–31.
18. Augustin N, Schmidt-Habelmann P, Wottke M, Meisner H, Sebening F. Results after surgical repair of Ebstein's anomaly. Ann Thorac Surg 1997;63:1650–6.
19. Carpentier A, Chauvaud S, Mace L, et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. J Thorac Cardiovasc Surg 1988;96:92–101.
20. Silva JP, Baumgratz JF, Fonseca L, et al. Ebstein's anomaly: results of the conic reconstruction of the tricuspid valve [in Portuguese]. Arq Bras Cardiol 2004;82:212–6.
21. Boston US, Goldberg SP, Ward KE, et al. Complete repair of Ebstein anomaly in neonates and young infants: a 16-year follow-up. J Thorac Cardiovasc Surg 2011;141:1163–9.
22. Sarris GE, Giannopoulos NM, Tsoutsinos AJ, et al. Results of surgery for Ebstein anomaly: a multicenter study from the European Congenital Heart Surgeons Association. J Thorac Cardiovasc Surg 2006;132:50–7.
23. Boston US, Dearani JA, O'Leary PW, Driscoll DJ, Danielson GK. Tricuspid valve repair for Ebstein's anomaly in young children: a 30-year experience. Ann Thorac Surg 2006;81:690–5, discussion 695–6.
24. Khositseth A, Danielson GK, Dearani JA, Munger TM, Porter CJ. Supraventricular tachyarrhythmias in Ebstein anomaly: management and outcome. J Thorac Cardiovasc Surg 2004;128:826–33.
25. Bockeria L, Golukhova E, Dadasheva M, et al. Advantages and disadvantages of one-stage and two-stage surgery for arrhythmias and Ebstein's anomaly. Eur J Cardiothorac Surg 2005;28:536–40.
26. Stulak JM, Dearani JA, Puga FJ, Zehr KJ, Schaff HV, Danielson GK. Right-sided maze procedure for atrial tachyarrhythmias in congenital heart disease. Ann Thorac Surg 2006;81:1780–4.

Key Words: Ebstein anomaly ■ surgery ■ survival ■ tricuspid valve surgery.